BSPGHAN

working for children with digestive and liver disorders

British Society of Paediatric Gastroenterology Hepatology and Nutrition

ANNUAL MEETING 2013

Wednesday 30th January - Friday 1st February 2013

The Midland Hotel, Manchester

Educational Grants provided by Gold Sponsors:

Mead Johnson Nutrition

Caring for young lives
Children's Liver Disease Foundation

MSD

NUTRICIA
Advanced Medical Nutrition

Local Organisers: Adrian Thomas; Anthony Akobeng; Andrew Fagbemi: John Bowen

E&OE and CPD Approved

Thanks to all of our sponsors

On behalf of the organising committee and the membership and counsel of BSPGHAN, we would like to express our profound thanks to all our sponsors for their very generous support given for this and every BSPGHAN winter meeting.

Care for children with paediatric gastroenterological, hepatological and nutritional disorders is best delivered by a strong well organised multidisciplinary team working with the best products made available by our partners in industry. These meetings are an opportunity to continuously improve that care. They are central to the effective operation of the society being one of our great opportunities to improve knowledge and skills, present our work and network with colleagues and yourselves, our partners in the pharmaceutical and scientific nutrition commercial arenas. They are always also great fun. We are therefore extremely grateful for your support and continuing contribution to our development as well as these opportunities to meet with you.

We therefore trust and hope that this meeting will be as beneficial and enjoyable for you as it is for us.

Thanks to our Principal Sponsors:

Mead Johnson Nutrition
Caring for young lives – Children's Liver Disease Foundation
NUTRICIA Advanced Medical Nutrition
MSD

Thanks to our Silver Sponsors:

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Orphan Europe
Abbott Nutrition

Charities:

NACC
CICRA

Abstract selection committee:

Dr Rajeev Gupta, Chair Education Group: Dr Julian Thomas, Chair Research Group; Dr Andrew Fagbemi and Dr Anthony Akobeng, Local Organising Committee: Dr Fiona Cameron, Chair Elect Trainee Members' Group; Mr Mick Cullen, Chair Associate Members' Group

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BSPGHAN
working for children with digestive and liver disorders

British Society of Paediatric Gastroenterology Hepatology and Nutrition

Welcome address from Local Organising Committee

Dear Colleagues

We wish you all a very warm welcome to Manchester for the 27th annual meeting of BSPGHAN. We hope that you will enjoy the scientific programme and the social events as well as finding time to see some of our wonderful city. Manchester began as Roman fort in 79AD but it didn't become a city until the industrial revolution. When the Manchester ship canal opened in 1894 it was the largest river navigation canal in the world and it enabled Manchester to become Britain's third busiest port despite the city being 40 miles from the sea. The world's first passenger railway ran between Manchester and Liverpool. Manchester is also where the atom was first split and the world's first computer was built. The city is the birthplace of socialism and it is where Marks and Engels drafted the communist manifesto. More recently music has had a great influence: The Bee Gees, 10cc, The Sex Pistols, Joy Division, New Order, Happy Mondays, The Stone Roses, Morrissey, The Smiths, Simply Red, Oasis and Take That all originated here. The legendary Hacienda nightclub (now apartments) was 3 minutes from the Midland Hotel.

Manchester city centre was rebuilt after the devastating IRA bomb in 1996 and today is unrecognisable from the post-industrial city of 20 years ago. Home to grand relics of the industrial revolution such as the Town Hall on Albert Square, just 2 minutes from here, the modern city is also studded with independent boutiques, top-flight shops, great hotels, restaurants, concert halls, cool bars and snug pubs. The museums house some of the UKs finest historic collections including the Manchester Museum, People's Museum, The Museum of Science and Industry, Imperial War Museum North and the National Football Museum. The Manchester Art Gallery, 2 minutes away on Moseley Street, is an impressive Neo-Classical building with an outstanding Pre-Raphaelite collection. Further afield The Lowry Arts Centre in Salford Quays is a spectacular waterside development housing theatres, art galleries and the Trafford Centre is a shopper's paradise. Manchester has the UK's largest single site university and hosts the two leading premiership football teams.

The Midland Hotel was built by the Midland Railway as the counterpart to the St. Pancras Hotel at the other end of the line in London. It was here that Henry Royce first met Charles Stewart Rolls, a meeting which led to the formation of Rolls Royce. It is close to the city centre shops and the bars in Castlefield, Deansgate and Deansgate Locks. For a great view of the city try Cloud 23 cocktail bar on 23rd floor of the Hilton hotel at sunset.

We have tried to organise a full and exciting scientific programme with the central theme for the meeting being evidence based practice. We hope that the new and old members alike will enjoy presenting their work and that there will be much lively discussion. Please also take time to visit the stands of our sponsors who have provided very generous support. We are also indebted to Carla for her advice, support and hard work in all aspects of the organisation. We are honoured to welcome you to Manchester and hope that you have a very enjoyable stay here.

From left to right: Andrew Fagbemi, Jo Price, Hannah Barlow, Abigail Swancott, Anthony Akobeng, Adrian Thomas, Jane Roberts, John Bowen

Wednesday 30th January 2013

POSTGRADUATE DAY CONFERENCE
The Midland Hotel, Manchester

8.00
Registration Opens - The Midland Hotel, Foyer

Visit Exhibitor Stands
Trafford Suite and Octagon Lounge

9.50
Meeting Opens - Alexandra Suite A

9.50 – 10.00
Welcome and Introduction
Dr Adrian Thomas - Consultant Paediatric Gastroenterologist, Manchester

Session 1
10.00 – 11.30
Evidence Based Medicine

Chairs:
Dr Mark Beattie
Consultant Paediatric Gastroenterologist, Southampton General Hospital
and
Dr Richard Hansen
Clinical Lecturer in Child Health, University of Aberdeen, Aberdeen

10.00 – 10.20

Basic Principles of EBM
Dr Bob Phillips
Paediatric Oncologist
Leeds General Infirmary
Leeds
LS1 3EX

10.20 – 10.40

How systematic reviews help in understanding the evidence
Dr A Akobeng
Consultant Paediatric Gastroenterologist
Royal Manchester Children's Hospital
Oxford Road
Manchester
M13 9QL

10.40 – 11.00

A case-based discussion - communicating the risk of lymphoma in IBD patients who receive biological therapy
Dr Tony Akobeng, Consultant Paediatric Gastroenterologist
and Dr Morris Gordon, Consultant Paediatrician
Royal Manchester Children's Hospital
Oxford Road
Manchester
M13 9QL

11.00 – 11.20

Clinical Guideline: Principles and Progress
Professor Mark R Baker
Clinical Adviser
National Institute for Health and Clinical Excellence
Manchester
M1 4BD

11.20 – 11.30

Discussion

Session 2
11.30 – 12.30
Free Papers
Plenary Session One

Chairs:
Dr Alastair Baker
Consultant Paediatric Hepatologist, King's College Hospital, London
and
Dr Susan Protheroe
Consultant Paediatric Gastroenterologist,
Birmingham Children's Hospital, Birmingham

11.30 – 11.40

Azathioprine and 6-mercaptopurine for maintenance of surgically-induced remission in Crohn's disease: A systematic review
Gordon M[1,2], Taylor K[3], Akobeng AK[4], Thomas AG[4]
[1]Blackpool Victoria Hospital, Blackpool; [1]University of Salford;
[1]North Manchester General Hospital; [1] Royal Manchester Children's Hospital

11.40 – 11.50

The Paediatric UK IBD Audit – What do 5 years of participation tell us?
Aimee Protheroe, Project Coordinator–UK IBD Audit, Royal College of Physicians;
Richard Russell,Consultant Paediatric Gastroenterologist, Yorkhill Hospital;
Sally Mitton, Consultant Paediatric Gastroenterologist, St Georges Hospital;
BSPGHAN IBD audit leads, various sites, UK.;
Emma Fernandez, Project Manager–IBD QIP, Royal College of Physicians;
Michael Roughton, Medical Statistician, Royal College of Physicians

11.50 – 12.00

Management of Eosinophilic Oesophagitis in children - a multi-centre retrospective study.
Charlotte Webb (Medical Student, Keele University), Naeem Ayub (Consultant Paediatrician, Shrewsbury and Telford Hospitals NHS Trust) and Mona Abdel-Hady (Consultant Paediatrician, University Hospital of North Staffordshire)

12.00 – 12.10

Nutritional issues of controversy in paediatric cystic fibrosis (CF) care; results of a European survey.
Chris Smith[1]; Helen White[2]; Dee Shimmin[3]; Paul Seddon[1]; Assad Butt[1]
[1]Royal Alexandra Childrens Hospital, Brighton; [2]Leeds Metropolitan University/St. James' Hospital, Leeds; [3]Belfast City Hospital; 4University of Brighton

12.10 – 12.20

The diagnostic value in paediatric small bowel assessment by wireless capsule endoscopy: A tertiary centre experience
Saliakellis E[1]*, Fotis L[1]*, St Louis D[1], Elawad M[1], Lindley K[1], Kiparissi F[1]
[1]Department of Paediatric Gastroenterology, Great Ormond Street Hospital NHS Trust, London, United Kingdom

* Doctors Saliakellis and Fotis have contributed equally to this work.

12.20 – 12.30

Survey of practices in UK centres looking after paediatric patients on home parenteral nutrition
Sophie Robertson, Caroline Cole, Mark Beattie, Mick Cullen, Kim Novell, Akshay Batra. Paediatric Gastroenterology Department, University Hospital Southampton, Tremona Road, Southampton. SO16 6YD

12.30 – 13.30

Lunch and Poster Viewing
Trafford Suite and Octagon Lounge

Session 3
13.30 – 15.00
Evidence based management and Quality Improvement

Chairs:
Dr Mark Dalzell
Consultant Paediatric Gastroenterologist,
Alder Hey Children's Hospital, Liverpool
and
Dr Ieuan Davies
Consultant Paediatric Gastroenterologist, University Hospital of Wales, Cardiff

13.30 – 13.50

Eosinophilic oesophagitis
Professor Simon Murch
Consultant Paediatric Gastroenterologist
Clinical Sciences Research Institute
Clifford Bridge Road
Coventry
CV2 2DX

13.50 – 14.10

Pancreatitis
Dr Alastair Makin
Consultant Gastroenterologist
Manchester Royal Infimary
Oxford Road
Manchester
M13 9QL

14.10 – 14.30

Viral Hepatitis
Dr Suzanne Davison
Consultant Paediatric Hepatologist
Children's Liver and GI Unit
Paediatric offices
Martin Wing, Floor E
Leeds General Infirmary
LS1 3EX

14.30 – 14.50

Quality Improvement
Dr Jenny Gordon
Programme Manager Evidence into Practice
Quality Standards and Innovation Unit
C/O RCN Learning & Development Institute
Room 203
20 Cavendish Square
London W1G 0RN

14.50 – 15.00

Discussion

15.00 – 15.20
AFTERNOON TEA

Session 4
15.20 – 17.00
Quality of Life and Compliance
Short Oral Poster Presentations

Chairs:
Dr David Rawat
Consultant Paediatric Gastroenterologist,
Chelsea and Westminster Hospital, London
and
Dr Loveday Jago
Consultant Paediatric Gastroenterologist, Macclesfield Hospital, Macclesfield

15.20 – 15.25

Short Oral Poster Presentation (1)

15.25 – 15.30

Short Oral Poster Presentation (2)

15.30 – 15.50

Quality of Life in children with IBD
Dr Adrian Thomas
Consultant Paediatric Gastroenterologist
Royal Manchester Children's Hospital
Oxford Road
Manchester
M13 9QL

15.50 – 16.10

Quality of Life in children with intestinal failure
Dr Sue Beath
Consultant Paediatric Hepatologist
Liver Unit
Birmingham Children's Hospital
Steelhouse Lane
Birmingham
B4 6NH

16.10 – 16.30

Quality of Life in children with constipation
Dr Manu Sood
Paediatric Gastroenterologist
8701 West Watertown
Plank Road
Milwaukee
WI 53226

16.30 – 16.50

Managing Compliance in Paediatric Populations
Dr Stewart Rust
Clinical Neuropsychologist
Central Manchester Children's Hospital
Manchester

16.50 – 17.00

Discussion

Symposium sponsored by Pfizer

17.00 – 18.00
Breast Milk and Immunity

Speaker:
Professor Alan Lucas
Institute of Child Health
London

18.00 – 19.00
Group Meetings - The Midland Hotel, Manchester

Open to all delegates

Associate Members AGM
Trainee Members
IBD QIP Meeting

Followed by
Football - Consultants v Trainees

or Cultural Event

19.30
Bar Open - Petersfield Suite, The Midland Hotel

20.00
Ice Breaker and Meet the Sponsors Supper
Alexandra Suite Room B, The Midland Hotel

22.00 – 1.00
Disco - Alexandra Suite Room B, The Midland Hotel

Thursday 31st January 2013

The Midland Hotel, Manchester

8.00
Registration Opens - The Midland Hotel, Foyer

Coffee and refreshments in the Octagon lounge

Exhibition
Trafford Suite and Octagon Lounge

Posters
Octagon lounge

7.45 - 9.00
Open Working Group Meetings:
Rooms to be confirmed

IBD Nurses
Hepatology
Nutrition
Gastroenterology
Education

Symposium sponsored by Nutricia

9.00 – 10.00 (this will take place in Alexandra Suite)
The challenges of improving nutrition status in infants with complex disease

Chair:
Professor David Candy
Consultant Paediatric Gastroenterologist,
Lead, Medicines for Children Research Network, Surrey and Sussex

Introduction : The background to this complicated patient group, the reasons for their faltering growth and the current nutritional management options.

Dr Assad Butt
Consultant Paediatrician
The Royal Alexandra
Hospital for Sick Children
Dept Paediatric Medicine
Dyke Road
Brighton BN1 3JN

Results from a multi centre study using a new ready to use peptide feed for infants, Chris Smith will present the preliminary data from a trial.

Case studies - 2 cases from the study will be presented for discussion.

Mr Chris Smith
Senior Paediatric Dietitian
Department of Nutrition and Dietetics
Royal Alexandra Children's Hospital
Eastern Road
Brighton
East Sussex

10.00
Main Meeting Opens - Alexandra Suite

10.00 – 10.05
Welcome - Adrian Thomas

Session 1

10.05 – 11.15
Hepatology (1) – what's new

Chairs:
Dr Sally Connolly
Consultant Paediatric Gastroenterologist, Sheffield Children's Hospital, Sheffield
and
Ms Safiya Mulla
Clinical Nurse Specialist, Children's Liver unit, Leeds

10.05 – 10.25

Acute Liver Failure
Professor Anil Dhawan
Consultant Paediatric Hepatologist
King's College Hospital
Denmark Hill
London, SE5 9RS

10.25 – 10.45

Portal hypertension
Dr Patrick McKiernan
Consultant Paediatric Hepatologist
Liver Unit
Birmingham Children's Hospital
Steelhouse Lane
Birmingham
B4 6NH

10.45 – 11.05

Metabolic liver disease
Dr Andrew Morris
Consultant in Metabolic Disease
Manchester Children's Hospital
Oxford Road
Manchester
M13 9QL

11.05 – 11.15

Discussion

11.15 – 11.45

Visit exhibitor stands
Coffee and poster viewing
Trafford Suite and Octagon lounge

Key Note Lecture
11.45 – 12.15

Chair:
Dr Mark Beattie
Consultant Paediatric Gastroenterologist, Southampton General Hospital, Southampton
and
Dr Alastair Baker
Consultant Paediatric Hepatologist, King's College Hospital, London

11.45 – 12.15

Keynote lecture - Autoimmune liver disease
Professor Giorgina Mieli-Vergani
Liver Unit
King's College Hospital
Denmark Hill
London

Session II
12.15 – 13.45

Functional GI disorders

Chairs:
Dr Nikhil Thapar
Consultant Paediatric Gastroenterologist, Great Ormond Street Hospital, London
and
Ms Kate Blakeley, Clinical Psychologist
Royal London Hospital, Whitechapel, London E1 1BB

12.15 – 12.35

Mechanisms of visceral pain in functional GI disorders and principles of management
Professor Qasim Aziz
Barts and The London NHS Trust
Turner Street
London
E1 2AD

12.35 – 12.55

Functional GI disorders in children
Dr Manu Sood
Paediatric Gastroenterologist
8701 West Watertown
Plank Road
Milwaukee
WI 53226

12.55 – 13.15

Hypnosis for IBS
Professor Peter Whorwell
Professor of Medicine and Gastroenterology
Wythenshawe Hospital
Southmoor Road
Manchester
M23 9LT

13.15 – 13.35

Evidence based management of severe constipation
Dr Jenny Gordon
Research & Development Fellow
Quality Improvement Programme
RCN Institute, Whichford House
Building 1400, Parkway Court
Oxford Business Park, Cowley
OX4 2JY

13.35 – 13.45

Discussion

13.45 – 14.40

Visit Exhibitor Stands
Lunch and poster viewing
Main exhibition and lunch is in The Trafford Room.
Poster viewing, charity stands and refreshments in Octagon Lounge

Session III
14.40 – 16.10

Gastroenterology and Hepatology
Plenary Session 1 and selected short oral poster presentations

Chairs:
Dr Girish Gupte
Consultant Paediatric Hepatologist, Birmingham Children's Hospital, Birmingham
and
Dr Keith Lindley
Consultant Paediatric Gastroenterologist, UCL, London

14.40 – 15.00

FAPS – an update on colorectal guidelines
Dr Warren Hyer
Consultant Paediatric Gastroenterologist
Northwick Park and St Mark's Hospital
Watford Road
Harrow
HA1 3UJ

15.00 – 15.10

The predictive value of ELF test in Biliary Atresia
Dr Nicola Ruth[1], Professor W Rosenberg[2], Dr Patrick McKiernan[1]
[1]The Liver Unit, BCH, [2]The Institute of Liver and Digestive Health, University College London

15.10 – 15.20

Hepatitis B Vaccination Failure in Children Born to Hepatitis B positive mothers
Legarda M[1], Brown M[1], Sira J[1], Boxall EH[2], Kelly DA[1]
[1]Liver Unit, Birmingham Children´s Hospital, [2]HPA West Midlands Public Health Laboratory.

15.20 – 15.30

Long term outcome of children following liver transplantation
Legarda M[1], Smith M[1,2], Lewis P[1,2], Lloyd C[1], Paris S[2], Kelly DA[1]
[1]Liver Unit, Birmingham Children's Hospital (BCH), 2Liver Unit, Queen Elizabeth Hospital Birmingham.

15.30 – 15.40

Propranolol in the management of hepatic haemangioendothelioma – King's College Hospital experience
Protima Amon , Palaniswamy Karthikeyan , Dominic Hughes, Mark Butler, Mark Davenport, Anil Dhawan, Sanjay Bansal
King's College Hospital, London

15.40 – 15.50

The BISCUIT Study: Exploring the "bacteriotype" of de-novo paediatric IBD
Richard Hansen[1,2], Richard K. Russell[3], Caroline Reiff[4], Petra Louis[5], W. Michael Bisset[6], Andy R. Barclay[3], Jon Bishop[3], Diana M. Flynn[3], Paraic McGrogan[3], Sabarinathan Loganathan[6], Gamal Mahdi[6], Emad M. El-Omar[1] and Georgina L. Hold[1]
[1]Gastrointestinal Research Group, Division of Applied Medicine, University of Aberdeen, Foresterhill, Aberdeen; [2]Child Health, University of Aberdeen, Royal Aberdeen Children's Hospital, Foresterhill, Aberdeen; [3]Department of Paediatric Gastroenterology, Royal Hospital for Sick Children, Dalnair Street, Glasgow [4]Max Planck Institute, Cologne, Germany. ; [5]Gut Health Programme, Rowett Institute, University of Aberdeen, Aberdeen; [6]Department of Paediatric Gastroenterology, Royal Aberdeen Children's Hospital, Foresterhill, Aberdeen

15.50 - 16.00

Identifying incidence of inherited metabolic disorders in patients with infantile liver disease
Gray Z[1]*, McKay K[2]*, Lloyd C[1], Hartley J[1], MacDonald F[2], Hendriksz CJ[3], Gissen P[4], Kelly D[1],
[1]Birmingham Children's Hospital NHS Foundation Trust, UK, [2]Birmingham Women's Hospital NHS Foundation Trust, UK [3]Salford Royal NHS Foundation Trust, UK, 4UCL Institute of Child Health, UK
*these authors contributed equally
On behalf of all collaborating centres

16.00 – 16.05

Short Oral Poster Presentation (3)

16.05 – 16.10

Short Oral Poster Presentation (4)

16.10 – 16.30

Coffee Break

Session IV
16.30 – 18.00

Nutrition

Chairs:
Dr David Wilson
Consultant Paediatric Gastroenterologist, Royal Hospital for Sick Children, Edinburgh
and
Ms Sarah Maconald, Dietitian, Great Ormond Street Hospital, London

16.30 – 16.50

Feeding the disabled child – what's new
Dr Peter Sullivan
Consultant Paediatric Gastroenterologist
University of Oxford
Dept of Paediatrics
John Radcliffe Hospital, Oxford
OX3 9DU

16.50 – 17.10

Vitamin D Deficiency – does it matter
Dr Zulf Mughal
Honorary Senior Clinical Lecturer in Medicine
Royal Manchester Children's Hospital
Oxford Road
Manchester
M13 9QL

17.10 – 17.30

Acquisition of tolerance in cow's milk allergy
Professor Berni Canani
(Speaker funded by Mead Johnson Nutritionals)
Dept of Paediatrics
University "Federico II" of Naples,
Via S. Pansini, 5
80131 Naples
Italy

17.30 – 17.50

Food allergy; making sense of it all
Ms Carina Ventner
Dietitian
School of Health Sciences and Social Work
White Swan Road
Portsmouth
PO1 2DT

17.50 – 18.00

Discussion

18.15 – 19.30
ANNUAL GENERAL MEETING

20.15
Reception and Gala dinner
at The Midland Hotel with Fake That
and dancing till late

Friday 1st February 2013

The Midland Hotel, Manchester

8.30
Registration Opens - The Midland Hotel, Foyer

Coffee and refreshments in the Octagon lounge

7.45 – 9.00
Open Professional Group Meetings

IBD
PeGHAN
Endosocopy
BIF WG
Research
Motility Working Group
(*Rooms will be confirmed at the meeting - please see notice boards for information)*

Meet Professor Berni Canani - Sponsored by Mead Johnson
Refreshments provided

Session V
9.00 – 10.40

Joint BSPGHAN and BAPS
What's new in Intestinal Failure

Chairs:
Dr Andrew Fagbemi, Consultant Paediatric Gastroenterologist,
Royal Manchester Children's Hospital, Manchester
and
Mr Naved Alizai, Consultant Paediatric Hepatobiliary Surgeon,
Leeds General Infirmary, Leeds

9.00 – 9.05
Welcome

9.05 – 9.30

Medical Management
Professor Oliver Goulet
Service de Gastroentérologie pédiatrique
Hôpital Necker Enfants malades
149, rue de Sèvres
Paris

9.30 – 9.55

Surgical Management
Professor A Pierro
Nuffield Professor of Paediatric Surgery and Head of Department of Paediatric Surgery
Institute of Child Health,
University College London Medical School.
30 Guilford Street, London
WC1N 1EH

9.55 – 10.15

Chronic Intestinal Pseudo-obstruction - "are we moving in the right direction?"
Ms Kelly Larmour
Dept of Dietetics
Great Ormond Street Hospital
Great Ormond Street
London
WC1N 3JH

10.15 – 10.30

British Intestinal Failure Survey
Dr Andrew Barclay
Consultant Paediatric Gastroenterologist
Royal Hospital for Sick Children
Yorkhill Hospital
Dalnair Street
Glasgow

10.30 – 10.45

Discussion

10.45 – 11.10

Coffee Break and Poster Viewing

Session VI
11.10 – 12.20

GI surgical issues – what's new?

Chairs:
Dr Huw Jenkins
Consultant Paediatric Gastroenterologist, University Hospital of Wales, Cardiff
and
Mr Simon Huddart
Consultant Paediatric Surgeon, University Hospital of Wales, Cardiff

11.10 – 11.30

Hirschsprung's disease
Mr Ian Sugarman
Consultant Paediatric Surgeon
Dept of Paediatric Surgery
Leeds General Infirmary
Georges Meet
Leeds
LS2 9NS

11.30 – 11.50

Surgical Management of Gastro-oesophageal reflux disease
Mr Matthew Jones
Consultant Paediatric Surgeon
Alder Hey Children's Hospital
Eaton Road
Liverpool
L12 2AP

11.50 – 12.10

Preventing Necrotising enterocolitis
Dr John Puntis
Consultant Paediatric Gastroenterologist
Leeds General Infirmary
Paediatric offices, off old A Floor
Great George Street
Leeds
LS1 3EX

12.10 – 12.20

Discussion

Plenary abstract session II
12.20 – 13.20

Chairs:
Professor Ian Sanderson
Professor in Paediatric Gastroenterology, Barts and The London, London
and
Mr John Bowen,
Consultant Paediatric Surgeon, Royal Manchester Children's Hospital, Manchester

12.20 – 12.30

Diagnostic and therapeutic utility of double-balloon enteroscopy
[1]Dr Arun Urs, MBBS, MRCPCH, Centre for Paediatric Gastroenterology and International Academy of Paediatric Endoscopy Training, Sheffield Children's NHS Foundation Trust, Sheffield, UK S10 2TH; [2]Dr Massimo Martinelli, MD, Department of Paediatrics, University of Naples, "Federico II", Naples, Italy; [3]Dr Prithviraj Rao, MBBS, MRCPCH, Consultant Paediatric Gastroenterologist, Centre for Paediatric Gastroenterology and International Academy of Paediatric Endoscopy Training, Sheffield Children's NHS Foundation Trust, Sheffield, UK S10 2TH; [4]Dr Mike Thomson, DCH, FRCP, FRCPCH, MD, Consultant Paediatric Gastroenterologist, Centre for Paediatric Gastroenterology and International Academy of Paediatric Endoscopy Training, Sheffield Children's NHS Foundation Trust, Sheffield, UK S10 2TH

12.30 – 12.40

Raised D-lactate – A surrogate marker of small bowel bacterial overgrowth
Dr Hemant Bhavsar, Paediatric Gastroenterology registrar; Dr Theodoric Wong, Consultant paediatric gastroenterologist; Dr Sue Protheroe, Consultant paediatric gastroenterologist. Birmingham Children's Hospital

12.40 – 12.50

Efficacy and Safety of Low Dose and High Dose Intravenous Methylprednisolone Treatment in Acute Ulcerative Colitis
R.Vora[(1,2*)], H. Finnamore[(2*)], K. Crook[(2)], AM. Dalzell[(2)], B. Krishnamurthy[(2)], V. Krishnappa[(2)], E. Whittle[(2)], T. Irvine[(2)], MKH. Auth[(2)].
*(joint first authors, equal contribution)
[1]Imperial College,London; [2]Alder Hey Children's NHS Foundation Trust, Liverpool.

12.50 – 13.00

Financial impact of Taurolock® during long term parenteral nutrition
Dr Veena Zamvar[1], Deirdre Kriel[2], Dr Jonathan Sandoe[3], Dr John W L Puntis[1]
Departments of Paediatric Gastroenterology[1], Pharmacy[2] and Microbiology[3], The Children's Centre, The General Infirmary at Leeds, Leeds LS2 9NS, UK

13.00 – 13.10

Paediatric-onset Crohn's patients relapse earlier than adult-onset after right hemicolectomy
Lung, J, Giles, EM, Sanderson, IR, Lindsay, JO, Naik, S.
Centre for Immunology and Infectious Disease, Blizard Institute, 4 Newark St. London E1 2AT

13.10 – 13.20

The Changing Epidemiology of Coeliac Disease in South Wales – a 28 year perspective
Whyte L A, Jenkins H R
Department of Paediatric Gastroenterology, Children's Hospital for Wales, Cardiff, CF14 4WX.

13.20 – 14.20

Lunch and poster viewing

Session VII
14.20 – 16.10

Complex Inflammatory bowel disease and Short Oral Poster Presentations

Chairs:
Dr Rob Heuschkel
Consultant Paediatric Gastroenterologist. Addenbrooke's Hospital, Cambridge
and
Mr Colin Baillie
Consultant Paediatric Surgeon, Alder Hey Children's Hospital, Liverpool

14.20 – 14.25

Short Oral Poster Presentation (5)

14.25 – 14.30

Short Oral Poster Presentation (6)

14.30 – 14.50

Nutritional Management: Case Based Discussion
Ms Jo Price
Chief Dietitian
Royal Manchester Children's Hospital
Oxford Road
Manchester, M13 9QL

14.50-15.10

Monoclonal antibody treatment
Dr Anthony Akobeng
Consultant Paediatric Gastroenterologist
Royal Manchester Children's Hospital
Oxford Road
Manchester
M13 9QL

15.10 – 15.30

Leukapheresis
Professor Tarja Ruuska
Dept of Paediatrics
Tampere University Hospital
Tampere
Finland

15.30 – 16.00

Surgical Management
Mr Bruce Jaffray
Consultant Paediatric Surgeon
Royal Victoria Infirmary
Royal Victoria Infirmary
Queen Victoria Road
Newcastle-upon-Tyne
NE1 4LA

16.00 – 16.10

Discussion

PRIZE PRESENTATION AND 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

Future Meetings:

2014
London
hosted by Dr Alastair Baker

WEDNESDAY SPEAKER INVITED ABSTRACTS

SESSION ONE

Basic Principles of EBM

Dr Bob Phillips

This short seminar will address key delegate questions relating to theissues of 'What is evidence based medicine?' and 'How is it undertaken?',
and introducing ideas to be developed through the day.

How systematic reviews help in understanding the evidence

Dr Tony Akobeng, Consultant Paediatric Gastroenterologist, Royal Manchester Children's Hospital, Manchester, UK

Evidence-based medicine is a systematic approach to clinical problem solving which allows the integration of the best available research evidence with clinical expertise and patient values. What is certain in today's medical knowledge may be tomorrow's bad practice. It is therefore important that we keep up to date with new research evidence for the benefit of our patients and for our own benefit. However, keeping up with evidence is not always easy as the volume of research data is constantly expanding. The quality of individual studies on a particular clinical topic may be variable and individual studies might have produced conflicting results. It is therefore important that healthcare decisions are not based solely on one or two studies without account being taken of the whole range of research information available on that topic. For this reason, review articles are frequently used as a source of summarised evidence on a topic. In this presentation, I will discuss systematic reviews and meta-analyses and highlight the numerous advantages that these have over the traditional, narrative reviews. Using specific examples from the medical literature, I will discuss potential human costs when we fail to perform or take account of the results of systematic reviews. Practical examples of how systematic reviews have contributed to our understanding of the evidence on some gastroenterology topics will also be discussed.

A case-based discussion - communicating the risk of lymphoma in IBD patients who receive biological therapy

Dr Tony Akobeng and Dr Morris Gordon
Royal Manchester Children's Hospital, Manchester

Communicating risk to patients can be a challenging task. The reasons for this include the fact that not all the various ways of reporting risk clearly show the benefits or harms of treatments in a clinically useful way. The interpretation of risk measures can also be daunting, and many measures of risk are frequently misunderstood by both health professionals and patients. In this presentation, we will briefly discuss some of the common risk measures highlighting those which allow a more effective communication of risk and relate them to the current evidence with regard to the risk of lymphoma in patients with IBD who receive biological therapy. A clinical scenario will be used to illustrate how the principles of evidence-based medicine could be applied to help improve risk communication and improve patient care. A practical demonstration of some of the ways of communicating risk and the importance of integrating evidence with patient's preferences will also be discussed.

Clinical Guideline: Principles and Progress

Professor Mark R Baker, Director, Centre for Clinical Practice,
The National Institute for Health and Clinical Excellence, Manchester

This presentation, by the Director of the Centre for Clinical Practice at NICE (the organisation responsible for issuing guidance on clinical and cost-effective health care for the NHS), outlines the structure of a clinical guideline and the steps taken to ensure it fulfils its role in summarising the best available evidence to provide recommendations for practice.

The duration of a full guideline is almost two years and includes separate phases for scoping, searching, analysis of the evidence, drafting recommendations, quality assurance (including consultation) and publication. The opportunity to discuss recent changes in the development of clinical guidelines will be offered.

SESSION TWO FREE PAPERS

Azathioprine and 6-mercaptopurine for maintenance of surgically-induced remission in Crohn's disease: A systematic review

Gordon M[1,2], Taylor K[3], Akobeng AK[4], Thomas AG[4]
[1]Blackpool Victoria Hospital, Blackpool; [2]University of Salford; [3]North Manchester General Hospital; [4]Royal Manchester Children's Hospital

Background:
There is no standard therapy for the prevention of postoperative recurrence or relapse in Crohn's disease (CD). A recent Cochrane systematic review found evidence to suggest 5-aminosalicylates (5-ASAs) may be effective and are well tolerated. Purine analogues, such as Azathioprine (AZA) and 6-mercaptopurine (6-MP), have been extensively used in the maintenance of remission of Crohn's disease. However, adverse events that limit their long term use are commonly reported in the literature. It is unclear whether AZA/6MP are more effective than other interventions or placebo. We performed a systematic review to investigate the use of AZA/6MP for prevention of post-operative relapses in CD.

Methods:
Randomised controlled trials (RCTs), published between 1966 and August 2012, which compared AZA or 6MP with either placebo or other interventions were included. Data sources were MEDLINE, EMBASE, Cochrane Central Register of Controlled Trials, Cochrane inflammatory bowel disease and functional bowel disorders Specialised Register and reference lists of retrieved articles. Data extraction and assessment of methodological quality were performed independently by two reviewers. Data was analysed according to the intention to treat principle.

Results:
5 RCTs met the inclusion criteria. 1 study compared AZA with Placebo (all patients received metronidazole), 3 studies compared AZA with 5-ASA and 1 study compared 6MP with 5-ASA and placebo. Risk of bias was variable, with concerns regarding reporting of dropouts, blinding and other sources of bias.

Meta-analysis of 4 studies with 390 participants comparing AZA/6MP with 5-ASA showed no significant difference in rates of relapse (Odds Ratio (OR) 1.49, 95% confidence interval (CI) 0.97 to 2.30). Serious adverse events that required medication to be discontinued were significantly more common in the AZA/6MP groups (OR 2.49, 95% CI 1.38 to 4.50).

An analysis of 3 studies that just compared AZA with 5-ASAs found a statistically significant reduction in relapse in the 5-ASA group (OR 1.86, 95% CI 1.13 to 3.06). There was still a statistically significant higher risk of serious adverse events requiring discontinuation of AZA (OR 3.00, 95% CI 1.46 to 6.06).

Meta-analysis of 2 studies with 168 participants comparing AZA/6MP with placebo showed a significant difference in rates of relapse favouring AZA (OR 0.41, 95% CI 0.20 to 0.85). There was significant clinical and methodological heterogeneity between these two studies, regarding the use of concomitant medications and the choice of purine analogue.

Conclusions:
There is some evidence to suggest benefit of AZA/6MP over placebo. There is no evidence that purine analogues have superiority over 5-ASA agents. It appears that 5-ASA agents may be more efficacious than AZA/6MP to maintain remission of CD post-operatively. However, individual studies were small and of varying quality. The side effect profile of AZA/6MP is significantly worse when compared to 5-ASA

The Paediatric UK IBD Audit – What do 5 years of participation tell us?

