

British Society of Paediatric Gastroenterology Hepatology and Nutrition

ANNUAL MEETING 2017

Wednesday 25th – Friday 27th January Double Tree by Hilton, Cambridge Street, Glasgow

Local Organiser:

Dr Richard Hansen, Consultant Paediatric Gastroenterologist



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Wae worth thy pow'r, thou curséd leaf! Fell source o' a' my woe and grief! Robert Burns, Lines Written on a Banknote

The annual meeting of our Society is not only the main event on our calendar, and a rare opportunity for us to come together, but it is also the most expensive activity of the Society by far.

The continued generous, loyal and committed support of our sponsors means that we can put on a world-class meeting in excellent facilities with expert faculty and also offer a tremendous social programme. We can do all of this whilst keeping the registration costs for Society members to a minimum. Whilst Mr Burns "curséd leaf" might well cause anxiety in the hosting of such a meeting, the support of our sponsors ensures that "woe and grief" within the Society is minimised.

The delivery of a state-of-the-art programme across paediatric gastroenterology, hepatology and nutrition for established clinicians, allied health professionals and trainees alike is a fundamental tenet of the Society's role, driving excellence in clinical practice, innovation through research and fostering multidisciplinary working. Engagement between commercial sponsors and Society members helps to drive similar innovation in the development of products for the children under our care, and ensures we remain up-to-date about what's available for our current and future patients, to better allow us to meet their needs. Please do visit the sponsors at all opportunities throughout the meeting. We simply wouldn't be here without them.

A special thanks to the charities who continue to support and enrich our annual meeting, support our patients, particularly those with chronic health problems, and develop resources used daily in routine clinical practice. When you have a chance, please visit them, say hello and see what's new this year... there's always something different and exciting.

Thanks to Colours for supporting our Wednesday night 80s disco.

Finally, specific thanks to two of our lead sponsors this year: Nestle, who have sponsored the Wednesday evening symposium on nutrition in inflammatory bowel disease and supported the Wednesday evening whisky tasting; and Nutricia, who have funded the Thursday morning symposium on milk allergy and the gut microbiome and subsidised the kilt hire and tartan sash prices to enrich our Thursday gala dinner. Both companies have been generous, not only with time and financial support, but with creative and educational direction, helping us to seamlessly blend the sponsored symposia into the programme. The organisational committee consider these symposia to be a core part of the programme. We hope you will too.

Please enjoy the meeting and enjoy a taste of Scottish hospitality during one of the most important weeks in the Scottish calendar.

Le gach deagh dhúrachd,

Richard Hansen

Chair of Local Organising Committee

Girish GupteBSPGHAN Treasurer

Canch Caupte

Welcome address

Kind Sir, I've read your paper through, And faith, to me, 'twas something new! How guessed ye, Sir, what maist I wanted? This mony a day I've grain'd and gaunted, Bairns guts and livers and nutrition, Michty me, there's nothin missin!

Robert Burns, To a Gentleman who had sent him a Newspaper, and offered to continue it free of expense Adapted for BSPGHAN (badly) by Richard & Moira Hansen

Well, here we are. The 31st annual meeting of the British Society of Paediatric Gastroenterology, Hepatology and Nutrition on our 30th anniversary. From the local organising committee, the BSPGHAN executive and council, welcome to Glasgow!

The motto of our city is "People Make Glasgow", perhaps marking Glasgow out as a peculiarity amongst cities for putting people at the very centre of what's great about the place, but hopefully by the end of the week you'll be convinced too. Certainly, People Make BSPGHAN Glasgow, and I'd like to start by thanking my family in PGHAN, and now your local organising committee: Lawrence Armstrong, Andy Barclay, Elaine Buchanan, Iain Chalmers, Diana Flynn, Vikki Garrick, Kostas Gerasimidis, Richard Russell, Rachel Tayler and our own organisational powerhouse Karen Fraser. Every aspect of this meeting from inception to completion has had eleven pairs of hands helping shape and perfect it. The result, we hope, is a robust, engaging and enjoyable programme with a social side that offers the best of Scottish hospitality and lets members sample the charms and joys of life in Glasgow.

It's fair to say that four previous annual meetings helped shape this one: Edinburgh in 2011 showed us what a Scottish meeting could be like - we've simply turned it up to eleven; Nottingham in 2012 gave the Society a rare Burns night celebration- sorry Charlie, nice try, but we're hoping to show you how it's really done this year!; Stratford-upon-Avon in 2015 tied the meeting inextricably to one nation's Bard- we're channelling another one in Robert Burns for this meeting; Bristol in 2016 showed us the way, provided a template and gave us a hard act to follow! Five people in BSPGHAN deserve special mention for helping us meet and exceed our ambitions for this meeting- Nick Croft as President, Nadeem Afzal as outgoing Convenor, Girish Gupte as outgoing Treasurer, Sandhia Naik as Education Chair, and of course Carla Lloyd as the glue that keeps the Society together and the engine of the annual meeting. Thanks to you all.

When looking for a quote from Burns to open this welcome, I was struck immediately by the one we've chosen. Whilst invited speakers make up most the programme for each annual meeting, and we have some truly tremendous and world-leading invited speakers over the next three days, the abstract presentations from our members provide the biggest surprises and often the most innovative ideas for shaping future practice within the Society. The abstracts, either in oral or poster form, are our lifeblood. Thank you to all those who have submitted abstracts and are presenting them to us this week. I'd encourage all our members to support and encourage those presenting their work at the meeting, particularly those taking their first steps in PGHAN.

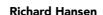
A new tradition for the Society has been the co-hosting of a portion of the annual meeting with a like-minded group or subspecialty. I'm honoured to welcome our friends and colleagues from the Scottish Paediatric Society, founded in 1922 as the first paediatric society of the UK, to co-host the final day of our programme. The theme for Friday is "the coalface interface" or where general paediatrics meets PGHAN. Before we get there though, there's much interest, learning, joy, dancing and friendship ahead of us.

As many of you know, my "wingman" Richard Russell and I have recently secured ESPGHAN for Glasgow in 2019 with the support of many Society members. Some of the ideas we have for that meeting are being tested here in smaller form. We hope most of you will be able to join us in 2019 for nicer weather, longer days and more of the same, albeit on a much bigger scale.

Finally, we've gone for a "less is more" approach to the programme to provide ample time for presentations and discussion. Every single talk in the meeting is open for questions, driven by our firm belief that peer review and polite scrutiny are fundamental facets of the scientific method. Please engage with our speakers and offer your insights, experience and comments. We hope to learn as much from each of you as you do from the meeting itself.

It's an absolute delight to welcome you to Glasgow, and to BSPGHAN our way.

On behalf of the organising committee, Fàilte!



Consultant Paediatric Gastroenterologist Chair of Local Organising Committee



Local organising committee

Lawrence Armstrong

Andy Barclay

Elaine Buchanan

lain Chalmers

Diana Flynn

Vikki Garrick

Kostas Gerasimidis

Richard Hansen

Richard Russell

Rachel Tayler

and the indefatigable Karen Fraser

With special thanks to the following people who have generously given their time in scoring abstracts and presentations.

Abstract Selection Committee 2017

Professor Stephen Allen, Chair of Research Working Group: Dr Richard Hansen, Local Organiser; Dr Paul Henderson, Trainee Member Representative; Dr Kelsey Jones, Chair of Trainee Members' Group; Ms Nicky Heather, Chair of Associate Members' Group

Oral Abstract Scoring Committee:

Dr Rafeeq Muhammed (Chair), Consultant Paediatric Gastroenterologist: Dr Alastair Baker, Consultant Paediatric Hepatologist; Dr Richard Russell, Consultant Paediatric Gastroenterologist; Dr Victoria Merrick, Paediatric Trainee; Ms Elaine Buchanan, Specialist Dietitian

Poster of Distinction Scoring Committee:

Dr Kelsey Jones: Dr Konstantinos Gerasimidis; Dr Marcus Auth; Dr Fiona Cameron; Dr Lucy Howarth; Dr Georgina Hold; Ms Nicky Heather; Dr Paul Henderson; Dr Assad Butt; Ms Joan Gavin; Dr Astor Rodrigues; Dr Anastasia Konidari; Ms Hazel Duncan; Dr Nicola Ruth; Dr Ieuan Davies; Ms Jackie Falconer

POST GRADUATE DAY Wednesday 25th January 2017

Double Tree by Hilton, Cambridge Street, Glasgow

Welcome and Introduction

10.20 - 10.30

Dr Richard Hansen Consultant Paediatric Gastroenterologist Local Organiser BSPGHAN Annual Meeting 2017

10.30 - 12.30

Session 1

Interactive IBD Masterclass

Chairs:

Dr Richard Russell, Consultant Paediatric Gastroenterologist, Royal Hospital for Children, 1345 Govan Road, Glasgow and

Dr Mike Cosgrove, Consultant Paediatric Gastroenterologist, Dept of Child Health, Singleton Hospital, Swansea

10.30 - 11.00

Logical Progression - how to get the best of IBD Medications in 2017

Professor David Wilson Child Life and Health University of Edinburgh 20 Sylvan Place Edinburgh, ED9 1UW

11.00 -11.30

Liver involvement in IBD: a user's guide

Dr Rachel Tayler Consultant Paediatric Gastroenterologist Royal Hospital for Children 1345 Govan Road Glasgow

11.30 - 12.30

IBD Case Discussions

Panel

Dr Richard Russell, Consultant Paediatric Gastroenterologist
Dr Paul French, Consultant Paediatric Pathologist
Dr Tom Savage, Consultant Paediatric Radiologist
Mr Gregor Walker, Consultant Paediatric Surgeon
Sister Lee Curtis, Clinical Nurse Specialist in Paediatric IBD

Royal Hospital for Children 1345 Govan Road Glasgow, G51 4TF

12.30 - 13.30

LUNCH

and opportunity to meet sponsors in Cambridge Suite Poster viewing in Robert Burns Suite

13.30 - 15.30

Session 2

Interactive Nutrition Masterclass

hairs:

Ms Elaine Buchanan, Dietitian, Royal Hospital for Children, 1345 Govan Road, Glasgow and

Dr Theodoric Wong, Consultant Paediatric Gastroenterologist, Dept of Gastroenterology, Birmingham Children's Hospital, Steelhouse Lane, Birmingham

13.30 - 13.50

Key points to management of Home Parenteral Nutrition (HPN)

Ms Christina McGuckin Paediatric Parenteral Nutrition Nurse Specialist Royal Hospital for Children 1345 Govan Road Glasgow

13.50 - 14.10

Oral versus enteral and novel feeding in intestinal failure

Ms Sarah Macdonald Dietitian Great Ormond Street Hospital for Children Great Ormond Street London WC1N 3JH

14.10 - 14.30

Non-transplant surgery in intestinal failure

Professor Mark Davenport Consultant Paediatric Hepatobiliary Surgeon King's College Hospital Denmark Hill London

14.30 - 14.50

Liver only versus combined transplant and why early transplant should still be considered

Dr Girish Gupte
Consultant Paediatric Hepatologist
Liver Unit, Birmingham Children's Hospital
Steelhouse Lane
Birmingham

14.50 - 15.20

Medical management of short bowel syndrome

Professor Palle Jeppensen Gastroenterologist Rigshospitalet, Blegdamsvej 9 DK-2100 København Ø Copengahen Denmark

15.20 - 15.30

Interventions for intestinal failure - Open discussion with interactive voting

Dr Andrew Barclay Consultant Paediatric Gastroenterologist Royal Hospital for Children 1345 Govan Road Glasgow

15.30 - 16.00

AFTERNOON BREAK

opportunity to visit sponsor stands in Cambridge Suite Poster viewing in Robert Burns Suite

16.00 - 17.00

Session 3

Plenary abstract session

Chairs:

Dr Diana Flynn, Consultant Paediatric Gastroenterologist, Royal Hospital for Children, 1345 Govan Road, Glasgow and

Dr David Devadason, Consultant Paediatric Gastroenterologist, Queens Medical Centre, Derby Road, Nottingham

16.00 - 16.10

The evolution of the BIFS registry to Paed eBANS: a 2016 update

Dr Andrew R Barclay, Consultant Paediatric Gastroenterologist, Royal Hospital for Children Glasgow; Dr Paul Henderson, Consultant Paediatric Gastroenterologist Royal Hospital for Sick Children, Edinburgh; On behalf of 31 participating nutrition centres; 10 Paed eBANS centres

16.10 - 16.20

Clinical and financial benefits of a Nutrition Support Team

Tracey Johnson, Senior Specialist Gastroenterology Dietitian; Michelle Butcher, Clinical Nurse Specialist; Haidee Norton, Senior Specialist Gastroenterology Dietitian; Amanda Scott, Senior Pharmacist; Adam Henderson, Senior Pharmacist, Theo Wong, Consultant Gastroenterologist, Sue Protheroe, Consultant Gastroenterologist; Birmingham Children's Hospital, Steelhouse Lane, Birmingham, B4 6NH

16.20 - 16.30

Exclusive enteral nutrition modulates the microbiota in childhood Crohn's disease independently of its effect on intestinal inflammation

Protima Amon¹, Gloria Serena², Alessio Fasano², Allan Walker², William Wade¹, Ian R. Sanderson¹ ¹Blizard Institute, Queen Mary University London, UK. ²Harvard Clinical Nutrition Research Centre, Massachusetts General Hospital, Boston, USA.

16.30 - 16.40

Diagnostic accuracy of neutrophil-lymphocyte ratio in suspected paediatric inflammatory bowel disease: a regional cohort study

Dr Iain Chalmers¹;Professor David C Wilson^{1,2};Dr Paul Henderson^{1,2}
¹Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh; ²Child Life and Health, University of Edinburgh, Edinburgh

16.40 - 16.50

Equivalent efficacy and short term safety, with reduced cost of Infliximab biosimilars in a national paediatric UK IBD induction cohort: time for universal adoption?

Neil Chanchlani, Specialist Trainee in Paediatrics, Royal Free London NHS Foundation Trust, London'; Kajal Mortier, Project Manager, Royal College of Physicians, London; et al

16.50 - 17.00

The State of Grid Training: Looking to the Future Training of Paediatric Gastroenterologists and Hepatologists in the UK

Dr Edward Gaynor, Trainee Representative of PGHAN CSAC & ST8 Paediatric Gastroenterology Registrar, Kings College Hospital; Dr Sue Protheroe, Chair of PGHAN CSAC & Consultant Gastroenterologist, Birmingham Children's Hospital

17.00 - 18.00

Session 4



Evening Symposium

Chair

Dr Richard Russell, Consultant Paediatric Gastroenterologist, Royal Hospital for Children 1345 Govan Road, Glasgow

The science of exclusive enteral nutrition

Dr Konstantinos Gerasimidis Lecturer in Human Nutrition School of Medicine, Dentistry & Nursing New Lister Building Glasgow Royal Infirmary

Nutritional therapy for IBD: an exclusive club or room for anyone?

Professor Arie Levine Paediatric Gastroenterologist Wolfson Medical Center, Israel

The anti-inflammatory power of exclusive enteral nutrition: from IBD guidelines to real life

Dr Frank Ruemmele Service de gastroentérologie-hépatologie-nutrition pédiatrique CHU Paris - Hôpital Necker-Enfants Malades 149 rue de Sèvres 75743 PARIS, FRANCE 18.15 - 19.15

Professional Group meetings

Associates Trainees

19.30 - 20.30

Annual Trainees v Consultants Football Match

or

Kelvingrove Art Gallery Tour
visit www.glasgowlife.org.uk/museums/kelvingrove/Pages/default.aspx

20.30 - 22.00

Opportunity Learn about the history of Whisky, the different processes and opportunity to taste different Whiskies. All welcome as non-alcohol drinkers will be catered for with a soft drink option

Sponsored by Nestle

21.00 - till late

Robert Burns Suite

Welcome Dinner and Disco "80s 'til Laties" STREETrave with DJ Jon Mancini from Colours

Thursday 26th January 2017

Double Tree by Hilton, Cambridge Street, Glasgow

7.45 - 8.55

Working Group Meetings
Please see notice boards for room details

Endoscopy Nutrition Motility Education Research

9.00 - 10.00

Session 5



Breakfast Symposium: Nutricia Advanced Medical Nutrition

Microbiota-Immune Interactions in Allergic Disease

Chai

Dr Richard Hansen, Consultant Paediatric Gastroenterologist Royal Hospital for Children, Glasgow

The microbiome in the development of allergic disease - what is the role of pre/probiotics?

Professor Simon Murch Consultant Paediatric Gastroenterologist University Hospital Coventry & Warwickshire Professor Emeritus Warwick University

Synbiotics can target microbial dysbiosis in the dietary management of cow's milk allergy

Dr Louise Michaelis Consultant in Paediatric Immunology& Allergy Great North Children's Hospital Newcastle

10.00 - 10.50

Session 6

Nutrition Meets the Microbiome

Dr Richard Hansen, Consultant Paediatric Gastroenterologist, Royal Hospital for Children, 1345 Govan Road, Glasgow and

Professor Ian Sanderson, Consultant Paediatric Gastroenterologist, Barts and The London, Turner Street, London

10.00 - 10.25

The beginner's guide to microbiome: from bench to bedside

Dr Georgina Hold Senior Lecturer in Gastroenterology University of Aberdeen Institute of Medical Sciences Foresterhill Aberdeen AB25 2ZD

10.25 - 10.50

Microbial therapeutics in inflammatory bowel disease

Dr Daniel Gaya Consultant Gastroenterologist Glasgow Royal Infirmary Castle Street Glasgow G4 0SF

10.50 - 11.20

MORNING BREAK

opportunity to visit sponsor stands in Cambridge Suite Poster viewing in Robert Burns Suite

11.20 - 12.30

Session 7

Liver

Chairs:

Dr Suzanne Davison, Consultant Paediatric Hepatologist, Leeds General Infirmary, Great George Street, Leeds and

Dr Indra van Mourik, Consultant Paediatric Hepatologist, Liver Unit, Bimingham Children's Hospital, Birmingham

11.20 - 11.50 Debate: Who does immunosuppression better in liver disease?

For Adult Heptology:

Dr Ewan Forrest Consultant Gastroenterologist Glasgow Royal Infirmary Castle Street Glasgow G4 0SF

For Paediatric Hepatology:

Dr Jonathon Hind Consultant Paediatric Hepatologist King's College Hospital Denmark Hill London

11.50 - 12.10

Supporting families awaiting liver transplant

Ms Julie Jeffery Transplant Coordinator Leeds General Infirmary Leeds, LS1 3EX

12.10 - 12.30

A pragmatic approach to acute liver failure

Dr Sue Beath Consultant Paediatric Hepatologist Liver Unit, Birmingham Children's Hospital Steelhouse Lane, Birmingham

12.30 - 13.30

LUNCH

and opportunity to meet sponsors in Cambridge Suite Poster viewing in Robert Burns Suite

13.30 - 14.30

Session 8

Plenary Abstract Presentations

Chairs:

Dr Akshay Batra, Consultant Paediatric Gastroenterologist, Southampton General Hospital, Tremona Road, Southampton

Ms Vikki Garrick, IBD Nurse Specialist, Royal Hospital for Children, 1345 Govan Road, Glasgow

13.30 - 13.40

Development of a Paediatric Endoscopy Global Rating Scale - Results of a National Pilot

Dr P Narula¹, Mr R Broughton², Dr R Bremner³, Dr A Piggott⁴, Dr D Rawat⁵, Mr M Cullen6, Dr NA Afzal⁶, Dr L Howarth⁷, Dr P Gillett⁸, Dr P Henderson⁸, Dr K Venkatesh⁹, Dr C Tzivinikos⁹, Ms J Maginnis⁴, Ms S Mckenna¹, Dr D Devadason10, Dr S Loganathan¹⁰, Dr M Stanton⁶, Dr J Green², Ms D Johnston², ¹Sheffield Children's Hospital; ²JAG; ³Birmingham Children's Hospital; ⁴Royal Stoke Children's Service; ⁵Royal London Hospital; ⁶Southampton Children's Hospital; ⁷John Radcliffe Children's Hospital; ⁷John Radcliffe Children's Hospital; ⁹Alder Hey Children's Hospital; ¹⁰Nottingham

13.40 - 13.50

Safety and efficacy of a novel haemostatic agent Hemospray® in the emergent endoscopic management of acute upper gastrointestinal bleeding in children.

Thomson M, Rao P, Narula P, Urs A, Campbell D, Belsha D. Sheffield Children's Hospital, Western Bank, Sheffield

13.50 - 14.00

Systematic review and meta-analysis of hepatic outcomes in non-alcoholic fatty liver disease after bariatric surgery in children.

Jake P. Mann: Department of paediatrics, University of Cambridge; Valerio Nobili: Hepatometabolic Unit, Bambino Gesu Hospital, IRCCS, Rome, Italy

14.00 - 14.10

Ethanol and Taurolidine line locks for the prevention and treatment of catheter related bloodstream infections in paediatric intestinal failure: a systematic review and meta-analysis Dr lain Chalmers, ST8 Paediatric Gastroenterology¹; Dr Paul Henderson, Consultant Paediatric Gastroenterologist¹.²; Dr Rachel Tayler, Consultant Paediatric Gastroenterologist³; Professor David Wilson, Consultant Paediatric Gastroenterologist¹.²; Dr Andrew Barclay, Consultant Paediatric Gastroenterologist³

¹Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh; ²Child Life and Health, University of Edinburgh, Edinburgh; ³Department of Paediatric Gastroenterology, Royal Hospital for Children, Glasgow

14.10 - 14.20

Hepatopulmonary syndrome in children – symptoms, clinical progression and outcome post Liver transplant

¹S. Warner, ¹PJ McKiernan, ¹DA Kelly, ¹ID van Mourik, ¹G Gupte, ¹J Hartley, ¹M Abdel-Hady, ¹K Sharif, ²D Mirza, ²P Muiesan, ²T Perera, ¹SV Beath.

¹Birmingham Children's Hospital and ²The Queen Elizabeth Hospital, United Kingdom.

14.20 - 14.30

Portal Vein Obstruction: The Challenge of Timely Diagnosis

D Belsha, S Davison, S Hodges, S Rajwal, P McClean. Children's Liver Unit, Leeds General Infirmary, Leeds LS1 3E

14.30 - 15.00

Research Session

Chairs:

Professor Stephen Allen, Professor of Paediatrics, Liverpool School of Tropical Medicine, Dept of Clinical Sciences, Liverpool and

Dr Julian Thomas, Consultant Paediatric Gastroenterologist, Royal Victoria Infirmary, Victoria Road, Newcastle

1. Research workplan for 2017

Professor Stephen Allen

2. Joint BSPGHAN/CORE grants 2015 - progress reports:

- "Anabolic resistance and abnormal muscle function across the nutritional spectrum: a pilot study in Crohn's disease and non-alcoholic fatty-liver disease"; Nottingham Digestive Diseases Centre: Amanda Walker
- "LiverMultiScanTM for the assessment of graft fibrosis in children post-liver transplant";
 King's College London: Serena Kyrana
- "Health informatics research in paediatric gastroenterology: nationwide datalinkage exploration of perinatal risk factors for and consequences of paediatric-onset Inflammatory Bowel Disease"

Child Life and Health, University of Edinburgh; Paediatric Gastroenterology, Royal Hospital for Sick Children, Edinburgh: David Wilson

15.00 - 15.30

BREAK

opportunity to visit sponsor stands in Cambridge Suite Poster viewing in Robert Burns Suite

15.30 - 16.30

Session 9

The Future of IBD

Chairs

Dr Dharam Basude, Consultant Paediatric Gastroenterologist, Bristol Royal Hospital for Children, Upper Maudlin Street, Bristol and

Dr Nadeem Afzal, Consultant Paediatric Gastroenterologist, Southampton General Hospital, Tremona Road, Southampton

15.30 - 16.00

Future Optimisation of IBD therapy

Dr Tariq Ahmad Consultant Gastroenterologist Royal Devon and Exeter Hospital Exeter

16.00 - 16.30

Predicting the future in IBD patients

Professor Arie Levine Paediatric Gastroenterologist Wolfson Medical Center Israel

16.45 - 19.00

BSPGHAN Annual General Meeting

19.30

Carriages to Oran Mor, Byres Road

20.00 - late

Burns Night Theme Gala Dinner

Rock Ceilidh

Music provided by Pacific www.pacificweddingband.com

Kilt Hire sponsored by Nutricia

Friday 27th January 2017

Double Tree by Hilton, Cambridge Street, Glasgow

7.45 - 9.00

Working Groups (Please see notice boards for rooms)

Quality Standards Hepatology PeGHANS

09.00 - 10.30

Session 10

The Coalface Interface: Part I

Chairs:

Dr Mike Bisset, Consultant Paediatric Gastroenterologist Royal Aberdeen Children's Hospital, Westburn Road, Aberdeen and

Dr Huw Jenkins, Consultant Paediatric Gastroenterologist, University Hospital of Wales, Heath Park, Cardiff

9.00 - 9.30

Investigating and managing abdominal pain

Professor Mark Beattie Consultant Paediatric Gastroenterologist Southampton General Hospital Tremona Road Southampton

9.30 - 10.10 Debate: Is invasive nutrition support indicated in severe neurodisability?

For:

Dr Andrew Barclay Consultant Paediatric Gastroenterologist Royal Hospital for Children 1345 Govan Road Glasgow

Against:

Dr Akshay Batra Consultant Paediatric Gastroenterologist Southampton General Hospital Tremona Road Southampton

10.10 - 10.30

Difficult end of life decisions and hospice support in paediatrics

Dr Patrick Carragher
Medical Director
Children's Hospice Association Scotland
CHAS Head Office
Canal Court, 42 Craiglockhart Avenue,
Edinburgh

10.30 – 11.00

BREAK

opportunity to visit sponsor stands in Cambridge Suite Poster viewing in Robert Burns Suite

11.00 - 12.30

Session 11

The Coalface Interface: Secondary meets Tertiary PGHAN

Chairs:

Dr Lawrence Armstrong, Consultant in Paediatrics, Darlington Road, Ayr and

Dr Naeem Ayub, Consultant Paediatrician, Royal Shrewsbury Hospital, Telford

11.00 - 12.30

Top 10 Calls to PGHAN

Featuring the full range of Gastroenterology, Hepatology, Nutrition and IBD Panel:

Dr Diana Flynn, Consultant Paediatric Gastroenterologist, Glasgow Dr Huw Jenkins, Consultant Paediatric Gastroenterologist, Cardiff Dr David Goudie, Consultant Paediatrician, Raigmore Ms Lindsay Hogg, Paediatric Specialist Nurse, Birmingham Ms Kathleen Ross, Paediatric Dietitian, Aberdeen

12.30 - 13.30

LUNCH

FINAL opportunity to meet sponsors in Cambridge Suite
Poster viewing in Robert Burns Suite

13.30 - 14.30

Session 12

Let Sleeping Babies Lie

Chairs

Dr Andrew Barclay, Consultant Paediatric Gastroenterologist, Royal Hospital for Children, 1345 Govan Road, Glasgow and

Dr Mohammed Mutalib, Consultant Paediatric Gastroenterologist, Evelina London Children's Hospital, St Thomas's Hospital, Westminster Bridge Road, London

13.30 - 14.00

Investigating and managing reflux in infancy

Dr Peter Gillett Consultant Paediatric Gastroenterologist Royal Hospital for Sick Children 9 Sciennes Road Edinburgh

14.00 - 14.30

Paediatric and infant sleep: What's normal?

Dr Mike Farguhar Consultant in Paediatric Sleep Medicine Evelina London Children's Hospital St Thomas' Hospital, Westminster Bridge Road London

14.30 - 15.30

Session 13

Plenary Abstract Presentations

Joint Scottish Paediatric Society and BSPGHAN

Dr Loveday Jago, Consultant Paediatric Gastroenterologist, Manchester Children's Hospital, Oxford Road, Manchester

Dr Kostantinos Gerasimidis, Lecturer in Clinical Nutrition, School of Medicine, University of Glasgow, New Lister Building, Glasgow Royal Infirmary

14.30 - 14.40

Awareness of ESPGHAN guidelines on coeliac disease amongst general paediatricians in **Southwest England**

Dr Siba Paul¹; Miss Helen Adams²; Dr Dharam Basude¹, ¹Bristol Royal Hospital for Children; ²University of Bristol

14.40 - 14.50

Diagnostic endoscopy in children with GI symptoms: Indications and Outcomes **Prospective Study**

A Kadir, S Naik, D Rawat, N Meadows, P Amon, N Croft Royal London Hospital, London

14.50 - 15.00

Prospective Paediatric Appendicitis and Risk of Perforation from Prehospital Delay

Dr David I Campbell¹; Alexander Labeit², ScHARR, Health Economist; Basil Bekdash³; Paediatric Surgical Trainee; Dipanker Dass1; Paediatric Surgical Trainee; Sean Marven1, Surgical Consultant; Tracey A Young ScHARR², Statistician

¹Sheffield Children's Hospital, Western Bank Sheffield; ²University of Sheffield;

³University of Oxford

15.00 - 15.10

Dietary Manipulation of the Healthy Human and Colitic Murine Gut Microbiome by CD-TREAT diet and Exclusive Enteral Nutrition; a proof of concept study.

V. Svolos1, R. Hansen², U.Z. Ijaz³, C. Quince⁴, D. Watson⁵, A. Alghamdi⁵, A. Brejnrod⁴, C. Ansalone⁶, S. Milling⁶, D. Gaya⁷, R. Russell², K. Gerasimidis¹ ¹Human Nutrition, School of Medicine, College of Medical, Veterinary and Life Sciences, University of Glasgow, Glasgow Royal Infirmary, Glasgow, United Kingdom; ²Department of Paediatric Gastroenterology, Hepatology and Nutrition, Royal Hospital for Children, Glasgow, United Kingdom; ³School of Engineering, University of Glasgow, Glasgow, United Kingdom, ⁴Warwick Medical School, University of Warwick, Warwick, United Kingdom, ⁵Strathclyde Institute of Pharmacy and Biomedical Sciences, University of Strathclyde, Glasgow, United Kingdom; ⁶Institute of Infection, Immunity and Inflammation, School of Medicine, College of Medical,

Veterinary and Life Sciences, University of Glasgow, Glasgow, United Kingdom; ⁷Department of Gastroenterology, Glasgow Royal Infirmary, Glasgow, United Kingdom

15.10 - 15.20

Blended Diet for Enteral Feeding - Identifying current practice in a paediatric community setting and developing a clinical tool to improve patient care

Mrs Charlie Bigwood, Senior Paediatric Dietitian, Chailey Clinical Services, Beggars Wood Road, Lewes, East Sussex, BN8 4JN

15.20 - 15.30

Scottish home parenteral nutrition longitudinal point prevalence data suggest a dramatic rise

Dr Iain Chalmers¹, ST8 Paediatric Gastroenterology; Dr Paul Henderson^{1,2}, Consultant Paediatric Gastroenterologist; Mrs Christina Mcguckin3, Parenteral Nutrition Clinical Nurse Specialist; Miss Catherine Paxton¹, Parenteral and Enteral Nutrition Clinical Nurse Specialist Dr Shyla Kishore⁴, Consultant Paediatric Gastroenterologist; Dr David Goudie⁵, Consultant Paediatrician; Dr David Mitchell¹, Consultant Paediatric Gastroenterologist; Dr Diana M Flynn³, Consultant Paediatric Gastroenterologist; Dr Andrew R Barclay³, Consultant Paediatric Gastroenterologist

¹Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh; ²Child Life and Health, University of Edinburgh, Edinburgh; ³Department of Paediatric Gastroenterology, Royal Hospital for Children, Glasgow; ⁴Department of Paediatric Gastroenterology, Royal Aberdeen's Children's Hospital; ⁵Department of Paediatrics, Raigmore **Hospital Inverness**

15.30 - 16.15

Session 14

Keynote Lecture

Dr Donald McGregor, President of Scottish Paediatric Society, Consultant Paediatrician Ninewells Hospital, Dundee and

Professor Nick Croft, President of BSPGHAN, Professor of Clinical Paediatric Gastroenterology, Barts and the London School of Medicine, London

15.30 - 16.15

New thoughts on weaning in infancy

Dr Michael Perkin Consultant in Paediatric Allergy St George's University Hospitals NHS Foundation Trust St George's, University of London Cranmer Terrace, London

16.15 - 16.30

PRIZES AND PRESENTATIONS CLOSE OF MEETING

Previous Prize winners

2008 Southampton

Alex Mowat Prize – Dr Andrew Barclay
Best Abstract Presentation – Ms Elaine Buchanan
Best Presentation – Dr Sherina Ross

2009 Sheffield

Alex Mowat Prize – Dr Johann van Limbergen Sean Devane Memorial – Dr Jenny Epstein Best Allied Health Professional – Ms Jackie Falconer

2010 Liverpool

Alex Mowat Prize – Dr Emer Fitzpatrick Sean Devane Memorial – Dr Rachael Taylor Best Poster Presentation – Dr Paul Henderson

2011 Edinburgh

Alex Mowat Prize – Dr Paul Henderson Sean Devane Memorial – Dr Emer Fitzpatrick Best Poster Prize – Ms Helen French

2012 Nottingham

Alex Mowat Prize – Mark Goddard Sean Devane Memorial – Anna Gregory Challenging Case – Lisa Whyte Best Poster – Ms Hannah Williamson

2013 Manchester

Alex Mowat Prize – Dr Protima Amon Sean Devane Memorial – Dr Lisa Whyte Best Poster Prize – Dr Rana Bitar

2014 - London

Alex Mowat Prize – Dr Vandana Jain Sean Devane Memorial – Dr Ed Giles Best Poster Prize – Dr Bradley Keller

2015 - Stratford upon Avon

Alex Mowat Prize – Dr Mona Abdel-Hady Sean Devane Memorial – Dr Kelsey Jones Best Poster Prize – Sarah Macdonald and Dr Katherine Fawbert Best PICO Presentation – Dr Huey Miin Lee

2016 – Bristol

Alex Mowat Prize – Dr Nicola Ruth Sean Devane Memorial – Y Koh Best Poster Prize – Martin Lister

Speaker Biographies



Dr Tariq Ahmad, M.B., Ch.B., D.Phil., F.R.C.P. (UK)

Dr Tariq Ahmad is a Consultant Gastroenterologist in Exeter. He qualified from Bristol University in 1992 and completed higher specialist training in Oxford where he developed particular interests in inflammatory bowel disease and nutrition. Dr Ahmad was awarded a D. Phil in 2002 for his research investigating the genetic basis for disease heterogeneity in IBD, and continues to work in this field at the University of Exeter Medical School. He is a member of the UK IBD genetics consortium and currently leads a series of UK-wide pharmacogenetic studies, PANTS and PRED4, investigating response and side effects of drugs used in gastroenterology. He is currently the PI or CI for a number of commercial clinical trials and is the Peninsula NIHR gastroenterology research lead. He spends his spare time on the phone hassling colleagues to recruit to the genetic studies, much to the annoyance of his family and friends.



Dr Andrew Barclay

Andy Barclay is 28th generation Caledonian.

He studied as an undergraduate at the University of Aberdeen before completing a post graduate degree at the University of Glasgow into the role of the gut microbiota in Inflammatory diseases of childhood. He has been a consultant at the Royal Hospital for Children Glasgow since 2011, and has a specialist interest in complex enteral nutrition and intestinal failure. He continues to have a research interest in intestinal failure epidemiology, systematic review of evidence in paediatric gastroenterology and has been the lead for the BSPGHAN Paed eBANS registry 2012-2017.

Andy became the chair of the Scottish Society of Paediatric Gastroenterology Hepatology and Nutrition in 2013, a position he has unilaterally converted into the quasi-democratic position of 'SSPGHANident'. He intends to occupy this position in perpetuate, via his own self-imposed 'emergency measures'.



Dr Akshay Batra

Akshay is a Paediatric Gastroenterologist at University Hospital Southampton NHS Trust. He joined UHS in 2011 after competing PGHAN training at Bristol and Birmingham Hospitals. He has special interest in Nutrition and is the locally leads the Nutritional Support Team and Intestinal Failure Service. He is also the Training Advisor for nutrition on College Speciality Advisory Committee.



Dr Sue Beath BSc; MB BS; DTM; MRCP; FRCPCH

Dr Sue Beath graduated from St. Mary's Hospital Medical School in London in 1983 and via junior appointments in Infectious Diseases and Paediatrics at St George's Hospital joined the Liver Unit in Birmingham in 1989. The Liver Unit was already well known for liver transplantation and was the only centre in the UK at that time to undertake liver transplants in children weighing less than 10kg. The Liver Unit at Birmingham Children's Hospital (BCH) started the first clinical small bowel transplantation programme in the UK in 1993 and Dr Beath was appointed as the first clinical lead for this programme in 1995-2004.

Currently the BCH Liver Unit carries out on average 40 liver transplants per year. Of the 20 children admitted with liver failure each year, approximately 10 have developed an acute decompensation as a result of chronic liver disease; 5 have multi-organ disease which precludes liver transplantation, and 5 require emergency liver transplantation.

We continue to work closely with colleagues at national and international level to improve the management and commissioning of care and research in liver disease and liver failure across the whole spectrum: from short term self-limiting cases of neonatal hepatitis in babies and children, to the long term patients who have established cholestatic syndromes with or without hepatic fibrosis, and those who require transplantation.



Professor Mark Beattie

Professor Beattie is a Consultant Paediatric Gastroenterologist in Southampton and the lead for the regional service. His main clinical and research interests include the Nutritional Management of Chronic Disease, particularly Inflammatory Bowel Disease. He has published widely in the area of Paediatric Gastroenterology and he is the author of multiple revision texts, and the Oxford Handbook of Paediatric gastroenterology, Hepatology and Nutrition. He was President of the British Society of Paediatric Gastroenterology, Hepatology and Nutrition from 2010-3. He is the Editor in Chief of Archives of Disease in Childhood, BMJ Publishing Group.



Dr Patrick Carragher

Dr Carragher has been the Medical Director for Children's Hospice Association Scotland (CHAS) since 2006. He worked as a General Practitioner in Scotland from 1987 to 2006, but was also lead doctor at Rachel House (children's hospice) from 1996 to 2006, and during this time completed a Diploma in Palliative Medicine from the University of Wales College of Medicine, where he is now an Honorary Lecturer.

His work includes "hands on" medical care for babies, children and young people with life shortening conditions in Rachel House, Robin House and in hospitals across Scotland, as well as local, national and UK representative strategic work, mixed with research.

Since 2012 he has been Chair of the Association of Paediatric Palliative Medicine, which is a UK group formed in 2009 to facilitate better education and support for doctors working with children with life shortening conditions.

He is the chair of Scottish Children's and Young People's Palliative Executive (SCYPPEx), which has developed a national children's resuscitation policy, as well as the Framework for the Delivery of Palliative Care for Children and Young People in Scotland. In 2013, he was elected as a Fellow of the Royal College of Paediatrics and Child Health (RCPCH).

Sister Lee Curtis

Lee Curtis (BSc) is one of three Paediatric Inflammatory Bowel Disease (IBD) Clinical Nurse Specialist (CNS) based at the Royal Hospital for Children in Glasgow. During a secondment as a Stoma CNS, her interest in IBD grew and she became an IBD CNS in 2010. Since joining the IBD team, she has achieved an Advanced Clinical Assessment qualification at Stirling University and has also completed the IBD specialist module in Salford University. She has a keen interest in transitional care and has led a local working group developing a guideline and pathway for this process. She has also audited varicella status within the paediatric IBD population and presented this in ECCO 2013. Further to this, she has developed local guidelines on her findings.

Lee is a member of the Nursing & Midwifery Council, Royal College of Nursing, Scottish IBD Nursing group, Scottish Society of Paediatric Gastroenterology Hepatology & Nutrition, the British Paediatric Society of Gastroenterology, Heptatology & Nutrition and the European Crohn's & Colitis Organisation. Within her IBD work she continues to work closely with the IBD charities and acts as a patient advocate.



Prof. Mark Davenport ChM FRCS (Eng) FRCPS (Glas) FRCS (Paeds)

Qualifications: MBChB (distinction in paediatrics) 1981, FRCS (Eng) 1985, FRCPS (Glas) 1984, FRCS (Paeds) 1994

Mark Davenport qualified in medicine in 1981 at the University of Leeds. Surgical training at Leeds, Sheffield and Manchester followed before arriving in London in 1989. A postgraduate research degree (ChM) was awarded from the University of Leeds on experimental aspects of liver transplantation in 1995.

I was appointed as a consultant paediatric surgeon at Kings College Hospital, London in 1994 and since 1998 have been Head of Department. I have developed a major research interest in all aspects of surgical hepatobiliary diseases supported by the Institute of Liver Studies and also antenatally-diagnosed surgical malformations supported by the Harris Birthright Centre for Fetal Medicine.

In 1994 and since 2008 I was honoured with a personal chair in paediatric surgery from Kings College, London.

I actively promote medical writing and have now edited three textbooks on general and hepatobiliary paediatric surgery, currently writing two more and have contributed over 250 papers and reviews together with over 50 chapters on a variety of surgical topics.

I am the UK editor of the Journal of Pediatric Surgery, and an editorial board member of Pediatric Surgery International. I am also the President of the British Association of Paediatric Surgeons for 2016-18.

I am married and my life is blessed with a teenage daughter. Life otherwise revolves around English rock music of the 1970s and a Triumph Tiger Explorer (a motorcycle)!



Dr Mike Farquhar

Dr Michael Farquhar trained in general children's medicine, children's respiratory medicine and children's sleep medicine at the Royal Hospital for Sick Children Glasgow, Nottingham Children's Hospital, The Children's Hospital at Westmead (Sydney), Sydney Children's Hospital and Great Ormond Street Hospital.

Dr Farquhar has been a consultant in the Evelina London children's sleep medicine department since 2012.



Dr Diana Flynn

Diana Flynn is a consultant paediatric gastroenterologist at the Royal Hospital for Children in Glasgow. Her main area of interest is in nutrition, and she is part of the nutrition team in Glasgow which runs the Paediatric Home Parenteral Nutrition Service for the West of Scotland and cares for children with complex enteral nutrition needs. Dr Flynn also runs the eosinophilic oesophagitis service for children with West of Scotland. She was clinical lead for development of the PYMS score for hospital malnutrition, and medical paediatric representative on the Scottish National Nutritional Care Advisory Board, which developed the Nutrition Standards for Scotland, published in 2015.



Dr Ewan Forrest

Consultant Hepatologist and Honorary Clinical Associate Professor at Glasgow Royal Infirmary.

Graduated from the University of Aberdeen. Research MD at Royal Infirmary of Edinburgh investigating Portal Hypertension. Clinical interests in Alcoholic Liver Disease and Nutrition. Co-developer of the Glasgow Alcoholic Hepatitis Score and the Glasgow Modified Alcohol Withdrawal Score.

Conflict of Interest related to Debate: married to a Neonatologist.



Dr Paul French

I am one of four Consultant Paediatric Pathologists based at the Queen Elizabeth University Hospital, Glasgow. I joined the department as a Consultant in August 2013 which receives approximately 5000 surgical specimens and undertakes 350 post-mortem examinations annually. The department reports on a wide range of surgical specimens including a large number of paediatric medical Gl biopsies, surgical Gl resections and a smaller number of medical liver biopsies. My postgraduate general histopathology and specialist paediatric pathology training was undertaken in Glasgow from 2007 to 2013 and undergraduate medical degree at Sheffield University from 1997 to 2001.



Sister Victoria Garrick

Sister Vikki Garrick MSc, BSc, RGN, RSCN, Non-medical prescriber Paediatric Inflammatory Bowel Disease Nurse Specialist Royal Hospital for Children Glasgow

Vikki became a Sick Children's nurse in 1995 after working as an adult nurse for 4 years. She was a staff nurse in the paediatric Burns unit for 5 years then took her first nurse specialist post in 2001. She set up the Tissue Viability Nursing service in the Royal Hospital for Sick Children, Glasgow and continued to run it for another 5 years.

She became the IBD nurse specialist in 2006 where she set up, delivered and managed the specialist nursing service in the Royal hospital for Sick Children in Glasgow. She has since designed and implemented several care pathways and has published her experience of teaching self administration of Methotrexate at home using one of these pathways. She is a keen proponent of MDT working and has worked closely with dietetic colleagues to design and implement an Exclusive Enteral Nutrition Care pathway; this co-ordinated approach has been shown to be of great benefit to the paediatric patient group. More recently she has completed her thesis on the 'Practical Management of Perianal Crohn's disease' and has also published a review on this subject.

Vikki is an advocate for the importance of effective MDT working in the delivery of high quality patient care and is keen to raise the profile of IBD Nurses particularly. She is currently on a working group with Crohn's & Colitis UK and the Scottish Government looking specifically at this role and is enthusiastic about the outcomes from this project.



Dr Daniel Gaya

Dr Daniel Gaya is a consultant gastroenterologist at Glasgow Royal Infirmary and an Honorary Associate Professor at the University of Glasgow Medical School.

Dr Gaya's sub-specialist interest is inflammatory bowel disease (IBD) and is the recipient of a research fellowship from the Chief Scientist Office (CSO) of the Scottish Government to undertake a comprehensive IBD research programme in the West of Scotland. His research interests include novel clinical trial work, faecal biomarkers and manipulation of the microbiome in IBD. He leads on a number of academic and commercial trials at Glasgow Royal Infirmary.

He receives tertiary referrals for the management of complex cases and helped set up the transition clinic for adolescents with IBD with colleagues at the Royal Hospital for Children in Glasgow.

Dr Gaya is board member of the Scottish IBD charity C³ (www.curecrohnscolitis. org) and a member of the British Society of Gastroenterology IBD Committee, the Greater Glasgow & Clyde Area Drug & Therapeutics Committee (ADTC) and a gastroenterology advisor to the Scottish Medicines Consortium (SMC).



Dr Peter Gillett

Is a Consultant Paediatric Gastroenterologist at the Royal Hospital for Sick Children, Edinburgh and was appointed in 2001. His training was in Newcastle and Edinburgh and the Children's Hospital in Vancouver between 1997 and 1999.

He won the Inaugural JA Campbell Young Investigator of the Year Award in 1999 from the Canadian Celiac Association jointly with his wife Helen Gillett (who established an anti- tTG assay in Edinburgh and then Vancouver) and completed research projects in children with Type I Diabetes and in Turner Syndrome.

His main interests are Coeliac disease, upper GI disorders (primarily gastrooesophageal reflux disease and H pylori), endoscopy and constipation and its improved management.

He has an interest in GI problems in neurodevelopmental disorders and is a Specialist advisor in Gastroenterology to the Cornelia de Lange Society and the CdLS World Federation.

