BSPGHAN
working for children with digestive and liver disorders

British Society of Paediatric Gastroenterology Hepatology and Nutrition

Silver Anniversary

ANNUAL MEETING 2012

Wednesday 25th – Friday 27th January 2012 at Albert Hall, Nottingham

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Dr Charlie Charlton, Consultant Paediatric Gastroenterologist

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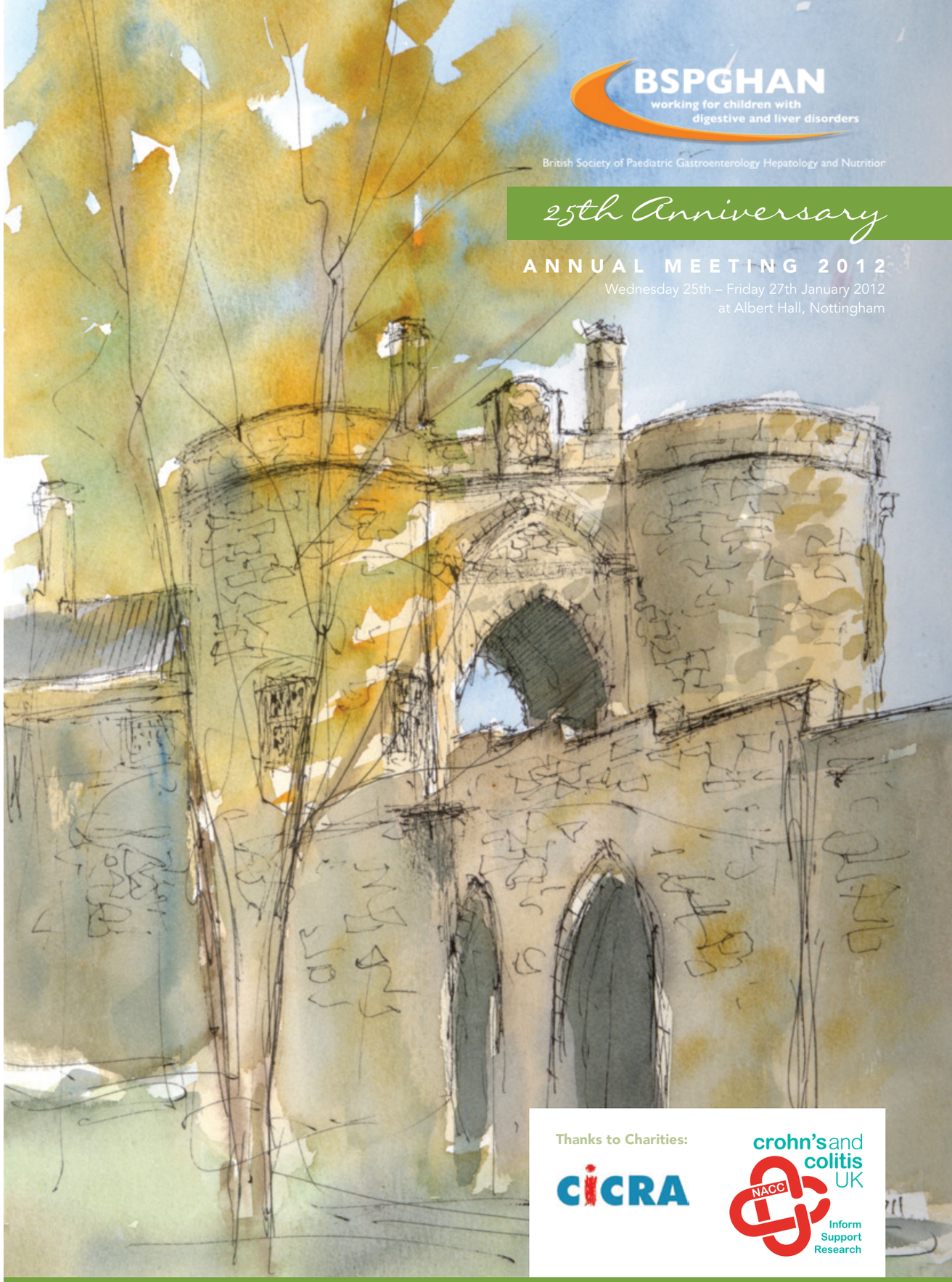

BSPGHAN

working for children with digestive and liver disorders

British Society of Paediatric Gastroenterology Hepatology and Nutrition

25th Anniversary

ANNUAL MEETING 2012

Wednesday 25th – Friday 27th January 2012

at Albert Hall, Nottingham

Thanks to Charities:

CICRA

crohn's and colitis UK

NACC

Inform Support Research

Local Organisers: Dr Charlie Charlton, Consultant Paediatric Gastroenterologist

E&OE and CPD Approved

Thanks to our Principal Sponsors

On behalf of The Society, we thank you all for renewing your most generous support for our meetings which are an essential element in the partnership we establish in managing children with gastrointestinal, liver and nutritional disorders. It is through mutual respect, understanding and cooperation that we have witnessed such major changes in recent times in the way we deliver education, particularly to our trainees and colleagues within the breadth of our speciality and in the way we deliver quality care to our patients. Long may this relationship continue to flourish.

Mead Johnson Nutrition

Caring for young lives
Children's Liver Disease Foundation

NUTRICIA
Advanced Medical Nutrition

Thanks to our Silver Sponsors

The Society is extremely grateful that you are willing to participate in our meeting and to offer such generous support. It is through your willingness to share with us your initiatives and ideas that we continue to move forward as a speciality. Thank you.

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Crohn's in Childhood Research Association
Orphan

Thanks to Exhibitor:

Aptamil

Thanks to Hedley

Carrington FNSA artist of Nottingham Castle on front cover.

Organising Committee

Dr Charlie Charlton; Dr Mark Beattie, Dr Sue Protheroe, Dr Mike Cosgrove, Carla Lloyd

Abstract Selection Committee

Dr Rajeev Gupta, Dr Sian Kirkham, Dr Suresh Babu, Dr Richard Hansen

Social Commitee

Dr Charlie Charlton; Dr David O'Neil, Dr Sarang Tamnhe, Sarang Tamhne

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Louise Dilloway

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Prestige Print & Design, Birmingham

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

Welcome to Nottingham for the Silver Anniversary Year of our society. The Nottingham team is honoured to host this special meeting. BSPGHAN was started by a small group of doctors who organized the gastroenterology session at the BPA spring meeting. They set up a winter meeting to encourage research and development of the specialty. We now are a large multidisciplinary and multiprofessional society with a strong emphasis on clinical standards, training, guideline development, quality improvement and advocacy for children's health and commissioning. These are all very relevant issues to us practicing in the NHS in the 21st century. I am delighted to have had so many abstracts submitted including challenging cases and hope you will enjoy the plenary and poster presentations.

I hope all participants will enjoy the meeting. It is an opportunity to network and exchange ideas. It is an opportunity to catch up with friends and colleagues.

I hope you enjoy the social programme and take the opportunity to view Nottingham and visit the famous Nottingham Castle particularly on the Wednesday night.

I have had fantastic advice help and support from the team and many people in developing the programme and organizing the meeting. You will all have observed the unstinting work that Carla does for the society and I must tell you that what you see is only the tip of the iceberg. I have seen her energy, dedication and conscientiousness at close hand and this is only her part-time work! There have been sterling contributions by Mark, Sue, Mike and Rajeev & the abstract committee.

I also thank my family for their patience over the last year.

The Sponsors from industry have provided indispensible financial support as well as advances in treatments and equipment, which improve our care of children. I look forward to thank them and to view their stands with you all.

I thank the speakers and chairs who have shown great flexibility in the ever-changing programme.

WELCOME TO YOU ALL AND THANKYOU FOR COMING! IT'S YOUR MEETING NOW.

Charlie Charlton

From bottom stairs: David O'Neill, Sharon Thomas, Mary Weston, Anette Richardson, Ruth Prigg, Jenny Creek, Richard Hastings, Lucy Davies, Sian Kirkham, Charlie Charlton, Sarang Tamhne

Missed from picture: Michelle Short, Amy-Jo Hart

Wednesday 25th January 2012

POSTGRADUATE DAY CONFERENCE
Albert Hall Nottingham

8.00
Registration Opens - Great Hall Foyer

Coffee and Exhibition in Osborne Suite

9.50 Meeting Opens

Session 1
9.50 – 11.00

Chairs:
Dr Mike Cosgrove
Consultant Paediatric Gastroenterologist, Singleton Hospital, Swansea
and
Dr Dharam Basude, Consultant Paediatric Gastroenterologist,
Royal Bristol Children's Hospital, Bristol

9.55 – 10.00

Welcome and Introduction
Dr Charlie Charlton
Consultant Paediatric Gastroenterologist
Queens Medical Centre
Derby Road
Nottingham
NG7 2NH

10.00 – 10.20

How can we prove Reflux and aspiration?
Dr David Rawat
Consultant Paediatric Gastroenterologist
Barts and The London
Turner Street
London
E1 2AD

10.20 – 10.40

Finances and developing tariffs
Dr Balaji Krishnamurthy
Consultant Paediatric Gastroenterologist
Royal Liverpool Children's NHS Trust
Alder Hey
Eaton Road
Liverpool, L12 2AP

10.40 – 11.00

Rectal Manometry and Ultrasound: How, When and Value?
Mr Shalinder Singh
Consultant Paediatric Surgeon
Nottingham Children's Hospital
Derby Road
Nottingham

Session 2
11.00 – 12.20
Challenging Case Presentations

Chairs:
Dr Sue Protheroe
Consultant Paediatric Gastroenterologist,
Birmingham Children's Hospital, Birmingham
and
Dr Mark Dalzell
Consultant Paediatric Gastroenterologist,
Royal Liverpool Children's Hospital, Liverpool

11.00 – 11.20

What should we recommend for a remarkably asymptomatic 6 year old child with extensive Crohn's Disease?
Whyte L, Jenkins H, Davies I
Department of Paediatric Gastroenterology
University Hospital of Wales
Heath Park
Cardiff
CF4 4XN

11.20 – 11.40

New association in systemic rotavirus illness: encephalopathy with vasculitis
Paul S[1] Candy D[2], ST4 in Paediatrics,
[1]Great Western Hospital, Swindon
[2]St Richard's Hospital, Chichester

11.40 – 12.00

The challenges of transfer from paediatric to adult care post liver transplantation
Henderson L, Milson C, Gouge J, McClean P.
Leeds Teaching Hospitals NHS Trust
Leeds General Infirmary
LS1 3EX

12.00 – 12.20

Coeliac Disease: To biopsy or not – NICE v ESPGHAN
Professor Simon Murch, Consultant Paediatric Gastroenterologist
Clinical Sciences Research Institute
Clifford Bridge Road, Coventry, CV2 2DX

12.20 – 13.20
LUNCH

Poster Viewing and Sponsor Exhibition
Osborne Suite, Osborne Foyer and Great Hall Foyer

Session 3
13.20 – 14.50
& Plenary abstract session one

Chairs:
Dr Rajeev Gupta
Consultant Paediatrician, Barnsley Foundation Hospital, Barnsley
and
Dr Richard Hansen
Clinical Lecturer in Child Health
University of Aberdeen, Royal Aberdeen Children's Hospital, Aberdeen

13.20 – 13.50

Speaker TBC
XXXXX
XXXXX
XXXXX
XXXXX
XXXXX

13.50 – 14.00

2 year experience of a catheter free, radio telemetric, oesophageal pH monitoring (Bravo®) system in children at a single UK tertiary centre
Rao N, Campbell I D, Taylor C.J., Narula P, Thomson M, Rao P
Department of Gastroenterology
Sheffield Children's Hospital
Western Bank
Sheffield
S10 2TH

14.00 – 14.10

Effectiveness of double-balloon enteroscopy facilitated polypectomy in children with Peutz-Jeghers syndrome
Rao P[1], Urs A[1], Despott EJ[2], Fraser C[2], Thomson M[1]
[1]Center for Paediatric Gastroenterology
Sheffield Children's Hospital NHS Foundation Trust
Western Bank
Sheffield
S10 2TH
[2]Wolfson Unit for Endoscopy
Polyposis Registry
St Mark's Hospital and Academic Institute
Imperial College London
United Kingdom

14.10 – 14.20

Faecal calprotectin for the diagnosis of paediatric inflammatory bowel disease: a meta-analysis
Paul Henderson[1], Niall H Anderson[2], David C Wilson[1]
[1] Child Life and Health
[2] Centre for Population Health Sciences
University of Edinburgh
Edinburgh
United Kingdom

14.20 – 14.30

A risk score for intestinal failure in gastroschisis
A Gregory [1,2], J M Wells[2], A R Bremner[1]
Paediatric Gastroenterology[1], Paediatric Surgery[2],
Birmingham Children's Hospital
Steelhouse Lane
Birmingham
B4 6NH

14.30 – 14.40

The epidemiology and natural history of paediatric inflammatory bowel disease in a UK region: a prospective 14-year study in SE Scotland
Paul Henderson[1,2], Pam Rogers[2], David Mitchell[2], David Devadason[2], Peter M Gillett[2], David C Wilson[1,2]
[1]Child Life and Health, University of Edinburgh and [2]Department of Paediatric Gastroenterology and Nutrition
Royal Hospital for Sick Children
Edinburgh
United Kingdom

14.40 – 14.50

Comparison between oral (liquid and modigraf) and intravenous induction with tacrolimus in intestinal transplantation patients
Jayachandran S, Sharif K, Taha A, Khalil B, Mirza D, Gupte G.
Liver unit (including small bowel transplantation)
Birmingham Children's Hospital
Steelhouse Lane
Birmingham
B4 6NH

14.50 – 15.20
AFTERNOON TEA

Poster Viewing and Sponsor Exhibition
Osborne Suite, Osborne Foyer and Great Hall Foyer

Session 4
15.20 – 15.35

Chair:
Dr Sue Protheroe, Consultant Paediatric Gastroenterologist,
Birmingham Children's Hospital, Birmingham

15.20 - 15.35

National IBD Audit
Dr Sally Mitton
Consultant Paediatric Gastroenterologist
Dept of Child Health
St George's Hospital Medical School
Cranmer Terrace
London, SW17 0RE

Symposium sponsored by MSD
15:45 – 16:45

Chair:
Dr Robert Heuschkel, Consultant Paediatric Gastroenterologist
Addenbrookes Hospital, Cambridge

A debate on getting the balance right with biologicals for children and adolescents with IBD

Professor Séverine Vermeire, MD, PhD
Department of Gastroenterology
University Hospital Gasthuisberg
Herestraat 49
3000 Leuven
BELGIUM

Dr Richard Russell
Consultant Paediatric Gastroenterologist
Yorkhill Hospital
Dalnair Street
Glasgow
G3 8SJ

16.50 – 17.50
Professional Group Meetings - Albert Hall, Nottingham

Open to all delegates
Associate Members AGM - Balmoral Gallery
Trainee Members - Great Hall
IBD Quality Improvement Feedback Forum - City Suite

18.00 – 19.00
Group Meeting - Park Plaza Hotel

IBD Nurses Working Group - Vista Suite

18.10
Football: Consultants v Trainees

Social
Anish Kapoor Retrospective Exhibition and Permanent
Exhibits at the Nottingham Castle 18.30 to 20.00
with a guided tour at 19.15 including reference to Robin Hood and castle history by
Deborah Dean Visual Arts Curator

The mirror to the right of the Albert Hall by the Playhouse Theatre
is also by Anish Kapoor.

20.30 – 23.30
Reception for delegates and sponsors with Lord Mayor of Nottingham,
Councillor Michael Wildgust - Nottingham City Council House

Followed by Burns Supper

Thursday 26th January 2012

Albert Hall Nottingham

8.00
Registration Opens - Great Hall Foyer

Coffee and Exhibition - Osborne Suite

8.00 - 9.15
Open Working Group Meetings:
Albert Hall, Nottingham

IBD - Great Hall | Endoscopy - Balmoral Gallery

Motility Working Group - Syndicate 4 | Nutrition - City Suite

9.30 Main Meeting Opens

Session I
9.30 – 11.15

Focus on Inflammatory Bowel Disease

Chairs:
Dr Sian Kirkham
Consultant Paediatric Gastroenterologist, Queen's Medical Centre, Nottingham
and
Professor David Wilson
Professor of Paediatric Gastroenterology, University of Edinburgh

9.30 – 9.35

Welcome and Introduction
Dr Charlie Charlton
Consultant Paediatric Gastroenterologist
Nottingham Children's Hospital
Derby Road
Nottingham

9.35 – 10.05

Bone marrow transplantation and other new treatments for IBD
Professor Chris Hawkey
Professor of Gastroenterology
Nottingham University
University Park
Nottingham
NG7 2RD

10.05 – 10.25

Quality Improvement Programme for IBD through inspection
Ms Emma Fernandez
Project Manager IBD Quality Improvement Project
Royal College of Physicians
11 St Andrews Place
Regents Park
London
NW1 4LE

Dr Ian Shaw and Dr Charlie Charlton

All units invited to review new proforma and join IBD QiP programme

10.25 – 10.45

What matters for patients and families
Mr Richard Driscoll
Chief Executive Officer
Crohn's and Colitis UK
4 Beaumont house
Sutton Road
St Alban's
Herts AL1 5HH

10.45 – 11.00

Preparing children for pouch surgery
Ms Ali Wright
Specialist Stoma Nurse
Nottingham Children's Hospital
Derby Road
Nottingham
NG7 2NH

11.00 – 11.15
Patient and family perspective of severe Crohn's involving liver disease
Howard and Huw Arthur

11.15 – 11.45
COFFEE

Poster Viewing and Sponsor Exhibition
Osborne Suite, Osborne Foyer and Great Hall Foyer

Session II
11.45 – 13.00

Hepatology

Chairs:
Dr Paddy McLean
Consultant Paediatric Hepatologist, Leeds General Infirmary, Leeds
and
Dr Patrick McKiernan, Consultant Paediatric Hepatologist,
Birmingham Children's Hospital, Birmingham

11.45 – 12.10

Drug induced liver injury
Professor Guru Aithal
Consultant Paediatrician Paediatrician
University Hospital NHS Trust
Derby Road
Nottingham NG7 2NH

12.10 - 12.35

Fatty liver disease where we are and what the future holds?
Professor Anil Dhawan
King's College Hospital
Denmark Hill
London

12.35 – 13.00

Therapy and advances In Alpha 1 anti-trypsin deficiency
Professor Noor Kalsheker
Nottingham University
University Park
Nottingham
NG7 2RD

13.00 – 14.00
LUNCH

Poster Viewing and Sponsor Exhibition
Osborne Suite, Osborne Foyer and Great Hall Foyer

Session III
14.00 – 15.15

Plenary abstract Session Two and Oral Poster Abstracts

Chairs:
Dr Jenny Gordon
Research & Development Fellow, RCN Institute, Oxford OX4 2JY
and
Dr Adrian Thomas
Consultant Paediatric Gastroenterologist,
Manchester Children's Hospital, Manchester

14.00 – 14.10

Identification of risk factors for metabolic syndrome and obesity in healthy school children (REACH Y6 cross-sectional pilot study)
Konidari A[1], Boddy LM[2], Newland P[1], Jones J[1], Didi M[1], Thomas NE[3], Hopkins N[2], Graves LEF[2], Foweather L[2], Gobbi R[2], Stratton G[2], Auth MKH[1]
[1]Alder Hey Children's NHS Foundation Trust
Departments of Gastroenterology
Biochemistry, Pathology, Endocrinology
Liverpool
[2]Liverpool John Moores University
Research Institute for Sport and Exercise Sciences, and for Education
Community and Leisure
Liverpool
[3]Swansea University
College of Health Sciences

14.10 – 14.20

Serum protein N-glycosylation as a biomarker of paediatric NAFLD
Blomme B[1†], Fitzpatrick E[2†], Quaglia A[3], De Bruyne R[4], Van Vlierberghe H[1*], Dhawan A[2*]
[1]Department of Hepatology and Gastroenterology
Ghent University Hospital
Ghent
Belgium
[2]Paediatric Liver, GI, and Nutrition Centre
King's College London School of Medicine at King's College Hospital
London UK
[3]Dept. Liver Pathology, Institute of Liver Studies
King's College Hospital
London UK
[4]Departent of Paediatric Gastroenterology, Hepatology, and Nutrition
University Hospital Ghent
Belgium
[†]Joint first authors
[*]Joint senior authors

14.20 – 14.30

Extra-hepatic Portal Venous Obstruction: The Scottish Experience
Cordiner D[1], Devadason D[1], Bishop J[2], Bisset M[3], Goudie D[4]
[1]Royal Hospital for Sick Children, Sciennes Road, Edinburgh
[2]Yorkhill Hospital, Dalnari Street, Glasgow
[3]Childrens Hospital, Aberdeen Royal Infirmary, Westburn Road, Aberdeen;
[4]Raigmore Hospital, Inverness

14.30 – 14.40

Neonatal acute liver failure (NALF: Review of 20 years experience
Ashok D[1], Pietrobattista A[1], McKiernan PJ[1], Kelly DA[1], Hartley J[1], van Mourik I[1], Lloyd C[1], Sharif K[1], Mirza D[2], Gupte G[1]
[1]Liver and Small Bowel Transplant Centre
Birmingham Children's Hospital
Steelhouse Lane
Birmingham B4 6NH
[2]Liver Unit
Queen Elizabeth Hospital
Edgbaston
Birmingham

14.40 – 14.50

Trace element levels in intestinal failure patients and response to parenteral nutrition
Saliakellis E[1], Pichler J[2*], Macdonald S[3], Costa N[4], Horn V[5], Hill S[6]
[1]Department of Paediatric Gastroenterology
Great Ormond Street Hospital NHS Trust
London, UK
[2]Paediatric and Adolescent Medicine
Medical University of Vienna
Vienna, Austria
[3]Department of Dietetics
[4]Department of Chemical Pathology
[5]Pharmacy Department
Great Ormond Street Hospital NHS Trust
[6]Department of Paediatric Gastroenterology
Great Ormond Street Hospital
London, UK

14.50 – 15.00

Six years of the British Intestinal Failure Survey (BIFS)
Gowen H[1], Lloyd C[1], Beath SV[1], Puntis JWL[2]
[1]Birmingham Children's Hospital
Steelhouse Lane
Birmingham, B4 6NH
[2]The General Infirmary at Leeds
Clarendon Wing
Belmont Grove
Leeds, LS2 9N