Aimee Protheroe, Project Coordinator–UK IBD Audit, Royal College of Physicians; Richard Russell, Consultant Paediatric Gastroenterologist, Yorkhill Hospital; Sally Mitton, Consultant Paediatric Gastroenterologist, St Georges Hospital; BSPGHAN IBD audit leads, various sites, UK; Emma Fernandez, Project Manager–IBD QIP, Royal College of Physicians; Michael Roughton, Medical Statistician, Royal College of Physicians

Introduction / Background
The participation of paediatric sites in the UK IBD audit in 2008 was a major step forward in helping to ensure that the desired consistent, high-quality care is available for all IBD patients independent of age. Involvement in the re-audit in 2010 enabled analysis of comparative data for the first time, including adult audit data.

Aim
The UK IBD audit seeks to improve quality and safety of care for all IBD patients in hospitals throughout the UK. The aim of this abstract is to summarise the development of the IBD audit and reflect changes noted in paediatric IBD care over a 5 year period.

Subjects and Methods
25 specialist paediatric gastroenterology units across the UK (sites), with an IBD service that routinely admits paediatric IBD patients acutely were invited to participate in both rounds of audit (rounds of audit referred to as 2008 and 2010). 2008–23 sites collected clinical data on consecutive UC and CD inpatients and undertook a one off assessment of their provision of service (as at 01/09/08). 2010–24 sites collected clinical data on consecutive UC/CD inpatients, all IBD patients newly started on biological therapies and undertook a one off assessment of their provision of service (as at 01/09/10). GPs were approached to provide information about each patient's treatment prior to the audited admission and a patient questionnaire was also distributed.

Results
Paediatric participation in the 2010 UK IBD audit was 92% and 96% in the organisational and inpatient care elements respectively (adult participation was 90% for each). Across 2 rounds of audit, inpatient data was collected on 1119 (424 UC/695 CD) cases.

Significant improvements were identified in: 1) Inpatients seen by an IBD Nurse (306/512 [60%] 2008 and 310/435 [71%] 2010 p=0.001). 2) Collection of stool samples (122/270 [45%] 2008 and 174/318 [55%] 2010 p=0.02). 3) Use of rescue therapy in UC (17/33 [52%] 2008 and 10/38 [26%] 2010 p=0.03). 4) CD patients weight measured (288/297 [97%] 2008 and 283/285 [99%] 2010 p=0.038). 5) Prescription of prophylactic Heparin (11/512 [2.1%] 2008 to 26/435 [6%] 2010 p=0.002). Across the two rounds there was a significant increase in the number WTE IBD/Gastro nurse specialists (median [IQR]) (1[0,1] 2008 to 1.5[0.8,2] 2010 p=0.017).

Areas identified for improvement: 1) 59/237 (25%) of CD patients had their pubertal status recorded in the previous 12 months in 2010-not asked in 2008. 2) PUCAI score was recorded on Day 1 in 13/66 (20%) of UC cases in 2010-not asked in 2008. 3) 15/25 (60%) eligible sites have entered data to the biologics audit. 78/202 (39%) of adult sites in 2010 reported that their service looked after patients aged 16 or less, in total this equated to 1023 patients (680 CD and 343 UC) aged <16 years looked after by adult IBD services. Paediatric patients were significantly less likely than adult patients see their GP prior to admission (60/135 [44%] vs 923/1516 [61%] p<0.001) and of the paediatric patients who were seen they were much less likely to have treatment started (15/60 [25%] paediatric patients, 407/920 [44%] adult patients p=0.004). Of the 167 paediatric patients that returned an inpatient experience questionnaire 97% reported their overall experience during their hospital stay as good, very good or excellent.

Summary and Conclusion
Clear improvements in care and service provision have been shown. The 3rd round of IBD audit is beginning and continued support of the paediatric community is vital to: monitor provision of biological therapies in young people as per NICE guidelines, re-audit provision of services against IBD standards, assess areas of care still known to be deficient eg recording of PUCAI scores, pubertal status, and inform the debate around anticoagulation. New foci on both adolescent care and out-patient care prior to admission both of which are of particular interest to those working with young people with IBD. Data from the 3rd round will be used to inform guideline development (local and national), drive service improvement and address inconsistencies in care.

Management of Eosinophilic Oesophagitis in children - a multi-centre retrospective study.

Charlotte Webb (Medical Student, Keele University), Naeem Ayub (Consultant Paediatrician, Shrewsbury and Telford Hospitals NHS Trust) and Mona Abdel-Hady (Consultant Paediatrician, University Hospital of North Staffordshire)

Background:
Eosinophilic oesophagitis (EoE) is a condition characterised by symptoms of oesophageal dysfunction and associated with predominantly eosinophil-mediated epithelial infiltration and inflammation, diagnosed histologically by >15 eosinophils per high powered field.

Younger children often present with feeding intolerance and symptoms of gastro-oesophageal reflux while older children and adults may have dysphagia and food impaction. If untreated, faltering growth and oesophageal stricture formation may ensue.

There is an increasing prevalence with a 3:1 male:female ratio. Incidence is greater among white ethnicities and in individuals with underlying allergy. There is a paucity of data on both the natural history and optimal treatment of EoE. Despite the recommendations of the First International Gastrointestinal Eosinophil Research Symposium (FIGERS) in 2007, there remains a lack of consensus towards management in children.

Aim of the study:
To review the management of EoE at three UK District General Hospitals.

Subjects and methods:
All children (≤ 18 years) with a diagnosis of EoE between 2009 - 2012 were identified from 3 hospitals: University Hospital of North Staffordshire, Royal Shrewsbury Hospital and Princess Royal Hospital, Telford. A retrospective review of case notes was carried out using a specifically designed proforma and data collected using excel sheet.

Results:
Twenty seven patients were identified, 5 female and 22 male. Median age at diagnosis was 8.5 years. Presenting complaints varied, with a proportion asymptomatic (22%) – other primary symptoms were gastro-oesophageal reflux ± vomiting (55.5%) and feeding difficulty (18.5%), often in combination or compounding one another. Amongst all patients with whom feeding was a problem, dysphagia was responsible for 54%.

There was a positive history of atopy in 37% of patients. More in-depth allergic investigations were carried out in all patients using skin prick testing (70%), radioallergosorbant testing (RAST) (15%), eosinophilia count (78%) and serum IgE levels (26%).
26 patients (96%) underwent diagnostic oesophageogastroduodenoscopy (OGD), 88.5% had biopsies from multiple levels. Other relevant investigations included barium studies(8/27) and pH studies (4/27)

Significant symptomatic improvement resulted from the use of proton-pump inhibitors and topical steroids, either alone or in combination. Eighteen (18) children received PPI's alone with a 50% response and 15 received topical steroids only (60% response). Only the children treated with both medications (10) responded fully to the treatment. Other therapies utilised included exclusion diets (14 patients), leukotriene receptor antagonists (11 patients) and sodium chromoglycate (2 patients).

Conclusion:
Treatment of eosinophillic oesophagitis remains variable. Although PPI's and topical steroids may improve symptoms, best responses are achieved with combination therapy. A variety of allergy tests were performed but these were not evaluated by an allergy specialist and their value remains uncertain.

Nutritional issues of controversy in paediatric cystic fibrosis (CF) care; results of a European survey.

Chris Smith[1]; Helen White[2]; Dee Shimmin[3]; David Crook[4]; Paul Seddon[1];Assad Butt[1];
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[3]Belfast City Hospital; [4]University of Brighton

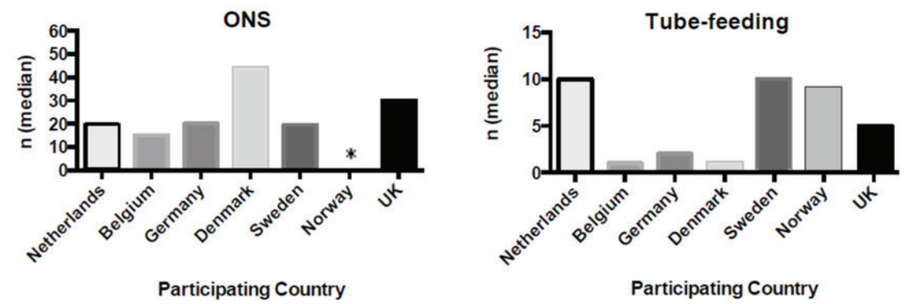
Introduction/Background:
Nutrition is a key aspect to CF care. Use of oral nutritional supplements (ONS) and tube feeding (TF) in CF remain controversial due to scarcity of RCT's. European guidelines on nutrition in CF give some direction on these aspects but are open to interpretation. Obesity is an emerging concern in this group of patients and as such there are limited data in the literature. Whilst some countries have national databases/ registries' these may be incomplete and a comparison between countries has not been done.

Aim:
To identify and compare the prevalence of 3 key nutritional aspects (ONS, TF and overweight) in CF patients across Europe.

Methods:
A questionnaire was developed and the content agreed with the chairs of the European Cystic Fibrosis Nutrition Group (ECFNG). This was circulated as a personalised email to all members of the group via the members email database. Questionnaires were then returned via email and the data was exported into spreadsheets and tabulated manually. Data collection took place over a 6 month period (February - August 2012).The questionnaire contained demographic data on the responder's clinic population looking at % prevalence of the following: use of ONS, use of TF and overweight status (defined as BMI >91st centile).

Results:
63 emails were sent out and a total of 39 Dietitians responded. The 39 responding dietitians were responsible for a total of 7333 patients (5163 adult, 2170 paediatric). 7 countries were represented providing paediatric data with 20 Dietitian responses: 10 UK (n= 1226), 3 Belgium (n=285), 2 Netherlands (n=70), 1 Germany (n=85), 1 Denmark (n=89), 1 Norway (n=55) and 2 Sweden (n=110)) Not all responders were able to give results for every section. Medians were used as histograms indicated uneven data distribution for several categories. Medians from the full paediatric data set (n=2170) were 20% prevalence of ONS use and 4.4% prevalence of tube feeding.

Graphs showing median % prevalence of paediatric CF caseloads using ONS or TF



*No data provided

The median for the full data for overweight status was 1.5%. Prevalence (medians) for overweight were as follows Netherlands, Denmark and Sweden 0%, Belgium 2%, and UK 5%. No data was provided for this category by responders from Norway and Germany.

Summary:
UK data is comparable with other European countries in terms of prevalence of ONS and TF use but had a noticeably higher prevalence of overweight CF patients.

Conclusion:
This is the first survey comparing and reporting topical dietetic issues to date. TF (1-10% of patient populations) indicates wide variation in use of this means of nutritional support across EU countries. Use of ONS (15-44%) emphasises their more routine use across patient populations. Obesity is an emerging area in CF which requires further investigation of genotype and phenotype to more fully understand the population who present with this new challenge. Although UK prevalence of overweight is higher than other countries this does not appear to be related to higher use of either ONS or TF. Responders were limited to ECFNG members therefore doesn't represent all European centres. This data provides a collaborative overview of practice and is an encouraging foundation for future projects.

The diagnostic value in paediatric small bowel assessment by wireless capsule endoscopy: a tertiary centre experience

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** Doctors Saliakellis and Fotis have contributed equally to this work.*

Introduction/Background:
Wireless capsule endoscopy (WCE) provides a method to assess small bowel pathology by filling the endoscopic gap between push-enteroscopy and ileocolonoscopy.

Aim:
The aim of this study was to assess the diagnostic value, tolerance and safety of WCE in paediatric patients referred to our unit.

Subjects and Methods:
This is a retrospective review of the WCE studies (PillCam SB, Given Imaging) that were performed during a 5.5-year period (May 2007-October 2012). Indications were confirmed/suspected IBD (n=114, 39%), obscure/occult GI bleeding (n=36, 12%), GI polyps syndromes/tumors (Peutz-Jegher's S., angiodysplasias, blue rubber bleb syndrome) (n=16, 5%), protein losing enteropathy(PLE) (n=15, 5%), recurrent abdominal pain (n=26, 9%), eosinophilic gastrointestinal disease (n=19, 7%), non-GI conditions with significant gut manifestations (autoimmune diseases, bone marrow transplantation, immunodeficiencies) (n=15, 5%) and other (coeliac disease, diarrhoea, failure to thrive etc.) (n=47, 16%).

Results:
291 children (140 male, median age 10.8 years (range: 6.5 months to 19.0 years) swallowed 102 capsules (35%), 188 were placed endoscopically into the duodenum under
General anaesthesia using an acorn-like device (n= 179)/"Roth net". 72 patients (25%) were under the age of 8 years. In 220 cases (76%) the WCE was seen in the coecum at end of recording (8 hours).The swallowed capsule did not leave the stomach in 6 patients. 2 patients retained the capsule, only one needing surgical removal of TI stricture with a normal contrast study pre WCE. Positive findings were observed in 184 (63%) of the studies of which 101 (34%) were diagnostic in terms of either establishing the diagnosis or altering the therapeutic approach of the patient. The diagnostic yield is highest in PLE (80%), polyposis syndromes/tumors (68%), Crohn's disease (39%) and bleeding (27%).

Summary and Conclusion:
Our experience - which is based on the largest cohort of paediatric patients and the youngest child undergoing WCE - demonstrates that with careful selection of patients, WCE is a useful and safe diagnostic modality in children with suspected small-bowel diseases.

Survey of practices in UK centres looking after paediatric patients on home parenteral nutrition

Sophie Robertson, Caroline Cole, Mark Beattie, Mick Cullen, Kim Novell, Akshay Batra. Paediatric Gastroenterology Department, University Hospital Southampton, Tremona Road, Southampton SO16 6YD

Introduction/Background
Home parenteral nutrition (HPN) is a specialist service and its use has increased significantly over the past decade. In 2010, 167 paediatric patients in 23 centres were identified as either receiving or in process of being set up to receive HPN (1). There are currently no national guidelines available. There is no published data reviewing practices in centres looking after patients on HPN in UK.

Aims
To capture variation in practice across centres looking after paediatric patients on HPN.
To share expertise and good practice amongst UK centres through description of practice.

Subjects and methods
We performed a survey of practices in all centres looking after patients on HPN. The survey was sent to all BSPGHAN members to ensure that all centres that look after patients on HPN received the survey. Responses were collected and collated electronically. The survey was developed by a multidisciplinary team including a Gastroenterologist, Pharmacist, Nurse Specialist and Dietician. Questions included free text spaces to allow explanation of responses where required. The name of the centre was included to allow combination and comparison of responses from within each unit.

Results
We received 22 responses to our survey from 15 different centres. Of these 4/15 (27%) had less than 5 patients on HPN, 5/15 (33%) had 5-10 patients on HPN and 6/15 (40%) had more than 10 patients on HPN.

Intralipid was the most commonly used form of lipid in children requiring PN for <28days. 9/15 centres used Intralipid for this indication, 3/15 used SMOF, 2/15 used ClinOleic, 1/15 used another lipid emulsion. In children requiring PN for > 28 days SMOF was used in 11/15 (73%) centres, Intralipid in 2/15, Clinoleic in 1/15 and other in 1/15. All (15/15) centres used SMOF for children with liver disease. 8/15 centres used lipid free days (days when no lipid was given in parenteral nutrition), of those 4/8 (50%) centres increased carbohydrate on fat free days. 9/15 centres used aqueous and lipid phase in a single bag, 6/15 used separate lipid and aqueous phase.

A third (5/15) of the centres did not use Taurolock. Of the 10 centres that used Taurolock, 3 used it in all patients, 7 used it in patients with multiple line infections and in addition two centres used Taurolock occasionally as part of treatment for line infections. 3 used Hep 100 and 7 used citrate. 8 centres reported choosing to withdraw Taurolock and 2 centres reported choosing to flush it.

11 centres responded to a question on the use of alcohol in central lines, of those 3 never used alcohol for lines, 2 used it for blocked lines, 4 used it for infection prophylaxis and 1 centre used alcohol for treatment of line infection.

Conclusion
This survey provides an important review of practice at 15 centres looking after patients on HPN. There is significant variation in practice at different centres in the UK. Collaboration between centres and reporting these details may help identify differences in outcome. There is need for a national guideline to be developed using the best evidence available, although the lack of high quality evidence in this field means that expert opinion is likely to form the basis of many aspects of care.

Beath SV, Gowen H, Puntis JWL. Trends in paediatric home PN nutrition and implications for service development. Clinical Nutrition 2011 Aug;30(4):499-502

WEDNESDAY SESSION THREE

INVITED SPEAKER ABSTRACTS

Professor Simon Murch

Dr Alastair Makin

Viral Hepatitis

Dr Suzanne Davison, Consultant Paediatric Hepatologist, Children's Liver and GI Unit, Leeds

Hepatitis B virus infection (HBV) and hepatitis C virus infection (HCV) lead to significant morbidity and mortality world wide due to their propensity to chronicity. Cirrhosis and hepatocellular carcinoma develop in 20% of those infected. The natural history is of slow progression of liver disease over decades: adults who have end stage disease, particularly due to HBV, are frequently those who were infected in infancy. There are however fundamental differences between HCV, a RNA virus, and HBV, a DNA virus, which impact on treatment efficacy and treatment aims.

This presentation will discuss the currently available treatment options for HCV and HBV, outline the NICE guidelines applicable to adults, the rationale and evidence for treatment in childhood, and also consider future developments in antiviral agents.

Quality Improvement

Dr Jenny Gordon, Research & Development Fellow, Quality Improvement Programme
RCN Institute, OX4 2JY

At present, prevailing strategies in healthcare rely largely on outmoded theories of control and standardisation of work. More modern and much more effective, theories seek to harness the imagination and participation of the workforce in reinventing the system' (Don Berwick, Former CEO IHI).

This session gives an overview of some key insights from the literature on large scale change and quality improvement. It will discuss common themes that may challenge what you think about what it takes to be successful in leading change and will end with some practical models and tools that may be useful in a range of settings to support using improvement methodology to make change happen.

WEDNESDAY SESSION FOUR

INVITED SPEAKER ABSTRACTS

Quality of life in children with IBD

Dr Adrian Thomas, Consultant Paediatric Gastroenterologist, Royal Manchester Children's Hospital

As in other chronic diseases children and young people with IBD are at risk of depression, anxiety, social isolation and altered self image which can all negatively affect health related quality of life (HRQoL) The World Health Organization defines Quality of life (QoL) as "an individuals perception of their position in life in the context of the culture and value systems in which they live in relation to their goals, expectations, standards and concerns". HRQoL has been defined "as a concept that includes the physical, emotional and social aspects of health perception and health functioning". Instruments to measure HRQoL can be generic or disease specific, generic instruments can compare HRQoL in different conditions and even in healthy children but may be too imprecise to reflect impairments in specific conditions or to assess response to therapy. Disease specific instruments are more sensitive to detect problems in specific conditions and to assess response to treatment. A review of HRQoL measures in children in 2001 revealed many problems including a lack of disease specific measures, a lack of measures for self completion, discrepancy between child & parent ratings and cultural inappropriateness of some measures.

The IMPACT questionnaire is a disease specific instrument (questionnaire) to measure HRQoL in children with IBD. It was originally developed by conducting interviews in Canadian children with IBD to identify areas of concern or worry. These issues were incorporated into a 96 item reduction questionnaire and a different group of Canadian children with IBD scored each item for importance & frequency. Items were ranked according to the sum of the scores and the 33 item IMPACT questionnaire was developed from the top ranking items. The item reduction questionnaire was subsequently ranked by children with IBD in the UK and a close correlation was seen between the highest ranking items in UK and Canadian children although most scores were higher in the UK children indicating a worse HRQoL.

HRQoL may be worse during a flare-up, in adolescents, certain personality traits those with negative coping strategies, a pessimistic outlook, sleep disturbances and poor social support networks.

Strategies employed to improve HRQoL have had variable degrees of success and include education, self-management, exercise, counselling, psychotherapy, antidepressant therapy, hypnotherapy, social support and of course treatment of the IBD itself which requires compliance with treatment.

Quality of life children with intestinal failure

Dr Sue Beath, Consultant Paediatric Hepatologist, Birmingham Children's Hospital, Birmingham. UK

What is quality of life

Good quality of life (QoL) can be an elusive concept, it is often defined as a state of well-being and happiness. It does not depend on absolute physical fitness and is intrinsically subjective eg some individuals who are disabled may report good quality of life and others who have minor restrictions may report having a terrible quality of life. Quality of life is perceived differently at different ages: young children desire to fit in with their peer group, whereas teenagers and young adults value independence. For example a 10 year old may be more concerned about how shiny her hair is and regard the side effects of chemotherapy with particular horror, whilst the teenager may be very concerned about the impact of a stoma on body image and a young adult will be most concerned about mobility.

Why measure quality of life

The perceptions of carers including parents can vary from the self assessment by the patient with the latter often being more optimistic and reporting better quality of life that their parent or doctor/nurse would expect [Wong C et al 2000, Sudan D et al 2004, Gottrand et al 2005]. For this reason it is essential that quality of life is taken in account in vulnerable people of any age, but especially children. The second key reason for measuring quality of life is to enable informed judgements to be made by clinicians and patients about choosing treatments. At the two ends of the spectrum are 1. a highly effective treatment in terms of disease eradication which is so damaging physically and or cognitively that a patient may decline it because of the impact on QoL, and 2. the treatment carries a high risk of mortality but the gain in quality of life is immense, but the patient may still decline treatment because of the short term risks. Although in many individual situations the quality of life gains are not clear cut, it is possible to use standardised instruments measuring quality of life to generate population data on quality of life and where the costs of treatment are incorporated a quality of life year can be calculated (QALY) [Brazier et al 2012].

How can quality of life be measured

The ideal quality of life measure (also known as instruments) should include generic and disease specific items [Wallender JL et al 2001]. The questions should have been developed jointly by professional and patient groups and include social and systemic functions. For rare disorders such as chronic intestinal failure the optimal approach is to use two of more questionnaires one of which is generic and the other is often an in-house development. The QoL instruments range from extensively validated questionnaires (eg. Short Form 30 (SF-30), General Health Questionnaire (GHQ-28), Child Health Check List (CHQ), the Euroqol) to in-house check lists, and semi-structured interviews [Emedo et al 2010]. In 1998, Sanders et al reported 66 quality of life instruments of which 14 are generic eg EuroQol; 16 psychological eg mental health index and 22 condition specific test. Evaluating quality of life in young children (less than 5yrs) remains a challenge and most studies rely on proxy ratings by parents and other adults, which are inherently unreliable. Tools using visual stimuli via a computer or a board game which do not require reading skills and no adult intervention and do not exceed attention span of 20-30 mins, have been developed but are not widely available [Christie D et al 2012].

Published studies of Quality of life in children with intestinal failure

There are very few studies in children, and most do not incorporate a generic tool, which makes comparison with other studies and with other disease groups difficult. Using an in house semi-structure questionnaire Emedo et al interviewed seven children aged 7-17yrs with long term intestinal failure and identified the following themes "Living with illness" and "Living a Restricted life" and explored social support, relationships and hopes for the future. This study provides insights into the experiences of children on PN and indicated that complications of the treatment and the underlying disease are more of a burden than the process of receiving the treatment itself. In another study of parents of children with intestinal failure the GHQ-28 was given to 11 parents of children on long term PN, and this showed that 7 parents had scores which exceeded the threshold for psychiatric morbidity [Wong et al 2000]. This adverse impact on parents' QoL has been observed by others [Unpublished observations Puntis JWL and Zamvar V]. Another study [Edge H et al 201] used parental satisfaction as a proxy indicator of improved quality of life after autologous bowel reconstruction – the end points measured included physical symptoms (bowel habit) and ease of caring for their child. Sudan et al carried out a direct comparison using the Child Health Questionnaire of children after small bowel transplant with children with renal failure and ordinary US school children and found that QoL in the small bowel transplant recipients was comparable to normal school children, although the parents' ratings were lower. A study comparing patients before and after small bowel transplant was done in adults [Rovera GM et al 1998] using the generic instrument Quality of Life Inventory - this suggested that QoL was rated similarly in the Home PN patients and small bowel transplant recipients although QoL improves with time in the small bowel transplant group.

Conclusion
Despite the difficulties of evaluating the perceptions of children and adolescents there are now many tools available to measure quality of life of children with intestinal failure on long term PN. For such a resource intensive treatment, it is unfortunate that this has not been performed more regularly in the past, but with alternative therapies for chronic intestinal failure and improved systems of management becoming available (e.g. bowel lengthening surgery and small bowel transplant multi-professional working) it is now imperative that such QoL studies take place.

Reading list
Brazier JE, Rowen D, Mavranezouli I, Tsuchiya A, Young T, Yang Y, Barkham M, Ibbotson R. Developing and testing methods for deriving preference-based measures of health from condition-specific measures (and other patient-based measures of outcome). Health Technol Assess. 2012;16:1-114.

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Dr Manu Sood

Quality of Life in children with constipation.

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Over fifty years ago, the World Health Organization defined health in its constitution as "a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity". This landmark definition broke from the myopic view that health is simply the absence of disease or pathology. Measuring health related quality of life (HRQoL) is important in clinical practice because physician based traditional measures of clinical success do not always coincide with patient perceptions. In chronic conditions, patients are more concerned about their HRQOL and potential disability than about their longevity. Moreover, poor HRQOL is a better predictor of increased health care use and resource utilization than reports of stool frequency or characteristics of impaired defecation. Thus, HRQOL is an important parameter to help define health care needs in a population. Childhood constipation and fecal incontinence can run a chronic course interspersed with periods of exacerbation in up to 15% of children with this disorder. It has been associated with psychosocial comorbidities, poor school performance and social maladaptation. Most experts think these are the consequence rather than a cause of constipation and fecal incontinence.

Instruments which measure HRQOL are increasingly being used to evaluate the outcome of therapeutic interventions and to assist with resource allocation. While disease-specific HRQOL instruments address the burden of a disease, they may not detect more general changes in a patient's daily functioning. Generic instruments to measure HRQOL are useful when comparing the impact of different illnesses on a patient population. While quality of life instruments have traditionally relied solely on parent perceptions, there has been a shift in recent years towards child self-reporting. In a case-controlled study by Youssef and colleagues 80 constipated children 5 to 18 years of age and their parents completed Peds QOL questionnaires at the time of their first visit to a Paediatric Gastroenterology clinic in the United States and these were compared to 46 control subjects from a primary care clinic. Children with constipation reported lower physical scores than did healthy children and their parents reported significantly lower scores than did their children with constipation. This was similar to an Australian study performed in patients attending a hospital gastroenterology clinic in which 51 children ages 8–18 years with an established diagnosis of slow transit constipation were compared to 79 control subjects. This study also used the Peds QOL to show that total QOL scores were substantially and significantly lower in constipated children. In a study of 57 Brazilian children ages 5–12 years with functional constipation and 29 children with functional fecal retention, CHQ-PF50 was used to evaluate QOL and compared with 314 age-matched controls. Decreased QOL scores were seen in both psychological and physical well-being in both the functional constipation and the functional fecal retention groups, compared to the non-constipated healthy children. Similar findings were reported in a study from the Netherlands in 114 children with fecal soiling.

We recently developed a disease specific instrument to measure HRQOL of children with constipation. We recruited 441 subjects from 5 centers across the United States. A Caregiver scale, consisting of 25 items and 6 subscales, was found to have good internal consistency which explained 61% of the variance associated with HRQoL. The 6 subscales of the Caregiver scale include; relationship with providers, effect on family, emotional adjustment, social adjustment, medical intervention and additional responsibilities. These subscales show a pattern of caregiver concerns which focus primarily on issues which affect the caregivers ability to successfully complete caregiving responsibilities (e.g., relationship with provider, managing additional responsibilities), and concerns regarding relationships within the family system (e.g., effects on family, emotional and social adjustments). The caregiver's perception regarding the child scale, consisting of 20 items and 5 subscales, also showed good internal consistency, and explained 62% of the variance associated with HRQoL. The 5 subscales of the caregiver's perception regarding child scale, child's social functioning, caregiver/child relationship, child's experience of pain, child isolation and effects on family, appear to represent aspects of the child's individual experiences of constipation (e.g., pain and isolation) and impairments across a variety of social systems (child's social functioning, caregiver/child relationship).

The emerging data suggests that in addition to objective endpoints such as frequency of bowel movements, HRQOL measures are important in determining treatment outcomes in children with constipation. It also seems conceivable that different laxatives might result in similar improvements in defecation but have different effects on QOL measurements. Such information might influence choice of treatments in the future.

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Managing Compliance in Paediatric Populations.

Dr Stewart Rust
Consultant Clinical Psychologist

Medical expectations regarding compliance / engagement with treatments often believe that lack of compliance is related to lack of understanding regarding the proposed treatment plan.

However a range of patient and lifestyle factors affect perceptions of medical treatments and further clinicians routinely fail when considering treatments to optimise health themselves.

Ideas will shared that encourage consideration of a wider range of factors when considering improving compliance and when to involve mental health professionals.

WEDNESDAY POSTERS

POSTERS OF DISTINCTION

Can gastrointestinal symptoms predict histological outcomes in food allergic children under the age of one?

Ru-Xin M. Foong[1], Eleni Volonaki[1], Neil J Sebire[2], Robert Dziubak[1], Osvaldo Borrelli[1], Keith J Lindley[1], Mamoun Elawad[1], Nikhil Thapar[1], Neil Shah[1]
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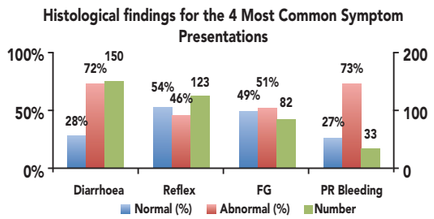
Background:
Approximately 2.1-4.2% of children between the ages of 0 and 3 years are diagnosed with a food allergy. It has been found that up to 60% of children who present with food allergy initially display gastrointestinal (GI) symptoms. The most common group of children affected are young infants. Endoscopy can be complex at this age and requires general anaesthesia. Clinicians are faced with the difficult task of assessing whether or not the procedure will be of value in the diagnostic process. At present, there are no studies that have looked at the gastrointestinal symptoms as a marker of histological outcome following endoscopy and biopsy.

Aims:
Our aim of this study was to see if there was correlation between the type of gastrointestinal symptoms in children diagnosed with food allergy and histological outcome from endoscopy.

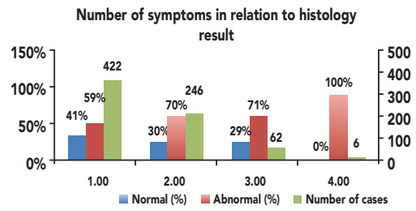
Methods:
All patients under the age of 1-year-old referred to our tertiary paediatric gastroenterology centre during the period of June 1987 to August 2007 who had an endoscopic procedure (oesophageal gastroduodenoscopy (OGD), colonoscopy or both) were included. The clinical symptom presentation and histological outcomes were reviewed.

Results:
A total of 736 endoscopic procedures were performed in food allergic children over this 10-year period. We analysed the data for the four most common symptoms (diarrhoea, faltering growth (FG), peri-rectal bleeding (PR bleeding) and reflux/vomiting) and their combinations with one another. We classified the histological findings as either "Normal" (normal biopsy) or "Abnormal" (the presence of any positive histology including eosinophils, inflammation or other histological findings). The 10 most common symptom combinations were reviewed. The combinations of the four main common symptoms (Diarrhoea, Reflux/Vomiting, FG and PR bleeding) were responsible for 575 of 736 cases (78.1%). Ten of the most common combinations were: Diarrhoea 150 (17%), Reflux 123 (17%), FG 82 (11%), Diarrhoea + FG 77 (10%), Reflux + FG 47 (6%), PR Bleeding 33 (4%), Diarrhoea + PR Bleeding 21 (3%), Diarrhoea + Reflux + FG 18 (2%), Diarrhoea + Reflux 12 (2%), Diarrhoea + Reflux +PR bleeding 6 (1%). In patients who presented with diarrhoea or PR bleeding alone, 28% and 27% of the patients had normal histological findings, respectively. Contrastingly, in patients who presented with reflux/vomiting or FG in isolation, 54% and 49% of the patients had a normal biopsy result (Graph 1). We also found evidence that the greater number of symptoms a child presented with the more likely they were to have an abnormal histological result for any endoscopic procedure performed (Graph 2).

Graph 1



Graph 2



Conclusion:
Diagnosing food allergy based on clinical symptoms alone especially in infants under the age of one is complex and challenging for the clinician. This data suggests that for children who present with diarrhoea or PR bleeding, having an endoscopic procedure higher chance of finding abnormal pathology in the biopsies taken which may be beneficial in the identification of food allergy. The evidence is not as convincing for the symptoms of reflux/vomiting and FG in isolation where there is a 50% chance of the child having a completely normal biopsy. Our data also shows that children who present with a combination of symptoms are more likely to benefit from having an endoscopic procedure to aid in the diagnostic process of food allergy than in children who present with a single symptom.

Cholangitis and outcome after Kasai portoenterostomy for biliary atresia.

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Aims:
The diagnosis of cholangitis following Kasai portoenterostomy (KP) for biliary atresia is difficult to prove and antibiotic treatment is often commenced on the basis of a high index of suspicion. This study aims to examine the clinical features of children treated as cholangitis, their response to antibiotics and need for subsequent liver transplant (LT).

Methods:
A retrospective case note review of 118 consecutive children with biliary atresia who underwent KP at a single centre between June 1994 and December 2011. Cholangitis was defined as "definite" in the presence of unexplained fever, deterioration in liver function tests (LFT's) with positive blood cultures or "probable" if unexplained fever accompanied either deranged LFT's and/or pale stools but without positive blood cultures. Children without fever but with deteriorating LFT's and/or pale stools were defined as having "possible" cholangitis. Response to antibiotics was defined as improvement in symptoms of cholangitis by the end of treatment.

Results:
One hundred and eighteen children had a KP and the data was available for examination on all patients. The median age at KP was 51 (10 - 144) days and median follow up after KP to last clinic visit, death or LT was 24 (10 -210) months.

80/118 infants (68%) cleared their jaundice (Bilirubin<20mmol). Thirty six of these children had 77 episodes of cholangitis (5 definite, 46 probable, 26 possible) and there was a response to antibiotics in 66/77 (86%) episodes. Ten children subsequently came to transplantation at a median of 23.5 (15-24) months post KP. This was precipitated by unresponsive cholangitis in 5 cases.

38/118 infants did not clear their jaundice post KP. One died 9 months later from cardiac causes and was never treated for cholangitis. One died following the second episode of cholangitis that was unresponsive to antibiotics. The 1st episode was included in this study. Of the rest 18 were treated for 31 episodes of cholangitis (7 definite, 17 probable, 7 possible). 19/31 (61%) episodes responded to antibiotics and all subsequently underwent a LT at a median age of 6.75 (4-32) months post KP.

Overall response to antibiotics was 89% (8/9) with definite, 82% (52/63) with probable and 6% (31/36) with possible cholangitis.

Conclusions:
In our experience most children who have cleared their jaundice post KP respond to prompt antibiotic treatment for cholangitis and do not come to early transplantation. Treatment of possible cholangitis still results in improvement in 86% of cases. A high index of suspicion is therefore justified.

STAMPing out poor nutrition. An audit of the nutritional status of paediatric outpatients.

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Background:
There is a recognised need to screen for malnutrition in paediatric patients and a number of tools have been developed. One such tool developed in the UK is the Screening Tool for Malnutrition in Paediatrics (STAMP) (1) which is designed and validated for inpatient settings. Several large studies throughout Europe have described the prevalence of malnutrition amongst hospitalised children but comparatively few have described the incidence in out patient settings where the population are more likely to suffer from chronic disease and therefore long term nutritional issues.