He established the SE Scotland regional coeliac service in 2001 and is an Advisor to Coeliac UK for the last 10 years and now sits on the Health Advisory Council of CUK. Research within the department has been funded partly by CUK and the Gloag family foundation. He is a member of the Scottish government groups developing the GFFS and DOIT programmes and was a member of the NICE Coeliac GDG which reported in 2015, the subsequent NICE Quality Standard published in 2016 and is a member of the BSPGHAN and ESPGHAN Coeliac guideline groups.



Dr David Goudie

I am a paediatrician with an interest in PGHAN working in Raigmore Hospital, Inverness in the Scottish Highlands. I work within the North of Scotland PGHAN network sharing care with the visiting team from Grampian. We look after children over a wide geographical area, which involves increasing use of telemedicine for patient reviews, but also visiting some pleasant Highland hotels.



Dr Girish Gupte

Consultant Paediatric Hepatologist, Liver Unit, Birmingham Children's Hospital Girish finished his undergraduate and postgraduate studies (M.D. paediatrics) in India and came to UK for his higher specialist training. He now works at Birmingham Children's Hospital (BCH) as a Hepatologist with special interest in small bowel transplantantion. He first came to Birmingham Children's Hospital as a Registrar and now practices as a consultant. Girish also practices at five other small bowel outreach clinics across the country.

Clinical interests:

- Managing complex liver conditions and small bowel transplantations
- Developing non-invasive markers of small bowel transplantation

Areas of expertise:

- Chronic liver disease
- Intestinal Failure associated liver disease
- Metabolic liver disease
- Liver Transplantation
- Intestinal Transplantation

Dr Huw Jenkins

Dr Huw Jenkins is a Consultant Paediatric Gastroenterologist at University Hospital of Wales in Cardiff.



Dr Jonathan Hind

Consultant in Paediatric Hepatology, Intestinal Rehabilitation and Transplantation King's College Hospital, London, Jonathan read medicine at Nottingham University where he was President of the Medical Society, as well as winning a university distinction in paediatrics and the British Paediatric Association student prize. He trained in hepatology and gastroenterology at Chelsea and Westminster and King's College Hospitals. During this time he was presented with a "rising star" award from the International Liver Transplant Society. After that, he gained further experience in intestinal transplantation at Birmingham Children's Hospital and the Children's Hospital Pittsburgh. Jonathan returned to King's College in 2008 to join the Consultant Paediatric Hepatology team.

Jonathan has developed and leads the growing intestinal rehabilitation centre at King's and is clinical lead for the supra-regional intestinal transplantation service. He serves on the UK Transplant Bowel Advisory Group, and the British and European Societies of Paediatric Gastroenterology, Hepatology and Nutrition Intestinal Failure working groups, as well as being a councillor for the Intestinal Rehabilitation and Transplantation Association. Current research interests include donor specific antibodies, microbiota and body composition in intestinal failure and transplantation



Lindsay Hogg

Lindsay commenced her training in 1989 as a General Nurse at the Southern General Hospital, Glasgow. She worked for a year in Aberdeen in long term geriatrics before starting her sick children's training at the Royal Hospital for Sick Children, Edinburgh. Following completion of her training she spent time working in renal and respiratory medicine as well as completing two secondments in Infection Control.

In 2000 she moved to Birmingham and joined the Liver Unit at Birmingham Children's Hospital as a Junior Sister. In October 2001 she joined the Specialist Nurse team. She has worked her way through the team to be appointed Principal Specialist Nurse in July 2011.



Dr Georgina Hold

Georgina Hold is a Senior Lecturer in Gastroenterology at Aberdeen University and runs a multi-disciplinary research team focussed on understanding the contribution of gut microbes to gastrointestinal disease. She has published over 85 peer-reviewed publications including in the highest impact journals in the gastroenterology field (Gastroenterology, Cell Host Microbe, American Journal of Gastroenterology, IBD) as well as a number of high ranking invited review articles (Nature Reviews Microbiology, Nature Reviews Gastroenterology and Hepatology, Gut) and book chapters. She has been cited > 3400 times. Her research interests are focused on the identification and functional characterization of microbial communities and understanding host/microorganism interactions during disease progression. She has created an internationally renowned centre for microbiome analysis in Aberdeen. Her group was the first to identify an overrepresentation of epsilon Proteobacteria (non-pylori Helicobacter and emerging Campylobacter) in ulcerative colitis. From these initial studies, an extensive research programme looking at the role of specific pathogenic organisms in colonic disease has been established. She has significantly advanced the field of defining microbial diversity associated with the colonic mucosa and appreciating its relevance in understanding mucosal diseases. Her reputation has seen her elected to numerous external consultancy and advisory roles, she is currently a member of the BSG Gut Microbiota for Health Expert Panel as well as the Bowel Cancer UK Critical Gaps project. She was recently elected the UK member for the European Helicobacter and Microbiota Study Group and was awarded a Fulbright Scholarship in 2014 – 2015.



Julie Jeffery BSc (Hon) MSc

Julie Jeffery is a dedicated transplant co-ordinator for the Live Donor Liver Transplant programme at the Leeds Teaching Hospitals NHS Trust. Julie has varied clinical background in both transplantation and organ donation, with experience working within the hepatobiliary/transplant team in the theatre department and then as a Donor Transplant co ordinator within the Yorkshire Donor co-ordinator Team.

During her career she has developed a special interest in liver transplantation. In 2006 she took up a part-time post as a Liver Transplant co-ordinator with a remit to develop the Live Donor Liver Transplant Programme within the Leeds Teaching Hospitals NHS Trust; going on to spend some time at the well-established Live Donor Liver Transplant Programme at the Toronto General Hospital, Canada.

In 2007 Julie took up her current full time post and is responsible for organizing and leading the assessment pathway of potential live liver donors. Julie also has responsibility for leading the Enhanced Recovery After Surgery (ERAS) team for liver surgery, as well as leading on the on-going development of the Live Donor Liver Transplant Programme.



Professor Palle Jeppensen

Professor Palle Bekker Jeppesen is the Head of the Research at the Department of Gastroenterology at Rigshospitalet in Copenhagen, Denmark. He is also an Affiliate Professor in the Department of Nutrition, Exercise and Sports at the unit for Clinical and Experimental Nutrition, University of Copenhagen.

His major interests include patients with short bowel syndrome and intestinal failure with a recent focus on novel therapies for their management, rehabilitation and care.

Professor Jeppesen graduated in medicine from the University of Copenhagen, Denmark, in 1989 and completed his residency in medical gastroenterology at the Rigshospitalet in Copenhagen. He completed his PhD thesis, entitled 'The significance of the fatty acid chain-length for the clinical effect in the enteral and parenteral nutrition in patients with malabsorption' in 1998 and his Doctor's Degree, entitled Intestinal insufficiency and failure in 2003.

PBJ has published more than 150 peer-reviewed papers and book chapters within this field, has more than 3000 citations and a H index of above 30.



Professor Arie Levine

Arie Levine is the director of the pediatric gastroenterology and nutrition unit at the Wolfson Medical Center and associate professor at Tel Aviv University, Israel. His clinical and research interests are focused primarily on IBD. Arie has chaired the ESPGHAN Porto group, and PECCO, the pediatric committee of ECCO. He is a founding member of the dietary committee of ECCO. He was instrumental in the classification schemes and diagnostic criteria for pediatric IBD, leading the Paris Classification and Revised Porto criteria for IBD. He also serves as a section editor on the IBD journal.

In recent years he has led numerous novel research concepts through clinical and translational studies including early clinical prognostication of IBD, treatment strategies targeting the microbiota in IBD, as well as the role of diet in pathogenesis and treatment of IBD. He is currently running several multicenter multination randomized controlled trials targeting the microbiota.



Sarah Macdonald

Sarah has worked in paediatric dietetics for the last 30 years and has gained experience in most clinical specialties. Her main areas of interest are Gastroenterology and Intestinal Failure and she is responsible for leading the service and training specialist dietitians in the management of the complex disorders seen at tertiary level.

Sarah is the author of the chapter on Gastroenterology in Clinical Paediatric Dietetics (Wiley Blackwell Publishing 2015) and is a dietetic representative of the BSPGHAN Nutrition Working Group as well as holding the post of secretary on the ESPGHAN AHP committee. She is co-course director of the annual GOSH Parenteral Nutrition Course. Sarah was involved in the Body Basics research study investigating the assessment of body composition in paediatrics.



Christina McGuckin

Paediatric PN CNS, RHC Glasgow.

I qualified from Western School Of Nursing in December 1988 as Registered Sick Childrens Nurse. As a result of no job vacancies within paediatrics, I worked with a nursing agency and was one of the first agency nurses booked by Royal Hospital for Sick Children in Glasgow and also as an Enrolled Nurse within adult hospitals until taking up a full time Staff Nurse post in a surgical neonatal unit in Royal Hospital for Sick Children, Glasgow. It was in this unit that I was successful in gaining promotion from, 'D' Grade Staff Nurse through to Acting Sister 'G' Grade, over a period of 13 years. Following successful interview, in November 2002, I commenced my present post, Paediatric Parenteral Nutrition Clinical Nurse Specialist which is now graded as a'Band 7' post. I have set up the Home Parenteral Nutrition Programme for RHC in Glasgow and I am an active stakeholder in the Scottish HPN Contract Advisory Board and in the development of Healthcare Improvement Scotland Complex Nutrition Care Standards.



Kathleen Ross

I am Lead Paediatric Dietitian at Royal Aberdeen Children's Hospital. I was originally a single handed dietitian covering everything and now manage a department of nine.

My main clinical interests are gastroenterology and inherited metabolic disease. Many of the specialist paediatric services in Scotland are delivered regionally or nationally and I have a special interest in managing complex patients at distance.

I am a past President of SSPGHAN (Scottish version of BSPGHAN) and currently Acting Clinical Lead for the North of Scotland gastro network.



Dr Richard Russell, MB ChB, MRCPCH, MRCP, PhD.

Current Occupation:

Consultant Paediatric Gastroenterologist and Honorary Clinical Associate Professor; RHC, Glasgow.

Qualifications:

MB ChB The University of Edinburgh 1988 – 1993; MRCPCH 1998; PhD The University of Edinburgh, 2007.

Papers (Peer Reviewed Journals): 90 papers, 19 letters and 4 book chapters published

Full Academic Profile www.gla.ac.uk/departments/childhealth/ourstaff/drrichardrussell

Current Grant Support:

Includes co-applicant on Medical Research Council Strategic Grant: (G0800675) The Paediatric-Onset Inflammatory Bowel Disease Cohort and Treatment Study (PICTS) and European Commission Horizon 2020 - Research and Innovation Framework Programme: Paediatric Inflammatory Bowel Diseases Network for Safety, Efficacy, Treatment and Quality improvement of care. Awarded NHS Research Scotland career fellowship 2012 with extension 2015.

Relevant previous Experience and Current Post:

I am currently one of 5 consultant paediatric gastroenterologists working in the new children's hospital in South Glasgow caring for children with PGHAN problems from across Scotland. I am the clinical lead within the department and have been involved with setting up a number of specific developments including: transition clinics, patient support days, and the development of specific patient treatment pathways. My main research interest is IBD and I have presented and published widely on this subject. I am actively involved in several clinical research trials at present. I am the departing chair of the BSPGHAN IBD working group, a member of the Paediatric ECCO committee and a member of the ESPGHAN IBD "Porto" group. I am one of 2 paediatric members of the UK IBD audit group.



Dr Tom Savage

Consultant Paediatric Radiologist with interest in body, cardiac and MSK imaging.

Post CCT fellowships in Dublin and Vancouver where I expanded my interest in gastroenterology and hepatobilliary imaging including the correlation between clinical and radiological in IBD and role of hepatic agents in paediatric MRI.

I work closely with my gastroenterology and surgical colleagues here in Glasgow providing a range of imaging guided solutions to problems our often complex GI patients face bringing this all together at MDT.



Dr Rachel Tayler

Rachel is a consultant paediatric gastroenterologist in Glasgow's Royal Hospital For Children. She graduated from The University of Aberdeen in 2005 and undertook paediatric training in the West of Scotland deanery. Rachel took up her current post in 2015 following PGHAN Grid training in Newcastle and Leeds. Her interests are Hepatology and IBD.



Mr Gregor Walker

Gregor Walker is a Consultant General Paediatric and Neonatal Surgeon in the Royal Hospital for Children in Glasgow. He works in close cooperation with the Paediatric Gastroenterology team in the management of children with inflammatory bowel disease (IBD), enteral feeding in complex patients with neurodisability, and children with intestinal motility disorders. The multidisciplinary IBD team working includes weekly planning meetings, monthly clinicopathological conferences, and joint management of patients with organised joint operating lists. His specialist interests include minimal access surgery which, along with adult surgical colleagues, he has successfully introduced for children with IBD in RHC, Glasgow.



Professor David Wilson, MB, BCh (Hons), MD, FRCP, FRCPCH

Professor of Paediatric Gastroenterology and Nutrition and Honorary Consultant in Paediatric Gastroenterology; Child Life and Health, University of Edinburgh and Department of Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh.

David qualified from Queen's University of Belfast and, following paediatric training in Northern Ireland, completed PGHAN training in HSC, Toronto. David started in Edinburgh in August 1997, setting up the new regional PGHAN service. His main clinical interest is IBD and he is clinical lead of the PGHAN service in RHSC, Edinburgh and of the regional SE Scottish PGHAN network. David's programme of academic activities includes clinical, epidemiological and translational research in IBD. He is PI of the MRC-funded PICTS cohort, a leading paediatric IBD cohort, aiding in collaborative Scottish nationwide IBD studies, including the Scottish PIBD biologicals registry. National and international collaborations include the UK and Irish Paediatric IBD genetics group, COLORS in IBD group, and both the UK and International IBD Genetics Consortia. He survived chairing the BSPGHAN IBD Working Group, and is member of ECCO and the ESPGHAN IBD Porto Group. David has gained most enjoyment from relationships with the 5 PGHAN trainees performing doctorates in IBD and 4 AHPs gaining PhDs in gastroenterology/ nutrition. IBD career grant support as PI includes MRC, CCUK, CICRA, and BSPGHAN-CORE, and as co-l includes the Wellcome Trust and EC. David has authored or co-authored >145 papers in peer-reviewed journals, over 65%

BSPGHAN 2017 Annual Meeting

PLENARY ABSTRACTS
WEDNESDAY 25TH JANUARY 2017

Dr Andrew R Barclay, Consultant Paediatric Gastroenterologist, Royal Hospital for Children Glasgow; Dr Paul Henderson, Consultant Paediatric Gastroenterologist Royal Hospital for Sick Children, Edinburgh; On behalf of 31 participating nutrition centres; 10 Paed eBANS centres

Background:

The need for accurate epidemiology regarding infants and children receiving parenteral nutrition (PN) for ≥28days (Type II intestinal failure (IF)) and long-term (type III IF) remains essential for regional and national service planning given the resource intensity, morbidity and potential need for organ transplantation in this group. Prior to 2011, BSPGHAN had performed epidemiology via the BIFS registry with occasional point prevalence survey and longitudinal registration of Type II and Type III IF. However paper reporting and the need for informed consent prior to patient registration were viewed as an obstruction to improve case ascertainment.

Aim:

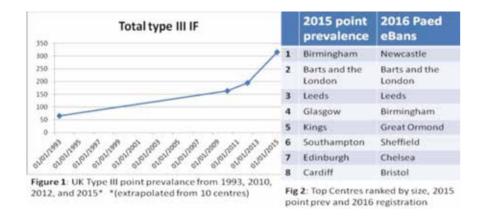
To describe the development of the Paed eBANS registry from 2011 and report epidemiological findings from this project up to 2016.

Subjects and Methods:

From election of an IF registry lead in 2011, a working group (WG) was formed to: (1) develop a primary (concise) and secondary (detailed) data set for collection;(2) explore potential new data sources, methods of collection with an aim to perform occasional point prevalence survey; (3) to start a new longitudinal registry with ability to generate point of care entry, importantly electronically and without obtaining informed consent. In 2012 it was agreed by the WG that creating a paediatric arm to the existing eBANS (www.ebans.com) was the best option with primary and secondary data agreed. Ethical extension for paed eBANS was sought and approved by the HRA in 2013 (England and Wales), and web design of the primary dataset was developed and tested over 2013-2014.

Results:

Paed eBANS went live in January 2015. Simultaneously point prevalence survey was performed in 2012 (complete ascertainment) and 2015 (incomplete). From 4 'early adopting' centres in 2015, technical support has increased this to 10 regular reporting centres in 2016 (Fig 2). These large centres represent >70% of the previous prevalent type III population in a fully ascertained survey. Point prevalence data was presented internationally and published [1]. Updated data, to included 2015 data, suggest a marked increase in HPN



127 and 144 episodes were recorded in 2015 and 2016 respectively to the registry. Short bowel syndrome remains the largest single diagnostic category (39%), but absolute number of neuromuscular disease patients also continues to grow.

Summary and conclusion:

The Paed eBANS project has demonstrated the ability to obtain complete point prevalent data on type II and type III IF. A longitudinal registry with the ability to collect ongoing data at point of care, without informed consent, now exists. The marked increase in numbers by 2015 point prevalence, suggest that ongoing accurate IF epidemiology remains a preeminent concern to BSPGHAN. Increased centre uptake and case ascertainment will be the immediate agenda for a new Paed eBANs lead from 2017 onwards.

Barclay et al Clin Nutr. 2015 Dec;34(6):1128

16.10 - 16.20 Clinical and financial benefits of a Nutrition Support Team

Tracey Johnson, Senior Specialist Gastroenterology Dietitian; Michelle Butcher, Clinical Nurse Specialist; Haidee Norton, Senior Specialist Gastroenterology Dietitian; Amanda Scott, Senior Pharmacist; Adam Henderson, Senior Pharmacist, Theo Wong, Consultant Gastroenterologist, Sue Protheroe, Consultant Gastroenterologist; Birmingham Children's Hospital, Steelhouse Lane, Birmingham, B4 6NH

Background:

The evidence-based standard for optimal nutritional support is the multidisciplinary team (MDT) approach of a Nutrition Support Team (NST). The NST at Birmingham Children's Hospital was devolved in 2007 and audit raised concern regarding lack of competence in prescribing PN, inappropriate use of PN, inadequate monitoring, high wastage and over and under provision of nutrition. The NCEPOD Report (1) and Chief Pharmacists Report (2) recommended that national standards in nutritional care needed to be improved and all children on PN should have access to a competent MDT. A business case was accepted in 2014 to fund the NST.

Aim:

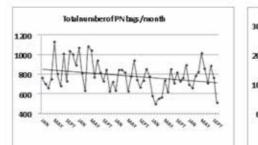
To assess improvements in clinical care and financial benefits before and after the inception of the NST at Birmingham Children's Hospital between 2012-2016

Methods:

The impact of the NST on patient care was assessed by review of biochemical monitoring, nutritional status and intake compared to an audit carried out in 2008 and educational strategies. Financial impact was assessed from total PN usage, standard bag usage and PN wastage before and after implementation of the NST.

Results:

>60% patients met their Estimated Energy Requirement and gained weight at an appropriate rate compared to only 38% meeting the EER and 26% gaining weight at an appropriate rate in 2008. Improvements in biochemical monitoring were seen. New educational strategies included the development of a Moodle providing support for staff to achieve competencies in the prescription of PN, a PN study day and a parent information leaflet. There has been a 20% reduction in the number of total PN bags/year (Fig 1) resulting from avoiding inappropriate use of PN and achievement of good nutritional status in an appropriate time frame. Standard bag usage has increased from 6% to 15% without compromising nutritional intakes. The combined cost saving is estimated at £144,000/ year. Wastage has fallen from 5% to 3% amounting to an additional cost saving of £19,000/yr.



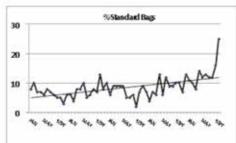


Fig 1 Fig 2

Summary and conclusion:

PN is a safe and effective treatment but management of children receiving PN requires a high level of knowledge and expertise. The interventions of the NST have resulted in sustained improvement in clinical care and cost savings.

References

- A Mixed Bag: A report by the National Confidential Enquiry into Patient Outcome and Death (2010)
- 2. Improving practice and reducing risk in the provision of parenteral nutrition for neonates and children: A report from the Paediatric Chief Pharmacists 2011

Protima Amon¹, Gloria Serena², Alessio Fasano², Allan Walker², William Wade¹, Ian R. Sanderson¹ ¹Blizard Institute, Queen Mary University London, UK; ²Harvard Clinical Nutrition Research Centre, Massachusetts General Hospital, Boston, USA.

Background:

The composition of the gut microbiota differs between healthy individuals and patients with Crohn's disease (CD) both in terms of species richness and abundances. Broad patterns of microbiota changes are emerging, including a decrease in bacterial richness, decreased proportions of taxa belonging to the Firmicutes phylum, and a rise in Gammaproteobacteria. These findings suggest that the gut microbiota plays a crucial role in the pathogenesis of intestinal inflammation. However, there still remains much that is not understood. For example, it is a challenge is to distinguish between cause and effect in the relationship between the microbiota and inflammation. Exclusive enteral nutrition (EEN) is recommended as induction therapy for active CD in children because it has strong anti-inflammatory effects. It has long been hypothesized that EEN acts by changing the microbiota. However proving this is difficult. While changes in the microbiota can be examined, it is difficult to show that they are central to its efficacy. This is because inflammation itself could, in theory, alter the microbiota. However, if children given an anti-TNF to treat inflammation did not demonstrate similar changes in the microbiota, this would be evidence that the enteral diets act primarily on the microbiota.

Aim:

The purpose of this study was to determine whether the changes in microbial composition are a result of giving EEN to children with CD rather than a consequence of reduced inflammation.

Subjects and Methods:

This was a prospective study which recruited a total of 79 children (aged < 16 years). The study period was March 2014 to September 2015. The patient groups comprised of 33 newly diagnosed CD patients started on EEN, 15 other CD children treated with anti-TNF and 31 control patients without intestinal inflammation. Stool samples were obtained prior to any treatment. The faecal microbiota was characterized by amplification and sequencing of the 16S ribosomal RNA gene V4 region using the Illumina Miseq platform. The data was analyzed using the mothur pipeline.

Results:

There were no significant differences in the disease phenotype between the newly diagnosed CD patients and the CD patients who were commenced on anti-TNF treatment. Both groups of patients had moderate to severe disease (PCDAI >30) and both treatments achieved comparable remission rates after 6 weeks of EEN (79% remission) or induction course of anti-TNF therapy (100% remission). CD patients were found to have significantly reduced species richness compared to controls (P<0.0001, Student's t test). Notably, there was no significant difference in the microbial community structure between the new-onset CD patients and the CD patients who were commenced on anti-TNF therapy (p=0.17 by AMOVA). Furthermore, the composition of the microbiota changed upon clinical remission in the group treated with EEN. Clinical remission correlated with a fall in the relative abundances of Prevotella and Faecalibacterium. The composition in disease remission remained significantly different to that of controls (p<0.001). By contrast, anti-TNF therapy was not associated with any significant changes in the microbiota composition before and after induction course of treatment (p=0.583).

Summary and conclusion:

Changes in microbiota composition are a true effect of EEN rather than a result of decreased inflammation upon disease remission. In addition, the description of specific changes in certain microbial species at disease onset and following EEN, may offer clues to disease aetiology and have potential therapeutic implications in the future.

16.30 – 16.40 Diagnostic accuracy of neutrophil-lymphocyte ratio in suspected paediatric inflammatory bowel disease: a regional cohort study

Dr Iain Chalmers¹; Professor David C Wilson^{1,2}; Dr Paul Henderson^{1,2}

¹Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh

²Child Life and Health, University of Edinburgh, Edinburgh

Background

Biomarkers such as faecal calprotectin have an established role during the investigation of paediatric inflammatory bowel disease (PIBD) but may not be available to all clinicians, especially in resource-poor settings. Availability for individual patients, especially adolescents, is often hampered further by non-provision of samples; long turnaround times for results also contribute. Therefore exploration of other readily available indices which may indicate a strong likelihood of PIBD in the absence of faecal biomarkers remains important. The neutrophil-lymphocyte ratio (NLR) can be simply calculated from a standard full blood count (FBC) panel and has been proposed as a useful marker of subclinical inflammation. Small studies in adults have demonstrated higher NLR in active compared to inactive ulcerative colitis.

Aim

We aimed to determine the diagnostic accuracy of NLR in suspected PIBD patients prior to their first endoscopic assessment.

Subjects and Methods

Children (aged <18 years) undergoing diagnostic endoscopic assessment for suspected PIBD were identified from our prospective regional cohort (IBD database and local endoscopy lists) from 2005-2011. Laboratory records were reviewed to identify the FBC performed closest and prior to endoscopy. Patients were eligible for inclusion if complete FBC data was available within 6 months prior to endoscopy and importantly during the initial diagnostic cycle. Statistical analyses were performed using Vassarstats and EasyROC v1.3.

Results

182 patients met the inclusion criteria with 89 cases of PIBD and 93 controls with non-IBD diagnoses or normal investigations. The PIBD group were older (12.6yr vs 8.8yr; p<0.0001) with no difference in sex ratio (p=0.575). Median time from blood sample to endoscopy was 16 days (IQR 0-49) and was lower in the PIBD group (4d vs 37d; p<0.0001). NLR was significantly higher in PIBD (median 3.04, IQR 2.03-4.18) than in non-IBD patients (1.4, 0.95-2.03; p<0.0001). There was no significant difference in NLR between patients with Crohn's disease (3.05, 2.26-4.20) and those with ulcerative colitis or IBDU (2.59, 1.72-4.01; p=0.503). The area under the receiver operator curve (AUROC) for NLR as a diagnostic test for PIBD was 0.81 (95% confidence interval 0.75-0.88). Using an optimal cut-off for NLR of 2.37 gave a sensitivity of 67% (95% CI 57-77) and specificity 85% (95% CI 76-92). In patients with otherwise normal blood markers (CRP, ESR, albumin, WCC, platelets), median NLR remained significantly higher in the PIBD group (2.32(1.74-3.28) vs 1.43(1.01-1.97); p<0.001, AUROC 0.77 (95% CI 0.65-0.89)). Using any abnormal test from CRP, ESR, albumin, platelets, WCC and NLR, sensitivity was 89% (95% CI 81-95), specificity 60% (95% CI 48-72), positive predictive value 72% (95% CI 62-80) and negative predictive value 83% (95% CI 70-92).

Summary and conclusion

NLR is significantly higher in patients with PIBD compared to controls with other non-IBD diagnoses or normal investigations. This remains the case in patients with otherwise normal inflammatory markers. NLR provides moderate sensitivity and specificity for the diagnosis of PIBD and sensitivity is improved by combination with other readily available blood parameters. We propose that NLR has a useful place in the diagnostic work-up of patients with PIBD, especially in the resource-limited setting and where there are barriers to obtaining faecal samples.

Neil Chanchlani, Specialist Trainee in Paediatrics, Royal Free London NHS Foundation Trust, London; Kajal Mortier, Project Manager, Royal College of Physicians, London; Mike Cosgrove, Consultant Paediatrician, Morriston Hospital, Abertawe Bro Morgannwg University Health Board, Wales; Marcus Auth, Consultant in Paediatric Gastroenterology, Hepatology, and Nutrition, Alder Hey Children's Hospital, Liverpool; Rafeeq Muhammed, Consultant Paediatric Gastroenterologist, Birmingham Children's Hospital, Birmingham; Huw Jenkins, Consultant Paediatrician, Cardiff & Vale NHS Trust; Andrew Fagbemi, Consultant Paediatric Gastroenterologist, Central Manchester and Manchester Children's University Hospitals NHS Trust, Manchester

Background

Infliximab biosimilar drugs have been available in the UK since February 2015.

Aim:

Routinely collected data from the UK IBD biologics registry is the first to include Infliximab biosimilar (IFX-B) drugs, (Inflectra $^{\text{TM}}$ and Remsima $^{\text{TM}}$), as alternatives to the originator Infliximab (IFX-O) (Remicade ®). We sought to summarise the short-term efficacy, safety, and cost of using IFX-B compared to IFX-O in biologic naive patients.

Subjects and Methods:

Prospective audit of 278 patients across 27 paediatric sites between March 2015 to February 2016; 1050 patients have been in the audit since its inception in 2011. Pre-treatment screening was with chest radiograph, Hepatitis B/C serology, and Mantoux or Gamma interferon. Disease severity, response to treatment, and remission rate was measured by Paediatric Crohn's Disease Activity Index (PCDAI) and Physician Global Assessment. Results are presented as number (5) or median (interquartile range). Cost was determined at current market value distributed and projected estimate of savings [1,2].

Results:

Between Mar 2015 to Feb 2016, 278 patients: 175 (63%), 82 (29%), and 21 (8%) were newly started on IFX-O, IFX-B, and Adalimumab respectively. Screening was adequately undertaken for 47% (109/230) of patients prior to starting treatment. Male distribution (61% in IFX-O vs 60% in IFX-B) (p>0.05), age at diagnosis (12 years), and age at biologic initiation (14 years) was similar between both IFX groups.

IFX-O was commenced in 61% (n=129) of patients with Crohn's disease (CD), 68% (n=32) of patients with ulcerative colitis (UC), and 70% of patients with inflammatory bowel disease unclassified (IBDU). IFX-B was commenced in 30% (n=63) of CD patients, 30% (n=14) of UC patients, and 25% (n=5) of IBDU patients.

At baseline, 86% (n=150) and 79% (n=65) in the IFX-O and IFX-B groups respectively received immunosuppressants and 29% (n=51) and 31% (n=25) in the IFX-O and IFX-B groups, respectively, received steroids (p>0.05 for both). Median PCDAI score was 36 (20,48) (n=42) in the IFX-O group and 28 (20,40) (n=29) in the IFX-B group.

At 3-month follow up, median PCDAI score was 5 (0,11) (n=19) and 0 (0,8) (n=15) in the IFX-O and IFX-B groups respectively. Response to treatment was 85% (n=28) and 86% (n=19) with remission rates of 68% (n=25) and 79% (n=19) in the IFX-O and IFX-B groups respectively (p>0.05). IFX-B was then compared to historical patients commencing IFX-O or Adalimumab (2011 – 2015), whose response to treatment at 3-months was 74% (n=158) (p=0.05) and remission rate was 65% (n=144) (p=0.37).

Adverse events (AE) were recorded at time of initial treatment in 3/175 patients and 0/82 patients in the IFX-O and IFX-B groups respectively. At 3 months, 4/76 and 2/39 AE were reported in the IFX-O and IFX-B groups respectively (both p>0.05). When comparing IFX-B data with IFX-O or Adalimumab (2011 – 2015), <0.05% patients reported AE on initiation (p=0.26) and <0.1% of patients reported AE on follow-up (p=0.45). With a conservative estimate £875,000 would have been saved with universal adoption of biosimilars in patients included in this audit alone.

Summary and conclusion:

At initiation and 3-month follow-up, IFX-B are as effective as IFX-O in treating IBD in comparable paediatric populations. No increase in AE was reported. Sites should adopt biosimilars for all new starts due to the cost reduction with no difference in any other parameters.

1] National Institute of Clinical Excellence. Infliximab and Adalimumab for the treatment of Crohn's disease. TA187. [Published May 2010] https://www.nice.org.uk/guidance/ta187/chapter/3-the-technologies

[2] Jha A, et al. The Budget Impact of Biosimilar Infliximab (Remsima®) for the Treatment of Autoimmune Diseases in Five European Countries. Adv Ther. 2015 Aug;32(8):742-56. doi: 10.1007/s12325-015-0233-1. Epub 2015 Sep 5.

16.50 – 17.00 The State of Grid Training: Looking to the Future Training of Paediatric Gastroenterologists and Hepatologists in the UK

Dr Edward Gaynor, Trainee Representative of PGHAN CSAC & ST8 Paediatric Gastroenterology Registrar, Kings College Hospital; Dr Sue Protheroe, Chair of PGHAN CSAC & Consultant Gastroenterologist, Birmingham Children's Hospital

Background

In 2002 the RCPCH introduced the 'National Grid' to provide equity of access to subspecialty training and aid workforce planning. The provision of high quality paediatric gastroenterology, hepatology and nutrition (PGHAN) training is overseen by the College Specialist Advisory Committees (CSACs) through site inspections.

Aim

As part of an engagement exercise with trainees and Grid Centres, CSAC requested qualitative feedback on current training in the UK, in order to provide clarity on the current challenges, and to maintain and improve the quality of PGHAN Grid training.

Subjects and Methods

Between June and December 2016, all PGHAN Grid centres, trainees and newly appointed consultants were approached to complete a questionnaire detailing the requirements expected to delivery quality subspecialty training.

Results

Feedback was received from 19 PGHAN Grid Centres (3 liver Centres, 16 of 18 gastroenterology centres) and 31 trainees or newly appointed consultants.

Centres reported differing service models in delivering PGHAN services, with all trainees and centres declaring that patient needs and safety were put first.

Supervision and staffing - All grid centres reported a minimum of 70% working time in speciality, however 32% of trainees self-reported training below this requirement. Similarly 29% of trainees claimed they worked more than 1/3 of hours in our of hours delivery of care. A theme of "poor staffing levels in acute or out of hour staffing" was commented on as a significant cause. Clinical supervision was positively reported with 87% of trainees able to meet with their education supervisor every 2 months and 100% having annual multi-source feedback.

Training and education - 97% of trainees reported sufficient clinical experience, with 94% of trainees having sufficient training in their subspecialty. Notably only 52% of trainees felt they had adequate training in anthropometrics and 58% in transitional care.

Clinical skills – although meeting CSAC requirements of one or more training lists per week, trainees reported differing access to endoscopy: an average of 9 upper GI endoscopies per month (range 1-20) and 5 colonoscopies per month (Range 1-10). Trainees commented that service and out-of-hour commitments, or sharing lists with other fellows or trainees limited access to endoscopy.

Trainees and trainers were asked to rate the opportunities in their posts, rating the overall PGHAN training positively – 8 out of 10 (range 4-10) and 9 out of 10 (range 8-10) respectively.

Summary and conclusion

Trainees and recently qualified consultants reported achieving subspecialty competencies during their three-year period of training, through rotation to more than one centre. In the face of competing service demands, access to specific training may become a challenge, and it will be important to ensure quality training is maintained across PGHAN Grid Centres in the future

BSPGHAN 2017 Annual Meeting

PLENARY ABSTRACTS THURSDAY 26TH JANUARY 2017

13.30 – 13.40 Development of a Paediatric Endoscopy Global Rating Scale - Results of a National Pilot

Dr P Narula¹, Mr R Broughton², Dr R Bremner³, Dr A Piggott⁴, Dr D Rawat⁵, Mr M Cullen⁶, Dr NA Afzal6, Dr L Howarth⁷, Dr P Gillett8, Dr P Henderson⁸, Dr K Venkatesh⁹, Dr C Tzivinikos⁹, Ms J Maginnis⁴, Ms S Mckenna¹, Dr D Devadason¹⁰, Dr S Loganathan¹⁰, Dr M Stanton⁶, Dr J Green², Ms D Johnston²,

¹Sheffield Children's Hospital; ²JAG; ³Birmingham Children's Hospital; ⁴Royal Stoke Children's Service; ⁵Royal London Hospital; ⁶Southampton Children's Hospital; ⁷John Radcliffe Children's Hospital; ⁹Alder Hey Children's Hospital; ¹⁰Nottingham

Background

The Endoscopy Global Rating Scale (GRS) provides a clear framework for service improvement and underpins the accreditation of endoscopy services. The GRS is supported by a web-based self-assessment tool and provides a structure for endoscopy services to prioritise tasks in the first instance and is supported over time with a web-based knowledge management system linking solutions directly to challenges. The endoscopy GRS was originally developed as a patient centred quality improvement tool in 2005 for the adult services to drive up standards. The adult experience suggests demonstrable improvement in quality and safety with embedding of standards through the accreditation process. Services are required to score a Level B in all standards in order to apply for and, once achieved, to maintain accreditation from the Joint Advisory Group in GI endoscopy (JAG). An adapted GRS was piloted by two paediatric tertiary GI endoscopy services in 2013 and this highlighted that further progress towards a paediatric accreditation required a more usable and appropriate quality assessment tool.

Aim

To develop a paediatric endoscopy GRS (P-GRS) as a quality improvement tool which would also define standards for accreditation of paediatric endoscopy services

Methods

A P-GRS working group was established in May 2015 and the membership of this group evolved to include representatives from the BSPGHAN endoscopy working group, district general hospital, paediatric surgery, allied health professional, and specialist input from the JAG. Feedback was actively sought from the patients and parents partnership group. A draft P-GRS was developed following extensive discussion and communication between the P-GRS working group and the JAG. This was sent out for consultation to the regional endoscopy leads and BSPGHAN council in October 2015. The regional endoscopy leads led on the next phase of consultation with the individual endoscopy leads in their region during November 2015. The draft version was revised based on feedback received following communication between the P-GRS working group and the JAG. Nine sites agreed to pilot the P-GRS (Alder Hey, Birmingham, Edinburgh, Nottingham, Oxford, Royal London, Sheffield, Southampton and Stoke on Trent) and all pilot sites underwent a training day organised by JAG in May 2016. The first assessment against the P-GRS was completed in June 2016. A second assessment is to be completed in December 2016 and the results of this will be available before the BSPGHAN annual meeting to share with the membership.

Results

Eight of the nine pilot sites completed a GRS assessment in June 2016, testing the standards in their service and identifying an action plan for their unit. The first results showed significant variation across the standards, which is in keeping with the first adult GRS submissions.

In the clinical quality domain, 12% of units achieved a level B or above in the leadership and organisation standard, 25% in safety, 63 % in comfort, 25% in quality, 38 % in appropriateness and 50% in results. In the quality of patient experience domain, 38% of the units achieved a level B or above in the respect and dignity standard, 24% in the consent process including patient information, 38% in the patient environment and equipment, 12% in access and booking, none in productivity and planning, 38 % in aftercare and none in patient involvement

In the workforce domain, 12% of the units achieved a level B or above in the teamwork standards, 26% in workforce delivery, 38% in professional development.

In the training domain, none of the units achieved a level B or above in the environment, training opportunity and resources standard, trainer allocation and skills standard or assessment and appraisal standard.

Conclusion

These results highlight a need for improvement in all paediatric endoscopy services. Overall, all units agreed this was a positive experience and this enabled the units to develop an action plan to improve their endoscopy services and share good practice. All pilot sites reflected on the measures in the P-GRS to ensure they were relevant to paediatric endoscopy services and fit for purpose.

Mike Thomson; Prithviraj Rao; Priya Narula; Arun Urs; David Campbell; Dalia Belsha All of the authors are consultant gastroenterologists at the Sheffield Children's Hospital.

Background

Acute upper gastrointestinal bleeding (AUGIB) remains virtually the last paediatric emergency that is still managed badly in many centres. Attendant mortality is mainly contingent on the lack of recognition that emergency endoscopy may be life-saving and the timing of such but, importantly, mortality is also predicated upon the skill mix and experience of the endoscopist. Advanced endo-haemostatic technique performance and experience is extremely variable in distribution amongst pediatric endoscopists in the UK and other countries, partly due to their sophistication and difficulty but also because of the sparsity of cases and contingent lack of familiarity of the endoscopists in their use. A novel endo-haemostatic topically applied powder (Hemospray®) has the advantage of extreme ease of use and hence may lower the threshold of competency required by the endoscopist thereby potentially reducing mortality. A recent adult study reported technical success of 88% and re-bleeding risk of 16% with no reported adverse events. (1)

Prospective evaluation of the efficacy and the safety of Hemospray® in paediatric AUGIB.

Subjects and Methods

Prospective enrolment of children fulfilling the Sheffield AUGIB score for likely need for endohaemostatic intervention of >8/24. (2) Hemospray® was applied using the Cook application device and a follow up endoscopy at 72 hours, 7 days and 1 month was offered to the families and occurred in those deemed to have clinical need pre-discharge. Regional Ethics Committee approval was obtained.

Results

14 applications of Hemospray® occurred in 12 patients (7 male), 7.1 (0.7-15.0) years, 25.1 (10.3-67.8) kg. Sheffield scoring system for likely requirement for endoscopic intervention due to ongoing active bleeding was applied with 8/12 having had blood transfusion and 8/12 with Hb drop of >20g/l. 10 patients had pre-existing conditions, one patient had excessive NSAID ingestion and one patient had no past history of note. Hemospray® application was easy, took 8 (5-15) minutes, was immediately successful in achieving haemostasis and no adverse events were observed. No other endo-haemostatic technologies were employed. Repeat endoscopy in 6 patients at 72 hours revealed healing of the bleeding lesion, however 2 patients required a second application due to ongoing bleeding - one also required endoclip application and one required laparotomy which was temporarily successful but subsequent interventional radiographic embolization of the afferent vessel was needed.

Pre-existing condition	Findings on endoscopy		
NSAID intake (4 hourly for 48 hours)	Gastric ulcer		
TPN dependant, short gut due to volvulus	Gastric ulcer x2		
Nil of note	Gastric ulcer		
Portal hypertension	Erosive gastritis		
Post-polypectomy of large gastroduodenal polyp	Post-EMR Gastric bleed		
Small bowel diaphragm disease	Post-web division bleed		
Duchene muscular dystrophy	Duodenal and gastric ulcer		
Acute pancreatitis	Erosive gastritis and duodenitis		
Glanzmann's thrombocythaenia	Actively bleeding angiodysplasia		
GVHD, ileal stricture, ileostomy	Bleeding ileal ulcers		
Hereditary spherocytosis, recent splenectomy	Gastric ulcer		
Cerebral palsy, seizures	Haemorrhagic gastritis		

Summary and conclusion

Hemospray® appears effective in the majority of paediatric AUGIB in this preliminary prospective series, is easy and quick and is associated with no observable adverse events. It has the potential to transform the management of this emergency due to its ease of performance, thereby opening the door to relatively inexperienced paediatric endoscopists.

References

- 1. Changela K, Papafragkakis H, Ofori E et al. Hemostatic powder spray: a new method for managing gastrointestinal bleeding. Therap Adv Gastroenterol. 2015;8(3):125-35.
- 2. Thomson M, Leton N, Belsha D. Acute upper gastrointestinal bleeding in childhood and endoscopic management: development of the Sheffield scoring system to predict the need for endoscopic intervention. Journal of Pediatric Gastroenterology and Nutrition. 2015:60:632-6.

13.50 - 14.00 Systematic review and meta-analysis of hepatic outcomes in non-alcoholic fatty liver disease after bariatric surgery in children.

Jake P. Mann: Department of paediatrics, University of Cambridge

Valerio Nobili: Hepatometabolic Unit, Bambino Gesu Hospital, IRCCS, Rome, Italy

Background

Bariatric (weight loss) surgery is indicated for morbidly obese patients with obesity-related complications, including non-alcoholic fatty liver disease (NAFLD). Data from adults is heterogeneous but surgery can result in histological improvement. Whilst there is clear evidence of improving metabolic outcomes (e.g. type 2 diabetes), the impact of bariatric surgery on liver-related outcomes in children is unclear.

Aim

We aimed to perform a systemic review assessing the impact of bariatric surgery on liver-related outcomes in paediatric NAFLD.

Subjects and Methods

A systematic review of MEDLINE for: "bariatric surgery" and "nonalcoholic steatohepatitis (NASH)" or "nonalcoholic fatty liver disease" or "fatty liver" in "children" or "adolescents" from any date until November 2016. Studies were excluded if there was no assessment of NAFLD post-operatively or if children (<18 years) were not included. Data extracted: surgical procedure, demographics, biochemistry, and histology (where available). Primary outcome was histological resolution of NASH. Secondary outcomes were improvement in fibrosis or non-invasive markers of NAFLD. Paediatric NAFLD Fibrosis Index (PNFI) was calculated as a non-invasive marker of fibrosis and AST/ALT ratio as a marker of NASH.

Results

87 studies were identified, of which 4 met all inclusion criteria. The population comprised 169 children (126 female (75%)), mean age 17.4 years and follow-up 15.2 months. 81/169 (48%) underwent baseline biopsy, demonstrating NASH in 32/81 (40%). Post-operative biopsy data was available for 41 patients, with complete NASH resolution in 27/41 (66%) and fibrosis improvement in 26/41 (63%).

There were no statically significant changes in pre- vs. post-operative mean PNFI (9.94 vs. 8.82) or AST/ALT ratio (0.89 vs. 0.94).

All studies were assessed as medium-high risk of bias due to: lack of randomization, no blinding, and incomplete reporting of outcomes. There was high heterogeneity of methods for assessing hepatic outcomes across studies.

Summary and conclusion

Bariatric surgery results in histological improvement in 2/3rd of children with biopsy proven NAFLD. Non-invasive assessment did not demonstrate improvement of NASH or fibrosis. Results are promising but there is currently insufficient data to recommend bariatric surgery as a primary treatment for NAFLD in children. Future trials should include validated end-points for paediatric NAFLD.

14.00 – 14.10 Ethanol and Taurolidine line locks for the prevention and treatment of catheter related bloodstream infections in paediatric intestinal failure: a systematic review and meta-analysis

Dr lain Chalmers, ST8 Paediatric Gastroenterology¹; Dr Paul Henderson, Consultant Paediatric Gastroenterologist^{1,2}; Dr Rachel Tayler, Consultant Paediatric Gastroenterologist³; Professor David Wilson, Consultant Paediatric Gastroenterologist^{1,2}; Dr Andrew Barclay, Consultant Paediatric Gastroenterologist³

¹Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh; ²Child Life and Health, University of Edinburgh, Edinburgh; ³Department of Paediatric Gastroenterology, Royal Hospital for Children, Glasgow

Background

Catheter-related bloodstream infections (CRBSIs) remain a significant concern in children with paediatric intestinal failure (PIF); that is dependency on parenteral nutrition (PN) for ≥28 days. These patients are dependent on central venous catheters (CVCs) and CRBSIs contribute to morbidity and mortality, notably increasing the risk of intestinal failure associated liver disease (IFALD) and the loss of suitable veins for central access. Line locks using 70% ethanol or taurolidine are increasingly used as part of strategies to reduce CRBSIs.

Aim

We aimed to systematically review the evidence for the use of ethanol and taurolidine line locks in the prevention and treatment of CRBSI in PIF. Primary outcome of interest was reduction in the rate of CRBSI per 1000 catheter days.

Subjects and Methods

A systematic review of the literature was performed using electronic searches of the Cochrane Library, MEDLINE (Ovid) and EMBASE (Ovid) from inception to 20th November 2016 with the keywords and MeSH terms "intestinal failure", "child", "ethanol" and "taurolidine". Hand searches and review of reference lists were also performed. Two authors independently assessed the level of evidence of included studies using established SIGN methodology (www.sign.ac.uk). Where possible results were combined in a meta-analysis using RevMan v5.3.