Oral poster abstracts
15.00 – 15.15

15.00 – 15.05

15.05 – 15.10

15.10 – 15.15

15.15 – 15.45
COFFEE

Poster Viewing and Sponsor Exhibition
Osborne Suite, Osborne Foyer and Great Hall Foyer

Session IV
15.45 – 16.20

Chairs:
Dr Mike Thomson, Consultant Paediatric Gastroentrologist,
Sheffield Children's Hospital, Sheffield
and
Dr Charlie Charlton
Consultant Paediatric Gastroenterologist, Nottingham

15.45 – 16.00

Bleeders come first; How common and who deals with it
Mr Simon Huddart
Consultant Paediatric Surgeon
University Hospital of Cardiff
Heath Park
Cardiff

16.00 – 16.20

Managing Severe Gastrointestinal bleeding in children
Dr Patrick McKiernan
Consultant Paediatric Hepatologist
Liver Unit
Birmingham Children's Hospital
Steelhouse Lane
Birmingham

16.30 - 18.00
ANNUAL GENERAL MEETING

19.30 for 20.00
Gala Dinner and dancing till late
The Albany Suite

Friday 27th January 2012

"MORE OF THE SAME"

8.30
Registration Opens - Great Hall Foyer

7.45 – 8.45

Open Professional Group Meetings
Albert Hall, Nottingham

Gastroenterology - City Suite | Hepatology - Syndicate 4

Education - Balmoral | PEGHAN - Great Hall

9.00 – 10.00

Symposium Sponsored by Mead Johnson

Welcome by Chair: Professor Jon Vanderhof

Cost effectiveness of using an extensively hydrolysed formula compared to an amino acid formula in the initial treatment of cow milk allergy in the community in the UK
Professor Julian F Guest,
Catalyst Health Economics Consultants,
Northwood,Middlesex UK and School of Biomedical Sciences
King's College, London, UK

Gastro oesophageal relux - an update
Professor Colin D. Rudolph
Vice President for Global Medical Affairs and
Clinical Medical Officer
Mead Johnson Nutrition
2400 West Lloyd Expressway
Indianopolis
USA

Session V
10.00 – 11.00

Gastroenterology Session
Chairs:
Dr Michael Green, Consultant Paediatrician, Children's Hospital Leicester
and
Dr Sabari Loganathan - Consultant Paediatric Gastroenterologist
Royal Aberdeen Children's Hospital, Aberdeen

10.00 – 10.05

Welcome

10.05 – 10.25

Post infectious irritable bowel syndrome
Professor R Spiller, Professor of Gastroenterology, Nottingham University Hospital
Queen's Medical Centre, Derby Road, Nottingham NG7 2NH

10.25 – 10.45

Should we exterminate *H. pylori?*
Professor John Atherton
University of Nottingham, Digestive Diseases Centre, Queen's Medical Centre, Nottingham NG7 2UH

Key Note Lecture
10.45 – 11.05

Chairs:
Dr Charlie Charlton, 2012 BSPGHAN Annual Meeting Organiser
Consultant Paediatric Gastroenterologist, Nottingham
and
Dr Mark Beattie, President of BSPGHAN,
Consultant Paediatric Gastroenterologist, Southampton

10.45 – 11.05

Paediatric Gastroenterology in Facing the Future: Standards for Paediatric Services
Professor T Stephenson, President
RCPCH, 5-11 Theobalds Road
London, WC1X 8SH

11.05 – 11.30
COFFEE

Poster Viewing and Sponsor Exhibition
Osborne Suite, Osborne Foyer and Great Hall Foyer

Session VI
11.30 – 12.30

Nutrition
Chairs:
Ms Jenny Creek, Paediatric Dietitian, Nottingham Children's Hospital, Nottingham
and
Dr John Puntis, Consultant Paediatric Gastroenterologist, The General Infirmary at Leeds

11.30 – 11.50

Management of really sick patients with anorexia nervosa (MARSIPAN)
Dr Dasha Nicholls, Consultant Child and Adolescent Psychologist
Great Ormond Street Hospital, Great Ormond Street, London

11.50 – 12.10

What is new in network commissioning
Dr Tim Bowling, BAPEN, Consultant Gastroenterologist
Nottingham University Hospital, Nottingham

12.10 – 12.30

Advances in feeding neonates in NNU
Ms Karen Hayes Advanced Neonatal Dietitian, Dept of Nutrition and Dietetics, Box 119
Cambridge University Hospitals NHS FT, Hills Road, Cambridge, CB2 0QQ

12.30 – 13.30
LUNCH

Poster Viewing and Sponsor Exhibition
Osborne Suite, Osborne Foyer and Great Hall Foyer

Session VII
13.30 – 14.00

Nutrition
Chair:
Dr Peter Sullivan
Consultant Paediatric Gastroenterologist, John Radcliffe Hospital, Oxford

13.30 – 14.00

Achieving Millennium Development Goal 4 targets: what will it take?
Zulfiqar A Bhutta, MB, BS, FRCPCH, FAAP, PhD
Founding Chair
Division of Women & Child Health,
The Aga Khan University,
Karachi, Pakistan

Session VIII
14.00 – 15.15

Plenary Abstract Three and Oral Poster Two

Chairs:
Dr Alastair Baker
Consultant Paediatric Hepatologist, King's College Hospital, London
and
Dr Suresh Babu
Consultant Paediatrician, Lincoln County Hospital, Lincoln

14.00 – 14.10

Pancreatitis in Children Presenting to a Tertiary Paediatric Gastrointestinal Centre
Mutalib Mohamed, Sunil B, Lindley K, Elawad, M, Fevronia K
Great Ormond Street Hospital / Institute of Child Health
Great Ormond Street
London

14.10 – 14.20

A STAT change in Inflammatory Bowel Disease? Altered signalling in intestinal T cells
E. Giles[1], J.O. Lindsay[2], I.R. Sanderson[2], T.T. MacDonald[1], A. J. Stagg[1]
[1]Centre for Immunology and Infectious Disease
[2]Centre for Digestive Diseases, Blizard Institute, Barts and the London School of Medicine and Dentistry, QMUL

14.20 – 14.30

Can anti-tissue transglutaminase antibody (tTG) levels predict mucosal inflammation in children with Coeliac Disease thus avoiding endoscopy? Experience in a single Paediatric Gastroenterology Centre
Yeop I, Shergill-Bonner R, Elawad M, Kiparissi F.
Great Ormond Street Hospital, Great Ormond Street,London.

14.30 – 14.40

Potential Impact of Revised ESPGHAN Guidelines for Diagnosis of Coeliac Disease
Abdulkarim D, Briar G, Morris M-A
Norfolk and Norwich University Hospitals, Colney Lane, Norfolk

14.40 – 14.50

Paediatric *Helicobacter pylori* practice in the United Kingdom: A BSPGHAN Survey
Goddard M[1], Lloyd C[2], Beattie RM[3], Hansen R[4].
[1]Medical Student, School of Medicine, University of Aberdeen
[2]Administrator, British Society of Paediatric Gastroenterology, Hepatology and Nutrition, Birmingham Children's Hospital
[3]President, British Society of Paediatric Gastroenterology, Hepatology and Nutrition, Southampton General Hospital
[4]Child Health, School of Medicine, University of Aberdeen

14.50 – 15.00

Suboptimal Vitamin D Status in Treated Coeliac Disease: Is Current Practice for Monitoring Bone Health Adequate?
Smith C, Hope B, Butt A
Royal Alexandra Children's Hospital, Dyke Road, Brighton, BN1 3JN

Oral poster abstracts
15.00 – 15.15

15.00 – 15.05

15.05 – 15.10

15.10 – 15.15

Session IX
15.15 – 15.55

Chair:
Dr Mark Beattie, Consultant Paediatric
Gastroenterologist, Southampton General Hospital, Southampton

Key Note Lecture

15.15 – 15.55

New Arrangements for National Speciality Commissioning for (Paediatric) Gastroenterology, Hepatology and Nutrition
Dr Edmund Jessop
Medical Advisor
National specialised commissioning team
NHS London
104 Victoria Street
London
SW1E 6QT

15.55 – 16.00
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

Future Meetings:

2013
Manchester
hosted by Dr Adrian Thomas

2014
London
hosted by Dr Alastair Baker

ABSTRACTS FOR WEDNESDAY 25TH JANUARY 2012

Invited Speakers'
Challenging Cases
Oral Plenary Session Abstracts
Challenging Cases Posters

How can we prove Reflux and aspiration?

Dr David Rawat, Consultant Paediatric Gastroenterologist, Barts and the London, Turner Street, London E1 2AD

The recognition of many of the clinical manifestations of extra-oesophageal reflux (EOR) has gained acceptance amongst our adult colleagues however the prevalence of otolaryngologic and respiratory disorders caused by GORD in paediatrics remains unknown (1, 2). Aspiration has been shown to be responsible for both acute and chronic respiratory disease. Though pharyngoaspiration is more easily and frequently detected, GORD aspiration provides an important underlying cause of respiratory disease, and therefore the confirmation of the diagnosis is essential for the treatment of many pulmonary disorders. Moreover, the current clinical practice of treating children with symptoms suggestive of EOR is increasing, despite the lack of data to support it. In part, this appears to be because currently used diagnostics for EOR often rely on testing methods and normative standards that were established for the diagnosis of classic gastro-oesophageal reflux disease (GORD), which may not be appropriate for use in diagnosing EOR disease.

There are various functional diagnostic tools available to assess oropharyngeal function and to diagnose aspiration, each with advantages and disadvantages. Currently videoflouroscopic swallow studies (VFSS) and flexible endoscopic evaluation of swallow (FEES) have been recognized as the gold standards for the detection of aspiration. Although used clinically, the limitations of these techniques are well known and relate to age, radiological exposure and the lack of measurable objective parameters. Although multiple diagnostic options exist, only nuclear scintigraphic and barium studies offer direct evidence of aspiration of gastric contents. Bronchial scintigraphy in conjunction with a meal offers greater sensitivity in detecting GOR related aspiration; the ability to perform delayed scanning, and decreased radiation exposure.

24-hour pH monitoring with double/triple probes (distal and proximal oesophageal and/or naso-pharyngeal probe) is still considered the "gold standard" for diagnosing extra-oesophageal manifestations. The advent of impedance technology has allowed the measurement of non-acid reflux and has shown a better symptom correlation with extra-oesophageal non-acid reflux (3-5). However there is ample evidence that normal amounts of reflux may also be associated with extra-oesophageal manifestations. Recent advances with high-resolution offer more promise in particular when used together with videoflouroscopy and pH/MII. Recently a novel technique, automated impedance manometry (AIM) was discovered. By combining pressure and flow measurement in one catheter, new swallow function variables can be derived. Pharyngo-oesophageal pressure-impedance profiles are derived using automated computational algorithms which then calculate objective swallow variables as a marker of deglutitive function. Alterations of these variables in relation to ineffective swallowing and to aspiration risk can thererefore by assessed using a swallow risk index (6-8). This technique has been validated in adult patients with dysphagia however recent paediatric studies have applied this technique for assessment of neurologically impaired children with suspected aspiration. Preliminary data looks promising in terms of predicting circumstances when aspiration is likely. However this novel technique is restricted to specialist centres and further multicentre studies and normative paediatric data is required.

Another alternative method to indirectly indicate the presence of gastric refluxate in the lungs is the use of biomarkers (3). Measurements of lipid laden macrophages and bile in respiratory tissue have been used in the past. Because pepsin plays a primary role in causing laryngeal and airway tissue damage and because it is a relatively large molecule, pepsin is an excellent clinical marker for EOR disease. However sensitivities and specificities of these biomarkers are not as high as expected, so new biomarkers are now being considered.

Despite our advances in the biomechanical understanding of swallow and extra-oesophaeal reflux, detection of aspiration remains challenging particularly in children with neurodisability. Further studies are required to improve the sensitivity and specificity of the methods used to establish a causal association between reflux and aspiration and to establish the best diagnostic approach for the diagnosis of GORD related aspiration.

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Finances and Developing Tariffs

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Rectal Manometry and Ultrasound: How, When and Value?

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What should we recommend for a remarkably asymptomatic 6 year old child with extensive Crohn's Disease?

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Introduction:
There is a paucity of evidence and guidance as to if, when and what immune suppression should be introduced for relatively asymptomatic children or those in remission with confirmed Crohn's Disease. We clarify this dilemma with the case of a 6 year old girl with histologically confirmed Crohn's Colitis who is remarkably well and whose parents are understandably reluctant to accept the diagnosis and agree to immune suppression at this stage.

Case:
R presented aged 2 years with some mild chronic diarrhoea that appeared to settle with the elimination of cows milk protein from her diet. She was re-referred at the age of 4 years with a confusing history more in keeping with constipation and over-flow soiling. After non-invasive investigation failed to reassure her physician and parents an upper endoscopy and colonoscopy was arranged. This was expected to be normal. The upper endoscopy was normal but despite a normal macroscopic appearance, the colonic histology was, surprisingly, suggestive of Crohn's Colitis. Explaining the likely diagnosis was challenging and the management was debated within our department. Given that R was remarkably well, we followed her parents' wishes and tried treatment with non-exclusive Modulen IBD. This was quickly stopped because the child found it unpalatable.

Follow up continued and by the age of 6 years R was showing increasing bowel frequency (up to 5 times a day) but she otherwise remained extremely well. Faecal Calprotectin was found to be significantly elevated and a subsequently arranged repeat colonoscopy showed extensive Crohn's Colitis both macroscopically and on histology. Other granulomatous disease was excluded. R's parents remain very reluctant to consider treatment with potentially toxic drugs and are currently pursuing a homeopathic remedy.

Dilemma:
Although R continues to thrive she is definitely now symptomatic. She has extensive Crohn's Colitis and her physicians are becoming increasingly anxious that she is not receiving any conventionally accepted treatment for this chronic inflammatory condition and anticipate she will soon begin to manifest complications.

This atypical presentation in a young and relatively asymptomatic child highlights a common dilemma faced by paediatric gastroenterologists. When is the right time to start immune suppression in cases of extensive Crohn's Disease in relatively well young children?

New association in systemic rotavirus illness: encephalopathy with vasculitis

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Introduction:
Rotavirus is increasingly recognised as systemic disease. Isolated associations such as encephalopathy, cutaneous vasculitis have been reported. We report a new association in systemic rotavirus illness: encephalopathy with vasculitis. 3 such cases are reported which initially provided a diagnostic challenge in light of the clinical presentation.

Aim
The aim of the case series is to make the clinicians aware that rotavirus should no longer be considered as being the commonest organism causing gastroenteritis but increasingly showing newer associations either in isolation or as a combination of previously demonstrated isolated clinical presentations.

Method:
3 children between the ages of 6 months and 3 years presented to the emergency department with an irritability/drowsiness, fever, non-blanching petechial spots (in non-SVC distribution), diarrhoea, vomiting and looking unwell.

Results:
In view of fever and non-blanching rashes all the 3 children were initially treated as suspected meningococcal disease. 2 of the children were initially acidotic which resolved within 12 hours. The blood inflammatory markers were reported within normal limits in all 3 cases. The children recovered within 48 to 72 hours and blood culture and meningococcal PCR were reported as negative. One child needed transferring to the PICU for 24 hours and all 3 suffered significant morbidity. Stool ELISA were reported as positive for rotavirus in all 3 cases.

Discussion:
- Encephalopathy has been reported as an association with rotavirus being demonstrated in the CSF
- Fluid resuscitation and replacement is necessary
- Vasculitis has been considered as a possibility in laboratory based reports and following rotavirus immunisation
- Only 11% of non-blanching rashes has been reported to be due to a meningococcal disease in previous studies
- Stool sample should always be sent if a child presents with encephalopathy, non-blanching petechial spots and gastroenteritis.
- Rotavirus vaccines have shown great success in the USA and European countries.

Conclusion:
- New dimension in systemic rotavirus illness is demonstrated which can mimic as serious bacterial pathologies such as a meningococcal sepsis
- It is important to recognise other causes of non-blanching petechial rash in children
- A new association with rotavirus is reported in the form of encephalopathy and vasculitis.
- Oral rotavirus vaccines will prevent these morbidity in systemic rotavirus illness

The challenges of transfer from paediatric to adult care post liver transplantation

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Despite improvements in the provision of transitional services between child centred and adult orientated liver transplant units, there remains an overall inconsistency in the level of medical and support services provided for young people. With no consistently applied standard of best practice for such patients, there are concerns that medical outcomes deteriorate after transition to adult services. This is particularly evident amongst vulnerable young adults who have displayed poor adherence to medication regimens and poor attendance at clinic, posing difficult ethical questions when considering retransplantation.

This case presentation describes a particularly immature young man with mild learning difficulties. He was diagnosed with biliary atresia and had a successful Kasai portoenterostomy in infancy. However he subsequently developed portal hypertension and needed banding of oesophageal varices. He finally received a liver transplant just before his 16th birthday and required addition of mycophenolate mofetil to his immunosuppression regimen to treat recurrent acute rejection in the immediate post operative period. After discharge, he initially appeared to be doing well, but within two years of his transplant he exhibited problems adhering to medication, was self harming and had been thrown out of the family home. Social services were heavily involved due to the young man's claim that he had been physically abused by his father. This culminated in an overdose of mycophenolate mofetil and tacrolimus. After extensive preparation, he was transferred to adult services at the age of 19 years. Non attendance at the adult clinic soon became an issue and sirolimus was added to his immunosuppression regimen to help salvage his deteriorating liver function. After much debate he was listed for a second liver transplant aged 20 years. Sadly he deteriorated within a month due to terminal graft failure and died soon after.

Non adherence to medication in adolescents with any chronic disease is well recognised and in some adherence improves as the young person matures. How do we encourage this?
Adult transplant patients who are non adherent usually will not be offered a re transplant, should we treat adolescents differently?

Coeliac disease: To biopsy or not – NICE v ESPGHAN

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2012 heralds a controversy of potentially Olympic proportions for Paediatric Gastroenterologists. This year sees the introduction of new ESPGHAN guidelines for the diagnosis of coeliac disease, in which – for the first time since small intestinal biopsy became feasible, diagnosis of this life-long disorder may be made on blood testing alone.

Successive ESPGHAN position statements have reduced the number of recommended diagnostic biopsies from three in 1970[1] to one in 1990[2] and now potentially to zero[3], albeit with strict criteria. Firstly the child or adolescent must be symptomatic and not simply screened on the basis of being genetically at risk. Secondly the titre of the specific blood test (IgA anti-TG2) must be 10 times the upper limit of normal for that assay system. Thirdly a repeated blood test is required, which must confirm both a positive IgA endomyseal antibody (EMA) and that the child is of either HLA-DQ2 or DQ8 tissue type. Such an approach is not mandatory, but can be offered by the paediatric gastroenterologist to the family in lieu of endoscopy if a high titre TG2 antibody is identified on initial testing.

These criteria are founded on changes in the conceptual framework in which coeliac disease is now considered, no longer just as a small intestinal enteropathy induced by gluten, but rather a systemic disease with chronic immune symptoms affecting a number of organ systems. They are also based on recognition of the imperfect data provided by duodenal biopsies, in which histological features may be non-specific or even limited to the duodenal bulb (such children would have tested negative in the days of capsule biopsies).

There are, of course, potential advantages and disadvantages of such an approach. In the ideal world, where guidelines are followed scrupulously and corners are not cut, no-one will be diagnosed on a single blood test and commenced on a gluten-free diet by a practitioner without adequate specific expertise. In the real world, this scenario may indeed occur, if the message is taken that biopsies are “no longer needed”. Leaving financial implications to one side (again, an ideal world position), there are potential advantages for the child in both avoiding an endoscopy and also potentially achieving greater diagnostic speed and specificity. Potential concerns include ensuring access to diagnostic modalities such as EMA and HLA typing and maintaining adequate quality control for serological testing.

NICE guidelines currently recommend small intestinal biopsy, in line with the previous ESPGHAN policy. These are being reviewed in 2012, and may be modified in the light of the new ESPGHAN recommendations. However the costing implications of such a change are likely to play a large role in their final decision. For the moment, the ability of the paediatric gastroenterologist to follow the new ESPGHAN guidelines may vary with locality.

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2 year experience of a catheter free, radio telemetric, oesophageal pH monitoring (Bravo ®) system in children at a single U.K. tertiary centre

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Introduction:
With the increased understanding of reflux disease exacerbating respiratory, ENT and even behavioural conditions, introducing a catheterless form of oesophageal ph monitoring to a younger or more challenging child has now become possible. The BRAVO pH system is a significant advancement in the evaluation of patients with gastroesophageal reflux because of better tolerability and the ability to record data over a 48-hour period.

Aim:
To evaluate performance, tolerability, safety and day-to-day variability in acid reflux patterns in using the BRAVO pH system.

Method:
Retrospective review of existing database and case notes of all children that underwent BRAVO pH capsule placement from June 2009 to August 2011 was done. All the capsules were deployed 4- 6 cm proximal to the Z-line of the oesophagus under the influence of general anaesthesia and under endoscopic vision. The data from the capsules was recorded by radio telemetry onto the pH boxes. This was subsequently uploaded onto a computer designated for this purpouse. The paired t test was used to analyze the pH values from 2 sub groups (group 1 with 24 hr recordings and group 2 with 48 hr recordings).

Results:
203 consecutive patients (122 male) of median age 9 (2-18 yrs) had the procedure. The youngest child weighed 9.29kg. Successful pH data over 24 hours was obtained in 93.6% of patients and over 48 hrs in 59% of patients. Commonest indications were reflux symptoms (45%), abdominal pain (33%) and regurgitation (19.5%). In 12% of our patients with an underlying behaviour disorder the wireless pH study was preferred over a standard naso-oesophageal probe for compliance reasons. The other common indication in our centre was assessment pre and post fundoplication with 7 of the Bravo pH study results indicating need for surgical management (3 laparoscopic and 4 endoscopic full thickness fundoplications). Nearly all studies (86%) were performed off acid-suppressing medications.

A failure rate of 6.4% (13/203) was noted. Of the 13, 4 failed to detach from the introducer to the oesophageal mucosa. 8 capsules detached within 1 hour of deployment and one capsule failed due to technical problems with the recording box

There was no statistical difference between the pH-measurements of the first 24 hours with the 48-hour measurements (p=0.56).

No adverse events were noted in any of our patients.