Aim:
To screen all medical out patient attendees at our centre for nutritional risk and describe the incidence of status by BMI Z score and STAMP category.

Methods:
We screened every medical patient attending the OP department at the RACH using the STAMP tool. The data collection took place over one week in January 2012. The screening was completed by 2 assessors and the weighting and measuring of children was consistently completed by a core group of experienced outpatient nurses. The main value for analysis was BMI for age z score, in accordance with WHO standards, a z score of <-2 was considered underweight, >1 was considered overweight, and >2 obese. STAMP scores were also calculated using height and weight centiles, subjective appetite rating and diagnosis.

Results:
Information was gathered from 18 condition clinic types, those with very small numbers, were grouped into the general paediatrics group, leaving a total of 11 clinic groups. The total population screened was 226 (male:female, 119:107), mean age 7.06 years, range 0.1-18.8. The average BMI for age z score over the whole population was 0.54. A total of 6 children (2.7%) were underweight, 70 were overweight (30.9%), and 30 were obese (13.3%). There was no significant difference between the mean BMI z-score of clinic groups.

The STAMP score was calculated for each patient, 41.2% of all patients were found to be medium risk and 5.8% high risk. Certain clinics had far higher proportions of patients in these groups, for example: Cystic fibrosis 73% medium, 27% high risk (n=11), and Cardiac 89% medium, 7% high risk (n=28), this can be compared to the general clinics where 25% were medium and 2.7% high (n=72).

Discussion:
This investigation showed a low proportion of underweight children attending our hospitals' outpatient department however, we recognise the importance of identifying them as they have a well documented risk of morbidity and mortality. The assessors found the screening tool quick and easy to use. A limitation is its lack of validation in outpatient setting. The proportionally large number of overweight and obese children identified in the exercise raises the question of the need for routine screening for obesity, also known to have significant morbidity in children and adults. Relying on clinical impression alone has been shown to be highly inaccurate both in the identification of over and underweight patients. Tools to draw health professionals towards care and attention surrounding growth assessment, if quick and practical to undertake are of importance. These however are not a replacement for sound clinical assessment including careful plotting on appropriate growth charts. Higher incidence of "at risk" identified by STAMP compared to the lower incidence of clinically underweight by BMI highlights the importance of a multi dimensional tool to provide a more comprehensive assessment of nutritional status.

Conclusion:
This tool, if formally validated would be useful in the outpatient setting to prompt clinicians to undertake full nutritional assessment, especially those whom this may not be their primary focus. However, any such future tool in the outpatient setting would ideally identify both malnutrition and obesity.

Use of an electronic communication system for patients on parenteral nutrition at home.

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Background
Children with chronic severe intestinal failure that does not respond to treatment are discharged home on treatment with intravenous nutrition. Care at home is complex with not only the need for the administration of parenteral nutrition (PN), but also management of the underlying gastrointestinal disease. The child's care is managed by a team specialising in paediatric gastroenterology (including nurse specialist, dietitian, doctors), a general paediatric team at the child's local hospital, community nursing team, GP, and most importantly the parents. Effective communication is essential. Since November 2010 we have trialled the use of an electronic communication system accessible to the family and all health professionals involved in the child's care who would like to use it.

Aim
The aim of this study was to review the electronic communication system used by the families and health professionals to converse with each other.

Subjects and Method
We retrospectively analysed all e-mails and discussions via the electronic system to see which healthcare professionals the family contacted, how often and the theme of each message. We also invited the families to complete an online survey or be interviewed in clinic using 10 pre-set questions to gain qualitative feedback on the electronic system.

Results
The families of 33 children with severe intestinal failure aged 2 - 18 (mean age 8) years signed up to the electronic system. 1572 messages were sent about 819 different enquiries. A new message thread was sent from families to professionals on average 1.5 times per month. The professionals contacted were the nutrition nurse specialists (459/1572), 29%, doctors (299/1572), 19%, administrator 18%, community and local hospital 18% and dietitians 10%. The three most frequent message themes were appointments and admissions (170/819), 21% followed by general advice (126/819), 15% and laboratory tests and scan results (125/819) 15%.

In response to the questionnaire 11/15, 73% families said they found the system 'extremely useful' with the discussion section the most helpful. 12/14, 86% of respondents said that the system had improved the management of their child's care.
The system has also helped the Clinical Nurse Specialists to organise and structure their workload more effectively.

Summary & Conclusion
Overall there was overwhelmingly positive feedback from parents, patients and healthcare professionals who used the electronic system, finding it an effective communication and information sharing tool.
Our impression is that the use of electronic communication has empowered families to make informed decisions regarding their child's health. We have found that electronic communication has provided ease of access and improved the connection between the family of the patient at home and health care professionals.

WEDNESDAY POSTERS

An exploration of perceptions and views of the key stakeholders about what constitutes successful transition for young people with liver transplants.

Jessica Arkley[1], Laura Elwell[1, 2], Carla Lloyd[1], Jo Wray[5], Janet McDonagh[1, 2], Patricia McClean[4], Patrick McKiernan[1], Catherine Arkley[6], David Adams[2, 3], James Ferguson[3], Deirdre Kelly[1, 2].
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Background:
The success of paediatric liver transplantation in transforming survival rates for patients with end stage liver disease means increased numbers of adolescent patients are transferred to adult services. An effective transition process is essential when patients are preparing to transfer from paediatric to adult care (Fredericks, 2011). The determinants of successful transition remain unexplored with liver transplant recipients. The information from this pilot study will help determine the important risks and protective factors in order to improve services and long-term outcomes for patients.

Aims:
1. Explore the experiences and views of patients, parents and healthcare professionals from paediatric and adult liver units at Birmingham Children's Hospital (BCH), University Hospitals Birmingham (UHB) and the Leeds Teaching Hospitals (LTH) about what constitutes successful transition.
2. Quantify the occurrence of non-adherence, graft loss and rejection in paediatric liver recipients from two national paediatric liver units in the UK (BCH and LTH), who have transferred to adult care between July 2006 and September 2011.

Subjects and methods:
Approximately 25-30 patients and parents and 12 health professionals will be interviewed to ascertain views and perceptions of transition. Interviews will be analysed using Interpretative Phenomenological Analysis (IPA). In addition, a retrospective case note review will be conducted to quantify the occurrence of non-adherence and graft survival in transferred patients.

Results
Case note review
A review of 109 case notes has highlighted that at the time of transfer, the median age was 18.6 (range 16.1-23.0) years. 78.9% of patients were on their first graft, 18.3% on their second, 1.8% on their third and 0.9% on their fourth. 9 (8%) patients have died since transfer. 2 patients died in the first year, 2 died in the second year, 4 died in the third year and 1 died in the fourth year post transfer.

Qualitative interviews
Twenty-nine interviews have been conducted to date and have been transcribed verbatim. IPA is currently ongoing.

Summary and conclusion:
The preliminary results show a reasonable outcome after transfer to adult-care. Patient deaths were highest between years 1 and 3 following transfer, suggesting this may be a particularly crucial period for young people in the transition process. Further analysis of the case note data is needed in order to quantify the occurrence of non-adherence, graft loss and rejection. It is anticipated that the results from the study will provide important new insights into the transition process and be instrumental in changing the management pathways for future patients and improving long term outcomes of young people with a liver transplant.

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Can Patients with Suspected Coeliac Disease with Very High tTG Levels and Positive EMA be Diagnosed without OGD and Biopsy? A review of cases at Alder Hey Children's Hospital over the past 5 years

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Coeliac Disease is an autoimmune disease affecting about 1% of the population, caused by gluten intolerance resulting in an enteropathy of the small bowel. A correct diagnosis is very important to prevent complications of the disease. It is important however to avoid an incorrect diagnosis, as a gluten free diet imposes lifelong restrictions on the patient. Diagnosis is traditionally confirmed by OGD and duodenal biopsy; however, recent research has suggested that serological tests including tTG and EMA have a sensitivity and specificity of >95%. Recently published ESPGHAN guidelines recommend that symptomatic patients with tTGs over 10 times the upper normal limit and a positive EMA do not require an OGD and duodenal biopsy for confirmation of the diagnosis. This study reviewed patients referred to Alder Hey Children's Hospital over the past 5 years with elevated tTGs and reviewed the histology reports to see whether a diagnosis of Coeliac Disease was confirmed. 130 patients were identified, 32 of whom were symptomatic and had a tTG of over 10 times the upper normal limit. 31 of these (96.9%) were diagnosed with Coeliac Disease following OGD and duodenal biopsy. There were a number of limitations to this study including a paucity of information of some patients referred from district hospitals and GPs and the fact that most patients hadn't been tested for EMA or HLA. However, the results are in keeping with recent research and ESPGHAN guidelines, therefore a recommendation is made that a multi-centre study be carried out and consideration be given as to a change in practice regarding the diagnosis of coeliac disease.

Duodenal Haematoma - a rare complication of upper gastrointestinal endoscopy

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Introduction:
The occurrence of intramural duodenal haematoma (IDH) after small bowel biopsy is rare & has only been reported in 18 children (1). Risk factors are thought to include the fixed position of the duodenum in the retroperitoneum, the relatively profuse submucosal blood supply of third part of the duodenum & abnormalities of haemostasis & coagulation. The literature also suggests that malnourished or growth retarded children may be at risk (2)

Aim:
We aim to highlight the risk of intramural duodenal haematoma formation after small bowel biopsy and the relative increased frequency of this complication in the haematology/oncology population by means of a case report & literature review.

Method / Case report:
We describe the case of a nine month old infant with disseminated Langerhans cell histiocytosis who underwent elective re-staging upper GI endoscopy following 3 cycles of cladrabine & cytarabine. Upper & lower GI biopsies obtained without event at the time of initial diagnosis at 10 weeks of age demonstrated extensive infiltration. Platelet count & coagulation profile were within accepted parameters at the time of both initial & follow up endoscopy. The endoscope was passed without difficulty to the third part of the duodenum. Best practice was followed with biopsies being taken close to the tip of the endoscope with limited projection of the biopsy forceps and no biopsies were taken in the proximity of the ampulla of vater (3) No excessive bleeding was noted from the biopsy sites & no macroscopic abnormality of the stomach or duodenum was identified.
The patient represented 48 hours later with bilious vomiting. A fall of 3gm/dl in haemoglobin was noted on re-admission screening bloods. Ultrasound of the abdomen demonstrated an intramural haematoma of the third part of the duodenum which was confirmed to be partly obstructing the duodenum by water soluble contrast. There was no clinical or biochemical evidence of an associated pancreatitis. The patient responded to conservative management (bowel rest, parenteral nutrition & nasogastric suction). Full enteral feeding was established within 14 days.

Summary / Conclusion:
The incidence of IDH following duodenal biopsy at upper GI endoscopy is unknown but an estimate of 1:1,250 has been proposed (3). Of the reported cases the majority have occurred in children & young adults. A significant proportion of these cases have been in children post oncology diagnosis or bone marrow transplantation. Surgical management has been required in a small proportion of patients following failure of conservative management.
We propose that every patient presenting with abdominal pain & vomiting within 48 hours of endoscopy should be examined for IDH & it's complications. We recommend that IDH is cited as a potential complication of endoscopy when written consent is obtained. Interestingly, our patient developed IDH despite normal haematological parameters and absence of disease infiltration of the gut at the time of biopsy. We hypothesise that the increased risk reported in the literature in this population may be due to persistant changes in gut vascularity post malignant infiltration, with a possible contribution from post chemotherapy retroperitoneal fibrosis. We suggest that a high index of suspicion is maintained for this complication particularly in the oncology & post bone marrow transplant population and that patient information is given accordingly

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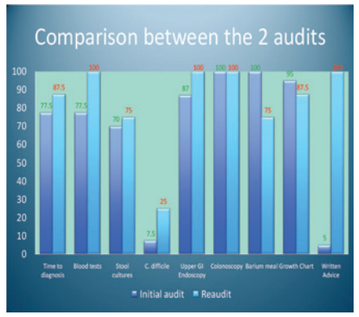
Evaluation of parent satisfaction of shared care within a regional clinical network.

Dr Siba Prosad Paul, Yeovil District Hospital, Previously GWH, Swindon
Dr Janet King, Great Western Hospital, Swindon
Professor Bhupinder Sandhu, Bristol Royal Hospital for Children



Introduction:
The BSPGHAN guidelines on the management of Inflammatory Bowel Disease (IBD) were published in 2008 to streamline the diagnostic work-up and management pathways. The South-West of England paediatricians have developed a regional clinical network whereby children, suspected of having IBD travel to the specialist centre based in Bristol for full diagnostic work-up. The rest of the care is managed by the local hospital paediatricians with an interest in gastroenterology, with advice from Bristol and a joint three monthly out patient clinic in Swindon. Children with very severe disease or needing surgery are dealt with by Bristol team. The effectiveness of this model was audited pre and post the introduction of BSPGHAN guidelines and found to be improved after the guidelines (Fig1).

Aim:
The aim of this pilot study is to assess parent and patient views as to quality of service being provided within the Swindon/Bristol regional network.

Subjects and methods:
All children and young people aged <16 years who had been diagnosed with IBD between 2010-2011, after endoscopy and other workup in Bristol, and managed by the shared care services by Swindon and Bristol teams, were identified. There were13 cases. A telephone questionnaire survey was designed which consisted of 12 questions and a section for free comments. Questions were aimed at length of time to diagnosis of IBD, information sharing pre and post diagnosis, satisfaction with the services and preferences re further follow-up care.

Results:
9/13 (69%) responded to the telephone survey, 3/13 (23%) had moved and 1/13 (8%) could not be contacted by telephone. Out of 9 patients, 4 had Crohn's disease, 3 Ulcerative colitis and 2 Indeterminate colitis. 4/9 (45%) parents felt their concerns were addressed adequately initially at Swindon whereas all parents were satisfied with the services provided at Bristol when they went for endoscopy and work up. 6/9 (67%) parents felt they were satisfied with the expertise available locally for post-diagnosis management. However 8/9 (89%) parents were happy with follow-up care by the joint care services at Swindon.

Parent's stated that 'they should be taken seriously when they recurrently report something being wrong with their child at initial presentation'. There were also requests for information leaflets at their GP surgeries/Swindon. 2/9 (22%) wanted more information about pharmacological agents and 3/9 (33%) felt more dietician input would be helpful. Nearly all parents felt happy with the shared care services and felt they can easily get in touch with the nurse specialist. Clinic timings were good and adequate for discussion and there was good support during investigations.

Summary and Conclusion:
This pilot study of one centre within a larger SW Clinical Network demonstrates that joint care provided by this model not only leads to care more concordant with BSPGHAN guidelines but is appreciated and valued by parents. There is scope for further improvements such as time to 1st referral to hospital to minimise time to diagnosis, better information sharing and more dietician input. This pilot study provides a template for ensuring and improving parent/patient involvement and satisfaction and there are plans to modify the questionnaire taking into account any suggestions for improvements and roll it out.

Infant weaning advice and practice: A population based study on weaning practices

Dr B F Gurreebun, Dr G Raptis*,Dr A. Date Consultant Paediatrician** , Dr EB Odeka Consultant interest in Gastroenterology *** (*The Royal Oldham Hospital, **Tameside General Hospital,*** Royal Manchester Children's Hospital & University of Manchester)*

Introduction/Background:
National recommendation in England is to delay weaning until 6 months. Though this has been widely debated, there is evidence that compliance with national guidelines is poor.

Aim:
We aimed to study weaning practice locally, to assess knowledge of and compliance with national guidelines and to look at sources of information available on weaning and the parents' need for further information.

Subjects and methods:
We designed a questionnaire that was randomly distributed to parents attending the Paediatric outpatient department, neonatal unit and the antenatal clinic at Royal Oldham Hospital. The local ethics committee approved this study. Several questions on weaning, information sources, need for further information and awareness of guidelines were asked.

Results:
162 questionnaires were returned. 54 % of parents breastfed, 33.1% of whom breastfed for over 6 months. Only 5.2% weaned at or after 6 months, 38.8% weaned between 4-6 months and 56 % weaned before 4 months. Rice was used by 73 % for weaning. 79.1 % of parents had received information on weaning. 54 % expressed an interest in further information, 54 % of which wanted information on weaning age. 50.6 % obtained information from family and friends, 60.7 % from antenatal clinics and midwives and less than 30 % from health visitors. Only 33.1 % of parents claimed to be aware of government guidelines and this was significantly less in ethnic minorities. Parents receiving weaning information were more likely to be aware of national guidelines (38% compared to 16%, p= 0.02). Those aware of the guidelines were less likely to want further weaning information (30 % compared to 65%, p=001) and more likely to wean later (76% of those weaning at 3-4 months were unaware of guidelines, dropping to 64% at 4-5 months and 40% at 5-6 months, p=0.02).

Summary:
Our data illustrates poor compliance with national guidelines. A large proportion of parents requested more information on weaning. A large number received weaning information from family and friends, as previously shown, and from midwives and antenatal clinics – an important finding which should be built upon. Fewer parents obtain weaning information from health visitors, a source which can be improved. We also show that the provision of weaning information and parents' awareness of the guidelines does affect weaning practice.

Conclusion:
We conclude that there is a demand for further weaning information from parents and an urgent need to improve the provision of this information. We recommend that midwives and health visitors should play a key role in this in order to improve compliance with national guidelines.

Inflammatory Bowel Disease Quality Improvement Project, progress after two years

Fernandez E, Charlton C, Nottingham Children's Hospital, Consultant Paediatric Gastroenterologist, IBD Quality Improvement steering group

Introduction
The IBD Quality Improvement Project (IBDQIP) is a national project involving adult and paediatric services from across the UK, that aims to support services in achieving the quality of care set out in the IBD standards.

Methods
Services meet as a multidisciplinary team to complete a web-based self-assessment (www.ibdqip.co.uk) that benchmarks their care in 4 different main domains (patient experience, clinical quality, organisation and choice of care, research education and audit). Results are presented using a hierarchical grading system in performance dashboards. Sites then are encouraged to prioritise action plans and utilise the wide range of resources available to support service improvement.

Results
Engagement has grown steadily, with a 78% increase in the number of paediatric sites participating in the last year.

75% of paediatric sites in 2012 managed to meet and discuss results with a multidisciplinary group of service members. There were significant improvements in quality of care for both adult and paediatric services who have been participating for two years.

Sections of IBDQIP with significant improvement in grades between 2011 and 2012		
	% of sites that improved	p value
Involvement of patients in service improvement	54.5%	0.016
Information and support for patient organisations	72.7%	0.007
Access to Nutritional support and therapy	63.6%	0.010
Surgery for IBD	36.4%	0.047
Referral of suspected IBD patients to services	72.7%	0.005
Training and education	36.4%	0.047
Service development	36.4%	0.047

The table above shows that many sites showed improvement in grades between 2011 and 2012 and also showed significant improvements in certain individual statements.

Comparison of number of 'yes' answers for 11 sites between 2011 and 2012	
Statement	p value
>30%of IBD patients are seen within 24 hours of admission, by a Paediatric IBD specialist or a Paediatrician with interest in gastroenterology working in conjunction with a paediatric IBD specialist, who is available for immediate telephone advice.	0.016
For the physically mature patient, who has completed their growth, is emotionally mature and without psychological or educational problems, investigations may be undertaken locally by the adult service. This should be by a Gastroenterologist experienced in the management of adolescents with IBD in discussion with a Paediatrician with appropriate expertise in gastroenterology.	0.016
All children with Ulcerative colitis patients who had the disease for more than 10 years should be formally identified and a surveillance plan made (with adult services)	0.031

Conclusions:
Growing number of sites are participating in IBDQIP data entry
Some evidence that sites are benefiting from involvement
This process encourages team discussion and patient/ carer involvement in services

Not inflammatory bowel disease after all…

Dr Lucy Backhouse, Dr Sandhia Naik, Dr David Rawat, Dr Binutha Bharathan, Dr Mike Millar
Whipps Cross Hospital, Whipps Cross Road, Leytonstone, E11 1NR

A case study of a 14 year old Orthodox Jewish girl who was referred to a tertiary paediatric gastroenterology centre for investigation of possible inflammatory bowel disease. She presented with a 2 month history of weight loss and fever with high inflammatory markers (CRP 114), significant anaemia (haemoglobin 6.1) and hypoalbuminaemia (albumin 27). She had also been experiencing abdominal pain and increase in stool frequency. Endoscopy was normal and subsequent MRI showed multiple, small splenic abscesses. There was no history of recent travel apart from a trip to Israel 2 years previously.

All cultures were negative and she was empirically treated with IV antibiotics. After 3 weeks she developed a pleural effusion and samples of the pleural fluid were analysed. A positive result for brucella species was found using 16s rRNA PCR analysis. This has since been retested and original results have been confirmed. Appropriate antibiotic treatment was commenced following this result.

Splenic abscesses are uncommon in the paediatric population and mostly results from disseminated infection. Surgical treatment is often necessary either with splenectomy or drainage of the abscesses.

Brucellosis is the commonest zoonotic infection worldwide. More than 500 000 new cases occur every year however the incidence in the United Kingdom is only 0.3 cases per million. Complications of the infection include arthritis, sacroiliitis, encephalitis and endocarditis. Treatment aims to shorten the duration of symptoms, avoid complications and prevent relapse. A meta-analysis of treatment options has determined that dual or triple antibiotic therapy including an aminoglycoside is the most effective in treating human brucellosis.

It is important to consider all possible diagnoses when investigating non-specific symptoms. The in depth microbiology analysis was key in the final diagnosis enabling correct treatment of the unusual condition.

Perforated duodenal ulcer in a 14 year old; A case presentation

Dr Holly Evans, Dr Sian Kirkham, Dr Kiron Kumar
Paediatric Department, QMC, Nottingham

The occurrence of peptic ulceration in children had been regarded as a rarity, but in recent years the condition has been diagnosed with increasing frequency largely due to a more general appreciation of the existence of the problem. It is still however a relatively uncommon condition in children with an estimated frequency of 1 case in 3000 hospital admissions. Usually classified as either primary or secondary depending on the underlying pathology. Primary ulcers occur with almost equal frequency in the duodenum or stomach and tend to be solitary with a male prevalence. Secondary ulcers are usually gastric and often multiple and affect younger children. Initial presentation is usually with GI bleeding.

We present the case of a 14 year old boy who presented with sudden onset epigastric pain having been found collapsed in the street. He also reported a history of several months of intermittent PR bleeding. He was found to have a perforated anterior wall duodenal ulcer at laparotomy which surgeons felt had perforated one or two weeks earlier. He was commenced on H. Pylori eradication treatment and 6 weeks later underwent endoscopy with a subsequent diagnosis of Crohn's disease being made.

He has made a complete recovery from the perforated ulcer and is currently well.

Profound Hypoalbuminaemia?: Consider protein losing gastropathy

Dr Sheela Sanmani, ST3 Paediatrics; Dr Lesley Peers, Consultant General Paediatrics; Sheffield Children's HospitalDr Fiona Shackley, Consultant ID and Immunology; Dr Prithvi Rao, Consultant Gastroenterologist; Dr Priya Narula, Consultant Gastroenterologist;Sheffield Children's Hospital

Case:
A 3 year old Caucasian boy was admitted with a six day history of puffiness of face, vomiting, lethargy, abdominal distension and swollen legs, preceded by a coryzal illness. There was no significant family or past history. On examination he had peri-orbital puffiness, pedal oedema and no pallor, jaundice or hepatosplenomegaly. Initial laboratory tests revealed hypoalbuminaemia(12g/L), normal clotting profile, hyponatremia and mild transaminitis with a normal bilirubin. Nephrotic syndrome was clinically suspected, but further tests included normal urinary protein-creatinine ratio. He had a normal coeliac serology but significantly low immunoglobulins (IgM 0.2g/L, IgA 0.3g/L, IgG 1.5g/L). He was fluid restricted with strict monitoring of fluid balance.

An abdominal ultrasound showed mildly enlarged spleen with small amount of ascites and normal dopplers. With synthetic liver function being normal and no protein loss identified from the renal tract, protein loss from his gastrointestinal (GI) tract was considered. He then underwent an upper GI endoscopy which revealed linear erosive oedematous folds in stomach with relative antral sparing. A clo test was negative. Gastric biopsy, serology and urine samples tested strongly positive for CMV PCR. He was reviewed by the immunology/infectious diseases team and as he was clinically improving no CMV specific treatment was indicated. GI histology showed hypertrophied gastric foveolae with acute and chronic inflammation and no evidence of H.pylori. A diagnosis of Menetrier's disease was made and the patient was discharged on proton pump inhibitors.

At 8week follow up he remains clinically well. His oedema has resolved and he is asymptomatic. His albumin levels and immunoglobulins have gradually normalised.

Discussion
Menetrier's disease is a rare hyperproliferative protein losing gastropathy of the gastric foveolar epithelium. Paediatric Menetrier's has an abrupt onset with spontaneous regression and has been linked to infection with CMV and H.pylori. This case highlights that in children presenting with oedema, hypoproteinaemia and hypoalbuminaemia without a renal or hepatic cause, protein losing gastropathy such as Menetrier's disease should be considered in the differential diagnosis. Paediatric Menetrier's disease runs a benign course with good prognosis on conservative management.

Retrospective study of nutritional intervention in primary intestinal lymphangiectasia

Dr Kiran Kumar; Dr Charles Charlton (consultant in paediatric gastroenterology); Dr Sian Kirkham (consultant in paediatric gastroenterology);
Queens Medical Centre, Nottingham University Hospitals, Derby Road, Nottingham, NG7 2UH

Background:
Primary intestinal lymphangiectasia is a rare digestive disease and few studies have focused on the therapeutic effect in Primary intestinal lymphangiectasia patients. This retrospective study was undertaken to evaluate nutrition-oriented intervention in children with Primary intestinal lymphangiectasia.

Methods:
Four children with Primary intestinal lymphangiectasia were studied. Their medical records, anthropometric measurements and blood tests performed during follow-up were reviewed.

Results:
During hospitalisation, all four patients were subjected to diet intervention. Parenteral nutrition support was also given to three of them. Clinical symptoms and laboratory parameters of the patients were significantly improved at discharge. After discharge, the patients continued diet control, two of whom received Parenteral nutrition during subsequent admission. All patients are managed via out patient follow up. Duration of follow up is from six months to seventeen years and they all kept in a stable condition without symptoms relapse. Weight, height and body mass index for age were normal during the follow up, as also biochemical parameters and immunoglobulin concentrations.

Conclusions:
Nutrition therapy is effective as a valid and safe therapeutic management for Primary intestinal lymphangiectasia patients. No growth failure was observed in these four children after the therapy, but they are still at risk of nutrient malabsorption. Therefore, they need long-term, regular monitoring and intensive nutritional care.

THURSDAY INVITED SPEAKER ABSTRACTS

SESSION I

Advances in the management of paediatric acute liver failure

Prof Anil Dhawan MD FRCPCH, Director, Paediatric Liver GI and Nutrition Centre,
King's College Hospital, London
Anil.dhawan@nhs.net

The lecture will focus on the developments over the last 10 years in the diagnosis and management of acute liver failure in children. The lecture will focus on the following points-

Consensus on definition –
Paediatric hepatologists have now agreed a functional definition of acute liver failure in children, which uses INR as an objective test and doesn't require encephalopathy to be a pre requisite as in adults. The agreed definition is "Multisystem disorder in which severe acute impairment of liver function (INR>2) with or without encephalopathy (INR .1.5) occurs in association with hepatocellular necrosis in a patient with no recognised underlying chronic liver disease.

Epidemiology - Children under one year of age have the highest incidence while the second peak being in adolescents. Slight increase in new cases is noted during winter months.

Diagnosis - Establishing the aetiology is important but following factors make this task difficult. Being a rare disease with a rapidly evolving course, presentation to a hepatology centre at variable time in the illness course with heterogeneous aetiologies. Specialist (long turn around time) and invasive investigations, limited by coagulopathy usually complicate and delay establishing the diagnosis. Liver biopsy particularly is not very helpful and could be counterproductive.

Prevention of ALF - Last decade has seen decrease in incidence of few conditions leading to ALF namely; Hepatitis A and B, Halothane induced hepatitis, Reye's Syndrome and Neonatal Hemochromatosis (GALD). Paracetamol overdose (accidental and suicidal) is an avoidable cause of acute liver failure

Medical Management - The lecture will address areas of neuro-protection, renal replacement therapy, sepsis and role of liver assist devices while waiting liver transplant or natural recovery.

Surgical Management - Liver transplantation is the only proven treatment for children with ALF. Prognostic models of spontaneous liver survival Vs. death, need for disease specific models and surgical advances like auxiliary liver transplant which could avoid whole liver replacement and subsequent need for life long immunosuppression in about 70% of patients who survive the operation.

Hepatocyte Transplantation - Given the success of auxiliary liver transplantation, preliminary data on human hepatocyte transplantation will be presented. The technique involves transplantation of alginate embedded human hepatocytes in the peritoneal cavity with out the need for immunosuppression.

Portal hypertension: What's new

Dr PJ Mc Kiernan, Consultant Paediatric Hepatologist, Liver Unit, Birmingham Children's Hospital NHS Trust, Birmingham B4 6NH
email Pat.Mckiernan@BCH.nhs.uk

Portal hypertension is most commonly due to cirrhosis, but the commonest single cause is extrahepatic portal vein obstruction (EHPVO).

New, non-invasive methods to predict the presence of oesophageal varices are being developed, but to date upper GI endoscopy remains the definitive diagnostic tool.

Risk factors for first variceal bleed:
1. Variceal size
2. The presence of red colour signs at endoscopy
3. The severity of liver disease
4. Severity of portal hypertension

Approximately 40% children with cirrhosis who have oesophageal varices will bleed over a 5 year follow-up. Bleeding is commoner in those with EHPVO where there is a life-long bleeding rate of 80%.

Management of acute variceal bleeding:
Emergency management is summarised in Table 1.

Pharmacotherapy:
Octreotide should be used as an adjunct to endoscopic treatment. Glypressin appears safe but there is little experience in children.

Endoscopy:
This should be carried out within 24 hours with a view to endotherapy. Variceal band ligation is the preferred method with sclerotherapy reserved for where this is impossible. Endotherapy should control acute bleeding in up to 90% of cases. If bleeding persists endotherapy should be repeated once.

Failure to control bleeding:
Balloon tamponade is an effective palliative to facilitate transfer or to temporise until definitive treatment.

Transjugular intrahepatic porto-systemic shunt:
This is a technically demanding radiologically created porto-systemic shunt. It is highly successful in controlling acute variceal bleeding and should be considered early if there is initial failure to control bleeding.

Secondary prophylaxis:
Recurrent bleeding rates are as high as 80%. All children should receive secondary prophylaxis. Available options include:
(i) endoscopic treatment
(ii) ß blockers
(iii) TIPS
(iv) Surgical treatment

Variceal banding is the preferred endoscopic method.

ß blockers: These are as effective as endotherapy in adults. They have not been shown to be effective in children but their use seems reasonable in post pubertal children. The effect of βblockers seems confined to those who achieve a reduction in hepatic venous pressure gradient (HVPG) to <12 mmHg or a reduction from baseline of >20%. HVPG measurements are invasive, requiring transjugular hepatic vein catheterisation. However with skilled operators this is feasible and safe, even in small children.

TIPS has a role in selected cases where repeated anaesthesia is best avoided, for example if cystic fibrosis liver disease.

Surgical treatments:
Liver transplantation: This remains the definitive treatment for children with portal hypertension due to cirrhosis. Portal hypertension however is rarely the sole indication for liver transplantation.

Mesoportal bypass. This provides a long term functional and anatomical cure for EHPVO. Where it is feasible this should be the treatment of choice.

Surgical shunts: Porto-systemic type shunts are now rarely indicated, but in skilled hands provide long term protection against variceal bleeding..

Primary prophylaxis:
ßßblockade and variceal banding are indicated for large oesophageal varices in adults. ß blockers are usually first line with banding reserved for where these are contraindicated or not tolerated.

Endoscopic treatment: Uncontrolled studies have shown this to be effective and safe. Definitive controlled studies are probably not feasible. Hence this is a reasonable option for large oesophageal varices.

ß blockers: These have not been shown to be effective in children but their use seems reasonable in post pubertal children. In prepubertal children they should only be used as part of a structured protocol and if used should be combined with HVPG monitoring.

Table 1.

Emergency management of suspected variceal bleeding

- Establish IV access
- Crossmatch blood (70 ml/kg packed cells)
- Consider passing nasogastric tube
- Start ranitidine/omeprazole
- Start octreotide (1µg/kg bolus and 1µg/kg/hour)
- Only correct severe coagulopathy/thrombocytopenia
- Start antibiotics

Metabolic causes of fulminant liver failure

Dr Andrew Morris, Consultant in Metabolic Disease, Manchester Children's Hospital, Manchester

Inherited metabolic diseases can present with liver failure, cholestasis, cirrhosis or hepatomegaly.

Specific treatment is available for many and diagnosis is particularly urgent for those with fulminant liver failure.

Clues to the diagnosis include the age at presentation and precipitating factors. Galactosaemia usually presents between 4 and 10 days of age. Urea cycle disorders (UCDs) and fatty acid oxidation disorders (FAODs) often present in the first 3 days; later presentations are usually precipitated by an infection or a change in diet. Hereditary fructose intolerance (HFI) only presents after fructose is introduced into the diet. Phosphomannose isomerase deficiency (MPI-CDG) presents in the first year. Mitochondrial disorders can cause liver failure at any age and so can tyrosinaemia type 1, after the first 2 weeks. Wilson's liver disease presents at 4-20 years of age.

The accompanying problems are another diagnostic clue. Hypoglycaemia, hyperammonaemia and lactic acidaemia are non-specific features of liver failure but, if severe, suggest an inborn error. Patients with UCDs and FAODs have a Reye-like encephalopathy. Renal tubular dysfunction occurs in galactosaemia, tyrosinaemia, HFI, Wilson disease and many mitochondrial disorders. A history of seizures suggests Alpers disease. Kayser-Fleischer rings and haemolysis suggest Wilson disease but are often absent in children presenting with liver failure.

The diagnostic tests can occasionally be misleading. In galactosaemia, urine reducing substances are negative after milk is withdrawn and red cell Gal-1-PUT is unreliable after transfusion. To exclude tyrosinaemia, urine succinylacetone should be requested specifically, as the organic acid sample requires 'oximation'. Mitochondrial studies may be normal in muscle in patients with hepatocerebral mtDNA depletion (including Alpers disease). No single test for Wilson disease is entirely reliable.

Treatment starts by excluding potential dietary toxins, such as galactose, fructose or protein, and providing enough energy to prevent catabolism. Treatment for UCDs includes sodium benzoate, sodium phenylbutyrate and arginine, with dialysis if there is severe hyperammonaemia. Other specific drugs include nitisinone for tyrosinaemia, mannose for MPI-CDG and penicillamine (±zinc) for Wilson disease.

Liver transplantation is needed for liver failure due to ∂-1-antitrypsin deficiency, some patients with Wilson disease and occasionally in galactosaemia or tyrosinaemia.

Liver transplantation is seldom offered for mitochondrial disorders but it has been done for MPV17 defects as neurological involvement may not occur for several years.