Results

The search strategy yielded a total of 3,572,413 hits. Combination of the search terms above reduced this to 2689 records. Review of title and abstracts identified 22 studies which were reviewed in detail. 7 studies were excluded as they contained only adult data or data relating specifically to PIF could not be extracted. 15 studies were analysed comprising a total of 196 patients and 98,295 catheter days. 13 studies used 70% ethanol and two used taurolidine; all studies showed a reduction in CRBSI rates. 11 studies were evidence level 2- (case control or cohort studies with a high risk of confounding or bias) and the remaining 4 evidence level 3 (non-analytic studies, e.g. case reports, case series) demonstrating that the evidence remains poor. A meta-analysis of 8 of the studies using ethanol locks demonstrated a reduction in CRBSI of 6.11 (95% CI 4.79 - 7.42; p<0.001) per 1000 catheter days (Figure 1). Data on CVC blockage and thrombosis with ethanol locks varied between studies.

	Pre-ethanol Post-ethan			ol	Mean Difference			Mean Difference		
Study or Subgroup	Mean	50	Total	Mean	SD	Total	Weight	IV, Fixed, 95% CI	Year	IV, Fixed, 95% CI
Mouw 2008	8.87	7.77	- 5	1.17	1.63	10	3.7%	7.70 [0.81, 14.59]	2008	
Jones 2010	10.05	5.63	23	2.95	2.78	23	26.3%	7.10 [4.53, 9.67]	2016	_
Wales 2011	10.2	6.2	10	0.9	1.8	10	10.8%	9.30 [5.30, 13.30]	2011	
Cober 2011		5.4	15	1.3	3	15	17.7%	6.70 [3.57, 9.83]	2011	
Pieroni 2013	10.2	5.9	13	2.8	2.4	13	14.5%	7.40 [3.94, 10.86]	2013	
Mezoff 2015	8.2	7.8	30	4.9	5.9	30	14.2%	1.30 [-2.20, 4.80]	2015	
Kawano 2016	6.77	2.48	4	3.13	3.2	4	11.0%	3.64 [-0.33, 7.61]	2016	-
Mokha 2015	10.4	8.98	29	4.6	17.12	13	1.8%	5.90 [-4.06, 15.66]	2016	
Total (95% CI)			129			118	100.0%	6.11 [4.79, 7.42]		•
Heterogeneity Chi*=	12.64, 1	f=70	P = 0.0	8); F = 4	5%				-	- t t t t
Test for overall effect										Favours Hepatin Locks Favours Ethaniol Locks

Figure 1: Meta-analysis of 8 studies on the use of ethanol for CRBSI prevention

Summary and conclusion

Existing studies of ethanol line locks are of low evidence level but consistently demonstrate reduction in CRBSI rates. The data available for taurolidine locks is less compelling, with only two small studies available. Future studies comparing ethanol and taurolidine locks, and comparing differing ethanol administration regimens are warranted. Given the overall reduction in global CRBSI rates in PIF, any prospective RCT would need to be multi-centred to be adequately powered.

14.10 – 14.20 Hepatopulmonary syndrome in children – symptoms, clinical progression and outcome post Liver transplant.

¹S. Warner, ¹PJ McKiernan, ¹DA Kelly, ¹ID van Mourik, ¹G Gupte, ¹J Hartley, ¹M Abdel-Hady, ¹K Sharif, ²D Mirza, ²P Muiesan, ²T Perera, 1SV Beath.

¹Birmingham Children's Hospital and ²The Queen Elizabeth Hospital, United Kingdom.

Background

Hepatopulmonary syndrome (HPS) is a rare but serious complication of chronic liver disease that carries a high morbidity and mortality.

Liver transplant (LT) is curative but presentation can be insidious and therefore a high index of suspicion needs to be maintained.

Aim

To evaluate presenting symptoms in patients with HPS and compare their outcome post LT.

Subjects and Methods:

Retrospective review from 1996 to 2016 of case notes and electronic database in patients diagnosed with HPS. The following factors were reviewed;

- presenting symptoms of HPS
- clinical progression during wait for LT
- Oxygen saturation in room air (SpO2 %) from HPS diagnosis to LT
- Haemoglobin (Hb g/L) from HPS diagnosis to LT
- Outcome post LT (survival, length of hospital stay and oxygen supplementation)

Results:

Pre- Liver transplant: 20 patients with HPS were identified between 1996 to 2016. Median age at HPS diagnosis was 10yrs. Symptoms at presentation were Dyspnoea with daily activities (n=17), cyanosis (n=8), and pneumonia (n=3). Diagnosis was confirmed by VQ scan (n=13) or Contrast Echocargdiogram (n=7). Seventeen patients were listed for LT. Three patients had co-morbidity which contra-indicated LT. One patient was removed from the wait list with optimization of medial therapy. Median wait from listing to transplantation was 8 weeks. 14 patients had a drop in oxygen saturation in room air from a median of 92% at diagnosis to 88% at LT, 9 required Home oxygen for progressive cyanosis and 2 had recurrent respiratory tract infections. 15 became more polycythaemic; Hb range at diagnosis was 91 - 157 g/L, compared to 119 - 180 g/L on the day of transplant.

Post Liver transplant: 15 patients made a full recovery and were discharged home from hospital; one patient died from sepsis within two weeks of LT. All 15 survivors were weaned off supplementary Oxygen (O2). Median parameters; PICU stay n=3days (days on assisted ventilation n=1), duration of supplementary oxygen requirement n=12days and hospital stay n=20days. Respiratory complications post LT were pneumonia (n=3), pulmonary oedema (n=2) and persistent intrapulmonary shunting (n=1)

Correlation analysis of paired data found the following to be of statistical significance:

- SpO2 <89% in room air on the day of LT (prior) and length of hospital stay (p 0.02)
- \bullet Hb >131 g/L on the day of LT and supplementary O2 post LT (p 0.01)
- Hb >131 g/L on the day of LT and the duration of assisted ventilation requirement post LT (p 0.01)
- Length of wait for LT and duration of supplementary O2 post LT (p 0.01)

Summary and conclusion:

Diminished exercise tolerance in the setting of chronic liver disease should prompt screening for HPS. Patients had progressive hypoxia and polycythaemia on the wait list which correlated with longer hospital stay post LT. Length of wait to LT also correlated with longer supplementary oxygen requirement post LT. Children with HPS have excellent outcome post LT and therefore should be considered for transplantation without delay.

48 48 4

14.20 - 14.30 Portal Vein Obstruction: The Challenge of Timely Diagnosis

D Belsha, S Davison, S Hodges, S Rajwal, P McClean. Children's Liver Unit, Leeds General Infirmary, Leeds LS1 3E

Background

Portal vein obstruction (PVO) due to portal vein thrombosis with portal cavernoma formation is a major cause of portal hypertension in children. Significant morbidity occurs due to oesophageal variceal bleeding. Early diagnosis permits appropriate counselling and provides an opportunity for prophylactic intervention. We aimed to evaluate the spectrum of clinical presentation of PVO, and identify opportunities for early diagnosis.

Methods

All children born after 1990 with a final diagnosis of PVO who were referred to a single centre between 2000 and 2015 were identified. A proforma was devised and data collected retrospectively from case notes including initial symptoms, signs and investigations, incidence of gastrointestinal (GI) bleeding and time to diagnosis.

Results

ifty-six children with PVO were identified, one was later excluded due to insufficient data. Mean age at presentation was 3y 10m (range 1 m -13y 8 m), 26 were male. Risk factors were identified in 28/55 (51%) including umbilical vein instrumentation (12), congenital heart disease (10: 4 also had non-cardiac abnormalities) and other congenital anomalies (6: renal, anorectal, tracheo-oesophageal fistula and Goldenhar syndrome). Four patterns of presentation were identified: (a) incidental finding of PVO on US (n=9); (b) neonatal with jaundice +/- ascites leading to US diagnosis of PVO (n=4); (c) upper GI bleed (n=22) and (d) "haematological presentation" with splenomegaly and/or symptoms/signs of hypersplenism (n=20). Of 22 who presented with GI bleeding (mean age 3y 6m, range 6m-10y6m) 15 had diagnosis of PVO made during their initial admission. The remaining 7 were discharged without PVO being identified. All were subsequently readmitted after a median of 1m (range 7d - 18m) with further GI bleeding, after which the diagnosis was established. Three children with GI bleed at presentation were initially referred to haematology for investigation of thrombocytopenia, two undergoing bone marrow (BM) assessment. Of 20 children (mean age 5y 2m, range 17m-13 y), without GI bleed who had a "haematological presentation", 13 (65%) were referred to specialists in haematology (11) or immunology (2). BM assessment by trephine or aspiration was performed in 9/20 (45%) and was normal. Only 9/20 (45%) in this group had diagnosis of PVO established within 2 weeks of presentation. In the remaining 11 median interval to diagnosis was 10 m (range 2m - 6y). Of these, 10 had a significant upper GI bleed during the interval from presentation to diagnosis. In those who had evidence of splenomegaly on initial US, PVO was not initially identified.

Conclusion

PVO has a wide spectrum of clinical presentation. Associated risk factors are seen in 51% and may provide a clue to diagnosis. In this series diagnosis was delayed in 18/42 (43%), most frequently in those with a "haematological" presentation (55%) but also in those presenting with GI bleed (32%). Diagnosis by US can be technically challenging even when splenomegaly is detected. Carefully focussed imaging by US with Doppler studies and/or other radiological modalities should be undertaken in a child with history, symptoms or signs compatible with PVO.

BSPGHAN 2017 Annual Meeting

PLENARY ABSTRACTSFRIDAY 27TH JANUARY 2017

14.30 – 14.40 Awareness of ESPGHAN guidelines on coeliac disease amongst general paediatricians in Southwest England

Dr Siba Paul, Bristol Royal Hospital for Children; Miss Helen Adams, University of Bristol; Dr Dharam Basude, Bristol Royal Hospital for Children

Background

Aims

1) To gain an understanding of awareness and use of ESPGHAN guidelines for diagnosing CD in children amongst general paediatricians

2) Provide recommendations to increase awareness if required.

Methods

A telephone/email survey was conducted of general paediatric consultants (n≈140) across Southwest England with 11 DGHs. Survey included 8 questions to assess awareness and use of ESPGHAN guidelines, incorporating 3 main themes: when non-biopsy diagnoses can be made, when HLADQ2/8 genotyping should be requested and whether asymptomatic children from high-risk groups with anti-tTG>10xULN can be diagnosed without a biopsy.

Results

85/140 (61%) responses obtained. 99% paediatricians are aware of ESPGHAN guidelines and non-biopsy/biopsy pathways for diagnosing CD. 83% of paediatricians were unable to state all conditions required for non-biopsy diagnosis. None could describe all appropriate situations where HLA-DQ2/8 genotyping should be requested. 33% of paediatricians responded that asymptomatic children with anti-tTG>10xULN can be diagnosed with CD without a biopsy while 24% said they were unsure or would seek advice.

Summary and Conclusions

Survey highlighted need for greater in-depth awareness of non-biopsy pathway and situations where HLA-DQ2/8 genotyping is indicated. There is possible misinterpretation regarding the ESPGHAN guidelines as 1/3rd of paediatricians considered non-biopsy pathway is applicable for asymptomatic children with anti-tTG>10xULN. There is need for improved understanding of the ESPGHAN guidelines amongst DGH paediatricians. A user friendly Apps is planned to improve the diagnostic process.

14.30 – 14.50 Diagnostic endoscopy in children with GI symptoms: Indications and Outcomes Prospective Study

A Kadir, Locum Consultant Paediatric Gastroenterologist; S Naik, D Rawat, N Meadows, P Amon, N Croft

Royal London Hospital

Background

Gastrointestinal endoscopy is an invasive procedure used to diagnose and/or treat diseases of the gastrointestinal tract. As with any invasive procedure, there is a small risk for complications which includes risk of general anaesthesia as well as the complications of procedure itself – pain, perforation, bleeding and infection. It is therefore important that due consideration is taken when reviewing the indications for endoscopy, particularly in children

Aim

The aim of the study was to prospectively evaluate the prediction of abnormalities for first upper GI endoscopies and colonoscopies done in children and to correlate the indication for the procedure with expected contributive yield in clinical practice. A second aim was to examine variation of practice in decision making for GI endoscopy nationally in the UK

Subjects and Methods

This study was a descriptive, prospective analysis, conducted at the department of paediatric gastroenterology, Royal London Children's Hospital Barts Health NHS Trust. All children who had first upper GI endoscopies and colonoscopies from April 2016 to June 2016 were included. Exclusion criteria were any interventional procedures. A standard questionnaire was filled in by the clinician at the time of requesting the endoscopy for prediction of abnormalities, which was compared to macroscopic and histology findings. At the time of endoscopy the endoscopist defined the macroscopic outcomes as normal or abnormal which were recorded. 2 consultant histo pathologist were involved in analysis of all biopsy specimens which were reviewed in a weekly multidisciplinary meeting To examine the possibility of differences in clinical decision making we also created an online survey of 6 different cases and through the endoscopy working group of BSPGHAN, a link for the survey was sent to paediatric gastroenterologist (consultants and trainees) throughout the United Kingdom. The survey was anonymized

Results

A total of 63 children had endoscopies in the period of 3 months, 14 children had repeat endoscopies therefore were excluded from the study leaving 49 children. Out of 49 children 28 (57.1%) were male and 21 (42.9%) were female. 26 children had upper GI endoscopies, 8 had colonoscopies and 15 children had both procedures. Our study suggested that the most common symptom for which upper GI endoscopy was requested were symptoms of upper abdominal pain (34%), for colonoscopy was rectal bleed (76%) and for request of both upper GI and colonoscopy was generalised abdominal pain (47%). Histological findings were abnormal in 46% of children with upper abdominal pain, 30% of children with reflux symptoms, 20% of children with rectal bleed and all endoscopies were normal in children with symptoms of vomiting. The predictability of a normal endoscopy (32.65%) was more accurate as compared to the predictability of abnormal histology findings (23%)

Summary and conclusion

Our study is the largest prospective clinical evaluation of paediatric OGD, colonoscopy and upper and lower endoscopy in a UK tertiary paediatric gastroenterology unit. We still need a larger prospective multicentre studies in order to develop appropriate guideline for patient selection to maximise diagnostic yield

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14.50 – 15.00 Prospective Paediatric Appendicitis and Risk of Perforation from Prehospital Delay

Dr David I Campbell SCH, Western Bank Sheffield; Alexander Labeit, ScHARR, Health Economist, University of Sheffield; Basil Bekdash, Paediatric Surgical Trainee, University of Oxford; Dipanker Dass, Paediatric Surgical Trainee, Sheffield Children's Hospital; Sean Marven, Surgical Consultant, Sheffield Children's Hospital; Tracey A Young ScHARR, Statistician, University of Sheffield

Background

Literature on whether prolonged pre-surgical time lead to a higher rate of appendiceal rupture or abscess formation are confounded by the methodology of those studies. Recall bias tends to overestimate the pre-hospital illness in life threatening acute illnesses. Confirming whether prehospital delays using more recent techniques to minimize recall bias (Time Line Follow Back) will more confidently quantify this association with disease severity and allow intervention studies to promote prompt medical attention.

Aim

To use near prospective methodology and time line follow back to identify when onset of acute appendicitis occurs to stratify risk of perforation or abscess formation (complex disease) in children presenting with lower abdominal pain.

Subjects and Methods

109 children (4-16 years of age) and their parents with acute onset of lower abdominal pain, with no background organic disease (IBD, liver or pancreatic disease, short bowel or previous abdominal surgery, oncology or immune deficient, TB, renal or rheumatological diagnosis) were interviewed by a research associate. When pain, feeling unwell and who was consulted for advice were documented. Time line follow back was used to confirm when these events occurred. Patients were then classified as a "case" if a diagnosis of acute appendicitis was made or a "control" if a diagnosis of non-appendicitis was made (Non specific abdominal pain, constipation, mesenteric adenitis, IBS). Cases were subdivided in to "complex" if operative or histological findings suggested perforation or abscess. "Simple" disease had confirmed acute inflammatory changes only.

REC committee approval was granted following formal submission

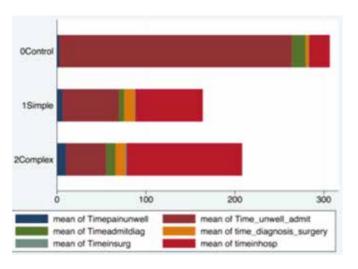
Results

57% of cases were male (62% in complex group) and 44% of controls. There was no significant difference in the age groups (10.6 vs 10.2 years). Figure 1 shows that from the onset of pain or from feeling unwell there is a shorter duration of time before attending hospital and getting a diagnosis of appendicitis in children with complex compared to simple appendicitis (34.5 v 57.4 hours respectively p=0.05). The time from onset of pain to hospital admission is significantly longer in the control group (274.4 (72.2) hours), but the duration of admission is significantly shorter than either the two appendicitis groups (p<0.05). Healthcare expenditure was higher in the complex appendicitis group (longer surgical time, LOS, IV fluids and antibiotics).

Summary and conclusion

Using near prospective methodology and timeline follow back, children with complex appendicitis have a more rapid presentation to hospital than those with simple appendicitis. We conclude that children with appendix perforation and abscess probably have a different disease from the outset.

Fig 1 Timing for stages in patient pathway to hospital and definitive treatment of appendicitis and non-appendicitis abdominal pain.



15.00 – 15.10 Dietary Manipulation of the Healthy Human and Colitic Murine Gut Microbiome by CD-TREAT diet and Exclusive Enteral Nutrition; a proof of concept study.

V. Svolos¹, R. Hansen², U.Z. Ijaz³, C. Quince⁴, D. Watson⁵, A. Alghamdi⁵, A. Brejnrod⁴, C. Ansalone⁶, S. Milling⁶, D. Gaya³, R. Russell², K. Gerasimidis¹ ¹Human Nutrition, School of Medicine, College of Medical, Veterinary and Life Sciences, University of Glasgow, Glasgow Royal Infirmary, Glasgow, United Kingdom; ²Department of Paediatric Gastroenterology, Hepatology and Nutrition, Royal Hospital for Children, Glasgow, United Kingdom; ³School of Engineering, University of Glasgow, Glasgow, United Kingdom, ⁴Warwick Medical School, University of Warwick, Warwick, United Kingdom, ⁵Strathclyde Institute of Pharmacy and Biomedical Sciences, University of Strathclyde, Glasgow, United Kingdom; ⁶Institute of Infection, Immunity and Inflammation, School of Medicine, College of Medical, Veterinary and Life Sciences, University of Glasgow, United Kingdom; ¹Department of Gastroenterology, Glasgow Royal Infirmary, Glasgow, United Kingdom

Backgroun

The extensive modulation of gut microbiome in children with Crohn's disease (CD) treated with exclusive enteral nutrition (EEN) offers clues about EEN's potential mode of action; but also on the development of novel therapies through dietary manipulation of the gut microbiota.

Aim

This proof of concept study compared the effect of a novel 'ordinary' food-based diet (CD-TREAT diet) and EEN on healthy human and colitic murine gut microbiota.

Subjects and Methods

A) Healthy adults followed two experimental diets for seven days with a 15 day wash out period in between; EEN and CD-TREAT, an "ordinary" food diet which has similar nutrient and food ingredient composition to EEN (e.g. fibre content, fatty acid composition, lactose and gluten free content). Participants were randomly allocated to start with EEN or CD-TREAT first. Fresh faecal samples were collected before and after each dietary intervention (4 different time points) and 16s rRNA sequencing, untargeted faecal and urine metabolomics (using LC-MS) were performed; B) In this study, 10-month-old HLA-B27 and HLA-B7 trangenic rats received EEN, CD-TREAT diet or regular rat chow for 4 weeks. Faeces were collected at baseline, 1, 2, 3 and 4 weeks post treatment initiation. Gut contents, ileal and colonic tissue were harvested at sacrifice. Disease activity was quantified by blinded histological scores and gut microbiota metabolic activity was measured by faecal short chain fatty acids (SCFA) quantification.

Results

A) 100 faecal and urine samples were collected from 25 healthy subjects. During EEN and CD-TREAT gut bacterial community structure (using Operational Taxonomy Units) significantly changed after both EEN (R-squared=0.14501, p-value=0.001) and CD-TREAT (R2=0.05016, p-value=0.003) and shifted towards the same direction. EEN's and CD-TREAT's impact on 3% OTU community structures was strongly correlated (Adjusted R-squared=0.3755, p-value<2.2e-16). Similarly, untargeted faecal metabolomics revealed a strong correlation between the changes during EEN and CD-TREAT (Pearson's R=0.31, p-value<10^-14).

B) 100 faecal samples were collected from 12 HLA-B27 and 8 HLA-B7 adult transgenic rats. Both dietary interventions increased the body weight of the HLA-B27 rats [Median(IQR)%weight change, EEN: +9.2(+6.5,+12.4) vs CD-TREAT: +15.7(+10.4,+17.7) vs Control: -2.1(-2.7,-0.3)] and decreased the weight of caecum and colon contents [Median(IQR) gut contents weight(g), EEN: 1(0.6,1.2), 0.1(0,0.26) vs CD-TREAT: 2.1(1.8,2.9), 0.5(0.3,0.7) vs Control: 3.5(3.1,5.2), 0.7(0.4,1.4)]. Faecal concentration of total SCFA, acetic, propionate decreased while iso-butyric and isocaproic increased during both dietary interventions [Δ Median μ mol/g, EEN: -324, -271, -44.8, +4.7, +2.2 vs CD-TREAT: -354, -292, -56.2, +3, +1.5]. Histopathology scores revealed that both dietary interventions benefited moderately ileal but not colonic inflammation.

Summary and conclusion

We have developed an "EEN composition alike" food based diet which induces similar effects on gut microbial composition and metabolites with EEN. This proof of concept study to support a subsequent pilot trial in people with active CD.

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15.10 – 15.20 Blended Diet for Enteral Feeding – Identifying current practice in a paediatric community setting and developing a clinical tool to improve patient care

Mrs Charlie Bigwood, Senior Paediatric Dietitian, Chailey Clinical Services, Beggars Wood Road, Lewes, East Sussex, BN8 4JN

Background

A growing number of parents and carers in the UK have adopted blended diet (BD) via feeding tube due to perceived health benefits. Current guidance advises against the use of BD. However due to its increasing parental use, it is essential to develop guidelines to facilitate safer practice, support informed choices and work in partnership with parents who want to give their children BD. The setting was a specialist residential school for children with complex neurodisability which has onsite clinical services.

Aims

- 1. Identify the prevalence of BD usage
- 2. Explore parents' views of perceived benefits
- 3. Develop a risk assessment and care planning tool for administering BD in a community setting

Subjects and Methods

Case notes of all patients on the dietetic caseload were reviewed and informal interviews were conducted during clinic sessions with parents, carers and key stakeholders to find out:

- How many families have already adopted BD?
- What are the families giving?
- How are families providing BD?
- Why have families started using BD?
- What are the associated risks for BD?

Risks were also identified by reviewing current literature. Discussions with multidisciplinary team members facilitated the development of the clinical tool.

Results

Use of BD among families:

Of a total of 54 patients receiving enteral nutrition, 15 (28%) were receiving BD. Eleven received BD in conjunction with commercial formula, ranging from BD providing the majority of nutritional requirements to supplementary fluids such as fruit tea or probiotic drinks. Four exclusively used BD. Various methods of administration were used but all used bolus feeding. BD consistency varied from single cream to thick custard.

Reasons parents gave for using BD:

- Give more 'naturally made' feed
- Normalizing meal times
- Feeling more able to nurture their child
- Regaining control
- Ability to provide bespoke nutrition and cater for specific requirements
- Supporting their child to develop food preparation skills
- Management of symptoms such as reflux, vomiting, diarrhoea, poor weight gain and oral food aversion

Risks of using BD:

- Tube occlusion
- Higher risk of microbial contamination
- Compromised nutritional intake due to unknown concentration of puree
- Poor volume tolerance
- Initial weight loss

A risk assessment and care planning tool was developed as a result of this work.

Summary and Conclusion

A significant proportion of children were receiving BD and parents reported a range of perceived physical and psychological benefits. There was a lack of clear guidance and support available to families and carers. In order to resolve this, a clinical tool was developed. This ensured each patient had a detailed individualised plan to enable nursing and non-clinical staff to safely deliver BD in this community setting.

A significant proportion of children were receiving BD and parents reported a range of perceived physical and psychological benefits. There was a lack of clear guidance and support available to families and carers. In order to resolve this, a clinical tool was developed. This ensured each patient had a detailed individualised plan to enable nursing and non-clinical staff to safely deliver BD in this community setting.

15.20 – 15.30 Scottish home parenteral nutrition longitudinal point prevalence data suggest a dramatic rise over the last 3 years.

Dr lain Chalmers¹, ST8 Paediatric Gastroenterology; Dr Paul Henderson^{1,2}, Consultant Paediatric Gastroenterologist; Mrs Christina Mcguckin³, Parenteral Nutrition Clinical Nurse Specialist; Miss Catherine Paxton¹, Parenteral and Enteral Nutrition Clinical Nurse Specialist; Dr Shyla Kishore⁴, Consultant Paediatric Gastroenterologist; Dr David Goudie⁵, Consultant Paediatrician; Dr David Mitchell¹, Consultant Paediatric Gastroenterologist; Dr Diana M Flynn³, Consultant Paediatric Gastroenterologist; Dr Andrew R Barclay³, Consultant Paediatric Gastroenterologist

Backgroun

Longitudinal data on home parenteral nutrition (HPN) are usually published by single centres of excellence with inherent recruitment bias. Previously published UK-wide point prevalence data suggest that the rise in HPN prevalence is accelerating (1). However these data have not been republished since 2012 and more recent surveys are incomplete.

Aim

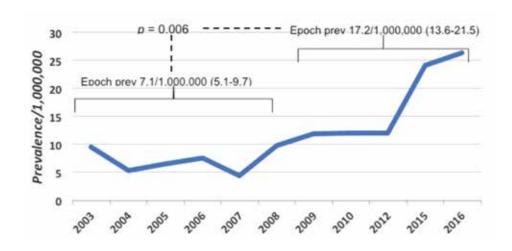
To present longitudinal, collaborative, Scottish HPN epidemiological data (representing 7.4% of the UK population <16yrs), which has been uniquely accrued since 2003.

Subjects and Methods

HPN prevalence and basic demographic data were prospectively collected annually from 2003-2010 by the Scottish HPN managed clinical network (SMCN), and subsequently through the SSPGHAN nutrition group in 2012, 2015 and 2016. Population data for Scotland from the National Records of Scotland provided population figures for children aged <16 years. Trend analysis was performed for the 2 epochs 2003-08 and 2009-16 using Poisson regression. Data were extrapolated to the entire UK population using publicly available data. Statistics were performed in Rv3.1.1.

Results

HPN point prevalence has increased from 9 cases in 2003 to 24 cases in 2016. Diagnoses in 2016 were: short bowel syndrome 13 (54%), neuromuscular disease 9 (38%) and enterocyte disorders 2 (8%). Scottish HPN point prevalence is currently 26.3/1,000,000 population at risk. Epoch analysis revealed a significant increase in point prevalence from 7.1/1,000,000 (95% CI 5.1-9.7) to 17.2/1,000,000 (95%CI 13.6-21.5) from 2003-2008 to 2009-2016 (p=0.006)(Figure 1). Extrapolating for the UK in 2016, data would suggest a total of 324 prevalent HPN cases.



Summary and conclusion

We present longitudinal, collaborative, Scottish HPN epidemiological data. Due to the unique close working practices of the SMCN and SSPGHAN nutrition group, we can be uniquely confident of complete case ascertainment. A secular trend in increasing neuromuscular disease was noted, this may reflect increased survival(2) or case recognition, but also the expansion of indications for HPN within neuro-disability. Given the insights provided by Scottish epidemiology, rejuvenation of UK-wide HPN epidemiology is a research priority. The sharing of such data is important for informing patients and the planning of services on a regional and national basis.

- 1. Barclay, et al Clin Nutr. 2015 Dec;34(6):1128-32
- 2. Barclay and Henderson. JPGN 2016 62(3):363-4

BSPGHAN 2017 Annual Meeting

POSTERS OF DISTINCTION

1 An investigation of azathioprine on autophagy pathway activity

Hooper K¹;Barlow PG¹; Henderson P^{2,3}; Stevens C¹

¹School of Applied Sciences, Edinburgh Napier University, Edinburgh; ²Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh; ³Child Life and Health, University of Edinburgh, Edinburgh

Introduction

Autophagy is an intracellular process that degrades damaged or aged proteins and organelles to maintain cellular homeostasis. Defective autophagy has been strongly linked to inflammatory bowel disease (IBD) pathogenesis, with evidence that enhancing autophagy may be therapeutically beneficial by regulating inflammation and clearing intestinal pathogens. Due to the high cost associated with the development of new drugs, a more comprehensive characterisation of commonly used IBD drugs and their mechanism of action are required.

Aims

To investigate the effect of azathioprine on autophagy pathway activity and to determine the molecular mechanisms involved.

Methods

The autophagy response to azathioprine was assessed in vitro using several complimentary methods. Live-cell confocal microscopy, flow cytometry and Western immunoblotting were used to assess autophagy in cells engineered to stably express the autophagy marker LC3 fused to GFP (GFP-LC3), or endogenous LC3 was assessed using specific antibodies. In addition cells were transiently transfected with dual GFP-RFP tagged LC3 to measure flux through the autophagy pathway. To determine whether mTORC1, a master regulator of autophagy activity, was affected by azathioprine the phosphorylation of S6 ribosomal protein (rpS6; a surrogate marker of mTORC1 activity), was monitored by Western immunoblotting and in-cell Western.

Results

A significant increase in autophagy was observed in response to 120µM of azathioprine, with optimal autophagy activity at 6 hours post-treatment. Confocal microscopy showed an increase in the percentage of cells exhibiting GFP-LC3 foci, and flow cytometry showed an increase in the fluorescent intensity of GFP-LC3 in cells treated with azathioprine compared to control cells. Western immunoblotting also showed that azathioprine treatment leads to an accumulation of LC3-II, the lipidated and active form of LC3. By monitoring cells transiently expressing the GFP-RFP-LC3 fusion protein we show that azathioprine stimulates autophagy pathway activity, and rules out the possibility that accumulation of LC3 positive autophagosomes is due to reduced fusion with lysosomes. Analysis of mTORC1 activity revealed that azathioprine treatment causes a decrease in phospho-rpS6, suggesting that azathioprine may stimulate autophagy via modulation of mTORC1 signalling.

Conclusions

We have used several complimentary methods to demonstrate that the immunomodulatory drug azathioprine strongly induces autophagy in vitro. Our results suggest that azathioprine may modulate autophagy via the mTORC1 signalling pathway. Work is now underway to further characterise the mechanism of action of azathioprine in the context of autophagy.

2 Prevalence of autoimmune diseases in a nationwide paediatric inflammatory bowel disease cohort

Dr VM Merrick¹, Dr P Henderson¹, Ms H Drummond², Dr J Van Limbergen³, Dr RK Russell4, Prof J Satsangi², Prof DC Wilson¹

¹Child Life and Health, University of Edinburgh, Edinburgh, UK; ²Gastrointestinal Unit, Centre for Genomic and Experimental Medicine, University of Edinburgh, Edinburgh, UK

³Paediatric Gastroenterology, Department of Paediatrics, Dalhousie University, Halifax, Canada; ⁴Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Glasgow, UK

Background

Autoimmune diseases (AIDs) affect up to 10% of individuals living in Europe and are a significant cause of morbidity. High rates of immune-mediated comorbidity and familial clustering suggest that genetic predisposition underlies AI disease susceptibility, yet few clinical studies have defined the prevalence rates of co-morbid AIDs in specific paediatric populations.

Aim

We aimed to document the occurrence of Juvenile Idiopathic Arthritis (JIA) and other AIDs in a Scotlandwide cohort of paediatric inflammatory bowel disease (PIBD; diagnosed <17 years of age) patients.

Subjects and Methods

The Paediatric-onset IBD Cohort and Treatment Study (PICTS) is a nationwide Scottish study of incident and prevalent PIBD patients, collecting a wide range of data, including rigorous phenotyping (Paris classification) with continuous long-term follow-up. The PICTS database was interrogated to identify patients enrolled up to 30/06/12 with a diagnosis of at least one associated AID by last follow-up; case notes were then reviewed with follow-up to 30/04/15. Cases believed to be related to use of anti-TNF2 treatment were excluded; atopic diseases were excluded due to their ubiquitous presence in the Scottish population.

Results

51 of 809 patients in the PICTS cohort had one or more associated AID; an overall co-morbid AID prevalence of 6.3%. 57% (29/51) were male; 59% (30/51) had Crohn's disease (CD), 37% (19/51) ulcerative colitis (UC), and 4% (2/51) IBD unclassified (IBDU). Median age (range) at PIBD diagnosis was 11.5 years (2.92-16.25).

Autoimmune liver disease was the most frequently occurring AID in 35% (18/51); psoriasis 24% (12/51); JIA 18% (9/51); spondyloarthropathy (SPA) 12% (6/51); coeliac disease 8% (4/51); type 1 diabetes 6% (3/51); thyroid disease 4% (2/51). 3 patients (6%) had multiple co-morbid AIDs; all psoriasis and joint disease with PIBD. Primary sclerosing cholangitis (PSC) was the predominant final liver diagnosis in 83% (15/18). Onset of PIBD preceded SPA in 100% (6/6) cases in contrast to JIA preceding PIBD in 89% (8/9) cases. There was a high prevalence of extensive disease on Paris classification; 83% (25/30) CD patients had extensive disease (ileo-colonic (L3) or greater) and 47% (14/30) had pan-enteric disease (L3+L4). 43% (13/30) had aggressive disease behaviour (B2 +/or B3); 33% stricturing (B2), 10% penetrating (B3). 71% (15/21) UC/IBDU patients had pan-colonic disease (E4); 19% (4/21) had at least one severe colitis episode (S1).

Summary and conclusion

6.3% of PIBD patients in this large cohort study have associated AIDs and the majority of these patients have an extensive IBD phenotype. Autoimmune liver disease (predominantly PSC) is the dominant comorbid AID, closely followed by joint disease (JIA and SPA) and psoriasis.

3 Rapid increase in pan-treatment refractory Crohn's disease after transition to adult services: A regional cohort study

Dr VM Merrick¹, Dr P Henderson^{1,2}, Ms P Rogers², Dr ID Arnott³, Prof J Satsangi³, Prof DC Wilson^{1,2}
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²Department of Paediatric Gastroenterology, Royal Hospital for Sick Children, Edinburgh, UK ³Department of Gastroenterology, Western General Hospital, Edinburgh, UK

Background

Inflammatory bowel disease (IBD) presents in childhood in up to 15% of cases. Paediatric onset IBD (PIBD) has a more extensive and dynamically changing phenotype and a faster rising incidence than adult-onset IBD.

Aim

We aimed to evaluate rates of treatment refractory disease at and then following transition to adult services in our regional cohort.

Subjects and Methods

A prospective PIBD database identified a cohort of all patients discharged from our regional service since 01/01/07. A retrospective study of patients graduating from paediatric to adult IBD services through a transition process, transition event (single joint clinic) or transfer until 31/12/13 was conducted with post transfer follow-up (FU) data at a minimum of 1 year to last adult FU (LAFU). Pantreatment exposure (PTE) was defined as exposure to all of azathioprine (AZA) or mercaptopurine (MP), methotrexate (MTX), infliximab (IFX) and adalimumab (ADA). Pan-treatment refractory (PTR) disease defined as those refractory (primary non-response [PNR], loss of response [LOR] or intolerance) to all of these therapies. We used the Montreal classification to describe disease location (L) and behaviour (B) phenotypes. Psychological co-morbidity was defined as a formal psychiatric diagnosis, regular psychiatry/psychology input (or intention for this if repeated family refusal), documented anxiety or depression and deliberate self-harm.

Results

138 patients graduated to adult services, 69% (95/138) had Crohn's disease (CD); 59% (56/95) male, 76% (72/95) with extensive disease (L3 or L3+L4) and 22% (21/95) B2 or B3 disease at time of transfer. Median (IQR) age at transfer 17.8 years (17.3, 18.4) and median (IQR) disease duration at transfer 5.4 years (4.6, 7.6). Median (IQR) length of FU post-transfer was 3.3 years (2.1, 5.1). 12% (11/95) had PTE with 4% (4/95) having PTR disease by time of transfer. PTE rates increased significantly to 26% (21/82) p=0.009 at LAFU and PTR disease to 18% (15/82) p=0.003; 13 patients lost to follow-up. 90% (19/21) of those with PTE had extensive disease and 48% (10/21) had B2 or B3 disease by LAFU. 80% (12/15) patients with PTR disease required bowel resection or a defunctioning stoma by LAFU, compared with 37% (30/82) of the whole CD cohort p=0.002. 24% (5/21) of those with PTE had significant psychological co-morbidity by LAFU.

Summary and conclusion

Our novel data show that pan-treatment exposure in paediatric-onset CD is already significant by time of transfer to adult services and continues to increase to affect 26% of this regional cohort within a relatively short period of adult follow-up. 18% of paediatric-onset CD patients have failed all medical treatments by LAFU and 71% with PTE require resectional or defunctioning surgery to manage disease.

4 A prospective cohort of patients receiving exclusive enteral nutrition (EEN) confirms high clinical response rates after 8 weeks of treatment: Initial results from the BIG study.

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Background

Exclusive enteral nutrition (EEN) is the first line treatment to induce remission in active luminal paediatric Crohn's disease (CD).

Aim

To prospectively characterise the efficacy of EEN in the West of Scotland during and after a course of EEN.

Subjects and Methods

Paediatric patients attending our hospital suspected of inflammatory bowel disease were recruited to our ongoing BIG (bacteria & inflammation in the gut) study between August 2014 to June 2016. Diagnosis was based on established clinical, endoscopic and histological criteria. Patients diagnosed with CD, who undertook a course of EEN, had data and biological samples taken before, during and on completion of an 8 week course of EEN. Disease activity was defined using the weighted paediatric Crohn's disease activity index score (wPCDAI). Anthropometry and systemic inflammatory markers of disease activity including faecal calprotectin, CRP, ESR, and haemoglobin were recorded at diagnosis and at end of EEN.

Results

41 patients (12 female, median age at diagnosis 12.3y (Q1- 10.0, Q3- 14.7) were identified. In 36/41 (88%) this was the first course of EEN, the remainder were undergoing a repeat course following a relapse. 34/41 (76%) presented with disease affecting both small intestine and colon (Montreal classification L3: n = 10, L3/L4: n = 18, L2/L4: n = 6). Seven children had isolated colonic disease (L2: n = 7). All participants were treated with polymeric feeds (Modulen: N = 40; Paediasure: N = 1); 31/41 (76%) had EEN orally, the remaining 10/41 (24%) required nasogastric tube. 10/41 (24%) were non-responders; 8/10 due to symptom escalation /poor response and 2/10 were unable to tolerate EEN.

At treatment initiation, median wPCDAI was 38.8 (range 10-85, Q1- 21.9, Q3, 57.5), 40/41 had a wPCDAI >12.5; 31/41 (76%) patients had entered clinical remission at the end of EEN (wPCDAI at EEN end <12.5). Before treatment initiation median inflammatory markers ESR, CRP, serum albumin and haemoglobin levels were 21 mm/h, 6 mg/L, 35 g/dL, and 10.3 respectively, with 63% of participants having a least one abnormal result. Three of these values had significantly improved by the end of EEN (ESR to median 7 mm/h, p = 0.012; CRP to median 6 mg/L, p = 0.004; albumin to median 38 g/dL, p = 0.0001).

All participants at time of treatment initiation had a raised calprotectin. Median faecal calprotectin concentration at treatment initiation was 1305 mg/kg and declined by the end of EEN to a median of 869.5 mg/kg (p = 0.006). There was a significant difference in calprotectin in those that respond to EEN (median 485.5) versus those who did not (median 1509.5) p = 0.019. In total, five participants had a faecal calprotectin below 250 mg/kg and, of these, four (10%) had dropped to below 100 mg/kg.

Summary and conclusion

8 weeks of EEN is associated with high rates of clinical remission in 76% of patients treated as assessed by the wPCDAI. This is paralleled by significant improvement in blood parameters and calprotectin but normalisation of calprotectin occurs in a minority. Consideration should be given to longer courses of EEN with the aim of driving calprotectin lower to see if this improves length of remission. The BIG study will now follow the clinical course of patients following EEN, specifically looking at patients who are and are not on nutritional supplements after their course of EEN whilst examining laboratory and clinical relapse and the contribution of the microbiome

5 Do self selected "Non Transitioned" referrals from paediatric services have lower treatment requires

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Background

Best practice guidelines stipulate children with long term health problems should have their care transitioned between paediatric services and adult health services. Our paediatric IBD patients are offered an appointment in a transition clinic, however non-attendance is high.

Aim

he aim of this study is to compare treatment requirement (as a surrogate marker of disease severity) and service engagement between patients choosing to attend transition clinic (transitioned) and those not (non-transitioned patients).

Subjects and Methods

All known IBD referrals from Birmingham Children's Hospital to University Hospital Birmingham aged 16-18 years from 2010-13 were collected. Baseline demographics, disease status and treatment history were collected from both adult and paediatric settings. Post referral procedures, changes in treatment and clinic attendance data were collected.

Results

57 patients were identified of which 33 were transitioned. Data regarding treatment prior to referral to adult services and changes post-referral are presented in table 1. Data is also presented for clinic attendance and follow-up length

		Non-transitioned patients	Transitioned patients	P value
Demographic	Number	24	33	-
	Male:Female ratio	13:11	14:19	0.385
	Ethnicity (cauc/black/ South Asian/ Unknown or mixed ethnicity)	14/1/1/8	14/1/7/11	-
	Crohn's Disease	17 (70.9%)	25 (75.8%)	-
	Ulcerative Colitis	7 (29.1%)	8 (24.2%)	0.679
Prior to referral	AZA/MTX	10 (41.7%)	22 (66.7%)	0.063
	Anti TNF use	4(16.7%)	8 (24.2%)	0.492
	IBD Surgery ^	5 (20.8%)	12 (36.4%)	0.210
Post-referral	New Anti-TNF use	5 (20.8%)	7 (21.2%)	0.963
	New course of steroids	6 (25%)	10 (30.3%)	0.927
	Surgical procedure	4(16.7%)	7 (21.2%)	0.670
	Endoscopic procedures	11 (45.8%)	15 (45.5%)	0.978
	Mean days follow-up per patient (months)	1361 (44.7)	1351 (44.4)	-
	Total attended Clinic appointments (per patient)	233 (9.7)	356 (10.8)	0.051
	Number of non-attendances (percentage of appointments)*	32 (13.7%)	16 (4.5%)	0.031

^{*}Hospital baseline 2015 DNA rate was 11.1% for new patients and 8.7% for IBD follow-up clinic patients overall. ^ Small bowel resection, stricturoplasty, and drainage of perianal abscesses. Summary and conclusion:

Fifty-eight percent of IBD patients referred from paediatric services chose to attend a transition clinic. Patients attending transition clinic are a self-selecting group in our cohort, as all are offered such a clinic appointment. Following referral both groups continue to have high therapy demands. A new course of steroids, starting Anti TNF therapy or surgical procedure was considered a surrogate for increased disease activity. Our data suggests that those attending a transition clinic are not less likely to flare, compared to those who did not attend. This is in contrast to other datasets which suggest that transition reduces disease flares. An assumption that patients choosing not to attend transition clinic have milder disease and need less intensive follow-up, is not supported by our data. www.nhs.uk/National-framework-for-continuing-care-england.pdf

6 The Caledonian Express; Novel hub and spoke model for 'regional' motility outreach services, experience, outcomes and service developments

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Background

Neuromuscular gastrointestinal disorders (NMD) provide a significant burden on regional GI services. This has been contributed to by increasing numbers of patients with Paediatric Intestinal Pseudo-Obstructive (PIPO) and colonic dysmotility (CD) disorders. The long-term survival of the most severely affected individuals and utility of an array of investigations, medical therapies and surgical interventions places a strain on the expertise of regional services. Equally the capacity for nationally funded specialist services to provide optimal ongoing medical and surgical support and follow up for the long-term is likely to be exceeded.

Ain

In 2014, due to an increased caseload, a formal shared care outreach model (SCO-GOS) was adopted between Scotland (SCO) and the Nationally funded motility service (GOS). We describe our experience in terms of referral, diagnosis, management, patient experience, cost saving and service development.

Subjects and Methods

Patients referred to GOS from 3 SCO regional services were prospectively characterised and an initial clinical review at GOS replaced by a combined SCO-GOS clinic. Investigation pathways were streamlined by completing any preceding prep/investigations in SCO to reduce the frequency of GOS visits and length of stay away from home. Tele-med multi-disciplinary clinics with both services and families, created a forum for maintaining communication and ongoing clinical care including complex surgical decisions. Reduction in patient travel, length of stay and costs were calculated from; post code, standard GOS motility investigation pathway; standard class rail travel (with 1 carer) and over-night bed days (OBD).

Result

24 patients had 19 new patient and 17 follow clinic visits in SCO-GOS

Patients	No (F)	End point surgery		
PIPO	7	3 high, 3 defunctioned ileostomies, 1 colostomy revision		
CD	6	3 defunctioned ileostomies, 1 left hemi-colectomy,		
Other	6	2 colostomy, 1 jejunal feeding		
Awaiting Dx	5			

Table: Demographics of 24 patients seen in SCO-GOS clinic 2014-2016

In all, from the 24 cases, 17 full thickness rectal biopsies, 9 full thickness serial GI biopsies, 7 colonic transit (radio-opaque marker) and 7 nuclear medicine gastric emptying studies were completed in SCO to inform specialist clinical assessment of cases. 8 patients travelled to GOS for further investigation. (5 seen directly in GOS but seen in SCO for review). These patients had a total of 37 specialist motility investigations cumulatively over 104 days. 7 patients had surgical procedures agreed by MDT forum and performed in SCO (4 defunctioned ileostomies, 1 subsequent colectomy, 1 extended left hemicolectomy/pull-through, 1 formation of left colostomy, 1 witzel jejunostomy). In the 2 years prior to SCO-GOS (2012-14) 2 patients completed surgery in GOS compared to none since (2014-16). Overall, establishing the SCO-GOS with increased local/regional delivery of investigations and surgical input has amounted, cumulatively, to reductions of 28,616 miles travel and 22 OBD, saving £5964 and £15,525 respectively. Improved local expertise has contributed to approval to extend and establish SCO GI motility investigations, including extended scintigraphy, oesophageal, antro-duodenal and ano-rectal manometry.