Conclusion:
The BRAVO pH system is a safe and effective method of recording oesophageal acid exposure. It is an acceptable alternative for children who are particularly unwilling or unable to tolerate nasopharyngeal pH catheter. Our results support the use of pH measurement for a period of 24 hours only.

Effectiveness of double-balloon enteroscopy facilitated polypectomy in children with Peutz-Jeghers syndrome

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Introduction:
Large small bowel (SB) polyps in Peutz-Jeghers syndrome (PJS) pose a high risk for intussusception, often necessitating laparotomy with intraoperative enteroscopy (IOE) for polyp clearance. This prospective series examines the effectiveness of double-balloon enteroscopy (DBE) facilitated polypectomy as an alternative therapeutic option for paediatric patients with PJS.

Methods:
Data on all patients with PJS referred for DBE facilitated SB polypectomy at two tertiary referral centers in the UK, were prospectively collected from February 2005 to April 2011.

Results:
12 pediatric patients with PJS (7 males, mean age 11.3; range 0.9-16 years) were referred for DBE facilitated SB polypectomy. In total, 20 DBEs (including 2 laparoscopically assisted DBE (Lap-DBE)) were performed. Significant polyps (≥1.5cm) were found in 11 patients. Successful clearance of SB polyps by DBE or Lap-DBE was achieved in 11/12 (92%) patients; with one requiring laparotomy with IOE. One patient suffered a post- Lap-DBE pelvic abscess from an infected port-wound. No other complications ensued and 10/12 children that underwent successful DBE facilitated polypectomy remained symptom and intervention free throughout follow-up (median 24 months). 2 children were lost to follow up.

Conclusion:
This series demonstrates that DBE facilitated polypectomy is an effective alternative to laparotomy with IOE in selected paediatric patients with PJS. DBE offers a less invasive approach which may reduce the need for surgery and associated morbidity and should be considered as an alternative therapeutic option.

Faecal calprotectin for the diagnosis of paediatric inflammatory bowel disease: a meta-analysis.

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Introduction:
Faecal calprotectin (FC) is increasingly used as a marker of intestinal inflammation, especially in diagnosis of inflammatory bowel disease (IBD). A previous meta-analysis (Rheenen et al. BMJ 2010) evaluated adult and paediatric studies to October 2009 and concluded that FC has good discriminative power to safely exclude IBD, but that this power was greater in adults than in children. However, the seven paediatric studies only included a total of 226 paediatric IBD (PIBD) patients, therefore small study effect may have existed.

Aim:
We aimed to determine the diagnostic utility of FC at presentation in children with suspected IBD by evaluating all the available literature.

Method:
A search was performed with keywords relating to IBD and calprotectin in several electronic resources from 1966 to November 2011. A hand search of articles was also performed, drawn from reference lists and meeting abstracts. Inclusion criteria were studies that reported FC levels prior to the endoscopic investigation of IBD in children less than 18 years old. There was no English language restriction. Each study was evaluated using the QUADAS tool and a meta-analysis of all included studies was performed using RevMan (v 5.1) and HSROC (R package). Pooled sensitivity and specificity was generated using a random effects model.

Results:
A total of 73 papers were identified during the initial search. All were reviewed but only 8 met the inclusion criteria (6 prospective and 2 retrospective case-control studies). Two studies in the original BMJ meta-analysis (Bunn 2001, Kolho 2006) were excluded as both the IBD and control groups did not represent children undergoing primary investigation for suspected IBD. In total the studies presented FC levels at initial presentation in 394 IBD patients and 353 non-IBD controls. Pooled specificity and sensitivity for the diagnostic utility of FC during the investigation of suspected PIBD were 0.998 (95% CI 0.960-1.000) and 0.665 (95% CI 0.537-0.820) respectively.

Conclusions:
FC has a high specificity but a modest specificity for the diagnosis of IBD in the paediatric population. The inclusion of two new larger studies and removal of two studies from the original meta-analysis led to an increase in the pooled sensitivity and reduction in pooled specificity. Furthermore, it is evident from other studies that use of a higher normal range (i.e. >200ug/g) would significantly increase this specificity and therefore further work is required that report multiple cut off levels to truly appreciate the value of this test.

A risk score for intestinal failure in gastroschisis

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Introduction:
Gastrointestinal functional outcome is extremely variable in infants with gastroschsis. Surgical management with primary closure, or silo with delayed primary closure, enables the majority to attain full enteral feeding. However, some infants have difficulties establishing enteral feeding, and require long-term parenteral nutrition (PN) with a resultant prolonged hospital stay.

Aim:
The aim of this study was to audit the nutritional outcome of infants with gastroschisis managed in a regional neonatal surgical centre, to determine predisposing risk factors predictive for intestinal failure and develop a Gastroschisis Intestinal Failure Risk Score (GIFS).

Method:
Risk factors for poor outcome were identified by a literature review. Data was collected from the Neonatal Network electronic database and case notes of 30 patients born between 1st Jan 2010 and 31st March 2011, managed in a single centre (15 were male). Outcome measures were the duration of PN and hospital stay, and analysis was with Spearman's correlation and t-tests. P-values <0.05 were considered statistically significant. GIFS was developed using the three most significantly associated factors, repeat or multiple surgery (scores 0-3), reduced bowel length (scores 0-2), and complications (scores 0-1), with a maximum score of 6.

Results:
The median duration of PN was 3 weeks (range 1 to over 52 weeks), and eleven (37%) required PN for more than 28 days. Seven (23%) required more than 3 months of PN, and three (10%) infants were discharged with home PN (two continue). There was no mortality. Gestation, timing of diagnosis (antenatal/postnatal), presence of antenatal bowel dilatation, mode of delivery, condition at birth, birth weight centile, time to admission to the referral unit, type of initial closure and co-morbidity was not associated with prolonged PN. Factors associated with prolonged PN were maternal age under 19 years (t-test, p=0.04), gastrointestinal complications (atresia, perforation, dehiscence, adhesions, necrotising enterocolitis) (t-test p=0.01), reduced bowel length (t-test, p=0.03) and abdominal operations (Spearman's rho 0.61, p<0.01). GIFS >1 identified 10 of 11 of cases requiring PN for more than 28 days. All 19 cases requiring PN for less than 28 days scored 0 or 1.

Summary:
Prolonged requirement for PN in gastroschisis was associated with younger maternal age, presence of intestinal atresia, post-closure complications, bowel resection/loss and multiple operative procedures. A simple scoring system provides a clinically useful predictor of intestinal failure (GIFS>1).

Conclusion:
Identification of predictive factors for prolonged enteral feed intolerance allows targeted interventions to encourage gastrointestinal autonomy or to initiate home PN training at an earlier stage. Prospective evaluation of GIFS is planned

The epidemiology and natural history of paediatric inflammatory bowel disease in a UK region: a prospective 14-year study in SE Scotland.

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Introduction:
An accurate understanding of trends in incidence and prevalence as well as disease burden of paediatric-onset inflammatory bowel disease (PIBD) within a large region are essential to plan services and unravel possible aetiological factors.

Aim:
We aimed to delineate these trends together with numbers of PIBD cases requiring surgery and immunomodultory/biological therapies.

Methods:
All incident and prevalent cases of PIBD in South-East Scotland were prospectively collected from service establishment in August 1997 to October 2011. All cases of Crohn's disease (CD), ulcerative colitis (UC) and IBD-unclassified (IBDU) were diagnosed by standard criteria and detailed demographic information and both medical and surgical therapies were recorded on a departmental database. Date of first attendance and of discharge from paediatric services was also recorded, allowing calculation of incidence and yearly point prevalence (1st April). To allow for accrual of cases during the establishment phase of the PIBD service, only data from 2000-2010 was used for incident and prevalent rates. Rates were sex-adjusted and statistical analysis carried out using GraphPad Prism and R.

Results:
A total of 318 children with IBD were cared for during the 14 year period. The cohort comprised of 206 CD (65%), 73 UC (23%) and 39 IBDU (12%) cases. The median age at diagnosis was 11.6yrs (IQR 9.1-13.4yrs) with an overall preponderance of males. Overall incidence of IBD in the period 2000-2010 was 6.4/100,000/yr with a significant rise from 5.6/100,000/yr to 7.2/100,000/yr between the periods of 2000-2004 and 2005-2010 (p=0.002). This was mainly driven by an increase in the incidence of CD in males from the earlier (4.0/100,000/yr) to the later (5.5/100,000/yr) epoch (p=0.002). The overall rates of UC (1.5/100,000/yr) and IBDU (0.9/100,000/yr) remained relatively stable. The point prevalence of IBD rose from 23.1/100,000 (2000-2004) to 48.7/100,000 (2005-2010). During the total follow up time of 1432 patient years, 68% of patients required azathioprine, 27% methotrexate, 18% biologicals and 21% underwent IBD-related surgery.

Conclusion:
Evaluating a well defined paediatric population over a 14 year period gives a clear indication of the rising trend in the incidence and prevalence of PIBD. In addition, knowledge of the requirement for medical and surgical therapies allows for the tailoring of local service provision.

Comparison between oral (liquid and modigraf) and intravenous induction with tacrolimus in intestinal transplantation patients

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Introduction:
Achieving optimal tacrolimus level (TL), either by oral liquid (o-t), IV route (IV-t) or Modigraf (m-t) early in post intestinal transplantation (Itx) is ideal to prevent acute rejection

Aim:
To report on single centre experience of tacrolimus induction with different forms of tacrolimus formulations in intestinal transplant patients

Method:
Records of three groups were retrieved from a prospectively maintained database. Group-1 was started on o-t dose of 0.075 mg/kg/dose; this was changed to IV-t 24-hour infusions of 0.15 mg/kg/day if low levels persisted. Group-2 was started on IV-t 24-hour-infusion of 0.05 mg/kg/day. Group 3 was started on modigraf (tacrolimus granules) at 0.3 mg/kg/day. Target TL (measured once daily) for all the groups were aimed at 15-20 ng/ml in the 1st 3 weeks, then 12-15 ng/ml till third month

Results:

Median (range)	Group 1 (n=10)	Group 2 (n=11)	Group 3 (n=7)
Age in months	24 (12 – 54)	40 (15.7 – 169.4)	38 (17 – 98)
Gender (M:F)	4:6	8:3	5:2
Days of IV-t in 1st 30 days	18 (1-26)	16.3 (4-30)	0
Median TL of each patient 1st 30 days in ng/l	18.1	8.5	17.0
Rejection episodes while on IV-t	3 (mild to moderate)	2 (severe)	Not applicable
Rejection episodes in the 1st 30 days	70% (7/10)	73% (8/11)	28% (2/7)
Post-transplant lympho-proliferative disease (PTLD) episodes (1st 6 months)	0	5	2

TL was in target range in 24%, 28% and 28% of days in groups 1, 2 and 3 respectively. Most of group 2 (IV-t) patients TL were higher than target range (47% of days) while most of group 1 (o-t) and group 3 (m-t) patients TL were lower than target range (42% and 38% of days respectively), despite frequent dose modifications (50% of days group 1, 60% of days group 2 and 42% of days group 3). 9/10 patients in group-1 were switched to IV-t on the 5th day (3-29). Group-2 patients were converted from IV-t to o-t within 5 days (0-14) while IV-t was not needed in group-3 patients. Higher incidence of PTLD and opportunistic infections were noted in children receiving IV-t at induction. Lesser incidence of episodes and severity of rejection were evident in the group 3 patients.

Conclusion:
Modigraf granules usage was associated with fewer, milder episodes of acute rejection in immediate ITx period and fewer incidences of PTLD in the first 6 months following ITx. Modigraf granules are recommended for tacrolimus induction in ITx. In addition, amongst solid organ transplant recipients who are having difficulty achieving TL, modigraf can be considered

National IBD Audit

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A Rare case of gastric outlet obstruction

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Aim:
To describe a rare case with an unusual rash and vomiting

Method:
Descriptive case study
Case history: This is a 6 year old boy who was referred to a tertiary Paediatric gastroenterology unit with a 1 year history of lethargy, 6 month history of vomiting and weight loss. He vomited once every day or alternate days. The vomitus was usually large containing partially digested food materials. He also had foul smelling belches.He had been seen by the dermatologists for 6 months for an unusual hyperpigmented rash on his hands and feet which was initially thought to be possibly psoriasis or eczema related dermatopathy and was treated with steroids. Subsequently it was thought to be pellagra due to his severe malnutrition and weight loss. He was born at term with no significant past medical history of note.On examination he was undernourished, with hyperpigmented rash on both hands and feet . abdomen examination was unremarkable.
Investigations : Barium meal showed delayed gastric emptying, but no evidence of obstruction or malrotation. His bloods showed ESR 24 mm/hr and albumin 33g/L. He had a gastroscopy done that showed ulceration of the pylorus with stricture and difficulty in intubating the duodenum. Histology showed oesophagitis, gastritis with duodenitis but no granulomas. It was thought that he had probably got an inflammatory mass in the proximal duodenum. He was tolerating some NG feeds and subsequently started on parenteral nutrition. A Nasojejunal tube failed to be passed endoscopically due to the pyloric stricture. MRI small bowel showed bowel wall thickening in terminal ileum. He was treated with iv steroids . A week later his symptoms improved .Subsequent colonoscopy that showed IleoCaecal Crohn's disease. He was then started on Modulen feeds.

Conclusion:
This was an unusual case of Crohn's disease who presented with incomplete gastric obstruction and pyloric stenosis that can be very challenging to manage.

A Rare Presentation of E.Coli 0157 Ischaemic Colitis in a Teenager

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Introduction:
E.coli 0157 is an enterohaemorrhagic gram negative rod commonly associated with HUS and TTP in children. It is as a causative pathogen in haemorrhagic colitis and colonic complications include necrosis, pseudomembrane formation, or perforation. However these complications present with bloody diarrhoea. We present a case of E.coli 0157 which did not present with PR bleeding causing an atypical colitis.

Aim:
To disseminate knowledge on a rare clinical presentation and discuss a challenging diagnosis.

Subject and eventual results
A 16 year old girl who is normally fit and well presented to A&E with a two day history of right iliac fossa pain, diarrhoea and vomiting. She had nil blood PR and nil urinary symptoms. Her βHCG and triple vaginal swabs were negative. On examination she was tender in the right iliac fossa. Her blood results showed a raised CRP of 153 mg/l, WBC of 6.69 x 109/l and Hb of 10.9 g/dl. C.Difficile toxin results were negative. An ultrasound scan showed a small amount of fluid in the pouch of douglas. A clinical diagnosis of appendicitis was made and she underwent a laparoscopic appendicectomy which found a macroscopically normal appendix confirmed by histology results later however the caecum had patchy inflammation with areas of early necrosis. An abdominal drain was left in situ and she was commenced on intravenous cefuroxime and metronidazole.

Post operatively on day two she continued to have abdominal pain, distension and vomiting. In addition she was septic with pyrexia, tachycardia, hypotension and a low urinary output. On examination there was guarding and large bilious nasogastric aspirates. Her abdominal drain was draining green coloured serous fluid. An arterial blood gas showed a raised lactate of 2.6 and a pH of 7.28. She underwent a plain abdominal xray which showed possible dilated small bowel loops and a small pneumoperitoneum. The working diagnosis at this stage was inflammatory bowel disease, most probably Crohns, causing bowel perforation.

She was transferred to a tertiary paediatric centre on day three due to anticipated technically difficult surgery where she was admitted in PICU and resuscitated before she underwent a diagnostic and therapeutic laparotomy. At laparotomy the patient had gross contamination of the peritoneum with faeces. There were two perforations at the proximal caecum and one perforation at the proximal transverse colon. The ascending colon was oedematous and thickened however the appendix stump was intact. A right hemicolectomy with a defunctioning ileostomy was performed. The patient was discharged day thirteen post laparotomy. The resected colon biopsy showed inflammation, ulceration, abscess formation and a chronic inflammatory cell infiltrate with scant focal cypt abscess formation. It concluded that there were several features of Crohns which were regarded as unusual, in part inflammation was predominantly large intestine rather than terminal ileum, there was no granulomatous component and cryptitits was not a conspicuous component. It was felt that it was prudent to explore and an infective cause of inflammation. The patient had a negative vasculitic screen, yersinia serology, campylobacter screen, and c.difficile screen yet had positive e.coli 0517 antibodies.

Conclusion:
To our knowledge this is a rare presentation of E.coli causing colits and perforation in the absence of HUS or TTP and excluded C.difficile infection. There has been one case described by Uc et al similar to this however in that case the child presented with PR bleeding and the histological diagnosis was more definitive. This case highlights an unusual histological presentation of E.coli colitis compared to data in current literature and draws attention to the fact that a clinical diagnosis of appendicitis is often difficult.

Acute upper gastrointestinal haemorrhage in a 15 year old patient.

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Introduction:
A 15 year old female patient was admitted with a 3 day history of exertion dyspnoea, general malaise and melaena. There was no history of retching, ethanol excess or recent NSAID or aspirin therapy. There was no significant family history and the only personal history was well-controlled asthma. Examination revealed pallor but no stigmata of disease.

Investigations:
The only abnormal laboratory investigation was a microcytic anaemia (Hb 5.5g/dL and MCV 62fL) and evidence of iron deficiency (ferritin 3µg/L). The patient was resuscitated initially with colloid and red blood cell transfusion.

Management:
An emergency gastroscopy was undertaken. This revealed four columns of grade 2 oesophageal varices with stigmata of recent haemorrhage, but no other stigmata of portal hypertension. Following variceal band ligation the patient was treated with antibacterial prophylaxis and terlipressin.

Further investigation:
A full chronic liver disease screen, including viral hepatitis serology, iron, β1-antitrypsin and copper studies, and hepatic autoimmune profile were normal. Abdominal ultrasound revealed a liver with normal parenchyma, normal spleen and no evidence of intra-abdominal varices or portal hypertension. A chest radiograph revealed mediastinal shift to the left with a hyper-expanded right lung and enlarged right main pulmonary artery. Echocardiography revealed right ventricular wall hypertrophy and pulmonary hypertension (RVP 35mmHg).

Ongoing therapy:
The initial bleeding was successfully treated with variceal band ligation and vasopressin therapy. Further outpatient endoscopic banding procedures were undertaken to achieve complete variceal eradication.

Outcome:
Computed tomography scanning of the chest and abdomen with contrast, followed by cardiac MRI were undertaken. Select images from these investigations will be displayed before the final diagnosis is presented and discussed.

Chylomicron Retention Disease… And What Else?

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Introduction:
DX is a 3 year old boy born at term after a normal pregnancy to Pakistani parents who are first cousins. A maternal uncle died aged 2 years in Pakistan due to chronic intestinal problems. DX was breastfed for 3 weeks and then bottle feed. He presented to hospital at 3 months of age with 3 to 4 weeks history of crying on feeding, reduced feeding, lethargy, floppiness and marked weight loss. He was commenced on nasogastric feeding, but developed vomiting, abdominal distension and diarrhoea. He did not tolerate hydrolysed feeds and was started on total parenteral nutrition (TPN). Symptoms settled on gut rest and full TPN.

He was found to have severe deficiency of fat soluble vitamins, metabolic acidosis and positive stool fat globules. Creatine kinase was normal. Lipid profile showed low cholesterol and normal triglyceride. He had a negative sweat test and had normal lipoproteins.

He underwent an endoscopy at age 4 months and histology showed fat laden enterocytes. A diagnosis of Chylomicron Retention Disease was made.

He was started on nasogastric monogen feeds, high dose Vitamin E, A and D. However, his diarrhoea and acidosis resumed when he was nearly on full feeds. He had to go back on TPN and thrived. He had a few line related sepsis.

At re-endoscopy after a 30ml monogen feed at 1 year of age, there was no obvious macroscopic abnormalities seen. Histology showed villous atrophy with some enterocytes showing both vacuolated and degenerate, detached enterocytes. Electron microscopy showed lipid droplets free in the cytoplasm and other lipid structures, which were consistent with Chylomicron Retention Disease.

Another attempt at taking him off TPN and increasing his low fat feeds failed. Genetic studies showed normal male karyotype. SAR1B gene is negative.

Currently, he is on TPN and around 300mls a day of monogen feeds. He eats tiny amounts of finger foods and has 5 to 6 runny Type 7 stools. This increases with food intake. His current clinical status is stable and he is thriving.

Discussion:
This case presents a diagnostic challenge. Current clinical picture and investigation support Chylomicron Retention Disease, with electron microscopy findings and triglyceride profile being characteristic. However, his symptoms recur when his feeds are increased, he remains TPN dependant and his SAR1B gene is negative. Is this a variant of chylomicron retention disease or is this something else?

Colitis in a child with Cystic Fibrosis.

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Introduction:
Children rarely suffer from pseudomembranous colitis although they often receive antimicrobials and some have predisposing factors. We report a boy with cystic fibrosis (CF) receiving Azithromycin prophylaxis who developed Clostridium difficile toxin positive pseudomembranous colitis.

Case Report:
A 13 year old boy with CF and atypically good respiratory function presented with a week long history of abdominal pain, weight loss and increasing abdominal tenderness. As a neonate he underwent a small bowel resection and right hemicolectomy due to jejunal atresia and micro-colon respectively. Subsequently he suffered multiple episodes of distal intestinal obstruction syndrome (DIOS), often requiring and responding to gastrografin treatment. On this admission he was receiving Azithromycin prophylaxis, Ursodeoxycholic acid, Creon, vitamins and DNase nebulisers.

Initial investigations revealed a significant neutrophilia and raised CRP. Confusingly, an abdominal ultrasound suggested a severe pancolitis. The differential diagnosis included DIOS, adhesive obstruction with bowel ischaemia or an infective colitis. Intravenous fluids and Metronidazole were commenced. He subsequently developed a temperature and diarrhoea. After a careful multidisciplinary discussion an urgent limited colonoscopy revealed oedematous and inflamed mucosa with raised white nodules typical of pseudomembranous colitis. Oral Vancomycin was added. Clostridia difficile toxin was found positive in his stools. He recovered and Azithromycin prophylaxis was discontinued.

Conclusion:
DIOS is quite commonly seen in CF and this unusual presentation led to a diagnostic dilemma which was clarified by colonoscopy. We present the literature that confirms pseudomembranous colitis is rare in children and has not often been reported in children receiving Azithromycin prophylaxis.

Diffuse Primary intestinal lymphangiectasia in a toddler: successfully treated by Double Balloon Enteroscopy and segmental resections of small bowel

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Primary intestinal lymphangiectasia (PIL) is a rare cause of fat-soluble vitamin deficiency and protein-losing enteropathy. The often diffuse nature of lymphatic abnormality within the small bowel makes the management challenging.