What's new in Autoimmune Liver Disease

Giorgina Mieli-Vergani, MD PhD FRCP FRCPCH, Paediatric Liver, GI and Nutrition Centre
King's College London School of Medicine at King's College Hospital, London

Autoimmune liver disorders in childhood include autoimmune hepatitis (AIH), autoimmune sclerosing cholangitis (ASC) and *de novo* AIH after liver transplant (LT). These inflammatory liver disorders are characterised histologically by interface hepatitis, biochemically by elevated transaminase levels and serologically by autoantibodies and increased levels of immunoglobulin G. AIH is particularly aggressive in children and progresses rapidly unless immunosuppressive treatment is started promptly. With appropriate treatment 80% of patients achieve remission and long-term survival. For non-responders and difficult-to-treat patients, novel and more effective therapeutic approaches are sought. ASC responds to the same treatment used for autoimmune hepatitis in regards to parenchymal inflammation, but bile duct disease progresses in about 50% of cases, leading to a worse prognosis and higher LT requirement; it has a high recurrence rate after LT. Progression of liver disease and recurrence after LT are more common in patients with associated poorly controlled inflammatory bowel disease. *De novo* AIH after LT affects children transplanted for non-autoimmune conditions and responds well to the same treatment schedule used for AIH, but not to the standard schedule used for acute rejection.

Though the mechanisms underlying the pathogenesis of liver autoimmunity are not fully understood, there is mounting evidence that genetic predisposition, molecular mimicry and impaired regulatory networks contribute to the initiation and perpetuation of the CD4+ T cell-mediated autoimmune attack. A deeper understanding of the pathogenesis of these conditions will contribute to the development of novel treatments, aimed ultimately at the restoration of tolerance to liver-derived antigens.

THURSDAY INVITED SPEAKER ABSTRACTS

SESSION II

'MECHANISMS OF VISCERAL PAIN IN FUNCTIONAL GI DISORDERS'

Professor Qasim Aziz, Neurogastroenterology Group, Barts and The London School of Medicine and Dentistry, Queen Mary University of London UK

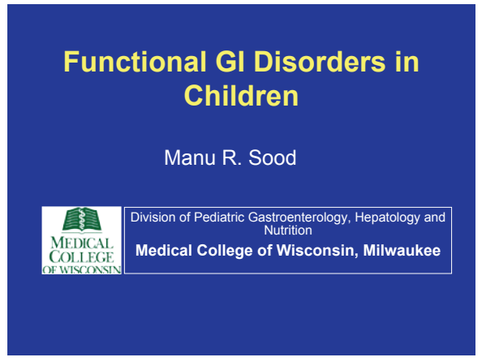
Introduction:
Heightened perception of gastrointestinal (GI) sensation (visceral hypersensitivity): is commonly observed in patients with unexplained abdominal pain. Studies have reproducibly demonstrated that patients with Functional Gastrointestinal Disorder (FGID) have lower gastrointestinal pain thresholds in comparison to healthy subjects. Evidence from animal and human studies clearly demonstrate that inflammation of the GI tract leads to visceral hypersensitivity due to increased sensitivity of afferent pathways. Furthermore, a third of FGID patients give a prior history of gut inflammation such as gastroenteritis. Based on current scientific evidence a number of hypotheses have been proposed to explain the mechanism of visceral hypersensitivity. These include (1) sensitisation of gastrointestinal afferent nerves (peripheral sensitisation {PS}), (2) sensitisation of spinal cord dorsal horn neurons (central sensitisation {CS}), and (3) misinterpretation of non-noxious sensation as noxious due to cognitive and emotional biasing (hypervigilance), the result of psychiatric / psychological disorders.

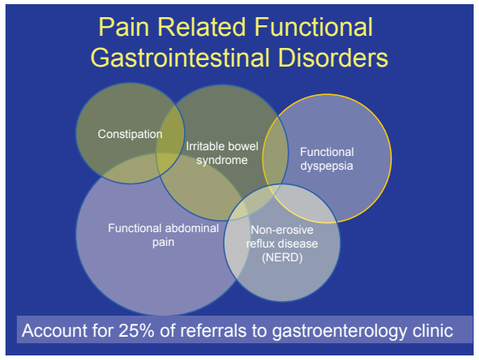
The brain gut axis in FGID:
Due to advances in functional brain imaging it has become possible to study the human brain gut axis in health and disease. These studies show that the brain processing of visceral pain is multidimensional and involves processing in the sensory discriminative, affective and cognitive areas of the brain. More recent studies demonstrate that the brain processing of visceral pain is modulated by psychological factors such as personality, anxiety, mood and attention. For instance the emotional context in which visceral sensation is perceived modulates the brain processing of that sensation. In addition the entire visceral pain network is activated during the anticipation of pain. In patients with FGID aberrant brain processing in the affective and cognitive evaluative brain regions is consistently demonstrated. Assessment and management of psychological aspects in FGID patients is therefore of paramount importance.

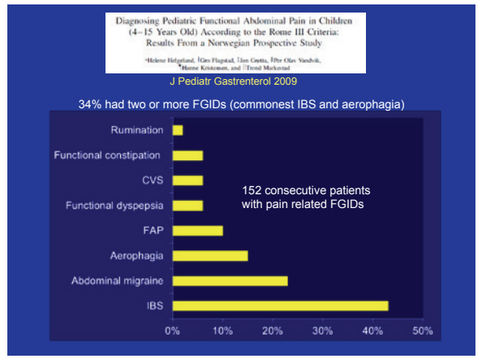
Management of FGID:
A number of management strategies can be considered for management of visceral pain in FGID. Modulators of peripheral and central sensitisation include low dose tricyclic antidepressants, gabapentin and pregabalin can be considered in appropriate patients. Psychological therapies such as cognitive behavioural therapy have been shown to have long term efficacy in FGID patients.

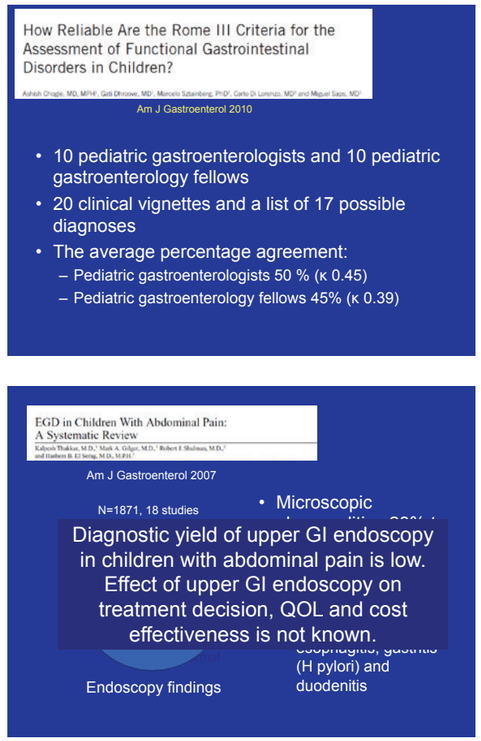
Functional GI disorders in children

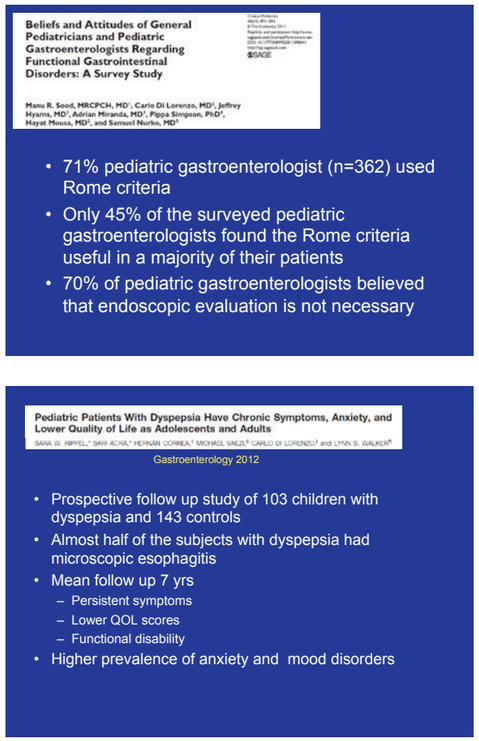
Manu R. Sood, Professor and Chief, Division of Pediatric Gastroenterology, Hepatology and Nutrition, Medical College of Wisconsin, Milwaukee, USA.











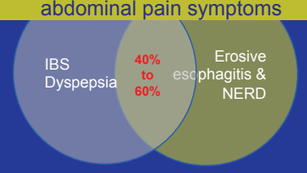
Pediatric Gastroesophageal Reflux Clinical Practice Guidelines: Joint Recommendations of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN)

Histology may be normal even in some patients with erosive reflux esophagitis; conversely, it may be normal or abnormal in non-erosive reflux disease (NERD)

Endoscopic biopsy cannot determine whether esophagitis, if present, is due to reflux.

Reflux disease and FGID overlap

We need to better categorize children with gastroesophageal reflux and upper abdominal pain symptoms



Neumann H, et al. Dig Dis 2008
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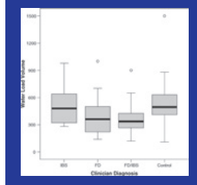
Diagnostic utility of water load test

Schurman JV, et al. JPGN 2007:44:51-7



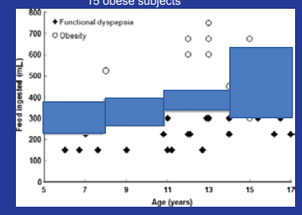
- Vol. of water consumed:
  - Chronic abdominal pain 395 ± 198 ml.
  - Controls 528 ± 257 ml. ($p < 0.01$)
- Using a cut off of 275 ml the sensitivity was 33% and specificity 100%

Assessment of gastric motor function in childhood functional dyspepsia and obesity

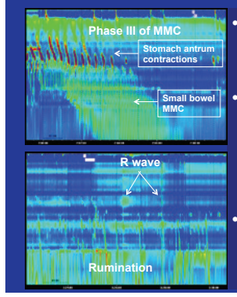
E. HOFFMAN* & J. TACK†

Neurogastroenterol Motil 2012

28 patients with dyspepsia
15 obese subjects



Adolescent Rumination Syndrome



- In a majority of patients diagnosis can be made by history
- Antroduodenal manometry is considered a "big convincer" but not always helpful
- Post prandial impedance-manometry

Dilated Intercellular Spaces as a Marker of GERD

Lori A. Orlando, MD, MHS, and Roy C. Orlando, MD

Refractory Heartburn: Comparison of Space Diameter in Documented GERD vs. Functional Heartburn

Intercellular space diameters of the oesophageal epithelium in NERD patients: Head to head comparison between light and electron microscopy analysis

Intercellular Space Volume Is Mainly Increased in the Basal Layer of Esophageal Squamous Epithelium in Patients with GERD

Gall bladder dyskinesia

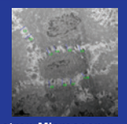
- Functional gall bladder disorder is a poorly defined entity
- No validated symptom based diagnostic criteria
- A validated standardized gall bladder scintigraphy ejection fraction test is needed
- Current data suggest that 40% - 50% patients screened for surgery using GB scintigraphy test continue to have symptoms following surgery
- Outcomes of medical and surgical management of functional gall bladder disorder are comparable

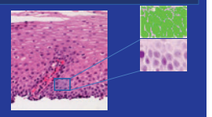
Factors affecting GB ejection fraction

- Duration of fast before the test
- Dose of CCK
- Infusion time
- Diabetes, celiac disease and IBS
- Medications:
  - opioid analgesics, calcium channel blockers, oral contraceptives, histamine-2 blockers, benzodiazepines

Esophageal epithelial intercellular space

Esophageal mucosal biopsies were obtained 3-5 cm proximal to GE junction





Electron Microscopy:
- Tissue was fixed in 4% glutaraldehyde solution and embedded in epoxy resin
- Electron microscopy images were obtained and digitized
- 100 measurements of ICS diameter were performed

Light microscopy:
- Tissue was fixed in formaldehyde
- 5 µm sections stained with H&E were used
- At least 4 images at 200X magnification were used to measure ICS spaces using Axial Cam and AXIO VISION software
- Phase contrast was used to calculate ICS area

Functional dyspepsia

A heterogeneous disorder

Hypersensitivity to distension: causes epigastric pain, belching and weight loss

Delayed gastric emptying: post prandial fullness, nausea and vomiting

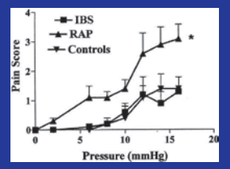
Reduced accommodation: causes early satiety and weight loss

Suggested protocol for gall bladder ejection fraction test

- Fast for 4-8 hours before the test
- Perform the test as out patient and NOT when the patient is acutely ill or in pain
- Opiate and anticholenergic should be held for at least 48 hours
- Nifedipine, indomethacin, octreotide, theophylline, benzodiazepines, phentolamine, isoproterenol and progesterone should be held for 24 hours
- Diabetes, celiac disease and IBS can result in a false +ve test
- Based on adult studies sincalide dose of 0.02 mcg/kg
- Infusion over 60 min
- Gall bladder ejection fraction ?
  - 35% cut off used in most pediatric studies
  - Using a lower value i.e. 15% can help better predict outcome of surgery

Gastric sensory function in children with RAP

Di Lorenzo et al J Pediatr 2001:139:838-43

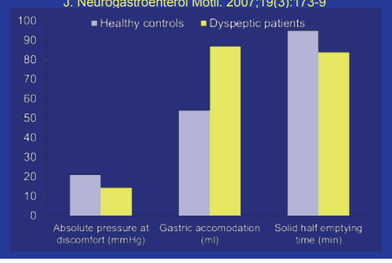


- Gastric pain perception in children with RAP lower than controls $p<0.005$ and IBS patients ($p<0.01$)

Assessment of gastric sensorimotor function in paediatric patients with unexplained dyspeptic symptoms and poor weight gain

I. HOFFMAN,* R. VOS† & J. TACK†

J. Neurogastroenterol Motil. 2007;19(3):173-9



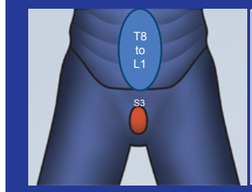
Rectal barostat in children with functional GI disorders

	R. Van Ginkel, et al 2001		C. Di Lorenzo, et al 2001		J. Castilloux et al 2006	
	Control n=9		Control n=7		Control n=10	
Rome category	IBS n=8	RAP n=8	IBS n=10	RAP n=10	IBS n=29	RAP n=18
Rectal pain threshold	+++	++	+++	++	++	++
Rectal compliance	Reduced in IBS No difference between disease groups		No difference between controls or disease groups		Not evaluated	

Viscero-somatic projection of rectal distension

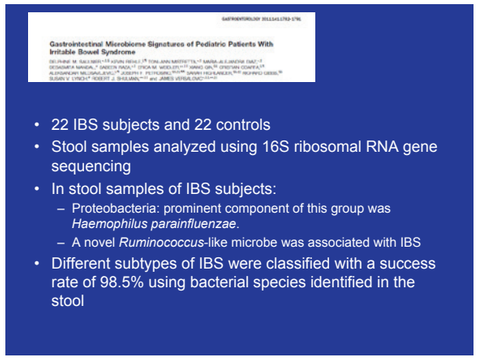
Faure C, Wieckowska A. J Pediatr. 2007;150(1):66-71.

No. of subjects: controls 10, IBS 21, RAP 8, FD 6



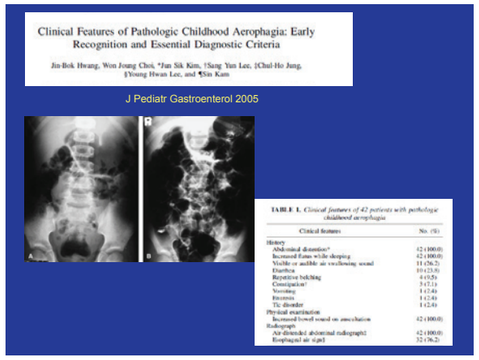
Rectal distension induced pain was similar to the pain at home:

- 90% of IBS
- 87.5% with RAP



Gastrointestinal Microbiome Signatures of Pediatric Patients With Irritable Bowel Syndrome

- 22 IBS subjects and 22 controls
- Stool samples analyzed using 16S ribosomal RNA gene sequencing
- In stool samples of IBS subjects:
  - Proteobacteria: prominent component of this group was *Haemophilus parainfluenzae*.
  - A novel *Ruminococcus*-like microbe was associated with IBS
- Different subtypes of IBS were classified with a success rate of 98.5% using bacterial species identified in the stool



Clinical Features of Pathologic Childhood Aerophagia: Early Recognition and Essential Diagnostic Criteria

J Pediatr Gastroenterol 2005

TABLE I. *Clinical features of 42 patients with pathologic childhood aerophagia*

Clinical features	No. (%)
History	
Abd-minal distention*	42 (100.0)
Increased flatus while sleeping	42 (100.0)
Visible or audible air swallowing sound	11 (26.2)
Diarrhea	10 (23.8)
Repetitive belching	4 (9.5)
Constipation†	3 (7.1)
Vomiting	1 (2.4)
Insomnia	1 (2.4)
Tic disorder	1 (2.4)
Physical examination	
Increased bowel sound on auscultation	42 (100.0)
Radiograph	
Air distended abdominal radiograph‡	42 (100.0)
Esophageal air sign§	32 (76.2)

Management of FGIDs

- Psychological and cognitive behavioral therapy
- Diet modification
- Probiotics
- Antispasmodics
  - Peppermint oil
- Tricyclic antidepressants and SSRIs
- Melatonin

Summary

- There is significant overlap between FGIDs defined using Rome criteria
- Studies validating Rome criteria are lacking
- Visceral hypersensitivity, motility abnormalities, altered gut microbiota, and ineffective coping mechanism have been implicated in the pathophysiology
- Objective measures of GI sensory function may be helpful

Hypnosis for IBS

Professor Peter Whorwell, Professor of Medicine and Gastroenterology, Wythenshawe Hospital Southmoor Road, Manchester, M23 9LT

Our unit has been undertaking research on the therapeutic potential of hypnotherapy in adults with functional gastrointestinal disorders for over twenty years. During this time we have demonstrated that it improves the symptoms of irritable bowel syndrome, functional dyspepsia and non cardiac chest pain as well as having the capacity to modulate some aspects of gastrointestinal the physiological function. More recently there have been some encouraging reports that hypnotherapy, and similar techniques, are remarkably effective in children with irritable bowel syndrome and functional abdominal pain, where the beneficial effects are sustained over a number of years.

Evidence based management of severe constipation

Dr Jenny Gordon, Research & Development Fellow, Quality Improvement Programme
RCN Institute, OX4 2JY

This session will discuss the diagnosis and management of chronic idiopathic constipation based on NICE Guideline 99 Constipation in children and young people (May 2010). It will include history taking, examination, clinical management, diet and lifestyle and information and support. It will also consider the challenges of implementing evidence based practice in the current healthcare landscape and the role of further research to improve outcomes for children and families in the future. www.nice.org.uk/CG99

THURSDAY INVITED SPEAKER ABSTRACTS

SESSION III AND FREE PAPERS

Familial adenomatous polyposis syndrome (FAP): current guidelines for endoscopic management of FAP and timing and choice of colectomy

Dr Warren Hyer, Consultant Paediatric Gastroenterologist, St Mark's Hospital Polyposis Registry, UK
Email: warren.hyer@nhs.net

Educational aims:

- Understand the genetics and endoscopic management of FAP
- To be able to counsel and advise on surgical choices for colectomy for FAP in adolescents
- Understand the timing for colonoscopy and colectomy in children and adolescents
- Access the European guidelines for managing FAP in adolescents (and adult)

FAP is an autosomal dominant condition caused by APC mutations on chromosome 5 with a correlation between genotype and severity of phenotype for some of the common mutations. Without surgical intervention, affected patients inevitably develop CRC (colorectal cancer) by age 40–50 years.

Children of affected parents are referred to paediatric gastroenterologists by Polyposis Registries, genetic counselors or colorectal surgeons for screening, surveillance and advice on therapeutic options. Once the family-specific mutation has been identified, DNA diagnostic techniques are used to predict FAP in other family members. The absence of the family-specific mutation is considered accurate in excluding FAP.

Present consensus recommends screening at risk children age 10-12 years, whilst the risk of CRC is very low before age 20 years. Those families with a more aggressive APC genotype (e.g.1309 & 1464), should undergo screening earlier age 7 -10 years. Symptomatic at risk children with diarrhea and/or bleeding should undergo colonoscopy at the earliest opportunity as they risk dense polyposis or severe dysplasia.

Full colonoscopy is advisable in gene positive children. Once adenomas are identified, annual colonoscopy is required until colectomy is performed. This is to assess adenoma burden and assess density of rectal polyps. In children of affected parents where the APC gene mutation has not been identified, annual sigmoidoscopy with dye spray is required until adenomas are identified, and then colonoscopy should be performed.

Colectomy is mandatory in affected patients before they develop CRC. This is usually necessary age 16-25 years, depending on genotype, polyp density identified at colonoscopy, and risk of desmoid disease. In general, a colectomy is indicated if there are large numbers of adenomas > 5 mm, including adenomas showing a high degree of dysplasia. The two options for prophylactic colectomy are:

1. colectomy with ileorectal anastomosis (IRA)
2. proctocolectomy with ileal pouch–anal anastomosis (IPAA).

IRA is a relatively simple and straightforward operation, compared with IPAA. The complication rate is relatively low and the bowel function postoperatively is good. After an IRA, the rectum is left in place and this requires annual surveillance and carries a risk of later cancer, or removal and conversion to an IPAA. For IPAA, more extensive surgery is needed including pelvic dissection with its risk of haemorrhage, and reduction of fertility in women..

The surgical decision should be shared with the patient. Those with rectal adenoma burden >15- 20 adenomas, should be offered an IPAA. Patients with a low rectal polyp burden (<15), can safely be offered an IRA with annual surveillance and treatment of the rectal adenomas. The preferred surgical choice depends will be influenced by age of the patient, the severity of rectal polyposis, the wish to have children, the risk of developing desmoids and the site of the mutation in the APC gene.

Chemoprevention with COX 2 inhibitors should presently be reserved for those patients who have had a colectomy by IRA as an adjunctive intervention to reduce rectal polyp density.

Ref: Guidelines for the clinical management of FAP. Vasen, et al. Gut 2008 57: 704-713.

The predictive value of ELF test in Biliary Atresia

Dr Nicola Ruth[1], Professor W Rosenberg[2], Dr Patrick McKiernan[1]
[1]The Liver Unit, BCH, [2]The Institute of Liver and Digestive Health, University College London

Background and aims:
Biliary Atresia (BA) is the commonest cause of serious liver disease in childhood. The primary treatment is surgical with the Kasai procedure. Where this is successful in clearing jaundice most children will subsequently survive without transplant into adult life. Where the Kasai procedure is unsuccessful, the only treatment is liver transplantation. There is a need for non invasive test to predict prognosis in individuals with BA. The extended liver fibrosis (ELF) test has been widely validated for staging hepatic fibrosis in adult patients and in some sub-groups of paediatric liver disease. Our aim was to determine whether serial changes in the ELF test or its components are predictive of long term outcome (and therefore survival with/without transplant) following the Kasai procedure

Subjects and Methods:
39 patients with BA presenting to a single centre between 2007 and 2011. ELF and its components were measured at time of Kasai (baseline), 3 months later and at 1 year. At baseline liver biopsy was performed, and analysed by degree of fibrosis as well as ISHAK score. Outcome was classified as survival with or without transplantation at 18 months.

Statistics used were multivariate and univariate logistic regression.

Results:
All are alive and 13 have undergone transplant at age <18 months. At baseline the only factor predictive of requiring transplantation was age at surgery (p=0.048). None of ELF, its components or baseline bilirubin were predictive of outcome.
At 3 months bilirubin level, ELF and 2 of its components (Hyaluronic acid and TIMP1) were predictive of outcome. Following multivariate analysis bilirubin was the only significant predictive factor (p=0.001).
In those surviving without transplant there was a significant fall in ELF from baseline mean of 12.16 to 10.65 and p<0.005.

Conclusion:
In biliary atresia: ELF at the time of Kasai is not predictive of early outcome. ELF at three months post Kasai is predictive of early outcome but does not improve on conventional measures. ELF levels fall significantly following successful Kasai.
Serial ELF measurements have prognostic value for the early outcome of biliary atresia. Longitudinal studies of serial ELF measurements in biliary atresia are necessary.

Hepatitis B Vaccination Failure in Children born to hepatitis B positive mothers

Legarda M[1], Brown M[1], Sira J[1], Boxall EH[2], Kelly DA[1]
[1]Liver Unit, Birmingham Children´s Hospital, [2]HPA West Midlands Public Health Laboratory.

Objectives:
To evaluate the reasons for HBV vaccination failure in children born to HBV positive mothers in the UK.

Background:
HBV vaccination is effective in preventing infection in infants of mothers with chronic HBV. Antenatal screening programmes identify HBV infected mothers in the UK and their infants are recommended an accelerated vaccination schedule at 0, 1 and 2 months with HBV serology and booster at 12 months. Children born to HBeAg positive mothers or with high HBV viral load also receive HBIG at birth.

Subjects and Methods:
We retrospectively evaluated patients referred between 1997 and 2012, who were infected following HBV vaccination at birth. We collected data on demographics, child and maternal HBV serology, viral load, genotype, vaccine escape mutations and vaccine schedule. Statistical analysis was performed using SPSS version 15.0. Results: 40 patients (16 girls) born to 33 HBV positive mothers in the UK were reviewed. Median age at diagnosis was 14 months (range 6 to 109 months). The majority of mothers originated from the Indian subcontinent and the Far East and none received antiviral treatment during pregnancy. All children were HBsAg positive. We divided the children in two groups: 21 children did not receive a correct vaccination schedule and 17 children had been fully and correctly vaccinated (true vaccination failure). Vaccination schedule was unknown in 2 children. The reason for incorrect vaccination was: unknown in the majority, failure to attend appointments in 3, no appointment given in 2, unknown HBV infection in 1, incorrect vaccination schedule in 3 and change of address in 1. The majority of incorrectly vaccinated children missed more than one dose of vaccine. Four children in the true vaccination failure had vaccine escape mutants. The majority of mothers were HBeAg positive.

The following table shows the characteristics of the children with true vaccination failure and with incorrect vaccination schedule and the p values obtained using chi^2.

	True Vaccination Failure (n =17)	Incorrect Vaccination Schedule (n=21)	Incorrect Vaccination Schedule
Maternal HBeAg + (n=29)	14 (82%)	15 (71%)	0.48
Maternal HBV DNA > 106copies/ml (n= 21)	13 (76.5%)	8 (38%)	0.01
HBV Genotype (n=17)	1A, 2B, 7C, 3D	3B,2D	0.001
Vaccine Escape Mutant (n=5)	4 (23.5%)	1 (5%)	< 0.001
HBIG (n=29)	17 (100%)	12 (57%)	< 0.001

Summary:
The majority of children who failed HBV vaccination had not received the correct vaccination schedule either from failure of primary care services or non attendance by parents. True vaccination failure was associated with high maternal viral load and genotype C, presence of HBV vaccine escape mutant and HBIG administration. The majority of mothers were HBeAg positive in both the true vaccination failure and incorrect vaccination schedule groups.

Conclusions:
It is essential that children born to HBV mothers receive adequate HBV vaccination, with appropriate communication, follow up and involvement of primary care. Mothers with high viraemia should receive antiviral treatment in pregnancy to reduce infectivity. True vaccination failures must be investigated to ensure that the use of Hepatitis B immune globulin is not selecting for vaccine escape mutants.

Long term outcome of children following liver transplantation

Legarda M[1], Smith M[1,2], Lewis P[1,2], Lloyd C[1], Paris S[2], Kelly DA[1]
[1]Liver Unit, Birmingham Children's Hospital (BCH), [2]Liver Unit, Queen Elizabeth Hospital Birmingham.

Aims:
To evaluate survival and outcome of patients more than 15 years following liver transplantation.

Background:
Liver transplantation (LT) is a successful treatment for end stage liver disease. There are little data on the long term outcome of patients who underwent LT during childhood.

Subjects and methods:
We retrospectively reviewed all patients who underwent LT more than 15 years ago. We reviewed their medical notes and patients who lived locally were interviewed personally during their routine outpatient appointments. We contacted physicians looking after those patients who had been referred elsewhere in order to complete the data collection. We collected data on liver & renal function, hypertension, dyslipidemia, bone disease, recurrent disease, rejection, de Novo Autoimmune Hepatitis, psychological disorders, compliance and social adaptation.

Results:
116/181 patients who underwent LT between 1985 and 1995 in BCH were alive more than 15 years after transplantation (median post-transplantation time was 19.5 years, range: 16 to 27 years). Median age at LT was 25 months (range: 15 days to 16 years) and the main indication for transplantation was extrahepatic biliary atresia (51%). 23/116 were re transplanted. The majority of re transplantations occurred during the first year post LT. 7 patients required a second transplant 5 to 11 years after their first LT due to chronic rejection. At > 15 years 89% had normal functioning grafts; 43% were on steroid immunosuppression and 54% were on monotherapy mainly with Mycophenolate/Tacrolimus or Cyclosporin. The main causes of graft dysfunction were chronic rejection (66%) and graft fibrosis (33%). Long term complications included renal dysfunction (22% had calculated Glomerular Filtration Rate (cGFR) or GFR < 60 ml/min/1.73 m^2 and 3 patients required a renal transplant), 39% required antihypertensive treatment, 9% had had post transplant lymphoproliferative disorders/lymphoma/leukaemia. 54% were compliant with treatment and appointments. 28% went to university; 43% to college; 29% were still at school. 50% are still in education and 50% are employed. 20 % are unemployed and not in education. 51% are married or in a relationship and 13% have had children.

Summary:
The survival rate of LT recipients more than 15 years after LT is 64%. Most have normally functioning grafts, are in employment or education and are married or in a relationship. The main reasons for graft loss were chronic rejection and graft fibrosis. The complications of long term immunosuppression include high blood pressure, renal dysfunction and lymphoproliferative disorders. Non-adherence was present in 46% of the group.

Conclusions:
The long term outcome of LT is excellent allowing patients to have normal social adaptation and functioning.

Propranolol in the management of hepatic haemangioendothelioma – King's College Hospital experience

Protima Amon, Palaniswamy Karthikeyan, Dominic Hughes, Mark Butler, Mark Davenport, Anil Dhawan, Sanjay Bansal
King's College Hospital, London

Background:
Hepatic haemangioendothelioma (HHE) are rare vascular tumours. Treatment can be conservative (corticosteroids, diuretics) or surgical/radiological (hepatic artery ligation (HAL)/embolisation, resection or liver transplantation). The rarity of the condition and lack of randomised controlled trials makes therapeutic decisions particularly challenging. Propranolol has been used in the treatment of cutaneous and solid organ haemangiomas or haemangioendotheliomas.

Aims:
To assess the efficacy and safety of Propranolol in the treatment of HHE.

Subjects and Methods:
42 patients were referred with HHE between May 1998 and September 2012. Our initial protocol was to use corticosteroids and/or diuretics and/or surgical/radiological intervention. Since April 2008, Propranolol was added to the protocol. Patients were categorised depending on whether they received Propranolol (Group 1) or not (Group 2). Patients were followed up with serial ultrasound scans. The starting dose of Propranolol used was 0.5mg/kg in 2 divided doses titrating up to a maximum dose of 2mg/kg in 2 divided doses.

Results:

		Group 1 n	Group 1 %	Group 2 n	Group 2 %
Number of Patients (n)		16	100	26	100
Sex	Male	5	31	13	50
	Female	11	69	13	50
Median Age at Presentation (days)		39		67	
Mode of Presentation	Symptomatic	5	31	8	31
	Incidental	7	44	9	35
	Antenatal	4	25	9	35
Severity	PICU Admission	8	50	4	15
Haematological	Thrombocytopenia	3	19	5	19
Treatment	Steroids	8	50	6	23
	Diuretics	7	44	7	27
	Surgical (ligation/resection)	2(2/0)*	13	10 (8/2)	38
	No Treatment	0	0	14	54
Radiological	Diffuse lesion	13	81	10	38
	Improvement (mean time in months)	3.9		6.8	

*Both these babies received propranolol after HAL. The first baby was listed for liver transplantation after he had developed ischaemic hepatitis following HAL. While he was on the waiting list, Propranolol was started 4 months after ligation. The second baby had a primary HAL as she was clinically unstable. Due to the diffuse nature of the tumour, it was assumed that ligation might not be as effective and listing for liver transplantation was considered. However, she was started on Propranolol a week after the ligation. Both the babies responded to propranolol and recovered completely and did not require liver transplantation.

Propranolol was very well tolerated. Only one patient discontinued treatment because of wheezing. Two children became hypotensive and one child became hypoglycaemic. All three continued Propranolol after a dose reduction and subsequently tolerated titration to maximal dose.

Summary and Conclusions:
Propranolol is a safe and effective treatment modality with or without steroids/diuretics for HHE and may obviate the use of surgical or radiological interventions.

The BISCUIT Study: Exploring the "bacteriotype" of de-novo paediatric IBD

Richard Hansen[1,2], Richard K. Russell[3], Caroline Reiff[4], Petra Louis[5], W. Michael Bisset[6], Andy R. Barclay[3], Jon Bishop[3], Diana M. Flynn[3], Paraic McGrogan[3], Sabarinathan Loganathan[6], Gamal Mahdi[6], Emad M. El-Omar[1] and Georgina L. Hold[1]
[1]Gastrointestinal Research Group, Division of Applied Medicine, University of Aberdeen, Foresterhill, Aberdeen; [2]Child Health, University of Aberdeen, Royal Aberdeen Children's Hospital, Foresterhill, Aberdeen; [3]Department of Paediatric Gastroenterology, Royal Hospital for Sick Children, Dalnair Street, Glasgow; [4]Max Planck Institute, Cologne, Germany; [5]Gut Health Programme, Rowett Institute, University of Aberdeen, Aberdeen [6]Department of Paediatric Gastroenterology, Royal Aberdeen Children's Hospital, Foresterhill, Aberdeen

Introduction
Paediatric Inflammatory bowel disease (IBD) incidence is rising in Scotland[1]. Recently the role of the gut microbiota has been recognised as pivotal in disease pathogenesis[2,3]. IBD microbial studies to date have focussed on established disease cohorts, with limited studies in treatment naive patients. We prospectively examined candidate bacterial triggers at disease onset, prior to any treatment, in children with IBD using several complementary methods focussing exclusively on mucosal biopsies. In doing so we set up the Bacteria in Inflammatory bowel disease in Scottish Children Undergoing Investigation before Treatment (BISCUIT) study.

Methods
Colonic biopsies were taken from a single site, from the distal colon in controls (rectum/sigmoid) or from the most distal inflamed site in those with colonic inflammation. 1-2 biopsies were used for microaerophilic culture work and 2-3 biopsies were collected for bacterial DNA analysis by PCR. Culture utilised selective plates, incubated in microaerophilic gas conditions, which were reviewed twice weekly for up to one month. Blood for serology and human DNA studies was also collected.

Results
100 children were recruited across three Scottish paediatric centres (Aberdeen, Glasgow and Dundee). These were 44 IBD (comprising Crohn's disease (CD) (29), ulcerative colitis (UC) (13) and IBD-type unspecified (2)); 42 normal colon controls (NCC) and 14 "others". 69 were male (30/44 IBD, 33/42 NCC and 6/14 "others"). Mean age (± standard deviation) was 11.0± 3.5 years (11.9± 2.9 IBD vs. 10.6± 3.5 NCC, p=0.067).