7 Therapeutic drug monitoring in Paediatric inflammatory bowel disease on maintenance Infliximab and Adalimumab treatment improves clinical remission with a proactive approach

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Aim and Background

The anti-TNF antibody Infliximab (IFX) and Adalimumab (Ada) are frequently used as maintenance therapy in Paediatric inflammatory bowel disease (pIBD). However, the role and frequency of monitoring trough levels and anti-drug antibodies (ADA) during maintenance treatment remains unclear in children, with two regimens being considered, with an either reactive or a proactive approach.

The aim of the present study was to investigate the trough levels of IFX and Ada, the presence of ADA and to identify correlation with inflammatory activity and clinical response.

Subjects and Methods

We conducted a retrospective study of all Anti TNF treated children with IBD (n = 67, Crohn's disease [CD] = 47, ulcerative colitis [UC] = 11, inflammatory disease unclassified [IBDU] = 7 and Early onset IBD [EOIBD = 2]; Male n=43, age range 4 years 3 months-17y; median 13y8m). Biologic monitoring at our institution was started in 2013 with ELISA assays. Demographics, CRP, ESR, albumin, activity indices PUCAI and PCDAI were recorded. Ada was started after Infliximab was discontinued for various reasons. All patients were on concomitant immunosuppressive treatment. 42 patients were on IFX only, 25 on Ada and 6 on Vedolizumab. 8 excluded because they had insufficient data. Children were on maintenance TNF treatment and had received treatment for 3-66 months with a median of 18 months.

Results

Group 1 Infliximab converted to Adalimumab; n= 25 patients, n=7 excluded as no data available (pre through level era availability); CD n=15, UC n=3, IBDU n=5, EOIBD n= 2. The lowest Ada trough levels in n=15 showed a median of 5.6, range 0.3-17, the highest a median of 9.1, range 3.7-12.7. ADA for Ada was negative in 16 patients, n=5 became positive over time, n=2 were positive at first measurement.

Group 2 Infliximab only; n=42; the lowest IFX through levels had a median of 1.4, range <0.8-32.5, with highest through levels median 5.2, range 0-45. ADA for IFX were negative in n=37, n=7 developed antibodies over time, median ADA of 61, range 10->200. 50 %(21/42) of patients with either low through levels and/or positive ADA received double doses to salvage treatment. Although there was clinical improvement, this did not correlate with a reduction of ADA. However in 81 %(17/21) of patients, double dosing led to an incremented of through levels above >2, median 4.1, range 2.4-21.9.

Although only 15/67 (22%) out of 67 patients had completely normal laboratory tests, 42/47 (89%) CD patients had normal PCDAIs, 10/11 (91%) UC patients had normal PUCAIs. 14/47 (30%) CD patients developed antibodies to IFX, 2/11 (18%) UC patients developed antibodies to IFX.

Summary and conclusion

The vast majority of our patients on either Adalimumab or Infliximab had an excellent clinical response to their treatment with our proactive approach regarding therapeutic drug monitoring, thus enabling us to optimize their treatment and bring them into clinical remission. We advocate therefore proactive biologic drug monitoring.

8 A prospective audit of biosimilar (Remsima) use in paediatric IBD: No change in clinical effectiveness but an opportunity for significant cost savings!

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Background

Biosimilars were licensed for use in Paediatric Inflammatory Bowel Disease in early 2015. A safety and efficacy audit of biosimilar Infliximab (Remsima) was carried out in our Tertiary Paediatric Gastroenterology unit in Glasgow August 2015 - June 2016.

Aim

To review the safety and efficacy when using the biosimilar Remsima

Subjects and Methods

Prospective clinical data was collected from laboratory reports, electronic patient records and case notes of patients starting a biologic for the first time. All of the statistics used in the results were calculated through Microsoft Excel. The weighted Paediatric Crohn's Disease Activity Index (wPCDAI) and Paediatric Ulcerative Colitis Activity Index (PUCAI) were used to document disease activity at initiation and follow up. For analysis, calprotectin samples and C.Reactive Protein (CRP) values were limited to maximum and minimum assay values respectively. Costings for biosimilar and originator Infliximab were obtained from National Procurement Scotland.

Results

There were 28 consecutive (N=28) patients (54 % (15/28 Male) commenced on Remsima equating to 110 infusions in total. 21 patients had a diagnosis of Crohn's disease and 7 UC/IBDU. The median age (IQR) at diagnosis was 12 (10, 14) years and 13.5 (12/16) years at biologic initiation. The primary reasons for treatment in CD were: Active Luminal 82% (17/21), Perianal – 9% (2/21) and other – 9 % (2/21). For IBDU/UC: Chronic Refractory Disease – 57% (4/7) and Acute Severe Colitis – 43% (3/7). All 28 (n/N) patients were on immunomodulator therapy and all were given pre-dosing steroids. At initiation 43 % (12/28) were on oral Prednisolone as acute therapy. Remsima was associated with a significant improvement in Crohn's disease post induction (see table).

Clinical Data	At initiation	At 12 week review	Comparative p value	
ESR - Median (IQR)	14(7.5,28.3) (N=28)	5(2,10) (N=15)	P=0.004	
CRP- Median (IQR)	7 (2.5, 24.6) (N=28)	1(1,3.8) (N=16)	P=0.01	
Albumin - Median (IQR)	33(28.8, 38) (N=28)	39(36.8,42) (N=16)	P= 0.004	
Calprotectin- Median (IQR)	1000(60, 1800) (N=23)	153(60,1000) (N=13)	P=0.03	
wPCDAI- Median (IQR)	28 (3.8, 51.3) (N=21)	0(0,8.8) (N=14)	P = 0.005	
PUCAI- Median (IQR)	45(27.5,52.5) (N=7)	49(33.1,64.4) (N=2)	P = ns	
Disease Classification (Crohn's) % (n/N)	Remission 38% (8/21) Mild 29% (6/21) Moderate 24% (5/21) Severe 9% (2/21)	Remission 86% (12/14) Mild 14% (2/14) Moderate 0% (0/14) Severe 0% (0/14)	P=0.005 P=0.32 P<0.05 P=0.23	

1 patient had an infusion reaction with a per infusion rate of 1/110 (1%) and per patient 1/28 (4%). Immediately after the 2nd Remsima infusion began, the patients face became flushed and throat felt tight. The infusion was then discontinued and therapy was changed. Infliximab levels were recorded as 25.9 mg/L with positive antibodies.

The average cost saving per vial during this period was £130 compared to originator Infliximab, so we estimate a total cost saving of around £40,000 during the 10 month audit.

Summary and conclusion

In summary, we have found that Remsima is as safe and effective as the originator, using our own local and comparative national audit data. There are associated significant cost savings. This baseline data is now enabling us to switch patients from originator to biosimilar Infliximab, adopting the same system of prospective audit to monitor results.

9 Surgical management of paediatric inflammatory bowel disease: a regional cohort study

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Background

The incidence of paediatric inflammatory bowel disease (PIBD) is increasing in Scotland and worldwide. With our own data demonstrating that a significant proportion of patients have extensive disease at time of transfer to adult services, progression to surgical management in childhood is often inevitable. However, at present population-based data regarding the surgical management of PIBD is lacking.

Aim

To investigate the prevalence and characteristics of surgery for patients with PIBD in a regional tertiary centre.

Subjects and Methods

An existing prospective database was used to identify incident and prevalent PIBD patients cared for in a regional treatment centre between 01.08.1997 and 31.12.2014. An electronic surgical audit database was utilised to extract detailed information on PIBD-related surgeries. Results are presented as median (interquartile range).

Results

394 patients with PIBD were identified. Age at diagnosis was 11.8yrs (9.4-13.5) and age at last follow-up was 17.8yrs (17.3-18.3). 65 (18%) patients had PIBD-related surgery while in paediatric services; 61% were male, 80% had Crohn's disease (CD), 17% had ulcerative colitis (UC), and 3% had IBD unclassified (IBDU). Prevalence of surgery in CD, UC and IBDU was 21%, 12% and 4% respectively. 44 (68%) patients had intestinal procedures and 30 (46%) had perianal procedures; 9 (14%) had both. Age at diagnosis in surgical CD patients was 10.9yrs (8.9-12.4) and for surgical UC patients was 10.5yrs (9.5-12.1). CD patients who underwent perianal procedures were younger at first surgery (12.5yrs [9.7-14.2] vs 14.8yrs [12.9-15.8]) than those requiring intestinal procedures, despite being a similar age at diagnosis. 5-year cumulative risk for intestinal surgery in CD patients was 9.6%, for perianal surgery was 9.2%, and for all surgery was 17.2%. 5-year cumulative colectomy risk in UC was 10%. The most common procedures for UC was total colectomy (n=11), and for CD were drainage of perianal sepsis (n=17) and right hemicolectomy (n=15). There were 2 IBDU surgical patients, one total colectomy and one drainage of perianal sepsis.

Summary and conclusion

This population-based, regional study demonstrates that almost one-fifth of PIBD patients undergo PIBD-related surgery, with the highest incidence of surgery in CD. This cohort also demonstrated a high proportion of perianal procedures as compared to previous UK-based data. Colectomy rate in UC was comparable to data from other centres.

10 Anaemia In Children Receiving Home Parenteral Nutrition

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Background

Children receiving long term Home Parenteral Nutrition (HPN) are at risk of developing iron-deficiency anaemia. Causative factors are inadequate absorption of enteral iron due to underlying gastrointestinal pathology, recurrent venopuncture and limited ability to add iron to the PN bag.

Aim

The aim of this study was to determine the incidence of iron deficiency anaemia in paediatric HPN patients, to establish what treatment symptomatic children received and determine the efficacy of each therapeutic approach to make a recommendation about future treatment strategies.

Subjects and Methods

Data was collected retrospectively from children receiving HPN at a large tertiary referral centre over a 12 months period (December 2014 - December 2015) and 41 patients were identified. Blood test including full blood count (FBC), Ferritin and C reactive protein was done at least 3 times for each patient in the follow-up period. The liver tests were checked at the beginning and at the end of the study. The number of blood transfusions and iron infusions was registered.

Results

41 HPN patients (61% females) were identified during the studied interval. The median age at the beginning of the study was 7 years 8 months and the median duration of home PN at the beginning of the study was 2 years 7 months.

The indications for home PN were an underlying motility disorder in 51% (21/41) of the cases, enteropathy in 29% (12/41) of the cases and short bowel syndrome in 20% (8/41) of the cases. 73% of the patients received oral/enteral feeds but none received oral iron supplements. The average amount of iron added to the PN bag was $0.45 \, \mu \text{mol/kg}$.

At the beginning of the study, 5% had severe anaemia, 54% moderate anaemia, 24% mild anaemia, and 17% had a normal haemoglobin (Hb) as per World Health Organization recommendations. On repeat testing, 10% had severe anaemia, 51 % had moderate anaemia, 20% had mild anaemia and 19% had normal Hb. At the end of the study, 5% of the patients had severe anaemia, 51% had moderate anaemia, 17% had mild anaemia and 27% had normal Hb. Iron deficiency anaemia (microcytic, low ferritin and iron) was most commonly seen.

Information about blood transfusions/iron infusions was available in 40 of the cases – 46% of them received blood transfusions and 29% iron infusions.

The patients who received oral/enteral feeds or had more than 0.5 µmol/kg of iron added to the PN didn't had a significant lower grade of anaemia at the end of the study. Children who received iron infusions didn't have significantly higher rate of improved haemoglobin compared with the ones who received blood transfusions (50% vs. 33%, p=0.657) and didn't have significantly higher difference in haemoglobin (g/L) compared with blood transfusions after the treatment (0.5 [-11.5 to 8] vs. -6 [-17 to 8], p=0.506)

From the group of children who had transfusion/infusion, the ones who had transfusions had significantly higher rate of abnormal liver function compared with the one who had iron infusions (93% vs. 37%, p=0.09).

Summary and conclusion

Iron deficiency anaemia, most commonly in the moderate severity range, is common in children receiving home PN. Many patients still receive blood transfusions if symptomatic. Intravenous iron may be a good alternative to prevent anaemia. Treatment guidelines are required to advice when and how much iron should be prescribed.

11 Anti-IL10 autoantibodies as a novel cause of very early onset inflammatory bowel disease (VEO IBD) with the resolution of bowel inflammation on B-cell-directed immunomodulatory therapy.

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Background

Anti-cytokine autoantibodies are increasingly recognised in disease pathogenesis. Homozygous loss of function mutations in interleukin-10 (IL10) and interleukin-10 receptors (IL10R) cause severe infantile (very early onset (VEO)) inflammatory bowel disease (IBD).

Air

To present a novel cause of VEO IBD and its targeted immunological therapy.

Subjects and Methods:

A female Caucasian infant, initially well, developed bloody diarrhoea and vomiting from age 3 months. Dietary restrictions initially induced mild improvement but sudden deterioration at 17 months required parenteral nutrition (PN) from aged 18 months. Referred to national service for evaluation at 19 months. Cytokine profiling identified high serum IL10. IL10R defects were excluded by functional testing. Anti-cytokine profiling revealed high titres of neutralising anti-IL10 autoantibody.

Clinical course, endoscopic findings and serial serum IL10 & IL10 autoantibodies:

Age /12	Clinical Data	Clinically	Endoscopic findings	IL-10 ▲ (pg/mL)	Anti IL10 auto- antibodies
20		PN dependant with vomiting, pain and bloody diarrhoea.	Gastritis & severe colitis with deep rolled edged ulcers	P=0.004	+ve 1/312500
22	1,2,3	PN dependant with vomiting, pain and bloody diarrhoea.		P=0.01	+ve 1/312500
23	1,2,3	PN dependant with vomiting, pain and bloody diarrhoea.		P= 0.004	+ve 1/162500
26	1,2,4,5*	Improving vomiting, non- bloody diarrhoea, improving pain. PN stopped.	Gastritis & colitis with improvement in size and depth of ulcers	P=0.03	+ve 1/312500
27	1,2,4,5	Stable. Off PN. No vomiting. Ongoing diarrhoea & abdominal pain	Gastritis & colitis with improvement in size and depth of ulcers	P = 0.005	
29	1,2,5	Well. No pain. One formed stool per day.		P = ns	-ve 1/100
32	1,2,5	Well. No pain. One formed stool per day.	Normal stomach and colon except areas of mild colonic scarring.	P=0.005 P=0.32 P<0.05 P=0.23	

Therapy: 1) IVIG 2g reducing to 0.5g/kg 2 weekly; 2) iv Rituximab 3 monthly; 3) iv methylprednisolone (MP) 2mg/kg/day then weaning; 4) oral prednisolone 1mg/kg/day then weaning; 5) Infliximab 10mg/kg 4-weekly, weaning to 5mg/kg 8-weekly. *Infliximab added due to clinical relapse on steroid taper.
A Normal Range 0-1 pg/mL.

Summary

The early onset severe colonic inflammation resembled that seen in IL10 pathway defects. We hypothesised functional deficiency of IL10 due to high titre of neutralising anti-IL10 antibodies. To target autoantibody production, we combined anti-B cell therapy with more conventional, gutdirected immunosuppression. The latter enabled rapid weaning from PN, but ultimate resolution of clinical symptoms and gastrointestinal pathology was not obtained until confirmed absence of anti-IL10 autoantibodies some 7 months after starting Rituximab. Infliximab is to be stopped and we plan to maintain the child on Rituximab and replacement scIG in the medium term.

Conclusion

Identification of an anti-cytokine antibody to IL10 as a novel pathogenic mechanism for VEO IBD has allowed a targeted immunological therapy with avoidance of long term immunosuppression and resolution, rather than control, of the inflammatory process.

12 Availability of laboratory investigations for Paediatric Inflammatory bowel disease; findings of a nationwide survey.

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Background

The early diagnosis, classification and appropriate management of paediatric inflammatory bowel disease (PIBD) continue to pose challenges to paediatricians and paediatric gastroenterologists. Laboratory (lab) investigations play an important role in guiding clinicians and prompt access to these is crucial in reducing the morbidity and complications associated with this chronic disorder. Labs in the NHS mostly invest in the resources for investigations independently at trust level. There are no national standards directing required investigations which leads to variability in their availability and hence in clinical practice. Paediatric gastroenterologists and Paediatricians often find themselves limited by the availability of these investigations and hence are forced to make less informed decisions whilst managing this condition.

Aim

To investigate the availability of IBD related laboratory investigations in NHS laboratories in England and to discern whether there are regional variations.

Subjects and Methods

A structured telephone survey was conducted in July 2016 by a single interviewer by contacting the clinical Labs in Acute NHS trusts across England with paediatric services. The available online handbooks for each lab were also accessed and where appropriate scientists were unavailable the survey questions were sent by email. The data was collected on a database and analysed using Microsoft excel. No ethical approval was required for this study.

Results

A response was obtained from 136 out of 139 laboratories (97.8%).

- Inflammatory markers (other than CRP): ESR is widely available at 98%, followed by Plasma viscosity (PV) at 71% and Orosomucoid (ORM) at 48%. Regional variations are significant with East of England and London having least access to PV and ORM.
- Faecal calprotectin was available in 89% of labs although only 51% offer in house testing. 84% allow any clinician to request the test whereas the rest allow only a few clinician groups to request.
- ANCA can be tested in 94% of labs but ASCA is available only in 29%.
- TPMT activity was available in 96% of labs with only 29% testing this on site.
- 6-Thioguanine metabolites was offered only by 58% of labs with 89% outsourcing it. This was most
 widely accessible in the south east.
- Infliximab serology is offered in only 61% of labs with only 14% able to test this on site. This is least
 accessible in the East Midlands.

Summary and conclusion

There is extensive regional heterogeneity in the availability of laboratory investigations for PIBD in England. There is also a significantly low level of on-site testing for a number of investigations which is likely to significantly add to the time lag in obtaining results. More research is needed to confirm the utility of the laboratory investigations in PIBD and establish their use. National guidelines should include standards for the investigations required and provide information on cost effectiveness to allow at least the regional units of each region to access the tests promptly.

13 Evaluation of Infliximab monitoring in paediatric IBD patients in a regional Paediatric Gastroenterology Unit.

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Background

Infliximab is a chimeric monoclonal antibody to tumour necrosis factor widely used in the treatment of Crohn's disease and severe Ulcerative Colitis. However, despite good initial response to treatment, reduced disease control and loss of response is common. Loss of response to infliximab may be multifactorial including; insufficient infliximab dosage, lengthy dosing interval or development of antibodies to the murine portion of the antibody. Evidence supports the use of infliximab drug and antibody monitoring in adults with IBD however; evidence to support its use in paediatric IBD is at present lacking.

Air

To evaluate the use of infliximab and anti-infliximab antibody levels in assessing suboptimal response to treatment and guiding future therapeutic decision making.

Subjects and Methods

A retrospective review of 82 consecutive infliximab levels sampled from 41 patients with active Crohn's disease (n=36), ulcerative colitis (n=5) and indeterminate colitis (n=1). Data was collected from samples taken between Oct 2014 and December 2016 from patients aged between 8-17 yrs (median 13.5) under the care of the Paediatric Gastroenterology team. Indications for infliximab levels and anti-infliximab antibody monitoring included; routine monitoring, loss of response, poor response and drug-related side effects. Incomplete data sets were excluded. Induction regime was 5mg/kg at 0, 2 and 4 weeks and routine dosing interval was 8 weeks, unless otherwise indicated. Drug levels were measured using the ELISA-based Theradiag© LISA-Tracker kits (Theradiag, France).

Results

Median serum infliximab levels were 2.3 (0.3-8.8) ug/mL.

Thirty four infliximab samples were below the lab reference therapeutic level (2.0ug/mL). Consequently 14 patients had a dose increase (to 10mg/kg), 7 terminated infliximab (commenced adalimumab), 5 reduced dose interval and in 8 no change was made.

A total of 8 patients had detectable anti-Infliximab levels (>10ng/mL), these were only seen in patients with sub-therapeutic or undetectable infliximab levels.

Forty seven infliximab levels were therapeutic. As a result, 32 had no changes made, 14 patients had changes in management and 1 stopped infliximab.

Summary and conclusion

Therapeutic monitoring of infliximab levels and measurement of antibody levels is useful aid to support the management of patients with Crohn's and Ulcerative Colitis who may be losing clinical response. Our data shows that almost half of all patients tested had sub-therapeutic levels, which allowed for dose escalation and treatment optimisation. Anti-infliximab antibody levels were similarly useful, with the majority of affected patients progressing to alternative biologics.

Whilst the cost of infliximab assays is not insignificant, integration of therapeutic monitoring for paediatric patients with IBD may help to rapidly and confidently identify the cause for non-response and support more informed treatment choices to be made.

14 Setting a benchmark for quality standards: The incidence of central venous catheter infections in children on home parenteral nutrition

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Background

The British Society of Paediatric Gastroenterology, Hepatology and Nutrition (BSPGHAN) Quality Standards working group and the British Intestinal Failure Alliance (BIFA) have this year proposed standards for patients on home parenteral nutrition (HPN). They advise regular audit of patient outcomes, including rates of inpatient catheter related sepsis below 3/1000 catheter days and outpatient rates below 1/1000 catheter days in adults. The expected incidence in children is not yet known.

Ain

This study reviews the incidence of central venous catheter (CVC) related infections and venous thromboembolisms (VTE) in patients under the care of Birmingham Children's Hospital (BCH) over the past 16 years, and suggests potential interventions to improve outcomes for patients

Subjects and Methods

Data were collected retrospectively using medical notes, microbiology and results, and HPN patient database kept by the nutrition team at BCH from January 2000 to December 2015. The number of CVC infections were collected in 2 yearly intervals and used to calculate the rate of infection per 1000 catheter days using the formula; (Number of catheter-related bloodstream infections/Number of central line days) X 1000. Catheter related sepsis was defined as a febrile episode diagnosed clinically as CVC infection by the admitting consultant and treated with a complete course of intravenous antibiotics, with or without growth on blood culture. As a secondary measure, we looked at the rate of CVC related VTE diagnosed clinically or detected on screening ultrasound of central veins.

Results

CVC related sepsis has declined remarkably over the time period, from 10 infections/1000 catheter days in 2000/01 to 0.8 infections/1000 catheter days in 2014/15. The only episode of VTE occurred in 2001 and there have been nil since.

Summary and conclusion

Rates of infection have reduced as a result of better education for patients and families and the use of agents such as chlorhexidine wash, although Taurolock is not routinely used. BCH have a dedicated nutrition nursing team who reinforce practical measures to keep line sites clean, such as proactively screening for skin colonization and eradication with topical treatments. Line site infections are swabbed and treated. The team has a preference for prompt treatment of suspected infection in the emergency department and removal of lines if the infection does not respond to antibiotics within 24-36 hours. The low levels of VTE are likely due to this practice, as well as improvements in clinical technique at insertion, particularly since a dedicated venous access team was set up, who use ultrasound guiding insertion techniques

15 Shifting the Shunters in a Paediatric IBD Population: Thiopurine Dose Splitting versus Allopurinol and Thiopurine Co-Therapy

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Aim and Background

The use of 6-Thioguanine nucleotide (6-TGN) levels as a method of adjusting Thiopurine dosing (both for Azathioprine and 6-Mercaptopurine), thus optimizing therapeutic effects, have dramatically improved the safety of their use in Paediatric Inflammatory Bowel Disease (pIBD). The aim of this study was to evaluate the therapeutic outcomes of pIBD patients treated with either Thiopurine dose splitting (Group 1) or Allopurinol and Thiopurine Co-Therapy (Group 2), (Thiopurine dose reduced to 25% of 2mg/kg, Allopurinol 50mg <30kg weight, 100mg >30kg) for either abnormal level outside the therapeutic range of 235 to 450 pmol/8x10E8 RBC and/or abnormal 6-TGN/MeMP ratios (>11). Both are effective treatment options and although data is available in adult IBD on either regimen, there is paucity of data in pIBD patients.

Subjects and Methods

136 patients (Male n=81, age range 4years 10months – 16y 8m, median 13 years) on Thiopurines with recorded metabolites were retrospectively identified over a 26 month period our IBD database.

101 (74 %) of patients had levels within the therapeutic range with normal ratios. The two regimens above were implemented on those with abnormal result, n=35 (26%).

Results

In Group 1, n=22 patients were identified; the pre-intervention 6-TGN levels had a median of 199, range 75-521; post-intervention 245, range 123-577. The pre-intervention ratio had a median of 14.5, range 2-32; post-intervention 5, range 0-18. 18 patients had a ratio of >11, in n=17 (77%) the ratio median drop was 11, range 4-31, the biggest drops were with pre-intervention ratios of >18, with 19/22 (86%) patients returning to ratios <11. The pre-intervention MeMP levels had a median of 3179, range 219-5902, post intervention 1496, range 143-3805.

In Group 2, n=13 patients were identified; the pre-intervention 6-TGN levels had a median of 186, range 75-387; post-intervention 309, range 156-578. The pre-intervention ratio had a median of 15, range 8-34; post-intervention 1, range 0-6; 12/13 (92%) patients had a ratio of >11, in those the ratio median drop was 14, range 6-34 with 11/12 having median ratio of 1, range 0-2; The pre-intervention MeMP levels had a median of 2539, range 648-6333, post intervention 246, range 116-2306. There was a statistically significant difference regarding 6-TGN levels in the slit dose versus Co-therapy (0.04) and in the drop in ratio (0.013) favoring the Co-therapy treatment. There was no statistically significant difference in the MeMP levels (0.073)

Summary and conclusion

Although both groups lowered the abnormal levels/ratios, Co-therapy treatment was superior to split dose regimens in our patient cohort. Low-dose Thiopurines and Allopurinol co-therapy is a safe and effective treatment option in pIBD, however needs close monitoring to avoid myelotoxicity. Larger patient numbers are needed to confirm our data.

16 Carnitine deficiency in long term exclusively parenteral nutrition (PN) dependent children

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Background

Serum Carnitine estimation is not part of routine monitoring for patients on long term parenteral nutrition therapy. Carnitine deficiency (both primary and secondary) can present with inability to maintain blood glucose during the fasting state. This can be an important cause of hypoglycaemia in these patients during previously tolerated fasting periods.

Case Report

Two and a half year old boy with short bowel syndrome secondary to gastroschisis was admitted with mechanical break in the central venous catheter. He was exclusively dependent on PN for 16 hours a day and 7 nights a week. His enteral intake had been minimal due to various reasons in spite of adequate measures to encourage oral and gastrostomy feeds. He developed symptomatic hypoglycaemia on the Children's assessment unit whilst waiting for line repair not exceeding his usual fasting limit. His blood sugar was found to be 0.8 mmol/L and this was treated with dextrose infusion.

Hypoglycaemia screen done during this episode was normal except for low serum Carnitine of 3 umol/L (normal level 15-53 umol/L), suggesting diagnosis of Carnitine deficiency. Further paired serum Carnitine and urine Carnitine ruled out primary Carnitine deficiency with low serum Carnitine of 5 umol/L and urine free Carnitine of 2 umol/L. His serum Lysine (precursor of Carnitine) level was checked and was also found to be low at 75umol/L (normal level 101-246 umol/L). This was thought to be consistent with secondary (nutritional) Carnitine deficiency. He was started on enteral carnitine supplementation via gastrostomy with normal Carnitine level (49 umol/L) restored within two weeks.

Discussion and Conclusion

Carnitine plays a key role in the beta oxidation of fatty acids and its deficiency can lead to poor fasting tolerance. Endogenous Carnitine synthesis depends on its precursor lysine and is insufficient in children on minimal enteral nutrition. Most solutions for parenteral nutrition do not contain Carnitine. This makes exclusively PN dependent patients prone to develop nutritional (secondary) Carnitine deficiency especially if they have minimal enteral feeding tolerance.

Enteral supplementation of Carnitine normalised Carnitine level in our patient within two weeks and also improved fasting tolerance when prospectively monitored. He tolerated Carnitine supplementation well and is currently monitored for serum Carnitine levels regularly.

We are currently screening our cohort of intestinal failure patients on PN for serum Carnitine levels and trying to co-relate this with their enteral intake. Low Caritine level in these patients may suggest routine screening for serum Carnitine in these complex patients to actively look for development of nutritional Carnitine deficiency. This finding is previously been reported in literature in the context of neonates with PN dependence and suggested routine supplementation of Carnitine on PN therapy of more than 2 weeks.

17 Fermentation capacity of gut microbiota in patients with inflammatory bowel disease compared to healthy controls

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Background

Gut microbiota in the colon ferment undigested dietary fibre to produce short-chain fatty acids (SCFA). SCFA have beneficial effects on colonic health. Differences in microbiota composition and metabolic activity have been described between IBD patients and healthy controls.

Air

This study explored the capacity of the gut microbiota of IBD patients to breakdown dietary fibre.

Subjects and Methods

Fresh faecal samples were collected from IBD patients in clinical remission and healthy controls (HC). In vitro batch culture fermentations were carried out for 5 carbohydrate/fibres and for a mixture of these 5 fibres together (hi maize, pectin, raftilose, wheat bran, cellulose). Aliquots were taken at 0 and after 48 hours of fermentation. Faecal SCFA (butyrate, propionate and acetate) concentration (umol/g) and their proportional ratio (%) were measured with Gas Chromatography.

Results

39 IBD participants and 19 matched HC were recruited. Following 48h batch cultures, total SCFA from hi maize and raftilose in CD patients (median (IQR) HC; 51.76 (22.02) vs. CD; 41.12 (23.28) vs. UC; 41.94 (14.72) p=0.02) and from hi maize in UC patients were significantly lower than in heathy controls (median (IQR) HC; 58.87 (17.69) vs. UC; 44.92 (21.49) p=0.008). The proportional contribution of butyrate to total SCFA following fermentation with mixed fibre was also significantly higher for healthy controls compared to UC patients (median (IQR) HC; 10.76 (8.26) vs. CD; 8.75 (4.74) p=0.044). In no cases were there any significant differences between the SCFA concentration or relative contribution in CD or UC patients.

Summary and conclusion

These data suggest that the microbiota of IBD patients has a lower capacity to break down fibre, compared to healthy people. The findings of this work should be complemented with changes in microbiota composition using next generation sequencing.

18 Management of Central Line Infections in Paediatric Home Parenteral Nutrition Patients: Audit in a Tertiary Care Centre

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Background

Parenteral nutrition (PN), given through a central line, risks catheter-related sepsis. There are no national standards for the management of suspected central line infections in paediatric home PN patients. The Newcastle upon Tyne Hospitals Trust Standards advise broad-spectrum IV antibiotics within 2 hours of admission.

2013 audit showed that a significant proportion of suspected infections (9/33, 29%) presenting to A&E were confirmed by blood culture, and that mean time to IV antibiotics was 3.5 hours.

Changes since 2013 were the introduction of the central line safety letter ("golden ticket") to be shown at A&E, reformation of presentation to A&E only rather than either A&E or the Great North Children's Hospital (GNCH), and 24/7 phone triage by PN nurse specialists.

Aim

To assess the frequency and antibiotic management of suspected and proven central line infections presenting to Royal Victoria Infirmary (RVI) paediatric A&E against trust standards.

Subjects and Methods

Retrospective audit over the period 1/1/2015-31/12/2015. Inclusion criteria: all patients of the GNCH on home PN presenting to RVI paediatric A&E with suspected catheter-associated sepsis.

Data collected from PN nurses' database, electronic records systems (E-Records, Mermaid, Diadem) and paper notes. Suspected infections were confirmed by positive blood culture. Management was compared to trust standards, with an aim of time to antibiotics under 2 hours.

Unpaired two-tail Student's T test (p<0.05) of 2013 versus 2015 data

Results

14 patients with 57 presentations to A&E with suspected central line infection, of which 27 (in 7 patients) were confirmed by culture (47.4%). Most organisms cultured originated as skin (44%) or gut commensals (39%). Mean time to antibiotics was 136 minutes (95% CI: 116.9-155.7), a statistically significant improvement to 2013. There was no statistically significant difference in times to antibiotics between the proven and suspected central line infection groups, but the standard deviation was greater in the proven group (80.1 vs 58.0).

Summary and conclusion

47% of patients with suspected catheter-associated sepsis grew organisms on culture, demonstrating the need for rapid assessment and treatment with IV antibiotics in A&E. Current mean time to antibiotics is 136 minutes, a significant improvement from 2013 audit but still above our 2 hour aim.

We recommend the measures taken between 2013-2015 to decrease time to antibiotics, those being: a fast-track for these high risk patients to present to specialists, and phone triage by specialist PN nurses. In addition, we consider the following to be of use for further improvement: prescription of stat doses of IV antibiotics before regular dosing; A&E staff trained in central line access available at all times; emergency healthcare plan or clinical alert on admission to increase recognition of current protocol; increase staff awareness of central line complications and management.

19 Outcome measures and quality of randomised controlled trials in paediatric non-alcoholic fatty liver disease

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Background

There is currently a lack of consensus regarding the most suitable primary outcome for clinical trials in paediatric non-alcoholic fatty liver disease (NAFLD), which in part is due to the difficult balance between quality, accuracy and minimising risk to the patient.

Aim

We aimed to perform a systematic review of all randomised-controlled trials (RCT) in paediatric NAFLD to assess heterogeneity in outcome measures, risk of study bias and compliance with CONSORT quality recommendations.

Subjects and Methods

The MEDLINE database was searched for RCTs in children with fatty liver disease, in English, using MeSH terms 'child/children,' 'fatty liver' and 'clinical trials' as topic. Published articles were excluded if participants were >18 years and had other causes of liver disease. Data collection included interventions, primary & secondary outcomes, the Cochrane Risk of Bias score, and CONSORT reporting quality checklist. The review was registered on PROSPERO (CRD42016048084).

Results

189 abstracts were reviewed, of which 18 RCTs met the eligibility criteria. In total 7 therapies were assessed in a median 59 (range 10-180) size of study population. 2 out of 18 RCTs used liver biopsy as their primary endpoint, namely improvements in the NASH Activity Score and fibrosis stage. Of the remaining 16/18 studies, the primary endpoints included: change in ALT or AST (6/16; 38%), ultrasonographic echogenicity (3/16; 19%), liver fat on magnetic resonance imaging (3/16; 19%), and biochemical markers of insulin resistance and/or serum lipids (4/16; 25%). No studies have used novel biomarkers of NASH or non-invasive scoring systems for NASH/fibrosis. Only 2 out of 18 RCTs (11%) were assessed to be of high validity with a Cochrane risk of bias score of greater than 5. The quality of reporting was assessed using the CONSORT checklist and the median score was 18.5/25 (range 12.5-24.5), with a lack of descriptions of: randomisation or blinding, dates during which the trials took place, limitations and potential harms, and information about the trial protocol and funding sources.

Summary and conclusion

Our review highlights the heterogeneity of primary outcome measures and risk of study bias in RCTs in paediatric NAFLD. Without uniform agreement of trial design and clinically meaningful endpoints, drug development in children with NAFLD will remain challenging.

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20 Graft vs Host Disease and Gut Involvement Following Paediatric Bone Marrow Transplant: 4 Year Experience In Manchester

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Background

Bone Marrow Transplant has been one of the most exciting medical developments in recent times. The Bone Marrow Transplant programme in Manchester was established in 1985 and is now one of the biggest in the UK. By providing a specialist service covering the North West we have developed expertise in providing successful transplantation. The procedure is not without risk and GvHD with involvement of the gut remains a diagnostic and management challenge.

Aim

To review all cases of bone marrow transplant over a 4 year period in a tertiary paediatric transplant unit and assess rates of GvHD and Gut GvHD as well as possible trends.

Subjects and Methods

retrospective analysis of Bone Marrow Transplants conducted in Royal Manchester Childrens Hospital over a 4 year period between 2010 and 2015. All the patients who received a Bone Marrow Transplant between January 2010 and January 2015 were selected from the local transplant database (143 transplants). The database was analysed and rates of GvHD (72) and Gut GvHD (15) were identified. Those with involvement of the gut were assessed against those without with respect to morbidity and mortality rates as well as the diagnosis, conditioning and matching for the transplant as well as virology results.

Results

Of the 143 patients who had received a transplant in the period, half developed acute GvHD and a quarter developed chronic GvHD. 10 patients required a second transplant and 22 patients in total died following transplant (15%). Matching for transplant was good (69% with 10/10) but 10 out of the 22 that died had a 10/10 HLA Match.

15/72 GvHD patients developed GvHD of the gut and like the rest of the GvHD sample, around half of transplants in this group had a 10/10 match. There were 4 deaths in the gut group (27% of the gut group). Overall rates of chronic GvHD rates were higher in this group (87% vs 20%). There were 2 ICU admissions in the gut group vs 6 in the non gut group (13% vs 4.7%). 7 patients in total were highlighted as having severe GvHD, of these 5 had GvHD of the gut.

There were a wide range of diagnoses as well as conditioning strategies prior to transplant but no differences were found between the 2 groups in this regard. There were no differences in the proportion of patients with CMV and EBV between the 2 groups at 75% and 50% respectively.

Summary and conclusion

43 patients received a bone marrow transplant in Manchester over a 4 year period. Bone Marrow Transplant does involve risk, however overall levels of GvHD and the need for second transplant were low in our cohort which likely reflects effective matching as well as adequate T-Cell depletion prior to transplant.

Only 10% of patients developed gut involvement, however levels of morbidity and mortality were strikingly higher in those that had gut involvement. Those with involvement of the gut were more likely to develop severe GvHD, Chronic GvHD, be admitted to ICU and die following transplant. We could not find any differences between the groups with regards to diagnosis, virology, matching and conditioning for transplant.

Our results suggest gut involvement in GvHD is a sign of significant GvHD and suspicion of gut involvement should be high. Clinical diagnosis can be difficult and early investigation and management may be merited. We could not attribute any potential cause for our results. Overall numbers are low and we plan on gathering further data to investigate the management of those with gut involvement to provide additional insight on management and highlight areas that can improve outcomes.

21 Contribution Of GI endoscopic biopsy for diagnosis of acute Graft-versus-host disease (GVHD) in children

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Background

GVHD remains a major complication of Hematopoietic stem cell transplant (HSCT) often with skin followed by GI tract involvement at the onset of acute (<100 days post-transplant) GVHD. GI acute GVHD (aGVHD) is diagnosed by histological examination of endoscopic mucosal biopsies. This study was undertaken to better guide the investigation of children with suspected GI aGVHD.

Aim

Our aim was to determine whether recto-sigmoid biopsies alone would be sufficient in establishing the diagnosis of GI aGVHD.

Subjects and Methods

A retrospective case chart analysis was performed on all consecutive children (< 18 years) who had undergone endoscopy for suspected GI aGVHD within 100 days of an allogeneic stem cell transplant from July 2008 to August 2015 at a tertiary paediatric haemato-oncology centre. Electronic histology reporting services, the HSCT database and operating theatre records were utilised to identify patients. GVHD was defined histologically as the presence of gland apoptosis, not explained by other inflammatory or infectious etiologies. The patient was diagnosed with GI aGVHD if at least one biopsy site was positive.

Results

22 patients with 24 endoscopic procedures (simultaneous UGI and LGI endoscopy), M14:F8 with a median age of 5.7 years (range 0.5 – 16.6) were included. Those children without histological evidence of GvHD or Clostridium difficile infection were excluded (n=7). The most common endoscopic finding was normal mucosa (27%) with diarrhoea (100%) being the commonest symptom. Nearly 60% of children presented with predominantly lower GI symptoms and 9% with upper GI symptoms only. The sensitivity at oesophagus, stomach, and duodenum were 13%, 42% and 67% respectively. The overall sensitivity for upper GI endoscopy was 79%. The sensitivity for recto-sigmoid biopsy was 96% whether patients presented with diarrhoea, nausea, and vomiting or abdominal pain. In a subgroup of 10 patients who had ileo-colonoscopy sensitivities at the terminal ileum, caecum, colonic and recto-sigmoid were 83%, 88%, 100%, and 100% respectively. In only one patient diagnosis was achieved on Upper GI endoscopy they then had a further endoscopy 3 weeks later showing GVHD on both upper and lower GI biopsies. No complications were identified secondary to endoscopy but one patient developed hypotension requiring fluid resuscitation and another bled secondary to liver biopsy.

Summary and conclusion

As suggested in other adult studies, biopsy of the recto-sigmoid was best in diagnosing GI aGVHD regardless of symptom presentation in our cohort. In patients with a high index of suspicion with negative sigmoidoscopy, a combined ileo-colonoscopy and upper GI endoscopy may be considered.

22 Service evaluation of oesophageal impedance-pH studies conducted in a paediatric tertiary gastroenterology service

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Background

Multichannel intraluminal impedance pH monitoring has developed as a tool for evaluating gastro-oesophageal reflux (GOR) in children. Impedance monitoring helps assess patients with non-acid reflux, feeding related reflux, persistent reflux despite being on proton pump inhibitors and those with extra-oesophageal symptoms. In the latest NICE Guidelines on Gastro-oesophageal reflux in children and young people they have recommended requesting impedance-pH studies in children with mainly extra-oesophageal symptoms.

Aim

Our aim was to evaluate outcomes of impedance-pH monitoring in patients referred to a tertiary gastroenterology unit.

Subjects and Methods

We retrospectively reviewed case notes of all patients who underwent impedance-pH monitoring at our institution over a 12-month period (Jan- Dec 2015).

Results

Over the 12-month period 110 impedance- pH studies were requested. The vast majority of the requests were made by the gastroenterology team (48%) and the respiratory team (47%). Co-morbidities of these patients included: respiratory (34%), gastroenterology (18%), oropharyngeal (8%), neurological (7%), genetic (7%) and immunology (2%). In a significant proportion of patients, the study was conducted as an outpatient (78%).

The main reason for requesting an impedance-pH study was due to oesophageal symptoms (56%) in which regurgitation was the prominent symptom. Referral for non-oesophageal symptoms was made in 44% of patients with recurrent chest infection being the commonest reason. Half of the patients who underwent an impedance-pH study also had another test done this included: OGD (n=28), gastric emptying (n=13), bronchoscopy (n=4), manometry (n=3) and breath test (n=2).

In 60% of patients' anti-reflux medication was stopped prior to the study. Most of the patients were orally fed (74%) rest were NG fed (14%), NJ fed (2%) and gastrostomy fed (10%).

The test was successfully completed in 71% (78/110) of patients. Of these 78 patients, 47 had a normal impedance-pH study and 31 had an abnormal study. Pathological acid reflux was reported in 22 of 31 patients (71%) with an abnormal study, 6 patients had non-acid reflux and 1 patient had both acid and non-acid reflux.

In 25 patients (32%) anti-reflux medication was optimised as a result of the study. In 10 patients anti-reflux medication was stopped and in 28 patients there was no change in management. 8 patients were referred for surgery – 6 for gastrostomy and 2 for PEG-J.

Impedance-pH study could not be performed in 32 patients (29%) because of the following reasons: failed insertion / patient pulled probe (n=13), technical error (n=7), unwell on the day (n=4) and did not attend (n=8).

Summary and conclusion

We found the cohort of patients referred for impedance-pH study is generally complex with significant comorbidities. The high rate of failure to perform this test highlights how difficult it is to conduct in children. However, in patients in whom the test was successfully performed the findings influenced treatment decisions. Our study has shown that impedance-pH study is useful in complex patients with multiple comorbidities presenting with a range of symptoms.

23 The use of combined oesophageal impedance-pH recording in the evaluation of gastrooesophageal reflux in patients with respiratory illness

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Background

Combined oesophageal impedance-pH recording has been increasingly used in patients with chronic respiratory illnesses including cystic fibrosis and recurrent chest infections to assess whether gastro-oesophageal reflux could be a contributing factor to their respiratory symptoms and illnesses.

Aim

To review our single centre's use of combined oesophageal impedance-pH recording in the evaluation of gastro-oesophageal reflux in patients with respiratory illness and to assess the value of the test on the therapeutic implications in respiratory patients.

Subjects and Methods

Data of all patients in a tertiary paediatric unit referred by the respiratory team to our gastroenterology team for combined oesophageal impedance-pH recording from July 2015 to October 2016 were retrospectively reviewed. Information including patients' clinical condition, anti-reflux treatment, results of combined oesophageal impedance-pH recording and subsequent management was collected.

Results

Over the 16-month period from July 2015 to October 2016, there were 94 referrals from the respiratory team to our gastroenterology team for combined oesophageal impedance-pH recording. Median age was 6 years old (range 6 months old – 16 years 11 months old). Of the 94 referrals, 38 referrals (41%) did not have the study carried out due to non-attendance (15 referrals), technical failure (7 referrals), and intolerance to intubation of impedance catheter (16 referrals). 41 of the 94 referrals were for impedance study off anti-reflux treatment, and 53 referrals were for impedance study on anti-reflux treatment. Of the 94 referrals, 15 patients were referred twice for combined oesophageal impedance-pH recording. Taking repeated referrals into consideration, there were 79 individual respiratory patients referred for combined oesophageal impedance-pH recording. Of these, 28 (35%) patients had diagnosis of cystic fibrosis, 24 (30%) patients had background of recurrent chest infections and 7 (9%) patients had asthma. Of the 56 referrals that successfully had the combined oesophageal impedance-pH recording, 24 had the impedance study off anti-reflux treatment: 8 were anti-reflux treatment-naïve whereas 16 were on anti-reflux treatment but had the treatment stopped for the purpose of the impedance study. Of the 56 referrals that had the impedance study, 2 had mildly elevated oesophageal acid exposure in recumbent position, 2 had increased acid exposure in recumbent position, 7 had pathological acid gastro-oesophageal reflux, 5 had pathological acid reflux in recumbent position, 3 had increased number of non-acid reflux episodes, 1 had pathological gastro-oesophageal reflux. Of these 20 referrals with abnormal results on impedance study: 12 had a change of their anti-reflux medications on follow-up, either with the addition of anti-reflux treatments or change of medications, 4 continued on the same medications, 1 had fundoplication and PEG-J inserted, 3 patients have not had their follow-up clinic yet hence no information on treatment. Of the 36 referrals with normal study or with no evidence of pathological gastro-oesophageal reflux: 15 had no change in their anti-reflux regimen, 4 had their anti-reflux regimen weaned or stopped, 3 had their anti-reflux medication dose increased, 6 remained on no anti-reflux treatment, 2 had change of anti-reflux medications and 6 had no follow-up information.

Summary and conclusion

Combined oesophageal impedance-pH recording remains one of the useful diagnostic modalities that may help guide respiratory physicians in managing their patients. However, there is no consensus about pathways of referral for diagnosis of gastro-oesophageal reflux disease in children presenting with chronic respiratory symptoms. Further multicentre prospective studies and better collaboration with our respiratory and ENT colleagues is necessary to more rationally utilise combined oesophageal impedance-pH studies to diagnose supra-oesophageal manifestations of gastro-oesophageal reflux disease.