We report a case of diffuse PIL presenting with abdominal pain, voluminous diarrhoea, hypoalbuminaemia, and hypogammaglobulinaemia in a previously healthy infant. Conventional upper and lower gastrointestinal endoscopic evaluations were inconclusive. At our institution, initial presumptive diagnosis was achieved with wireless capsule endoscopy (WCE). Medical treatment with low-fat diet associated with supplementary medium chain triglycerides was unsuccessful. Therefore, at the age of two, the boy underwent double balloon enteroscopy (DBE), which confirmed diffuse areas of lymphangiectasia in the jejunum and in the ileum. The areas were defined with dye injection during DBE. Tattooing of the bowel allowed for laparoscopic segmental resection of the two most macroscopically affected ileal segments and end-to-end anastomoses were carried out. Histology showed lymphatic dilation in the mucosa of jejunum (biopsies) and ileum (biopsies and resected segments) confirming the diagnosis of PIL. At 1-year follow up the child was asymptomatic and on a normal diet. To the best of our knowledge, this case of diffuse PIL is only the fifth report in the English literature describing a successful response to segmental resection.

Do you think it's zinc? A new variant of acrodermatitis enteropathica (AE): what is the pathogenesis and how to manage?

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Patients and Methods
Two sisters presented at 2 years old and 9 months old with symptoms and signs of zinc deficiency including perioral and perineal eczematous rash, frequent infections, low mood and dry, sparse hair and slow growth but no GI symptoms. Serum zinc concentrations were 9.2 and 9.1µmol/l (11-24). Neither had the genetic defect of gene SLC39A4 on 8q24.3 diagnostic of AE. Because of the phenotypic resemblance to AE they were commenced on zinc supplements with complete disappearance of their clinical zinc deficiency and improved growth. Their maternal grandfather, who lives in Holland, was reported as having sparse hair and depression. His serum zinc was 9.5µmol/l and his symptoms also improved with zinc supplements. Parent's zinc concentrations were normal.

Both children remain well after 8 years on zinc supplements, currently 150mg and 120mg of zinc citrate and gluconate respecively. Zinc concentrations are monitored to avoid toxicity. The median zinc concentrations were 15.7 (n=14) and 16.5 (n=15) respectively; both children have had two zinc measurements above the normal range. Whenever their serum zinc concentrations fall into the lower end normal range they develop minor symptoms including slow weight gain, anorexia and facial and perineal eczema.

Discussion
These children appear to have increased requirements for zinc, and developed florid symptoms of zinc deficiency at concentrations which should be asymptomatic. AE has been excluded by genetic studies. Without a definitive diagnosis the prognosis is uncertain.

Questions which remain include:
What is the basis of their increased zinc requirements?
How can their zinc status best be monitored?
Could their excess zinc intake in the face of normal serum concentrations be harmful?

EBV induced erythrophagocytosis in a Crohn's patient on immunosuppressant

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Introduction:
13 year old boy with Crohn's disease
Previous hemicolectomy for stricture and loop ileostomy - revised
On azathioprine 75 mg OD (TPMT carrier)
PN dependent, functional short bowel

Presentation:
Admitted with high grade fever, maculopapular rash, deranged liver function and new onset hepatospenomegaly.
Developed pancytopenia and coagulopathy in hospital. Daily vitamin K corrected APTR.
Treated with empirical antibiotics for 7 day. Azathioprine was stopped on admission.
He was discussed and monitored regularly by regional liver unit.
Rash settled (thought to be reaction to tazocin) on the ward and clinically improved with resolution of hepatospenomegaly over 2 week period.

Investigation:
Pancytopenia (Neutrophils 0.6), AST 316, bilirubin 161 (conjugated 93), ALT 93, Ferritin 578, GGT 166, Haptoglobulin 0.88, APTR 1.61 (0.8-1.2 normal range), CRP 49, triglycerides 2.4, LDH 1687
EBV PCR – 3.16 x 10˜3 copies/ml
Negative CMV, hepatitis B and C and parvovirus
Blood cultures x 2 negative, Normal chest XR and abdominal USS
Bone marrow aspirate - cellular with all haemopoietic cell lines represented. Increased macrophages, some of which showed erythrophagocytosis. No phagocytosis of neutrophils or magakaryocytes seen.
Granule release assay - negative

Conclusion:
1. This patient presented with febrile illness and rash with background of immunosuppression related to treatment of Crohn's disease.
2. He was noted to have recent EBV infection. He clinically improved over 2 weeks.
3. He did not completely fulfil diagnostic criteria for hemophagocytic lymphohistiocytosis (HLH) syndrome
4. This case illustrates possible EBV induced immune reaction in immuno-compromised host. It is important to consider it clinically as delay in treatment (if necessary) can be fatal.

Gastrointestinal Cytomegalovirus Infection Complicating Crohn's Disease

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Introduction:
Cytomegalovirus (CMV) is well established as a cause of gastrointestinal tract disease in immunodeficient patients, particularly in those with acquired immune deficiency syndrome, or in transplant recipients. The possible association of CMV infection with a flare up of inflammatory bowel disease (IBD) in children is uncommon but well known. In adults many cases are severe, antiviral treatment is not always effective and colectomy may be required. CMV colitis in IBD patients produces symptoms, mimicking a flare up of the disease.

Report
We report a 13-year-old girl who presented with a 6 week history of bloody diarrhoea and left upper quadrant pain. Blood inflammatory markers were raised and amylase levels (865) were elevated. She underwent a blood transfusion for anaemia. Abdominal ultrasound and small bowel series were unremarkable apart from slight splenomegaly. Nasal swab was positive for Respiratory Syncytial Virus. Endoscopy & ileocolonoscopy revealed chronic duodenitis, mild ileal inflammation & active pancolitis consistent with crohn's disease. She started on a course of steroids and pancreatitis was managed conservatively. She presented with raised transaminases a month later & MRCP & liver biopsy didn't show signs of sclerosing cholangitis, pancreatic abnormalities or liver disease. Blood CMV IgM at that stage was negative. Endoscopy & ileocolonoscopy revealed chronic gastritis & nuclear atypia (sigmoid) with pericryptal granulomas in ileum & chronic active colitis consistent with crohn's disease. She was started on mercaptopurine in view of her active disease present whilst weaning steroids.

14 months later she presented with vomiting, abdominal pain, weight loss and loose stools. Blood inflammatory markers were markedly elevated and she was anaemic. Blood CMV IgG was positive. Blood CMV polymerase chain reaction (PCR) was 299 copies/ml & faeces CMV PCR (975 copies/ml) was positive. Upper GI endoscopy revealed patchy gastritis. Colonoscopy revealed patchy inflammation and cobblestoning in the distal colon. Histology revealed nuclear atypia (sigmoid), chronic active inflammation in distal colon, crypt abscess formation and cryptitis, but none of her biopsies showed inclusion bodies and immunohistochemistry was negative for CMV. However CMV PCR was positive in all gastric and colonic biopsies (740,000 copies/ml in sigmoid and 26,400 copies/ml in descending colon). She was treated with oral steroids and IV ganciclovir.

Her symptoms improved quickly after starting the treatment and her further blood CMV PCR was negative. Mercaptopurine was continued but subsequently stopped because of high mercaptopurine metabolite levels and side effects suggestive of mercaptopurine toxicity.

Discussion
In our case, CMV infection was diagnosed on mucosal biopsies with positive CMV PCR on biopsies. The actual gold standard for CMV infection complicating IBD is CMV PCR positive on biopsies with or without the presence of CMV IgM antibodies and/or CMV antigenaemia in blood. There were no Inclusion bodies on haematoxylin/eosin staining of biopsies in our patient.

Conclusion:
Clostridium difficile, Salmonella, Shigella, Campylobacter jejuni, are well established infectious causes of IBD exacerbations. CMV infection should also be considered when symptoms of IBD do not respond to steroid therapy. A common underlying factor is the use of immunosuppressive therapy, particularly high doses of corticosteroids. In our case this could also be due to mercaptopurine. In conclusion, CMV infection in children with IBD can occur with severe pancolitis, usually resistant to steroids. CMV colitis should be considered in children presenting with relapse of colitic symptoms. Systematic early detection and treatment seem to improve the prognosis and could avoid surgery, as reported in the adult literature.

Idiopathic small bowel diaphragm disease identified by laparoscopic assisted double balloon enteroscopy in a child: an integrated successful definitive therapeutic method.

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Small bowel diaphragm disease (SBDD) is a rare complication in adults of small bowel enteropathy usually secondary to the use of non-steroidal anti-inflammatory drugs (NSAID).

The main clinical manifestations are gastrointestinal (GI) bleeding and sub-acute obstruction. We present a case of a 5-year old girl with a three year history of recurrent abdominal pain and persistent microcytic hypochromic anaemia despite iron supplementation. The conventional laboratory screening tests and upper and lower GI endoscopies were normal. Wireless capsule endoscopy (WCE) examination revealed multiple narrow strictures throughout the ileum. Laparoscopic assisted trans-oral Double Balloon Enteroscopy (DBE) confirmed the presence of several narrow, diaphragm-like strictures in the ileum. The most distal lesion reached was tattooed with methylene blue via DBE. Mini-laparotomy allowed resection of the involved segment of ileum: the clinical features suggested SBDD, but histology revealed just non-specific chronic inflammatory changes. Medical treatment failed to control symptoms related to assumed residual disease in the terminal ileum. Eight weeks later, laparoscopic assisted on table enteroscopy confirmed the presence of further diaphragms in the distal ileum. The narrowest lesions were resected with endoscopic sphincterotome (Tapertome©), while the remaining lesions were dilated. Histology of diaphragm's biopsies showed an evolved picture characterized now by fibrotic and hamartomatous features typical of SBDD.

At 6 months follow-up, the child remains asymptomatic, on a normal diet and not requiring any treatment. To our knowledge this is the first case of idiopathic (no history of NSAID use) SBDD in the paediatric age group. This report describes an integrated successful definitive therapeutic method of Laparoscopy assisted DBE for small bowel pathology.

Primary or secondary lymphangiectasia- can you believe the cardiac echo?

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Introduction:
Constrictive pericarditis is an uncommon disease in childhood and very rare under the age of 10 years. We present a case of 6 year old boy who developed intestinal lymphangiectasia secondary to unrecognised constrictive pericarditis.

Case:
A 6 year old boy presented with a 3 month history of progressive peripheral oedema, abdominal distension and lethargy following a prodromal illness that caused a prolonged cough. He had no diarrhoea. On examination, he had hepatomegaly, ascites, peripheral oedema and bilateral pleural effusions. Tests revealed low albumin and lymphopenia. Duodenal biopsy was normal but video capsule endoscopy revealed lymphangiectasia. He was commenced on parenteral nutrition in view of hypomagnesaemia and inability to maintain satisfactory electrolyte balance, and referred to our centre for training for home parenteral nutrition. Further investigations were planned as his lymphangiectasia was unexplained, which included a cardiac opinion. His echocardiography was reported as structurally normal heart with good function and a small pericardial effusion. A CT scan unexpectedly revealed a large clot in his superior vena cava. This was removed during open cardiac surgery, and a thickened pericardium adherent to the heart was found, restricting cardiac function. He recovered well after pericardiectomy on an MCT diet and his ascites resolved over the next 2 months. Histology of the pericardium subsequently confirmed constrictive pericarditis, presumed viral in origin.

Conclusion:
Disorders of the heart including constrictive pericarditis should be considered in cases of unexplained protein losing enteropathy, but these may be difficult to diagnose despite clinical examination, Chest X-ray and cardiac ECHO. ECG findings are contributory and information for cardiologists must be directed towards the suspected cardiac pathology. Tuberculosis should be also sought as a cause for constrictive pericarditis.

Ref
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Successful endoscopic pyloromyotomy with topical Mitomycin C for recurrent pyloric stenosis: a therapeutic option.

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Introduction:
Idiopathic hypertrophic pyloric stenosis is a common surgical problem in infants and pyloromyotomy is almost always successful in alleviating the obstruction. There are few reports in the literature that discuss recurrent pyloric stenosis.

We present a case of a 2 yr old boy from Africa that was referred to our centre for a second opinion. He was born in Honduras and presented with persistent vomiting from 2 weeks of age. This was initially treated with gaviscon and subsequently proton pump inhibitors for presumed gastroesophageal reflux disease. At 17 months of age he presented with haematemesis and an emergency endoscopy done at the time showed gastritis and Barrett's oesophagus on histology. Endoscopy also indicated a pre-pyloric membrane which was confirmed and resected on laparotomy. Post operatively his vomiting persisted with associated weight loss. A barium meal confirmed delayed gastric emptying with pyloric narrowing. A repeat endoscopy confirmed a stenosed pylorus and histology showed pyloric fibrosis. He then went onto have multiple (x6) endoscopic balloon dilatations with no clinical improvement. As his regurgitation to solid foods persisted, nutrition was largely maintained with high calorie milk and drink supplements.

He was then referred to Sheffield Children's Hospital for a second opinion. On initial assessment he was noted to have no dysmorphic features and was well nourished (weight on 50th centile). A clinical diagnosis of gastric outlet obstruction was made which was confirmed with a barium meal and Ultrasound (US). US of the pylorus showed a stomach distended with fluid and an irregular and narrowed pylorus measuring 2.5 cm in length and 1.3 cm in diameter.

He then had an endoscopy confirming a stenosed pylorus and underwent an endoscopic pyloromyotomy with a single use 3-Lumen endosocopic Needle Knife (Olympus®) making 3 mm incisions in 4 quadrants of the pylorus . Post operative recovery was uneventful. There was resolution of vomiting and weight gain with radiological improvement of gastric stasis on barium meal. He re-presented 7 months later with regurgitation to solid foods. A CT scan done at this time excluded an annular pancreas and any 'extrinsic' obstruction to the pylorus. He then underwent a repeat second endoscopic pyloromyotomy using a sphincterotome knife. Radial endoscopic ultrasound guided the incision depth. 5 incisions of 2 mm depth each were made on the pylorus. He remained asymptomatic for 10 further months and warranted a repeat endoscopic pyloromyotomy using a sphincterotome knife in July 2011 followed by topical application of 0.5 ml of the anti-fibrotic agent mitomycin C. He has remained asymptomatic so far with resolution of vomiting, tolerance of normal diet and weight gain.

Conclusion:
We report the first case of successful endoscopic pyloromyotomy alongwith topical application of Mitomycin C for idiopathic recurrent pyloric stenosis. Endoscopic pyloromyotomy appears to be a safe and effective procedure offering a less invasive approach. It may thus reduce the need for surgery and associated morbidity and should be considered as an alternative therapeutic option.

Sweet's Syndrome in a child with ulcerative colitis

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Aim:
To report the association of Sweet syndrome in a patient with ulcerative colitis treated with azathioprine.

Method:
Retrospective case note review

Results:
A 9 year old boy who was diagnosed with Ulcerative Colitis in January 2008 presented with recurrent episodes of painful erythematous macules over his lower limbs and buttocks which developed for the first time in September 2009. He was commenced on azathioprine therapy in September 2008, i.e. a year before he began to develop the rash. This rash was usually associated with joint swelling, mainly in the ankles but occasionally in the feet and hands. With the onset of the rash, the patient was not usually systemically unwell but did experience some abdominal pain and severe pain at the sites of the lesions. His colitis remained well controlled during these episodes. There was often an associated rise in his CRP and a neutrophilia. The rash seemed to reappear when his oral steroid treatment was weaned. The rash improved on each occasion with a three day course of intravenous hydrocortisone.
A biopsy of the rash revealed neutrophilic inflammation of vessels, papillary dermal oedema and focal degeneration of collagen, therefore supporting the diagnosis of Sweet's syndrome.
Azathioprine therapy was discontinued temporarily in view of the medication being the possible precipitant for the condition. However, development of the rash associated with swelling continued to occur episodically.

Discussion:
Sweet's syndrome is diagnosed on the basis of major criteria which include an abrupt onset of tender and painful erythematous plaques or nodules and a predominantly neutrophilic infiltration in the dermis without leukocytoclastic vasculitis. Minor criteria include general malaise and fever, raised inflammatory markers, leukocytosis, an excellent response to steroids and association with chronic inflammatory conditions. It can be drug related and can occur due to azathioprine hypersensitivity. There are few case reports of Sweet's syndrome occurring in the paediatric population. This case highlights an important association with inflammatory bowel disease and the importance of differentiating from other inflammatory conditions which are more commonly associated with Inflammatory Bowel Disease. An awareness of this condition is important to initiate appropriate investigation, including skin biopsy and management of the patient.

Tube it and glue it. Oesophageal perforation following button battery ingestion mistaken for a coin.

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Several complications have been reported as a result of ingested button battery (BB) including oesophageal perforation and strictures, tracheo-oesophageal fistulation, vocal cord paralysis, ex-sanguinations after fistulation into blood vessels and death.

We present the case of a 3-year-old boy with an ingested BB whose initial radiographic appearance was presumed to be a coin lodged in the distal oesophagus.

The child, remaining dysphagic, was transferred on the following day to the regional Paediatric Surgical team. The AP view of his chest X-Ray showed the double-rim typical of a BB. Rigid and flexible oesophagoscopy nearly 20 hours after ingestion confirmed a BB impacted just above the gastro-oesophageal junction which had caused moderate damage to the oesophageal mucosa. The next day the child was pyrexial and dyspnoeic: chest X-Ray showed a right sided hydro-pneumothorax. An emergency right tube thoracostomy drained air and saliva, while flexible oesophagoscopy revealed a 2 cm oesophageal perforation where the burn was noted earlier. Right thoracotomy was performed. In view of inflammatory reaction around the perforation, primary repair was not judged safe. A latex rubber T-tube, usually used for biliary tree operations, was inserted into the perforation to create a controlled oesophago-pleura-cutaneous fistula to let it heal gradually by secondary intention. The T-tube was brought out through the lateral chest wall, and connected to free drainage. A mini-laparatomy allowed gastro-jejunal tube insertion for feeding. On post-operative day 14, a tubogram showed good flow of contrast into the stomach and oesophagus with no leak of contrast from the site of perforation. The child was discharged home with the T-tube in situ, on full jejunal feeds and free fluids by mouth. Three weeks later the T-tube was removed by flexible oesophagoscopy under general anaesthesia. Tissel©, a fibrin sealant consisting of human fibrinogen and thrombin, was applied endoscopically over the internal opening of the oesophageal fistula as an extra safety measure to prevent leakage. Three days later, a swallow contrast study showed normal oesophagus with no leak and no stricture. The child was discharged home the same day after normal solid diet was well tolerated.

Key learning points:
1. Signs of BB should be carefully checked on the AP (double-rim and halo-effect) and lateral (step off) X-Rays. 2. In the absence of a history of observed ingestion, it should be assumed that coin-like foreign bodies are button batteries until proven otherwise. Management strategy of such a patient depends on the extent of perforation and the time interval between perforation and diagnosis. The use of a T-tube to treat delayed oesophageal perforation resulted in complete successful resolution in this case with no further need for surgery. Future case reports may highlight the value of this technique in treating delayed oesophageal perforation in children

Two to tango? Intestinal Spirochaetosis : A rare entity report of 2 cases

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Introduction:
Intestinal spirochaetosis(IS), although a veterinary problem, is gaining recognition as a human quandary. There are only two reported cases in children. We present two further cases with varying presentation.

Case 1:
A 11 year old boy presented with intermittent pain in the epigastric area and bilateral iliac fossae. There was no reported alteration in his bowel pattern. There was no history of foreign travel or contact with any pets or farm animals. A review of systems, physical examination, laboratory and radiology workup did not elicit a cause for his symptoms. His endoscopy was macroscopically normal, however histopathology of colonic biopsy specimens showed a basophilic band along luminal surface of the biopsies on H & E stain without inflammatory infiltrates. A confirmatory silver (Warthin-Starry) stain demonstrated the spirochaetes along brush border discontinuously. Patient received a 2 week course of metronidazole and on completion became asymptomatic and remains so.

Case 2:
9 year old boy presented with abdominal pain, watery diarrhoea, loss of weight and per rectal bleed for couple of months leading to severe anaemia with haemoglobin of 70g/L and hypoalbuminaemia (serum albumin 23g/L). Physical examination and laboratory workup did not yield a cause. His endoscopy was macroscopically normal. Histology of colonic biopsy specimens was studded with brush border picket fence like organisms, confirmed on sliver (Warthin-starry) stain. He received 2 weeks of metronidazole with successful resolution of symptoms. He is currently on oral iron supplementation showing improvement in his iron indices.

Conclusion:
The clinical importance of intestinal spirochaetosis is controversial. The learning points were:

1. Presenting symptoms such as diarrhoea, abdominal pain and rectal bleeding can be non-specific and thus the diagnosis requires a high index of suspicion particularly in children with suspected inflammatory bowel disease.
2. Even though no mucosal inflammation was seen on histology, symptom resolution is achieved with antibiotics thus justifying a low threshold for it.

ABSTRACTS FOR THURSDAY 25TH JANUARY 2012

Invited Speakers
Oral Plenary Sessions
Posters of Distinction
Posters

Bone Marrow Transplantation and other new treatments for IBD

Professor Chris Hawkey, Professor of Gastroenterology, Nottingham University, University Park, Nottingham, NG7 2RD

Quality improvement programme for IBD through inspection

Dr Emma Fernandez, Dr Ian Shaw, Dr Charles Charlton

Background
The 2006 and 2008 National IBD Audit identified large variations in the quality of care across the UK and limited action planning between audits. As a result a National Service Standard was developed (http://www.ibdstandards.org.uk) to support quality improvement.

Aim
The IBD Quality improvement project aims to support services in achieving the National IBD Quality Standards through a web-based system (www.ibdqip.co.uk), comprising a self-assessment tool and a shared repository of best practice ('Shared Document Store'), which provides services with the tools to implement meaningful change.
The assessment tool has 4 different main domains (patient experience, clinical quality, organisation and patient choice and research, education and audit), with a hierarchical grading system.

Subjects and Methods
Over 70 adult and paediatric sites participated in the project. Sites were asked to meet as a multidisciplinary team, with a patient to discuss their service assessment. They provided feedback on the statements, to allow refinement of the tool.
Sites shared best practice using an on line document store and through regional meetings involving staff and patients.
Several sites were then chosen for a visit by a small team, to validate data entry and discuss in details results and action planning.