Microaerophilic bacterial culture work from all 100 patients, produced 414 individual bacterial isolates, of these, 114 were formally identified by DNA sequencing after screening out aerobes and Gram-positives. The most common isolate was *Sutterella wadsworthensis* (61 isolates from 32 recruits). Multiple non-*jejuni Campylobacter* species were also isolated (8 distinct patient isolates).

Colonic biopsy DNA from all 100 patients was then used to examine the prevalence of *Helicobacter* and *Campylobacter* genera and *S. wadsworthensis*. When comparing IBD vs. NCC, the prevalences were not significantly different (*Helicobacter* 11.4% vs. 11.9%, p=1.00; *Campylobacter* 75.0% vs. 76.2%, p=1.00; *S. wadsworthensis* 81.8% vs. 71.4%, p=0.312).

Biopsy DNA from 34 patients (11 CD, 11 UC, 12 NCC) was analysed by next-generation deep sequencing to examine overall bacterial diversity[4]. No significant changes were noted at phylum level among the Bacteroidetes, Firmicutes, or Proteobacteria. A significant reduction in bacterial βdiversity by Shannon index was noted in CD vs. controls (5.15± 0.46 vs. 5.59± 0.30, *p*=0.017) but not in UC vs. controls (5.54± 0.64 vs. 5.59± 0.30, *p*=0.835). This finding was replicated by two further methods (Simpson index and phylogenetic diversity). An increase in *Faecalibacterium* was observed in CD vs. NCC (mean 16.7% vs. 9.1% of reads, *p*=0.02) and replicated by specific *F. prausnitzii* RT-PCR (36.0% vs. 19.0% of total bacteria, *p*=0.02).

Conclusion
By using robust methodology we have characterised the IBD "bacteriotype" at diagnosis in children. This includes reduced diversity in CD with increases in *F. prausnitzii* but is not significantly altered in UC patients against controls. It is possible that exclusive enteral nutrition may reduce the colonic substrate for, and metabolic activity, of *F. prausnitzii*. The UC findings are against the adult literature which describes a reduction in bacterial diversity. This may suggest a "window of opportunity" to maintain a diverse colonic bacterial ecosystem, perhaps through probiotics, and influence disease chronicity.

Future work should focus on how to beneficially modify the IBD "bacteriotype" using established and novel IBD treatments.

References
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3 Jostins L, et al. Nature. 2012; 491(7422): 119-124.
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Identifying incidence of inherited metabolic disorders in patients with infantile liver disease

Gray Z[1], McKay K[2*], Lloyd C[1], Hartley J[1], MacDonald F[2], Hendriksz CJ[3], Gissen P[4], Kelly D[1], [1]Birmingham Children's Hospital NHS Foundation Trust, UK, [2]Birmingham Women's Hospital NHS Foundation Trust, UK [3]Salford Royal NHS Foundation Trust, UK, [4]UCL Institute of Child Health, UK*
**these authors contributed equally On behalf of all collaborating centres*

Introduction/Background:
The incidence of liver disease due to rare inherited metabolic disorders, including Niemann Pick type C (NPC), Citrin Deficiency and Progressive Familial Intrahepatic Cholestasis (PFIC), is unknown. New sequencing methods, including next generation sequencing (NGS), permit analysis of multiple genes simultaneously, reducing time to molecular diagnosis and cost. Accurate timely diagnosis is essential to optimise clinical management, improve targeted therapy for liver disease in infants, and avoid inappropriate liver transplantation.

Objectives:
To identify the incidence of inherited metabolic disorders, in patients with infantile liver disease, and to make this molecular information available early on in the diagnostic pathway.

Subjects/Methods:
A prospective study from 13 centres worldwide recruiting infants under 2 years presenting with cholestasis, acute liver failure or splenomegaly, using targeted NGS, for mutations in 6 genes (NPC1, NPC 2, ATP8B1, ABCB11, ABCB4 (PFIC 1-3), and SLC24A13 (Citrin Deficiency)).

Results:
202 patients have been recruited, and DNA sequenced in 87. Of those sequenced 66 presented with cholestasis, 12 acute liver failure, 7 isolated splenomegaly, 19 isolated hepatomegaly and 24 hepatosplenomegaly. Diagnosis was confirmed (homozygous pathogenic mutation or compound heterozygous pathogenic mutations) in 9 patients (10%): NPC1 (1); PFIC-1 *(ATP8B1)* (2); PFIC2 *(ABCB11)* (3); PFIC3 *(ABCB4)* (2); and in 1 patient two pathogenic mutations were identified in both PFIC-1 and PFIC-3 causing genes *(ATP8B1/ABCB4)*. 4 patients had single heterozygous pathogenic mutation identified. 29 variants of unknown significance were detected. There were no mutations detected in 55 of the 87 tested. Of these patients 12 had idiopathic cholestasis, 7 biliary atresia, 7 neonatal hepatitis, and 3 congenital cytomegalovirus infection.

Summary:
This unique study has recruited 202 patients with infantile liver disease, and so far successfully sequenced DNA in 87 of these. It has identified a mutation in 26/66 with cholestasis, 7/12 with acute liver failure, and 7/24 with hepatosplenomegaly. A genetic diagnosis was confirmed in 9 patients, and a heterozygous pathogenic mutation in 4 further patients. 29 novel variants were detected, of currently unknown pathogenicity. There were no mutations detected in 55 of the 87 sequenced. Most patients presented within 6 months of birth with cholestasis

Discussion/Conclusion:
These data show promising results for this sequencing method as a screening tool. Further analysis will allow us to establish the true prevalence of these disorders among patients with infantile liver disease, correlation between phenotypes and genotypes and clarify clinical indications for screening for these disorders.

No conflicts of interest declared. It is an investigator initiated study funded by Actelion Pharmaceuticals.

Selected short oral poster presentations

To be selected on day of meeting

THURSDAY INVITED SPEAKER ABSTRACTS

SESSION IV

Feeding And Nutritional Problems In Children With Neurological Impairment

Dr Peter Sullivan, Consultant Paediatric Gastroenterologist, University of Oxford, Dept of Paediatrics, John Radcliffe Hospital, Oxford, OX3 9DU

Feeding and nutritional problems are common in children with cerebral palsy and occur in up to 85% of those with severe spastic quadriplegic cerebral palsy. These feeding problems lead to growth failure and are associated with decrease in cerebral function, immune function, decrease in circulation time and respiratory muscle strength. Oro motor dysfunction underlies these feeding problems and is associated with profound feeding inefficiency and makes feeding these children slow and extremely difficult and stressful for their caretakers. Feeding difficulties in these children are associated with a significant reduction in the quality of life for their caregivers. Nutritional management of these children is complex and best undertaken in the context of a multidisciplinary feeding team with input from paediatricians, dietitians, speech and language therapists and clinical nurse specialists. Improvement in nutritional state can come from dietary supplementation but often adjunctive tube feeding is required. Gastrostomy feeding is not a panacea for the management of these problems and can be associated with significant complications not least of which is the potential for over-feeding. Careful pre-operative assessment before gastrostomy insertion as well as adequate post-operative follow-up is required.

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Vitamin D Deficiency – does it matter?

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Vitamin D is crucially important for calcium homeostasis and musculoskeletal health. Children with gastrointestinal (GI) conditions such as inflammatory bowel disease (IBD), coeliac disease, short bowel syndrome, pancreatic insufficiency and chronic liver disorders are at risk of developing vitamin D deficiency. Therefore, screening and treatment of vitamin D deficiency in patients with these GI disorders may help to prevent musculoskeletal consequences, such as rickets & osteopaenia. The goal of such treatment is to achieve normal serum concentrations of calcium, phosphate, alkaline phosphates, 25-hydroxyvitamin D and parathyroid hormone.

Evidence from epidemiological and laboratory studies suggest that vitamin D deficiency may play a role in the development of IBD. It is known that the risk of developing IBD is higher for individuals living in northern latitudes compared with those in southern latitudes. Thus, exposure to sunlight may provide protection against development of IBD through increased cutaneous vitamin D synthesis. The vitamin D pathway is a regulator of the immune system and there is emerging evidence that it plays an important role in the signalling between intestinal flora and the host. Mice that lack the vitamin D receptor have chronic, low-grade inflammation in response to non-pathogenic bacterial gut flora. Furthermore, treatment of IBD in IL-10 knockout mice with $1,25(OH)_2D3$, the biologically active metabolite of vitamin D, ameliorates symptoms and signs of IBD. Further studies are needed to confirm these relationships. Clinical trials of vitamin D supplementation in subjects at increased risk of developing IBD are warranted

My talk will deal with:

- Sources and metabolism of vitamin D.
- Musculoskeletal consequences of vitamin D deficiency in children.
- Assessment and treatment of vitamin D deficiency in children.
- Briefly review of the evidence linking the development of IBD to low body stores of vitamin D.

Acquisition of tolerance in infants with cow's milk allergy

Roberto Berni Canani, MD, Ph, Chief of Pediatric Nutrition and Food Allergy Unit, Pediatric Gastroenterology Hepatology and Nutrition Section, Department of Pediatrics and European Laboratory for the Investigation of Food Induced Disease, University of Naples Federico II, Naples, Italy

Cow's milk allergy (CMA) is the most common food allergy in early childhood, with an estimated incidence ranging between 2% and 3% in infants and marginally lower in older children. The majority of children regain tolerance to cow's milk proteins within the first 5 years of life. But, recent studies suggest that the natural history of CMA is changing, with an increasing persistence until later ages. The etiology of CMA is not yet completely defined, although numerous studies indicate that gut-associated immunity and microbiota may play a crucial role, and it has been suggested that an altered composition of gut microbiota results in an unbalanced local and systemic immune response to food allergens. There are qualitative and quantitative differences in the composition of gut microbiota between patients affected by CMA and healthy infants. Based on these findings, it has been proposed that specific beneficial bacteria from the human intestinal microflora, denoted "probiotics", could potentially restore intestinal homeostasis and prevent or treat food allergy. *Lactobacillus rhamnosus* GG (LGG) is the single probiotic with the greatest number of in *vitro and in vivo* evidences on possible effects in pediatric allergic disorders.

We recently demonstrated that an extensively hydrolyzed casein formula containing LGG is able to significantly accelerate the development of tolerance acquisition in infants affected by CMA. The mechanism of this beneficial effect are multiple, ranging from modulation of intestinal microflora composition, to direct effect on intestinal mucosa structure and function, and on local and systemic immune response. The results of this study on the beneficial effects elicited by LGG in children with CMA open the way to a possible "nutritional immunology approach" in these patients able not only to efficiently cure the symptoms but also to accelerate tolerance acquisition.

Food allergy and making sense of it all

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Food Hypersensitivity is the umbrella term recommended by the World Allergy Organisation to describe food allergies and non-allergic food hypersensitivity (also referred to as food intolerance). Food allergies are once again divided into IgE mediated (immediate onset) and non-IgE mediated (delayed onset) food allergies.

It is estimated that about 6% of young children in the UK suffer from food allergies, with the majority outgrowing their food allergies by the age of 5 years. Different food allergies are common in different age groups; milk and egg allergies being common in early childhood and nut and fish allergies being common in adults.

IgE mediated food allergies, presenting as urticaria, angioedema and potentially anaphylaxis, are predominantly seen with egg, nuts and seafood allergies. However, cow's milk and soya allergies present in most cases as non-IgE mediated food allergies with symptoms of vomiting, diarrhoea, eczema and gasto-oesophageal reflux disease. Taking an allergy focused clinical history forms the cornerstone of the diagnosis of food allergies. Other tests or investigations such as skin prick tests, specific IgE tests, food challenges or endoscopies should be performed as and when appropriate.

The management of food allergies is based on avoidance of the offending food(s) and the level of avoidance will depend on the clinical characteristics of the individual involved. The role of the dietitian in the diagnosis of food allergies is increasingly acknowledged. The dietitian also plays a crucial role in giving avoidance advice to the individual including safe shopping tips and free from recipes, at the same time ensuring sufficient nutritional intake.

Pharmaceutical management of food allergies and co-morbidities such as eczema, asthma and allergic rhinitis, may include the use of anti-histamines, oral/topical/inhaled steroids and injectable epinephrine.

Regular follow-up of patients is important to diagnose the development of tolerance in a timely fashion and also to monitor growth (in children) and adequate/optimal nutritional intake.

THURSDAY POSTERS

POSTERS OF DISTINCTION

A regional study in enteric neural stem cells

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Background and aims
Enteric neuropathies have so far been a challenging group of clinical entities with their current management being mainly palliative, thus driving research towards enteric nervous system (ENS) cell replenishment therapies using enteric neural stem cells (ENSCs-progenitors). Human ENSCs have been isolated only from colonic full thickness and mucosal biopsies, while mouse ENSCs have usually been derived from either colonic or small intestine. In order to determine which region of the gut would be a good source of cells for transplantation, this study attempted to establish which region contained a higher percentage or proportion of ENSCs. The accessibility of tissue from each region was also considered.

Methods
The studied regions were the colon, caecum , small intestine, stomach and oesophagus from wild type and the transgenic mouse line (Rosa26YFPstop:TgWnt1Cre) , where all neural crest cells (NCCs) , inclusive of ENSCs , express yellow fluorescent protein (YFP). By using fluorescence-activated cell sorting (FACS) the percentage of YFP positive cells was calculated for each region and clonal cultures were established to see how many single cells gave rise to colonies indicating percentage of ENSCs. Several methods of investigations were conducted to characterise the cells such as immunochemistry and FACS. . Either the entire gut or the outer muscle peels from the transgenic and wild type mouse gut were used for wholemount immunohistochemistry. Immunohistochemistry was also performed on sections of gut using various markers.

Results
The study revealed qualitative and quantitative differences in ENS across the regions. Wholemount on entire gut or muscle peel and section immunochemistry revealed more GFP, p75NTR, Tuj1, Sox10 and S100 stained cells in the colon and caecum followed by small intestine with much less percentages in stomach and oesophagus. A uniform difference in favour of myenteric plexus instead of submucosal was noted in all these markers. FACS sorting demonstrated highest percentage of YFP positive cells in colon with a peak in the caecum followed by a reducing pattern in the rest of the gut regions. Cell counting of Sox10 only stained cell in wild type mouse Sox10 /S100 co-labelling revealed the same pattern with a peak at caecum and its appendix.

Summary and conclusion
In conclusion, by using FACS and immunohistochemistry, we have demonstrated a predominance of ENSC-related ENS components in the colon and caecum .We have also shown a gradual reduction in the percentage across the gut from colon to the ileum to the stomach and oesophagus. Considering these findings and the balance between which region would contain more progenitors as well as be easily accessible to take tissue, we suggest that future ENSCs studies should continue to focus on the colonic region . In particularly, our study indicated that the appendix, as part of the caecum, may contain a more advanced niche of ENSCs, not yet elucidated.

Liver failure in young infants: aetiology, presentation and outcome

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Background:
Liver failure in infancy is rare but carries a significant mortality. The clinical presentation and results of initial investigations may help direct clinicians to the possible aetiology and guide treatment decisions. These infants require early referral to a paediatric liver centre were liver transplantation can be considered.

Aim:
This study highlights the aetiologies, associated features, helpful investigations and outcome of young infants presenting with liver failure.

Methods:
All infants presenting within 90 days from birth with a hepatic-based coagulopathy (INR ≥2) or encephalopathy were included in a retrospective case note review over a 19 year period (1993 - 2012).

Results:
81 infants (M=51) presented with liver failure; aetiologies included metabolic disease 30, (galactosaemia 11, tyrosinaemia 3, mitochondrial cytopathy 11, transaldolase deficiency 1, inborn error of bile salt metabolism 2, long chain 3-hydroxyacyl-CoA dehydrogenase deficiency 1, probable cholestatic liver disease 1), neonatal haemochromatosis (NH)7, infection 13, (Herpes simplex virus (HSV) 9, congenital CMV infection 2, enterovirus 1, E.Coli 1), infiltrative 6 (haemophagocytic lymphohistiocytosis 5, acute lymphoblastic leukaemia 1), hypoxic/ischaemic 15, hypopituitarism 3, and unknown 6.

Infants with metabolic disease were mainly jaundiced at presentation (77%, median bilirubin 207(16-767umol/L)) with mild transaminitis (Median ALT 85(16-1996 IU/L)). They had associated raised lactate (median 4(1.4-22 mmol/L)) however patients with mitochondrial cytopathy had markedly raised lactate (median 6.2(3-22 mmol/L)). Infants with infectious aetiology were mostly term (median gestation 39 weeks) but with low birth weight (median 2.7 kilograms). They had marked transaminitis (median ALT 895 (111-2726 IU/L)) and coagulopathy (median INR 6 (2.6-20)) at presentation. Infants with infiltrative disease mainly developed jaundice (83%), acidosis (83%), with moderate transaminitis (median ALT 562(36-1592 IU/L)) and a significantly raised lactate (median 10.6(6.9-12.6 mmol/L)). They also had the poorest outcome with 100% mortality. Infants with hypoxic insult had associated renal impairment (73%), acidosis (60%) and a high lactate (median 4.3(1.3-16 mmol/L)). Only five patients were transplanted (2 NH, 2 HSV and 1 mitochondrial) three of whom died post transplant. The overall survival rate was 59%.

Conclusion:
Aetiology of liver failure was established in 93% of infants. Careful attention to history, presenting symptoms and associated biochemical abnormalities may help guide in determining the aetiology of liver failure and therefore treatment decisions and prognosis.

Management of infantile gastroesophageal reflux in a district general hospital

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Introduction/Background
Gastroesophageal reflux disease (GORD) is the passage of gastric contents in to the oesophagus with or without regurgitation or vomiting; it covers a wide spectrum, from 'physiological' infantile to complicated GOR with severe oesophagitis.

The diagnosis of GORD in infancy remains clinical due to lack of sensitive and specific routine diagnostic tests, however there is no pathognomonic characteristic; management is empirical to a large extent, due to the lack of randomised controlled trials and large cohort studies in this age group. Local practice can therefore be variable.

Cow's milk protein intolerance (CMPI) is a well recognised condition in infancy which can mimic or aggravate GORD, and responds to dietary dairy elimination.

Aim
To assess management of infants diagnosed with GORD in a large district general hospital from April to September 2010.

Subjects/Methods
Retrospective audit. N=55 infants (0-12 months old) were diagnosed with GORD during this time period. Data were recorded from electronic medical records in a predesigned pro forma and were analysed with Microsoft Access.

Results
22% of all patients treated for GORD were eventually diagnosed with CMPI following resolution of symptoms after introduction of hydrolysed or aminoacid based formulas. 96% of all symptomatic infants presented within 4 months of age; 40% had attended the paediatric department twice, 38% once and 11% had attended more than three times. 25% had been admitted.

Main risk factor for GORD was prematurity and neurodevelopmental disorders; main symptom was vomiting or regurgitation (90%), followed by irritability and feeding difficulties (about 50%), back arching or colic (30%); about 15% had respiratory symptoms, 10% presented with failure to thrive and 5% had blood in stool.

In up to 90% of cases diagnosis was made clinically, however in10% of cases diagnosis was confirmed with upper GI endoscopy, pH or barium study.

96% of babies were given sodium alginate (gaviscon), 20% had thickened feeds and 30% had a trial of hypoallergenic feed ranging from one week duration up to 6 months (50% hydrolysed and 50% aminoacid based). 62% had acid suppressants for up to nine months (majority had ranitidine), 38% had prokinetics, mainly domperidone.

In all but 5% of cases followed up in hospital, symptoms resolved by first birthday.

Summary/Conclusion
Delay (up to five weeks) in diagnosis of CMPI and empirical introduction of dairy free diet in 22% of babies initially diagnosed with GORD, resulted in persistence of symptoms and repetitive visits to hospital. First line treatment for infantile GORD was sodium alginate, whereas regular (up to nine months) and frequent (62%) use of unlicensed anti-reflux medicines as second line for management of a self-limiting disease was noted, in spite of lack of evidence showing increased benefit/risk ratio from their prolonged use in infancy. Only in up to one third of cases hypoallergenic or thickened formulas were tried prior to administration of anti-reflux medication.

Need for adherence to evidence based guidelines advocating parental education, anticipatory guidance and alteration of feeding composition, including 2 week trial of hypoallergenic formula in unresponsive cases, is recommended in uncomplicated GORD; such management will also be optimal for treatment of underlying CMPI.

Objective criteria that help identify infants with GOR and CMPI are needed so as to facilitate early diagnosis and effective management.

Safety and efficacy of two forms of parenteral iron in children – regional experience

Dr Arun Urs, MBBS, MRCPCH[1], Dr Prithviraj Rao, MBBS, MRCPCH[1], Consultant Paediatric Gastroenterologist; Miss Valda Forbes, Specialist Gastroenterology Nurse[1]; Miss Natalia Iglesias, Paediatric and Neonatal Nutrition Pharmacist[2]; Dr John Puntis, Consultant Paediatric Gastroenterologist[2], [1]Sheffield Children's NHS Foundation Trust, Sheffield Children's Hospital, Sheffield, UK S10 2TH; [2]Leeds Teaching Hospital NHS Trust, Leeds General Infirmary, Great George Street, Leeds, LS1 3EX

Abstract:
Background: Iron deficiency anaemia is a common finding in gastroenterology patients as a result of dietary insufficiency, malabsorption and occult or overt gastrointestinal bleeding. Due to poor tolerance of oral iron supplementation some patients require parenteral administration. There is limited experience with use of parenteral iron in the paediatric population with gastrointestinal disease.

Aim:
To evaluate two centres' experiences of safety and efficacy of IV low molecular weight dextran (Cosmofer®) and IV iron maltoside 1000 (Monofer®) at 8-12 weeks after treatment in children with gastroenterological conditions.

Methods:
Children (< 16 years) received Cosmofer® at centre 1 and Monofer® at centre 2 over a two year period. Data abstracted included primary indication, underlying diagnoses, dosing and administration, laboratory values before and after therapy, and adverse reactions. The decision for parenteral iron was made on an individual basis. The total iron deficit was calculated using the Ganzoni formula in both groups with maximum dose of 20mg iron/kg as single infusion in the Monofer® group. In the Cosmofer® group, doses were split and a test dose over 1 hour given prior to infusion over 5 hours. Monofer® is a rapid one-visit total dose intravenous iron preparation that can be given over 1 hour without test dose. The main indication in both groups was malabsorption from short gut, and inflammatory bowel disease. There is no difference in licensing between either of the preparations.

Results:
There were 13 infusions of Cosmofer® (11 children) and 9 of Monofer® (8 children). The groups were comparable except the Cosmofer® group smaller in age/wt. and received less iron (Chart 1). Mann Whitney test was used to evaluate statistical difference for pre and post haemoglobin rise in both groups ($p<0.01$) Chart 2. One patient from each group experienced allergic reaction within few minutes of infusion but made fully recovery. There were no delayed effects reported on follow up.

(Chart 1) Baseline characteristics

	Cosmofer	Monofer
No. of children	11	8
% Male	72.7	62.5
Age yr Median (range)	8.4 (0.41-15.41)	12.3 (5.58-14.5)
Weight kg Median (range)	19 (6.36-37)	32.9 (18.7-56.4)

(Chart 2) Increase from baseline in Hb and MCV (8-12 weeks)

	Cosmofer	Monofer
Hb (g/dl) Median (range)	2.35 (0 to 5.5)	3.65 (0.1 to 5.7)
MCV (fl) Median (range)	4.5 (-4 to 17)	7.9 (-1.3 to 17.2)

Conclusion:
In children with gastrointestinal disease who are intolerant of oral iron, parenteral therapy is effective in correcting anaemia. The best formulation of parenteral iron is uncertain, and serious allergic reactions were observed with both preparations used. Monofer offers the potential advantage of a single, rapid infusion and simple dose calculation.

THURSDAY POSTERS

Cholesterol ester storage disease – a single centre experience

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Introduction:
Cholesterol ester storage disease (CESD) is a rare autosomal recessive disorder due to late onset lysosomal acid lipase (LAL) deficiency. There are relatively few documented clinical reports hence the clinical phenotype is not well defined and its natural history uncertain. We describe a series of 4 cases seen in a single centre.

Subjects and methods:
Patients presented between the ages of 1 and 13 with either organomegaly (n=4) and/or abdominal pain (n=3). At presentation all had hepatomegaly and one had splenomegaly. All had abnormal transaminases and three had hyperlipidaemia. All were confirmed to have LAL deficiency and were compound heterozygotes for 2 pathogenic mutations in the *LIPA* gene.
Three have residual abdominal pain suggestive of gastro-oesophageal reflux with partial response to drug treatment. Three developed biliary dyskinesia which responded to surgery. All were treated with a low fat diet which resulted in resolution of diarrhoea where present. Three were treated with statins with a good initial lipid lowering effect.

Results:
During a mean follow up of 11 years; two developed new onset splenomegaly, of whom one subsequently died following a road traffic accident at the age of 16. The other remains well age 22 and continues on statin treatment. One underwent liver transplantation aged 9 due to hepato-pulmonary syndrome and remains well with normal lipid levels six years on. The final subject also remains well aged 15 with persistent hepatomegaly and good lipid control on statin treatment.

Summary:
In summary CESD has a heterogeneous clinical presentation with hepatomegaly being a uniform feature in our series. There is a high incidence of biliary and abdominal symptoms. Hyperlipidaemia usually responds to statin therapy. Splenomegaly develops commonly and liver disease appears progressive. Liver transplantation is highly successful where indicated.

Does prednisolone improve outcome in non-A-E hepatitis?

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Introduction
Non-A-E hepatitis (NA-EH) is a leading cause of acute liver inflammation in children. It has a presumed infective aetiology, is associated with aplastic anaemia, and may lead to acute liver failure (ALF) requiring liver transplantation (LT). Diagnosis is based on clinical features and exclusion of other causes of hepatitis. Anecdotal evidence and small studies of prednisolone treatment for acute liver failure are inconclusive.

Aim
To describe the outcome of non-A-E hepatitis and assess the effect of prednisolone on clinical course.

Method
A retrospective analysis of consecutive children (<16 years) transferred to a supraregional liver centre, subsequently diagnosed with NA-EH from 2001 to 2012. ALF was defined as INR > 1.5 (PT > 18) or presence of encephalopathy. Children received a trial of prednisolone (2mg /kg maximum 40mg) according to clinical judgement and evolving unit experience. Children were listed for supra-urgent liver transplantation when they fulfilled strict criteria, which include severity of coagulopathy and encephalopathy.

Results
25 children, M16: F9 median age 8.1 years (range 1.1 – 15.66 years) were identified. Six children without ALF recovered without LT; two had received prednisolone. Of 19 with ALF, 10 reached listing criteria and underwent successful LT (median age 7.25 years, range 1.83 yr – 15.83 yr). Three had received prednisolone, of whom two were listed for LT within 24 hours and the third within 3 days of treatment. Of seven undergoing LT without prior prednisolone, admission INR was median 3.2 (range 1.5 – 10.1) and interval to listing was median 2 days (range 1-11). Five were listed within 2 days, and the other two 6 and 11 days after admission. Nine with ALF recovered without LT (median age 11.91 yr, range 6y – 14.6 yr), of whom six had received prednisolone. Median INR before treatment was 1.8 (range 1.7 – 2.2). In three who recovered spontaneously, all had peak INR of 1.6. Nine children (36%) developed aplastic anaemia. One had low platelets at onset of liver dysfunction. In eight, features of AA developed after onset of NA-EH. In two AA developed after LT and in six after improvement with steroids (5) or spontaneous recovery (1).

Conclusions
Prednisolone may have a beneficial role in the treatment of children with ALF due to NA-EH if started early and before encephalopathy ensues. Prednisolone treatment and immunosuppression after LT do not prevent subsequent aplastic anaemia.

Eosinophils in the colonic mucosa (CE): clinical and epidemiological relevance in children: a 4 year follow up cohort study

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Background:
Primary eosinophilic gastrointestinal disease is characterized by eosinophilic inflammation which is idiopathic i.e. in the absence of parasitic infection, drug reaction, or malignancy.

Objectives:
Our primary aim was to identify the estimated incidence, prevalence and clinical significance of colonic eosinophilia (CE). Secondary aims: To evaluate our services and identify what treatments worked more effectively for CE and to identify any histological features which may indicate evolution to inflammatory bowel disease
Setting: Tertiary care.
Study period: January 2006 to December 2008 (3 years) with follow up till September 2012.
Participants: All consecutive cases with colonic eosinophilic infiltration were examined.and were followed for 4 years until sep 2012.

Results:
55 (35 male) patients with CE were identified, Median age 10.8 (0.5-17) years. 43 % presented with diarrhoea, 32% with abdominal pain and 24% with per-rectal bleeding.
Initial diagnoses included :

a) 22 cases of Inflammatory bowel disease (IBD) (9 Crohn's, 8 UC, 5 indeterminate colitis). In the same time period the incidence of IBD at our centre was 87 patients thus CE was seen in 25 % of our patients with IBD;
b) 17 cases of allergic colitis;
c) 9 were determined as an incidental finding and
d) 7 were classed as idiopathic

Follow up data was available on all 22 patients with IBD and 16/22 (72%) remained stable (in remission) on initial maintenance therapy and did not warrant any escalation beyond second line maintenance medication.
In the allergic colitic group complete clinical response was seen to dietary elimination of antigens in 8/17 (47%). In the 1 child with colonic eosinophilia that evolved into IBD there were no particular predictive histological features.

Summary and Conclusion:
The estimated incidence of paediatric colonic eosinophilia in the South Yorkshire region is 7/100000 children. 87% of cases with CE were associated with an underlying organic pathology. CE was seen in 25% of patients with IBD and most (72%) did not require escalation of treatment beyond a second line maintenance agent. CE thus may be a predictor of a relatively benign course in children with IBD. Dietary elimination of antigens is associated with a good response in children with allergic colonopathy. We need further multicentred longitudinal studies to have a understanding of this condition

Facial morphology in Alagille Syndrome patients; Stereophotogrammetry Three-Dimensional Anthropometric (Pilot Study)

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Introduction
The facial features of Alagille Syndrome ALGS (triangular fascies, pointed chin, broad forehead, deep set eyes, bat ears) [1] represent one of the 5 major diagnostic criteria. However, there remains a debate as to their specificity for the syndrome and there is no information as to the significance of each of the components.[1]

Our first aim was to establish the methodology for our research application. We wanted to define the Alagille face numerically and to establish the facial dimensions recognised using the facial anthropometry method that corresponds to the ALGS facies. We also wanted to determine whether there was a statistically significant facial phenotype. An anthropometric model could be used as a diagnostic tool with 3D facial recognition software.

Methods
We measured the absolute and relative dimensions of 52 facial landmarks (12 Midline landmarks plus 20 Paired side landmarks) in 7 participants.

Equipment: A Three-Dimensional image capture and analysis system was used to map the facial soft tissue, with four linked cameras capturing simultaneous photographs of the patient. A specific dimensional imaging software program allows a 3D image to be assembled from the data recorded by the individual cameras[2]

3D photographs were obtained from patients with Alagille Syndrome AGLS and assembled through a custom built 3D software program (Cloud, Robin Richards, London, UK) which calculates distances and angles between landmarks, and measures surface areas prompting detailed analysis of the facial soft tissue features[2]

Results
We were able to measure facial dimensions that could display volume differences for patients with ALGS. Preliminary analysis of the dimensions and distances have shown promise in development of a formula to use for ALGS. Based on the characteristic facies the fallowing dimensions could be expected to define the ALGS phenotype

Phenotype	Anthropometric Measurement
Triangular Face	Distances: Δ pogonion -frontotemporale (Rt) pogonion -frontotemporale (Lt) frontotemporale (Lt) - frontotemporale (Rt)
Prominent / Broad Forehead	Distances: frontotemporale (Lt) - frontotemporale (Rt) trichion – glabella / nasion
Pointed Chin	Angle:< Tragion (Rt) gnathion Tragion (Lt)
Deep-set Eyes	Distances: Exocanthion (Rt) - Tragion (Rt) Exocanthion (Lt) - Tragion (Lt)
Moderate hypertelorism (Eyes)	Distances: Endocanthion (Rt) - Endocanthion (Lt) Exocanthion (Rt) - Exocanthion (Lt)

Table 1 – Example of anthropometric landmarks facial features studied

Fig. 1(a) Fig. 1 (b):
An example of measuring the distances between anthropometric facial landmarks

Conclusion
Preliminary results suggest that individuals with AGS display distinct facial phenotypes. Study of additional individuals will show whether there are differences between AGS patients, patients with other cholestatic diseases and healthy controls based on these facial characteristics.

References:
Binita M. Kamath, Kathleen M. Loomes, Ian D. Krantz; Facial Features in Alagille Syndrome: Specific or Cholestasis Facies? American Journal of Medical Genetics 2002,112:163–170
Trevor J. Coward, Robin Richards, Michael Fenlon, Brendan J. J. Scott. Development of a Stereophotogrammetry Technique to Assess Facial Change Following Surgery for Head and Neck Cancer' Int J Prosthodont, 2011 24 (4), pp. 342-344.

Glycogenic hepatopathy – an avoidable complication of insulin dependent diabetes mellitus or a consequence of mitochondrial dysfunction?

E Fitzpatrick[1], C Cotoi[2], A Quaglia[2], N Hadzic[1]. [1]Paediatric Liver Centre and [2]Institute of Liver Studies, King's College London School of Medicine at King's College Hospital, Denmark Hill, London SE5 9PJ

Introduction:
Mauriac disease, the association of growth failure, massive hepatomegaly and Cushingoid features in children with poorly controlled type 1 diabetes should be obsolete with modern insulin regimes and monitoring. Over the last 10 years more than 30 children have presented to our paediatric liver centre with the syndrome however. Though the condition is closely associated with a high HbA1C, the aetiology is not fully understood.

Methods:
Patients diagnosed with glycogenic hepatopathy, the hepatic manifestation of Mauriac syndrome, were prospectively identified. Clinical, histological and biochemical data were then reviewed. SPSS 17.0 was used for statistical analysis.

Results:
Thirty one children (16 boys) with a median age of 15.1 years (IQR 14, 16) presented within the study period. The median age of diagnosis of diabetes was 10 years of age (IQR 11, 16). Median units of insulin per kilogram per day was 1.33 with a median HbA1C in the group of 11% (IQR 9.9, 12.4%). The growth of the children in the cohort was impaired with a median height z-score for age of -1.01 (IQR -1.73, 0.4) and for weight -0.11(IQR -0.8, 0.17) with a median BMI z-score of 0.28 (-0.12 0.67). Hepatomegaly was universal with splenomegaly present in 5 (16%). In all children, abdominal ultrasound demonsted an echogenic liver. Transaminases were abnormal in the majority with a median AST of 76IU/l (IQR 44, 187), ALT of 76IU/l (IQR 69, 177) and median GGT of 71IU/l (IQR 47, 114). Though no child was ketoacidotic at time of measurement, lactate was abnormal in 16 children, one child had a blood lactate of 9mmol/l. Liver biopsy was undertaken in 19 children (61%). All showed the presence of enlarged hepatocytes wth clear cytoplasm with glyogenated nuclei in 17 of 19. Steatosis was also present in the majority; this was mostly macrovesicular in nature. Interestingly, inflammation was present in 8 (42%); this was mainly periportal. Fibrosis was seen in 14 (73%) and was mild in the majority of cases though 2 children had bridging fibrosis. Steatosis, inflammation and fibrosis have not been well described previously in this condition. Megamitochondria were seen in 7 biopsies. One boy with an especially high serum lactate was found to have mitochondrial depletion of 29% of normal in liver biopsy. It is not clear whether this depletion was a primary or secondary phenomenon. The presence of megamitochondria correlated with AST (p=0.026) and presence of fibrosis on biopsy. At follow-up to 6.5 years following presentation, 17 children had normal or improved transaminases, in 13 there was no change. Transaminases tended to follow the trend of the child's HbA1c.