24 Audit of complications of gastrojejunal feeding- a single centre experience

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Background

Jejunal feeding is becoming more common within paediatric complex enteral nutrition (CEN) as an alternative to gastric feeding in those with poor feed tolerance. There is a paucity of information on complications associated with the various devices available and a lack of clinical experience with their use, particularly outside of paediatric gastroenterology

Aim

The aim was to examine the frequency and types of complications associated with gastrojejunal (GJ) feeding within our CEN service from 01.01.2013 to 31.10.2016.

Methods

All patients within the Complex Enteral Feeding clinic at the Royal Hospital for Children were included. Patients were identified using the CEN database and confirmed through case note review. Basic demographics were collected including: age, gender, underlying medical conditions and reason for gastrojejunal feeding, type of device, complications and outcome of GJ feeding.

Results

38 patients were identified, 8 excluded (1 non jejunally fed and 7 not under the CEN service), 22/30 (73%) were male. The most common underlying reason for GJ feeding was neurological impairment:10 cerebral palsy, 8 developmental delay and 6 underlying neurological syndromes. Other conditions included GI dysmotility in 5, short gut in 3, complex cardiac in 3 and gastroesophageal reflux in 15. The most common reasons for GJ feeding were: vomiting in 18, poor weight gain/failure to thrive in 11 and poor enteral feed tolerance in 6.

Fourteen patients (47%) had Corflo™ inserted as a primary device (16Fr with 6Fr jejunal extension), 10 (33%) had a gastrojejunal button (14Fr or 16Fr) and 6 (20%) had a primary Freka™ device (15Fr with 9Fr jejunal extension). In those with Corflo™ the most common complication was blockage of the device occurring in 8/14 compared with 2/10 with a GJ button and none with a Freka™. 4/14 had their device blocked at least once (2 Corflo™ and 2 GJ button), 2/14 (both Corflo™) blocked twice, 3/14 (all Corflo™) blocked 3 times and 1/14 blocked their Corflo™ device on 4 occasions. The other most common complication was dislodgement of the device occurring in 19/30 patients requiring reinsertion up to four times, 11/19 Corflo™ and 8/19 GJ button. Other complications included disconnection of the device, GJ button twisted on itself and problems with end connectors.

At the study end 13/30 remained jejunally fed although 3/13 proceeded to formal Roux-en-Y jejunostomy, 2/13 had witzel jejunostomy and 8/13 still had a GJ device. 12/30 were de-escalated to gastric feeding, 2/12 underwent fundoplication and one patient died of an undiagnosed neurological condition. Two patients were nil enterally due to underlying disease.

Conclusion

In a selected group of patients GJ feeding improves symptoms of vomiting and discomfort as well as improving enteral tolerance. However, complications of the devices are common leading to a significant burden on the MDT as well as families in managing the devices. In our single centre experience, CorfloTM devices blocked and dislodged more frequently than other devices despite significant input from the MDT. Consequently, we have changed our practice to use FrekaTM devices as our primary GJ device give the unacceptable rate of complications observed with other devices. Further multi centre collaboration is needed to determine if these results are applicable to other centres using gastrojejunal devices

25 Diagnostic evaluation and management of obscure gastrointestinal bleeding in children- a single centre experience

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Background

Failure to identify the cause of gastrointestinal (GI) bleeding at upper GI endoscopy and ileocolonoscopy poses a diagnostic and therapeutic challenge to the clinician. We present our 6- year experience on the management of obscure GI bleeding (OGIB) in children.

Subjects and Methods

The records of paediatric patients with OGIB who underwent diagnostic evaluation and management in our centre between 2010-2016 were reviewed.

Results

14 patients (6M) aged 2-16y (median 9y) were identified. The presenting symptoms included hematochezia (5), melaena (7), hematemesis (3), collapse secondary to GI bleed (2), refractory iron deficiency anemia (3) and recurrent abdominal pain (3). 5 patients received blood transfusions and 4 octreotide/ terlipressin. 8/14 had been investigated in other tertiary centres with upper GI endoscopy +/- ileo-colonoscopy (14), wireless capsule enterosocopy (WCE) (5), Meckel's scan (2), CT angiogram (2), technetium-99m red blood cell scan (1), laparotomy (2) and laparoscopy (2). 2 had had segment of the colon removed. The interval between initial presentation and referral to our centre ranged from 4d to 10 years (median 16m). All patients had endoscopic reassessment with upper GI endoscopy and ileo-colonoscopy in our Unit. The diagnostic work up included WCE (13), antegrade and retrograde double balloon enteroscopy (DBE) (8), laparoscopic assisted enteroscopy (4), CT angiography (1), CT abdomen (1) and Meckel's scan (1). In 2 patients no cause was identified and in one the bleeding was attributed to severe esophagitis. The diagnosis, diagnostic modality and management in 11/14 patients are shown in table 1.

Patient	Diagnosis	Diagnostic Modality	Management
1	concentric stenosis and ulcers in small bowel	Lap assisted enteroscopy	Referred for genetic testing SLCO2A1
2	blue rubber bleb naevus (gastric body)	Upper GI endoscopy	Argon plasma coagulation, endoclip, proximal gastrectomy
3	Apthoid ulcers (distal ileum)	DBE	Treatment for Crohn's disease
4	Polyps in small bowel (Peutz-Jeghers)	WCE	Polypectomy/ endoclip
5	Angiodysplastic lesions (colon)	Colonoscopy	Referral for resection
6	Meckel's diverticulum	Lap assisted enteroscopy	Excision
7	Perforated gastric duplication cyst and fundic type mucosa, colic fistula	CT abdomen	Resection
9	Threadworms in small bowel	DBE	Mebendazole
9	Meckel's diverticulum	Lap assisted enteroscopy	Excision
10	Lymphangectasia	DBE	Conservative treatment
11	Jejunal varices	DBE	Banding

Conclusion

In our case series the cause of GI bleeding was identified in 80% of the cases. Advances in small bowel imaging with WCE, DBE, radiographic imaging and close collaboration between gastroenterologists and surgeons have improved the diagnosis and outcomes of OGIB in children. These diagnostic modalities are available in centres with special expertise.

26 Epidemiology of genital lymphoedema as the initial presentation of paediatric Crohn's disease

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Background

Genital lymphoedema is caused by inflammation and granuloma formation in the lymphatic system and is recognised as a presentation of cutaneous metastatic Crohn's disease (CD). It can precede luminal presentation by months to years, particularly in younger patients; many children with cutaneous CD have genital involvement. Despite several case reports, there is a scarcity of epidemiological studies demonstrating the incidence of genital lymphoedema in a paediatric population with Crohn's disease.

Aim

We aimed to identify the incidence of genital lymphoedema as the initial presentation of paediatric CD within a population-based cohort.

Subjects and Methods

Using a prospective, regional database, demographics and phenotypic data of all incident and prevalent paediatric inflammatory bowel disease (PIBD) patients in South-East Scotland between 01.08.97 and 31.12.11 were reviewed. Case notes of all CD patients were reviewed and those with genital involvement identified. Using all CD as the denominator, the incidence of genital lymphoedema as the initial presentation of CD in all patients (incident and prevalent) in this cohort was calculated.

Results

A total of 204 incident and prevalent cases of CD diagnosed less than 17 years of age were recorded in SES during the study period. 5 patients (2.5%) were identified as having genital involvement prior to, or at the time of, CD diagnosis. These patients were aged 4-15 years at presentation (median 9 years); 3 were male. One patient was diagnosed with CD despite normal endoscopic examination after developing perianal abscesses and fissures one year after histologically proven granulomatous genital oedema. Of the other 4 patients, only one presented with concomitant gastrointestinal and genital disease. The other patients were diagnosed with CD on endoscopy 8 months, 1 year and 3 years after initial presentation with genital oedema. 4 patients with genital oedema had concurrent perianal disease and one had oral disease.

Summary and conclusion

To our knowledge, this is the first paediatric, population-based, study of genital lymphoedema as an initial presentation of CD. In this cohort, 2.5% of paediatric CD within a regional PIBD cohort at diagnosis had prior or concurrent genital lymphoedema due to CD. This significant proportion highlights the importance of considering CD as one of the many differential diagnoses of genital oedema, particularly in the presence of perianal disease or other gastrointestinal symptoms.

27 Introduction of solids in infants with short bowel syndrome

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Background

In the current literature, there is minimal evidence available outlining ideal timing and type of food introduction for infants with short bowel syndrome. A previous unpublished audit conducted by Elaine Buchanan (presented at ESPGHAN 2007) showed variable practice for introducing solids in short gut infants across the United Kingdom (UK). Varying practices across the UK and the rest of the world led to a decision to audit present practice.

The objectives of the audit were to determine:

- 1. Current practice for introducing solids in infants with short bowel syndrome.
- 2. Current first and second line formula choice in infants with short bowel syndrome, when breast milk is not available.
- 3. Current practice for food allergy testing in infants with short bowel syndrome.

Methods

A literature review was conducted using 'EMBASE, CINAHL, PUBMED' to determine the current evidence available, for introduction to solids, formula choice and allergy testing in short gut infants. A short survey, created with Survey Monkey, was emailed to ESPGHAN and BSPHGAN allied health professional members, as well as individual gastroenterology dietitians across multiple centres. The survey incorporated 10 questions (multiple choice or free text) pertaining to the above objectives. Survey responses were collected over an 11 week period, from 19th April to 4th July 2016. Results were collated in August 2016.

Results

A total of 36 survey responses were received, from 35 different children's hospitals across the UK, Europe, Israel, United Arab Emirates, South Africa and Australia.

Introduction to solids and food exclusions: The majority of respondents (58%) state their weaning and food exclusion practice would be different, depending on length/quality of bowel remaining. Only 11% have a guideline for introducing solids in short gut infants, however 33% have a guideline for food reintroductions/challenges post food exclusions. All respondents would aim to introduce solids between four and six months; however timing for introducing solids would also depend on tolerance of enteral feeds. Eighty nine percent of respondents would routinely exclude one or more foods during weaning; the most common food exclusions were dairy (39%) or four food exclusion (dairy/egg/wheat/soya), 28%. Twenty five percent reported avoidance of other carbohydrates/foods such as sugar, lactose, fruit and fibre.

Formula choice: If breast milk is not available, as a first line feed, the majority of respondents choose extensively hydrolysed (89%) and 11% choose whole protein cow's milk formula. As a second line feed choice, most respondents choose amino acid (72%), while 19% choose extensively hydrolysed, 6% modular, and 3% whole protein cow's milk formula.

Allergy testing: The vast majority of centres (89%) did not carry out any allergy testing during weaning; while 6% undertake specific IgE testing.

Summary and conclusion

There is some consensus between centres in the UK and worldwide on first line feed choice and timing for introducing solids, however clear differences were noted, particularly for second line formula choice, and routine food exclusions. This is a complex patient group where individual assessment is required; however there is a clear need for consensus statements for introducing solids and feed choice in short gut infants.

28 Screening for malnutrition on admission to hospital using the Paediatric Yorkhill Malnutrition Score - a reality check

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Background

National guidelines recommend that all patients be screened for risk of malnutrition on admission to hospital and at regular intervals during an inpatient stay. The Paediatric Yorkhill Malnutrition Score (PYMS) is a screening tool developed to identify children at risk of malnutrition at the time of acute medical/surgical admission, and includes assessment of BMI as well as questions regarding recent weight loss, nutritional intake and predicted effects of the current medical condition. Our UK tertiary children's hospital has been using PYMS since 2010 and is required to be completed by ward nursing staff for all in-patients 1-16 years old within 24 hours of admission.

Aim

To investigate current practice in our hospital by: (1) Auditing completion rates of the PYMS charts, including assessing accuracy of completion and appropriate subsequent action taken; and (2) Assessing awareness of and attitudes towards the PYMS screening tool amongst medical staff.

Subjects and Methods

- 1. All nursing folders from all medical and surgical inpatient wards were audited on three separate occasions during August/September 2016, and were examined for the presence and completeness of a PYMS chart. Exclusion criteria included: age <1 or >16 years; duration of admission less than 24 hours; undergoing palliative care. The body mass index (BMI) calculation was checked using the PYMS BMI wheel. The final PYMS score was noted and checked for accuracy. The documented response to the score was compared against the action suggested on the PYMS chart.
- 2. An anonymised survey was distributed via email amongst all levels of medical staff including questions on awareness of the tool, how often high scores had been highlighted to them by nursing staff, whether they would like to be more aware of/involved in nutritional care and whether they felt this would have an impact on patient outcome. Incomplete responses and those from staff who were not part of a team directly responsible for inpatient care were excluded.

Results

- 1. 88 patient files were audited and included for analysis. Only 66 files (75%) contained a PYMS chart. 65% were completed within 24 hours of admission. 52 59% were completed with an accurate PYMS score. 51% showed an accurately calculated BMI. Six patients (7%) were identified as being at significant nutritional risk (PYMS score 2 or more). Referral to dietician (the appropriate response) was documented on the PYMS chart for only two of these six patients. Of those with a completed chart, 22 remained an inpatient for a sufficient duration to require recalculation of the PYMS score. 7 (32%) files recalculated the PYMS score within the correct time interval.
- 60 members of eligible medical staff successfully completed the questionnaire. 52% knew the purpose
 of PYMS, and most of these were familiar with the acronym. 77% had never been informed of a high
 PYMS score by ward staff. 70% would like to be more aware of/involved in their patients' nutritional care
 and 80% thought that doing so would be beneficial to their outcome

Summary and conclusion

An audit of current practice 6 years after introduction into our hospital shows that rates of full and accurate completion of the PYMS score should be improved in order for it to remain a useful and reliable nutritional screening tool. We plan to use the findings of our survey to empower our nursing staff to consider PYMS to be more than just a form-filling exercise but rather a useful tool which can be the starting point for multidisciplinary discussions about the nutritional needs of our patients. Our finding of 7% of admitted children being at risk of malnutrition is in line with published rates from similar centres.

29 Experience of managing poorly controlled constipation in paediatric age-group using home based 'rectal washout' (Peristeen) in the nurse-led consultant supervised constipation clinic of a District General Hospital.

Dr. Tushar Banerjee, Consultant Paediatrician; Dr. Antima Banerjee, Consultant Paediatrician; Mrs. Maria Cooke, Specialist Constipation & community Nurse; County Durham & Darlington NHS Foundation Trust

Background

Constipation is a chronic condition best managed in the community. Several patients fail to respond to standard NICE guideline-based management strategies. These patients may need recurrent hospital admissions for disimpaction. The major problem faced by these children & young people is persistent soiling leading to poor quality of life and low self-esteem. The inability to manage constipation at home may lead to significant frustration among patients and parents.

Aim

To analyse patient characteristics, outcome, and adverse incidents related to home based rectal washout.

Subjects and Methods

Retrospective case note review of 7 Patients (4 boys and 3 girls) managed with a home based rectal washout system. The patients were between 7 to 16 years of age. The clinical decision of starting rectal washout was made by a consultant with gastroenterology interest along with patients and parents after detailed discussion about the risk and side effects. All patients had a history of faecal soiling, palpable fecolith, poor response to optimal oral treatment, recurrent hospital admissions for disimpaction and significant behavioral issues requiring clinical psychology input.

Results

- All patients responded to the initial treatment (within first 4 weeks) with reduction of the frequency
 of soiling and reported either complete cessation or only occasional episodes of soiling (within
 12 weeks).
- Six patients continued to be compliant. One non-compliant patient has significant behavioral issues with associated encopresis.
- Out of the six compliant patients, one continued to have abdominal distension with soft stool constipation & radiologically proven dysmotility syndrome.
- All patients and parents reported that the rectal washout was easy to perform at home & were satisfied with the improvement in social aspects of life.
- Only one non-compliant patient required retraining and reassessment.
- All patients were managed in the nurse-led constipation clinic without any report of adverse incidents.
- The frequency of rectal washout usage in all patients reduced from daily to 2-3 times a week without exacerbation of soiling.
- No hospital admission was required after commencing on home based rectal washout in six patients; only one case with bowel dysmotility needed hospitalisation for faecal disimpaction while on 'rectal washout regimen'.
- After training and initiation, all 7 patients were able to set & administer the treatment without parental supervision.

Summary and conclusion

The home based rectal washout is an effective & easy to use alternative to ACE (Antegrade Colonic Enema) procedure for children and young people with severe constipation and soiling. This treatment could be safely managed and delivered through the nurse-led constipation clinic. This is also more cost effective, less labour intensive, discreet and user-friendly. This intervention also reduces consultant follow-up clinic visits as well as hospital admissions with sustained clinical improvement. The overflow incontinence responded in 5/7 cases with some initial improvement in 2/7 cases. All children had a positive impact on their social aspects of life. Further studies are required to develop a clinical guideline for considering home based rectal washout as a standard treatment for faecal impaction with overflow incontinence in the paediatric age group. It is highlighted here that this home-based management is effective to control faecal soiling, which is a socially embarrassing situation. However, it is essential to assess the compliance issues before offering this modality for management of difficult to treat constipation.

30 Measured energy expenditure of non-ambulant, artificial fed, neurologically impaired patients: How predictive is the Oxford equation?

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Background

Recent reviews have once again highlighted the poor predictive values of estimating caloric requirements with calculations. It is further recognised that patients with chronic disease and altered body compositions are most likely to have the biggest range of error. The caloric requirements of non-ambulatory patients with severe neurological impairment are very poorly investigated. There are limited studies comparing true resting energy expenditure (REE) measured by indirect calorimetry (IC) with predictive equations such as FAO/WHO/UNO. These studies predate the introduction of The Oxford predictive equation, the validity of which is yet to be assessed in this specific group. It has been proposed the Oxford equation may prove to be the most accurate and generalizable predictive equation due to its design.

Aim

To compare REE measured by IC with the Oxford equation for BMR.

Subjects and Methods

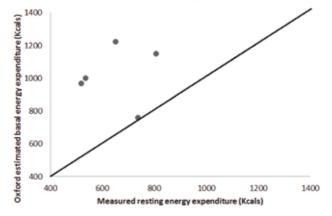
Patients were identified from the nutrition team complex case load and dietetic referrals between Jan – Nov 2016. Patients were included if they were non ambulant (GMFCS 5 or equivalent), exclusively artificially fed and had neurological impairment. Preparation and collection of data from IC followed standard recognised guidelines and vales of RQ between 0.67-1.3 were used as markers of validity of the measurement.

Result

5 patients were identified. The Oxford BMR equation overestimated the REE in all patients by 46.5%, median, (range 3.3%-47.0%).

Subject	Age (years/	BMI Centile	Resting energy expe	Resting energy expenditure (Kcals)		
	months)		Measured (IC)	Predicted (Oxford)	(%)	
1	6y /4m	<0.4	736	761	3.2	
2	7y/7m	>91st	518	969	46.5	
3	8y /2m	>25th	534	1002	46.7	
4	8y/10m	>98th	649	1225	47.0	
5	15y/4m	9th	806	1150	29.9	

Correlation of measured REE (IC) and predicted BMR (Oxford)



Summary and conclusion:

In our group of patients with severe neurological impairment and exclusive reliance on artificial feeding, the Oxford equation markedly overestimated the REE. Notably 2 patients had a BMI centile >91st suggesting historical excess caloric delivery (the practical worry in this group).

This comparison data is of concern as when resting energy expenditure is estimated rather than measured (which is most common in clinical practice) the risk and impact of overfeeding is increased. IC can assist in better estimating caloric needs in this vulnerable group.

BSPGHAN 2017 Annual Meeting

POSTERS

WEDNESDAY 25TH JANUARY 2017

W1 Crohn's disease duodenal stricture; mitomycin and endoscopic balloon dilatation.

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Background

Approximately one third of Crohn's disease (CD) cases develop a stricture within 10 years of diagnosis. [1] Endoscopic balloon dilatation (EBD) is being widely used for dilatation of oesophageal and intestinal strictures in IBD. [2] However, there are no case reports or series published about the use of EBD for duodenal stricture in paediatric CD and the evidence is limited to adult literature only. [3-4] A case of 12 year old boy with CD with duodenal stricture managed effectively with EBD is presented here.

Subjects and Methods

A 12 year old boy first presenting at 11 years of age with 7 months history of abdominal pain, vomiting and diarrhoea. The initial endoscopy showed mild thickening of duodenum and terminal ileum (TI) with histological findings of ulceration and acute inflammation with villous architectural distortion in the duodenum and TI. A diagnosis of CD was made and the patient was commenced on appropriate treatment. The MRI was not suggestive of any stricture.

Poor compliance was an issue. A reassessment endoscopy after 8 months continued to show thickening in duodenum and ulcers in TI but the histopathology showed acute duodenitis only. Regular Infliximab was commenced but diarrhoea and abdominal pain persisted. Further endoscopy 1 year later revealed a 2mm diameter stricture in the first part of the duodenum. Adalimumab was started, however vomiting quickly ensued. Duodenal EBD was then undertaken to 10mm under radiological control. This resulted in immediate symptom resolution with consequent weight gain. Mitomycin C anti-fibrotic (0.5mg/ml) was applied topically post-dilation via the endoscope. The duodenum was reassessed with repeat endoscopy after 6 weeks and demonstrated a 10-12mm calibre. (Figures)

Summary and conclusion

To our knowledge this is the first ever reported case of EBD of the duodenum in paediatric Crohn's disease including the application of topical Mitomycin C and hence avoidance of more invasive options of surgical resection or stricturoplasty was made possible.

W2 To establish if Carnitine supplements are required for those on long term Parental Nutrition (PN) as currently, due to the stability of PN it is not possible to add Carnitine. In addition to assess if Carnitine screening should become standard practice for those receiving long term PN.

Jenny Brecknock, Specialist Nurse¹, Anna Hughes, Advanced Nurse Practitioner (Trainee) ¹, Emily Swallow, Specialist Nurse¹, Dr Anne Willmott, Paediatric Consultant², Dr Hemant Bhavsar, Consultant Paediatric Gastroenterologist², Dr Maureen Cleary, Metabolic Consultant². Kelly Lamour, Principle Dietician¹, Dr Jutta Koeglmeier, Gastroenterology Consultant¹, Dr Susan Hill, Gastroenterology Consultant¹,

¹Great Ormond Street Hospital. ²Leicester Royal Infirmary With thanks for increasing our awareness of Carnitine within our patient group.

Background

Parental Nutrition (PN) is used for patients who are unable to obtain adequate nutrition through oral or enteral diet. It is a nutritional formula given intravenously via a central venous catheter (CVC) and is used either as the main or a supportive form of nutrition for those children and young people with Intestinal Failure (IF). Our Tertiary centre has developed a two week intensive training programme for parents/carers which enables patients requiring long term PN to be discharged home and cared for in the home. Recently it was discovered that a number of our patients had a low blood Carnitine level and this initiated screening of our whole cohort. Although Carnitine is naturally produced by the body an additional source is obtained through diet and absorbed in the small bowel. Given that our patient cohort have altered levels of gut absorption and have restrictive dietary requirements it raised the question regarding Carnitine supplementation. This study will be a long term study and is currently in its preliminary stages due to this only 20 results have been obtained. It is acknowledged that all current patients will be screened and as the number of patient's requiring long term PN are increasing so will this research cohort.

Air

To establish if Carnitine supplements are required for those on long term Parental Nutrition (PN) as currently, due to the stability of PN it is not possible to add Carnitine. In addition to assess if Carnitine screening should become standard practice for those receiving long term PN.

Subjects and Methods

All patients who have been discharged home on PN were included within the study. Those who are receiving home PN but awaiting training were excluded. Patient details obtained were: age, sex, Carnitine result, amount and frequency of PN, oral/enteral diet and intake and if supplements were commenced. Blood spot Carnitine levels were obtained as patients were seen in outpatient clinics or when they were inpatients. The parameters for treatment were: 10 and above no treatment needed, 10-5 monitor, 5 and below to commence supplements.

Result

In 2016 our patient cohort is 46, out of these 20 patients were screened, (11 females and 9 males) with an age range of 1 year 4 months – 14 years 3 months, mean age was 7.4 Years. 18 were on cyclical PN over 12-20hrs, 2 were on continuous PN due to hypoglycaemia. 11 (55%) received some form of enteral nutrition, 9 (45%) were Nill By Mouth (NBM). From the 20 results collected 1 (5%) had a high Free Carnitine, 7 (35%) had a low free carnitine and 12 (60%) were normal. Out of the 11 who were NBM 3 (27.3%) had a low free carnitine 8 (72.7%) were normal, from the 9 who received some form of enteral nutrition 4 (44.4%)had a low free carnitine 5(55.6%) were normal. When examining the individual markers of carnitine the most significant results were found in relation to the 7 patients with low free carnitine where 4 (57.1%) had a low Tetradecenyl carnitine and 3 (42.3%) were normal. All those with a normal free carnitine had normal tetradecenyl carnitine. For the 2 patients who require 24hr PN both had normal Free Carnitine.

Summary and conclusion

In conclusion due to the small cohort size it is not possible to get a significant p value resulting in an inability to draw statistically meaningful conclusions. However, our results do demonstrate a carnitine deficit within our patient group. Further research is needed with a bigger cohort of patients to distinguish if there is a common factor leading to a low blood carnitine level in patients on long term PN. The number of patients receiving long term PN will only continue to grow in numbers and complexity; therefore it is important to establish is there is a specific patient group who require routine screening or supplementation in order to standardise care.

W3 Epidemiology of Monogenic Forms of Paediatric Onset Inflammatory Bowel Disease the United Kingdom

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* COLORS in IBD group investigators (appendix); ** OxfordIBD cohort study investigators (appendix)

Background

Multiple rare monogenic diseases can present with inflammatory bowel disease (monogenic IBD). There are no population-based epidemiologic data on the spectrum of monogenic IBD.

Method

We performed a survey of paediatric gastroenterology, tertiary immunology, dermatology, metabolic and haematopoietic transplant centres in the United Kingdom to identify prevalent patients with monogenic IBD aged less than 16 years between 2010 and 2014. We surveyed a total of 53 monogenic disorders and recorded gene defect, IBD diagnosis, need for stem cell transplantation and mortality. We compared the age of IBD onset with classical IBD (Crohn's disease, ulcerative colitis and IBD unclassified) in several paediatric UK IBD datasets. We compared numbers to the at-risk and overall paediatric IBD populations.

Results

We recorded monogenic IBD patients with causative mutations in 24 different genes. The median age at onset of intestinal inflammation in monogenic disorders was four years. In 60% of monogenic IBD cases, haematopoietic stem cell transplantation was performed. The case fatality rate during the 5-year period was 16%. Although there is a significant enrichment of patients in the patient group with infantile onset of IBD a large proportion presents with IBD beyond the infantile age.

Conclusion

Our data provide an epidemiological rationale for providing early immunologic and genetic screening for monogenic diseases, particularly in those children with infantile and very early onset IBD.

W4 Comparison of Immunogenicity of Biosimilar Infliximab and Originator Infliximab in Children with Inflammatory Bowel Disease in Real Life Setting

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Background

Biosimilar Infliximab (CT-P13) has been used for the treatment of paediatric Crohn's Disease (CD) and Ulcerative Colitis (UC) since 2015 in the United Kingdom. There are no randomised controlled trials (RCT) in children with Inflammatory Bowel Disease (IBD) assessing the immunogenicity of biosimilar Infliximab.

Aim

To compare the immunogenicity of Biosimilar Infliximab and originator Infliximab in children with IBD.

Subjects and Methods

We performed a single centre evaluation of 220 patients (185 patients with CD and 35 with UC) who had received Infliximab for IBD. We analysed the rate of anti-Infliximab antibody positivity, clinical remission and co-immunosuppression.

Resul

60 children with CD received treatment with Biosimilar Infliximab. 34/44 (77%) had anti-Infliximab antibodies. 125 children with CD received treatment with Originator Infliximab. 55/78 (71%) had anti-Infliximab antibody positivity. There was no difference between disease characteristics, rate of clinical remission at 6 months and co-immunosuppression between these two groups.

20 children with UC received treatment with Biosimilar Infliximab. 7/11 (64%) had anti-Infliximab antibodies. 15 children with UC received treatment with originator Infliximab. 3/6 (50%) had anti-Infliximab antibodies. There was no difference between disease characteristics, rate of clinical remission at 3 months and co-immunosuppression between these two groups.

Summary and conclusion

The rate of anti-Infliximab antibody formation was not different with the use of biosimilar and originator Infliximab. Most patients with anti-Infliximab antibodies achieved clinical remission. These findings are reassuring when considering change in clinical practice to the use of biosimilar infliximab, which has the advantage of significant cost savings for the health care systems.

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W5 Percutaneous Endoscopic Gastrostomy placement in paediatric patients with Crohns Disease improves both nutrition and growth

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Background

Gastrostomy feeding has been used in the management of paediatric patients with Crohn's Disease (CD) but there is limited supporting literature on the impact on patients.

Ain

To describe the outcomes of gastrostomy feeding in Patients with CD.

Methods:

Patients with CD who received gastrostomy feeding for at least two years between 2003-2010 were identified. Data recorded included anthropometric data, CD phenotype, surgical technique used, complications, medication, feed type, median calories and volume of feed received as well as clinical outcomes.

Results

Sixteen patients were identified (14 male). Median age at CD diagnosis was 9.74 years (IQR: 7.57-11.28); median age for gastrostomy insertion was 12.63 years (IQR: 9.45-14.03). Follow up was for a median of 2.29 years (IQR: 0.33-6.5) following gastrostomy insertion. All patients had percutaneous endoscopic gastrostomy tube insertion using a pull technique, with laparoscopic assistance in 2 patients. 9/16 (56%) experienced short-term complications. Overgranulation (38%) and localised infection requiring oral/topical antibiotics (38%) were the most common complications, with leakage reported by 3 patients (19%). One patient had long-term issues with overgranulation and repeated infection. No patient developed evidence of Crohn's disease at gastrostomy sites, or fistula formation.

Anthropometry significantly improved at follow up compared to baseline: BMI at baseline was -0.92 sds (IQR: -1.97- -0.50) and had improved at T+6 to -0.12 (IQR: -0.58- 0.61) (p=0.005). BMI z score at T+12 was -0.19 (IQR: -0.94- 0.38) (p=0.04). Body weight z score had also improved significantly by T+6 (p=0.03) baseline z score -2.00 (IQR: -2.34- 0.68). Height z score at baseline was -1.85 (IQR: -2.32-1.10) and at T+24 was -1.03 (IQR: -2.15- -0.43) and T>+24 was -0.51 (IQR: -2.00- 0.29) (p=0.04 and p=0.03 respectively).

The median volume of enteral nutrition delivered per day prior to PEG insertion (10/16 pts) was 400ml (range 0-550ml) which increased to 738ml (392-1300ml) following gastrostomy insertion (p=0.009). Median daily calorie intake prior to gastrostomy insertion was 705kcal (410-1080kcal) compared with 860kcal per day post gastrostomy insertion (642-1392kcal) (p=0.01).

Conclusion

Gastrostomy feeding in paediatric patients with CD is associated with improved growth. Early complications are common although the majority are self-limiting. PEG tube insertion is useful to promote growth and guarantee access for delivery of supplemental enteral nutrition.

W6 Adrenal suppression in inflammatory bowel disease patients treated with glucocorticoids: a systematic review

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Background

Oral glucocorticoids are a mainstay of inflammatory bowel disease (IBD) treatment, with a 10-week tapering course frequently used to induce remission or control flares. Impairment of the hypothalamic-pituitary-adrenal (HPA) axis, resulting in adrenal suppression (AS), is an established side effect of exogenous glucocorticoids. However there is currently no standard guidance or recommendation to suggest routine screening of IBD patients for this potentially fatal complication following steroid use.

Aim

To systematically review the published evidence for risk of AS in both adult and paediatric IBD patients treated with glucocorticoids.

Methods

An electronic literature search using PubMed, Ovid Medline and Embase was performed using MeSH terms relating to IBD and adrenal function/insufficiency/suppression up to November 2016. A hand search of references of relevant papers was also performed. Papers presenting original data, including biochemical evidence of adrenal function during or after glucocorticoid therapy for IBD, were included. Evidence was assessed using GRADE recommendations.

Resul

198 papers were initially retrieved and reviewed; 10 papers met the inclusion criteria (6 randomised controlled trials, 4 case series). Four of the randomised trials (one paediatric) compared oral budesonide with prednisolone or placebo and measured AS as a secondary outcome. Results varied, with one study showing no increase in AS after budesonide, two suggesting AS in up to 62% of patients after budesonide, and one study showing AS in 89% of patients treated with prednisolone. Two retrospective case series (one paediatric) specifically investigated adrenal function following oral prednisolone: one measured morning cortisol and found 20% to be low; the other demonstrated AS in 60%. Both papers measured the time taken for the HPA axis to return to normal (5.6 weeks and 7.2 months respectively). The remaining 2 randomised studies and 2 case series measured adrenal function after different steroid enemas and found some degree of AS after prednisolone and betamethasone preparations.

Conclusion

Adrenal suppression in patients with IBD after or during therapy with either oral or rectal glucocorticoids has been shown to occur in up to 89% of patients. However good quality, adequately powered, studies are lacking and methods of measurement of adrenal function, as well as timing of testing in respect to courses of steroid therapy, varied across the studies. Stimulation testing with ACTH (adrenocorticotropic hormone) analogues is the most sensitive method but is a more complex procedure than a morning cortisol assay. Clinical significance of AS had not been explored in the selected studies but case reports and experience from other patient groups suggests that it can be non-specific, mimic IBD symptoms or even lead to death.

W7 First UK report of Crohn's-like colitis in children with oculocutaneous albinism - Hermansky-Pudlak syndrome (HPS)

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Background

Recent advances in genetic mapping and sequence techniques have identified "orphan "disorders which may cause monogenic IBD-like disease. Although most cases of IBD are caused by polygenic contribution toward genetic susceptibility, there is a diverse spectrum of rare genetic disorders that produce IBD-like intestinal inflammation. Hermansky-Pudlak syndrome (HPS) is an autosomal recessive hyper inflammatory disorder (types 1, 4 and 6), consisting of the triad of oculocutaneous tyrosinase-positive albinism, prolonged bleeding time secondary to platelet storage pool defect and ceroid depositions within the reticuloendothelial system. Schindella et al (1) first reported a granulomatous colitis associated with this syndrome.

Case 1

A 7 year old girl presented with abdominal pain, diarrhoea and rectal bleeding. She had oculocutaneous albinism, moderate sensori neural hearing loss and headaches, treated with Pizotifen. Gastrointestinal biopsies revealed pancolitis with mild to moderate architectural distortion, increased lamina propria inflammation, eosinophilic and neutrophilic crypt abscesses. Her neutrophil burst test, T&B lymphocyte subsets, immunoglobulins, full blood count, CRP and ESR were normal. Exclusive liquid diet achieved a clinical remission, and currently she is well without any treatment.

Case 2

A 16 year old girl presented with abdominal pain, mouth ulcers and diarrhoea containing blood. She had oculocutaneous albinism, polycystic ovaries, a gallstone and focal segmental glomerulosclerosis, treated with Cyclosporin. ESR 45mm/hr, CRP 3 mg/L, a high IgG and a negative PCR for CMV/EBV. At endoscopy, microscopically there was a mild colitis with focal minimal active inflammation of the lamina propria in sigmoid and rectum. The architecture was preserved and the findings were nonspecific, but Cyclosporin may have modified the inflammation. Her symptoms escalated with urgency and abdominal cramps, she was started on Mesalazine and Budesonide in combination with Prednisolone enemas were added. Repeat endoscopy after treatment did not reveal a diagnostic abnormality although mild bowel symptoms persisted with ESR 80mm/hr and a normal CRP.

Discussion

This is the second report of Crohn's-like colitis in children with oculocutaneous albinism (Hermansky –Pudlak syndrome). Monogenic or single gene defects have been found to alter intestinal immune homeostasis via several mechanisms, such as hyper inflammation of auto inflammation or disrupt T-and B- cell selection and activation (2,3). As a group, these diseases have a high morbidity and may require different treatment strategies than most cases of IBD. Access to genotype - specific therapies is important as may avoid adverse effects of medical therapy or surgery in patients who are unlikely to benefit from conventional IBD therapies in the long term.

Abdominal pain and rectal bleeding were the prominent features in both cases and these should raise suspicion of HPS in children with oculocutaneous albinism. Since this is a rare disease, we recommend enrolling affected children in the 100'000 Genome project and collecting data nationally.

References

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W8 Transition: not just an event – Using a transition clinical practice benchmarking tool1 in an IBD transitional service in a tertiary children's hospital

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Background

Transitioning from the children's services to adult services can be a difficult time for young people (YP) with Inflammatory Bowel Disease (IBD) and their parents. Lack of "being prepared" was a key finding from the recent report on transition by the Care Quality Commission (CQC 2014). Subsequently a clinical practice benchmarking tool 1 for transitional services was developed and published.

Aim

To evaluate the quality of transitional care for YP with IBD and their parents in a tertiary children's hospital and identify areas of improvement.

Subjects and Methods

All YP with IBD who were undergoing transition or transferred care during January 2013-January 2016 were identified from a transition database. A patient and parent questionnaire was developed using a transition clinical practice benchmarking tool 1 and approved by the local PALS team. Two rounds of questionnaires were sent in January 2016 & March 2016. Case notes of all YP were reviewed to assess transition related documentation.

Results

72 YP with IBD were identified from the transition database during the study period. The questionnaire response rate was 33% (24) for parents and 29% (21) for YP. High levels of satisfaction were reported with information giving in clinic and transition clinic experience.

Questions to YP with IBD	DGH (n=11)	Local (n=10)
Did you feel you understood your condition at transition?	91%	100%
Did you have a chance to visit the adult facilities including inpatient areas?	18%	30%
If not would you have liked to?	44%	43%
Do you feel information was given to you in a way you could understand?	91%	100%
Were you invited to have the consultation without parents present?	64%	60%
Was readiness for transition discussed with you?	82%	70%
Did you feel ready for transition in terms of knowledge and confidence in managing your condition?	91%	80%
Questions to parents of YP with IBD	DGH (n=12)	Local (n=12)
Did you feel you were given help to take a step back but still support your son/daughter?	75%	92%
Were you given any written info on this?	25%	75%
Were you informed about the plan for transition?	75%	100%
Were you given a point of contact to make enquires/raise concerns?	92%	92%
Did you feel you knew how the adult service was going to work?	83%	83%
. ,		

53 sets of notes were retrieved. Of the YP that had transferred care, 93% had documentation of transition discussion, 33% of written information given, 74% had a documented transition plan and 87% had documentation of assessment for readiness for transition.

Summary and conclusion

Overall there were high levels of satisfaction with information giving and transition clinic experience for both YP and their parents. Parents of YP being transitioned to local adult services were better informed and felt more supported. Areas of improvement identified included need for improved documentation, offering visits to adult services, independent consultations for YP and multi-speciality transition co-ordination.

W9 Copper deficiency in children receiving Home Parenteral Nutrition

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Background

A child with short gut syndrome and dependence on parenteral nutrition (PN) from birth; presented at 2 years of age with neutropenia.

Blood parameters showed: Haemoglobin 85 g/L, Platelet Count 114 \times 109/L White Blood Count 4.12 \times 109/L; Neutrophil Count 0.26 \times 109/L. Copper 1.2 umol/L (10-30 umol/L) Zinc 13.7 umol/L (5-15 umol/l). Serial blood cultures were negative and all other blood parameters were within normal limits.

His home parenteral prescription delivered over 15 hours daily provided; in addition to electrolytes and vitamins: 150% of his copper requirements, 100% of zinc and manganese requirements along with 1.1mg iron.

The cause of the neutropenia and copper deficiency was not initially apparent. His parents reported brown discolouration of the filter following administration of Home PN. This had also previously been reported in 2 other patients, in whom serum copper levels were below the reference range for age. The copper levels had previously been normal despite filter discolouration for all three patients.

Aim

To investigate the cause of low serum copper levels in 3 children receiving their total estimated energy and nutrient requirements as parenteral nutrition

Subjects and Methods

3 children with low serum copper levels and reported discolouration of the aqueous filter following the administration of cysteine containing home PN solution were identified.

Results

In the cysteine containing solutions there was an obvious discolouration to the filters and a reduction in the post filtration copper content. The amount of reduction differed for each solution by 45%, 51% and 60%. The zinc content remained stable. Repeating the process with a non -cysteine amino acid source showed no discolouration in the filter or reduction in copper levels.

Summary and conclusion

Pancytopenia is a recognised consequent of copper deficiency, but rare in patient receiving PN supplemented with adequate amounts of copper.

Brown discoloration of filters associated with a possible incompatibility between a cysteine-containing paediatric amino acid solution and copper sulphate in PN has been reported.

All three patients were converted to a non-cysteine PN solution with no alteration in copper or zinc prescription. Biochemical and haematological resolution was noted in all cases. Investigation as to the chemical nature of the precipitant in the filters is ongoing.

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W10 Micronutrients in children receiving Home Parenteral Nutrition – A Single Centre Experience

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Background

Parental nutrition (PN) is a well-established mode of nutrition for neonates, infants and children. In addition to providing calories it provides micronutrients required for the prevention of disease and for normal physiological functioning.

Aim

We retrospectively reviewed the micronutrients in 14 children receiving bespoke home PN from our unit.

Subjects and Methods

Serial results of Zinc Copper Selenium Manganese Vitamin A & E were retrospectively reviewed in all children over a 2 year period.

Children were split into two groups - 7 children in whom PN provided 100% of their estimated energy requirements (Group 1) and 7 children in whom it provided no more than an average of 70% of their estimated requirements (Group 2).

Only children who required home PN for at least 2 years were included in the study.

Children were excluded from the study if intestinal failure requiring PN was thought to be primarily due to an inflammatory cause (for example Crohn's disease or autoimmune enteropathy). Children with abnormal liver function tests at the time of sampling were also excluded from the study.

Results

Group 1

Vitamin A and E levels were noted to be higher than the upper limit of the reference range for age when receiving greater than 85% of the recommended dose of fat soluble vitamins.

Vitamin D levels were uniformly low in this group and required additional supplementation.

Three children were noted to have high manganese levels at a period when they were noted to have recurrent central line sepsis, with manganese normalising on resolution of the central line sepsis. Two children were noted to have low copper levels associated with normal zinc levels.

In one child low copper levels despite supplementation to 150% of estimated requirements resulted in neutropaenia, which resolved on changing the amino acid source from Vaminolact to Aminoven.

Group 2

Children required an average of 70% of the nutritional calories as PN – ranging from 19% to 85%. Vitamin A and E levels were within the reference ranges for age in children receiving less than 70% of the requirements as PN.

One child was noted to have a persistently raised manganese level despite removing manganese from his PN; this was thought to be due to manganese in his enteral nutrition (receiving 1.5mg/day).

Summary and Conclusion

Aside from one child presenting with neutropenia all children were asymptomatic. Vitamin D was uniformly low in both groups and required additional supplementation.

Changes in Manganese levels noted require further investigation.

Current recommended dosing of micronutrients in children receiving parenteral nutrition does not ensure optimal blood levels.

Frequent and meticulous monitoring of micronutrients in children receiving parenteral nutrition is essential to prevent side effects from over or under dosing.

W11 A Single Centre Experience in managing small bowel bacterial overgrowth in patients with intestinal failure.

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Background

Small bowel bacterial overgrowth (SBBO) and colonic fermentation (CF) are processes that may cause malabsorption and prevent weaning from PN in patients with short bowel syndrome. It presents with non-specific gastrointestinal symptoms. A number of antimicrobial treatments are often given simultaneously. Our approach is to introduce a single antibiotic and monitor clinical response and D-lactic acidosis as a marker of SBBO. A second agent may be cycled according to clinical response.

Aim

To describe the clinical efficacy of single antibiotic regimens in patients with SBBO/CF in improving symptoms and reducing raised d-lactate.

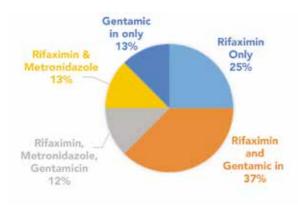
Subjects and Methods

Subjects who were treated with oral antibiotics for SBBO/CF were selected from the cohort of patients on home parenteral nutrition (PN) at Birmingham Children's Hospital. Post prandial D-lactate results were collected from the chemistry system and clinical details were collected from medical records.

Results

8 out of 40 home PN patients were prescribed antibiotic for clinical suspicion of SBBO/CF. 7 had short bowel syndrome (SBS) secondary to multiple small bowel atresias, gastrochisis, malrotation and volvulus; the majority (6) had no ileocaecal valve. The bowel length ranged from 8cm to 70cm with median of 30cm. One patient had chronic pseudo-obstruction. Criteria for starting a trial of antibiotics were based on symptoms in combination usually with raised DL.

Treatment was initiated based on symptoms in 2 out of 8 did not have raised d-lactate and 1 improved symptoms. 1 patient with raised d-lactate did not respond to treatment (d-lactate level or symptoms). 5 (63%) of those with raised d-lactate and symptoms improved within 5 months of treatment. Gentamicin cycled with Rifaximin was the commonest combination (60%) and it reduced d-lactate effectively within 5 months; 20% were on gentamicin only and 20 % on rifaximin cycled with metronidazole.



Syptoms	No of Patients (n= 8)	Improved Symptoms
Vomiting	1	1 (100%)
Abdominal Pain	3	2 (67%)
Gaseous Distension	1	0
Diarrhoea	3	3 (100%)
Belching	1	1 (100%)
Flatus	1	1 (100%)
Drowsiness	1	1 (100%)
Feed Intolerance	2	1 (50%)

Chart 1: Number of Patients (%) Using Different Antibiotic Regimens (either single antibiotic or cyclical antibiotic regimens).

Summary and conclusion

We have identified that absence of ICV is a risk factor for developing SBBO/CF in patients with SBS on HPN. Symptoms may improve by reducing carbohydrate load in diet. This approach isn't always successful and oral antibiotics may be beneficial. We have shown that treatment with single oral antibiotic in a rotating cycle was successful in the majority and monitoring of symptoms and D-lactate has been clinically useful.

W12 Granulomatous Interstitial Nephritis in Crohn's Disease Mesalazine induced or extraintestinal manifestation of Crohn's Disease?

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Background

Granulomatous Interstitial Nephritis (GIN) is a rare complication of Inflammatory Bowel Disease (IBD), with an unknown cause or pathophysiology, which raises treatment challenges. It has been regarded as either an extraintestinal manifestation of IBD with a disturbance of both systemic immunity and T-Cell activation against renal interstitium antigens, or as a side effect of aminosalicylates (5-ASA) medication. There are no current guidelines for the investigation or management of this entity, but immunosuppressive agents have been used with good effect.