Results
- Initial analysis shows that teams are scoring as predicted and that data mirrors that of the 2010 UK IBD Audit organisational data.
- Feedback from teams and validation visits has ensured refinement of questions.
- Teams involved have provided positive feedback on the tool and its usability.

Summary
- During regional meetings, patient participation focussed discussion away from the needs of the teams towards change that would improve patient experience.
- There remains large regional variation, and also variations between services within the same Trust.
- Service involvement in the refining of the assessment tool has been important to ensure accurate interpretation of the questions.
- Teams welcomed the sharing of documents.
- Sites are now using their results to prioritise action plans and will meet at further regional meetings to discuss these. The next round of self assessment will take place in March 2012.

Conclusion
- An on line benchmarking tool like this is low cost and easy to implement. This approach could be translated to many other long term conditions.
- Teams have welcomed a resource which they can use to support commissioning.
- Patient involvement from the outset gives improves decision making.
- This approach is has less burdensome data entry and reporting, with immediate results. This speeds up the audit cycle and maintains clinical engagement.

What matters for patients and families

Mr Richard Driscoll, Chief Executive Officer, Crohn's and Colitis UK, 4 Beaumont House, Sutton Road, St Albans, Herts, AL1 5HH

Parents and teenagers have been invited to give their comments on what matters and the themes of their responses will be presented. Crohn's and Colitis UK has also run a number of family days and assisted with parent and family involvement in a number of paediatric IBD services. The issues raised at those events will be considered and the process of involving parents and families in providing IBD care will be discussed.

Preparing children for pouch surgery

Ms Ali Wright, Specialist Stoma Nurse, Nottingham Children's Hospital, Derby Road, Nottingham, NG7 2NH

My presentation aims to give an insight into my role as a stoma nurse working with children and young people who have Ulcerative Colitis.

When surgery is discussed as a treatment option to a child or young person; as part of the multi disciplinary team I explain about surgery for a colectomy, formation of ileostomy and ileoanal pouch. I spend time with the patients looking at stoma care products and talk about stoma care and how to balance this with school, work friends and family life. Fulfilling the pre operative needs of a young person considering surgery can differ vastly from their parents needs and at times can be challenging; but can have a positive effect on post operative recovery.

Patient and family perspectives of severe Crohn's involving liver disease

Howard and Huw Arthur

'Huw was diagnosed with Crohns at five years of age and has lived with Crohns for the past fifteen years rather than suffered from it even though Huw's experience of Crohns is at the severe end of the spectrum. Huw and his father, Howard, will speak about the experiences of family life, social, education, healthcare systems and processes and some thoughts and reflections on how the systems could be further developed and improved.'

Drug-induced Liver Injury

Guruprasad P Aithal MD, FRCP, PhD

Drug-induced liver injury (DILI) is the most frequent reason cited for the withdrawal of approved drugs from the market and accounts for significant proportion of cases of acute liver failure. DILI can mimic variety of clinical syndrome and yet is commonly misdiagnosed. Although considered uncommon in children, recent studies have described the frequency of DILI, common drugs associated with DILI and risk factors that increase susceptibility to DILI in the paediatric population. International consensus on case definition and diagnostic criteria has improved phenotyping and characterization of a spectrum of clinical syndromes that constitute DILI. There is increased understanding of interaction between drug, host and environmental factors. Recent genome wide association studies have highlighted the role of immune system in the pathogenesis of DILI. In addition, features of hypersensitivity may influence the natural history and prognosis in DILI. Early detection and withdrawal of the offending agent is the most important step in the prevention of adverse outcome. Biomarkers that pre-empt and detect potentially serious DILI are desperately needed.

Fatty liver disease where we are and what the future holds

Professor Anil Dhawan, King's College Hopsital, Denmark Hill, London

Advances in Alpha1-antitrypsin deficiency

Professor Noor Kalsheker
School of Molecular Medical Sciences and Centre for Genetics and Genomics, Nottingham University.

Alpha1-antitrypsin (AAT) is the major circulating serine protease inhibitor in human plasma. Genetic deficiency of AAT is associated with predisposition to developing liver disease in childhood and lung disease in early adult life, particularly in cigarette smokers. Severe deficiency occurs throughout the world and is most prevalent in northern Europeans affecting about 1 in 3000 of the population. 5-10% of children with severe deficiency present with liver cirrhosis with an initial presentation of prolonged neonatal jaundice. AAT deficiency represents the commonest metabolic cause for liver transplantation in children, currently the only effective treatment option for advanced liver disease. AAT deficiency is under-recognised and under-diagnosed. The mean interval from presentation with symptoms to diagnosis in adults is about 8 years and children are more likely to be diagnosed as part of screening for prolonged neonatal jaundice. The liver disease is caused by abnormal polymerisation of the protein in hepatocytes, the major site of AAT synthesis and the lung disease is due to the deficiency of AAT in the circulation, resulting from poor anti-protease activity. For the latter, replacement therapy has been used in clinical trials for AAT deficiency. For the liver disease, compounds which prevent polymerisation have been developed and are being tested for therapeutic use. Correction of the genetic defect for the most common form of severe AAT deficiency in pluripotent stem cells, has been successfully achieved and should offer new therapeutic options in the future.

Identification of risk factors for metabolic syndrome and obesity in healthy school children (REACH Y6 cross-sectional pilot study)

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Introduction:
Non-alcoholic fatty liver disease (NAFLD) has evolved as the most common referral for chronic liver disease in children. Although an association with obesity and insulin resistance is established, risk factors for development, of NAFLD, progression, long-term data and interventions are inconclusive in children.

Aim:
The aim of our study was therefore to analyse the correlation of non-invasive measures and novel bio-markers with standard measures of risk for metabolic syndrome.

Method:
A cross-sectional study in a healthy population of 10-11 year old school children (REACH Y6 cohort) was performed, to which 62 participants voluntarily agreed Blood samples were taken and assayed for transaminases, lipid profile, fasting insulin (and HOMA-IR), adiponectin, and high sensitivity CRP. Body composition included BMI standard deviation score(sds) and anthropometry. Cardiorespiratory fitness level was assessed by peak oxygen uptake (VO2peak), by incremental treadmill test. Statistical analysis of non parametric data was performed by SPSS and Spearman's correlation coefficients were calculated.

Results:
In this cohort, 7 children (11.3%) had a BMI greater than 25 (>99.6th centile for this age group), 6 were female. 1 child demonstrated raised ALT of 65 U/l and was obese.
HOMA-IR correlated positively with the BMI sds ($p=0.001$, $r= 0.428$) and negatively with VO2peak ($p=0.0001$, $r= -0.498$), suggesting the association of body mass index, fitness level, and metabolic syndrome in this cohort.
High sensitive CRP correlated positively with the BMI sds ($p=0.001$, $r= 0.416$) and negatively with VO2 fitness level($p=0.0001$, $r= -0.45$).
Adiponectin was negatively correlated with HOMA-IR ($p<0.05$, $r= -0.302$) and also with BMI sds ($p=0.0001$, $r=-0.468$). These results underline the impact of physical activity in both obese and non obese children. They also show that both hs-CRP and adiponectin appear to play a role in the pathways contributing to the metabolic syndrome in children, and indicate sensitive changes in obese children, with normal transaminases.

Conclusion:
Our findings illustrate a clear correlation between fitness level, BMI sds, and HOMA-IR values, markers of the metabolic syndrome. Our results indicate the value of novel biomarkers including hs-CRP and adiponectin in the early identification of risk factors leading to metabolic syndrome, obesity, and possibly the development of NAFLD.

Serum protein N-glycosylation as a biomarker of paediatric NAFLD

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Introduction:
Serum protein N-glycosylation has previously been shown to distinguish adult patients with simple steatosis from non-alcoholic steatohepatitis (NASH). The pattern of disease in paediatric patients is distinct from adults.

Aim:
The aim of this study was to characterise the glycomic profile of children with varying degrees of NAFLD to identify potential biomarker profiles of disease. Children with biopsy proven non-alcoholic fatty liver disease (n=51) were recruited from a tertiary paediatric hepatology unit. Liver biopsy was scored according to NAFLD activity score.

Method:
Blood was taken on day of biopsy for analysis. Serum protein N-glycosylation patterns were assessed with DNA-sequencer assisted fluorophore-assisted capillary electrophoresis (DSA-FACE) and compared with histology.

Results:
Median age at biopsy was 13.3 years (range 4.5-17.4). 31 were male. Median BMI z-score was 1.81. 23 children scored as simple steatosis / borderline NASH and 28 as true NASH. 18 children had no / minimal fibrosis (<F2) and 33 had significant fibrosis (≥F2). Similar to previous work in adult patients with NAFLD, peak 1 (NGA2F) was the most significantly raised N-glycan in paediatric NASH patients with peak 5 (NA2) demonstrating the greatest decrease. The logarithmically transformed ratio of peak 1 to peak 5 (Glycomarker) was -0.85 (SD 0.22) in simple steatosis / borderline NASH and -0.73 (SD 0.12) in NASH ($p=0.02$).

The biomarker correlated well with the amount of lobular inflammation with a consistent increase with ascending grade of inflammation. There was also a trend towards significance in differentiating patients with significant fibrosis ≥F2; -0.74 (SD 0.13) from patients with no / minimal fibrosis <F2; -0.86 (SD 0.24), ($p=0.06$). Glyco-analysis of immunoglobulin G (IgG) confirmed the undergalactosylation status with a significant increase in peak 1 (NGA2F; $p=0.024$) and a significant decrease of peak 6 (NA2F; $p=0.01$) on IgG. In multivariate analysis of the Glycomarker, GGT, AST and INR, only the Glycomarker displayed a significant result for distinguishing simple steatosis form NASH ($p=0.019$)

In conclusion, the findings in this study are novel in that they represent the first Glycomic analysis of paediatric NAFLD. They validate findings in adults in that a Glycomarker can serve reliably as a biomarker of severity of disease in NAFLD. The same N-glycosylation alterations are observed in paediatric NASH patients when compared to an adult population and therefore the same biomarker can be used. B cells play a dominant role in the N-glycan alterations of NASH patients, both in an adult and paediatric population.

Extra-hepatic Portal Venous Obstruction: The Scottish Experience

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Introduction:
Extra-hepatic portal vein obstruction (EHPVO) is an uncommon condition. It is however a common cause of portal hypertension and significant gastrointestinal haemorrhage in children. Population based data on this rare condition is limited from the developed world. Opinion is divided on the most appropriate management strategy with regards to serial therapeutic endoscopy versus the relatively new Meso-Rex shunt surgery.

Aims:
(1) To identify all cases of EHPVO in Scotland, estimate prevalence and describe the early presentation and diagnostic work up. (2) To study the long term outcome of this condition in the era of the Meso-Rex shunt.

Method:
Multicentre collaborative study with retrospective longitudinal collection of data based on case note review. Information collected included demographic data, mode of presentation, endoscopic and radiological findings, and transition to adult care where relevant.

Results:
Thirteen patients were identified (8 male, 5 female). After one moved abroad and was therefore lost to follow-up, the remaining twelve were followed up for a mean of 9.5 years (range 1-18 years). This represented 114 years of cumulative follow up. 6/13(46%) presented with significant GI bleeds under the age of 5. 10/13(77%) had significant perinatal factors that may have contributed to the origin of portal venous thrombosis (including prematurity, UVC placement, omphalitis and congenital anomalies). 8/13 (62%) had at least one abnormality on the thrombophilia screen. There were 25 episodes of GI bleeding that required hospitalisation and 20% of these episodes required blood transfusion. 7/13(54%) have either already had a Meso-Rex shunt or are currently being considered for this surgery. Interestingly, none of the patients who were treated early with betablockers have proceeded to a shunt procedure. Ultrasound scanning was usually successful in making the diagnosis but often needed to be repeated on one or more occasions.

Conclusion:
The diagnosis of EHPVO requires a high index of suspicion and repeat sonography should be considered. Unexplained gastrointestinal bleeding, particularly in the very young child, should alert the clinician to the possibility of EHPVO. Perinatal risk factors may contribute to the origin of this rare disease highlighting the importance of a high standard of care in the neonatal period. Early consideration of the Meso-Rex shunt eliminates the need for therapeutic endoscopy and may result in reducing the risk of GI bleeds. Further collaborative study investigating this rare condition will help refine current practice.

Neonatal acute liver failure (NALF): Review of 20 years experience

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Introduction and Aim:
Neonatal acute liver failure defined as coagulopathy (INR >2) uncorrected by Vitamin K, is a rare disorder with high mortality. We aim to describe the aetiology and outcome of NALF in a nationally designated centre:

Methods:
Data were collected retrospectively from medical records and liver unit database fro 1991-2011.

Results:
67 neonates were included (term 53, preterm14); (Male:Female= 38:29). Aetologies of NALF were: infections (n=17) – HSV (1)), enterovirus (3), CMV (1), Staph Aureus (1), E Coli (1), other (1). Metabolic (n=22). Neonatal haemochromatosis (NH) (n=14), Miscellaenous (n=5) and Indeterminate (9). 21 children (31%) recovered with medical management alone. 28 (42%) died despite intensive medical management. 18 children (27%) underwent liver transplant (LTx) of whom 10 are alive and 8 died. The overall survival in NFALf was 46%. Mortality (n=36) according to aetiology due to: Infectious (n=11, Herpes 6, Entervirus 2), Metabolic (n=13, mitochondrial 8, Niemann Pick C 2, OTC deficiency (1), galactosaemia (1), tyrosinaemia (1), NH (3), Miscellaenous (9). 4 neonates died due to severe co-morbidities and 4 neonates died on LTx waiting list. Immediate post transplant mortality (5) was due to disseminat4ed herpes infection (2), vascular complication (2) and sepsis (1). Late post transplant mortality was due to progressive mitochondrial disease (2) an Niemann-Pick C (1).

	1991-1995 (n=10)	1996-2000 (n=11)	2001-2005 (n=15)	2006-2011 (n=31)
Median age of presentation (range)	6 d (1-25)	6 d (1-24)	3 d (1 – 28)	4 d (1-28)
Survival with medical treatment	0	4	6	11
Number of liver transplants	3	4	2	9
Median age in days and weight in kg at transplant	31d (data NA)	14 d 2.7 kg	48, 3.3 kg	21d, 3.5 kg
Median donor:recipient weight ratio	(data NA)	6.7	4.3	11.4
Survival with liver transplant	2	3	0	5
Death following liver transplant	1	1	2	4
Overall survival	2 (20%)	7 (64%)	4 (40%)	16 (52%)

31% of neonates were rescued by medical treatment alone, 60% (6/10) of children with HSV died of advanced disease. Disseminated herpes infection is associated with a high mortality and caution should be exercised in considering them for LTx in context of multi organ failure. Mitochondrial cytopathies and Niemann-Pick C can present as NALF and should be excluded before considering children for LTx. In our patient group, NH had a better prognosis.

Conclusion
The outcome of NALF is dependent on underlying cause and liver transplantation can be used as a rescue treatment once multi-systemic metabolic disease has been excluded.

Trace element levels in intestinal failure patients and response to parenteral nutrition

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Introduction:
Critically ill children with intestinal failure (IF) are unable to absorb sufficient fluids and nutrients to gain weight and grow. Micronutrient deficiencies may develop.

Aim:
The aim of this study was to assess the effect of parenteral nutrition (PN) supplementation of trace elements in children with severe IF.

Method:
All children treated with PN for more than 28 days at a tertiary paediatric centre over a 14-month period were included. Children were prescribed the manufacturer's recommended trace element doses in line with ESPGHAN guidelines (1ml/kg/day Peditrace ® -Fresenius Kabi if body weight ≤15kg or 0.1ml/kg/day Additrace ® - Fresenius Kabi if body weight >15kg). Serum concentrations of copper, selenium and zinc were recorded at start and after about 28 days of PN (range: 14-31). Trace element levels were compared using one-way ANOVA analysis in different disease groups (gastroenterology, surgery, haematology/ oncology, intensive care, other).

Results:
140 children (77 male, 55%), median age 1.7 years (range: birth to 16.5), presented from January 2010 to March 2011. Median duration of PN treatment was 41 days (range: 28-70). Median copper concentration at presentation was 17.4mmol/l (range: 7.6-34.8) and after PN 14.8mmol/l (range: 8.1-30). At presentation 18% had copper deficiency with levels <12.6mmol/l and 38% after 28 days. Median zinc concentration before PN was 12.0mmol/l (range: 4.8-21.9) and after 28 days 13.1mmol/l (range: 9.8-16.6). 33% had a low zinc level (<11.0mmol/l) and in 18% the level was still low after treatment. The median selenium concentration was 0.5mmol/l (range: 0.3-1.7) and 0.6mmol/l (range: 0.4-1.4) at presentation of PN and after 28 days respectively. Prior to treatment 36% of patients had serum selenium below the normal range and in 31% remained low after 28 days on PN. Paired samples t-test revealed no significant differences in the levels of trace elements before and after treatment with PN. No child had clinical signs of trace element deficiencies. There were no significant differences in trace element levels between the different groups.

Conclusion:
We have demonstrated that low blood trace element levels are common in children presenting with severe IF and persist after 4 weeks of PN when prescribed at current manufacturer's and ESPGHAN regulations. Copper and selenium deficiencies were most common. It is important to adjust PN trace element content in response to plasma levels Our next aim will be to audit the amount of PN the patients actually received and whether current trace element recommendations need to be increased during the first 28 days of parenteral nutrition.

Six years of the British Intestinal Failure Survey (BIFS)

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Introduction:
Intestinal failure (IF) has been changed from a fatal condition to a manageable one with the advent and development of parenteral nutrition (PN). There remains a paucity of information on the prevalence of IF across the UK. BIFS was established in 2005 to determine incidence and outcome of paediatric IF in the UK to establish the number of children who may require small bowel transplant (SMBTx).

Method:
Subjects: Inclusion Criteria: Children (<=18years old) who were on PN ≥28 days as inpatient. Exclusion Criteria: Pre-term neonates on PN solely because of immature gut function.
Methods: Data captured prospectively (at local participating centres) include baseline demographics recording diagnosis, length of time on PN, age at commencement of PN with six monthly follow up to confirm data is current and accurate. Data are submitted to BIFS Administrator and recorded on a centrally held database.

Results:
538 subjects enrolled between July 2005 and October 2011. Two year post date of first PN episode is available for 337 of them. 21 of these were excluded from further reporting as they started PN pre-2004 so reliable follow up data was unavailable. 39 satisfied the criteria for recruitment into BIFS but did not have a diagnosis of "intestinal failure". 277 are reported on.

At two years post commencing PN, 211 (76%) had been weaned off PN, 39 (14%) had been discharged on HPN, 7 (3%) were on PN as an in-patient, 2 (1%) had transferred to adult services and 18 (6%) had died. Reported events are given in Table 1

Table 1.

Diagnosis Description	Disorder of Motility (n=40)	Enteropathy (n=36)	Short Bowel Syndrome (n=201)
Line Sepsis*	18 (45%)	12 (33%)	88 (43%)
Jaundice*	2 (5%)	8 (22%)	45 (22%)
Non-Tx Surgery*	20 (50%)	3 (8%)	77 (38%)
Tx Assessment	4 (10%)	6 (16%)	35 (17%)
Listed for Tx	4 (10%)	5 (13%)	21 (10%)
Transplanted	3 (7%)	4 (11%)	13 (6%)

*At least 1 episode reported within two years of starting PN

Conclusion:
BIFS has shown itself to be a successful model of collaboration for centres involved in the management of children with IF across the UK. Recruitment of subjects has been made difficult by the need for consent and the BIFS reliance on voluntary contribution of information.

Posters of Distinction

A five year follow-up of children with home parenteral nutrition

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Aim:
The aim of this study was to review 5-year outcome of patients established on long term home parenteral nutrition (PN) in 2006 with regard to survival, resolution of intestinal failure, complications of PN and growth.

Method:
All patients attending our specialist intestinal failure clinic on treatment with PN at home in 2006 with care by parents were reviewed. Nutritional intake was regularly reviewed and protein and calorie intake increased if weight gain or growth were impaired. The main aetiology of intestinal failure, survival, continuing dependence on PN, need for transplant, growth and liver function were recorded.

Results:
33 children (55% male) were identified. The median age in 2006 was 11.9 years (0.6-18). Underlying aetiologies were enteropathy in 17 or 52% (2 with a primary immunodeficiency), intestinal dysmotility in 8 or 24% and short gut syndrome in 8 or 24%. PN was initiated as a neonate in 64%, infancy in 12%, 1-5 years in 21% and > 5 years old in 3%. Median PN duration was 13.5 years (1.5-20.7) with an overall follow up period of 13 years (1.6-22.8). 11 or 33% children successfully weaned off PN after a median period of 2.8 years (1.5-18.7). The weaning rate of PN was highest in enteropathy (41%), followed by short gut syndrome (38%) and intestinal dysmotility (12.5%). Four patients had significant life-threatening complications and underwent intestinal transplant (2 for liver failure; 2 for excessive intestinal fluid losses). Two other patients had abnormal liver function tests (one transaminitis, one cholestasis) whilst still on PN at 5 years. A further two patients with immunodeficiency underwent bone marrow transplant and died. 22 patients (15/17 enteropathy, 4/8 short gut and 3/8 dysmotility) had sub-optimal growth with height on or below 0.4th centile (UK-WHO centile chart) that did not respond to good intake of PN protein and calories. None of the patients died on long-term PN without transplant.

Conclusion:
This study demonstrated that children who are well established on PN at home with care by parents have a 1 in 3 chance of weaning from PN over a 5-year period. Overall growth is poor and not improved by increasing protein and calorie intake. All 27 children who did not undergo intestinal or bone marrow transplant survived.

Bowel preparation for paediatric colonoscopy: A systematic review

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Introduction:
Adequate pre-colonoscopy bowel preparation is crucial to ensure complete visualisation and thus successful diagnostic and therapeutic colonoscopy. Administration of bowel preparation agents can be problematic in children, with reduced tolerance to agents often used in adults and poor compliance, as well as increased rate of complications. We carried out a systematic review to summarize the available evidence investigating the optimum bowel preparation agents for colonoscopy in children using the Cochrane Collaboration format.

Method:
Randomised controlled trials (RCTs), published between 1966 and July 2011, which compared agents for bowel preparation before elective colonoscopy in children 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.