Discussion:
Despite modern insulin regimens and monitoring in children with type 1 diabetes, Mauriac syndrome still exists. Although previous reports have not found inflammation or fibrosis in liver tissue, we have found both in significant amounts. The association of megamitochondria and high lactate levels in children with the condition lend suggest possible mechanism for the pathophysiology of the condition.

High prevalence of vestibular dysfunction in childhood cyclic vomiting syndrome.

Fernanda Christofori, Efstratos Saliakellis, Jutta Köglmeier, Osvaldo Borrelli, Ewa Ragulan & Keith J Lindley
Great Ormond Street Hospital for Children NHS Foundation Trust, London WC1N 3JH

Introduction
Cyclic vomiting syndrome (CVS) in childhood is usually a benign condition which improves with age. Triggers of the emetic episode include intercurrent infections, travel, tiredness, excitement and in older girls menstrual periods. Once an emetic episode is triggered there follows a commonly stereotypical sequence of events during the emetic phase followed by a rapid return to normality.

The vestibular apparatus provides important sensory input to the brainstem vomiting centre and modulates the emetic threshold. We hypothesised that vestibular dysfunction might lower the threshold for emesis in CVS.

Methods
Retrospective review of medical records of consecutive referrals with CVS to a single tertiary centre who had undergone formal vestibular function testing as part of their diagnostic evaluation.

Results
Records of 55 patients with CVS, 31 female and 24 male, average age of onset 66 months were reviewed. In 7 children CVS episodes were triggered by travel, in 11 stress / excitement, in 5 infections, in 3 menstruation and in 30 no trigger was identified. 23 suffered from motion / travel sickness, 4 from tinnitus and 9 had hearing problems. 16 reported poor balance and 23 reported headaches.

5 had conductive hearing loss on pure tone audiometry.

11/33 had an abnormal Unterberger's test consistent with vestibular dysfunction, and 9/16 had abnormal bithermal caloric testing. All 55 had undergone either electronystagmography (ENG) or videonystagmography (VNG), a form of video-oculography (VOG). 6 had abnormal smooth pursuits, 2 optokinetic nystagmus, 12 had directional preponderance on rotational impulse testing, and 14 abnormalities on sinusoidal chair rotation.

Overall 19/55 had evidence of vestibular dysfunction and 12 of these underwent therapies aimed at improving vestibular function. 44/55 had cranial MRI scans of which 5 were abnormal, although none with progressive pathology.

Conclusion
Vestibular abnormalities are prevalent in childhood CVS in the context of a tertiary referral unit. Appropriate treatment of these abnormalities could potentially modulate emetic threshold favourably and lead to a reduction in frequency of emetic episodes.

Improved patient experience in young people with Inflammatory Bowel Disease undergoing transition following changes to the transition pathway

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Department of Paediatric Gastroenterology, Hepatology and Nutrition, Sheffield Children's Hospital

Background:
A transition service is an essential part of quality care for young people (YP) with IBD and helps to effectively transfer a growing cohort of YP with IBD from a family focussed child centred model of care to adult orientated care.

We reorganised our transition pathway two years ago and the main changes included starting a YP clinic which helped initiate the process of transition and ensure readiness for transfer with an adolescent checklist (annual adolescent transition plan), improved documentation and a choice of transition clinics with two adult teams held at the Children's hospital. Care is then transferred after a handover clinic at the adult hospital for the majority of young people.

Aim:
To assess the impact of the new transition pathway on patient experience in YP with IBD.

Methods:
A retrospective comparative postal-questionnaire based survey was done in two groups of YP with IBD. Group 1- had been transitioned and care transferred to the adult services in 2008-2010 and Group 2- YP who underwent transition and transfer in 2011 after the transition pathway was reorganised.

Results: 26 (45%) YP returned the questionnaires in Group 1 and 14(56%) returned the questionnaires in Group 2.

Survey questions	YP transitioned 2008-2010 (n =57)	YP transitioned 2011 (n=25)	Impact
Age at transfer of care to adult services	median age 17.6 (16 – 19 years)	Median age 17 (16-17.9 years)	
Was transition process discussed prior to transition clinic?	23 (88%)	14 (100%)	
What age did this discussion take place?	Median age 16.7 (15 – 19 years)	Median age 16 (15-17.6 years)	
Were your questions about transition answered satisfactorily? (scale 1-5) Scale 2-3* Scale 4-5* Not scored	Median 4 (range 2-5) 19% 77% 4%	Median 5 (range 4-5) 0% 100%	19% improvement
Would you have liked more information before transition clinic?	Yes (30%)	No	Adequate information provided in 100%
What was your experience of the transition clinic? (scale 1-5) Scale 2-3* Scale 4-5* Not scored	Median 4 (range 2-5) 31% 62% 7%	Median 5 (range 4-5) 0% 100%	31% improvement
Would you have liked to visit the adult hospital before the first clinic appointment?	No 10 (38%) Yes 14 (54%) Had visited 2 (8%)	No 12 (86%) Yes 1 (7%) No answer 1(7%)	

*Scale 1- unacceptable, 2- Poor, 3-Average, 4-Good, 5-Excellent

Summary and Conclusions:
Whilst there's a degree of recall bias, the new transition pathway had a measurable positive impact on patient experience. We continue to seek feedback from our YP to further improve our transition services and are now seeing YP at a younger age in the YP clinic to initiate earlier discussion about the transition process.

Interventions for the treatment of iron deficiency anaemia in patients with inflammatory bowel disease.

Sophie Tudor Jones[1], Morris Gordon[2], Anthony Akobeng[2]
[1]University of Manchester; [2]Department of Paediatric Gastroenterology, Royal Manchester Children's Hospital, Manchester, UK.

Background:
Iron deficiency anaemia (IDA) is common in patients with inflammatory bowel disease (IBD). There are many different medications to treat IDA in IBD, however the optimal preparation and route of administration is not clear. We set out to systematically evaluate the efficacy and safety of interventions for treating IDA in patients with IBD.

Methods:
Randomised controlled trials comparing iron therapy to placebo or other interventions in patients with IBD and IDA were included. Data sources were MEDLINE, EMBASE, Cochrane Central Register of Controlled Trials, Cochrane inflammatory bowel disease and functional bowel disorders Specialised Register and reference lists of retrieved articles. Data extraction and assessment of methodological quality were performed independently by two reviewers. Data was analysed according to the intention to treat principle.

Results:
7 studies involving 907 participants met the inclusion criteria. 3 papers compared Intravenous (IV) iron sucrose with oral iron preparations, two compared subcutaneous (S/C) erythropoietin (EPO) with placebo, one paper compared IV ferric carboxymaltose with oral iron and one compared IV ferric carboxymaltose with IV iron sucrose. Risk of bias amongst included studies was variable, with some concerns regarding blinding and selective reporting of results.

In three studies, patients receiving either IV or oral Iron preparations had statistically significant rises in haemoglobin (hb) and Ferritn levels, although meta-analysis showed hb increases were marginally higher in patients receiving IV iron (Weighted mean difference 0.28, 95% confidence intervals (CI) 0.05- 0.50). Adverse events requiring withdrawal of therapy were statistically significantly lower in patients receiving IV Iron (Odds Ratio 0.11, 95% CI 0.03- 0.43). The overall body of available evidence showed no worsening of disease activity in patients who receive oral iron therapy compared to those who receive IV iron.

There was individual trial data to suggest erythropoietin may be more effective than placebo at increasing Hb levels and no change in disease activity was reported. Due to heterogeneity of data, no meta-analysis was possible. EPO was generally safe and well tolerated.

Conclusions:
Both IV and oral Iron preparations appear effective at improving hb and Ferritin levels, although the increase is marginally higher with IV Iron. The available evidence suggests that IV iron is better tolerated than oral iron, but the data set is small and of variable quality. There is some evidence that EPO is effective and safe, but again samples were small and studies were heterogeneous, limiting the strength of these findings. Conclusions about other forms of treatment or comparisons cannot be made due to limited available data. Further research appears warranted.

Paediatric GI Endoscopy: A Qualitative Study

A Wahid; Paediatrics, Watford General Hospital K Devarajan: Paediatric Gastroenterology, Addenbrooke's Hospital, Cambridge M Zilbauer: Paediatric Gastroenterology, Addenbrooke's Hospital, Cambridge R Heuschkel: Paediatric Gastroenterology, Addenbrooke's, Cambridge

Introduction:
Gastrointestinal endoscopy is an invasive procedure used to diagnose and/or treat diseases of the gut. As with any invasive procedure there is a small but not insignificant risk of complications and it is therefore important that due consideration is taken when reviewing the indications for upper and lower GI endoscopy, particularly in children. Despite this, there remains a wide variation nationally in clinical practice amongst paediatric gastroenterologists.

Background:
The paediatric gastroenterology service at Addenbrooke's Hospital carries out approximately 500 procedures a year with almost all children investigated as day cases under general anaesthesia. In the absence of a standard, we reviewed our current practice with a view to developing an audit tool that could facilitate the comparison of endoscopy 'conversion' rates nationally i.e. number of endoscopies performed / new patient referral. Although the primary intention was to look at the proportion of new patients requiring endoscopy, we also reviewed the clinical impact of the procedure on management.

Method:
We collected data retrospectively using electronic medical records (eMR) and the endoscopy database kept by the Dept of Paediatric Gastroenterology. A list of new clinic patients who had endoscopy within three months of their appointment was obtained, dating from 1st January to 31st December 2011. Individual eMR records were reviewed for the type of endoscopy, indications, interval to endoscopy, histology findings, diagnoses, and impact on management.

Results:
18% of the 705 new referrals made to clinic over the year went on to have an endoscopy; half of these had both OGD and colonoscopy. 39% were diagnosed with inflammatory bowel disease accounting for 3% of the patients referred. All of the OGDs reviewed in this study were diagnostic in nature; 39% were for raised anti-tTG antibody, and these were positive for coeliac disease in each case. However, a large proportion of non-coeliac OGDs were reported to be normal and 1/5 patients with a normal OGD were referred for psychological assessment.

Discussion:
This study sets an initial benchmark for endoscopy practices in a regional paediatric gastroenterology service. It should provide a useful dataset against which similar units can validate their own endoscopy service and help inform commissioners planning future services. A standardised audit tool should also help identify and explain variations in care and hence improve standards of care across the country.

The use of Sirolimus for Refractory Inflammatory Bowel Disease: a case series

Blackstock J, Mutalib M, Kiparissi F, Lindley K Great Ormond Street Hospital, Great Ormond Street, London WC1N 3JH

Background:
Refractory inflammatory bowel disease (IBD) in children is challenging to treat as there are limited therapeutic options. Animal models and case reports suggest a therapeutic benefit of Sirolimus in IBD. Sirolimus inhibits the mTOR pathway preventing T cell proliferation and B cell activation by inhibiting the response to IL-2. It also promotes immunoregulatory T cells which play an important role in mucosal healing in IBD.

Objectives:
The primary outcome of this study was to assess the therapeutic efficacy of Sirolimus in children with refractory IBD. Secondary outcomes included Sirolimus tolerance and duration of therapy.

Method:
We retrospectively reviewed medical and nursing records of all patients with IBD on Sirolimus (23 patients) between 2006 and 2012. Clinical response was assessed by PUCAI or PCDAI. All patients had an activity index at baseline and 3 months after induction of Sirolimus. Endoscopic and histological findings before starting Sirolimus were compared to the next available endoscopy at least 3 months later. A total of eight cases were excluded, 6 due to incomplete data and 2 due to short treatment time (less than one month).

Results:
All 15 cases had severe refractory IBD, 4 were Crohns disease (CD), 11 were Ulcerative colitis (UC). Nine were males, 6 were females. The mean average age of onset of disease was 8.7 years. The mean number of years after diagnosis until induction with Sirolimus was 3.1 years. The oral daily dose range was between 0.5mg and 5mg with a targeted serum trough level of 5-10ng/ml. The mean trough level was 7.5ng/ml. Before commencement of Sirolimus all 16 patients had failed standard medical therapy and all stayed on conventional treatment while on Sirolimus. All 15 patients had received steroids and thiopurines and 5-ASAs, 13 had received infliximab, 8 had received adalimumab, 5 had received methotrexate, 2 had received tacrolimus and two had received basiliximab prior to starting Sirolimus. 8/11 patients with UC had a response shown by improvement in their PUCAI score at 3 months, 3/11 had no response. Of the patients who responded 2 achieved remission with concomitant administration of Basiliximab, one was started on Mycophenalate while the fourth child required complete gut rest and parenteral nutrition for 6 weeks. 3/8 responders and 2/3 non responders went on to have Colectomys. One patient was weaned off adalimumab after commencement on Sirolimus. Of the patients with CD 3 had a clinical response according to their PCDAI scores. One CD patient was successfully weaned off prednisolone and one was weaned off methotrexate. We were unable to create scores for one patient however no response was achieved. Endoscopy results did not appear to correlate well with PCDAI/PUCAI scores however these were not performed at standard intervals, many were performed during flare ups and well after the 3 month period when the scores were calculated. 5/11 patients with UC achieved histological mucosal healing while 4 patients still had some disease activity. Of the CD patients one achieved mucosal healing while 3 demonstrated ongoing inflammation, No side-effects were reported to Sirolimus in the included patients.

Conclusion:
Our study was limited by its retrospective design; however we demonstrated clinical and histological response to Sirolimus in children with refractory IBD who failed conventional medical therapy. With careful patient selection, Sirolimus (as an adjunct immunosuppressant) appears to be safe and effective medical therapy to be considered in children with severe refractory IBD.

UK regional variations in nutritional issues of controversy in paediatric cystic fibrosis (CF) care.

Chris Smith[1]; Helen White[2]; Dee Shimmi[3]; David Croo[4] ; Paul Seddo[1]; Assad Butt[1]
[1]Royal Alexandra Childrens Hospital, Brighton; [2]Leeds Metropolitan University/St. James' Hospital, Leeds; [3]Belfast City Hospital; 4University of Brighton

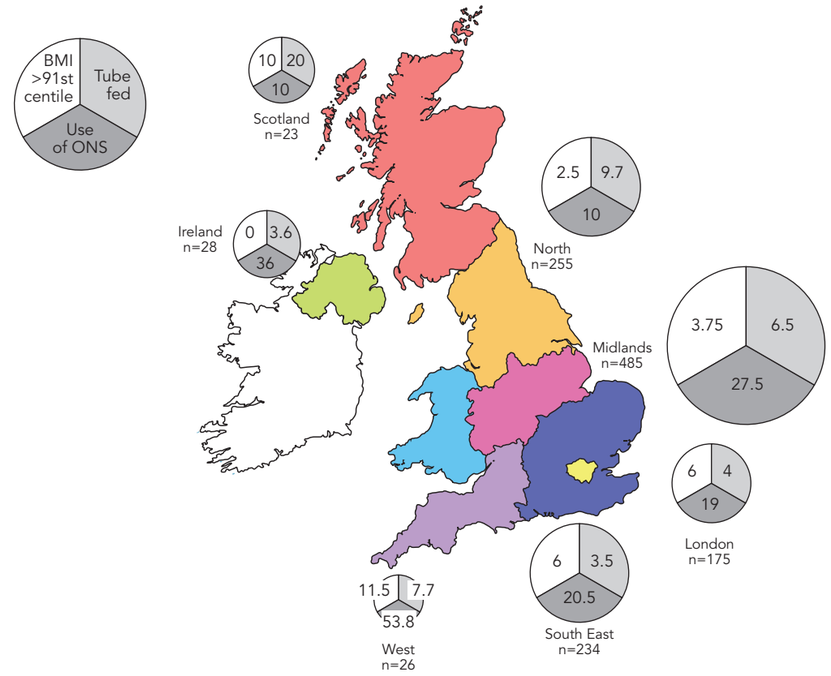
Introduction:
Nutrition is a key aspect to CF care. Use of oral nutritional supplements (ONS) and tube feeding (TF) in CF remain controversial due to scarcity of RCT's. Obesity is an emerging concern. The CF registry collects data but does not report results geographically.

Aim:
To examine differences in prevalence of use of ONS, use of TF and overweight status (defined as BMI >91st centile) across the UK.

Methods:
As part of a pan European survey a questionnaire was developed with the chairs of the European Cystic Fibrosis Nutrition Group (ECFNG). This was circulated as a personalised email to all members via the groups email database. Questionnaires were then returned via email and the data was exported into spreadsheets and tabulated manually. Data collection took place over a 6 month period between February 2012 and August 2012. The questionnaire contained demographic data on the respondent's case load.

Results:
63 emails were sent out with 39 Dietitians responding from across Europe. 10 responses were received from the UK (8 England, 1 Scotland, 1 N. Ireland) representing a total of 1226 patients.

Size of circle for each region represents sample size Figures are % prevalence



Regional variation is noticeable in the prevalence of BMI >91st centile with higher numbers seen in the South of England and Scotland. There is a considerable range of supplement use from 10 - 53% of case loads. Tube feeding prevalence in Scotland (1 centre) was double that of any other region.

Summary and Conclusion:
Reports of prevalence of BMI >91st centile in CF are limited in the literature and this is the first report to describe regional variation. Regional differences in TF and ONS practice are also unreported. The survey was limited to members of the European CF group: however our overall data on prevalence are very similar to the CF registry figures for 2011 (ONS: 20.5% vs. 20.1% and TF 6.5% vs. 5.9% respectively) suggesting this was a representative sample. This explorative study suggests further work is needed to clarify and unite practice in these areas.

Where did the salt go A challenging case of bronchiolitis

Dr Siba Prosad Paul, Yeovil District Hospital (Previously at GWH, Swindon); Dr Sarah HICKS, Bristol Royal Children's Hospital (Previously at GWH, Swindon); Dr Lucy Grain, Great Western Hospital, Swindon

Introduction:
Bronchiolitis is common and usually managed in the community. Some infants may require hospitalisation for feeding support or oxygen therapy. A small minority of infants requires CPAP or ventilator support.

Extrapulmonary manifestations of bronchiolitis include hyponatraemia and SIADH. We present a challenging case of bronchiolitis complicated by severe hyponatraemia and highlights the need for consideration of a dual pathology in such cases.

Methods:
A 4 week old ex-35/40 week infant with an uneventful neonatal period and did not require an admission to Neonatal Unit. She was admitted with RSV bronchiolitis for oxygen therapy. She was tolerating bottle feeds at 100ml/kg/day. 30 hours into admission she desaturated acutely after feeding and was seen to have a focal seizure.

Results:
Capillary blood gas showed moderate respiratory acidosis with severe hyponatraemia (114mmol/L). She had borderline low serum osmolality (248mosmol/kg) with inappropriately high urine sodium (55mmol/L) and urine osmolality (390 mmoL/L). Serum cortisol was appropriately raised. Sodium levels normalised over 48 hours after treatment with 2.7% hypertonic saline.

She subsequently deteriorated requiring transfer to PICU for ventilatory and inotropic support. Prior to transfer she had a bilious vomit with an abnormal abdominal X-ray. A GI contrast study few days later in PICU showed malrotation which was surgically treated. Initial results were consistent with inappropriate ADH secretion but the hyponatraemia appeared to be out of proportion to the severity of her respiratory illness at that time and remained a challenge to explain. Retrospectively, the malrotation could help explain this unexpectedly low serum sodium seen.

Discussion :

Hyponatraemia and SIADH are recognised complications of bronchiolitis
Important to investigate severe hyponatraemia: osmolality, blood glucose, cortisol, etc
Important to consider other causes, including surgical, when clinical picture and biochemistry do not correlate
Dual pathology presented a clinical challenge as an apparently stable infant with bronchiolitis suffered desaturation secondary to convulsion.
Further raises a query regarding the use of hyposmolar fluids (0.45% NaCl with dextrose) in infants with bronchiolitis

Conclusion:
This case demonstrates that 2 serious co-existing pathologies can present a clinical challenge in a relatively common paediatric condition such as bronchiolitis. There is a need to revisit the initial diagnosis if clinical and biochemical features remain unexplained.

FRIDAY SESSION V
INVITED SPEAKER ABSTRACTS

Intestinal Failure Associated Liver Disease

Olivier J. Goulet MD, PhD, Department of Pediatric Gastroenterology, Hepatology and Nutrition, Intestinal Failure Rehabilitation Center, National Reference Centre for Rare Digestive Diseases

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"Intestinal failure" (IF) is caused by the critical reduction of functional gut mass below the minimal amount necessary for adequate digestion and absorption to satisfy body nutrient and fluid requirements for maintenance in adults and growth in children. The advent of parenteral nutrition (PN) resulted in a dramatic improvement in life expectancy of patients suffering IF especially those with short bowel syndrome. PN made possible the emergence and the diagnosis of rare congenital digestive diseases, as those involving the development of intestinal mucosa (microvillous inclusion disease, tufting enteropathy or syndromic diarrhea) or severe intestinal pseudo-obstruction syndromes. However, PN has its own complications, such as PN related sepsis, thrombosis, cholestasis or bone disease. IF associated liver disease (IFALD), is a common and potentially life-threatening problem for pediatric patients receiving long-term PN for IF. In neonates and young infants sepsis, catheter related or originating from the intestine through translocation, play a major role in the development of IFALD. New approach consisting in targeting harmful cytokine responses can be expected to reduce the severity and frequency of IFALD.

Appreciation of risks factors, improvements in surgical procedure, ICU management, involvement of nutrition support teams, as well as progresses in the type and mode of delivery of PN, including the use of fish oil based lipid emulsions, and enteral feeding, may contribute to decrease the incidence of end-stage IFALD. Intestinal and liver transplantation have an important role in avoiding or treating end-stage liver disease, especially in young children, although a shortage of suitable donors means that the mortality remains high. ITx has became a well established procedure achieving long-term intestinal function. Isolated liver transplantation for SBS infants with end-stage IFALD was reported by several groups, but should no longer be necessary. Reversal of cholestasis and fibrosis were shown after longitudinal intestinal lengthening and tailoring (LILT), serial transverse enteroplasty or isolated ITx and should be considered in selected infants and children with SBS.

Surgical Management of Intestinal Failure

Professor A Pierro, Nuffield Professor of Paediatric Surgery and Head of Department of Paediatric Surgery Institute of Child Health, University College London Medical School, 30 Guilford Street, London WC1N 1EH

Chronic intestinal pseudo-obstruction (CIPO) – are we moving in the right direction?

Miss K Larmour, Principal Dietitian for Neurogastroenterology, Great Ormond Street Hospital for Children NHS Trust, London

Chronic intestinal pseudo-obstruction is a severe syndrome characterized by a profound derangement of intestinal propulsive motility that resembles mechanical obstruction (although there is no mechanical obstruction), and is one of the main causes of intestinal failure. In 2010 Great Ormond Street Hospital secured funding to provide a national diagnostic service for infants and children with CIPO disorders. The service is delivered by a multidisciplinary team including paediatric gastroenterologists, a nurse specialist, a psychologist and a dietitian.

Nutritional management is of crucial importance in the treatment of this population of mainly very young children involving the administration of special formulas by nasogastric tube, percutaneous gastrostomy or jejunostomy. In the most severe cases, parenteral nutrition is mandatory in order to satisfy nutritional requirements and manage obstructive episodes. During the planning phase of this new service, a delivery pathway has been created to outline the process of dietetic care from the point of referral to the service until discharge, and to assess the impact of recommended interventions on selected nutritional parameters. A Standardised Nutrition Assessment Tool has been produced so that consistent information can be collected for each patient.

The nutritional assessment is carried out twice: at baseline at the point of referral to the service, and after any intervention such as changes to feeding plan (oral/enteral), stoma formation, or starting home parenteral nutrition. If no intervention is made, the assessment is repeated within a year of the initial assessment. The information collected is transferred to a database. A number of nutrition related outcomes are considered including a. nutritional status (as assessed by anthropometry and nutritional bloods); b. administration of nutrition, such as route of feeding and type of feed; c. change in quality of life, regarding symptom change and hours/day spent feeding.

Between May and December 2012, 22 patients have been assessed and we now report preliminary results.

British Intestinal Failure Survey

Dr Andrew Barclay, Consultant Paediatric Gastroenterologist, Royal Hospital for Sick Children, Glasgow

Prolonged parenteral nutrition (PN) and home parenteral nutrition (HPN) are the established treatments of choice for children with established (type II) intestinal failure. Long-term survival on HPN has been readily described and patient populations have risen year on year. Major complications of PN; catheter related blood stream infection (CRBSI) and intestinal failure associated liver disease (IFALD) have limited morbidity and mortality for patients, with intestinal transplantation being a potentially life-saving intervention, predominantly for patients with end-stage IFALD The development of specialist multidisciplinary nutrition support teams and 'intestinal rehabilitation programmes' has seen outcomes improve consistently primarily through sepsis and IFALD prevention and reconstructive surgery (although the evidence base for individual interventions remain poor). Attempts to characterise paediatric IF epidemiologically have been hampered by poor ascertainment due to the disperate spread of patients across services (neonatal, surgical, medical) and their relative small numbers across local regions. The high costs of prolonged PN and HPN treatment with, now expected improved outcomes, means that accurate national epidemiology on type II IF and HPN are required for service planning and benchmarking standards of care. We outline the planned changes to the british intestinal failure registry to accurately describe the demographics, treatments and outcomes for children who require prolonged PN.

SESSION VI

INVITED SPEAKER ABSTRACTS AND SELECTED ORAL PRESENTATIONS

Uncertainties in the management of Hirschsprung's Disease

Mr Ian Sugarman
Consultant Paediatric Surgeon, Leeds General Infirmary, Leeds

This short presentation will look at a few topics that still cause the Paediatric Surgeon difficulties in the management of patients with Hirschsprung's disease. These will include which operation to perform, what do we understand by the 'transition zone, and why results may differ in similar groups of children.

The importance of follow-up will be discussed and examples will be given to help understand the problems encountered.

One hopes, by the end of the presentation, the audience will have gained a little understanding into how and why we treat these children as we do.

Surgical Management of Gastro oesophageal reflux disease

Mr Matthew Jones, Consultant Paediatric Surgeon, Alder Hey Children's Hospital, Eaton Road, Liverpool L12 2AP

THE SURGICAL TREATMENT OF GASTRO-OESOPHAGEAL REFLUX

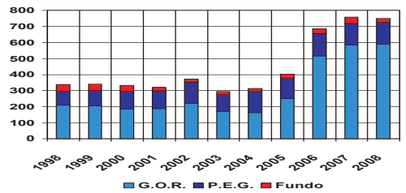
CATCHMENT AREA

MULTIDISCIPLINARY CLINIC

- Key role of multidisciplinary teamwork.

MOST "REFLUXERS" DO NOT NEED ANTI-REFLUX SURGERY

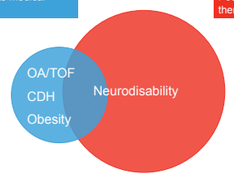
Patients treated at AHCH from 1998-2008



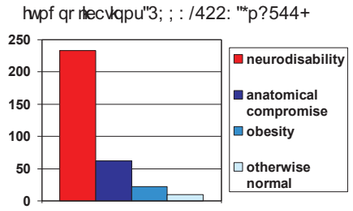
SPECTRUM OF "REFLUX"

Anatomical compromise:
Investigations positive
Good response to medical therapy

Functional disorder:
Investigations often negative
Poor response to medical therapy



WHICH PATIENTS TEND TO NEED ANTI-REFLUX OPERATIONS?



BEWARE THE "NORMAL" CHILD WITH APPARENTLY INTRACTIBLE "REFLUX"

- Functional disorder.
- Intracranial pathology.
- Missed syndromes.
- N.A.I. / Fictitious illness / Non-compliance.

The clue is the normality of investigations and the failure of response to medical treatment.

THE ROLE OF INVESTIGATIONS

- Contrast studies (delineate anatomy; vital for surgery).
- OGD and biopsy (gold-standard for diagnosing and monitoring pathological reflux).
- 24 hr pH monitoring (good for linking episodes of reflux to unexplained symptoms).
- Isotope milk scan (hmm…).

ROLE OF SURGERY

- Benefits include resolution of:
  - Poor nutrition;
  - Oesophageal pathology;
  - Recurrent aspiration;
  - Unacceptable symptoms.

BUT…

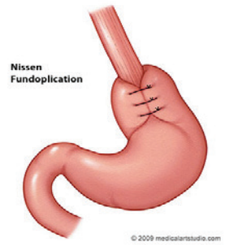
- Risks include:
  - Complications of surgery;
  - Failure to resolve symptoms;
  - Dysphagia / gas bloating;
  - Further debility or death.

CHOICE OF OPERATIONS

- Simple fundoplication (+/- laparoscopic).
- Fundoplication with vagotomy & drainage.
- Oesophago-gastric dissociation.
- Roux-Y feeding-jejunostomy.

SIMPLE FUNDOPLICATION

SIMPLE FUNDOPLICATION



SIMPLE FUNDOPLICATION

Traditional surgical treatment for G.O.R.
Treatment of choice for simple reflux.

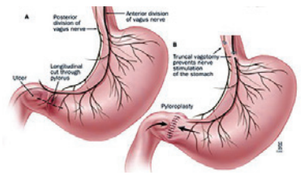
Difficult to assess efficacy because of:
- Lack of symptomatic follow up.
- Widely variable patient selection.
- Commercial bias?

But:

Studies suggest failure rates of 10-50% in neurologically impaired children.

FUNDOPLICATION, VAGOTOMY AND PYLOROPLASTY

FUNDOPLICATION, VAGOTOMY AND PYLOROPLASTY



FUNDOPLICATION, VAGOTOMY AND PYLOROPLASTY

- Increasingly the operation of choice for the neurodisabled child.
- Treats reflux but also denervates stomach and aids gastric emptying.
- 36 cases in last five years (now 77).

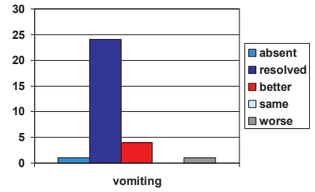
Mean age 2 years (range 2 months – 17 years)
Mean weight 11.4 kg (range 2.3 kg – 52.6 kg)

FUNDO + V&P

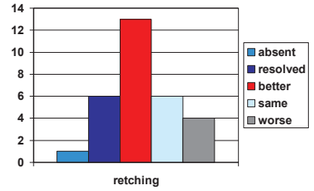
Complications:

Death	1 (Respiratory arrest; DNR status)
Line infection	3 (8%)
Bleeding	0
Leakage	0
Splenic injury	0
Tight wrap	0
Oesophageal injury	0
Respiratory problems	12 (33%)
Minor wound infection	4 (11%)
Wrap herniation	1 (3%)

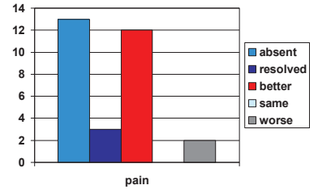
RESULTS OF FUNDO + V&P



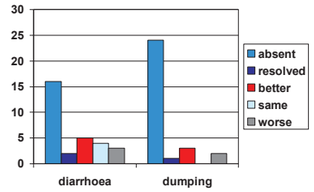
RESULTS OF FUNDO + V&P



RESULTS OF FUNDO + V&P



RESULTS OF FUNDO + V&P



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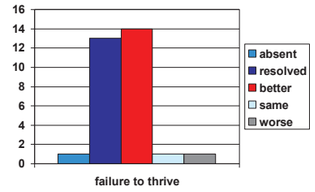

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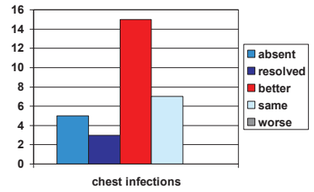
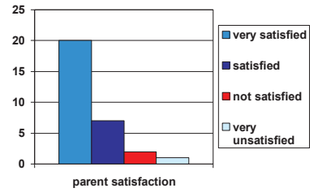
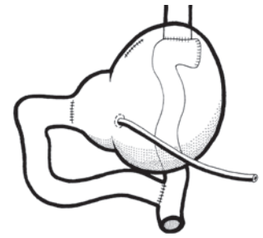


RESULTS OF FUNDO + V&P



OESOPHAGO-GASTRIC DISSOCIATION (OGD)

OESOPHAGO-GASTRIC DISSOCIATION (OGD)



OESOPHAGO-GASTRIC DISSOCIATION (OGD)

- Severe neurodisability.
- Severe hypertonia.
- Severe pain on feeding.

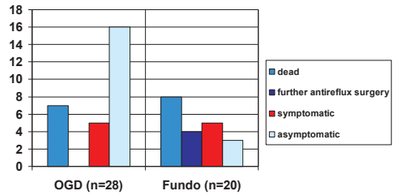
But:

- Beware the “gas-bloaters”.

OGD vs SIMPLE FUNDOPLICATION

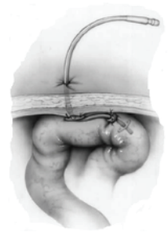
	OGD group (28 patients (now 35))	Fundoplication group (20 patients)
Operating time	*238 minutes (120-470 minutes)*	*150 minutes (60 -240 minutes)*
Start of feeds	*5.0 days (4-7 days)*	*2.7 days (1 – 4 days)*
Full feeds	*8.4 days (6 - 21 days)*	*9.1 days (4 – 49 days)*
Intensive care stay	*1.6 days (0 - 9 days)*	*4.1 days (1 – 18 days)*

OGD vs SIMPLE FUNDOPLICATION OUTCOME



ROUX-Y JEJUNOSTOMY

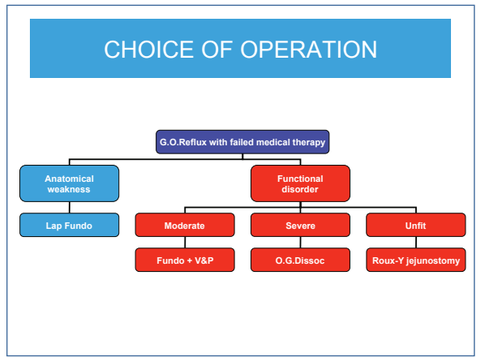
FEEDING JEJUNOSTOMY



ROUX-Y JEJUNOSTOMY

- 35 patients (mostly unfit or unsuitable for fundoplication), of whom 32 established full enteral feeds. BUT…

- 8 required re-operation (6 for volvulus and 2 for adhesions).
- 4 tube dislodgement.
- 5 major stoma leakage.



Prevention of necrotising enterocolitis.

Dr John Puntis, Consultant Paediatric Gastroenterologist, Leeds General Infirmary, Leeds

Necrotising enterocolitis (NEC) is the most common severe gastrointestinal disorder on the neonatal unit. It can progress rapidly in some cases, from mild abdominal distension to septic shock, necrosis of the entire intestine, and death. Mortality ranges from 20-50% and morbidity includes strictures, adhesions and short bowel syndrome.

The ORACLE Children Study Cohort follow up has shown that survivors are more likely to have functional disorders and continuing bowel dysfunction in middle childhood. The primary risk factor for NEC is prematurity, with incidence varying inversely with gestational age; hypoxia, infection and feeding are thought to play important roles.