Air

We report a case of a 19 year old female patient, in the process of transition to Adult services, who developed acute kidney injury while on 5-ASA treatment. There are only 12 reported cases of GIN associated with IBD (only 2 female patients), most of them presented with nephritis at the onset of IBD, only 3 cases had treatment with 5-ASA beforehand.

Case Repor

Our patient had a biopsy proven diagnosis of Crohn's pancolitis at 15 years of age and had been initially on total enteral nutrition, followed by Pentasa (500 mg TDS) in association with Azathioprine (75 mg OD).

Due to a flare up she required a step up in her therapy – Pentasa 2 grams BD, and Azathioprine 100 mg OD, 18 months after disease onset. Infliximab was introduced soon after and had to be increased to 10 mg 6 weekly due to a relapse of symptoms. Pentasa was then changed to Mezavant (2.4 grams OD) and patient also reported having taken NSAIDs for headaches.

Renal function started to deteriorate 2 years after diagnosis, at 18 years of age, with raised creatinine, proteinuria and haematuria. Renal function initially seemed to improve after stopping 5-ASA. However, creatinine levels started to rise again. At that time upper endoscopy showed mild reactive changes in the stomach and lower endoscopy revealed mild active right-sided Crohn's colitis. Renal biopsy identified severe diffuse tubulointerstitial nephritis, predominantly lymphocytic infiltrate, with a granulomatous appearance of the inflammation, a few tubules contain neutrophil casts, moderate numbers of plasma cells and eosinophils which are all features in keeping with the diagnosis of GIN. Azathioprine was then stopped, Infliximab was continued as monotherapy. A high dose course of steroids was commenced, which improved her renal function, controlled the bowel symptoms, but rather unfortunately induced Cushingoid features. Her next suggested treatment line will be to continue monitoring the renal function closely and adding on Tacrolimus to the Infliximab once steroids have been weaned.

Discussion

In the case presented above GIN appeared 2 years after diagnosis of gut related Crohn's Disease, while having mildly active gastric and colonic disease on optimum doses of Azathioprine and Infliximab. Etiology of this condition is unknown and previously it was thought to be related to 5-ASA treatment. This does not exclude a contribution of Crohn's disease to this inflammation driven process – as the patient was still having active bowel disease at the time of the renal biopsy and her condition only improved when immunosuppressive therapy was increased. There is no current marker to diagnose renal involvement in Crohn's disease, nor is there a specific dose or length of 5-ASA treatment proven to be harmful. Therefore the only way to prevent and reduce kidney injury is by monitoring renal function in all patients with IBD, whether or not they are undergoing treatment with 5-ASA.

W13 Prospective cost comparison pilot study of (PEG) percutaneous endoscopic gastrostomy placement of Single Step Low profile (Mic-Key®) Button device VS traditional 2 stage technique of Corflo PEG placement followed by replacement to a button device.

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Background

The standard practice in our unit for long term enteral feeding via gastrostomy currently requires two endoscopic procedures under general anaesthetic (GA). The one first to insert a Corflo peg to allow tract establishment followed by a swap to a low profile device (Mic-key or Mini button). It is now possible to opt straight for a low profile device thus eliminating the need for a second anaesthetic/endoscopy. The safety profile of this procedure has been established in a 3 year monocentric study 1 done in Lille, France. The reduced number of procedures, anaesthesia and time in hospital for the family alludes to a potential cost improvement opportunity.

Aims

A pilot study to compare costs of (PEG) percutaneous endoscopic gastrostomy placement of Single Step Low profile (Mic-Key®) Button device VS traditional 2 stage technique of Corflo PEG placement followed by replacement with button device.

Methods

Prospective service evaluation study that was commenced after approval from the governance dept. within the trust. Inclusion criteria: 20 consequent consented patients requiring long term nutritional support via a gastrostomy feeding device from Jan-September 2016. Exclusion criteria: Children needing prolonged hospital stay (>7days) for non procedure related complications. The children were assigned to each arm of the study group. Group A constituted the "Single" step button group and Group B was the "2- stage" Corflo PEG and secondary button replacement group. The risk benefits were explained to each patient and the parents chose which procedure they would opt for. The study was prospectively done in collaboration with coding and finance managers with financial data captured from the time of admission to discharge (upto 4 days post op).

Item Corflo Cost Conversion cost (Corflo to button)			One Step Cost
Device insertion costs	Corflo insertion £540	Removal of Corflo and insertion of low profile device £180	One step insertion £540
	Corflo kit £99.60	Snare retrieval kit £31 Mic-key £183.78	X 1 One Step insertion kit £297.55
Average cost/night of ward bed	£231/night 3 nights = £693	Day case £603	£231/night 3 nights = £693
Total cost of each procedure	f1332.60 +	f997.78 = f2330.38	£1714.33
Average Cost savings to trust per patient	2330.38-1714.33=616.05 per patient (N=9) 5544.50£		
Trust generated cost code to CCG and resultant savings	£2262 per patient (N=9) 20,358£		

The duration and drug usage of both antibiotics and analgesia in both groups was similar. The 2 children excluded from the study in each group had non-gastrostomy related complications (Crohn's disease in group B and Myotonic dystrophy needing ventilation in Group A) warranting a prolonged inpatient stay.

Conclusions

The single step procedure is both cheaper to perform and costs less to the CCG than the traditional 2 stage technique of inserting a Corflo and then converting this to a button device. In our trust we were able to demonstrate a cost saving of an average of £616 per patient. Further, the difference in the HRG coding invoice to the CCG and the actual cost highlighted that this difference was £2262 per patient. Thus the overall savings of £20,358 has contributed to the CIP (cost improvement programme) commitment of our trust to the CCG.

Reference

1.Safety of the One-Step Percutaneous Endoscopic Gastrostomy Button in Children June 2015Volume 166, Issue 6, Pages 1526–1528 Anne Jacob, MD, Frédéric Gottrand, Et Al. The Journal of PAEDIATRICS W14 Prospective patient satisfaction comparison pilot study of (PEG) percutaneous endoscopic gastrostomy placement of Single Step Low profile (Mic-Key®) Button device VS traditional 2 stage technique of Corflo PEG placement followed by replacement with button device.

Rhona Hubbard, Gastroenterology and Hepatology CNS: Lisa Bellamy, Gastroenterology CNS; Professor Thomson, Paediatric Gastroenterology Consultant; Dr Urs, Paediatric Gastroenterology Consultant; Dr Narula, Paediatric Gastroenterology Consultant; Dr Rao, Paediatric Gastroenterology Consultant

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Background

For children requiring gastrostomy direct insertion of a low profile device (Mic-key or Mini button) is now possible. Previously two endoscopic procedures under general anaesthetic were required (one to insert a Corflo peg to allow tract establishment followed by conversion to a low profile device). The one- step method eliminates the need for a second endoscopy/anaesthetic but there is limited experience of this procedure in UK paediatric patients in the UK.

Aims

A pilot study to compare overall patient satisfaction of (PEG) percutaneous endoscopic gastrostomy placement of Single Step Low profile (Mic-Key®) Button device VS traditional 2 stage technique of Corflo PEG placement followed by replacement with button device.

Method

Prospective service evaluation following approval from trust clinical governance dept. 10 consecutive consented patients from January-September 2016 were assigned to each arm of the study group. Group A was the Single step button group and Group B was the Corflo PEG group. The patients could not be randomly assigned as the risk benefits were explained to each patient and the parents chose which procedure they would opt for. Patients completed a satisfaction survey post-surgery. A follow up telephone call was made 2 weeks later to ask about pain, complications, experience of using gastrostomy device and overall satisfaction.

Subjects

Paediatric patients aged 1-16 years old with a range of medical conditions resulting in the need for long term nutritional support via a gastrostomy feeding device.

Results

20 patients with 10 in each group. Corflo group consisted of 3 males: 7 females. One step group consisted of 7 males: 3 females.

	One step group	Corflo group
Satisfied with pre-op information received	50%	100%
Pain well managed in post op period (paracetamol and morphine)	100%	100%
Family felt confident about looking after feeding device at home	100%	100%
Family found feeding device easy to care for and use	90%	100%
Family happy with device as a method of feeding their child	100%	100%

Summary

Prior to surgery both groups either asked about or would have liked to have known about procedure length, risks, method of insertion, information about gastrostomy device and recovery time. Overall the One Step group reported slightly higher average pain levels for the first 3 days post op than the Corflo group but all felt pain was well managed. Reports of post discharge pain were roughly the same across both groups. The number of post op complications reported by families was even across both groups. There was one patient in each group requiring either readmission to hospital or return to theatre for intervention to manage post op complications. Any other complications were managed by the community team or GP. There were 2 Corflo site infections managed with oral antibiotics, one blocked Corflo, one Corflo with an anchor site that wouldn't open. The gastropexy anchor points fell off before 2 weeks in 2 one step patients and 2 one step patients had red/sore gastrostomy sites that healed and resolved without intervention. By the two week follow up call 70% of the Corflo patients had already decided they wanted to convert to a gastrostomy button.

Conclusions Overall patient/parent satisfaction was equal across both groups. This level of satisfaction could be increased further by spending more time with families before either procedure to ensure they fully understand all aspects of gastrostomy insertion and the follow up care required. Following this study a gastrostomy nurse clinic has been set up facilitate this and the gastrostomy leaflets are being updated to include information about the One Step gastrostomy button.

W15 Prospective safety comparison pilot study of (PEG) percutaneous endoscopic gastrostomy placement of Single Step Low profile (Mic-Key®) Button device VS traditional 2 stage technique of Corflo PEG placement followed by replacement with button device.

Rhona Hubbard, Gastroenterology and Hepatology CNS: Lisa Bellamy, Gastroenterology CNS; Professor Thomson, Paediatric Gastroenterology Consultant; Dr Urs, Paediatric Gastroenterology Consultant; Dr Rao, Paediatric Gastroenterology Consultant

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Aims

A pilot study to compare overall safety of (PEG) percutaneous endoscopic gastrostomy placement of Single Step Low profile (Mic-Key®) Button device VS traditional 2 stage technique of Corflo PEG placement followed by replacement with button device.

Methods

Prospective study over a 10 month period from Jan-Sep 2016 was commenced following approval from trust clinical governance dept. 10 consecutive consented patients from January-September 2016 were assigned to each arm of the study group. Group A was the Single step button group and Group B was the Corflo PEG group. The procedure and associated risk benefits were explained to each family and the consenting parent chose which procedure they would opt for. Data was prospectively collected in real time during procedure, after procedure and up to a 6 week follow up period including telephone follow up. Data was extracted from purpose designed audit forms.

Subjects

All Paediatric patients aged 1-16 years old with a range of medical conditions resulting in the need for long term nutritional support via a gastrostomy feeding device.

Result

20 patients with 10 in each group. Corflo group consisted of 3 males: 7 females. One step group consisted of 7 males: 3 females.

Variable	Corflo insertion + conversion	One-Step
Average total length of surgical procedure(s)	34 minutes	39 minutes (First 3 procedures >50 mins)
No. of surgeons present	2 (endoscopist and surgeon)	2 (endoscopist and surgeon)
Complications during procedure	0	2 (20%)- Non significant 1.Small stoma site bleed (resolved with digital pressure alone) 2.Fall out of gastropexy suture needing reinsertion
Complications post procedure PEG Site infection post	4	0
prophylaxis period	1	1
Prolonged pain Other	1 patient pale, clammy with tachycardia requiring IV fluids and oxygen only	1 case of Pneumoperitoneum resolved with short laparoscopic procedure to vent air 2 cases of Gastropexy sutures falling off prematurely but with no effect on stoma tract
Average duration of analgesia	3.7	4

Average discharge time was 3-4 days in both groups.

Conclusions

The safety profile of the single step technique is similar to that of the traditional 2 stage technique with lesser risk of PEG site infections. Whilst this is only a pilot study this initial data is encouraging enough to pursue a longer study duration. The authors noted a slightly longer average procedural time for the single step but this was skewed by the initial procedures being longer in duration. As with any surgical skill, the average time for this procedure is likely to be lesser in comparison to the 2 stage technique as the surgeons gain more experience with this technique. The only significant complication in the one step group was the case of pneumo-peritoneum which was resolved and subsequent practice was changed to use CO2 insufflation in all cases of single step button placement with no further similar complications. We believe that for patients with a high risk of anaesthetic related complications the one-step procedure is a suitable alternative with a similar or lesser risk profile in comparison to the traditional 2 stage technique

BSPGHAN 2017 Annual Meeting

POSTERSTHURSDAY 26TH JANUARY 2017

T1 Role for isoamylase electrophoresis in paediatric patients with raised amylase levels

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Background

Case 1: 15 year old boy with severe ulcerative colitis with frequent flare ups was found to have significantly raised amylase level (1083 U/L) on routine monitoring for Azathioprine treatment. There was no baseline (pre Azathioprine treatment) amylase level recorded. Abdominal pains associated with his flare ups caused diagnostic difficulties and continuation of treatment. He was changed to 6 Mercaptopurine (6 MP) but his hyperamylaesemia continued posing difficulty with clinical management. He had normal imaging of pancreas and normal lipase level.

His paired serum amylase (3703 U/L) and urine amylase (125 U/L) confirmed low urinary amylase. Electrophoresis for amylase iso-enzymes confirmed presence of macroamylase. This helped to restart and continue treatment with Azathioprine monitoring his amylase level with his previous baseline levels.

Case 2: 6 years old girl with neuronal migration disorder, global developmental delay and intractable epilepsy on treatment with 3 antiepileptic drugs and gastrostomy feeds was admitted with abdominal pains probably suggesting constipation. Her amylase level was 2846 U/L. She was conservatively managed for possible acute pancreatitis in view of treatment with antiepileptic drugs known to be associated with pancreatitis. Her amylase level fluctuated with no correlation to clinical status. Her imaging and serum lipase level was normal.

Her electrophoresis for amylase iso-enzymes suggested rise in predominantly salivary amylase (see table 1 below). She was noted to have background of excessive salivation and recent injection of botulinum toxin in to her submandibular gland had not helped her.

Table 1

Serum Total Amylase	6730 U/L (<100)
Serum Pancreatic Amylase	260 U/L (<50)
Serum Salivary Amylase	6470 U/L (<50)
Serum Pancreatic/Total Amylase	0.4 (0.0-0.75)

Discussion

In both these cases, isoamylase electrophoresis helped to establish underlying diagnosis and avoided alteration to the ongoing treatment in view of raised amylase level of doubtful clinical correlation.

Summary

Raised amylase level is seen frequently in complex patients treated with multiple medications and is attributed to acute pancreatitis. Due to aetiological uncertainty of acute pancreatitis, this often leads to unnecessary modifications to the treatment. Imaging pancreas, checking lipase and estimating urinary amylase are useful investigations in these patients to narrow down differential diagnosis especially when clinical course is unusual. Isoamylase electrophoresis is helpful to look for rise in specific isoenzyme component in these patients to guide long term approach.

T2 Neonatal jaundice screens- More thought less action?

Mahrukh Mirza, Medical Student, University of Aberdeen, Foresterhill Campus; Dr Richard Hansen, Consultant Gastroenterologist, NHS Glasgow and Clyde; Dr Steve Turner, Senior Clinical Lecturer, University of Aberdeen

Background

Neonatal jaundice (NNJ) affects up to 60% of term and 80% of preterm infants and is usually a benign phenomenon. Profound early NNJ is a cause of kernicterus and prolonged NNJ can be a sign of life-threatening conditions, the most common of which is extrahepatic biliary atresia (EBA). Our hypothesis was that the number of referrals for NNJ has risen nationally and that the incidence of kernicterus and EBA have remained constant.

Aim

Our aims were to find out if the number of neonates referred with neonatal jaundice was rising and if the ratio of neonatal jaundice screens to incident cases of extrahepatic biliary atresia had changed.

Subjects and Methods

All admissions to hospitals in Scotland between 2000 and 2013 for individual aged <16 years were analysed. Admissions with NNJ, kernicterus and EBA were identified from coding. Individuals born before 2000 were not included in the analysis. The incidence of NNJ admissions and incident cases of EBA were standardised to number of deliveries per annum. Only the first NNJ admission for an individual was included in the analysis.

Results

There were 830,401 paediatric admissions of which 3147 (0.38%) were for NNJ and 43 (0.005%) for EBA (including 10 cases born before 2000). There was one case of kernicterus. The incidence of NNJ admission was 2.2/1000 live births in 2000 and this rose to 7.0/1000 in 2013. The incidence of EBA was 0.42/10000 live births (or 1 case for each 23,687 live births) between 2000 and 2013, and did not increase during this period. The ratio of NNJ admissions: incident EBA cases rose from 25:1 to 164:1 between 2000 and 2012.

Summary and conclusion

This whole-population study demonstrates that there has been a threefold rise in admissions with NNJ but no rise in the incidence of EBA. Kernicterus remains very rare. The rise in NNJ admissions most likely reflects a change in the threshold for admission with NNJ. Given the continuing rise in NNJ referrals, guidelines for referral may require amendment.

T3 Growth and bone health in children following liver transplantation

Faris Alkhalil, Paediatric Resident Post-Graduate Year 3; Rana Bitar, Paediatric Gastroenterology Consultant; Amer Azaz, Paediatric Gastroenterology Consultant; Hisham Natour, Paediatric Gastroenterology Specialist; Noora AlMeraikhi, General Paediatric specialist; Mohamad Miqdady, Paediatric Gastroenterology Consultant. Sheikh Khalifa Medical City, Abu Dhabi, United Arab Emirates

Background

Children with liver transplantation are achieving very good survival and so there is now a need to concentrate on achieving good health in these patients and preventing disease. Immunosuppressive medications have side effects that need to be monitored and if possible avoided. Glucocorticoids and calcineurin inhibitors are detrimental to bone and mineral homeostasis in addition steroids can also affect linear growth. Steroid sparing regimes in renal transplant children has shown to improve children's height.

Aim

We aim to review the growth and bone health of children post liver transplant by measuring bone mineral density (BMD) using dual energy X-ray absorptiometry (DEXA) scan and assessing if there is a clear link between poor growth and impaired bone health and use of long term steroids.

Subjects and Methods

This is a single centre retrospective Cohort study, we reviewed the medical notes of children (0-16 years) who underwent a liver transplantation between November 2000 to November 2016 and currently being followed at our centre.

Results

39 patients were identified (25 males and 14 females), the median transplant age was 2 years (range 9 months - 16 years). Four patients received a combined transplant, 2 kidney and liver transplant and 2 received a liver and small bowel transplant.

The indications for transplant included, Biliary Atresia (31%), Acute Liver failure (18%), Progressive Familial Intrahepatic Cholestasis (15%), transplantable metabolic disease (10%), TPN related liver disease (8%), Primary Hyperoxaluria (5%), Hepatocellular carcinoma (3%) and other causes (10%). 36 patients (95%) were on a calcineurin inhibitor (34 patients were on Tacrolimus and 2 on Cyclosporin). The other two patients were on Sirolimus. Low dose long-term steroids was used in 21% of the patients.

A considerable proportion of the patients had poor growth. 15% were below the 3rd centile for weight for age and 21% were below the 3rd centile for height for age. Most of our patients with poor growth were not on long term steroids.

49% of patients had a DEXA scan post transplantation. 21% of these children had low bone mineral density, one patient had met osteoporosis criteria with a vertebral fracture. Most of our patients with impaired bone health were not on long term steroids.

20% of the patients who did not undergo a DEXA scan developed long bone fractures and 50% of them were on long term steroid use which may suggest impaired bone health in these patients.

Summary and conclusion

The incidence of impaired bone health, although studied in limited number of patients; was high. Early recognition and treatment should be instituted to avoid fractures and improve bone health. Many of the patients were below the 3rd centile for weight and height however there was no clear relationship between steroid use and impaired bone health, reduced weight and reduced linear height.

T4 Overseas liver transplantation in children; One centre experience

Faris Alkhalil, Paediatric Resident Post-Graduate Year 3; Rana Bitar, Paediatric Gastroenterology Consultant; Amer Azaz, Paediatric Gastroenterology Consultant; Noora AlMeraikhi, General Paediatric specialist; Hisham Natour, Paediatric Gastroenterology Specialist; Mohamad Miqdady, Paediatric Gastroenterology Consultant

Sheikh Khalifa Medical City, Abu Dhabi, United Arab Emirates.

Background

Paediatric liver transplantation is the most widely accepted treatment for end stage liver disease. Many children requiring liver transplantation don't have access to a liver transplant unit in the country they live in and travel to overseas specialist centres for treatment. Their long-term subsequent post transplant care, however, tends to take place in their home country. This may influence the success of transplantation. The outcome and long term survival of these children has not been reviewed previously. in the United Arab Emirates (UAE) there is no liver transplant service for children or adults. Patients travel to overseas liver transplant units for transplantation. However, almost all pre and post liver transplant patient care takes place in Sheikh Khalifa Medical City; the main paediatric hospital in the UAE with an established paediatric gastroenterology unit.

Aim

We aim to review children living in the UAE underwent liver transplantation looking at the diagnosis, country of transplant, type of transplant, immunosuppressive treatment, complications and outcome including long term survival and graft survival.

Subjects and Methods

This is a single centre retrospective cohort study. We reviewed the medical notes of children (0-16 years) who underwent a liver transplantation outside the UAE from November 2000 to November 2016 and are followed up in SKMC.

Resulte

39 patients were identified (25 males, 14 females). The median transplant age was 2 years (range 9 months - 16 years). Four patients received a combined transplant, 2 kidney and liver transplant and 2 received a liver and small bowel transplant. The indications for transplant included, Biliary Atresia (31%), Acute Liver failure (18%), Progressive Familial Intrahepatic Cholestasis (15%), transplantable metabolic disease (10%), TPN related liver disease (8%), Primary Hyperoxaluria (5%), Hepatocellular carcinoma (3%) and other causes (10%). 33% of patients had their transplant in the United Kingdom (UK), 15% in India, 10% in the United States of America (USA), 10% in Germany, 8% in Korea, 5% in France, 5% in Singapore, 3% in Philippines, 3% in Belgium, 3% in Saudi Arabia, 3% in Egypt and 3% in Taiwan. 64% of the patients received a living related donor transplant, 33% were cadaveric and only one patient received a non-related donor transplant. All patients going to Germany, France, Belgium and most going to the USA received a cadaveric liver. However, most patient receiving a liver transplant in the UK and India received a living related donor transplant. Tacrolimus was the main Immunosuppressant used. 56% of patients were on monotherapy (22/39 patients); 20 on Tacrolimus, 1 on Sirolimus and 1 on Cyclosporine. 36% were on double immunosuppression; Tacrolimus and Mycophenolate Mofetil (MMF) or Tacrolimus and low dose Prednisolone and 8% were on triple immune suppression. Complications included chronic rejection 13%, hypertension 10%, Post-Transplant Lymphoproliferative disease 8%, end stage renal disease 8%, portal hypertension 5%, portal vein thrombosis 5%, hepatic artery thrombosis 2%, non-Hodgkin's lymphoma 2%, Brain hypoxia 2% and biliary stricture 2%. The overall survival rate was 92%, There were 3 deaths, and all were due to sepsis. Only one patient had a graft failure shortly after transplant and needed retransplantation.

Summary and conclusion

Our patients received liver transplantation in multiple overseas centres, and one third were in the UK. The overall indications and complications of liver transplantation in this group of patients were similar to already published reports. Despite being managed in a non transplant centre; the overall survival rate was high. This suggests that performance of liver transplantation in overseas centres with subsequent main post transplant care in home country can be safe and can carry a good survival. Sepsis was the main cause of death. This highlights the need for timely and aggressive treatment of infection in these patients

T5 Use of Exome Aggregation Consortium data to identify pathogenicity of mutations and prevalence estimations in ultra-rare monogenic hepatic disorders

Jake P. Mann, Department of paediatrics, University of Cambridge; Anna Carter, Medical school, University College London; Patrick McKiernan, Children's Hospital Pittsburgh, Pennsylvania, USA

Background

Identification of pathogenic mutations in ultra-rare conditions (those affecting <1/100,000) may be based on only a handful of exome sequences. Using small case series of a single ethnicity may result in incorrectly categorising a non-pathogenic polymorphism as a causative mutation.

Aim

We aimed to demonstrate how large whole-exome databases may be employed to exclude polymorphisms as causative in rare paediatric liver diseases.

Subjects and Methods

We searched MEDLINE, Online Mendelian Inheritance in Man (OMIM) database for ultra-rare (prevalence <1/50,000) paediatric, monogenic hepatic disorders. Conditions were then sorted into those primarily causing hepatocyte-mediated pathology. All reported pathogenic mutations were extracted from available literature, including ClinVar. Mutations were compared against population data (60,706 individuals) using the Exome Aggregation Consortium (ExAC) database (http://exac. broadinstitute.org) to classify mutations into: 'congruous', 'borderline', or 'incongruous' with the mutation's population prevalence. Mutations were analysed through polyphen-2 and SIFT (Sorting Intolerant From Tolerant) to determine the pathogenicity of mutations. Combination of population allele frequency with method of inheritance allowed for estimation of prevalence.

Results

Database search yielded 98 ultra-rare paediatric monogenic conditions involving the liver. Mutations were analysed for 10 disorders that primarily affect hepatocytes, encompassing 190 mutations. From these, we found 175/190 mutations to be 'congruous', 8 'borderline', and 7 as 'incongruous'. For example, the suggested incidence of autosomal dominant polycystic liver disease type 2 was approximately 1/152 due to the high allele frequency (0.0033) of p.Glu568del mutation in SEC63. The calculated incidence of autosomal recessive infantile hypertriglyceridaemia was 1/26,000, from the p.lle54Val mutation in GPD1 with an allele frequency of (0.0041).

Summary and conclusion

These data illustrate how population-level whole-exome sequencing databases can be used to identify mutations that are more likely to be pathogenic in ultra-rare conditions and common. Several mutations may need to be re-classified as non-pathogenic. This process can be utilised by clinical genetics and genomics research to aid in correct identification of common mutations and to aid in the estimated prevalence of rare conditions.

T6 Serum alanine aminotransferase levels in children: what is normal?

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Background

Serum alanine aminotransferase (ALT) is widely recognised for its role in detecting liver disease and injury. Despite routine use in clinical practice, normal reference ranges are difficult to establish and are likely to be both age and gender specific. Elevation of ALT above the adult reference range (female < 19IU/I male < 30IU/I) has been proposed as threshold for decision making in management of chronic hepatitis B (HBV) in both adults and children. However, current paediatric reference ranges suggest that ALT up to 60IU/I may be normal. In contrast, need for treatment of chronic hepatitis C (HCV) infection is not determined by ALT elevation. After successful eradication of HCV RNA and in the absence of co-morbidity, ALT would be expected to be normal. Monitoring ALT before and after therapy could help determine whether pretreatment values within the laboratory reference range are truly normal, or whether a significant decrease after HCV clearance suggests that a lower range is the true normal.

AIM: To determine ALT values in children before and after HCV eradication to determine whether current paediatric ALT reference range or the adult reference range, which is significantly lower, is more appropriate as a guide of liver inflammation.

Methods

A retrospective study of consecutive children treated for chronic HCV infection at a single centre. Response to treatment was determined by HCV RNA level, and ALT values before and at end of treatment were ascertained. ALT values were defined as either (i)"abnormal" if above the laboratory reference range, (ii) "laboratory normal" if within the laboratory reference range but above adult reference range and (iii) "adult normal" if within adult reference range.

Results

Of 53 children who received treatment for HCV infection, 10 were excluded because of co-morbidity (4) or lack of treatment response (6). Forty-three children were therefore studied: 23F:20M, median age at starting treatment was 8y2m (4y5m -15y11m). Before treatment median ALT was 42 IU/I (11-140), and was "abnormal" in 16 (37%), (median 1.8 x ULN, range 1.3-2.9), "laboratory normal" in 21 (49%) and "adult normal" in 6 (14%). At the end of treatment, ALT had improved in all children, and was "abnormal" in none, "laboratory normal" in 14 (33%) and "adult normal" in 29 (67%). Of 21 with "laboratory normal" values pre-treatment, ALT decreased to "adult normal" values at end of treatment in 15 (71%). In the remaining 6, all female, ALT fell from median pre-treatment level of 40 IU/I (21-45) to median end of treatment ALT of 19.5 IU/I (19-24).

Conclusion

This study revealed that ALT levels improved in children following eradication of HCV. The decrease in ALT, even in those who had "laboratory normal" ALT before treatment, suggests that the upper limit of normal in paediatric reference ranges may be too high. In children with chronic liver disease in whom the need for treatment (such as HBV infection) or treatment response (such as autoimmune hepatitis) is determined by ALT values, these findings may be significant.

T7 Similar presentation but heterogenous clinical course, gastrointestinal pathology and immunological defects in 5 children presenting with subsequently confirmed tricho-hepatic-enteric syndrome (THE)

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Background

THE, also known as phenotypic diarrhoea, is a rare autosomal recessive condition affecting exosome-mediated RNA surveillance usually presenting in first months with diarrhoea and intestinal failure with variable associated features. The GI dysfunction is felt to be a disorder of enterocyte function rather than immune-mediated damage and no treatment modalities have been identified. Aim: To compare clinical features, immunological defects, gastrointestinal histology and outcome in 5 children presenting with THE.

Subjects, Methods and Results

We present 5 children with THE (1 male) subsequently confirmed to have mutation TTC37. All examined had trichorrhexis nodosa.

Sex	PN	Liver disease	Immunology	Other features	Gastrointestinal histology	Outcome
1	2/12 – death	Increasing transaminitis and synthetic dysfuction Hepatosplenomegaly	PCP and disseminated adenovirus, Hypogammaglobulinaemia, Neutropaenia. Lymphopaenia		Neutrophilic oesophagitis. Normal duodenum. Colonic focal cryptitis and apoptosis	Died 8/12 acute liver failure + sepsis
2 ♀	3/12 - 15/12	Moderate transaminititis Resolved aged 21/12	Hypogammaglobulinaemia Treated sclg	Hypo- pigmented skin patches	Normal duodenum Normal colon	Aged 3 1/2 years. Orally fed. 50th centile. Remains on sclg
3	3/12 - 5/12	Prolonged conjugated hyperbilirubinaemia as infant Mild transaminititis Resolved aged 27/12	Enterovirus encephalitis Pneumococcal septicaemia Specific antibody deficiency IgM increased Abnormal cytokine profile with no production of IL12	IUGR Dandy Walker malformation VSD and ASD Eczema	Normal duodenal mucosa Minimal focal active coltis	HSCT aged 28/12. Now 4 years. Immunology normalised after HSCT but diarrhoea & FIT continue. Orally fed & no enteral / parenteral supplemental feeding (parental choice)
4	2/12 - 12/12	None until 10/12 when started immunosuppression then persistent transaminitis	DCT+ve. Weak anti-islet cell ab +ve Specific antibody deficiency Treated prednisolone, Tacrolimus and sclg from 10/12	Hypo- pigmented skin patches	Duodenal villous atrophy and focal erosion. Colonic apoptosis	Diagnosed as immune dysregulation and auto- immune enteropathy at 10/12. HSCT aged 17/12. Died adenovirus pneumonitis day+46 post HSCT
5 ♂	3/12 - 12/12 & 32/12- 46/12	Icterus prolongatus - florid hepatitis with microgranulomas on liver biopsy aged 3/12	Normal Hypergammaglobulinaemia aged 12 years		Upper GI - 'GVHD like' picture oedema, focal inflammation and frequent apoptoses. Colonic ulceration ++, patchy chronic active inflammation	Short stature and chronic diarrhoea with blood aged 12 years Oral supplemental feeds Immunosuppression with anti-TNFs being trialled.

Summary and conclusion:

The presentation of THE is consistent, but the clinical course, GI pathology, immunological defects and outcome are variable. The immunodeficiency may be corrected by HSCT but the intestinal features appear unchanged. Distinguishing from autoimmune enteropathy is critical and identifying trichorrhexis nodosa whilst genetic tests awaited may aid diagnosis and prevent potential harmful treatment choices.

T8 Adrenal insufficiency in paediatric liver graft recipients

Dr Anastasia Konidari; Dr Xanthippi Tseretopoulou; Dr Talat Mushtaq; Dr Marumbo Mtegha; Dr Stephen Hodges; Dr Suzanne Davison; Dr Patricia McClean; Dr Sanjay Rajwal; Children's Liver Unit, Leeds General Infirmary Great George St LS1 3EX

Background

Adrenal insufficiency has been a reported feature in adult patients with cholestasis (Quinn et al), cirrhosis (Thevenot et al)-referred to as 'hepato-adrenal'syndrome- (Marik et al Crit Care Med. 2008)-, as well as post liver transplant (Karagiannis et al, World J Hepatol. May 2015), but not well described in children. The postulated mechanisms reported are adrenal reserve depletion due to adrenal haemorrhage, (Marik et al, Int care med 2006), increased pro-inflammatory cytokines suppressing ACTH secretion (Bornstein et al, N Engl J Med 2009), low cholesterol levels as substrate for steroid-genesis, in vivo suppression of the hypothalamic-pituitary-adrenal axis through bile acid mediated inhibition of glucocorticoid metabolism (McNeilly et al, J Hepatol. May 2010), aberrant central modulation of hypothalamic peptides (McMillin et al, Mol endocrine Dec 2015), or iatrogenic due to glucocorticoid administration post liver transplant.

Aim

To report the clinical experience of adrenal suppression post liver transplant in a cohort of 250 paediatric patients managed in a supra regional tertiary liver centre over the last 15 years.

Subjects and Methods

We performed a retrospective review of the departmental database of 250 paediatric patients who underwent liver transplant for any indication. We identified patients diagnosed with adrenal insufficiency post liver transplant and described characteristics such as age and indication for transplant, presenting symptoms, time of diagnosis of adrenal insufficiency and outcome at last paediatric follow up.

Results

Five patients were diagnosed with adrenal suppression, defined as rise in cortisol less than 250, 30 minutes post administration of low dose synacthen (Marik et al, Crit Care. 2006). Two out of five patients had been diagnosed with biliary atresia, one with progressive familial intrahepatic cholestasis and two with undiagnosed chronic cholestatic disease. Patients (decimal age at transplant range 0.23 to 12.3 years) presented at variable time intervals ranging from 6 weeks to 2.3 years after transplantation, whilst patients were managed on the maintenance immunosuppression dose of 0.1 mg/kg/day. Symptoms reported from tremors, lethargy, 'funny episodes' to hypoglycaemic seizures, usually at early morning, after fasting or during inter-current illness. All patients had received same dose /kg of corticosteroids as per unit protocol at and following transplantation; no patient required treatment for acute graft rejection. Diagnosis was established at the earliest three months post 'non-specific' symptoms', pointing to undetected hypoglycaemic episodes. Increasing maintenance prednisolone dose or adding hydrocortisone replacement dose during inter-current illness was commenced with good effect. At date of last follow up all five patients still required steroid replacement with endocrinological input (range 3months -6 years) after diagnosis of adrenal insufficiency.

Summary and conclusion

Adrenal insufficiency may develop in paediatric liver transplant recipients of any age at variable time intervals after liver transplantation and may give rise to increased morbidity; there is insufficient data about pathophysiology, aetiology and risk factors that predispose patients to adrenal insufficiency, however by reporting our experience we would like to raise clinicians' index of clinical suspicion so as to facilitate early recognition of a potentially serious condition with non-specific symptomatology. Further basic and clinical research is required for identification of incidence, aetiology and clinical course of adrenal insufficiency in children post liver transplant.

T9 What is the main limiting factor to feed progression requiring feed changes post liver or combined liver and kidney transplant?

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Background

38 patients underwent a transplant in 2015 with many requiring formula changes when establishing enteral feeds after a transplant. There was an increased use of parenteral nutrition and several patients required a prolonged inpatient stay post-transplant (>30days).

Aim

To identify any limiting factors affecting feeding post liver or combined liver and kidney transplant, and implement new guidance in the dietetic protocol as required.

Subjects and Methods

All patients who underwent a liver or combined liver and kidney transplant in 2015 had a retrospective review of their dietetic case notes.

Results

Of the 38 patients, who underwent a transplant in 2015, 32 patients had a liver transplant and 6 patients had a combined liver and kidney transplant. The median day feeds were commenced post-transplant was day 4 (range day 2-10) with a median day to reach full feeds of 11 days (range 5-56 days). 12 patients (32%) required parenteral nutrition primarily for post-surgical ileus and where enteral feed progression was delayed.

17 patients (45%) received enteral feeds pre transplant rising to 25 patients (66%) discharged on enteral feeds post operatively.

22 patients (58%) required feeds changes in the post-operative period, but only 3 of these were due to concerns around large diarrhoeal losses. The majority of feed changes were required for chyle leaks and where additional calories were required to meet requirements. The findings are summarised in the table below.

Reason feed changed	Number of patients
lleus therefore put Nil By Mouth	1
Chyle leak	7
Diarrhoea	3
Increased calories to meet requirements	7
Vomiting	1
Large aspirates	2
Bilirubin normalised so MCT ceased	1

Summary and conclusion

The majority of feed changes in our patients post-transplant are made for chyle leaks where an MCT feed and diet are required, or changing to an energy feed to enable nutritional requirements to be met.

Based on the findings all children commencing enteral feeds post-transplant will be given a standard whole protein 1kcal/ml non fibre containing feed. Once a patient has demonstrated that they have reached their full feed volume to meet requirements they can be changed to the energy version of the feed. The aim being to reduce feed volumes required and encourage oral intake. The management of chyle leaks will remain unchanged with a 6 week period of MCT feeds and diet.

T10 Progressive Liver Failure and its complications in Glycogen Storage disease type IV with a novel GBE1 mutations: A case report

Dr Hina Rizvi, Liver Grid trainee; Dr Girish Gupte, Paediatric Hepatologist; Liver Unit, Birmingham Children's Hospital

Introduction

We report a case of Anderson disease; a rare subtype of glycogen storage diseases presenting with rapidly progressive liver failure and its complication.

Case

A 11 month old referred to his local hospital with concerns about jaundice. At our tertiary centre child was found to have hepatosplenomegaly, cholestatic jaundice, coagulopathy and ascites. He was stabilized with intravenous vitamin K, diuretics and albumin infusions. His abdominal ultrasound scan showed hepatosplenomegaly, free fluid. His CT abdomen angiogram showed thrombus in the portal vein along with narrowing of hepatic veins. Diagnostic work up revealed normal viral serology, acyl carnitines, lactate and negative for galactosemia and tyrosinemia screen. Bone marrow aspiration showed no evidence of storage disorder. This combined with CT findings initially raised suspicion of Budd Chiari. Niemann Pick and mitochondrial disorders were unlikely as he was developmentally normal. However, genetic confirmation was awaited. He had liver biopsy followed by cardiorespiratory arrest. Following which he developed fulminant hepatic failure, hepatic encephalopathy and had MRI head which confirmed PRES syndrome. He was not able to recover and died at 13months. His biopsy subsequently showed cirrhotic liver with PAS resistant storage material and abundant foamy macrophages, confirming GSD IV.

Discussion

Glycogen storage disease type IV (Andersen disease) is a rare autosomal recessive metabolic disorder with mutations in the GBE1 gene that encodes the 1,4-alpha-glucan-branching enzyme resulting in deficient glycogen branching enzyme activity causing abnormal, amylopectin-like glycogen deposition in multiple organs. Its clinical presentation is variable, however GSD IV presents itself in two main forms. The neuromuscular form of GSD-IV varies in onset (perinatal, congenital, juvenile, or adult) and severity. Hepatic form has progressive and non-progressive subtype. Our case had progressive hepatic subtype of this rare disorder. Genetic analysis showed that our patient was a compound heterozygous for the p.(Arg515His), c1544G>A mutation and p.(Arg524*), c1570C>T in exon 12 of the GBE1 gene.

Conclusion

In literature, mortality for hepatic variant is reported around 5 years without liver transplantation. However, our case highlights a patient with compound heterozygotes for mutations in the GBE1 gene, making it difficult to predict genotype—phenotype correlations, disease progression and hence expected mortality

T11 Nutritional Needs and Chylothoraces

Dr Rosalind Rabone, Paediatric Registrar; Dept of Paediatric Gastroenterology; Mrs Tracy Johnson – Birmingham Children's Hospital.

Background

Chylothoraces are a known complication following cardiothoracic surgery. They develop when chyle builds up around the lungs causing respiratory compromise. Conservative treatment includes total parenteral nutrition or a fat-restricted oral diet, supplemented with medium-chain triglycerides to reduce chyle production.

Aim

To explore the role of parenteral nutrition in the management of chylothorax, we present a case report with a discussion of management approaches using current literature.

Case Report

A 9 month old girl with complex congenital heart disease was admitted to PICU following elective surgery. During recovery she developed a MAPCA bleed which produced a haemothorax; causing hypovolaemia and cardiac arrest. After open chest exploration and occlusion of the MAPCA, there was a re-accumulation of the pleural effusion which was noted to be chylous. After 9 days on PICU she was discharged to the ward and referred to Nutritional Support and Intestinal Failure team (NSIF).

After exclusive PN she was commenced slowly on monogen feeds. Despite being fat-restricted, the chylothorax continued to accumulate and cause respiratory compromise. Weight became difficult to interpret as she developed oedema secondary to hypoalbuminaemia. To achieve her targeted kcal/kg whilst fluid restricted was a challenge. As a multidisciplinary team we addressed her nutritional needs without worsening respiratory distress, inducing IFALD or contributing to her oedema.

Discussion

From reviewing the literature, it is clear there are definitive management aims. These include, relieving respiratory distress, preventing re-accumulation of chyle and treating or preventing malnutrition and associated immunodeficiency, electrolyte disturbances and thrombotic events. Treatment is with surgery, nutrition and pharmacological agents.

Conservative management is recommended as 80% have resolution of symptoms with low fat enteral feeds (EN). Our patient was commenced on Monogen which is 80% MCT 20% LCT; it also contains added essential fatty acids; other formulas are commercially available. A case series reported resolution of chyle by day 3-6 with EN. There are no clear predictors for whom MCT feeds are effective.

After 3-6 week EN trial, there should be an indication for parenteral nutrition. This is controversial as it is generally reserved for intestinal or growth failure. In this case it is a treatment modality as PN bypasses the lymphatic system to prevent chyle production. This is not without risk due to CVL access and infections, fluid management and long term risk of cholestasis or IFALD, as well as high cost.

In PICU or NICU settings, fluid restriction often creates a challenge to meet nutritional requirements of the child, as found in this case. PICU often use variations of nitrogen sources to maximise PN protein. This could potentially lead to semi-essential amino acid depletion in children under 1 year of age (i.e. Vamin18 vs. Vaminolact). The PEPaNIC trial has suggested that PN is not essential in acute phase of illness whilst on PICU and withholding may produce superior outcomes.

Monitoring nutritional growth can be challenging in these patients. Weight is often unreliable due to oedema. Anthropometry should be the gold standard, but requires a skilled and consistent individual to record measurements. Serum albumin is often used as a marker of nutritional gain however its value in chronic conditions or acute illness is limited.

The addition of pharmacological agents such as Ocreotide have been shown to aid chyle resolution in 70% and can reduced the number of days of PN. The role of immunoglobulin infusions and trace elements and vitamins deficiencies (i.e. selenium) has also been explored in the literature.

Summary

Nutritional needs can be met with a Nutritional MDT approach in complex post-surgical patients with chylothoraces. It is unknown which nutritional methods are most effective. Parenteral nutrition can treat chylothoraces as well as addressing the child's nutrition. Practice is varied and on-going research on Chylothorax Treatment in Infants & Children will hopefully give us more evidence on the best practice.

T12 Development of an annual endoscopy audit plan using measures in the P-GRS (Paediatric Global Rating Scale for endoscopy) in a tertiary paediatric endoscopy service to facilitate Quality Improvement

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Background

A paediatric global ratings scale for endoscopy (P-GRS) is currently being piloted nationally, and this will provide a quality and safety framework for service improvement in Paediatric endoscopy units. An annual endoscopy audit plan is essential to help units identify that they are meeting the required measures and identifying areas of improvement in their unit.

Air

To develop an annual endoscopy audit plan to facilitate quality improvement in the endoscopy service in a tertiary centre.

Subjects and Methods

A retrospective audit of all procedures done by the Paediatric gastroenterology team during 1/10/16-15/10/16 was done. We used measures from the P-GRS to develop standards for the audit plan. Data was collected via EDMS (electronic document management system) and included review of letters of correspondence, consent, operation notes, anaesthetic charts, nursing documentation and biopsy reports. Patient feedback questionnaires were also included.

Resul

Data was retrieved on 46 patients (age range 8 months to 17 years, median age 9.5 years). 46 patients had endoscopies during the study period and data was collected on all these patients. 78%(36) of these had elective procedures. Out of the 22%(10) who had non-elective procedures, 18%(8) were urgent and 4%(2) were emergency procedures.

100% of procedures had a clearly documented indication, and had completed consent forms in the notes, all of which were 2-stage. The procedure completion rate was 100%, and bowel preparation was adequate in 98%. One patient developed post-operative oxygen requirement that required an admission; otherwise there were no other post-procedure complications noted. There were no deaths within 30 days of the procedure. Patient feedback questionnaires showed 78% of respondents rated their overall endoscopy experience as "excellent" or "good".

One patient had an endoscopic assessment for Upper GI bleeding during the audit period. This patient was risk assessed at time of admission and had an endoscopic assessment appropriately.

Summary and conclusion

The audit showed that our Unit is performing well against a number of the quality and safety measures in the P-GRS. Areas that require improvement include developing procedure-specific after care patient information leaflets, better documentation on anaesthetic needs of the patient and procuring an endoscopy reporting system (ERS). This also highlighted the need for close collaboration with other stakeholders such as anaesthetics and theatre admissions staff to share findings and implement change.

T13 The role of malnutrition screening tools and body composition in predicting surgical site infections in patients after scoliosis surgery.

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Background

Malnutrition screening tools (MSTs) are used in the paediatric setting, with the aim of identifying malnourished patients or those at risk of becoming so, to intervene at an early stage. However, despite evidence suggesting they may predict the length of hospital stay, there is a lack of data on their ability to predict clinical outcomes in specific patient groups. Surgical site infection (SSI) is a serious complication of spinal surgery, associated with prolonging hospital stay, by weeks to months.

Aim

To investigate the ability of MSTs and body composition to predict a specific clinical outcome - SSI in patients with diverse diagnoses and spinal pathology, admitted for elective spinal surgery.