Results:
7 RCTs investigating different interventions met the inclusion criteria. Oral sodium phosphate (NaP) was investigated in 4 studies. Meta-analysis of 2 studies with 63 participants comparing polyethylene glycol (PEG) with NaP showed no significant difference in the quality of bowel preparation (Odds Ratio 2.90, 95% confidence interval 0.91 to 9.24). In one of these studies, a nasogastric tube (NGT) was passed in all patients receiving PEG, while in the remaining study, 53% of participants in the PEG group were unable to finish taking the solution. 1 study compared NaP with Magnesium citrate and enema, again concluding NaP was better tolerated and equivalent in preparation quality. The final study compared NaP with a diet kit and magnesium citrate/bisacodyl, finding comparable preparation quality and tolerance. 2 studies with 165 participants investigated Sodium picosulphate + Magnesium citrate, comparing it to PEG and bisacodyl + enema respectively. Both studies reported better acceptability of Sodium picosulphtate, with lower rates of abdominal discomfort and higher satisfaction respectively. Better quality bowel preparation compared to biscacodyl and equivocal bowel preparation to PEG was seen. Once again, PEG was found to be more difficult to tolerate, with 75% of participants requiring an NGT, compared with 2.5% in the sodium picosulphtate group. No serious adverse events were reported in any of the studies.

Conclusion:
The available evidence suggests that Sodium phosphate and Sodium picosulphtate with Magnesium citrate are well tolerated and effective for bowel preparation in children. PEG also appears effective, but seems to be poorly tolerated in children. The evidence base is extremely small and heterogeneous. Further research is needed to investigate the most appropriate agents for bowel preparation in children.

Duodenal haematoma: A rare complication of upper Gastro Intestinal Endoscopy and biopsy.

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Introduction:
Intramural duodenal haematoma was first reported in 1838. The commonest cause is blunt abdominal trauma with other causes such as coagulation disorders and pancreatic disease also documented. Rarely, iatrogenic haematomas can occur as a complication of diagnostic endoscopy. We present a case of iatrogenic intramural duodenal haematoma following upper gastrointestinal (GI) endoscopy and review the current literature.

Case report:
A 5 year old boy with cerebral palsy weighing 17kg with symptoms of gastro-oesophageal reflux underwent a diagnostic upper GI endoscopy and pH study for objective assessment of ongoing pain thought to be because of gastro-oesophageal reflux despite anti-reflux medication. The procedure was uncomplicated and he recovered well and was discharged home that day after receiving a feed. That evening he began vomiting and became very distressed. His parents thought he would settle but 36 hrs later brought him to Accident and Emergency department where he required fluid resuscitation because of hypovolaemic shock and was admitted. He was treated for possible aspiration pneumonia due to chest signs and raised inflammatory markers (CRP 229). Abdominal examination was normal and abdominal xray (AXR) did not suggest GI perforation. He had a normal serum amylase and normal liver function tests. His vomiting did not settle and 48 hours later became bilious. An abdominal ultrasound and upper GI contrast were performed revealing an intramural duodenal haematoma. He was commenced on TPN and kept nil orally. By day 14 post procedure bilious aspirates via NG tube had stopped and resolution of the haematoma was confirmed on USS. By day 24 post procedure the patient was tolerating full enteral feeds. He was investigated for a possible coagulation disorder but all tests were negative.

Literature Review:
We conducted a review of the literature as we had not seen this complication before. This revealed 32 cases: 84% occurred in children under 18 years with males and females equally affected. . It appears more likely to occur in children than adult. 7 cases occurred in leukaemic bone marrow transplant recipients, 2 cases had Noonan's Syndrome and 11 cases reported clotting abnormalities or thrombocytopenia. All cases presented with vomiting and abdominal pain with onset 2 to 48 hours post procedure. 44% of cases were associated with a hyperamylasaemia. Management was mostly conservative with monitoring by ultrasonography. Surgical management varied from transcutaneous drainage to laparotomy and the indications for surgical management were not clear. Mean length of stay in hospital was 20 days (7 – 41 days) and surgical intervention did not shorten this. There were 3 deaths reported, all in leukaemic patients.

Discussion:
Intramural duodenal haematoma is a rare complication of diagnostic upper GI endoscopy and the incidence is unknown. It is not listed as a complication in guidance from the American Society of Gastroenterology or British Society of Gastroenterology. Haematomas may occur due to the fixed retroperitoneal position and rich submucosal blood supply of the duodenum and may be related to the shearing forces applied when taking biopsies. It can cause pancreatitis and obstructive jaundice from compression of surrounding structures.

Conclusion:
It is important to be aware of this rare but serious complication and should be mentioned when taking consent for diagnostic upper GI endoscopy.

Ileal intubation in paediatric ileo-colonoscopy in a large centre in the UK

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Introduction:
Sheffield Children's Hospital is a supra-regional centre for paediatric endoscopy, and a significant number of ileo-colonoscopies occur. We randomly audited ileal intubation rates in our centre from 2008-10. The reasons were primarily to audit if ileal intubation provided any diagnostic advantage over routine colonoscopy in detecting ileal changes in inflammatory bowel disease (IBD). The reasons for non-intubation were determined.

Aims:
1. Retrospectively and randomly determine the success rate of ileal intubation.
2. To determine if ileal intubation at ileo-colonoscopy offered any advantages in differentiation and diagnosis of inflammatory bowel disease in children.

Method:
Retrospective analysis of 100 case notes selected randomly of colonoscopy and ileal intubation performed by experienced paediatric endoscopists over a 3 year period.

Results:
1. Successful ileal intubation occurred in 89% of colonoscopies.
2. Technical difficulties precluded intubation in 11% - out of which 10 % were due to poor bowel preparation, and 1% was due to bleeding, where safety issues arose in acute colitis.
3. 24 children were diagnosed with IBD (10 UC with no ileal abnormalities).
4. Ileal findings contributed to the diagnosis in 5 out of the 11 Crohn's disease identified, of whom 1 child had ileal disease alone.
5. 3 had a diagnosis of indeterminate colitis with ileal changes but no granulomas, hence ileal intubation altered the diagnosis from UC in 16.7%.
6. Poor bowel preparation was the only significant reason for non-intubation - exclusively in children with recurrent abdominal pain who, in retrospect, may have had colonic stasis and would have required more bowel preparation than others.

Conclusion:
We have found in our experience, ileal intubation is useful in differentiating IBD type in 16.7%, and contributes to making the diagnosis in 21%.

Immune dysregulation and defects in mucosal B cell homeostasis in patients with PTEN hamartoma tumor syndrom

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Introduction:
The tumor suppressor phosphatase and tensin homolog deleted on chromosome 10 (PTEN) is the central regulator of the PI3K/AKT signaling pathway. Mice with defects in this pathway develop multiple alterations in T and B lymphocyte homeostasis leading to gastrointestinal lymphoid hyperplasia, autoimmunity and lymphomas. The immunological consequences of PTEN deficiency in humans have not been systematically analyzed.

Method:
We investigated 34 patients with PTEN hamartoma tumor syndrome (PHTS) for clinical immune dysregulation. In a subgroup of 14 patients with gastrointestinal lymphoid hyperplasia a functional analysis of the immune activation via the PI3K pathway was performed.

Results:
In patients with PHTS (Bannayan Riley Ruvalcaba or Cowden syndrome) we recorded immune dysregulation including gastrointestinal lymphoid hyperplasia (n=16), extensive hyperplastic tonsils (n=3) and thymus hyperplasia (n=1). Autoimmunity is indicated by autoimmune lymphocytic thyroiditis (n=6) and autoimmune hemolytic anemia (n=1). Functional analysis of the gastrointestinal lymphoid hyperplasia revealed increased mTOR signaling including S6 phosphorylation within CD20+CD10+ germinal center B cells resulting in increased proliferation. Furthermore, we found reduced apoptosis of germinal center cells. By contrast proliferation in T cell areas in situ was normal.

Conclusions:
Deficiency of PTEN in humans has a marked functional impact on B cell homeostasis by modulating the PI3K/AKT signaling pathway via mTOR and anti-apoptotic signals. Our results suggest that gastrointestinal and systemic immune dysregulation is part of the disease spectrum in patients with heterozygote PTEN deficiency.

Langerhans Cell Histiocytosis presenting with gastrointestinal involvement

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Aims:
Langerhans Cell Histiocytosis (LCH) is a rare disorder characterised by abnormal proliferation of the Langerhans cell1. While single system disease has a good prognosis, multisystem disease often requires intensive combination chemotherapy2,3. Gastrointestinal (GI) involvement in LCH remains rare4 and not criteria for such treatment. The clinical picture can vary but most often consists of bloody diarrhoea, non bloody diarrhoea or constipation, protein losing enteropathy, hypoproteinaemia and anaemia. It almost exclusively affects children below two years of age and it has been noted that over half of patients with LCH and GI involvement die within one and a half years of diagnosis4,5. From our experience of this case we recommend that combination therapy is considered for cases of LCH with GI involvement.

Methods:
A six month old female infant initially presented FTT and diarrhoea later developing a non-blanching erythematous rash involving the abdomen and groin and bloody diarrhoea and intermittent bilious vomiting. She was pale, wasted and developmentally delayed with a painless cystic lump on the back of her head. There was lymphadenopathy and an enlarged spleen. Investigations revealed anaemia, thrombocytosis, elevated fibrinogen, raised ESR and hypoalbuminaemia. Florid histiocytic inflammation of the small bowel was identified at endoscopy and a barium swallow showed narrowing of the distal duodenum and proximal jejunum without obstruction. Gut biopsies and skin samples confirmed the diagnosis of LCH.

Results:
Dual therapy consisting of vinblastine and prednisolone was commenced with quick regression of her bone and skin disease. However, the addition of mercaptopurine and methotrexate was eventually required to control her GI disease. Almost three years post diagnosis she remains on this quadruple regimen as maintenance therapy. Her current disease status is that of regressing disease although her symptoms return towards the end of treatment intervals.

Discussion:
The prevalence of GI involvement may be underestimated given its presentation; mild cases may be overlooked without endoscopic examination and biopsies. We may see a rise in recorded cases with increased availability of endoscopy. . From our experience of this case we recommend that combination therapy is considered for cases of LCH with GI involvement.

References:

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Management of children with Inflammatory bowel disease-a district general hospital experience.

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Introduction:
Inflammatory Bowel Disease (IBD) encompasses two related but distinct disorders of as yet unknown cause. Many believe the
incidence has increased substantially in recent years – 2.2 to 6.8/100 000 in
paediatric/ adolescent population worldwide. About 25% of IBD onset is in the paediatric age range particularly in adolescence but with 5% occurring before age10.
Current therapeutic goals in children and adolescents are to diagnose/ treat relapses early, improve clinical management and reduce morbidity.

Aim:
To review the management of children with Inflammatory Bowel disease presenting to the outpatients department of a district general hospital in UK.

Method:
A retrospective clinical audit.
Recommendations of the Working group on Inflammatory Bowel Disease of the British Society of Paediatric Gastroenterology and Nutrition, 2008, formed the standards to evaluate our local practice. Data on establishing the diagnosis (history taking, examination, investigations) and treatment was collected retrospectively and compared with the standards.

Results:
A total of 14 patients presenting to the children's outpatient department over a 5 year period were identified at random. Local practice maintained 100% standards for history taking, examination, blood tests, upper GI endoscopy with biopsy and colonoscopy. Only 28% of the patients had Barium meal and follow through and 92% had faecal or use of elemental nutrition as first line treatment and Azathioprine as first line for maintenance tests. For their treatment children with Crohn`s disease maintained 85% standards f of remission. Children with Ulcerative colitis fared better at achieving 100% standards. Need for replacing Barium meal follow through with Ultrasound abdomen was suggested.

Conclusion:
Our clinical audit has helped to review the management of children with inflammatory bowel disease in a district general hospital setting. Local protocol helps to maintain good management standards as recommended by the working group. Introduction of service provision in the form of abdominal ultrasound was identified to improve quality of care.

Reference:
"Guidelines for the management of Inflammatory Bowel Disease
In children in the United Kingdom", produced by the IBD working group
Of British Society for Paediatric Gastroenterology, Hepatology
& Nutrition, November 2008

Parenteral Nutrition (PN) referral form' can improve PN practice

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Introduction:
Short term PN is widely used by various paediatric sub-specialities. We audited PN practice during acute admissions in a tertiary Paediatric department using the'NCEPOD' toolkit. Poor documentation was a consistent finding along with difficulty in monitoring patients whilst on PN. A 'PN referral form' was therefore designed on the lines of NCEPOD toolkit.

Aim:
To evaluate if introduction of 'PN referral form' helps to improve documentation and PN practice in paediatric acute admissions.

Method:
PN referral forms were made available to PN requesting teams by Paediatric PN pharmacist. The form consisted section A (clinical details prior to use of PN) completed prospectively by requesting teams and section B (monitoring whilst on PN) completed by retrospective analysis of case notes and biochemistry results. Consecutive patients were studied to avoid bias over six weeks. Recommendations in the NCEPOD report and local pharmacy guidelines were used as standards.

Results:
9 patients (7 males; 2 females) were studied. Specialities requesting PN were medical oncology (3), paediatric gastroenterology (3) and intensive care (3). PN indication and treatment goal were documented in 9/9 cases (100%) compared to 37% and 62% respectively in previous audit. All patients had enteral nutrition considered as an alternative before starting PN and had nutritional assessment with risk of re-feeding. Details related to insertion of central venous catheter were completed in all 9 patients. PN prescription was reviewed daily by PN pharmacist in a working week with appropriate plans made for weekend. Clinical and biochemical monitoring of patients on PN was done daily. Biochemical abnormalities were studied at prior to commencing PN, within 36 hours of starting PN (Early phase) and between 36 hours and 7 days of starting PN (late phase). It was noted that about half (5/9) patients had biochemical abnormalities prior to starting PN and same number of patients continued with biochemical abnormalities in early as well as late phase of PN. PN was stopped after clinical improvement in 8/9 patients and one patient got transferred to referring hospital on PN.

Conclusion:
PN is widely used by different paediatric sub-specialities. Documentation in all areas improved significantly after introduction of PN referral forms. Delivery of PN and monitoring during PN is of high standard (Pharmacy). Biochemical abnormalities are seen in at least half of the patients and therefore monitoring is important. The relationship of biochemical abnormalities to use of PN needs further evaluation.

What has this audit added?
'PN referral form' can improve documentation in patients receiving PN during acute admissions
Ensures consistent practice
Relatively easy to complete (by the requesting team)
Can act as a reminder for several aspects of PN use in relatively inexperienced staff
Can act as an audit tool

Rapid Rise in Incidence of Irish Paediatric Inflammatory Bowel Disease

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Aim:
To describe the change in incidence of paediatric inflammatory bowel disease (IBD) observed at the National Centre for Paediatric Gastroenterology, Hepatology and Nutrition (NCPGHAN), and to determine whether the presenting phenotype and disease outcomes have changed during the past decade.

Method:
The annual incidence of IBD in Irish children aged < 16 years was calculated for the years 2000-2010. Two subsets of patients, Group A (diagnosed between 1st January 2000 - 31st December 2001), and Group B (diagnosed between 1st January - 31st December 2008) were retrospectively assigned disease phenotypes using the Paris Classification. Phenotype at diagnosis and two year follow up were then compared.

Results:
406 new cases of IBD were identified. The incidence was 2.5/100,000/year in 2001, 7.3 in 2008 and 5.6 in 2010, representing a significant increase in the number of new cases of both CD and UC. There were 238 cases of Crohn disease (CD); 129 of ulcerative colitis (UC); and 39 of IBD-unclassified (IBDU). Comparing Groups A and B, no differences were found in disease location at diagnosis or, for CD, in its behaviour.

Conclusion:
The incidence of both childhood UC and CD in Ireland has increased significantly over a relatively short period of time. However, disease phenotype at diagnosis has not changed. At two years follow up, CD appears to progress less frequently than in some neighbouring countries. Prospective longitudinal studies in this defined population will help to elucidate further the changing epidemiology of childhood IBD.

The prevalence and indication for paediatric gastrostomy feeding in Cardiff and Vale University Health Board

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Introduction:
Feeding via gastrostomy is recognized to be relatively common but there is a paucity of data regarding the extent, indication and outcome of gastrostomy tube feeding in children.

Method:
Retrospective observational data were collected on children with a gastrostomy in situ within one Local Health Board. Cases were identified through the record kept by the specialist nurse and were cross checked against a commercial data base of children requiring replacement devices. The study received local approval as an audit.

Results:
69 children (19 years or under) were identified as being fed by gastrostomy (local prevalence of 61 cases per 100,000.) The median age at gastrostomy insertion was 2 1/3 years (range <1/12 - 12 years). The main indications were cerebral palsy (39%), other neurological disorders (13%), syndromes associated with faltering growth (12%), metabolic disorders (7%), congenital heart disease (5.8%) and cystic fibrosis (5.8%).

Detailed surgical data were not found in all cases. Procedures were a percutaneous endoscopic gastrostomy (PEG) or an open surgical approach (with a simultaneous fundoplication in around 1/3 of procedures). Only one laparoscopic procedure was undertaken. Three major early complications occurred; significant haemorrhage (2) and gastro-colic fistula (1). One patient who had died (and was excluded from the main sample) needed conversion to jejunal feeding prior to death.

Conclusions:
The local paediatric population receiving gastrostomy feeding has been defined and described. This contributes to the information available to share with parents, may help shape services and will act as a start for reflection on current practice.

The Use of Aprepitant (EMEND®) in Children with Gastrointestinal Conditions in a tertiary Gastroenterology Centre

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Introduction:
Aprepitant (Emend®), a Neurokinin receptor-1 antagonist has been used effectively in Paediatric practice in Chemotherapy-induced nausea and vomiting in both acute and delayed phases1. Little is known about its use in non-oncology patients.

Aim:
The aim of this study was to evaluate the use and effectiveness of Aprepitant in children with varying underlying gastrointestinal conditions.

Method:
We retrospectively reviewed medical case notes on patients treated with Aprepitant in our institution between November 2004 and March 2011. 39 patients were studied; the median age of the study population was 11 years, range 1-17 years. Patients received doses according to their age, 3 - 10 years (80mg /m2 /dose), 10 - 12 years (80mg per dose) and >12 years (125mg per dose)

Results:
Aprepitant was used in Cyclical Vomiting Syndrome (CVS) (n=15), colitis and enteropathy (n=5), foregut dysmotility (n=2), Pseudo-obstruction (n=3), Intestinal failure (n=3), Post-Nissen's/gastric transposition (n=3), Intractable Nausea/Vomiting (n=3), Coeliac & Crohn's disease and/or nausea & CVS (n=2), others (n=3). 67% (n=10) of patients with CVS had resolution/improvement of symptoms, 40% improved in the colitis and enteropathy group, 50% with foregut dysmotility, there were no responders in Pseudo-obstruction and Intestinal failure. 100% of patients in post Nissen's group responded, whereas there was no response in post gastric transposition group. None responded in the Intractable Nausea/Vomiting group, also there was no response in 2 patients with Coeliac and Crohn's disease. Only 33% responded in the others group.

Summary:
Aprepitant has been used in gastrointestinal patients for nausea, retching and vomiting mostly after trying different other medications like Ondansetron, Granisetron, Omeprazole and Ranitidine or exclusion diets. Cyclical Vomiting Syndrome was the commonest indication of using Aprepitant with a response rate of 67%, the combined response rate to the other conditions was only 41%. The drug was tolerated well with no adverse effects being reported.

Conclusion:
Aprepitant is a safe and efficacious drug in CVS, particularly if used in the prodromal phase of cyclical vomiting. Further randomised controlled trials are needed to study the optimal efficacy of Emend.

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Management of chemotherapy- induced nausea and vomiting.
Indian Pediatr. 2010 Feb;47(2):149-55.

Very Low catheter infection rates are achievable in Paediatric Home Parenteral Nutrition Patients

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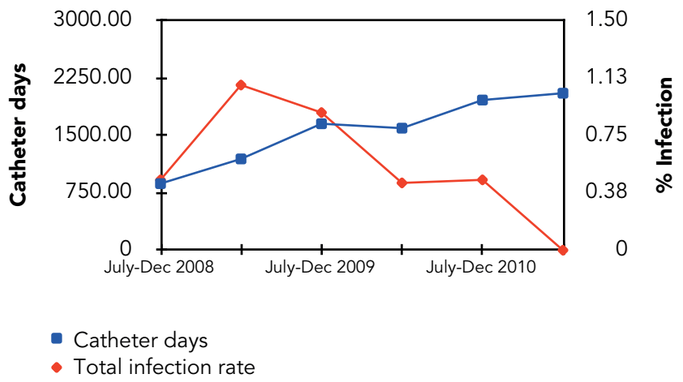
Introduction
A multi-disciplinary team is recommended to facilitate home parenteral nutrition (PN). Prior to May 2008 children requiring home PN were supervised on an adhoc basis. From May 2008 a specialist nurse was appointed to co-ordinate their care, working closely with two consultant paediatric gastroenterologists, paediatric surgeon and specialist pharmacist and specialist dietician to provide care for these children.

Methods
Our aim was to increase the use of home PN and decrease the rate of central line infection. We did this by:
Developing protocols
Training parents and staff on protocols
Specific strategies, such as introducing Taurolock as a routine
Developing individualised plans for patients with recurrent line infections

Results
The use of central venous catheters for home PN increased throughout July 2008 to June 2011. The total number of central line infections and infection rates showed an initial increase and then declined.

Figure 1



Most line infections in 2009 to 2010 were due to repeated line infections in 3 patients. We therefore employed individual strategies to decrease infections rates in these 3 patients which included the use of 70% alcohol locks, manipulation of cyclical antibiotic therapy and stopping gastric acid suppression.

2 patients continued to have repeated line infections despite these interventions and therefore they had a planned line replacement before continuing on this intensive treatment. All 3 patients remain on home parenteral nutrition and have been infection free for more than 6 months.

Conclusion
The use of PN has increased, as shown by the number of home parenteral nutrition catheter days. The initial increase was accompanied by a rise in infection rates but modification of protocols and strategies has enabled us to reduce the infection rate despite an increase in catheter usage. It is possible by using a multi-disciplinary approach, targeted strategies and protocols to manage a home PN service in children that has very low catheter infection rates.