Every year worldwide, 15 million babies are born preterm, of whom 1.1 million die. Prevention of prematurity has proved difficult, and for two decades there have been increasing rates in almost all countries with reliable data. The absolute numbers of preterm infants at risk for the illness have increased with advances in neonatal intensive care.

Established risk reduction strategies include use of antenatal steroids, standardised feeding protocols and early preferential feeding with breast milk, while probiotics represent the most promising newer approach to prevention. Delayed enteral feeding, frequent use of antibiotic therapy and altered acquisition of normal gut microflora are believed to contribute to the risk of NEC and provide the rationale for probiotic ("live microorganisms that when administered in adequate amount confer a health benefit on the host") supplementation.

While recent meta-analyses suggest a significant benefit in reducing death and disease, the heterogeneity of studies (different probiotics, mixtures and doses among other factors) and methodological critiques have divided neonatologists between those convinced that now is the time for a change in practice and those who urge caution and the need for more data, referencing past negative experiences with high oxygen intake causing blindness and dexamethasone for lung disease inducing cerebral palsy.

Diagnostic and therapeutic utility of double-balloon enteroscopy

[1]Dr Arun Urs, MBBS, MRCPCH; [2]Dr Massimo Martinelli, MD; [1]Dr Prithviraj Rao, MBBS, MRCPCH, Consultant Paediatric Gastroenterologist;, [1]Dr Mike Thomson, DCH, FRCP, FRCPCH, MD, Consultant Paediatric Gastroenterologist:
[1]Centre for Paediatric Gastroenterology and International Academy of Paediatric Endoscopy Training, Sheffield Children's NHS Foundation Trust, Sheffield, UK S10 2TH; [2]Department of Paediatrics, University of Naples, "Federico II", Naples, Italy

Introduction
The diagnostic and therapeutic benefits of double-balloon enteroscopy (DBE) have now been largely documented in the adult population with as yet little published in the paediatric population. This study aimed to evaluate the diagnostic and therapeutic utility of DBE in the setting of a paediatric tertiary-referral centre.

Methods
Prospective assessment of consecutive children younger than 18 years undergoing DBE for a variety of suspected small bowel disorders from Jan 2008 to August 2012 in a single paediatric tertiary referral centre was carried out. All the children had undergone prior upper GI endoscopy, ileo-colonoscopy, and in the majority wireless capsule endoscopy. The clinical and histological findings/treatment by DBE was followed by active treatment or followed up without any treatment and are presented.

Results
113 DBE were performed in 58 children (M=36, F=22, median age 12.7 years, range 1-18 years). 61 (54%) procedures were performed via the trans-oral approach with a median time of 90 minutes (range 26-245), and 47 (42%) procedures via the trans-anal approach (4 via ileostomy) with a median time of 45 mins (range 15-100), and 5 (4.5%) were with laparoscopic assistance. The median estimated insertion length of the small bowel distal to the pylorus (DTTP) was 230 cm (range 80-450), and proximal to the ileo-caecal valve (PICV) was 80 cm (range 5-275). The overall diagnostic yield for relevant lesions in the small bowel was 67% (n=39). The most common findings were polyps (n=19, 32%), mucosal ulcers and erosion (n=8, 14%), sub mucosal elevations with white nodules (n=4, 7%), and angioma/angiodysplasia (n=2, 4%). The indications included hereditary polyposis syndromes (Peutz-Jeghers' syndrome (PJS), blue rubber bleb naevus syndrome (BRBN), Cowden's syndrome, and familial adenomatous polyposis (FAP) (n=21, 36%), obscure GI bleeding (n=16, 28%), Crohn's disease (CD) (n=8, 14%), persistent intractable diarrhoea (PID) (n=5, 9%), recurrent abdominal pain (RAP) (n=4, 7%), and others (n=5, 9%). In children requiring endo-therapeutic treatment this was performed with a successful therapeutic yield of 48% (n=28). The mean duration of therapeutic small bowel enteroscopy was 85 min (range 20-245), compared to 69 min (range 20-135) for diagnostic procedures. Therapeutic procedures included polypectomy (n = 19), argon plasma coagulation of vascular lesions (n = 8), stricture dilation (n = 1), endoclip haemostasis (n=1), and small bowel variceal banding (n = 1). The overall usefulness of DBE in contribution to treatment including endoscopic, medical, and surgical was 72.5% (n=42). Three complications (5%) were noted, one perforation 2cm proximal to the stoma in a patient who had undergone a small bowel transplant, pelvic abscess from an infected pelvic wound secondary to laparoscopic assisted DBE and hypotension secondary to sepsis.

Conclusion
As previously reported DBE is not only safe and feasible but has a high diagnostic and therapeutic yield. The diagnostic yield of DBE was comparable to WCE with the added advantage of therapeutic possibility, histological diagnostic yield, and minimal complications and we believe that this technique should serve as a valuable addition to existing endoscopic techniques. The results of DBE had a substantial impact on subsequent management decisions. DBE offers a less invasive approach, which may reduce the need for surgery in children, particularly with polyposis syndromes. DBE should be considered as an alternative diagnostic and therapeutic option in the small bowel in children.

Raised D lactate – A surrogate marker of small bowel bacterial overgrowth

Dr Hemant Bhavsar, Paediatric Gastroenterology registrar; Dr Theodoric Wong, Consultant paediatric gastroenterologist; Dr Sue Protheroe, Consultant paediatric gastroenterologist
Birmingham Children's Hospital

Introduction:
Diagnosis of small bowel bacterial overgrowth (SBBO) may be difficult and delayed diagnosis can have significant morbidities. Excess luminal bacteria, such as lactobacilli, can produce D-lactate (DL) which can be detected in serum. We hypothesised that DL measurements is a clinically useful non invasive marker of SBBO and helpful in monitoring response to treatment. We present the first large scale cohort study of raised DL in infants and children with intestinal failure (on parenteral nutrition (PN) > 28 days) in a large paediatric tertiary referral centre.

Aim:
1. To identify Intestinal Failure (IF) patient characteristics as risk factors for raised DL
2. To review use of DL in monitoring response to treatment.

Methods:
A retrospective review of patient demographics, symptom(s) at the time of first DL measurement, feeding regimen, treatment, biochemical parameters and radiology from 01/01/2009 to 31/12/2011 was performed. Children rehabilitating from IF from 1-18 years of age with new onset diarrhoea, vomiting, abdominal pain/bloating and neurological symptoms were included in the study. Length of remaining small bowel was converted to percentage length appropriate for age using a published formula. DL measurement of >20 µmol/L was defined as a raised DL. A recurrence is defined as raised DL at least 4 weeks from the last measurement with standard treatment (rehydration, withholding or alteration of feeds and/ or bicarbonate supplementation and antibiotics).

Results:

Characteristics		Raised DL (N=25)	Not raised DL (N=24)	p value
Mean age (years) at first DL estimation (range)		4.65 (0.27-11.86)	4.63 (0.10-13.07)	NS
Bowel resection		20	15	NS
ICV absent (in bowel resection group)		11	6	NS
% small bowel length corrected for age (%SBL)	Mean	34.66 %	76.45%	0.020
	median	29.6%	91.4%	
PN duration (months) and range		7.97 (0-36)	5.64 (0-36)	NS
Oral feed ad libitum (+/- Enteral Nutrition)		17	18	NS
More than 1 predominant symptom		6	1	NS

NS= not significant

49 out of 209 patients with IF met the inclusion criteria. 28 pts were male. The group mean gestational age was 36 weeks with 49% being preterm (<37 weeks gestation). Aetiologies for IF were grouped into bowel resection due to congenital malformation (17/49), necrotising enterocolitis (15/49), dysmotility (3/49) and enteropathy (10/49). The commonest symptom was chronic diarrhoea (39/49). Other symptoms like vomiting, abdominal pain/bloating and neurological symptoms were seen in both groups and did not satisfy statistical significance.

Patients with less than 35% of SBL corrected for age, had 80% sensitivity for developing raised DL. No other factors such as duration of PN, absence of ileo-caecal valve (ICV), anatomical abnormalities on barium study, and use of Proton pump inhibitors were found to have a statistical significance as risk factors for raised DL. Relationship to feed could not be analysed due to lack of accurate information on carbohydrate content in the patients' diet. 50% (12/24) of the raised DL group satisfied our definition of a recurrence. DL was monitored in 12/25 patients during standard treatment and all had fall in DL with reduction in symptoms.

Conclusion:
Patient with <35% of bowel length estimated for age has an increased risk of SBBO in patients with symptoms as evidenced by raised DL. Measurement of DL is a useful non invasive method to detect SBBO. Its estimation should become routine in at risk patients to provide prompt treatment.

Efficacy and Safety of Low Dose and High Dose Intravenous Methylprednisolone Treatment in Acute Ulcerative Colitis

R.Vora[1,2], H. Finnamore[2*], K. Crook[2], AM. Dalzell[2], B. Krishnamurthy[2], V. Krishnappa[2], E. Whittle[2], T. Irvine[2], MKH. Auth[2]. *(joint first authors, equal contribution)*
[1]Imperial College,London, [2]Alder Hey Children's NHS Foundation Trust, Liverpool.

Acute severe colitis is a serious complication of ulcerative colitis (UC) in children with a reported colectomy rate of 61% in the pre-biologic era. According ECCCO and ESPGHAN guideline, intravenous methylprednisolone is the recommended first line treatment, however associated with a pooled steroid refractory rate of 35% in protocols using 1-1.5 mg/kg/day. While the role of calcineurin inhibitors and infliximab for salvage therapy is established, controversial reports about high dose pulse steroids suggested the need for dose-finding studies for intravenous methylprednisolone (ivMP). We compared the results of low dose (LDMP) or high dose (HDMP) intravenous methylprednisolone from two dosage protocols non-randomised for the treatment of acute severe and moderate-severe ulcerative colitis with regard to remission, morbidity, and colectomy rate.

Subjects and Methods:

Retrospective case note review on children (n=34) admitted for moderate- or acute-severe UC requiring ivMP over an 8 year period in our tertiary children's hospital. High dose MP was defined as >15mg/kg/day and low dose < or = 2mg/kg/day. Disease activity was correlated using actual PUCAI score since the score was published, and retrospective PUCAI score calculations for patients admitted earlier. Two-tailed student's t-test was used to compare results. Patient characteristics were comparable.

Children with acute UC	High Dose MP (HDMP)	Low dose MP (LDMP)
n = 34 patients	20	14
Median age at diagnosis in years (range)	9.3 (2.7-15.3)	11.8 (4-15.3)
Median age at start of ivMP (range)	9.3(2.7-15.3)	12.2(4-15.8)
Pancolitis	15 (75%)	11 (78%)
Left sided Colitis	5 (25%)	3 (22%)
5-ASA use pre admission	6(30% of HDMP)	4(28.6%of LDMP)
Azathioprine use 3 months before	3 (15%)	2 (14%)
Steroid use 1 month pre admission	13 (65%)	8 (57%) (p<0.05)

Results:

PUCAI during intravenous treatment of acute colitis

Median PUCAI score	High dose MP	Low dose MP	p-value
Day 1 of iv MP	60	55	p=0.58
Day 3 of iv MP	25	20	p=0.85
Day of Discharge	6	7	p=0.85
Median length of stay in hospital	7 days	6 days	p=0.87

Salvage Therapy- within 1 year of discharge after acute ulcerative colitis

Salvage therapy	High dose MP (n=20)	Low dose MP (n=14)
Infliximab at Discharge	n=2 (10%)	n=2 (14%)
Ciclosporin within 1month	n=0	n=1 (7%)
Colectomy within 1 year	n=0	n=4 (28.5%)

Notably, the four colectomies occurred in children treated with low dose iv MP (14% vs 0%) (p<0.05). Overall 85.2% of patients were in remission upon ward discharge, with higher requirement for salvage therapy for LDMP (21% vs 10%, not significant). Side effects of iv MP were only minor, temporary and comparable between both groups and comprised raised glucose levels (n=2) or arterial hypertension (n=3).

Summary and Conclusion:

Our results demonstrate a very low overall colectomy rate (11.7%), and high remission rate (85.2%) for children with acute or moderate severe UC. Notably, patients with HDMP required no colectomy, and their requirement for salvage therapy was not more frequent than in those with LDMP. With a median stay of 6.5 days, patients treated with iv MP had no persistent or severe side effects from this medication, suggesting that HDMP treatment is effective and safe. Further studies are required to define the role of high dose intravenous steroids in the management of acute severe colitis.

Declaration of interest: MA and KC received travel grants from Dr Falk Pharma and MSD; VK from Dr Falk Pharma.

Financial impact of Taurolock® during long term parenteral nutrition

Dr Veena Zamvar[1], Deirdre Kriel[2], Dr Jonathan Sandoe[3], Dr John W L Puntis[1]
Departments of Paediatric Gastroenterology[1], Pharmacy[2] and Microbiology[3], The Children's Centre, The General Infirmary at Leeds, Leeds LS2 9NS, UK

Background and aims:
Evidence suggests that use of Taurolidine-citrate lock solution (Taurolock®) has a major impact on central venous catheter (CVC) related bood stream infection (CRBSI) during long term parenteral nutrition (PN); Taurolock® may also prevent catheter occlusion. The aim of this study was to evaluate the cost implications of using Taurolock® in a group of PN-dependant patients .

Patients and Methods:
Out of 16 home PN patients, five received daily Taurolock® because of previous recurrent CVC sepsis, and one because of frequent CVC occlusion necessitating catheter replacements. A retrospective case note review identified the number of CRBSI/1000 catheter days and the number of CVC changes. Based on projections from pre-Taurolock® experience, we estimated the theoretical cost savings from reduced hospital admissions and antibiotic usage observed in our patients over one year post-Taurolock®.

Results:
13,028 catheter days (8371 pre- and 4657 post- Taurolock®) were evaluated. Pre-Taurolock®, CRBSI/1000 catheter days for patients 1 - 6 were 0, 0.87, 4.4, 5.26, 8.7, and 10.9; post-Taurolock®, there were no episodes of CRBSI. In 2 patients with recurrent CVC occlusion, patency was maintained during Taurolock® use. The total number of antibiotic days one year pre-Taurolock® was 387 and one year after the introduction of Taurolock® had reduced to 79. The number of inpatient hospital days pre-Taurolock® was 816 (approximate cost £489108) and one year post-Taurolock® 136 (approximate cost £68000). Expenditure on antibiotic treatment for CRBSI pre-Taurolock® was £14088 compared with £2146 post-Taurolock®. The total expenditure (antibiotics, hospital in patient stay and Taurolock® was calculated as £503196 pre- and £94236 post-Taurolock®, representing a total cost saving of £408960. No adverse effects related to use of Taurolock® were seen.

Conclusions:
The use of Taurolock® during cyclical PN was associated with a dramatic reduction in CVC sepsis rates in a small group of children with long term PN dependency. Taurolock® also appeared to lower the rate of CVC occlusion. The reduction in sepsis has large cost saving implications for the NHS, and if used widely, Taurolock® might reduce intestinal failure associated liver, venous thrombosis and need for intestinal transplantation.

Paediatric-onset Crohn's patients relapse earlier than adult-onset after right hemicolectomy

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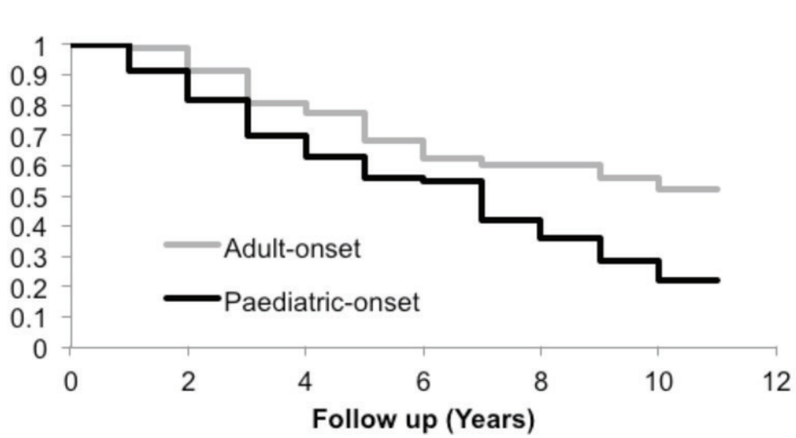
Backround and Aim
Crohns disease (CD) is a relapsing remitting disease with no known cure. It presents in childhood in up to 25% of cases. Right hemicolectomy (RH) is recommended for localised disease refractory to medical therapy, and when performed in childhood can promote growth. The outcomes from separate studies of paediatric-onset CD (P-CD) and adult-onset (A-CD) after RH suggest A-CD has a lower relapse rate. Our IBD database contains longitudinal data of both children and adults, and our aim was to do a direct comparison.

Subjects and Methods
All CD patients who had RH were identified on the IBD database of a single tertiary referral centre for children and adults. This was cross-referenced with theatre data for the last 10 years. Case notes were reviewed for date of diagnosis, time to RH, time to relapse, or duration of follow up if no relapse had occurred. Relapse was defined as recurrent disease identified at endoscopy or by radiological imaging, and/or clinical relapse defined by an escalation in immunosuppression or further Crohns-related surgery.

Results
Sixty-seven P-CD patients underwent RH from 1982-2011, with a median time from diagnosis to RH of 3 years (mean 3.5, range 0-13). There were 116 (A-CD) patients with RH from 1969-2011, with a median time from diagnosis of 1 year (mean 4.7, range 0-26 years). Forty-one (61%) of P-CD patients relapsed, with median time to relapse or (relapse-free) follow-up of 4 years. This compared to 81 (71%) A-CD patients, after median 6 years. The Kaplan-Meier curve shows that P-CD patients relapsed earlier than A-CD ($p<0.001$).

Conclusion
This is a large retrospective longitudinal study of relapse after RH in CD. Relapse rates were comparable to previous studies, and in this direct comparison P-CD relapse earlier than A-CD. This may reflect a more severe phenotype of paediatric-onset Crohn's Disease.



The changing epidemiology of coeliac disease in south Wales – a 28 year perspective

Whyte L A, Jenkins H R
Department of Paediatric Gastroenterology, Children's Hospital for Wales, Cardiff, CF14 4WX.

Introduction
The diagnosis of coeliac disease (CD) has increased in frequency, particularly since the accuracy of serological antibody testing has improved. Previous studies from our region have shown an increasing incidence of CD from 1983 to 2004, with a change in clinical presentation and decrease in specific gastro-intestinal symptoms, as well as an increase in age at diagnosis.

Aims/Methods
We reviewed all patients with CD presenting to the Regional Centre between 2005-11 and compared the incidence, age and documented mode at presentation with previous published data from the same area between 1983-2004.

Results
163 cases (23 per year) of CD were diagnosed between 2005 to 2011, with a median age at diagnosis of 14 years (range 0.8 to 16 years) compared with 50 cases (8 per year) a median age of 8 years between 1999 to 2004. 41% presented with classical symptoms, 23% with non-classical symptoms and 36% asymptomatic and diagnosed after serological screening of high risk group. Compared with the most recent previous study from the same population, the percentage of patients presenting with gastro-intestinal symptoms remained similar (42% vs 41%) but patients diagnosed after targeted screening had increased from 26% to 36%.

Conclusion
Frequency of diagnosis of CD in this stable and well defined population has risen dramatically in the last 7 years, although it is likely that many patients still remain undiagnosed in childhood. The median age at diagnosis has increased and over 50% of patients present with few or no symptoms.

SESSION VII
INVITED SPEAKER ABSTRACTS

Ms Jo Price

Monoclonal antibody in inflammatory bowel disease

Dr Tony Akobeng, Consultant Paediatric Gastroenterologist, Royal Manchester Children's Hospital, Manchester, UK

Inflammatory bowel disease (IBD) comprises of the chronic relapsing inflammatory disorders Crohn's disease (CD) and ulcerative colitis (UC). There is no medical cure for IBD and traditionally, the goals of treatment are mainly the induction of clinical remission and the maintenance of clinical remission. The conventional treatments that allowed the achievement of these goals over decades were the corticosteroids, 5-ASAs, and immunosuppressive agents such as azathioprine, -6-mercaptopurine, and methotrexate. However, despite the use of these treatments, the natural history of IBD did not really change. For instance, the long-term evolution of CD remained structural damage and the rate of surgery in patients with CD did not change over time. In the past 15 years or so, increasing understanding of how the inflammatory cascade works in the intestine has allowed the development of monoclonal antibodies which are targeted at key immune and inflammatory mediators. These drugs have had a considerable impact on the management of IBD and it is believed that they may offer the potential to modify the natural history of the disease. So instead of just aiming for the induction and maintenance of remission, perhaps our new treatment goals should include a sustained corticosteroid-free remission, mucosal healing, prevention of complications, reduction in surgeries, and reduction in hospitalisations. A number of monoclonal antibodies have been investigated or under investigation for IBD but the ones that have so far been proven to be effective and approved for the treatment of IBD are three TNF-alpha antagonists (infliximab, adalimumab, and certolizumab), and an alpha 4-integrin inhibitor, natalizumab. Currently, infliximab and adalimumab are approved for IBD in both Europe and the US whilst certolizumab and natalizumab are approved for IBD in the US but not in Europe. Despite the fact that the TNF-alpha antagonists have been used as treatments of IBD for about 14 years, there remain a number of important unanswered questions on their use. In this presentation, I will discuss the monoclonal antibodies that are used for treating IBD and discuss a number of day-to-day practical issues that face gastroenterologists who use these agents. Practical issues / questions that will be discussed include:

- which IBD patients should receive TNF-alpha antagonists?
- If we decide to use a TNF-alpha antagonist, which one should we start with?
- monotherapy (i.e. TNF-alpha antagonists alone) versus combination therapy?
- episodic treatments versus regular maintenance treatments
- what should we do when a patient loses his/her response to a TNF-alpha antagonist?
- how long should we continue TNF-alpha antagonist treatment for?
- safety concerns.

Leukapheresis

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Treatment of pediatric IBD patients is often demanding as the diseases can be chronic, active and almost half of the patients are corticosteroid dependent. GMA treatment has been used in Europe for some years and it has been shown to be effective in 70% of moderately severe cases of IBD. In GMA, granulocytes and macrophages are removed from circulation using Adacolumn and thus also the levels of inflammatory cytokines are lowered. There is so far limited experience in pediatric patients and mostly limited to UC.

European multicenter study (ADAPT) was conducted recently to evaluate the efficacy and safety of GMA treatment in moderately-severe UC children. Apheresis treatment was given once a week, 5-8 times, and the patients were followed for 15 months altogether.

The efficacy of treatment was evaluated by using PUCAI scores and reduction in corticosteroid dosage. By week 12 of main phase 76% of patients responded to the treatment. At 15 months follow-up PUCAI score was reduced by the mean of 25.3 points compared to baseline. Corticosteroid dosage was weaned from the mean of 14,6 mg to zero. Sixty percent got re-treatment during the follow-up after the main phase.

As also ADAPT study showed, relapsing is a common feature of UC. Therefore we have used GMA in Finland as a maintenance therapy. After the initial treatment period, 11 UC patients (mainly corticosteroid-dependent) have got GMA treatment once a month (3-6 weeks). 9/11 (81 %) of patients were in remission at the end of the initial treatment and 50 % remained in remission through the 18 month follow-up thereafter. The mean PUCAI scores remained stable after the initial phase. GMA seems to be effective also as a maintenance therapy in UC children with chronic, active disease. Side effects have been mild; mostly headache and tiredness. Therefore GMA could be considered as one treatment option in pediatric UC patients.

Speaker sponsored by travel grant from Otsuka

Surgery for complex inflammatory bowel disease

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The increasing incidence and severity of inflammatory bowel disease (IBD), among British children has led to more children undergoing surgery for their condition.

The presentation, surgery, complications and outcomes of 100 consecutive intestinal resections for Crohn's disease, and 56 children receiving an ileo-anal pouch procedure for colitis by one surgeon are discussed.

Particular emphasis is placed on the development of techniques specific for children to allow any age of child to receive an ileal pouch. The use of laparoscopic techniques for both Crohn's disease and colitis is discussed.

FRIDAY POSTERS

POSTERS OF DISTINCTION

Developing Azathiaprine therapy monitoring policy

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The immunosuppressant Azathiaprine is in common use in the management of gastrointestinal inflammatory conditions. Its potential complications of heptotoxicity and myelotoxicity mandate a program of blood test surveillance. It has been anticipated that dosage and thiopurine methyl transferase activity (TPMT) influence the likelihood of toxicity.
Since 2009 our policy has been that patients commence Azathiaprine therapy on a dose of 1mg/kg/day if they are TPMT deficient or TPMT status unknown, and 2mg/kg/day otherwise. This is followed by blood count and liver function testing weekly for month 1, fortnightly for month 2 and then monthly. A similar monitoring program follows dosage increases.

Aim
1) To Audit adherence to the monitoring regime in the initial four months of therapy.
2) To examine the relationships between monitored indices, TPMT, dosage and therapy duration which underlie the rationale for this monitoring regime
3) To report outcome of abnormalities detected by the monitoring regime.

Subjects
All patients attending paediatric gastroenterology clinic who commenced or increased their azathioprine therapy between June 2009 and 2011.

Methods
Audit: For each of 8 monitoring samples patients were considered compliant if a sample was taken within +/- 3 days of the schedule date and outcomes recorded for each monitoring frequency.
Rationale testing: The indices Lymphocyte and Neutrophil counts and ALT were recorded for each patient sample irrespective of it qualifying as a schedule compliant sample. Rates of change of each index between consecutive samples was calculated, so that variations in these which may justify more frequent early tests, could be assessed through the schedule. Mean index values and rates of change were compared for each monitoring frequency period by ANOVA using schedule compliant samples.
To assess the influence of TPMT, dosage and time since dosage increase, as continuous variables, these data were entered as independent variables into 6 multiple regression models with each index and each index rate of change as the dependent variable using all samples. The probability that the regression coefficient for each variable was different from zero was calculated. Non-zero values indicating some influence on the index concerned.

Results
Of 59 Dosage change events, complete schedule compliance was achieved for 29% of Weekly and fortnightly sampling periods and 39% for the monthly period.
ANOVA on schedule compliant samples showed no significant difference between the sampling periods for rates of change of ALT , lymphocyte count and neutrophil count.
In the 6 linear regression models on 278 blood test intervals, no coefficient for TPMT, dosage or dosage duration reached significance in its ability to predict rates of change of the three indices, or the indices themselves. No instance of myelotoxicity was detected. Six patients had hepatotoxicity, one requiring cessation of therapy.

Summary and Conclusion
These data highlight the challenges of developing both a child friendly safe Azathiaprine monitoring program and a robust audit method. They provide no supporting evidence that justifies our current TPMT, dosing and monitoring policies

How the new guidelines for the diagnosis and management of Coeliac Disease in children will potentially impact on practice in the North East of England and potential cost implications.

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Introduction/Background

Guidelines for the diagnosis and management of Coeliac Disease in children were updated by the Coeliac working group of BSPGHAN in 2012. They have focused on how the diagnosis of Coeliac Disease is confirmed.

Aim
We have retrospectively reviewed the impact of these guidelines on our practice and the potential cost implications.

Subjects and methods
We reviewed the notes of all children with possible Coeliac Disease who had an upper endoscopy and small intestinal biopsy from 1st January 2011 to 31 December 2011. We specifically looked at symptoms experienced, IgA tTG level, IgA-EMA and biopsy results. HLA DQ2/DQ8 is not performed routinely.

Results
During the year 2011, 42 children with suspected Coeliac Disease had an upper GI endoscopy and small intestinal biopsy. They were divided into 4 groups

1. Symptomatic ; tTG positive > 10x upper limit normal n =13
2. Symptomatic ; tTG positive < 10x upper limit normal n =20
3. Asymptomatic ; tTG >10x upper limit normal n = 7
4. Asymptomatic ; tTG < 10x upper limit normal n= 2

All patients in group 1 & 3 had biopsies consistent with a diagnosis of Coeliac Disease. 11/20 patients in group 2 had biopsies consistent with a diagnosis of Coeliac Disease. Both patients in group 4 had normal small intestinal biopsies.

Summary and Conclusion
Using BSPGHAN guidelines 13/42 children would not have necessarily required upper GI Endoscopy and small intestinal biopsy. Not having to have an invasive procedure will benefit many of our children greatly. There are also cost implications. Tariffs currently stand at
1.Day case Upper GI Endoscopy; biopsy and histology £869
2.EMA £16.03
3.DQ2/DQ8 £97

By following the new guidelines, children with a tTG greater than 10 x the upper limit of normal, a positive EMA and HLA DQ2 or DQ8 positive, would not require upper GI Endoscopy and Biopsy saving £756 per patient.

Liver Disease in Inflammatory Bowel Disease.

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Introduction/Background:
The association between inflammatory bowel disease (IBD) and liver disease, particularly sclerosing cholangitis is well known.

Aim:
To describe the features of liver disease in patients with IBD.

Subjects and Methods:
Retrospective review of patients with IBD referred to our Paediatric Liver Centre, between September 2004 and 2009, because of abnormal liver function tests (LFTs).

Results:
43 patients were identified (31M: 12F). Median age at IBD diagnosis was 10.9 yrs (2-15); 27 had ulcerative colitis, 13 indeterminate colitis, 2 Crohn's disease and 1 indeterminate colitis and coeliac disease. LFTs were abnormal at IBD diagnosis in 17, while became abnormal 20 mths (1-81) after diagnosis in 26. Median time for referral to our centre was 9 mths (0-117) after IBD diagnosis.

At presentation blood tests were: AST 69 IU/l (range 15-1245), ALT 15 IU/l (range 13-251), GGT 206 IU/l (range 6-498) and INR 1.07 (range 0.86-2.23). Total bilirubin was >20 µmol/l in 8. IgG was increased in 21. 6 patients were positive for antinuclear antibody, 5 for anti-smooth muscle antibody and 2 for both. No one was anti LKM-1 positive. 26 were ANCA positive.

Of 41 who underwent biliary imaging (MRCP/ERCP), 31 had cholangiopathy 28 of whom had bile duct damage (BDD) on liver histology; of these 18 who also had interface hepatitis (IFH) and/or positive auto antibodies were diagnosed as autoimmune sclerosing cholangitis (ASC), whilst 10 with no IFH and/or positive autoantibodies as sclerosing cholangitis (SC). Two with cholangiopathy on MRCP and no BDD on histology, but IFH and/or positive autoantibodies were diagnosed as ASC. 1 did not undergo liver biopsy.

Of 10 with normal biliary imaging, 6 had BDD on histology (small duct disease), 3 with concomitant IFH and/or positive autoantibodies. 3 with no histological BDD but IFH on histology and positive autoantibodies were diagnosed as autoimmune hepatitis. 1 child did not have a liver biopsy.

Of 2 patients with no biliary imaging, 1 had evidence of BDD and IFH on histology and 1 did not have a liver biopsy.

At diagnosis 23 patients were treated for IBD (prednisolone +/- azathioprine; mycophenolate mofetil +/- mesalazine; sulphasalazine) with ursodeoxycholic acid (UDCA) added in 14. 6 patients with cholangiopathy on biliary imaging were not on treatment. At follow up all patients were on treatment including UDCA in 37. One patient presenting simultaneously with UC and ASC required a liver transplant 5 yrs later. 7 patients, all male, required colectomies.

Overall cholangiopathic features either on imaging and/or histology were found in 38 (88%) of patients. All patients are alive at median follow up of 4.9 yrs (2.5-7.5).

Summary and Conclusion:
The high incidence of cholangiopathy in children with IBD and abnormal LFTs in our study highlights the need for prompt referral to a specialised paediatric liver centre for diagnosis and appropriate management.

When are young adults with Inflammatory Bowel Disease (IBD) ready for transfer to adult care?

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Introduction:
Transition is the planned move of adolescents and young adults with long-term physical conditions from child-centred to adult-orientated health care. The need for a well planned transition process has become increasingly recognized and guidelines have been produced to aid health care professionals through transition. The Inflammatory Bowel Disease Transition to Adult Health Care Guidance for Health Professionals was produced in 2008 to facilitate this critical process.

Aim:
We studied transition of Inflammatory Bowel Disease (IBD) patients in a tertiary paediatric gastroenterology unit to provide a baseline on our current performance and to further understand this fascinating process.

Method:
We prospectively assessed the Paediatric Gastroenterology department transition process against the Inflammatory Bowel Disease Transition Guidance. Parents and young adults were provided with a questionnaire in the IBD transition clinic. The questionnaire addressed the stages of transition and readiness for transfer.

Results:
36 questionnaires were completed. Patients seen in clinic were 14-17 years old. The mean age at diagnosis was 12 years 3 months (Range: 4-16 years). The mean age of discussing transition with young adults was 15 years (Range: 13-16). Despite all patients meeting the adult gastroenterology consultant at least once in the paediatric transition clinic only 55% of patients and 39% of parents felt the young adults were ready for transfer. There is a significant association between young adults being responsible for taking own medications and feeling ready to be transferred to adult follow up care ($p=0.0015$), furthermore most patients who were able to discuss their symptoms and management in the absence of their parents were happy to be transferred to adult care ($p=0.04$).

Recommendation:
Well planned transition is considered a standard element of care for young adults with IBD. They need to be encouraged early to participate in their medical care. Young adults may be ready to be transferred to adult services once they are able to discuss their own symptoms and management without parental support and are responsible for taking your own medications.

FRIDAY POSTERS

A National MDT Chronic Intestinal Pseudo-Obstruction Service: A Six Month MDT Review

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Introduction/Background:
In 2011, supported by BSPGHAN, the neurogastroenterology and motility service at Great Ormond Street Hospital was designated a national centre for the diagnosis of paediatric Chronic Intestinal Pseudo-Obstruction (CIPO). The aim of the service was to ensure that all infants and young children (<5) in the UK with a potential diagnosis of CIPO received expert multidisciplinary diagnosis and management guidance in a coordinated and timely manner. Given a paucity of CIPO data (incidence, diagnostic protocols, nutrition, psychological burden) available pre-launch, the service model relied upon a number of 'educated predicitions' of activity. Actual data is now available from the service.

Aim:
The aim of this paper is to report, 6 months into its formal structured phase, the progress, achievements of the national CIPO service, as well as the comparisons of actual versus predicted activity and the challenges that remain to ensure an optimal CIPO service.

Subjects and methods:
Each of the 4 key MDT companions reviewed data from the CIPO diagnostic service over the last 6 months including specialist-specific interventions that were undertaken on patients referred to the service. This was then compiled and compared to that predicted in the original submission to AGNSS (formerly NCG). 4 key areas reviewed were:
1) Gastroenterologist: reviewed 5 key aspects namely a) Diagnosis; b) Primary motor disturbance; c) Extent of GI involvement. d) Extra-intestinal involvement and e) Aetiology.
2) Nurse: Aim that patients should have 'streamlined & timely admission' therefore admitted for 2 weeks with all investigations completed. Patient experience monitored by complaints.
3) Dietician: A standardised nutritional assessment document was completed for all patients at the point of referral and again post intervention eg surgery, referral for home PN or within six months if no intervention. The information was entered into a database.
4) Psychologist: All patients planned to receive a standardised (age appropriate) screening assessment to determine baseline data as well as identify those in need of further intervention.
Results: 15 patients (mean 4.6yrs: 10 aged <5yrs) were admitted during the 6-month study period. 7 remain on the waiting list for admission.
Gastroenterologists: 11 out 15 were diagnosed with CIPO (7 aged <5yrs). Of the remaining 4, one had Intestinal lymphangiectasia, one possible long segment Hirschsprung disease and two still awaiting confirmation. Of the CIPO cases all appeared primary and showed neuropathic or mixed neuropathy/ myopathy motor patterns, 8 had generalised and 2 regional GI involvement. Three CIPO cases had urinary tract and 2 neurodevelopmental problems.
Nurse: 7 patients had one outstanding procedure at the end of the planned 2 week admission. One patient complaint has been received, and their comments appropriately actioned upon.
Dietician: Standardised nutritional assessments have been carried out for all 15 patients at the point of referral and the first follow up assessments are due in November.
Psychologist: Of the 15 cases, 5 had psychologist input, of which 3 cases referring centres had raised concerns of FII before admission for investigation.