Subjects and Methods

Data on SSI were collected and analysed retrospectively in post-operative children aged ≥5 years, who had undergone spinal surgery from January 2014 to March 2015. Anthropometry (age, sex, weight, height), information on diagnoses and body composition measurements (e.g. bioelectrical impedance, BIA) had been prospectively obtained, within 48 hours of admission (BodyBasics Study). MSTs (STAMP, STRONGkids, PYMS and the GOSH nutrition screening flowchart) were completed, classifying patients as low, medium or high risk when applicable; later re-categorised as "low-medium" and "high" risk, since high-risk patients are referred for dietetic input. Questionnaires provided data about patient activity, daily intake, feeding patterns and dietetic input. Diagnoses comprised three key groups; neuromuscular diseases, syndromic conditions and idiopathic scoliosis. Patients were "fully orally fed, independently", "fully orally fed, with the help of a carer" or "artificially fed" (partial enteral feeding via tube or parenteral nutrition). For all children surgery involved metalwork implants, thus perioperative antibiotics were administered as per hospital protocol. Definitions for SSI and surveillance periods were established in accordance to the "Protocol for the Surveillance of Surgical Site Infection" by Public Health England, and two variables are noted; the presence or absence of SSI.

Results

32 patients were included in the study; 12 males (37.5%) and 20 females (62.5%). The median age was 14.49 years (IQR 4.0). 31.3% (10) of the patients comprised the "neuromuscular group", 40.6% (13) the "syndromic diseases" group, and 28.1% (9) the "idiopathic scoliosis group". Mean weight was 41.88kg (SD 15.03) and mean weight SD score was -1.16 (SD 1.67). Median BMI was 18.85 (IQR 4.95) and median BMI z-score was -0.65 (IQR 1.97). 7 (21.9%) of the patients developed SSI. BIA mean SD score was -1.84 (SD 1.30). BIA SD score was not associated with SSI (p 0.253). STAMP, STRONG-kids and PYMS screening categories were not significantly associated with SSI (p. 0.750, 0.859 and 0.809, respectively). When re-grouped into "low-medium" and "high" risk groups, no significant difference was apparent between STAMP, STRONG, PYMS or GOSH the risk category and the development of SSI (p 0.454, 0.872, 0.583 and 0.327, respectively). The relative risk of SSI was 1.4 times higher for children with a more complex diagnosis (RR 1.44, 95%CI [1.1, 1.9]), compared to children who suffered only from idiopathic scoliosis, however this was not significant (p 0.061). Patients who were orally, independently fed had a significantly lower risk of SSI compared to patients who were orally fed with help, and to patients with artificial nutrition (p 0.011 and 0.021 respectively), and this effect was also observed when lean mass (BIA values) and STRONG ("low-medium", "high") or GOSH- flowchart risk categories were taken in to account in multivariate analysis (p 0.014 and 0.025 for STRONG, p 0.043 and 0.033 for GOSH).

Summary and conclusion

STAMP, STRONGkids and PYMS malnutrition screening tools and lean mass measurements did not predict SSI, in this specific patient group. "Simpler" variables such as underlying diagnosis and the ability to feed independently were related to SSI and may better identify those at risk in this patient group, although this needs confirmation in a larger sample.

T14 Setting up a Paediatric Anorectal Physiology Service - a new focus for an unsolved

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Faecal incontinence (FI) and constipation affects 5-30% of children in the UK (NICE, 2010) and remains a difficult condition to treat. Improvements in investigation of the structure and function of the anorectum in adults, has led to better outcomes and quality of life. We sought to set up a service that could offer physiological and structural investigation in children.

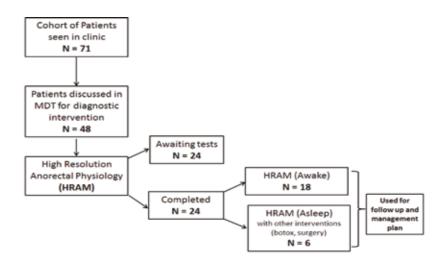
Our aim was to set up a Children's Anorectal Physiology Service (CAPS), using high resolution anorectal physiology (HRAM), endoanal ultrasound, colonic transit, MRI proctogram and validated questionnaires to assess bowel and psychosocial function. The data we produced will be used to identify novel phenotypes of children with FI and management pathways.

Subjects and Methods

A Health Foundation, Innovating for Improvement grant is funding the project for one year and seeks to demonstrate the efficacy of the service. All patients with FI and constipation, from neonates to 16 years are eligible. The project is designed as a longitudinal observational study, comparing data before and after investigation and treatment. Patients will be screened based on the following: St Mark's Incontinence Score; Cleveland Constipation Score; Pediatrics Quality of Life; Pediatric Index of Emotional Distress; Strengths and Difficulties Questionnaire, a multidisciplinary team (MDT) will decide on the need for physiological and structural investigations. Further decisions will be discussed in our weekly MDT meeting and a bespoke management plan will be formulated.

Results

Pilot data collected from August 2016 to December 2016 (4 months) shows that 71 patients had screening questionnaires, 48 patients were discussed in MDT and 24 have had investigations with 24 patients awaiting investigation. Mean age was 9 years; range 0-3 years (n=2); 4-7 (n=7); 8-12 (n=8) and 13-16 (n=6). Awake HRAM offers more information than done under general anaesthetic. Input from play specialists has allowed all patients selected to undergo awake HRAM. A play specialist was involved in 39% (7/18) patients who underwent HRAM. All patients had abnormal bowel function scores. Most patients had abnormal psychosocial scores.



All physiological and structural data obtained was regarded as useful in managing patients. A quarter of patients are now receiving psychological input

Summary and conclusion:

- Awake HRAM provides superior physiological data.
- Awake physiology can be achieved with careful and skilled play specialist input.
- A quarter of patients are receiving additional psychological input.
- Physiological and structural information and MDT was useful in all patients.
- Preliminary data supports the benefit of the service and seek to offer the service to patients outside our centre.

T15 Ischaemic Pneumatosis Coli following Head trauma

Dr Raj Singh Parmar; Dr Sarang Tamhne Alder Hey Children's Hospital, Liverpool)

Background

Ischaemic colitis (IC), first described by Boley et al, is the most common form of ischaemic injury to the gastrointestinal tract representing more than half of the cases with gastrointestinal ischaemia. The incidence of IC is underestimated because it often has a mild and transient nature. It is fundamentally characterized by circulatory insufficiency of the colon, resulting in varying degrees of local tissue necrosis and systemic manifestations. Ischaemic colitis can be broadly classified into non-gangrenous or gangrenous. We would like to report a very rare case of ischaemic pneumatosis coli seen in a paediatric patient following a severe head injury.

Subjects and Methods

A 15 year old previously fit and well boy presented following a road traffic accident having fallen off a quad bike. He suffered fractures to his skull with subdural hematoma and fracture of left arm. He required prolonged stay in Paediatric intensive care unit including need for multiple inotropes to manage his systemic hypotension but was fortunate to escape acute neurosurgical intervention.

4 weeks later, while recovering patient was commenced on pureed enteral feed following assessment by SALT team. Few days after introduction of enteral feeds patient developed an episode of acute vomiting, abdominal distension and diarrhoea. Abdominal radiograph showed multiple bowel loops filled with air and the most prominent feature being evidence of pneumatosis coli with air seen in the wall of the ascending colon and the caecum. Bloods showed mildly raised inflammatory markers.

Discussions with our Surgical team led to some dilemma for any invasive diagnostic or therapeutic surgical interventions with no evidence base to guide management.

Following discussions with the patient and his family, we took a decision to manage him conservatively with emphasis on bowel rest. He was treated with iv fluids and intravenous antibiotics (ciprofloxacin and metronidazole) and in view of poor nutrition and unresolved symptoms, he also received period of parenteral nutrition. Patient continued to show signs of recovery and a repeat X rays done 2 and 4 weeks later showed partial and complete resolution of intramural gas. Stool cultures were negative for Clostridium difficile, Yersinia, Salmonella, Shigella, Camplylobacter, E Coli 0157 and rota virus.

Results

Ischaemic colitis with features of pneumatosis coli managed conservatively.

Summary and conclusion

On extensive literature search, we could not find any reported case of ischaemic pneumatosis colitis in paediatric age group following traumatic brain injury. We hypothesise that either severe head injury per se led to neuro vascular dysregulation or the systemic hypotension led to bowel hypoperfusion which contributed to bowel wall ischemia causing pneumatosis coli and symptoms of feed intolerance and acute bowel obstruction. This unique case highlights and reiterates the usefulness of simple abdominal radiograph in cases of suspected bowel obstruction and increases awareness into possibility of ischaemic pneumatosis colitis following significant trauma.

T16 Complicated appendicitis is associated with very high CRP levels in children but not IBDs

Dr Siba Paul, Yeovil District Hospital; Miss Joanna Barnden, University of Bristol; Dr Megan Eaton, Yeovil District Hospital

Introduction

NICE guidance for managing children with fevers recommends that C-reactive protein (CRP) is measured in all children with red flag features at presentation or with an unknown focus for their infection. RCPath (UK) highlighted that a CRP>300mg/L should be a red flag for laboratory staff1. Two previous studies have demonstrated that CRP>300mg/L to be associated with significant pathologies such as pneumonia with pleural effusion, complicated appendicitis, sepsis often with atypical organisms but not with IBD flare-ups or toxic megacolon1,2.

Aims

This study of CRP>300mg/L is aimed at identifying underlying pathologies and their outcomes in a small DGH setup.

Methods

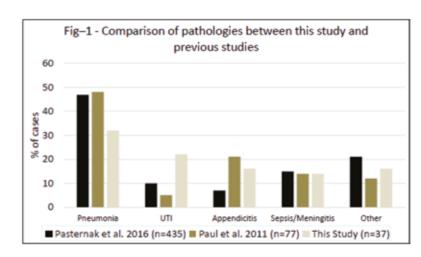
Patients aged <17-years with CRP>300mg/L were identified from the electronic pathology records between January'2011 and July'2016. Retrospective review of case notes and relevant electronic records were undertaken. Final diagnoses and outcomes were recorded.

Resul

37 patients identified. Three main pathologies were: pneumonia (n=12), appendicitis (n=6), and UTI (n=8). 27% patients had normal temperature (≤37.4½C) at initial presentation. Analysis showed 4/12 children with pneumonia had associated pleural effusions, all 6 patients with appendicitis had associated gangrene/perforation. No IBD associated pathologies were identified. No deaths were recorded in the study group. Fig-1 shows comparison between this and the 2 previous studies1,2.

Conclusions

- CRP>300mg/L in children is most likely to be associated with serious pathologies.
- Complicated pneumonia and appendicitis are most commonly associated with CRP>300mg/L, similar to that noted in previous studies1,2.
- Consider other pathologies including appendicitis in children with known IBD.



References

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T17 Comparison of ileal intubation rates and diagnostic yields in ileocolonoscopy: A multicentre, retrospective cohort study

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Background

Terminal ileum intubation (TII) rate at ileo-colonosocopy has been gaining increasing importance, and indeed now constitutes a pivotal mandate in the revised Porto Criteria to improve IBD diagnosis. It also has a critical role in contributing to the diagnostic yield of ileo-colonoscopy, both by its positive and negative findings.

Aim

Several service evaluation studies have looked at TII rates and diagnostic yields in individual centres, and the factors influencing the same. In our study, we have compared this data amongst four major paediatric gastroenterology centres in the UK, in order to identify practice variations and local factors that may influence these outcomes, with a view to using this knowledge to further improve practice.

Subjects and Methods

A service evaluation study was originally conducted in Sheffield Children's Hospital, UK, where data was collected randomly in 147 patients to evaluate the diagnostic yield of ileocolonoscopy in children. Subsequently, further data was collected as per an agreed proforma across three other paediatric gastroenterology training units in the UK – Bristol, Chelsea & Westminster (London) and Kings College Hospital (London). 50 consecutive cases were selected from each of these 3 centres and data collected retrospectively for demographic details, indication for ileocolonoscopy, macroscopic and histopathological findings, ileal intubation rates and scope details.

Results

The data was compiled and analysed on excel sheet and TII rates and other parameters, including normal findings, were evaluated. While the arithmetic mean was used as the measure of central tendency, the diagnostic yield was calculated from the Sheffield data and the sensitivities, specificities, positive predictive values(PPV) and negative predictive values(NPV) from all the other centres were calculated for comparison.

Summary and conclusion

Our data reveals that TII rates and positive diagnostic yields are similar across the four UK centres in the cohorts that were studied. This evaluation suggests that full paediatric ileo-colonoscopy in UK tertiary centres seem to be adhering well to established guidelines, with an overall high ileal intubation rate across all centres, without any obvious impact of the number of procedures done per list. Appropriate selection of cases is paramount, as is small bowel imaging in appropriate settings in order to further improve the diagnostic yield.

The average number of normal colonoscopy was also comparable among the various centres

T18 Abdominal Ultrasound Scanning in Acute Abdominal Pain: a Retrospective Service Evaluation

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Background

Acute abdominal pain (AAP) is a common presenting complaint in paediatrics. The role of abdominal ultrasound scan for AAP is debatable; it is known that most causes of AAP can be diagnosed on history and examination alone. It was considered useful to evaluate departmental use of abdominal ultrasound scans for AAP work-up, and if possible to answer the following question: "What is the value of an abdominal ultrasound scan in the management of children presenting with AAP?"

Aim

This project was intended to help clarify when abdominal ultrasound scans may be useful in children presenting with AAP.

Specific aims were to identify any key features in clinical cases that would indicate that an abdominal scan would be useful, or conversely, if any features indicated when it would not be of benefit, with a view to improving efficiency in the use of radiology resources.

Subjects and Methods

We looked retrospectively at all cases of children attending the Children's Assessment Unit, Princess Royal Hospital, Telford, with AAP, between February 22nd and May 16th 2016.

Cases were identified from the admissions diary as referrals with 'abdominal pain'. Electronic resources were used to gather data according to a proforma. Information gathered included patient's age, gender, investigations done, and final diagnosis made.

A pilot study of 10 cases was undertaken to refine the proforma, before further data was collected.

Data was uploaded onto an Excel spreadsheet for analysis.

Results

A total of 103 cases were identified. These comprised of 49 boys and 54 girls. The age range was 1 to 15 years, (median age 10 years.)

17% of all cases (n=18) had a scan.

- 32% of children ≥11 years had a scan, versus 8.5% of those younger than this.
- Only 10% of boys had a scan vs 24% girls.
- Of all the scans done, 72% were done in girls.
- Only 3 out of 18 scans were abnormal (16%). One showed features of unclear significance (later the patient was diagnosed with Lymphoma). Another showed unrelated pathology already known to exist in the patient. The third showed changes associated with underlying gynaecological pathology.

In terms of the discharge diagnosis, scans were most commonly requested when a diagnosis of non-specific or idiopathic abdominal pain was ultimately made. No cases of appendicitis were associated with an abdominal ultrasound scan request.

Summary and conclusion

- Abdominal ultrasound scans are rarely abnormal in acute abdominal pain.
- Scans may be helpful to reach diagnoses of exclusion.
- Abdominal ultrasound scans may be more useful in older girls where gynaecological pathology may exist.
- Appendicitis can be confidently diagnosed without the use of a scan.

T19 Patient and family experience of Endoscopy at a tertiary Paediatric Gastroenterology unit

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Background

A paediatric global ratings scale for endoscopy (P-GRS) is currently being piloted and this will provide a quality and safety framework for service improvement in Paediatric endoscopy units. An important aspect of this is patient involvement and an annual survey on the patient's experience with gastrointestinal endoscopy.

Aim

A patient/parent feedback survey was used to evaluate the endoscopy experience for our patients and family, as part of an annual endoscopy audit plan.

Subjects and Methods

A patient/parent questionnaire that has previously been approved by PALS and our clinical governance team in 2013 was used for this study. Questionnaires were distributed to patients and parents over a 3 week period (24/10/16-11/11/16). Data was then collated and analysed on Microsoft Excel.

Results

28 questionnaires were returned, including an even spread between age groups. The results are illustrated in Table 1 (below).

Table 1: Results of patient/parent questionnaire

Preparation before procedure		No (%)	Not recorded (%)
Was the procedure explained during consent?	100	0	0
Did you feel you had opportunity to ask questions?	100	0	0
Were you given information leaflets about the procedure?	75	18	7
In those who had colonoscopies, were you explained the importance of bowel preparation?	100	0	0
Were you informed of waiting time in advance?	71	25	4
Did you have an opportunity to discuss options with the Anaesthetist?	82	11	7
Overall preparation rated as "excellent" or "good"	79	14	7
Experience post procedure			
Did the patient experience post-operative pain?	29	61	10
Did the patient experience post-operative bleeding?	4	86	10
Did the patient experience post-operative vomiting?	4	86	10
Were the endoscopy findings discussed and explained?	71	11	17
Were follow up arrangements given at discharge?	71	4	25
Was advice given about complications after discharge?	46	7	47
Overall experience			
Overall patient comfort rated as "excellent" or "good"	75	7	18
Overall experience rated as "excellent" or "good"	75	7	18

Summary and conclusion

Overall, patients and families have had a good experience of endoscopy at our Unit, which is in line with previous studies. Areas for improvement include a need for specific endoscopy information and post procedure advice leaflets, and adolescent care.

T20 - Evaluation of the Liver Transition Services for Young People at a UK Paediatric Hospital

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Background

An effective transition process should prepare, equip and support young people in a holistic manner as they make the change from paediatric to adult services to prevent them becoming 'lost' and disengaged with health services (Department of Health, 2008).

The Care Quality Commission (2014) report reveals that many young people are not experiencing coherent transitional care to adult services which is having a negative impact on their long term health. Local services should involve young people in service evaluation, planning and delivery (NICE, 2016).

The nurse led Young Person's and Transition Clinics aim to prepare liver patients and their families for transfer to adult services. The transition process begins at age twelve with transfer around age sixteen. By addressing the current service limitations it is hoped that more tailored, robust transition services can be developed to benefit future patients.

Δim

To involve service users in identifying the strengths, limitations and potential methods of improving a hospital's liver transition service.

Methods

Seventeen young people and parents participated in service user evaluation questionnaires and interviews. These explored the strengths and limitations of the current transition service and identified areas for improvement. Data from the questionnaires was collated and presented in Likert scales to identify trends. Interview transcripts were analysed using thematic analysis.

Subjects

A predefined inclusion/exclusion criterion was used to identify participants for the study. This included patients with chronic liver conditions who had been through the Young Person's Clinics and at least one Transition Clinic. Using this criteria twelve patients and families were eligible to participate. Some of the participants were about to transfer to adult services while others had already done so. Unlike other published studies looking at the transition experiences of young people with liver conditions, only one participant in this study had received a liver transplant. All of the remaining young people in the study had chronic liver conditions but did not require transplant. These conditions included Autoimmune hepatitis, Allagile's syndrome, Non-alcoholic fatty liver disease, portal vein thrombosis, portal hypertension and TPN related liver disease.

Results

Overall patients and parents were satisfied with the current service. Suggested areas for improvement included a guided tour of the adult hospital prior to transition to help families feel more at ease in their new clinic environment. Feeling more confident about being seen independently in consultations is a factor that many young people strongly associate with being ready to transfer to adult services (Van Staa et al, 2011). Only 20% of young people felt confident about seeing the consultant independently in clinic. Families wanted their paediatric consultant to attend the adult hospital transition clinic. Families also suggested use of a transition website.

Summary

This was a small scale study therefore results may not be transferable to other centres or specialities. However the number of participants involved in this study was akin to other similar studies and were representative of the service being evaluated. The results are in keeping with those of similar studies across a variety of specialities both nationally and internationally. This study may also be the first to evaluate the transition experiences of non-transplant patients with chronic liver conditions.

Conclusions

This study has provided valuable insight into the transition needs of liver patients and their families. As a result service improvements have been implemented. A tour of the adult hospital prior to transfer has been introduced. Young people are now invited to see their paediatric consultant independently for part of their paediatric clinic consultation. A hospital transition website has been set up and we are hoping to facilitate paediatric consultant presence at the adult transition clinic. Once implemented these changes will be re-evaluated for effectiveness.

BSPGHAN 2017 Annual Meeting

POSTERSFRIDAY 27TH JANUARY 2017

F1 Eosinophilic Oesophagitis Management and Outcomes in a regional centre- Three Years On

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Background

Eosinophilic Oesophagitis (EO) is a chronic immune/ antigen-mediated disease with clearly evidenced links to atopy and allergy. Treatment options remain limited, with reliance on long term medication or food restricted diets. Three years ago we presented clinical presentations, diagnosis, treatment and outcomes of our cohort of patients diagnosed with EO between 2008 and 2013 from our regional paediatric population.

Aim

Here we investigate longer term outcome of these patients and summarise their management and outcomes after 3 further years of follow up.

Subjects and Methods

All patients diagnosed with EO in from our 2013 study were included (n = 30). Electronic patient records were available for all patients. A Clinical Research Form was developed and data on treatment and outcomes was collected and analysed. Emphasis was on: type and number of treatment modalities, clinical and endoscopic response to treatment, adherence to therapy, and follow-up.

Results

23 patients still met diagnostic criteria for EO. In 7 an alternative diagnosis was made: 2: progression to eosinophilic gastroenteritis; 3: reclassified as GORD; 1: Crohn's disease and 1: ulcerative colitis. 10 patients had trialled 2 treatments, and two patients 3 treatments prior to symptom improvement. Clinical response to treatment was present in 18/23 cases. The average number of endoscopies undergone in this time period was 1.63 (range 1-5). Endoscopic correlation with clinical response to last recorded treatment was available in 12 patients. Of these, 8/12 (75%) had a successful or partially successful response to treatment (where success is defined as asymptomatic + 0 eosinophils; partial success 0-15 eosinophils; and failure > 15 eosinophils regardless of clinical response). Successful treatment was seen with PPI alone in 2 cases, dietary change in 5 cases, and swallowed steroids (SS) in 2 cases. Adherence issues were found in 12/23 (52%) patients, occurring with all treatment modalities, 8 with diet, and 6 with PPI or SS. As of May 2016, 7 of the patients were undergoing active management within the paediatric service, 10 had been transitioned to adult services, four were lost to follow-up, and two were discharged. Four new allergies were identified between the initial data collection period and the latter, those being to barley, banana, milk and poppy seeds.

Summary and conclusion

Patients in this study tend to have good clinical outcomes, but over half had more than one treatment attempt and many had several endoscopies. Endoscopic correlation with outcome was not always available and adherence problems were high with all treatment modalities. Overall, the results highlight potential challenges in the management of EO. We recommend strategies be developed for improving concordance, including written patient and parent information leaflets, and use of symptom and Quality of Life scores. In recognition of this we have now established and are running a monthly EO clinic to harmonise investigative and management approaches

F2 Paediatric IBD patients do not meet the daily recommendations of Vitamin D and calcium intake: survey based analysis in a tertiary centre

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Background

Achieving optimal levels of vitamin D (VitD) and calcium (Ca) is essential for developing children, especially in patients with inflammatory bowel disease (IBD). VitD and Ca play a major role in bone health and recently VitD has shown to potentiate the effect of anti-inflammatory treatments. However, achieving a sufficient oral intake is difficult in this group taking into account young age, modern eating habits and the nature of IBD itself. The purpose of this study was to evaluate if children with IBD seen in our centre achieve optimal Vit D and Ca intake according to recommendations made by the British Scientific Advisory Committee on Nutrition and the UK Department of Health, Dietary Reference Values.

Methods

A prospective dietetic survey was conducted among sequential IBD children seen in clinics over a 12 month period. Ca and VitD intakes were assessed through a 24-hour recall of dietary intake and food frequency questionnaire. Children who had been placed on restricted diets for allergic disease were excluded as well as children under 4 years. Included patients were classified according to age into 2 groups: 4-10 and 11-18 years. Sources of VitD were divided into dairy, oily fish, fortified cereals and egg. Analysis was performed using absolute values, percentages and means in Microsoft Excel.

Results

Survey was conducted in 151 patients; this represents 68.3% of all IBD patients under follow-up. 94 patients were included for analysis and 57 were excluded. 43/94 (45.7%) were females. Overall, only 26.6% and 21.3% of the surveyed population achieved the current recommended intake for Ca and VitD respectively. In the younger group, only 7/31 (22.6%) met the current VitD recommendations, the same figure repeats with regards Ca intake. In the older group, only 13/63 (20.6%) and 18/63 (28.6%) met the Ca and VitD recommendations respectively. In both groups dairy was the main source of vitamin D (61.3% young ones and 58.7% older ones). Less than 1/3 of the patients have an optimal intake of oily fish (intake 19% for children and 30% for adolescents).

Summary

3.4% (age 4-10yrs) and 78.7 %(age 11-18yrs) of the population surveyed did not achieved the current recommended intake amounts for Ca and VitD respectively.

Conclusions

Paediatric IBD patients living in the UK do not meet the minimum requirements of VitD and Ca intake and therefore are at risk of having poor bone health, calcium homeostasis imbalance and VitD deficiency. In the great majority, Ca and VitD sources come from diary whereas the contribution of oily fish and egg as a VitD source is minimal. We recommend that paediatric IBD patients receive frequent counselling on healthy eating habits and proactive intake monitoring. Routine VitD supplementation recommended by local authorities must be followed as there is an insufficient VitD taken orally among these populations.

F3 Faecal Short Chain Fatty Acids in Untreated and Treated Children with Coeliac Disease

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Background

Recent literature indicates that the gut microbiota and its metabolic activity might be altered in children with coeliac disease (CD). However, the role of the gut microbiota in CD is unclear and the results among studies remain inconclusive. SCFAs are produced from bacterial fermentation of the fibre. They are important for the whole body metabolism and mediate immune response.

Aim

The aim of this study was to measure the faecal concentration of SCFA in treated and untreated children with CD and compared them with healthy controls.

Subjects and Methods

Faecal samples were collected from CD children on GFD for more than a year (Long Standing patients - LS); from newly diagnosed untreated CD children (ND), and following treatment with GFD for 6 and 12 months and from healthy controls (HC). SCFAs were measured using Gas Chromatography (GC). The data were analyzed using Mann – Whitney and Kruskal - Wallis tests and expressed as medians (Q1–Q3).

Results

Faecal samples were collected from 45 LS, 27 ND and 57 HC children. Among the 27 ND children, 13 adhered on GFD and gave us samples 6 and 12 months later. The relative abundance of acetate, isobutyrate, butyrate, isovaleric and valeric acids was significantly different between the LS, ND and HC (p<0.05). Regarding the absolute concentrations per wet matter it was found that only isovaleric and valeric acids were significantly different between the LS, ND and HC (p=0.041 and p=0.036, respectively). Specifically, isovaleric acid (µmol/gr wet sample) was significantly decreased in the group of LS (2.70 (1.75 - 3.65)) compared to ND (3.74 (2.60 - 4.68)) and HC (3.42 (2.57 - 4.56)), (p=0.040 and p=0.024, respectively), whereas valeric acid (µmol/gr wet sample) was significantly decreased in the LS (2.22 (1.55 - 3.05)) only opposed to the HC (2.85 (2.13 - 3.86)), (p=0.010). The relative abundance of acetate, isobutyrate, butyrate and isovaleric acid was significantly different during the 12 months period of treatment with GFD (p<0.05). Only acetate acid (%) was significantly increased 6 months after the adherence on GFD (74.01 (67.82-76.95)) compared to the ND on Gluten Containing Diet (GCD) (65.09 (61.27-68.65)), (p=0.007), while isobutyrate, butyrate and isovaleric were significantly decreased 6 months after the adherence on GFD compared to ND on GCD (p=0.046, p=0.028 and p=0.046 respectively). The relative abundance of isobutyrate and isovaleric acids was still significantly decreased 12 months after the treatment with GFD (p=0.039 and p=0.033, respectively).

Summary and conclusion

In this study differences concerning the relative abundance and absolute concentration of SCFAs between the HC and the ND were not observed. However, differences among the LS and the ND or the LS and the HC were found. Furthermore, differences on the relative abundance of certain SCFAs were obvious 6 and 12 months after the adherence on GFD. Thus, it may be the GFD responsible for the altered composition of the produced SCFAs from the gut microbiota of children with CD and not the factor of the CD itself. Although these differences were significant in our results, further research is needed to conclude to such a conclusion, since the first part of our study is cross – sectional designed and we cannot assume causality based on that.

F4 Feed choice when initiating jejunal feeding in children

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Introduction

There is limited current evidence regarding optimal feed choice when initiating jejunal feeding in paediatrics. Concerns include concentration, osmalilty and the protein base of feeds which may affect tolerance. Most recent evidence suggests peptide based feed is best tolerated however current practice varies.

Aim

To review feed type and enteral tolerance when initiating jejunal feeding in our centre.

Method

Patients who commenced jejunal feeding between April 2015 and November 2016 were identified from a prospectively recorded database. The choice of feed commenced, any changes made and clinical outcomes were retrospectively reviewed from electronic records.

Results

18 patients (12 male, median age 2.9yrs, range 0.4-15.8yrs) were identified. Prior to jejunal feeding 13/18 had a gastrostomy in situ the remaining 5/18 had nasogastric tube. Jejunal feeding was employed for foregut dysmotility associated with neurological impairment in 10 patients, isolated forgeut dysmotility in 6 patients and in 2 patients with significant cardiac anomalies where gastrooesophageal reflux (GORD) was a concern. 15/18 had PEGJ for jejunal feeding with the remaining 3/18 having NJT. 12/18 (66%) were already established on a whole protein feed prior to jejunal feeding. Of this group, 5/12 (42%) were established on 1kcal/ml feed and the remaining 7/12 (68%) on 1.5kcal/ml feed. 5/18 were established on peptide based feed and 1/18 on elemental feed whilst being feed gastrically. Once jejunal feeding was established 8/18 (44%) of patients tolerated a whole protein feed, 62% (5/8) of these tolerated a 1kcal/ml feed, but tolerance of 1.5kcal/ml feed reduced to 38% (3/8). 3/18 required a feed change to peptide based feed due to tolerance issues which included pain and loose stools. 1/18 remained on an elemental feed. The patient group most likely to require a change to peptide feeds were those with isolated foregut dysmotility concerns (table 1).

Table 1: Peptide feeds per patient sub group

Diagnosis	Numbers	% Requiring Peptide Feeds	
		Before jejunal tube	After jejunal tube
Complex neurodevelopment	10	30%	40%
Isolated GI dysmotility	6	17%	67%
Cardiac with GORD	2	50%	50%
All Patients	18	28%	50%

Conclusion

The majority of patients in our study tolerated their established feeding formula following initiation of jejunal feeding. Patients with GI motility concerns were the group most likely to require a change to peptide feeds. Following this audit the clinical practice within the unit is to recommence patients on their pre-existing feed when starting jejunal feeding. We would consider a change to peptide based feed only if there were clinical concerns regarding tolerance.

F5 MDT led jejunal feeding. Nurse and dietetic led pathways in the development of a MDT complex enteral feeding service

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Background

Pathways for the management of jejunally fed patients are often variable between medical and surgical sub-specialities due to the small numbers of specialist staff appropriately experienced to make management decisions, resulting in delays in treatment for patients who require care unexpectedly or out of hours.

Aim

We describe the development of our nurse and dietetic led pathways which have improved our service by standardising care and creating a trouble-shooting guide for jejunally fed patients within our regional service.

Subjects and Methods

A comparison of the service structure and pathways prior to, and after, appointment of a clinical nurse specialist into the complex enteral feeding service (August 2013), and the development of a multi-disciplinary complex enteral feeding clinic.

Results

Pre August 2013, patients for jejunal feeding or fundoplication were seen in isolation by either paediatric gastroenterology or paediatric surgical services from a variable referral pattern. Patients were seen in OPD a median of >2 occasions, with investigations coordinated in-between assessments, the decision for jejunal feeding was made in isolation by single clinician and feeding regimen, selected device (jejunal extension, GJ button), insertion method (endoscopic, radiological) feed escalation, length of stay and follow up all varied amongst clinicians. Responsibilities for unplanned tube troubleshooting were not clear. Key documents and nurse/MDT developed pathways are presented in table.

Document	Description	Resulting systemic change
Nurse-led clinic proforma (2014)	Pre-clinic patient evaluation, Rx, investigation pathway	Nurse led history gives greater relevant clinical detail, complex investigations and medical manipulations all performed prior to single MDT clinic visit where decision on jejunal feeding made
Jejunal tube placement flow chart (2014)	Agreed pathway for decision on tube type and method placement of tubes	Standardisation of tube use, ability to make tube replacement decision in absence of primary clinician, improved ability to optimise stock use and nurse referral to radiology
Jejunal pump feeding competency (2015)	Jejunal feeding traning pack	Standardises training and assessment of jejunal competency of families prior to discharge
Jejunal tube trouble-shooter (2016)	Actions for non specialist staff for blocked/displaced tubes and contacts	Increased jejunal tube awareness and competence in non-specialist staff, patient specific information on clear referral pathway onto specialist staff
Jejunal tube de-escalation plan (2016)	Patient specific pathway for unplanned blockage or displacement	Patient specific information with reference to short term tolerance of gastric clear fluids or feeds and urgency of tube replacement reduces unnecessary use of out of hours specialist services and allows patient discharge prior to jejunal tube replacement
Jejunal starter feed pack(2016)	Dietetic feed plan for new jejunal feeder	Complete feed escalation for new jejunal tubes reduces specialist input prior to discharge

Summary and conclusion

We present our nurse and dietetic led pathways which have developed, streamlined and standardised care, with greater clarity and ease of access to out of hours care. To our knowledge, this is the only complete package of such MDT pathways to be presented in the UK for such patients.

F6 Blenderised Diet in Schools - A Risk Assessment Approach

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Background

Increasing numbers of families are opting to put blenderised food down their child's gastrostomy tube. NHS Grampian has no guideline on the administration of blenderised food via a gastrostomy tube. The British Dietetic Association does not recommend the administration of liquidised food via enteral feeding tubes. Care givers within education cannot administer blenderised food without written guidance and training. Some parents were insistent their child had blenderised food whilst at school and asked the Royal Aberdeen Children's Hospital Dietetic Department to facilitate this.

Ain

We opted to produce a formal risk assessment using the NHS Grampian Risk Assessment matrix. This was to meet requirements of education, ensure safe practice for the child and to protect the managing Dietitian professionally.

Subjects and Methods

The subjects were gastrostomy fed children starting school. The risk assessment was designed using information from the Dietetic Department, Royal Hospital for Sick Children, Edinburgh and The British Dietetic Association Practice Toolkit*. It incorporated information from the NHS Risk Assessment matrix.

Areas covered in the risk assessment include:

- potential hazards to the patient e.g. tube blockage, food hygiene risks
- feeding plan agreed by parents, education, Dietitian and Nutrition Nurse
- care givers all to receive standard gastrostomy training
- parent to give a bolus feed in school in presence of Nutrition Nurse
- all stakeholders sign off the agreed risk assessment.

Results

A risk assessment has been successfully produced and used within two schools. The risk matrix combines the potential consequence and likelihood for any given risk e.g. a blocked extension set is a minor outcome which is unlikely to occur giving a medium risk. All risks scored low or medium. Parents took overall responsibility for the blenderised food and accepted responsibility for any identified risk.

Summary and conclusion

Facilitating families to enable their children to receive blenderised food via gastrostomy in schools is challenging. The risk assessment has been successful in supporting the patient, parents, care givers and the Dietitian. It would be adaptable for use in other settings e.g. respite care.

References

* The British Dietetic Association Practice Toolkit Liquidised foods via Gastrostomy tube, May 2015.

F7 Changes over time in patterns of prescribing for symptoms of gastro-oesophageal reflux in infants in Scotland.

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Background

Gastro-oesophageal reflux (GOR), a normal physiological condition, is a common concern among parents of young infants. Although symptoms of GOR resolve spontaneously over time, observations from practice and anecdotal evidence from discussions with practitioners suggest that in Scotland the number of infants presented with, or prescribed medicine for symptoms of GOR has increased over the years. More robust evidence from the United States (US), indicates that there has been an increase in prescribing for symptoms of GOR in infants in the US over the last 15 – 20 years (Hassell, 2012), whilst recent evidence from the UK also reveals that the prescribing of alginate (Gaviscon) for GOR cost NHS England £5.2 million in 2015 and is likely to increase (Jones, 2016). ISD Scotland data reveals a similar growth in prescribing patterns for medicines used to manage symptoms of GOR in infants aged 0-12months in Scotland. Given that the evidence regarding the efficacy of these medicines is equivocal, this is cause for concern.

Aim

- 1. To determine patterns of prescribing for GOR in infants aged 0-12 months in Scotland.
- 2. To assess changes over time in the management strategies used to address GOR in infants.
- 3. To explore what underpins these changes.

Subjects and Methods

This is a two-stage mixed methods study as follows:

Phase 1: National prescribing data for the key medicines used to manage symptoms of GOR in infants aged 0-12 months, was obtained from the Information Services Division (ISD) of NHS Scotland between the years 2009 – 2016. These data were analysed using Minitab to determine changes in the patterns of prescribing for alginate (Gaviscon), motility stimulants (Domperidone), protein pump inhibitors (Omeprazole) and H2-receptor antagonists (Ranitidine) at a National, and NHS Board level over time.

Phase 2: Phase 2 of the research study involves semi-structured interviews with health visitors, general practitioners, and parents of infants who experienced symptoms of gastro-oesophageal reflux in order to gain an understanding of what underpins these changes in prescribing patterns in the management of GOR.

Result

Phase 1: Secondary analysis of the prescribing data from ISD Scotland indicates that in Scotland between 2009 and 2016, the prescribing rates of Gaviscon increased from 15.7% to 24.7%, Ranitidine from 2.3% to 9.7%, and Omeprazole from 0.9% to 3.2%. Marked regional variation in prescribing patterns were also observed.

Phase 2: Preliminary thematic analysis of the data obtained from the semi-structured interviews with health visitors suggest that these changes in patterns of prescribing may be influenced by factors such as culture, the internet and social media, and higher expectations of parenthood.

Summary and conclusion

The upward trend in the prescribing of Gaviscon, Ranitidine and Omeprazole in Scotland and the marked regional variation in prescribing are a cause for concern given the uncertainty regarding the efficacy of these medicines. It may also have financial implications for NHS Boards during this period of austerity.

Peference

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F8 Genital Crohn's disease case series - natural history

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Background

Crohn's Disease (CD) is an inflammatory bowel disease with an increasing prevalence and a reported incidence of 4.75 - 5.85 per 100000 in the UK (1). Extra intestinal manifestations of CD are present in 25-40% of cases (2). Genital CD lesions are rare in childhood with a few reported cases in literature. The management of genital CD is challenging.(4)

Δim

Our aims are to review the natural history of genital CD symptoms and its management.

Subjects and Methods

It is a retrospective case note review of the patients with genital Crohn's disease attending our tertiary Paediatric Gastroenterology Service.

We analysed the medical notes of patients with genital CD. We also collected data on inflammatory markers, histology, radiological investigations and CD phenotype (PARIS classification). The clinical response to various treatment modalities on luminal CD and genital CD was noted. All data was collated on an Excel spreadsheet.

Results

Our cohort included 5 patients over last 15 years. The male: female ratio was 3:2.

Genital CD onset ranged from age 18months to age 10 years 3months. Interestingly 4 patients had genital involvement several months to years before the onset of gastrointestinal symptoms. Genital CD symptoms consisted of: scrotal oedema and erythema (3 patients), penile swelling (2 patients) with deviation of penis (1 patient) and restricted micturition (1 patient) for males and labial non-tender, erythematous oedema (2 patients) and perineal induration and inflammation (1 patient) for females.

Luminal CD onset ranged from age 6years to 11years 9months. Onset of luminal CD consisted of: diarrhoea (2 patients), bleeding per rectum (3 patients), abdominal pain (4 patients), decreased appetite (2 patients), weight loss (1 patient), lethargy (1 patient), recurrent mouth ulcers (1 patient). All had associated perianal disease of varied severity with skin tags most common (4 patients), but also perianal fissures (2 patients) and perianal fistula and abscess (1 patient). None of our patients had fistulating genital disease, however one had fistulating disease of the sigmoid.

Other extra intestinal manifestations consisted of: seronegative arthropathy (1 patient), psoriasiform dermatitis (1 patient), hepatic cysts (1 patient), granulomatous lung disease (1 patient), delayed puberty requiring testosterone injections (1 patient). Two patients developed significant psychological problems leading to deliberate self-harm episodes and depression.

- 1. Treatment for genital lesions included local and systemic agents. Local treatment used consisted of: topical calcineurin inhibitors, topical corticosteroids, intralesional steroid injections and compression underwear. Some minor clinical improvement of genital symptoms (mild decrease in labial erythema for 1 patient) was noted following topical calcineurin inhibitors and some major improvement (normal colour vulva and significantly less swelling noted for 2 patients) following intralesional steroids. However, these effects were not sustained once treatment with local agents was discontinued. Genital CD response to systemic treatment was seen when Infliximab (all 5 patients) and Adalimumab (only 1 patient) were used. On Anti-TNF treatment significant decrease in genital erythema and oedema was seen. Azathioprine and Methotrexate had no effect on genital CD lesions for any of our patients.
- 2. Luminal CD treatment included: liquid diet (1 patient), oral steroids (3 patients), Azathioprine (4 patients), Methotrexate (1 patient), Infliximab (5 patients) and Adalimumab (2 patients). Luminal CD remission was achieved using: liquid diet (1 patient), oral steroids (3 patients), Infliximab (5 patients) and Adalimumab (2 patients). Azathioprine induced gastrointestinal remission for one patient, however escalation to biologic treatment was required within 6 months. Methotrexate induced gastrointestinal remission for 1 patient, however this effect lasted for 8 months only.

One patient required bowel surgery acutely and still has an ileostomy in situ.

Summary and conclusion

Genital CD is a rare, extra intestinal manifestation of CD. The onset of genital CD symptoms is early and has poor response to conventional CD treatment.

Most of our patients had their genital CD onset months to years before gastrointestinal CD onset. We didn't identify a significant increase of extra intestinal manifestations other than genital involvement for most of our patients. There was no clear correlation between flare ups of gastrointestinal disease and flare ups of genital CD lesions. Genital CD symptoms responded mostly to Anti-TNF treatment with some benefit from local treatment agents. Early escalation of treatment may be considered in CD patients presenting with genital symptoms.

F9 Cutaneous manifestations of paediatric Crohn's disease

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Background

Paediatric patients with Crohn's disease (CD) commonly present with dermatological disorders. Despite this strong association, the dermatological manifestations and their relationship to intestinal disease have not been extensively studied.

Aim

To document the clinical features of cutaneous lesions of five children with CD referred for a specialist dermatology opinion at a tertiary paediatric centre, and to review the literature for correlations between cutaneous disorders and CD.

Subjects and Methods

Medical records of five children referred to our department for cutaneous lesions associated with CD were reviewed retrospectively. Relevant publications on cutaneous disorders associated with CD identified from a PubMed search were reviewed.

Resul

Five children, 3 females and 2 males, age 10 to 17, presented with skin lesions associated with CD over a four year period. 2 patients had a single skin disorder, and 3 patients presented with two or more different types of skin lesions. The dermatological disorders seen were recurrent sterile pustular rashes, erythema nodosum (EN), oral and genital ulcers, pyoderma gangrenosum (PG), psoriasiform scalp inflammation and alopecia, an erythematous patch with telangiectasia, and a non-specific blotchy rash affecting the upper body in one patient, and the feet in another. Four patients had clinical or histological evidence of active intestinal disease at the onset of their skin lesions. For these patients, their skin disorders improved following treatment with systemic corticosteroids and immunomodulators for their intestinal CD. One patient was on treatment with infliximab with quiescent intestinal CD at the onset of development of psoriasiform scalp inflammation and alopecia. An improvement with good hair regrowth was observed following cessation of infliximab for an unrelated reason and treatment with a potent topical steroid.

Summary and Conclusion

A review of the literature shows that cutaneous disorders associated with inflammatory bowel disease are seen in up to 30% of children with CD, with erythema nodosum most frequently implicated. Cutaneous disorders can be classified into 5 groups: metastatic or cutaneous extension of intestinal CD, immunoreactive skin disorders, dermatoses associated with inflammatory bowel disease, skin manifestations secondary to malnutrition, and skin disorders related to drug therapy for CD. The diversity in cutaneous manifestations seen, mean that treatment needs to be individualised. Skin lesions which are an extension of intestinal disease (e.g. oral and genital ulcers) or immunoreactive in nature (e.g. EN and PG) often improve with treatment of the underlying intestinal disease, as seen in four of our patients, suggesting a common inflammatory pathway. Skin disorders secondary to immunosuppressive or biologic therapies, such as the psoriasiform rash induced by infliximab in one of our patients, may improve following cessation of the offending drug, although this will limit therapeutic options for CD for the patient.

F10 Inflammatory Bowel Disease In Children Is A Challenging Diagnosis When Overlaps With Coeliac Disease

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Background

Inflammatory bowel disease (IBD) and coeliac disease (CD) are conditions associated with chronic inflammation of the gastrointestinal tract. Underlying aetiology includes genetic susceptibility, abnormal immune response and various environmental factors not fully understood to date. Both entities can overlap in paediatric patients although this is uncommon and difficult to confirm. We present our experience with patients who developed both conditions during early years of life.

Methods

We retrospectively reviewed all IBD patients seen in our centre who also had a diagnosis of coeliac disease over a 10-year- period. Data were collected from electronic notes and laboratory registries. Demographics, consultations, laboratory and endoscopic findings were extracted to a STATA database. Descriptive analysis was performed using absolute value, percentage and mean functions trough STATA software version 14.

Results

Only 8/578 patients were found to have both diagnosis, this accounts for 1.4% of all paediatric IBD patients seen in our centre (mean 57 new patients per year, past 10 years). 5 of them were female, 1 male had Down syndrome, and 1 patient had incomplete records. Mean age of diagnosis was 7.1 and 8.9 years for CD and IBD respectively. In terms of the IBD subtype, 4 patients suffered of Crohn's disease, 3 of ulcerative colitis and 1 of IBD unclassified. 3 patients were diagnosed with both entities within 3 months, other 4 had a previous history of CD and developed IBD years later (mean in years 3.0), despite having a well-controlled disease. Positive anti-transglutaminase (TTG) serology was found in 4/7 patients. Endoscopic findings were difficult to interpret, complementary specific biopsy immunostaining, small bowel imaging (MRI, CT) and video capsule endoscopy were required in order to support both diagnoses. Endoscopic assessment when there was a previous diagnosis of CD obeyed to persistent gastrointestinal symptoms despite normal TTG values, these 4 known CD patients had significant IBD features including granulomata and cryptitis in small bowel (3/4) and pancolitis (1/4).