Posters

Audit: Parenteral Nutrition use in children from 2007-2010

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Introduction:
In 2010 the National Confidential Enquiry into Patient Outcome and Death (NCEPOD) published findings of a national audit. This highlighted that many patients were receiving PN with insufficient assessment, and a lack of monitoring to ensure appropriate use. The risk of complications was discussed, with infection singled out as the most common serious side effect (NECPOD 2010). Recommendations were produced by NCEPOD to aid clinicians making decisions about the use of PN.
Previous audits had found that between 2003 and 2006 there was a significant rise in the use of PN, going from 228 PN days in 2003 to 1296 PN days in 2006.
The current audit was conducted in the light of the NCEPOD recommendations, to ensure that, where applicable to children, our service was of a high standard.

Aims:
1. Identify number of children treated with PN from 2007-2010
2. Compare numbers of children with previous audits
3. Review internal reporting system to identify CVC infections in children receiving PN
4. To use reporting systems to look at the long term outcomes for children treated with PN for more than 28 days.
5. To review NCEPOD recommendations and evaluate our service against the self-assessment checklist

Results:
- The current audit showed that PN use has not continued to rise, but has remained fairly constant since 2006
- There were 177 children who received PN in the four year period, of these 35 had CVC infections.
- The number of children receiving PN for more than 28 days peaked in 2007 and since then has reduced
- From 2007 to 2010 36 children received PN for more than 28 days. Of these 16 were started on PN during the neonatal period, 15 were premature babies and 6 were one of twins (this includes one set of twins)
- Review of the NCEPOD recommendations found that they are generally followed within the paediatric unit

Conclusions:
This audit showed that the long term prospects for children who required PN for more than 28 days was generally good. The majority were weaned off PN and discharged on full enteral feeds. Two patients were transferred to adult care for on-going PN. Two young children were discharged on home PN, and subsequently had small bowel transplants. Only two children were still being treated with PN when they died due to their underlying conditions.
Perhaps because children are less likely to require this intervention, when it is required there is a greater awareness and debate about the process.

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Caseload Audit Of The Use Of Pancreatic Enzyme Replacement Therapy (PERT) In The Paediatric Gastroenterology Service, Leicester

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Aims:
1. To examine the reason for PERT use
2. To investigate the biochemical and dietary monitoring in patients on PERT
3. To explore the dosage and length of PERT use.

The dosage of PERT depends on the level of residual pancreatic function and diet consumed (particularly fat and protein). It is calculated per meal or snack or drink according to the amount of fat despite the fact that the enzymes are also needed for protein and carbohydrate digestion. The titration of dosage is assessed from stool output (frequency/colour/consistency) and the presence of abdominal pain.

Method:
A retrospective audit was undertaken of all known cases of PERT. 9 medical and dietetic records (1988 to 2011) were reviewed in terms of PERT dosage, biochemistry, the use of a food diary and length of use.

Results:
At presentation all but 1 case had persistent loose stools and 4 had poor growth and weight gain when faecal chymotrypsin/faecal elastase was undertaken and found to be low. PERT was started. Further tests revealed the diagnosis in 5/9 subjects including Schwachman Diamond Syndrome (2), Angelman's Syndrome (1), Rowinsons Syndrome (1) and Coeliac Disease (1).
Starting dose and maintenance dose of PERT varies greatly as does the period of use. The average starting dose is 3547iu lipase/kg daily (range 1450 – 5000iu lipase /kg) and maintenance dose is 4612iu/kg daily (range 1930 – 10000iu lipase/kg). The majority of cases (6/9) stop using PERT after an average of 1037 days.
1/9 had no dietetic assessment before starting PERT and 4/9 did not have an initial food diary. 1 subject had serum fat soluble vitamins measured on starting PERT. 2/6 of the long term patients were monitored with 7 day food diaries and 4/6 annual serum fat soluble vitamins. All cases were on appropriate fat soluble vitamin supplements.

Conclusion:
PERT is used for a variety of conditions but decision to use is led by symptomatology rather than objective need. In this group pancreatic insufficiency tends to be a temporary condition with only a few patients needing to continue on PERT. Regular faecal elastase and fat soluble vitamin monitoring is suggested to assess the ongoing need for PERT.
Initial assessment should include a 7 day food diary to assess PERT dose and fat soluble vitamin measurement. Detailed review of diet alongside symptomatology is essential to gauge PERT titration.

Citrin Deficiency: variation in phenotype with identical genotype

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Abstract:
Citrin deficiency, due to mutations in the SLC25A13, gene can present in infancy as neonatal intrahepatic cholestasis and/or in adulthood with hyperammonaemia (citrullinaemia type II). It has been well described in the Far East but is now being recognised in other ethnic groups.

We report 7 cases of Citrin deficiency, from 4 unrelated families, of Pakistani origin, presenting to the children's liver and metabolic services in Leeds and London. All are homozygous for the c.1763G>A(R588Q) mutation in the SLC25A13 gene.

Five were investigated for neonatal jaundice. Of these, 4 had raised plasma citrulline, 3 galactosuria, 3 raised phenylalanine on newborn screening, and 3 had low serum albumin and mild coagulopathy at presentation. Two had liver biopsies which showed cholestasis, bridging fibrosis and micro and macrovesicular steatosis. Liver function tests and amino acid profiles normalised in all 5 over 2-18 months. The sixth case is a 10 year old asymtopmatic sibling, identified on family screening, who is homozygous for the same mutation.

A seventh child, initially investigated in 1996, at 6 months of age, for failure to thrive, poor tone and rickets, had hepatomegaly, generalised aminoaciduria and galactosuria. A liver biopsy at 1 year of age showed cirrhosis and focal mild macrovesicular steatosis. The diagnosis of citrin deficiency was only made in 2010 following an infant cousin being diagnosed in London. The mother of this cousin is also homozygous for the same mutation but she has no history of neonatal cholestasis or hyperammonaemia.

All children are well at 1-15 years of age.

Citrin deficiency should be considered in the differential diagnosis of prolonged neonatal jaundice, especially if citrulline is raised or the liver biopsy shows steatosis. However this series suggests a wide variation in clinical phenotype even with the same mutation. Whether patients with this mutation are at risk of hyperammonaemia in adulthood remains to be seen.

Coeliac Disease in Children presenting as Liver disease

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Introduction:
Coeliac disease (CD) is a common condition. The incidence according to our prospective longitudinal study of 14000 children based on CD serology is 1% although by the age of 7 years 90% of them remain formally undiagnosed and possibly asymptomatic (ref.1). There is poor awareness that CD may present with symptoms and signs of liver disease.

Aim:
To document the clinical picture of children presenting to a regional paediatric gastroenterology unit with liver disease as the first manifestation of CD.

Method:
Case notes of children presenting with liver disease to a regional paediatric gastroenterology unit who were subsequently diagnosed as having CD were analysed .

Results:
Between 2004-20011 four children presented with significant liver dysfunction and were subsequently diagnosed with CD. One presented with ascites, transaminitis, hypoalbuminaemia and abnormal clotting which resolved on gluten free diet .Second presented with tiredness and had transaminitis, diagnosed as autoimmune hepatitis requiring prednisolone treatment. In the third, oesophageal varix was noted at the time of endoscopy and liver biopsy confirmed cirrhosis and portal hypertension. The fourth presented with liver failure leading to hepatic encephalopathy requiring her to be listed for an urgent liver transplant. She had no GI symptoms, negative coeliac serology but had a low IgA. CD was only diagnosed after she underwent upper GI endoscopy with biopsy as a part of routine liver transplant work up. She improved on gluten free diet without liver transplant.

Summary:
CD associated liver disease can produce transient transaminitis in up to 40% of cases. Rarely it may present with autoimmune hepatitis; acute liver failure and cirrhosis with portal hypertension. Gluten free diet normalises liver enzymes in most children but some may need additional treatments

Conclusion:
CD should be considered early in differential diagnosis of children presenting with liver disease.

Reference:
Ravikumara M, Nutigattu YKT, Sandhu BK, Ninety percent of celiac disease is being missed. J. Ped. Gastroenterol Nutr. 2007; 45:497-99

Coeliac disease the ESPGHAN way: biopsy not needed?

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Introduction:
Proposed ESPGHAN guidelines recommend that coeliac disease can be diagnosed in patients with a positive history of suggestive symptoms, positive serology markers for coeliac disease and a response to a gluten-free diet without the need for a duodenal biopsy. In contrast, current NICE guidelines advise that a biopsy showing histological features compatible with coeliac disease is required for a diagnosis.

Aim:
To investigate whether using NICE guidelines versus using proposed ESPGHAN guidelines is likely to significantly impact the management of children with suspected coeliac disease.

Method:
A retrospective cohort of 70 children with positive serology for coeliac disease who underwent upper intestinal biopsy were investigated. Results of serology testing and histopathology findings discussed at clinicopathological conference were examined.

Results:
Of the 70 patients, 43 were positive for autoantibodies to both endomysial (EMA) and tissue transglutaminase (tTG); 20 were positive for tTG but not EMA.
A total of 6 patients (9%) had biopsies which were not suggestive of coeliac disease. Two patients with positive serology to both markers were biopsy negative for coeliac disease. One of these patients had a background of Down syndrome and the other was later diagnosed with fructose intolerance. Three children with negative biopsies were EMA positive but tTG negative. These included one child with a low IgA who was IgG EMA positive and one child with insulin-dependent diabetes mellitus. One patient with a biopsy that was not suggestive of coeliac disease was EMA negative and tTG positive. This child went on to show improvement with a gluten-free diet.

Conclusion:
This small study examined children with positive serological markers for coeliac disease who under NICE guidelines warranted upper GI endoscopy and biopsy. Almost 10% showed histopathological findings that were not compatible with coeliac disease. We suggest that using the proposed ESPGHAN guidelines could potentially prevent unnecessary biopsies in a substantial number of children

Hyperplastic polyposis syndrome in childhood – an incidental finding with consequences

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Introduction:
Endoscopy and autopsy findings in the adult population commonly confirm the presence of hyperplastic polyps in the recto-sigmoid colon. Hyperplastic Polyposis Syndrome (HPS) however, is a rare condition almost exclusively described in the adult literature (1) and first defined by WHO criteria in 2000 (2). Modification of the WHO criteria has been suggested to improve the identification of patients who have increased risk of malignancy (3). In the largest adult multicentre cohort study to date (4) colorectal cancer (CRC) was detected at initial endoscopy in 22 out of 77 patients with HPS.

Method:
LW, a 15 year old girl, presented to our department with a one-year history of abdominal pain and intermittent rectal bleeding. We performed colonoscopy and histological work up of all detected polyps. Literature was reviewed for evidence of best management and surveillance of paediatric patients with HPS.

Results:
Past medical history, extended family history, physical examination and investigations of blood and stool samples were unremarkable. Colonoscopy revealed six small dew-drop like lesions (3-5mm), which were biopsied and one large caecal polyp (20 mm), which was snare resected (see images). Histology of these polyps revealed hyperplastic changes in the epithelium with serration and proliferation with numerous mitoses but no nuclear atypia, consistent with the diagnosis of HPS of the proximal colon.

Conclusion:
HPS is an extremely rare condition, scarcely mentioned in the paediatric literature and unknown to the vast majority of paediatric gastroenterologists. Following recent publications of case series and a multicentre cohort study of adult patients (4) gastroenterologists generally aim for complete resection of all right sided lesions.
No consensus or guideline is available for paediatric patients with HPS, however evidence from adult data suggests surveillance endoscopy one to three yearly depending on the size of hyperplastic polyps and presence of adenomatous or neoplastic lesions (5).

References:
1. Williams GT et al: Metaplastic polyps and polyposis of the colorectum. Histopathology 1980; 4: 155-70
2. Burt RW, Jass JR: Hyperplastic Polyposis. WHO classification of tumors. Lyon, IARC Press 2000: 135-6
3. Kalady et al: Hyperplastic polyposis syndrome: a call for broader diagnostic criteria. Hereditary Cancer in Clinical Practice. 2010; 8(S1) P8
4. Boparai et al: Increased colorectal cancer risk during follow-up in patients with hyperplastic polyposis syndrome: a multicentre cohort study. Gut. 2010; 59: 1094-1100
5. Ferrandez et al: Phenotypic Characteristics and Risk of Cancer Development in Hyperplastic Polyposis: Case Series and Literature Review. Am J Gastroenterol 2004; 99: 2012-2018

Paediatric Cardiac transplantation and the Gut: Histological intestinal changes in patients with Post-transplant lymphoproliferative disorder (PTLD)

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Introduction:
PTLD is a recognized complication after heart transplantation (HT) with manifestations including the GI-tract (GIT). EBV infection is central role in the pathogenesis.

Aim:
Our aim was to describe symptoms and intestinal mucosal changes and their resolution after treatment.

Method:
We report 4 patients with PTLD post HT. All patients had received standard post-transplant treatment with Tacrolimus, MMF, steroids.

Results:
Patient 1 underwent HT aged 12 years. 4y later, she presented with anaemia and hypoalbuminaemia. EBV PCR DNA was negative. Her initial endoscopy was normal, her second endoscopy 6 months later showed deep infiltrations of jejunum/ileum with lymphocytes, plasma cells and large blasts suggesting EBV negative PTLD, with CD20 and CD79a positive cells. She received Rituximab/COPP and is now well on low levels of Tacrolimus. Patient 2 underwent HT aged 15 months. 4 months later he presented with anaemia/meleana. He had EBV viraemia. Endoscopy showed nodules in stomach/duodenum, histology revealed a dense lymphocytic infiltrate including large cells with features of immuno-blasts and CD20 and CD79a positive cells. There was strong EBV positivity within the infiltrate, suggestive of diffuse large B-cell lymphoma. He received Rituximab with reduction of Tacrolimus achieving complete PTLD remission as indicated by a repeat endoscopy 5 weeks later. 6 months later he developed PTLD recurrence without GIT involvement, he received Rituximab/COPP. 2y after treatment he remains well. Patient 3 underwent HT aged 14y. Four months later she developed a sore throat, intermittent fevers, abdominal pain, vomiting and a painful groin lump. EBV PCR DNA was positive. Node biopsy revealed large B cells and plasma cells strongly positive for EBV. PLTD diagnosis was established and the patient received Rituximab, with Tacrolimus reduction. The lesions resolved, but abdominal pain and vomiting persisted. An endoscopy revealed diffuse lymphocytic infiltrates of the gastric body, mostly with B-lymphocytes and EBV positive staining. She received COPP, we are currently awaiting treatment outcome. Patient 4 underwent HT aged 2y. 6y later she presented with watery diarrhea, foul smelling mucousy stools. EBV PCR DNA was positive. Histology of the GIT revealed increased plasma cells in the gastric body, mononuclear infiltrates with plasmatocytoid differentiation in the small bowel mucosa and TI and lymphoid tissue infiltration with eosinophils in the caecum/ascending colon. Plasmacytoid cells were strongly CD79a positive and CD20 negative and the immunostaining for EBV was negative. She received no treatment, other than reduction of Tacrolimus and is currently well.

Conclusion:
PTLD is a serious complication after HT which involves the GIT, mainly stomach and small intestine. Persistent GIT symptoms, even after a longer period post HT, warrants early assessment.

Quality assurance of a regional service for the management of jaundice in the neonatal and early infant period

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Jaundice in the neonatal period is common, reflecting normal physiological adaptation to extrauterine life. The differential diagnosis of "physiological jaundice" includes the "neonatal hepatitis syndrome", which is a heterogeneous group of diseases. The goal of management is to identify specific causes where possible, and to provide suitable treatment in a timely fashion to avoid progression to chronic liver failure. One particular cause associated with fatal outcome without treatment is that due to extrahepatic large duct biliary obstruction (EHLDO). This has a prevalence of 1 in 15000 and an incidence of 50 new cases per year in the UK. The preferred treatment comprises surgical relief of biliary obstruction. Long term prognosis is determined by the age at which the obstruction is relieved; prognosis is poor if this is delayed beyond 100 days. Prognosis for salvaging liver function also depends on the experience of those who manage this condition. Consequently, in the United Kingdom, infants with EHLDO are managed at only three specialised centres. The identification of patients with EHLDO from the relatively large number of infants with physiological jaundice or other types of neonatal hepatitis at a sufficiently early stage is considered a "high stakes" activity. Gastroenterologists and pathologists who do not work at the referral centres must remain vigilant for cases of a rare disease with a low incidence, which require early referral to maximise their chance of a successful outcome.

The diagnosis of liver disease requires multidisciplinary cooperation at clinicopathological conference (CPC), particularly in the neonatal and infant period. Histological examination allows classification into a relatively small number of morphologic categories. Precise diagnosis requires ancillary investigation using other modalities. If the histological appearances suggest EHLDO, then urgent referral for surgery is required. Gastroenterologists retain the prerogative to refer for second opinion regardless of histology. Patients without features of EHLDO are typically managed by local gastroenterologists. Histological assessment assists in the diagnosis but the investigation of non-obstructive liver disease requires a relatively broad range of ancillary investigations. A variety of possible aetiologies are sought, some of which might become apparent only as the condition evolves. The referral of cases to tertiary centres, together with the histological slides, provides a means to audit the performance of the regional centre in the diagnosis of EHLDO. Long term surveillance of those not referred, or the opinion of pathologists at the tertiary centres in the case of histological slides referred without the patient provides a means to audit the performance of management of neonatal hepatitis without EHLDO.

A search of electronic archives identified 146 specimens of liver submitted over a 12 year period. Of these, 31 cases were taken during the first 6 months of life under circumstances which raised EHLDO as a plausible differential diagnosis. 6 cases were referred to tertiary centres (together with histological slides) for suspected EHLDO. Concurrence in diagnosis was present in every case. In 4 cases, the histological slides were forwarded for second opinion. Concurrence in opinion was present in every case. None of the remaining patients developed features that suggested that EHLDO had been missed. The eventual diagnosis was the same as the original one in 7 cases. The clinical features of hepatitis resolved in 14 cases. In 5 cases, a specific diagnosis became apparent through ancillary investigation, or from the emergence of more specific features in due course. No cases were identified in which the diagnosis of EHLDO had been delayed by inappropriate interpretation of histological appearances of a liver biopsy specimen combined with discussion at CPC. No cases were identified in which the subsequent clinical progress suggested that the conclusions drawn at the time of initial assessment were incorrect. This study suggests that safe practice is possible in a regional centre providing the hepatologist works closely with the pathologist at regular CPC and the pathologist seeks second opinion from the tertiary centre in challenging cases.

Quality outcome measures in children with Coeliac Disease – a regional approach

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Introduction:
The East of England Paediatric Gastroenterology Network (EEPGN) was established in 2009. It includes two core centres, Addenbrooke's Hospital (AH) and Norfolk and Norwich University Hospital (NNUH) and a network of 15 other hospitals across the East of England. Currently there are no nationally recommended quality outcome indicators (QOI) for children with coeliac disease. As this is a life-long condition requiring adherence to a strict gluten-free diet to minimise long term complications, follow up should support compliance through education and monitor patients for complications or associated conditions.

Aim:
To develop and audit a series of quality measures for service and outcome in coeliac disease. These measures should be applicable to different clinical service models and provide an initial benchmark for clinicians and commissioners.

Method:
Following acceptance of a regional coeliac pathway in 2009, the EEPGN recognised a need for QOI to improve quality of care and equity of access across a large network.
Based on national guidelines and local best practice, the network developed QOIs by multi-professional consensus. Final outcome measures ranged across 4 domains, each with different standards: 6 'diagnostic', 10 'annual review', 2 'transition' and 2 'patient satisfaction'. A patient satisfaction questionnaire was devised in conjunction with the audit department at NNUH and used at both centres. A proforma was developed by the network steering group to standardise data collection. Audit standards were initially set to meet targets of 90% (except for children with 'normal tTG within two years of diagnosis' (75%) and 'satisfaction with clinic' (75%).
The study was carried out at NNUH and AH. Both centres hold monthly annual review clinics for children with coeliac disease. At AH patients are seen by a Dietitian and Gastroenterology Specialist Nurse, at NNUH they are seen by a Dietitian and Consultant. Over a six month period (November 2010 – May 2011) both units undertook prospective data collection on all coeliac patients seen for annual review.
To assess diagnostic standards, a retrospective audit of all registered, biopsy-proven, children with coeliac disease was undertaken using medical records.

Results:
A total of 99 patients with coeliac disease were seen during the study period (50 NNUH; 49 CUH – age range 1.6years – 16.8 years. The audit was carried out in two centres to allow comparison of two different service models. There was no significant difference in results between the two centres. Diagnostic standards were met in 94.5% children, annual review standards in 99%, transition standards in 95% and patient satisfaction standards were met in 98% children. The lower score in diagnostic standards represents the difficulty one site had in ensuring timely access to biopsy.

Conclusion:
This audit demonstrated that QOIs for coeliac disease in children, set by a regional network, were met in almost all cases, exceeding initial audit standards. This data has since supported a business case to improve a diagnostic pathway. Multi-centre, multi-professional collaborative working has allowed development of some standardized quality outcome measures for coeliac disease. Given the increasing focus on outcome-based commissioning, it is important that clinicians actively engage in providing clinically-relevant outcome measures.

Rapid reintroduction diet (3 day) versus slow food reintroduction (5 week) in the maintenance of remission after exclusive enteral nutrition in paediatric Crohn's disease.

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Introduction:
Exclusive enteral nutrition (EEN) has been shown to be effective treatment in paediatric Crohn's Disease (CD). To the best of our knowledge there is currently no single recommended method for food reintroduction in paediatric CD patients after EEN and practice appears to vary between centres.

Aim:
To assess the difference between slow food reintroduction (Slow FR) and rapid food reintroduction (Rapid FR) after induction of remission with EEN.

Method:
In this retrospective study the duration of maintained remission was compared in two groups of paediatric CD patients. Both groups of paediatric patients completed EEN for 6-8 weeks. Group 1 (n=17) embarked on a Slow FR (5 week food reintroduction with new foods reintroduced daily). Group two (n=18) followed a Rapid FR for 3 days before re-establishing normal eating.

Results:
9/17 patients in the Slow FR group and 8/18 patients in the Rapid FR group relapsed in the first year after EEN. There was no statistically significant difference in the number of relapses between the two groups in the first 12 months after EEN (P=0.09).

Conclusions:
We thus propose a Rapid Food Reintroduction process for paediatric CD patients in the hope of preventing possible nutritional inadequacy resulting from a restricted diet. A more convenient and shorter reintroduction may also improve compliance without adverse affect on outcome.