Summary and Conclusion:
The predicted caseload was 30 referrals per annum of <5yr olds, of which 12-15 cases would be diagnosed with CIPO. In 6 months therefore, although the service activity mirrors that predicted for the <5yrs, additional activity was performed in investigating those > 5yrs. This may be accounted for in a 'backlog' of those awaiting early and accurate diagnosis. Indications are that there is a national pressure to admit a greater number of cases every year in order to capture these older patients. 11 confirmed cases of CIPO have been made. All of these have had further definition of disease phenotype, which has instructed further management and prognosis. All patients have required robust and reactive dietary protocols. The requirement for psychology input accounts for greater than the predicted 30%, underlining the significant often 'hidden' burden of disease in CIPO. The service is exploring improvements in the tools and protocols available for psychological assessment. The service has made positive steps towards resolving a historical precedent of unclear diagnosis and management for CIPO as well as practical issues of admission delays and poor communication. Improvements are still needed but will require collaboration from across secondary and tertiary paediatric UK centres.

Adalimumab in children and adolescents with moderate/ severe ulcerative colitis: single tertiary UK center outcome

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Background:
Adalimumab has been approved for the treatment of ulcerative colitis (UC) in adult patients. Data on its role in paediatric UC are lacking.

Aim:
The aim of our study was to evaluate the efficacy of Adalimumab treatment in children and adolescents with moderate-to-severe ulcerative colitis who failed other treatments.

Subjects and Methods:
All patients with UC who received Adalimumab between April 2008 and October 2012 in our hospital were identified. Clinical response and long term outcomes were assessed.

Results:
Ten patients (7 females) with median age 14 years (6.8y-16.6y) at the time of first Adalimumab injection were included in our study, with median follow up 1.9 years (1y-3y). All patients had failed Infliximab after 6.5 months median duration of treatment (2-24 months). Three patients (3/10, 30%) showed sustained clinical response to Adalimumab with histological evidence of mucosal healing (follow up 16, 22 and 24months respectively). All three received concurrent treatment with Azathioprine or Methotrexate and one of them was successfully weaned off Adalimumab after 21 months, without relapse to date. For the remaining 7 patients, medical treatment was escalated with other immunosuppressive agents, with 4 of them ending up with colectomy (4/10, 40%).

Summary and Conclusion:
Overall, Adalimumab was efficacious in 30% of children and adolescents with moderate/ severe ulcerative colitis unresponsive to Infliximab and should therefore be considered as treatment option in refractory disease, prior to surgery.

Care of children under the age of 16 years in adult IBD services

Emma Fernandez, Project Manager, Royal college of Physicians, UK IBD Audit steering group

Introduction:
25% of IBD cases present before the age of 18 years and the impact of IBD in children is disproportionately high on the patient, their family or other carers and society. Poor care provision can damage young patient's trust in their clinicians, impacting on long term care.

Methods:
A review of results of adult IBD services 2010 organisational audit data, was done to look at the service provision for patients under 16 years of age, being admitted to adult services.

Results:
109 (54%) of all adult IBD services admitted 1 or more patients aged 16 and under, between 1st Sept 09 and 31st Aug 10, with a primary diagnosis of Crohn's Disease or Ulcerative Colitis. 27% (29/109) admitted 10 or more patients in this time. The median (IQR) number of paediatric admissions was 4 (1-9). Of these 109 services, only 57 (52%) had indicated that they look after patients under 16.
The table below shows the level of service provision available in adult services that did have a defined service for patients less than 16 years.

Service provision for those Adult IBD Services who stated that they did look after any patients aged 16 or under and who admitted at least one patient between 1/9/ 09 and 31/8/10 (n=57)

Area of IBD service provision	Number (%)
Care is provided by or in conjunction/discussion with either a paediatric gastroenterologist or a paediatrician with an interest in gastroenterology.	43 (75%)
Inpatients are looked after in an age appropriate environment	48 (84%)
Patients undergoing endoscopy, have an appropriate endoscopy area with age appropriate facilities	29 (51%)
For patients undergoing endoscopy, there is someone with training or extensive experience in paediatric endoscopy.	31 (54%)
For paediatric patients undergoing endoscopy, there is an Anaesthetist with paediatric training.	38 (67%)
For paediatric patients undergoing endoscopy, there is no access to an appropriate endoscopy area with age appropriate facilities, a person with training or extensive experience in paediatric endoscopy or an Anaesthetist with paediatric training.	13 (23%)
The IBD Service has a surgeon with suitable paediatric experience.	25 (44%)
The IBD Service has a radiologist (performing and reporting) with suitable paediatric experience.	33 (58%)
The IBD Service has Does your IBD Service a dietitian (including the use of exclusive enteral feeding) with suitable paediatric experience.	41 (72%)
The IBD Service has an IBD/GI Nurse Specialist with suitable paediatric experience.	17 (30%)
The IBD Service has none of the following personnel with suitable paediatric experience: Surgeon, radiologist, dietitian, IBD/GI nurse.	10 (18%)

Conclusions:
- Adult sites are treating very small numbers individually but collectively these represent a significant number of paediatric patients.
- Although the organisation of care in these adult settings does not meet that set by the IBD standards (A12) it is unclear whether these result in worse patient outcomes.
- It is not possible from current audit data to state that all paediatric services meet this standard of care.
- The next audit will specifically focus part of the patient experience questionnaire on adolescents in all UK settings to see whether adolescent patients rate their hospital experience differently in different care settings.

Changes in weight standard deviation scores of children admitted to a tertiary paediatric hospital

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Background;
There are very few studies examining the nutritional outcome of children admitted to hospital. Poor nutrition is known to impact in the short term on wound healing, immune function and pressure sores in bed bound patients and on growth and development in the long term.

Aim:
As part of the GOSH CQUIN audit for nutrition the weights of a representative sample of children discharged between 01.06.11 to 24.02.12 were compared to their admission weights as a proxy for nutritional sufficiency during their hospital stay. It was hoped that this information would be useful in the future to guide dietetic interventions and service to areas with particularly vulnerable patients.

Subjects & Methods
Patients were selected that were discharged on a particular day of the week .with length of stay>than 7 days and who were known to the dietetic department. The following information was obtained from the dietetic / medical notes: wt on admission, date taken, sex, gestational age if < 2yrs, the last wt taken during the hospital admission, date taken. The LMS growth programme was used to calculate the weight standard deviation (SD) scores on admission and discharge.

Results
Data was collected for 175 children over a 37 week period. The number of children with an identifiable weight on admission and discharge was 154 (88% of the total sample) The mean variance in SD score for weight from admission to discharge was -0.17. Sub-analysis for different age groups;0–1 year, 59 children(38%), -0.35, 1–2 years, 13 children(8%) -0.08, > 2 years 82 children(53%) -0.06. The specialties with the greatest variance (>-0.25) were: BMT 8 children(5%) -0.55, Cardiology /CICU 27 children(17%) -0.5, Endocrinology 5 children (3%)-0.29 NICU 11 children (7%)-0.26

Summary and Conclusion
The data suggests that infants under the age of 1 year have a poorer nutritional status on discharge than those aged >1 year. This is to be expected due to their high nutritional requirements and difficulties maintaining these during the hospital admission.
Because of the small numbers of patients in each specialty it is difficult to draw firm conclusions. Cardiology / CICU patients are known to be nutritionally vulnerable due to increased energy requirements often coupled with fluid restrictions. Patients undergoing BMT have increased LOS, severe mucositis and rely on parenteral nutrition for several weeks of their admission. The current GOSH protocol does not allow IV lipid to be given to these patients for one month post BMT due to the risk of VOD, thus restricting the energy that can be provided during this time.

This data cannot be extrapolated to all children admitted to GOSH as it is unknown whether the most nutritionally vulnerable patients are referred to the Dietetic Department. The Trust wide introduction of a nutrition screening tool in January 2012 should now ensure that all children at nutritional risk are referred and will be included in our future audits. It may be that the very complex nature of the children admitted to GOSH and the procedures that they undergo precludes maintenance of nutritional status.

Coeliac disease is more common in children with high socio-economic status

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Introduction
There are a number of genetic and environmental factors which are associated with an increased risk of developing coeliac disease (CD). Socio-economic deprivation is one of these but it is not clear how this increases or reduces the development of CD.

Methods
A cross-sectional study identified all children <16 years old diagnosed with CD in the same tertiary paediatric centre between January 1995 and December 2012. Data, including age at diagnosis and post code, were collected and these were linked with the quintile rank of the Welsh Index of Multiple Deprivation score 2008, a robust measure of socio-economic status.

Results
The overall prevalence of CD in the population studied was 0.75 over 1,000, with a median age at diagnosis 8 years (range 0.8-16 years). There was a graded association between the prevalence of CD and the rate of socio-economic deprivation, with the rate higher in children living in more affluent areas (OR 0.48 in 95% CI 0.279-0.597) with the difference between the lowest deprivation quintile and highest deprivation quintile the most significant.

Conclusion
In our population it is clear that CD is more common in children in the higher socio-economic group, despite the higher rates of breast feeding and lower rates of infection in affluent communities. The reasons for this are not clear but perhaps both the 'hygiene hypothesis' and the health seeking behaviours of parents with high socio-economic status are possible factors in the more frequent diagnosis of CD in this group.

Early presentation of Achalasia

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16 months old male child with history of recurrent wheezing episodes and vomiting presented multiple times to accident and emergency department. Initially treated for viral upper respiratory tract infections. However subsequently given the increasing association of symptoms of feeding and nocturnal cough he was empirically commenced on anti reflux therapy. Despite of antireflux medication he remained symptomatic and then started to falter in growth so was referred to a general paediatrician. A chest X ray was done which showed patchy changes bilaterally predominantly on right side and a dilated esophagus. Given these findings he was presumed to have micro aspirations as a result of supraesophageal manifestation of gastro esophageal reflux disease. His antireflux medications were escalated and was referred for a tertiary paediatric gastroenterology opinion.

He had a normal antenatal and a postnatal course. Feeding was not problematic in first 6 months and weaning was started at 7 months without any particular problems with progression of his solids. There was no significant past medical history however he required two courses of oral antibiotics for presumed lower respiratory tract infections. He had no notable manifestation of atopic disease and there was no family history of note.

He was further investigated with a barium swallow which confirmed dilated esophagus with tapering of distal esophagus and reported intraesophageal reflux. He had a videofluroscopy swallow study which excluded any aspiration or penetration in the pharyngeal phases, however the esophageal phase of the study was noted to be abnormal.

He proceeded to have had an upper gastrointestinal endoscopy. At endoscopy he was noted to have a dilated distal esophagus with sa fluid level however normal esophageal mucosa macroscopically. He was noted to have a tight lower esophageal sphincter and gastric intubation was failed with a paediatric endoscope. A guide wire was ultimately passed into the stomach and he underwent a pneumatic balloon dilatation under fluoroscopy. Under radiological screening he had 3 dilatations with an 8 mm balloon. Given his respiratory symptoms and X-ray changes a nasogastric tube was opportunistically passed at the time of endoscopy and post endoscopy he underwent a tuboesophagogram which excluded tracheo – esophageal fistula. Esophageal biopsies were histologically normal and specifically excluded reflux oesophagitis. His symptoms improved for the first week post dilatation but were not sustained and he proceeded to have repeated dilatation with a larger diameter balloon a month later which failed to have a sustained clinical response. MRI thorax excluded any external compression. Given his failure of response to dilatation, normal histology and radiological features all of which were suggestive of achalasia he was then referred to paediatric surgical colleagues. He underwent laparoscopic Heller's myotomy. At laproscopy the appearances were compatible with the working diagnosis of achalasia. He proceeded to have Heller's myotomy with a partial anterior wrap (usual practice for surgeons with a vast experience of patients with paediatric achalasia). His post operative recovery was unremarkable and he was discharged on day 3 of surgery.

Subsequent follow-ups revealed that he continued to vomit and the feeding was very problematic so proceeded to have a repeat endoscopy with dilatation. At the endoscopy it was now possible to intubate his stomach with a high resolution manometer catheter. Post dilatation after endoscopy when the child had recovered from anesthesia he underwent a dynamic high resolution manometry study this revealed hypertensive lower esophageal sphincter which was non relaxing on repeated swallows and also showed dysfunctional esophageal peristalsis. This was consistent with diagnosis of Type II Achalasia according to Chicago classification.

Efficacy of Infliximab in Paediatric Ulcerative colitis: single tertiary UK centre experience

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Background:
The role of Infliximab in the treatment of paediatric ulcerative colitis (UC) is increasingly recognised.

Aim:
he aim of our study was to evaluate the efficacy of Infliximab in children with ulcerative colitis who failed conventional treatment.

Subjects and Methods:
All children with UC who received Infliximab between April 2006 and October 2012 in our centre were identified. Clinical response and long term outcomes were assessed.

Results:
Twenty eight (28) patients with moderate/ severe UC were included in the study (17 males). Median age at histological diagnosis was 11.6 years (range 2.6y-15.2y), with median duration of disease prior to first Infliximab infusion 1.6y (5 weeks-5.5y) and median follow up post Infliximab regardless of outcome 1.6y (0.5y- 5.4y).

Fifteen patients (15/28, 53.5%) responded initially to the treatment. Nine patients (9/15, 60%) had sustained clinical response at 5mg/kg 8 weekly with histological evidence of mucosal healing, with median duration of treatment 1.6y (0.7y-5.4y). In six patients (6/15) Infliximab had to be discontinued due to either loss of initial response or allergic reaction.

Out of 13 patients with partial or no response to Infliximab (13/28, 46.4%), 6 failed escalation of medical treatment with other immunosuppressive agents , including Adalimumab and Sirolimus, and underwent colectomy 1 month-11 months later (median 0.6y), at median age 13.7y (8.9y-15y).

Summary and Conclusion:
Infliximab was efficacious in 53.5% of children with ulcerative colitis who failed conventional treatment, with response maintained in 60% of those patients. Therefore, Infliximab should be considered not only as rescue treatment, but also as maintenance treatment in children with refractory disease.

Eosinophilic Gastrointestinal Disease preceding Inflammatory Bowel Disease: A Paediatric Case series

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Introduction
Eosinophilic Gastrointestinal Disease (EGID) is a gastrointestinal inflammatory disorder dominated by mucosal eosinophil infiltration in the absence of known causes of eosinophilia. The presenting symptoms are often similar to those of inflammatory bowel disease (IBD).

Aims
We present 3 paediatric cases of EGID that later developed IBD. To our knowledge there are no other cases in the literature.

Case 1
A four year old boy presented with a history of abdominal pain, weight loss, constipation and rectal bleeding. Total IgE was elevated (283). Endoscopy was macroscopically normal; however histology revealed an increased eosinophil count in the colonic lamina propria with focal eosinophilic infiltration of the surface mucosa and crypt epithelium. He was commenced on an exclusion diet. He re-presented two years later. Repeat endoscopy revealed gastritis and aphthoid ulcers in the rectosigmoid region. Histology showed increased plasma cells in the gastric biopsies, patchy blunting of the villus architecture in the duodenum and an increase in plasma cells and eosinophils. Colonic biopsies were normal. Video capsule endoscopy revealed severe enteritis with skips lesions of ulceration and cobble-stoning consistent with small bowel Crohn's disease (CD).

Case 2
A thriving 6 year old boy presented with a 3 month history of diarrhoea and vomiting. Laboratory investigations showed elevated Total IgE (657). He was commenced on an exclusion diet with successful resolution of his symptoms. He re-presented a few months later. Endoscopy revealed mild gastritis and a macroscopically normal colon. Histology showed an increase in eosinophil density in the duodenum and throughout the colon. He continued on an exclusion diet. He represented 14 months later and endoscopy revealed gastritis and colitis with ulcers in recto-sigmoid region. Histology showed an increase of inflammatory cells in the colon with granuloma formation.

Case 3
A 13 year old boy presented symptoms of diarrhoea, abdominal pain and intermittent rectal bleeding. Laboratory investigations revealed mildly elevated inflammatory markers. The total IgE was 197. Endoscopy revealed moderate gastritis, prominent lymphoid follicles in the transverse colon and patchy rectal erythema. Histology revealed chronic gastritis with eosinophil dominated inflammation in the colon. Eighteen months later he re-presented and repeat endoscopy revealed gastritis, duodenitis and severe pancolitis. Histology confirmed chronic active inflammation within the stomach and duodenum, focal ulceration and distortion of the crypt architecture, there was severe active chronic inflammation with cryptitis and crypt abscess formation. No granulomata were identified. Subsequent endoscopies were consistent with ulcerative colitis.

Discussion
The complex role of eosinophils in gastrointestinal inflammation is not fully understood. These cases could support the hypothesis that eosinophils are involved in the initiation of IBD. Further research is needed to ascertain whether EGID is a pre-IBD condition or if patients with EGID are more predisposed to IBD. These cases also highlight the importance of surveillance in children with EGID.

Gastrointestinal manifestations of Ehler-Danlos Syndrome type 3: A Paediatric cohort.

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Background:
Ehler-Danlos syndrome(EDS) is a group of inherited connective tissue disorders, caused by a defect in the synthesis of collagen. EDS type 3 also known as hyper mobility syndrome, characteristically has associated gastrointestinal disorders. The clinical phenotype of EDS type 3 is well recognised in adults but there is very little data in children.

Aim:
To describe the clinical presentation of gastrointestinal symptoms associated wth EDS type 3 in a paediatric cohort of patients.

Methods:
Retrospective review of notes and electronic patient records of all children less than 18 years old diagnosed with EDS type 3 (Beighton score>4/9) referred to the paediatric neurogastromotility clinic. Patient symptom questionnaire was also used. The study period was January 2011 to March 2012.

Results:
A total of 56 children were recruited with a median range of 13yrs (range 2yrs-18yrs). Gender distribution: female n=37(66%), male n=19(34%). Ethnic origin: White British n=42, MWBC n=4, Asian n=3, AsianBritish n= 1,Other 6. 89% presented with symptoms of abdominal pain associated with either GOR(n=27), bloatedness(n=35) or constipation (n=46). 33 patients had nausea and 23 patients had associated vomiting. 6 children complained of dysphagia and 2 children had oesophageal spasm. 11 patients had IBS type symptoms. 82% had constipation and in 3 patients this was complicated by perianal fissures with prolonged healing times, 4 patients had faecal incontinence and soiling, 2 patients had painful rectal spasms suggestive of rectal evacuatory disorder. All patients had hypermobility and 52% has persistent chronic joint pain. 14 children had balance problems and this along with joint pain led to 9 patients using wheelchair for mobility. 15 children reported recurrent joint dislocation and 6 had fractures. 20 children reported chronic fatigue. Weight loss was noted in 18 children.28 patients had autonomic dysfunction, 17 had postural orthostatic tachycardia syndrome and orthostatic hypotension was found in 20 patients.8 of the 20 children who had impedance study showed pathological reflux while 5 out of 15 children showed reflux on barium study. 13/16 children had delayed gastric emptying. High resolution oesophageal manometry showed dysmotility (hypertensive peristalsis) in 6/11, small bowel manometry abnormal in 3/5 and anorectal manometry abnormal in 2/3, delayed colonic transit with megarectum noted in 7/15 children.

Medical management was with antireflux medication, prokinetics, laxatives, antiemetics and probiotic. Orthostatic hypotension was treated with fludrocortisone and sodium chloride.13 were on amitryptilline and gabapentin for pain. Nutritional management with exclusion diet and FODMAP were used in 6 children with some effect. Nutrition rehabilitation due to feeding difficulties and weight loss were required in the form of enteral feeds in 15 children either via NG, NJ, PEG or PEG-J. Parenteral nutrition was required in 5 children.Surgical treatment was required in 6 patients: defunctioning ileostomy n=1,colostomy n=1, colectomy with ileostomy n=1, gastric pacing n=1.Multidisciplinary input through dietician n=15, pain team n=8, physiotherapist n=17, occupational therapist n=4, psychologist n=18 and rheumatologist n=9 were required.

Discussion:
Hypermobility syndrome and the associated GI symptoms are being more and more recognised. There is preponderance in Caucasian females. 41% have a strong family history. There is increased incidence of dysmotility. Medical, nutritional and/or surgical management is useful in achieving symptomatic relief. Nutritional rehabilitation is very essential to maintain growth and development.

Conclusion:
Children with a diagnosis of EDS type 3 present with a wide spectrum of foregut, midgut and hindgut motility disorders. Associated autonomic dysfunction and hypermobility with lax joints can be quite debilitating. A holistic approach to management through the multidisciplinary team is required.

Hereditary Folate Malabsorption Effect of systemic folate supplements on myelination

1) *Dr Siba Prosad Paul, Yeovil District Hospital (Previously at SRH, Chichester)*
2) *Prof David Candy, St Richard's Hospital, Chichester*

Introduction:
Folic acid is a vital substrate for normal development of the foetus and infant. Folate deficiency antenatally leads to spina bifida. This case highlights the importance of folate in children for cortical myelination.

Methods:
4 month male infant, presented with history of poor feeding and pallor. Physical examination was unremarkable. Parents were non consanguineous and the infant was healthy till this presentation.

Results:
Blood investigations revealed megaloblastic anaemia with a haemoglobin 7.1gm/dl and MCV of 96fL. Further investigations showed a serum folate 0.1mcg/L. Started on IV folinic acid, however his respiratory status deteriorated and was transferred to PICU for respiratory support. CXR was indicative of PCP pneumonitis and Septrin® was started. BAL (Broncho Alveolar Lavage) confirmed presence of PCP. He received immunoglobulins for a short period of time and the immune status improved. Currently he is managed on IM folic acid. The serial MRI Brain scans showed delayed myelination pattern but no demyelination. The child recently started having difficult epilepsy and responded to sodium valproate.

Discussion :
- HFM is pan-ethnic, autosomal recessive.
- SLC46A1 gene demonstrated in HFM
- Folate may be absent in the CSF.
- Presents from 2 month with anaemia, immune deficiency, neurologic manifestations.
- FBC, serum folate, Vitamin B12 level are initial investigations.
- Transient immunodeficiency may be seen at the start of treatment.
- Red cell folate and CSF folate necessary for monitoring
- Serial MRI scans necessary to demonstrate myelination pattern
- Lifelong folate replacement therapy necessary
- Involvement of metabolic specialist, immunologist, gastroenterologist and developmental paediatrician necessary.

Conclusion:
This case highlights the importance of investigating rare causes of macrocytic anaemia in the infancy. A normal neurological and developmental outcome may be achieved by aggressive folate replacement therapy.

How common are trace element and vitamin deficiencies during long term parenteral nutrition?

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Aims:
Trace elements and vitamins are routinely provided during parenteral nutrition (PN), but precise requirements are uncertain. We investigated abnormalities of zinc, copper, selenium, ferritin, and fat soluble vitamin status in children receiving home PN (HPN), how often additional supplementation was deemed clinically necessary, and whether current monitoring practice is justified.

Methods:
HPN patients (n = 13) under the care of a tertiary referral centre in 2012 were identified through the nutrition support team, and results of routine 3 monthly biochemical monitoring recorded together with changes in micronutrient supplementation. Abnormality was defined using the hospital laboratory reference ranges. Underlying diagnosis was neonatal short bowel syndrome (n = 12) and pseudo-obstruction (n =1).

Results:
Monitoring took place over a total of 17.7 years of HPN. 8/13 (61%) patients had an abnormal result that prompted a change in supplementation. The frequencies were as follows - vitamin A low x 5; vitamin D low x 30, high x 3; vitamin E - 0; clotting prolonged x 3; ferritin low x 12; selenium low x 13 ; zinc low x 1, high x 3; copper low x 4, high x 7. Nutrient supplementation was adjusted as follows: vitamin A x 1; vitamin D x 3; ferritin x 5; selenium x 3; zinc x 1; copper x 2.

Conclusions:
Important micronutrient abnormalities requiring intervention occurred during HPN but were relatively infrequent (1 per 14 months). Since deficiencies have potentially important clinical consequences, regular 3 monthly monitoring is justified.

Influenza Vaccination Uptake in Inflammatory Bowel Disease- Is there room to improve?

Dr Damien Cheema, Dr Rafeeq Muhammed, Department of paediatric gastroenterology, Birmingham Children's Hospital NHS Foundation Trust.

Aim:
The aim of our study is to assess the seasonal influenza vaccination uptake in patients with inflammatory bowel disease (IBD)

Methods:
We have conducted a telephonic survey of our IBD patients in February 2012 to assess the influenza vaccination uptake for winter 2011-2012.

Results:
140 children had responded to this survey (61.6% of our IBD patients). 84 children had Crohn's disease, 35 had Ulcerative colitis and 21 had IBD unclassified. Majority of these children (90/140) were on immunosuppressive treatments. 61 children (44%) had received seasonal influenza vaccination in that winter. 21 of them received in October, 20 in November, 13 in December and 3 in January. Out of the 79 children who have not received the influenza vaccine, 42 were not aware of the need for vaccination and did not have the influenza vaccine in the previous winters as well. 10 children were aware of the need for the influenza vaccine; however they opted not to receive the vaccine. 14 children intended to receive the vaccine, however this was deferred due to various reasons like intercurrent illness, family bereavement and difficulties experienced the General Practice surgery. Only one IBD patient needed hospitalisation in 2011 and 2012 with Influenza infection, however this was in July before the vaccination had started.

Discussion:
Department of Health advises influenza vaccination for immunosuppressed individuals and also for children with medical conditions, who may need treatment with steroids for more than a month. European Crohn's and Colitis Organisation (ECCO) recommend influenza vaccination for IBD patients on immunomodulators. Experience from Philadelphia, Boston and Poland show that good, but variable, antibody response occurs after influenza vaccination in children and better protection occurs against type A strains. Side effects, both local and systemic, are generally mild. Experience from Australia and Germany show that the seasonal flu vaccination uptake in IBD patients are generally low, 10% and 16% respectively. We would like to hear from other centres about their experience of influenza vaccination uptake in IBD patients. Further efforts need to be done to increase the awareness of influenza vaccination in patients with IBD.

Conclusion:
Influenza vaccination uptake in our IBD patients are better than reported from other centres, however further work needs to be done both locally and nationally to improve the influenza vaccination uptake.

Lower Gastro-intestinal bleeding in children without inflammatory bowel disease: A single centre endoscopy experience

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Introduction:
Lower gastrointestinal bleed is a common indication for colonoscopy in children. Even though the diagnostic yield of colonoscopy is high in children presenting with bloody diarrhoea, it is not as useful in identifying aetiology in painless rectal bleeding in absence of diarrhoea1.

Aims:
We describe our experience in children who presented with painless lower GI bleeding in absence of diarrhoea. Our study focuses primarily on presentation, aetiology, endoscopy findings and the overall outcome.

Methods:
A retrospective review was conducted on children presenting with painless lower gastro-intestinal bleed without diarrhoea. Data collected included demographics, clinical spectrum, endoscopic assessment, histology and management with outcome. Total number of patients assessed were 26 over a 12 month period (June 2011- June 2012). The number assessed did not include patients who had a final diagnosis of inflammatory bowel disease.

Results:
A total of 26 children were identified. The mean age of presentation was 9.17 years with male female ratio of 0.73:1.The duration of bleeding at first assessment was less than a week in 11% (3), less than a month in 7% (2), between one to six months in 26% (7), longer than 6 months in 52% (14) and not known in one patient. Haemoglobin at the time of presentation was < 12 mg/dl in 31% (8), > 12 mg/dl in 56% (15) and no data was found in 3 patients. Associated symptoms of constipation and abdominal pain were found in 31% (8) and 35% (9) respectively. Colonoscopy was performed in 69% (18) of the total cases. On colonoscopic assessment, polyps were found in 2 cases. In both the cases the polyps were removed and histologically diagnosed as juvenile polyps. Solitary rectal ulcer was diagnosed in 1 patient. The colonoscopic assessment of the remaining 14 patients did not show any abnormality on macroscopic examination. Increased density of eosinophils in colonic mucosal biopsy was seen in 6 patients. There was no evidence of food allergy, helminthic infection or peripheral eosinophilia found in these cases. The average duration of symptoms prior to diagnosis in this group of patients was 9 months as compared to 4 months in children with no evidence of eosinophilia, which was significantly longer (p 0.02). Histological examination in two patients showed features of non specific inflammation. **Outcome at 6 months FU:** Bleeding resolved in all but 1 patient who did not undergo colonoscopy. The single patient with persistent bleeding in this group had associated features of constipation and anal fissure. The group of patients, who underwent endoscopy and polypectomy had no further bleeding. The child with solitary rectal ulcer continued to bleed and developed rectal prolapse. Bleeding resolved at the end of 6 months in 6 out of 7 (85.7%) children with normal histology. In children with tissue eosinophilia seen in colonic mucosa, bleeding was persistent at 6 months in 3 patients (50%).

Conclusions:
An obvious source of bleeding was found in 16% (3) of patients who underwent endoscopy. This number is small when compared to the number of procedures performed to identify aetiology. The prevalence of colorectal polyps in the study was 11% which is comparable to figures of 12% previously reported2. Tissue eosinophilia was found in 33% of cases who underwent colonoscopy for persistent rectal bleeding and was associated with persistence of symptoms. It is debatable as to whether increased eosinophils in colonic biopsies are contributory to aetiology of bleeding or whether it is purely an incidental finding. Larger study is required to establish a causal link between rectal bleeding and colonic tissue eosinophilia.

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Survey of dental health in children who are on home parenteral nutrition

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Introduction
Administration of Home Parenteral Nutrition (HPN) in adults is known to be associated with poor oral health1. Little is known about the incidence of dental problems in children who are on long term home parenteral nutrition.

Aim
To determine the presence of dental problems in children who are on long term home parenteral nutrition.

Methods
Long term HPN was defined as administration of HPN for a period of more than 3 months. Information about the duration of PN, methods of enteral feeding, breast and bottle feeding in infancy, frequency of dental visits, dental problems, frequency of brushing and use of tooth paste were collected by emails, questionnaires and telephone consultations.

Results
Dental health on twenty five children who were on HPN were analysed. The age group of study population was 1-18 years with a median age of 5.5. The average duration of PN administration was 4.3 years. Just under half had oral feeding concomitantly. 76% of patients have had breast and bottle feeding in infancy. 56% of children reported dental problems. 28% had teeth staining, 8% had gum infection, 16% had teeth decay and delayed dentition. 96% of children brushed regularly and 68% reported using tooth paste. 64% visited dentist on 2-12 monthly intervals.

Discussion
Concerns exist between poor oral hygiene and catheter related infection. 60% of adults on HPN are reported to have problems with oral health1. In England, 41-54% of children have tooth decay and 67% of children have non carious dental condition3. When compared to the above two, incidence of dental problems was lower in our study group. Micronutrient supplementation is known to improve oral inflammation4. Children on HPN are regularly monitored and supplemented with micronutrients and thereby deficiency is uncommon in them. This also could possibly explain the lesser incidence of dental problems on children who are on HPN.

Conclusion
Children on HPN have better dental health when compared to national statistics on children and adults on HPN.

Reference

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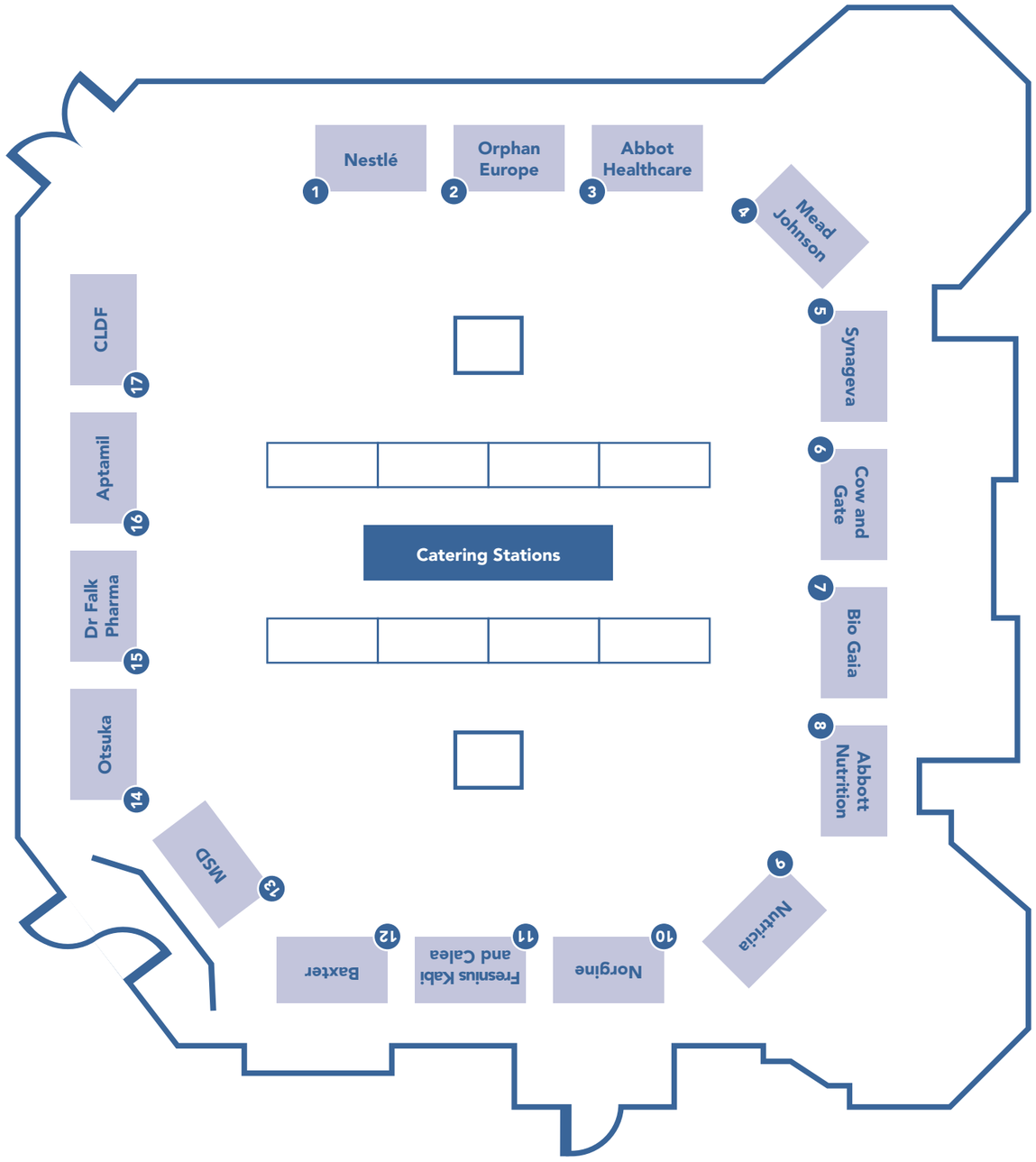
Exhibitors Floor Plan



NACC and CICRA will be in the Octagon lounge

Notes