Conclusions

Inflammatory bowel disease can overlap with coeliac disease in paediatric IBD patients although this association is rare. IBD can follow the appearance of CD years later despite TTG normalization and can also present at the same time of CD. Proving the coexistence of IBD and CD in children is a challenge, and requires of a multidisciplinary team involving expert histopathologists, gastroenterologists, dieticians and clinical laboratory scientists. IBD must be considered in CD patients with new onset of gastrointestinal symptoms or in CD patients whose gastrointestinal symptoms do not seem to respond to a gluten-free diet.

F11 Border-line Coeliac Serology in Children - Outcome & Correlation with histology

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Background

Coeliac disease is an immune-mediated condition that affects the lining of the intestine when exposed to gluten. The British Society of Paediatric Gastroenterology, Hepatology and Nutrition guideline on Coeliac Disease (2013) recommends that patients with Tissue Transglutaminase antibodies (TTG) less than 10 times normal should undergo duodenal biopsy to confirm a diagnosis of Coeliac Disease. However, there is a paucity of data on how often this correlates with a final diagnosis of Coeliac disease.

Aim

To measure the sensitivity of borderline Tissue Transglutaminase Antibody (TTG) levels (greater than normal but less than 10 times of normal values) in diagnosing Coeliac disease.

Subjects and Methods

The Study Population was children aged 1-16 years with TTG levels less than 10 times of normal seen at The Shrewsbury & Telford Hospitals NHS Trust. The study period was from January 2010 to September 2016.

Patients were identified from the biochemistry database. Relevant data including the clinical symptoms, TTG levels, IgA levels, HLA status and histology results were collected using the hospital data base. The histology results were classified according to the Modified Marsh Criteria.

Results

A total of 52 patients had TTG levels between 2-19.9 U/ml which is less than ten times of the normal range used by our biochemistry laboratory. The median age of presentation was 10 years while the median TTG value was 4.2U/ml. 10 patients with Type 1 Diabetes Mellitus were detected to have border-line positive coeliac serology on routine testing.

Thirty-nine patients underwent endoscopy with duodenal biopsies. Ten patients had neither endoscopy nor HLA testing performed because of various reasons including parental choice, symptom resolution prior to endoscopy while still on a normal diet, or repeat TTG levels within the normal range.

29 children (56%) out of the cohort of 52 had histological changes consistent with coeliac disease and were diagnosed as such. Seven of these (24%) had Grade 1 Marsh criteria, 3 children (10%) had Grade 2 Marsh criteria and 19 (66%) had Grade 3 Marsh criteria.

There was no significant difference in symptomatology between the children who were diagnosed with coeliac disease and those with normal biopsies, apart from recurrent abdominal pain which was commoner in coeliac disease.

Summary and conclusion

56% of children with borderline TTG levels were diagnosed with coeliac disease based on biopsy changes. Symptomatology was of poor discriminatory value apart from recurrent abdominal pain which was commoner in coeliac disease. Children with border-line positive coeliac serology should have duodenal biopsies to confirm a diagnosis of coeliac disease.

F12 Chorioretinitis - an unusual presentation of Crohn's disease

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Background

Extraintestinal manifestations of Inflammatory Bowel Disease are well described and routinely include joint, skin and other systemic features. Ocular complications of IBD occur in around 10% of cases in adult series1, and may precede systemic symptoms and include uveitis, complications of therapy, such as cataracts, or glaucoma from steroid use.

Aim

Case report of an unusual manifestation in Early Onset IBD and to raise awareness of visual loss as a presenting feature of IBD.

Subjects and Methods

A 14 year old girl, with a background of Asperger's syndrome and on medication for Attention Deficit Hyperactivity Disorder (ADHD), presented with short history of gastrointestinal symptoms suggestive of IBD. Although macroscopic and microscopic features at endoscopic assessment suggested Crohn's disease, there were no granulomas present on any of the biopsies. Small bowel imaging was normal. Exclusive modulen was commenced on the day of endoscopy as an inpatient and she was discharged on Exclusive Enteral Nutrition (EEN). She re-presented to A&E a few days following discharge with worsening symptoms of IBD. In addition, she also complained of poor vision and specifically a reduced field of vision.

Results

In view of the non-response to EEN, she was commenced on IV steroids. As an inpatient, she was reviewed by the ophthalmologist who diagnosed severe right-sided choroiditis affecting the retina. The visual acuity in the right eye was 6/36 and in the left eye of 6/6. Serial eye examination has revealed persistence of diminished visual acuity despite improvement in the rest of the symptoms of IBD.

Summary and conclusion

Evaluation of the eye should form part of the evaluation in the care of children with IBD. While classical features of Uveitis are easy to recognise, posterior uveitis requires a degree of suspicion. Systematic inquiry for cases with IBD should include questions about visual acuity and the field of vision. Conversely, the presence of choroiditis may be a manifestation of IBD, symptoms of which in children may be difficult to recognise.

Reference

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F13 Clinical effectiveness of Faecal Calprotectin (FC) in children with suspected IBD – a single tertiary centre experience

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Background

NICE in October 2013 published "Faecal Calprotectin diagnostic tests for inflammatory diseases of the bowel" and made recommendations to use faecal Calprotectin as an option to help clinicians differentiate between IBD and IBS in children who have been referred for a specialist assessment if: appropriate quality assurance and care pathways are in place for testing.

Aim

To determine whether Faecal Calprotectin testing results in changes to treatment strategy

Subjects and Methods

A retrospective evaluation identified FC testing on total of 78 patients with 81 samples from May 2013 to May 2015. 20 patients were enrolled into study after the exclusion criteria. Exclusion criteria included - Endoscopy > 3-month interval between FC collection and endoscopy, previous ileo-colonoscopy, previous GI diagnosis (including IBD), FC for monitoring activity or response to treatment and if Colonoscopy preceded by > 2 weeks from FC collection.

Results

The median age in inflamed group was 5.08 (1.41-14.91) and 12.29 (3.16-17.00) in non-inflamed group. Median FC level in inflamed group was 187 (100-1500) and 56 (8-748) in non-inflamed group. All children had complete colonoscopy including terminal ileum. Data on endoscopies in the two groups and final diagnosis are presented in table. In 7/20 (35%) patients who had positive FC showed normal histology which was reassuring and helped in clinical management.

Inflamed (n=7)		Non-inflamed (n=13)			
FC 50-150 (n=1)	FC >150 (N=6)	FC<50 N=6	FC 50-150 N=4	FC>150 N=3	
Pancolitis - UC	Indeterminate colitis	Lactose intolerance, thickened ileum on MRI, IBS	Non coeliac gluten sensitivity, IDA	Spastic quadriplegic CP, exprem 24 weeks, cotical visual impairment, feeding difficulties	
	Crohn's disease	Familial short stature	IBS	Pinworm infestation	
	Crohn's disease	Significant uveitis necessating infliximab, family h/o Crohn's disease,	Hyper-eosinophilic gastroenteritis, multiple food intolerance, Primary Immunodeficiency	Post appendisectomy dysmotility with reflux, and constipation	
	Eosinophilic colitis	Constipation with over flow incontinence	HSP like illness, recurrent abdominal pain and rectal bleeding		
	Very early onset IBD	IBS, Thread worm infestation			
	Crohn's disease	Constipation with faecal incontinence			

Summary and conclusion

At the recommended cut-off (FC<50), FC excretion correlated well with histopathologic assessments of colonic inflammation. A negative test seems to indicate low probability of mucosal inflammation in colon and other diagnosis may be considered if child has vague symptoms. A positive test facilitates a decision to proceed with endoscopy and prioritization of urgent cases. In children with FC values between 50-150, and associated risk factors progress to endoscopy or with persistent symptoms to repeat in 3 months.

F14 Single centre Service Evaluation to determine appropriate use of faecal calprotectin testing in children

F14 Single centre Service Evaluation to determine appropriate use of faecal calprotectin testing in children

Background

Faecal calprotectin (FCP) has been used for several years as a non-invasive marker for intestinal inflammation. The normal values of FCP differ with different kits. Paediatric data has shown that high FCP levels seen in patients <2 years is not thought to be clinically significant. The current evidence suggests a cut-off value of 50, with higher FCP values indicative of inflammatory bowel disease (IBD).

Aim

There are currently no formal paediatric FCP request guidelines. We informally adopted the adult guidelines with amendments including age > 5 years with chronic abdominal pain and diarrhoea and annual assessment of IBD patients on anti-TNF/ immunosuppressants to continue or stop treatment. We evaluated FCP requests made in children in 2015 in a tertiary gastroenterology unit as per amended in-house adult FCP guidelines mentioned below:

- 1. Chronic abdominal pain with non-bloody diarrhoea and weight loss in patients aged >5 years.
- 2. Known IBD out-patient with diarrhoea
- 3. Annual assessment of patient with Crohn's disease or UC on an anti-TNF/ immunosuppressant with no symptoms and normal bloods (including CRP)
- 4. Known IBD with non-specific symptoms (eg fatigue, abdominal pain, anaemia) and normal CRP

Subjects and Methods

We retrospectively reviewed clinic letters, reasons for FCP requests and investigations (bloods, OGD, colonoscopy reports) of patients <18 years in whom FCP was requested over a 12-month period (Jan- Dec 2015).

Results

A total of 219 FCP requests were made. Nine patients were excluded as there was incomplete information available. The vast majority of FCP requests (90%) were made in outpatients with 56% made by the paediatric gastroenterology team. Requests were also made by general paediatricians (20%), GP's (20%) and other specialities (4%) – surgeons, oral health, rheumatology. The main indication for requesting FCP was chronic abdominal pain and diarrhoea in both the gastroenterology team and other specialities. Other indications for requesting FCP by the gastroenterology team included known IBD with possible flare and known IBD for assessment regarding weaning or stopping medication.

Appropriate FCP requests were made in 132 patients (63%) and inappropriate in 78 patients (37%). Thirty five percent (41/117) of the FCP requests made by the gastro team and 40% (37/93) requests by non-gastro teams were inappropriate.

The main reason for inappropriate requests in the gastroenterology team was ordering scope simultaneously with FCP (17 patients) and in the non-gastro teams was PR bleeding (18 patients). 48 patients (23%) underwent a scope and in 18 patients FCP and scope were simultaneously requested.

51 requests (24%) were ordered in patients known to have IBD. In this cohort 26 patients avoided colonoscopy; 18 patients were treated for a relapse and 8 for no relapse on the basis of symptoms, bloods and FCP result. In 4 IBD patients' treatment was either stopped or weaned.

Summary and conclusion

Our results show there is a need to develop formal paediatric FCP request guidelines in order to reduce the number of inappropriate requests and colonoscopies. A guideline will improve cost effectiveness and optimise the use of FCP.

F15 An unusual case of Heiner's syndrome in a five week old infant

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Introduction/Background

Heiner's syndrome (HS) is a rare, delayed, non- IgE mediated food hypersensitivity which primarily affects infants causing pulmonary symptoms and is mostly caused by an allergy to cow's milk. To the best of our knowledge, this is a case report of the youngest child who was diagnosed with Heiner's syndrome (1).

Aim

To report an unusual case of Heiner's syndrome in a previously fit and well five week old child who presented acutely with pulmonary haemorrhage and improved subsequently on a Cow's milk protein free diet.

Subjects and Methods

A case notes review was done on a five week old infant who was previously fit and well and was brought by ambulance to A&E with unresponsiveness, hypoxia, tachycardia and bleeding from mouth. He was initially breast fed and was topped up with formula from day 6 of age and there was no other significant history other than a strong family history of Cow's milk protein allergy.

Assessment showed moderate increase in work of breathing with a good air entry. There wasn't any evidence of external injury or internal bleeding including his eyes. Initial Chest X-ray showed bilateral opacification. He was resuscitated with IV fluids due to poor perfusion and was treated with IV antibiotics. Due to respiratory deterioration, he was intubated and bleeding was noted in the trachea. He was ventilated and was transferred over to the nearest intensive care.

His subsequent investigations showed microcytic, hypochromic anaemia, normal platelets and clotting screen. His infection screen in blood and CSF, metabolic work up, skeletal survey, CT angiogram thorax and CT brain were normal. On extubation, he was started on an extensively hydrolysed formula (Peptijunior) and he became asymptomatic.

Results

Heiner's syndrome was diagnosed in this infant after extensive work up and he was put on extensively hydrolysed milk. He had a significant improvement and was found to be thriving well on the 99.6th centile at his follow up in five months when he continued to be asymptomatic.

Summary

Heiner's syndrome is a diagnosis of exclusion which improves rapidly with a dairy free diet. Radiologic pulmonary infiltrate is a fairly consistent finding in most children. There could be supporting evidences of eosinophilia, anaemia, positive IgE to cow's milk protein as well as haemosiderin laden macrophages in Broncho-alveolar lavage and/or gastric washings (2).

Conclusion

Even though Heiner's syndrome is rare, it should be suspected in young children who present with chronic pulmonary disease of unknown causes and it could be potentially life threatening in severe cases due to pulmonary haemorrhage.

1,2. Moissidis I, Chaidaroon D, Vichyanond P, Bahna SL. Milk-induced pulmonary disease in infants (Heiner syndrome). Pediatr Allergy Immunol. 2005 Sep;16(6):545-52.

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Background

In 2013 the coeliac working group of the British Society of Paediatric Gastroenterology, Hepatology and Nutrition (BSPGHAN) produced a new guideline which has resulted in some children being diagnosed with coeliac disease due to symptoms and bloods alone instead of upper gastrointestinal endoscopy. Since 2011 there has been dietitian led service in Lanarkshire. Following release of these guidelines and in conjunction with the West of Scotland Paediatric Gastroenterology, Hepatology and Nutrition group (WOSPGHAN) the guideline was implemented in June 2013.

Aim

To study the number of children diagnosed by bloods versus scope, to examine age, level of coeliac serology, HLA typing and marsh grading at diagnosis from June 2013 – 2016.

Subjects and Methods

There have been sixty nine children diagnosed with coeliac disease since the dietitian led service began in 2011. There are currently 84 active patients, eleven of whom have diabetes. Data was collated from the coeliac database, SCI Acute results system, medical and dietetic records.

Results

Between 2011 and 2016 sixty nine children have been diagnosed with coeliac disease in Lanarkshire with a mean of 11.5 per year (range 8 – 17). In 2015 twelve children from a population of 118,158 were diagnosed which meant a rate of diagnosis 1 in 9847 or 10.15/100,000. Whilst in the South East of Scotland, White et al2 (2013) found that there were 11.7/100,000 (2005-2009). The mean age of diagnosis has reduced over this time period from: 2014 – 9.3 (range 2-12), 2015 – 7.9 (range 1-15) and 2016 – 5.9 (range 2 – 15). There has also been an increase in classical presentation with poor growth, muscle wasting, distended abdomen and lethargy in younger children.

Since June 2013 twenty one children have been diagnosed by bloods and twenty three by scope. Of those forty eight scoped (since 2011) eleven had Diabetes Mellitus, two were IgG Endomysial Antibody positive, two were diagnosed by adult services but would not have required an endoscopy and one was under two years of age. One of those diagnosed by bloods alone was scoped one year after diagnosis due to ongoing abdominal pain. The scope indicated minimal chronic inflammation, a slight increase in intraepithelial lymphocyte cells and mild villous architectural changes. This therefore has saved four theatre lists of upper scopes with fewer children requiring a hospital investigation and general anaesthetic with the anxiety that can accompany this.

Reviewing results of anti tissue Transglutaminase antibody there was twenty three being over 70U/ml and fifteen over 128U/ml. For Endomysial antibody results thirty two were IgA positive one had a positive and negative IgA result, one was IgG positive and one had no test. Reviewing results of HLA Typing there were twenty nine with DQ2 positive, two with DQ8 positive, two were positive for DQ2 and DQ8 with seven having no test. In terms of Marsh grading from pathology samples, three had no grade (two scoped by adult services) and three no grade was given (two with a diagnosis made following gastroenterology and pathology meeting, one was one year post diagnosis), one was 2-3a, two were 3a, eight were 3b and three were 3c.

Summary and conclusion

A third less endoscopies have been required since the introduction of the new guidance. There appears to be a rise in the numbers of children with coeliac disease. There may be a trend of children being diagnosed at a young age which may be due to increased testing as well as greater awareness of the condition.

References:

- 1. Murch S, Jenkins H, Auth M, et al. 2013. Joint BSPGHAN and Coeliac UK guidelines for the diagnosis and management of coeliac disease in children. Arch Dis Child 98:806–811
- 2. Lois E. White, Victoria M. Merrick, Elaine Bannerman, Richard K. Russell, Dharam Basude, Paul Henderson, David C. Wilson and Peter M. Gillett. 2013. The Rising Incidence of Celiac Disease in Scotland. Pediatrics 2013. 132 (4) e924-931

F17 Long term outcome 6 -12 months after treatment with hospital PN for >27 days in a specialised children's hospital.

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Air

Parenteral nutrition is an essential supportive treatment for severe intestinal failure. However long –term parenteral nutrition carries the risk of developing life threatening Liver disease. Our aim was to review all hospitalised children on PN for more than 27 days to determine incidence, aetiology and outcome of intestinal failure-associated liver disease (IFALD).

Methods

All paediatric inpatients requiring PN for more than 27 days were included in this study. Data was collected from patients' notes and electronic data record of hospital on a pre-set required format. We reviewed and analysed data from June to November 2015.

Results

A total of 61 patients including 11(18%) neonates were reviewed (M: F; 28:33). There were 9 preterm neonates including 2 extreme preterm babies. Mean age was 4.8 years. Mean duration of PN was 72 days. Primary digestive disorder (PDD) was noted in 31 (51%) children with 11(18%) children having enteropathy and 2(3%) dysmotility. 17(28%) children had surgical diagnosis; commonest diagnosis was NEC in 11(18%) children followed by gastroschisis in 2(3%), hirschsprung disease in 2(3%) and atresia in 2(3%) children. Thirty (49%) children had Primary non-digestive disorder; the commonest group was oncology children 20(36%) followed by cardiology 4(6%), Immunology 2(3%) and other 4(6%). Thirteen (21%) children developed Intestinal failure-associated liver disease (IFALD). One child progressed to stage 2 and another to stage 3 IFALD. 11(85%) children with IFALD were less than one year old. Only 4 (6%) patients were noted to have sepsis. 42 (69%) children were noted to have hypoalbuminemia (<34g/L) and 15 (25%) severe hypoalbuminemia (<25g/L) before PN was started. After four weeks of parenteral nutrition treatment hypoalbuminemia (<34g/L) was noted in 32 (52%) children and severe hypoalbuminemia (<25g/L) in 4(6%) children. IFALD was associated with younger age, p=0.0005, prematurity, p= 0.003, surgical diagnosis, p=0.002 and hypoalbuminemia before starting on PN, p=0.039. IFALD was not associated with sepsis. All patients were followed-up between 6 months and 12 months of starting on PN. On follow-up, 5(8%) children were reported to have died, all with Primary non-digestive disorder (PNDD). The cause of death in these children were complications related to primary diagnosis but not PN related problems. Out of 56 alive children, sufficient enter feeds were established in 48(86%) children. Eight (14%) children were on long term home PN, all with Primary Digestive Disorder (PDD). No death was reported related to PN complications.

Conclusion

Although Parenteral nutrition is associated with life threatening complications it can now be considered relatively safe lifesaving supportive treatment for severely unwell children with intestinal failure associated with a wide range of underlying diseases in a specialist hospital setting. Mortality is related to the underlying disease as opposed to the PN itself.

F18 Long term outcome of Intestinal Rehabilitation in children over a period of 15 years – A single centre experience

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Background

Home Parenteral Nutrition (HPN) is the primary treatment for patients with irreversible intestinal failure (IF). Outcomes have improved over recent years, attributed to provision of care via a multidisciplinary nutritional support team.

Aim

To describe the efficacy and safety outcome measures of intestinal rehabilitation (IR) in children over a period of 15 years.

Subjects and Methods

Efficacy of IR was measured using the following parameters - growth and nutritional status, proportion of total enteral calories, dependence on enteral tube feeding (ETF), duration between initiation of PN and discharge home. Parameters used to measure safety of HPN included mortality, prevalence of intestinal failure associated liver disease (IFALD) and catheter related blood stream infections (CRBSI); and number of inpatient days because of complications of PN. Data was collected from patient notes and hospital records.

Results

31 patients (17 females) received HPN between 2001 and 2016. 3 of these patients were administered PN as part of palliative care package therefore not included. 17 (60%) patients had short bowel syndrome (SBS), 2 of these had ultra-short bowel syndrome (bowel length <10cms. 6(21%) had chronic intestinal pseudo-obstruction (CIPO). 5 (18%) enteropathy.

12 patients remain on HPN (2016) and 10 achieved enteral autonomy. 2 patients died because of complications of PN (1 on waiting list for liver and small bowel transplant) and 1 died post transplant. 3 patients were transitioned to adult services on PN. Among those on PN 3 patients were receiving less than 50% calories via PN, 7 were on 50-80% and 2 were on >80% PN calories. Growth and nutritional intake was assessed for patients being followed up at the time of the study (n=22). WFH (z-score for weight for height) was within 2 SD for all patients and HFA (z-score for height for age) was within 2 SD for all except 2 patients where it was -2.16 and -4.49. In only a third (8/22) of the patients enteral tube feeding was needed to support their nutrition.

The median length of stay from initiation of PN to discharge home was 152 days (Interquartile range IQR 128). Median number of days of readmission into hospital because of complications of PN was 14 days (IQR 31). The commonest reason for readmission was CRBSI followed by blocked catheter. Measures of safety of PN were compared between 2 defined periods, first 10 years (before establishment of NST) and subsequent 5 year

	2001-2011 (n= 13)	2011-2016 (n=15)
Total PN days	10213	18446
Mortality	2 (15%)	1 (7%)
IFALD	2 (15%)	2 (13%)
CRBSI/1000days	4	1.92

Summary and conclusion

The number of IF patients being managed on HPN has increased. There has been a reduction in morbidity and mortality. Most children achieved good linear growth and none of them were classified as obese.

20/22 current patients ate a varied diet and only a third of the patients needed support by enteral tube feeding.

F19 Glucose-galactose malabsorption - a rare cause of severe congenital diarrhoea

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Background

Congenital glucose-galactose malabsorption is a rare autosomal recessive disorder caused by a defect in glucose and galactose transport across the intestinal brush border. It is characterized by severe watery diarrhoea, dehydration and can result in early death without appropriate dietary management. Glucose-galactose malabsorption is caused by mutation in the SLC5A1 gene which results in a dysfunctional sodium/glucose cotransporter protein.

Case presentation

A two months old boy of consanguineous parents presented with persistent watery diarrhoea 2 days after birth, faltering growth and metabolic acidosis. He was born at term with a birth weight of 3.58 Kg (50th centile), and his weight at initial assessment was 3.81 kg (0.4 centile). His sibling had died at the age of 3 months with suspected pneumonia but had a history of unexplained chronic diarrhoea too. A full septic screen was performed which was normal. Parenteral nutrition was commenced pending investigations.

The diarrhoea was felt to be osmotic as it ceased when his feeds were stopped and the calculated stool osmotic gap was elevated, and further diagnostic evaluation excluded disorders of villous architecture such as microvillus inclusion disease and intestinal epithelial dysplasia, autoimmune enteropathy, acrodermatitis enteropathica and cystic fibrosis.

Despite several dietary manipulations with lactose-free, partially hydrolysed and elemental formulas the diarrhoea persisted, however he responded to a trial of a fructose based formula - Galactomin 19. This raised the possibility of glucose-galactose malabsorption which was subsequently confirmed by a homozygous SLC5A1 missense mutation, p.lle467Thr (c.1400T>C). The parenteral nutrition was stopped and oral feeding was fully established using Galactomin 19. At subsequent clinic follow ups, his parents reported that he was well, feeding on demand and passing formed stools. His growth and development soon caught up.

Summary and conclusion

Glucose-galactose malabsorption is a rare disorder that can present with severe life threatening diarrhoea in the early days of life. It can present a significant diagnostic challenge to neonatologists, paediatricians and paediatric gastroenterologists and should be considered early in the differential diagnosis of infants presenting with severe osmotic watery diarrhoea.

F20 Superior Mesenteric Artery syndrome as a presentation of Crohn's Disease.

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Background

Superior Mesenteric Artery Syndrome (SMA) is a rare cause of upper GI obstruction due to compression of the third part of the duodenum between the abdominal aorta posteriorly and superior mesenteric artery anteriorly. It causes postpradinal epigastric pain, nausea, vomiting and weight loss.

Subjects and Methods

15 yr old female presented with a 3 month history of vomiting, lethargy and weight loss. There had been a preceding gastroenteritis. She initially vomited overnight but this progressed in frequency and time of day. She was constipated but had no associated abdominal pain, blood or mucus PR. She was having migraines but had no other symptoms. Omeprazole was started by the GP with no effect. Academically she was very bright and doing well. Excellent family and social networks. No change in mood or life events. Enjoyed gymnastics 5 x a week but had been unable to attend due to weakness. There were no body image issues and she was trying to gain weight.

She weighed 44.9Kg (9th) Ht 165cm (50th) BMI 2nd. A loss of 9 Kg or 15% in 3 months. Was bright and interactive but looked malnourished, no skin or joint involvement. Systems examination was normal. Happily chose and ate a meal during examination.

Results

Initial hypokalaemia and hypophosphatemia were corrected and ranitidine was started. MRI head excluded brain pathology. Initial abdominal USS was normal. Barium Swallow was suggestive of SMA and an abdominal x ray 24 hours after showed stomach distension with barium that was consistent with delayed gastric emptying and supported the diagnosis. She was started on omeprazole and slowly enteral feeds were introduced. Further targeted abdominal USS showed the duodenum obstructed by the superior mesenteric artery. Initial upper GI endoscopy revealed normal oesophagus, but a large distended stomach with oedematous mucosa but no evidence of ulceration In view of the size of the stomach and the oedema the pylorus couldn't be intubated,. Pathology showed gastritis with some infiltrate of plasma cells in the lamina propria and some focal attenuation and possible granuloma. H pylori was not identified. She was commenced on high dose omeprazole and exclusive enteral feed with an aim to reduce the vomiting and improve the gastric distention. Repeat endoscopy again couldn't pass into the duodenum and ulcers were visible. Faecal calprotectin was 748. Ileo-colonoscopy demonstrated a normal colon with mild inflammation in the terminal ilieum. Crohn's disease was diagnosed 2 months after initial presentation. Small bowel with contrast showed terminal ileum inflammation with some proximal dilatation, there was notable gastric and proximal duodenal distension at the level of the mesenteric vessels. She failed to respond to exclusive enteral nutrition and while her disease improved on oral prednisolone she became dependent despite the introduction of azathioprine. Treatment was escalated to combination infliximab and azathioprine and she responded very well.

Summary and Conclusions

15 year old athletic female presenting with vomiting diagnosed with upper GI Crohns disease complicated by SMA syndrome. 5 months after presentation she gained 12 kg and had a weighted paediatric Crohns disease activity index (wPCDAI) of 0. She continues to do well.

F21 A Teaching Challenge in Home Parenteral Nutrition

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Background

Home parenteral nutrition is increasingly used to address the nutritional needs of those with intestinal or growth failure. Morbidity and quality of life is directly influenced by the quality of education and training given to parents learning to administer parenteral nutrition, as well as a potential financial benefit. Though the established best practice is to lead training with the multidisciplinary nutritional support team, there is limited evidence to suggest the best methods to train parents and care givers.

Aim

By reporting a case of a challenging teaching scenario and evaluating some of the literature on best methods used in teaching parents, we aim to illustrate the difficulties faced, and innovative techniques used when teaching home parenteral nutrition (HPN) and the invaluable support required from the nutritional care team to enable the safe management of patient's in their home environment.

Subjects and Methods

A 6 month old with failure to thrive secondary to osmotic diarrhoea was admitted for assessment of nutrition and consideration of parenteral nutrition (PN). The fourth child of consanguineous Pakistani parents, he had been closely followed up in outpatients with dietetic support but did not gain significant weight off PN.

Main challenges highlighted before commencing HPN were his mother's lack of English language, being innumerate and also illiterate. His father, though he had a better grasp of the English language, had learning difficulties and little enthusiasm to learn HPN techniques. Training more than one parent is normally encouraged.

Through Mum's dedication and the Nutritional Care Support team's diligence they began the painstaking task of training.

A consistent Urdu translator was present for all training sessions. Standardised HPN training kits and contract had to be adjusted to facilitate Mum's learning. Particular areas of difficulty were reading the names of the Gastro Team Emergency Contacts, reading the rates and volumes of infusions from the prescription and learning the simple task of putting on sterile gloves.

Further complications arouse when it became clear Mum's social standing was undermined within the family hierarchy and despite other family members being integrated into UK life, she had been held back from education and gaining language skills. She was isolated and with little social support. This made the situation less than ideal to train HPN.

Pictorial representations of procedural steps and photos of key team members were added to training documents. Basic Urdu words were used to subtitle pictures and phone numbers in the hope Mum would recognise these more than English words, or gain better engagement from Urdu educated family members. When signing off procedures, repetitive examples were required before being deemed competent. Healthcare at home community nurses were drafted in for setting up and flushing off of all PN infusions to support Mum.

Results

He was discharged on four nights a week of parenteral nutrition after several weeks in hospital and rigorous discharge planning. This was achieved by the hard work and effort from both parties; improving quality of life, encouraging the child's development and reducing costs. As a consequence of this interaction, the Nutritional support team have created an illustrated HPN training pack to aid learning when educating parents in HPN.

Summary and conclusion

HPN training can be achieved with a strong Nutritional care support team. Innovative methods for training are sometimes required to achieve training goals even in challenging scenarios. Learning from experience helps develop future training methods.

F22 Audit of screening and confirmation of diagnosis of coeliac disease in type 1 Diabetes patients

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Background

In recent years, there has been significant change in the guidance for screening and confirmation of coeliac disease (CD) in patients with type 1 IDDM (Insulin Dependent Diabetes Mellitus). ISPAD (International Society for Paediatric and Adolescent Diabetes) Clinical Consensus Guidelines 2014 (http://c.ymcdn.com/sites/www.ispad.org/resource/resmgr/Docs/CPCG_2014_CHAP_19. pdf) suggest screening of type 1 IDDM patients for CD at diagnosis with HLA DQ2/DQ8 and Immunoglobulin A tissue transglutaminase antibody (tTG-A). BSPGHAN (British Society of Paediatric Gastroenterology, Hepatology and Nutrition) guidelines 2013 (https://bspghan.org.uk/sites/default/files/guidelines/Coeliac%20Guidelines%202013_0.pdf) provide clear pathway for confirmation of diagnosis of CD in symptomatic and asymptomatic patients. Data for this audit was collected from 2011 when ESPGHAN (European Society of Paediatric Gastroenterology, Hepatology and Nutrition) guidance for this screening and confirmation of coeliac disease was first available and adopted in our centre.

Aim

- 1. To evaluate practice of screening of type 1 IDDM patients for CD by diabetes team prior to referral to gastroenterology team.
- 2. To evaluate compliance with ESPGHAN/ BSPGHAN guidance in confirmation of diagnosis of CD in these patients.

Methods

Retrospective data collected from type 1 IDDM and coeliac databases over 5 year period (2011-2015). Information on first screening for coeliac disease (including HLA) as well as frequency of screening and patient reporting of gastrointestinal symptoms until referral to gastroenterology team was collected. Retrospective reporting of gastrointestinal symptoms in gastroenterology clinic and method of confirmation of diagnosis of CD was noted to compare compliance with BSPGHAN guidance.

Results

12 patients (4 males and 8 females) with type 1 IDDM were diagnosed with CD with age range of 3-16 (mean 9.75 years). All had screening with TTG at diagnosis of type 1 IDDM however HLA was not done on any of these patients. Referral to paediatric gastroenterology team was as per BSPGHAN guidance in all the patients. Reporting of gastrointestinal symptoms increased from 3/12 in diabetic clinics to 8/12 in gastroenterology clinics. CD was confirmed by endoscopy in 5/12 (One transferred from other centre, one had TTG >3 times but <10 times UNL (upper normal limit) and 3 had raised tTG-A with no gastrointestinal symptoms. CD was confirmed by blood tests including HLA in 7/12 patients who were symptomatic and had TTG >10 times UNL.

Summary and conclusion

Screening of newly diagnosed patients with type 1 IDDM by diabetic team did not involve HLA testing. This could potentially avoid regular screening in minority of patients. There was significant increase in retrospective reporting of gastrointestinal symptoms in these patients when they attended with raised tTG-A suggesting possible subjective nature of the history. Diagnosis of CD was confirmed as per BSPGHAN guidelines with appropriate use of endoscopy where indicated.

Based on this small study, we suggest survey for review of practice in other centres to assess compliance to current guidance by both Diabetes and gastroenterology teams.

F23 Implementation of an International Quality Improvement Initiative (ImproveCareNow – ICN) for Children with Inflammatory Bowel Disease (IBD) – A UK site perspective

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Background

IBD are conditions causing chronic relapsing inflammation of the gastrointestinal tract and encompass two main distinct clinical entities; Crohn's Disease (CD) and Ulcerative Colitis (UC) [1]. Although IBD can be diagnosed at any age, a significant proportion occurs in childhood or adolescence, where IBD has a profound impact on growth and development as well as psychological and educational needs [2].

Given the complex nature of IBD, standard of care varies substantially amongst centres, which is likely to influence long term outcome. ICN is the largest IBD registry worldwide [3] aiming to improve and standardise the care of children diagnosed with IBD by creating a collaborative community of patients, families and health care providers.

Aim

To present an overview of the experiences of the CUH paediatric IBD team with joining ICN, challenges encountered and future directions for improving outcomes for children and young people with IBD through quality improvement initiatives and patient-centred research.

Results

Challenges faced by the team included; securing agreements for data transfer to the US, ethical approval, obtaining funding, designing appropriate information for children and families and organisation of the recruitment process. Electronic Health Records (EHR) in the form of the Electronic Privacy Information Centre (EPIC) were installed in 2014. Although this provided a major advantage for facilitating data transfer and standardised data collection, we also faced challenges to implement ICN within EPIC at our site.

At the time of approval, 218 patients were eligible for registration for ICN. So far, 82 children have been registered (39%) and 61 (80%) of these have also given informed consent for their data to contribute towards future research.

Summary and conclusion

We are currently aiming to reach a registration target of 75% of our IBD population, as this will allow us to generate meaningful reference data and benchmark our performance against leading US centres. ICN has great potential to transform the care of paediatric patients, as well as providing a unique dataset to perform future research studies ultimately improving the life of children and young people with IBD.

References

- 1. Henderson P and Wilson D (2012) 'The rising incidence of paediatric onset inflammatory bowel disease' Archives of Diseases in Childhood Vol. 97
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- 3. Melmed G and Siegel C (2013) 'Quality Improvement in Inflammatory Bowel Disease' Gastroenterology and Hepatology Vol. 9 No. 5 Page 286-292

F24 Neonatal polyuria; be suspicious.

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Background

In neonatal units, increased output in nappies is recorded as urine output unless semi-formed or formed stools are noted. Watery stools are likely be recorded as increased urine output prompting renal line investigations delaying the diagnosis of congenital diarrhoeas.

Aim

We present a case of congenital watery diarrhoea to highlight the diagnostic and treatment approaches in a neonate.

Subjects and Methods

Reviewed the presentation and progress of a neonate admitted in NICU with abdominal distension and significant metabolic and electrolyte disturbance.

Results

The neonate had diagnostic laparotomy for intestinal obstruction, he was suspected to have possible necrotising enterocolitis. By day 9 of life, he developed severe hyponatraemia, hypochloraemia and metabolic alkalosis and was identified to have high urine output. Had extensive renal investigations for polyuria which was unremarkable. With faltering of growth and need for TPN, gastrointestinal loss was suspected and profuse watery diarrhoea became apparent. Hypochloraemic, Hyponatraemic metabolic alkalosis with high stool chloride confirmed the diagnosis of congenital chloride diarrhoea.

Treatment will involve (i) life-long salt substitution; (ii) management of acute dehydration and hypokalaemia during gastroenteritis or other infections; and (iii) recognition and treatment of other manifestations of the disease, such as intestinal inflammation, renal impairment and male sub fertility.

Summary and conclusion

Congenital chloride diarrhoea is a rare autosomal recessive disease characterized by life-long watery diarrhoea of prenatal onset with high faecal chloride concentration. The diagnosis may easily be missed unless there is a high index of suspicion in a neonate with increased stool or presumed urine output.

F25 Breastfeeding in Samoa: A Study to Explore Women's Knowledge and the Factors which Influence Infant Feeding Practices

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Backgroun

A decline in breastfeeding rates in Samoa has been reported over the last century. This has coincided with the introduction of western diets, progressive urbanisation, and migration of families to urban areas with more work available for women.

Aim

To assess the length of time women breastfeed, their knowledge of both the advantages of and recommendations for breastfeeding, and the factors that influence their decisions to continue or discontinue breastfeeding.

Subjects and Methods

A questionnaire was distributed at Tupua Tamasese Meaole Hospital using convenience sampling. Eligible participants were Samoan women aged 18-50 years with a baby > 6months of age. One hundred and twenty-one eligible participants were included (mean age 28.2).

Results

Ninety percent of participants initiated breastfeeding, and the majority (78%) of babies were exclusively breastfed for at least the recommended 6 months. Many mothers introduced complementary (solid) foods later than World Health Organisation (WHO) and United Nation's International Children's Fund (UNICEF) recommendations of 6 months. Awareness of the advantages of breastfeeding was mixed. The most widely known advantage was "the development of an emotional bond between mother and baby" (67%). Other advantages were less widely known. Only a small minority were aware that breastfeeding reduces risk of maternal diabetes and aids weight loss postpartum. Doctors and healthcare workers were listed as the top factors encouraging breastfeeding. Participants' comments revealed a generally positive attitude towards breastfeeding, a very encouraging finding. Participants identified that the number of breastfeeding breaks available at work and the length of their maternity leave were factors discouraging breastfeeding. Other discouraging factors identified were tobacco use, difficulty initiating breastfeeding and concerns about transmitted infections.

Summary and conclusion

Future studies are necessary to determine if problems identified in this study are applicable on a national level. These could be important to determine measures to improve breastfeeding practices in Samoa, for example legislating increased paid maternity leave and health education.

F26 Provision of Corstop® at Bristol Royal Hospital for Children

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Background

In February 2016, Bristol Royal Hospital for Children (BRHC) implemented the use of the Corstop® device in children who had been fitted with a gastrostomy or button, in the event of tube displacement. In the incidence of simple tube displacement, the stoma can close within a relatively short time period (90 to 120 minutes), which may necessitate an additional invasive intervention to form a new stoma. The Corstop® is designed to maintain the stoma patency until a trained person can insert a new feeding tube (in the community or hospital), thus avoiding a visit to the emergency department and an unnecessary procedure. A guideline was written and placed on the hospital document management system along with a parent/carer information leaflet and various sizes of Corstop® devices were ordered in.

Subjects and Methods

Between February and June 2016 an audit was completed looking at all patients who had had a gastrostomy or button fitted and whether the guidelines were followed and the parent information given. A list of patients was provided from the community home enteral feeding team including all relevant discharges between this period. The dietetic and medical notes were checked to find the following information:

- 1. Did the child discharge with a Corstop[©]
- 2. Was the Corstop® given an appropriate size for the gastrostomy device insitu?
- 3. Was the patient/carer Corstop[®] training completed prior to discharge?
- 4. Was the patient information sheet for home given to the family prior to discharge?

All audit questions were based on the standards of care where the percentage required was 100.

Results

33% of patients were given a Corstop® device on discharge. None were given the correct size. 17% of parents and carers had training provided. None were given the patient information leaflet

Summary and conclusion

The provision of Corstop[®] is poorly documented within the medical and dietetic notes. The completion of patient training is low and the correct size of the device is not given. The responsibility of who gives out the device is unclear and there was no obvious place to record that the device, size and information for carers given within the notes.

Dissemination of the guideline with a clear pathway is required to ensure that all patients leaving the hospital with a gastrostomy or button have; training on how to use the Corstop[®] device, are given supporting written information and the correct sized device. A hospital wide action plan to include; the development of an electronic information form attached to the patients detailing the Corstop[®] standards and will be put into place through the enteral feeding strategy group to take these actions forward. A reaudit will be performed in early 2017.

F27 Head Scarf Pins.. Be Aware!!

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Background

Sharp objects ingestion is one of the most challenging clinical scenarios in paediatric gastroenterology. Cultural factors play important role in the incidence rate and the type of ingested objects1.

Δime

- 1. To report two similar cases of straight pin ingestion following hair scarf re-adjustment which has been managed according to the available guidelines at the time.
- 2. To emphasise the potential consequences of delayed sharp objects removal.
- 3. To demonstrate the importance of the available evidence in guiding clinical practice.

Subjects and Methods

Two cases, 12 and 13 year old girls, presenting with accidental ingestion of hair scarf pin while holding it between the lips and adjusting the scarf.

Results

The first case was managed prior to the publication of the paediatric gastrointestinal endoscopy guidelines by the European Society of Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN) and European Society of Gastrointestinal Endoscopy (ESGE) September 2016. As the patient was asymptomatic, the surgical team advised to watch and wait. However, the pin did not pass after 2 weeks and needed endoscopic removal (the pin head was embedded in the mucosa near the pylorus causing necrosis and damage). The second case attended after the guidelines were published with identical pin accidently ingested and removed on the day of ingestion as per guidelines (the pin head was also embedded in the mucosa but no necrosis was found).

Summary and Conclusion

The endoscopic removal of ingested foreign bodies remains a challenge facing paediatric endoscopists. The paediatric gastrointestinal endoscopy guidelines published by ESPGHAN and ESGE in September 2016 provide guidance to aid clinical decision; and in the second case it resulted in early endoscopic removal to prevent long-term mucosal damage and necrosis.

The pins used to fix head scarves in young girls can lead to life threatening incidents. This can be avoided by raising awareness and following safe removal technique.

Local council public health department has been informed to look further into this matter.

References

1. NASPGHAN Guidelines April 2015

F28 Vitamin B12 Deficiency is Common in Children with Ulcerative Colitis as well as Crohn's Disease

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Background

Crohn's disease is a risk factor for vitamin B12 deficiency due to frequent involvement of terminal ileum.

Ain

To assess the prevalence of vitamin B12 deficiency in children with Crohn's disease (CD) and Ulcerative Colitis (UC).

Subjects and Methods

We performed a single-centre service evaluation of 157 patients with CD and compared them with 88 patients with UC. In patients with CD, ileal inflammation on endoscopy and MRE were recorded.

Results

Prevalence of B12 deficiency in patients with CD was 31% compared with 32% in ulcerative colitis. 6/55 (11%) patients with CD had vitamin B12 deficiency at the time of diagnosis or within 3 months of diagnosis. 20/66 (30%) had low vitamin B12 levels at 1 year of follow up. Ileal inflammation seen in endoscopy or MRI was a risk factor in the development of B12 deficiency.

5/27 (19%) patients with UC had low vitamin B12levels at the time of diagnosis or within 3 months of diagnosis. 28/88 (32%) had low vitamin B12. 8/38 (21%) had low vitamin B12 level at 1 year of follow up.

Summary and conclusion

Vitamin B12 deficiency is common not only in patients with Crohn's disease but also in children with ulcerative colitis. Further studies are needed to look for the reasons behind vitamin B12 deficiency in children with ulcerative colitis.

F29 Vitamin D levels in Paediatric IBD patients living in England

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Background

Children living in England are prone to develop low Vitamin D (VitD) levels due to a limited sun exposure and often a limited oral intake. These factors are greatly exacerbated in children suffering of inflammatory bowel disease (IBD) as the disease usually relates to malabsorption, lack of appetite, food refusal, indoor resting and little sun exposure as a skin cancer precaution when using immunosuppressors. Vitamin D level monitoring has become a standard of quality in IBD care over the past years as there is good evidence that low vitamin D levels are detrimental in children for their bone health and possible for the disease itself.

Aim

To assess guidelines compliance for yearly VitD monitoring and determinate the frequency of VitD deficiency and insufficiency among our patients.

Method

We retrospectively reviewed all VitD levels performed locally and from district hospitals for IBD patients diagnosed over the past 5 years. Data were collected from electronic notes and laboratory registries. VitD insufficiency was defined when levels were 25-75nmol/L and deficiency when <25nmol/L. We compared the proportion of patients who were yearly assessed, patients with normal and insufficient levels, and IBD subtype (Crohn's disease (CD), ulcerative colitis (UC) and IBD unclassified (IBDU)). Demographics, laboratory findings, and treatment were extracted to a STATA database. Descriptive analysis was performed using absolute value, percentage and mean functions.

Results

51% (n=96) of patients under follow-up had VitD determination at least once over the past 3 years. When grouped by year, 21 patients had vitamin levels in 2014, 43 in 2015 and 66 in 2016. This represents a 3 fold increase considering an average IBD population of 188 patients. 15.6 % (15/96) were VitD deficient (6 CD, 5 IBDU, 4 UC) at least once, 60.4% (58/96) were VitD insufficient (26 IBDU, 24 CD, 8 UC) at least once during the past 3 years. All patients who were VitD deficient had normal levels after 2-3 months of VitD treatment. Not all the patients were advised to continue on prophylactic vitamin D after recovering from low levels. 56% of patients with levels greater than 100 nmol/L were on prophylactic supplementation (400-800IU).

Conclusions

A considerable improvement in the standard of care has been achieved by increasing the yearly rates of VitD determination among our patients. VitD deficiency and insufficiency were common among our IBD patients as described in other regions. VitD monitoring enable us to identify patients at risk of abnormal bone health and to promptly commence vitamin D supplementation when low levels are found.

F30 Hypnotherapy for the Management of IBS: A Case Study

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Background

Hypnotherapy is a form of complimentary therapy that uses the power of suggestion to bring about a sub-conscious change to thoughts and behaviour. We considered this technique could be well suited to gastroenterological symptoms which are refractory to conventional treatments

Aim

To show how hypnotherapy can successfully be used to markedly reduce gastro-intestinal symptoms which are otherwise refractory to medical treatment.

Subjects and Methods

Case report: Boy with irritable bowel syndrome with severe abdominal pain and cramps which was refractory to treatment and was hugely impacting on his quality of life and school attendance.

Results

The patient had 6 sessions and showed a response to treatment after the first session (Pain score reduced from 10 on arrival to 6 at the end of the session) By the 3rd session his pain score was 0. Techniques used included: the imaginary pain dial; the timeline and ego strengthening.

By the end of the sessions his school attendance was back to full-time.

Summary and conclusion

Children are perfect candidates for this therapy. Most children can easily access their sub-conscious and may go into a dream state at some time in a day. Despite the benefits of hypnotherapy we have found challenges in expanding its use due to lack of a professional qualification or organisation to accredit its use.

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