Recurrent Staphylococcal infection and IBD

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Introduction:
The role of staphylococcal superantigen in IBD has been considered but not well described. This case had recurrent staphylococcal infections and confirmed IBD on endoscopy needing full treatment. Her IBD symptoms were exacerbated with staphylococcal infections.

Aim:
Highlight complexity of the case and the complications

Method:
A 15 years old girl presented with multiple abscesses of arm needing drainage and drained 30 ml of fresh pus. A course of antibiotics was given and usual causes of multiple infection including diabetes etc were excluded . She however continued to have recurrence of serious infections for a year and most of these were staphylococcal skin infections. She has also developed a vasculitic rash on lower legs with ankle swelling and was investigated. No clear cause identifiable. She also had prolonged diarrhoea and endoscopy revealed inflammatory bowel disease. She had E028 liquid nutritional therapy and high dose steroid followed by prednisolone maintenance. There has been suggestion about IBD been triggered by Staphylococcus sepsis and she has been on maintenance Azathioprine. She continued to have several other serious infections including mastoiditis on left side needing mastoidectomy in May 2009 (mastoid cavity grew Pseudomonas Aeruginosa & Klebsiella pneumoniae). Neuroimaging revealed a small subperiosteal collection. She has been on various antibiotics and during the process. She has developed axonal peripheral neuropathy of sensory nerves affecting both the legs and it was thought to be related to Linizolid. She has been on Amytryptilin and Gabapentin for that. She also had fungal infections including Candida Glabrata sensitive to fluconazole and she was started on Fluconazole prophylaxis after full treatment.

Results:
- Reduced mannan binding lectin
- Normal extended lymphocyte subsets with a normal percentage classed sweet memory b cells
- TOLL receptors appear normal – TOLL like receptor4 (LPS) and TOLL like receptors 7 and 8 (CL097)
- T cell receptor – V Beta no significant abnormality
- Normal response to PHA and OKT3
- Previous normal immunoglobulin (IgG 10.5g/L, IgA 2.3g/L, IgM 0.67g/L)
- Previous normal vaccine response to tetanus,HIB,pneumococcal
- Normal complement pathway
- Persistently positive pANCA

Conclusion:
Association of staph infections and IBD has been considered but aetiological correlation not well established in literature. We would like to present this case that had complex presentation and tortuous course.

Reference:
Chiba M, Hoshina S, Kono M, Tobita M, Fukushima T, Iizuka M, Watanabe S. Staphylococcus aureus in inflammatory bowel disease. Scand J Gastroenterol. 2001 Jun;36(6):615-20.

Sensorineural hearing loss in gastrointestinal disorders

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Introduction:
Sensorineural hearing loss and enteropathy is a known association in mitochondrial diseases, while other conditions that linked sensorineural hearing impairment to gastrointestinal disorders are infrequently described outside the context of syndromes. Few case reports are published describing an association between celiac disease and inflammatory bowel disease with sensorineural hearing loss. To the best of our knowledge there was no study looking at the association between gastrointestinal diseases and sensorineural hearing loss.

Aim:
To describe the various gastrointestinal disorders in children with combined sensorineural hearing loss and gastrointestinal diseases.

Method:
We describe a series of 29 patients with variety of gastrointestinal disorder and sensorineural hearing loss who presented to our department between 1996 and 2011.

Results:
18 patients (62%) were males and 11(38%) were females. 7 patients (24%) had GI motility disorders (5 congenital dysmotilies and 2 slow transit constipation) while another 7 (24%) had Food allergy. Of those with food allergy 6 patients have increase in esinophils counts in their biopsies mainly in the colon
5 patients (17%) were part of syndromes; three of them were CHARGE syndrome.
The rest of the group, 10patients (34%) were divided equally between mitochondrial disorders, cerebral palsy, malignancies and autoimmune enteropathy.
Although gastrointestinal involvement in children with sensorineural hearing loss tend to fall under the umbrella of autoimmune and mitochondrial disorders, we describe a stronger association between food allergy in particular eosinophilic enterocolitis and gastrointestinal motility disorders with sensorineural hearing loss. A larger study to look into the link between sensorineural deafness and the gut is warranted.

Space-time Clustering of Childhood Inflammatory Bowel Disease Suggests influence of Environmental Factors on Disease Expression

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Introduction:
There has been an increase in the incidence of Paediatric Inflammatory Bowel Disease (IBD) in Northern England as there has in the rest of Europe over the past two decades.
We sought evidence for environmental influences that may have contributed to this increase by looking for clustering of birth and diagnosis in time and space within our patient population.

Aim:
Seek evidence for space-time clustering of new diagnosis of childhood IBD

Method:
All patients with new diagnosis of childhood IBD living in the North of England (corresponding to old Northern region Health Authority excluding Darlington and Tees Valley) were eligible for inclusion. Patient lists were compiled from January 2003 to August 2009 using the databases of the Paediatric Specialist IBD Nurses covering this patient population and cross checked with other record sources to ensure complete ascertainment. Space-time clustering was examined using 4-digit grid references of centroid of postcode, DOB or date of diagnosis by K function analysis with nearest neighbour (NN) threshold adjustment to allow for differences in population density between areas.

Results:
214 cases of IBD identified. 3 cases who only had Orofacial Granulomatosis and 4 cases with incomplete data were excluded from the analysis. There was evidence of significant space-time clustering based on date of birth and place of residence at diagnosis for Crohn's Disease (CD) and all IBD diagnosis, but not for Ulcerative Colitis (UC)alone (see table).

Table: K function analysis with NN threshold correction

Diagnosis	Date of birth and place of residence at diagnosis	Date of diagnosis and place of residence at diagnosis
UC n = 63	p = 0.125	p = 0.593
CD n = 124	p = 0.042	p = 0.517
All IBD n = 207	p = 0.030	p = 0.757

Conclusions:
Date of birth and place of residence at diagnosis were associated with clustering of IBD in childhood, particularly CD. This suggests that environmental factors influence the expression of IBD in susceptible individuals.

Transaldolase deficiency presenting as neonatal liver failure – a case report

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Transaldolase deficiency is a recently recognised metabolic error in the pentose phosphate pathway. To date eleven patients has been described in seven families worldwide. Presentations have ranged from neonatal hydrops through neonatal liver failure to slowly progressive liver cirrhosis.

We report an infant who presented with neonatal liver failure, cardiomegaly, high lactate and thrombocytopenia in the first few days of life. Initial investigations including bone marrow aspirate for haemophagocytosis failed to find a cause, however she gradually improved over the next few weeks. She was readmitted at 2 months of age because of poor weight gain, hepatosplenomegaly, ascites, gall stones and nephrocalcinosis. Further investigations particularly aimed at excluding mitochondrial disorders were normal. Urine was sent for examination of polyols and demonstrated significant excretion of erythritol, arabitol, ribitol, and sedoheptulose, which is highly suggestive of transaldolase deficiency. The enzyme assay on skin fibroblasts showed severely deficient transaldolase activity. A mutation was identified in the *TALDO1* gene.

Unfortunately she had a rhinovirus infection causing severe pulmonary haemorrhage leading to cardiorespiratory arrest. Following this care was withdrawn in the intensive care unit.

We present this case to highlight another cause of neonatal liver disease. Measurement of urinary polyols should be considered in infants with liver disease associated with cardiac, renal or haematological abnormalities.

Bleeders Come first: How common and who deals with it?

Mr Simon Huddart, Consultant Paediatric Surgeon, University Hospital of Cardiff, Heath Park, Cardiff

Acute gastrointestinal bleeding, both upper and lower, is a rare emergency in children. Three patterns can be seen:
a; Those patients known to have hepatic disease and portal hypertension, where acute blood loss is likely to be from oesophageal varices. Such patients are best treated within the regional liver service.
b; Steady bleeding which settles on conservative management, but may require endoscopic diagnosis or therapy.
c; Severe bleeding requiring open surgery for control - very rare and (Meckel's excepted), a surgeon would still require endoscopic siting of the source of blood prior to surgery.

Gastroenterologists are the acknowledged experts in diagnostic and therapeutic upper and lower GI endoscopy so it would make sense that such expertise is utilised in an emergency. Paediatric surgeons receive little training in upper GI endoscopy, and essentially none in lower GI endoscopy. However, many centres are unable to provide a 24/7 OOH gastroenterology service.

The incidence and causes of upper and lower GI bleeding, along with possible management pathways will be discussed.

Managing Severe Gastrointestinal bleeding in children

Dr Patrick McKiernan, Consultant Paediatric Hepatologist, Liver Unit, Birmingham Children's Hospital, Steelhouse Lane, Birmingham

ABSTRACTS FOR FRIDAY 27TH JANUARY 2012

Mead Johnson Symposium
Invited Speakers'
Oral Plenary Session

Cost-effectiveness of using an extensively hydrolysed formula compared to an amino acid formula in the initial treatment of cow milk allergy in the community in the UK

Prof Julian F Guest, Catalyst Health Economics Consultants, Northwood, Middlesex UK and School of Biomedical Sciences, King's College, London, UK.

Abstract

Objective:
To estimate the cost-effectiveness of using an extensively hydrolysed formula (eHF; Nutramigen) compared to an amino acid formula (AAF; Neocate) as first-line treatment for cow milk allergy (CMA) in the community in the UK, from the perspective of the National Health Service (NHS).

Method:
A decision model was constructed depicting the treatment paths and associated resource use attributable to first-line management of CMA with the two formulae. The model was based on the case records of 145 AAF-treated patients and 150 matched eHF-treated patients from the THIN database (a nationally representative database of patients registered with general practitioners (GPs) in the UK). The model estimated the costs and consequences of patient management over 12 months following their initial GP visit for CMA.

Result:
Patients with a combination of gastrointestinal (GI) symptoms and eczema accounted for 44% of the cohort. Those with GI symptoms alone and eczema alone accounted for a further 39% and 13% respectively. Those with urticaria and failure to thrive accounted for <5% and <7% of all patients respectively. There were no differences in the distribution and severity of symptoms (using the incidence of reflux, failure to thrive and anaphylaxis as a proxy), age and body weight between the two groups. Patients' age and weight at presentation was a mean 2.6-2.8 months and 4.4kg respectively. It took a mean 2.2 months for a formula to be prescribed after the initial GP visit. Time to symptom resolution after starting treatment with eHF and AAF was a mean 1.2 months in both groups, hence the mean number of symptom-free months during the 12 months following the initial GP visit was an estimated 8.6 months in both groups. Patients treated with an eHF had a mean 13.1 GP visits over the 12 months compared to 17.5 visits made by AAF-treated patients ($p<0.001$). The NHS cost of managing a CMA infant over the first 12 months following initial presentation to a GP was estimated to be £1,853 and £3,161 for an eHF-treated and AAF-treated patient respectively.

Conclusion:
First-line treatment of newly-diagnosed infants with CMA with eHF instead of AAF affords a cost-effective use of NHS resources. Moreover, in the absence of published evidence showing superiority of one formula over the other, the eHF is the preferred first-line treatment in newly-diagnosed infants receiving their first clinical nutrition preparation, except in the more severe cases.

Gastro oesophageal relux - an update

Professor Colin Rudolph, Vice President for Global Medical Affairs and Chief Medical Officer, Mead Johnson Nutrition c/o Ms K Goans, MJN, 2400 West Lloyd Expressway, Indianopolis

The 2010 NASPGHAN/ESPGHAN guidelines provided an updated approach to the management approach to several symptoms and signs of GERD. This presentation will focus on these changes including a discussion of the reliability of symptoms in infants and children to guide treatment of GERD; the use of biopsy for diagnosis of GERD; and the role of GERD in respiratory tract disorders.

Post infectious irritable bowel syndrome

Professor Robin Spiller, Professor of Gastroenterology, Nottingham Digestive, Diseases Centre & NIHR Biomedical Research Unit, University of Nottingham

Post Infectious Irritable Bowel Syndrome (PI-IBS) represents a natural experiment and provides a model for understanding mechanisms applicable to all IBS. Gastrointestinal infection is one of the strongest risk factors for developing IBS with a relative risk of 7.3 of developing IBS in the year following gastroenteritis compared with uninfected individuals1. PI-IBS accounts for about 1 in 5 of all IBS patients and is predominantly of the IBS with Diarrhoea (IBS-D) phenotype 2. Risk factors include the severity of the initial insult, the mucosal response, age and gender and adverse psychological factors. There is evidence of persistent low grade inflammation and increased gut permeability 3. There is also evidence of increased serotonin availability at the mucosal level with impairment of the serotonin transporter which is likely mediated by inflammatory cytokines. 5HT3 antagonists are logical and successful therapies for this condition as well as IBS-D.

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Should we exterminate *H. pylori*

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H. pylori should be treated in patients with peptic ulcer disease and low grade marginal zone lymphoma of the stomach. These conditions resolve following treatment in the vast majority of cases and do not recur. Best regimens for treatment are changing but where primary clarithromycin resistance is below 15% (as in most of the UK) clarithromycin-containing triple therapy is still most commonly used.

The most recent European "Maastricht" guidelines (as yet unpublished) also recommend *H. pylori* testing and where positive treatment in: young and middle-aged adults in the community presenting with dyspepsia and without "alarm" symptoms; functional dyspepsia (which will resolve in 1 in 12 treated cases); iron-deficient anaemia or vitamin B12 deficiency where no other cause is identified; and adult idiopathic thrombocytopaenic purpura.

At a population level, the biggest health problem attributable to *H. pylori* is gastric adenocarcinoma, the second biggest cancer killer worldwide with about 0.8M deaths/year. Worldwide extermination of *H. pylori* would save at least 80% of these deaths in the future. Vaccine programmes have still not delivered, but treatment of infected individuals as children or young adults, before they develop gastric atrophy, will prevent gastric cancer. After this stage treatment does not abolish the cancer risk but may reduce it to some extent - although it is difficult to demonstrate this from the current medium term follow-up studies.

Controversial studies suggest a possible weak contribution of *H. pylori* to atherosclerotic diseases and to low weight and height in childhood; however causality is hotly debated.

There has been much recent interest in whether *H. pylori* might protect against some diseases. After all, it has co-evolved with humans throughout our evolution and our physiology and immunology have adapted to it. The absence of *H. pylori* from the human stomach for the first time in our evolutionary history may create a change to which we cannot perfectly adapt. Thus there is now considerable evidence that the absence of *H. pylori* has contributed to the rise in oesophageal reflux-related pathologies, including oesophageal adenocarcinoma. More controversial but increasing evidence suggests that *H. pylori* absence may be contributing to the rise in asthma and atopy, particularly in childhood, through a major contribution to the hygiene hypothesis.

The current balance of evidence supports extermination of *H. pylori* from communities to reduce the incidence of gastric cancer, peptic ulcers and possibly other diseases. It seems unlikely that disadvantages will outweigh these advantages. However, we need to know the risks and benefits in order to plan sensible population management strategies in countries where *H. pylori* is still common.

Reference

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Paediatric Gastroenterology in Facing the Future: Standards for Paediatric Services

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Paediatrics in the UK is facing huge challenges staffing hard pressed services. One way to argue for more paediatricians is to set standards. I will refer to general and specialty paediatrics and draw on the experience of the Mid-Staffordshire inquiry and the Royal College of Paediatrics and Child Heath's *Facing the Future* publication in discussing the way forward

Management of really sick patients with anorexia nervosa (MARSIPAN)

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The Junior MARSIPAN report extends the work of the MARSIPAN (www.rcpsych.ac.uk/files/pdfversion/CR162.pdf) report providing guidance for the care of seriously ill adults with anorexia nervosa (AN). Separate guidance for children and adolescents was developed for a number of reasons: risks in young people differ; admission of children and adolescents with AN to paediatric wards is a relatively common event (partly reflecting service organisation, and partly changes in treatment approach over recent years); specialist Eating Disorders services for young people are variable in form, nature and availability; the legal and ethical issues surrounding treatment are multifaceted in young patients, and the role of parents is central.

The report covers the following areas: risk assessment, examination and investigations, location and transition of care, and management issues in specific settings, such as on paediatric wards. One of the most controversial areas was around the management of refeeding in high risk patients. The need for coordinated care between medical and mental health services for this high risk group is core to the report's recommendations.

What is new in network commissioning?

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Advances in feeding neonates in NNU

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Feeding and Nutrition on the Neonatal Unit can be challenging for all infants, but especially so for those undergoing gastrointestinal surgery.

The principle aim of preterm nutrition support is maintaining adequate growth in infants who are born with little nutritional reserve and have higher energy and protein requirements than their term counterparts. As a result they are poorly equipped to deal with the degree of nutritional compromise that often accompanies gastrointestinal surgery.

As with all infants who undergo gastrointestinal surgery, the success of post-operative feeding is very much dependent on the indication and site of surgery, the degree of gut resection, the position of any stomas and the quality of the remaining bowel.

If available, fresh maternal breastmilk is encouraged as first post-operative feed in all infants. In cases of an isolated perforation with a low stoma, post-operative feeding should be well tolerated. If maternal breastmilk is unavailable, then evidence supports the use of an appropriate preterm formula. This avoids the need for specialised term formulas, which fail to meet the preterm's nutritional requirements. However in some extremely low birth weight infants (<1000g), despite the presence of a low stoma, there may still be signs of functional Short Bowel Syndrome.

Approximately 15% of infants who require surgery for Necrotising Enterocolitis (NEC) develop Short Bowel Syndrome. These infants will require longer term Parenteral Nutrition (PN) and therefore be at high risk for PN related liver disease (PNRLD). Use of third generation lipids (SMOF), minimal enteral feeding and the regular recycling of stoma losses can all be helpful in minimising the risks of PNRLD. However, the use of other protective measures such as cyclical PN are difficult to implement in this population due to low gestation, body weight and labile glycaemic control.

Where maternal breastmilk is not tolerated, a hydrolysed protein feed is indicated in preference to elemental amino acid based formulations. This is due to the enhanced gut adaptive properties of hydrolysed proteins compared to those of amino acids. Hydrolysed feeds with a percentage of fat as medium chain triglycerides (MCT) are indicated where there is a risk of cholestasis or other liver disease.

Specialised hydrolysed and amino acids feeds are designed to meet the nutritional requirements of term infants, and so will often require concentration to meet the energy and protein needs of a preterm infant. Concentrating these feeds may still not meet all the micro-nutrient needs of the preterm infant and additional hyperosmolar supplementation will often be required.

For infants with high stoma outputs who are not tolerating standard bolus feeds, the use of continuous feeds is advocated. The infusion of small volumes of feeds, especially maternal breastmilk, can be problematic so specific enteral feeding syringe driver feeding pumps are recommended. When using continuous feeds early oral feeding experiences will be compromised. Hence early intervention from appropriately trained Speech and Language Therapists can be helpful in minimising long-term consequences to feeding. In order to achieve optimal nutrition and feeding outcomes in post-surgical neonates, input from a broad multi-disciplinary team is essential.

Achieving Millennium Development Goal 4 targets: what will it take?

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The millennium development goal (MDG) 4 targets reduction in child mortality by two thirds from 1990 base figures by the year 2015. Although worldwide mortality in children younger than 5 years has dropped from 11·9 million deaths in 1990 to 7·7 million deaths in 2010, consisting of 3·1 million neonatal deaths, 2·3 million post-neonatal deaths, and 2·3 million childhood deaths (deaths in children aged 1–4 years), many countries will not be able to achieve MDG 4 targets at current rates of reduction. It is estimated that a third of all deaths in children under 5 years occur in south Asia and 49·6% occur in sub-Saharan Africa, with less than 1% of deaths occurring in high-income countries.

Recent estimates also provide insight into causes of child deaths. Of an estimated 8·8 million deaths in children younger than 5 years worldwide in 2008, infectious diseases caused 68% (5·970 million), with the largest percentages due to pneumonia (18%, 1·575 million, uncertainty range [UR] 1·046 million–1·874 million), diarrhoea (15%, 1·336 million, 0·822 million–2·004 million), and malaria (8%, 0·732 million, 0·601 million–0·851 million). 41% (3·575 million) of deaths occurred in neonates, and the most important single causes were preterm birth complications (12%, 1·033 million, UR 0·717 million–1·216 million), birth asphyxia (9%, 0·814 million, 0·563 million–0·997 million), sepsis (6%, 0·521 million, 0·356 million–0·735 million), and pneumonia (4%, 0·386 million, 0·264 million–0·545 million). In 2008, nearly half (49%) of all child deaths occurred in five countries: India, Nigeria, Democratic Republic of the Congo, Pakistan, and China.

As part of the Countdown to 2015 for Maternal, Newborn, and Child Survival (CD) we reviewed progress from 1990 to 2010 in coverage of 26 key interventions in 68 countries accounting for over 90% of maternal and child deaths globally. Of the 68 CD priority countries, 19 were on track to meet MDG 4, in 47 there was acceleration in the annual rate of under five mortality reduction, while in 12 countries there has been a deceleration of progress since 2000. Progress in reducing neonatal deaths is limited and in most CD countries maternal mortality remains high with little evidence of progress. There are wide and persisting disparities in the coverage of different interventions between and within countries, but some have successfully reduced long standing inequities.

These recent data provide evidence from several countries showing that rapid progress is possible and that focused interventions and targeting can reduce inequities related to socioeconomic status and gender. However, much more can and should be done to address maternal and newborn health and improve coverage of interventions related to family planning, care around childbirth and case-management of childhood illnesses. This presentation will also present a framework for innovations to accelerate progress and achieve MDG4 and 5 targets globally, especially those that relate to delivery strategies to poor and marginalized populations

Pancreatitis in Children Presenting to a Tertiary Paediatric Gastrointestinal Centre

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Introduction:
Pancreatitis is a rare disease in Children and the diagnosis is often delayed. It is defined as the histological presence of inflammation within the parenchyma of the pancreas. Acute pancreatitis is a reversible process; by contrast chronic pancreatitis causes irreversible damage.

Aim:
To report the different clinical presentations of children with pancreatitis and suggest a diagnostic work up and look at genetic predisposition.

Method:
Retrospective review of case notes of patients with pancreatitis between 2004 to 2010, presenting with raised levels of either Serum Amylase under Gastroenterology. Presenting symptoms, age, laboratory investigations, pharmacological therapies, hospital stay and outcome were evaluated from the medical records. 20 children were identified with raised Amylase. Male n=14, mean age of presentation 11.02 years, range 1.2 year to 17.3 years.

Results:
The commonest complaint was mild to severe abdominal pain in 18 (90.0%) children, 4 (22.2%) had additional back pain. Vomiting was seen in 12 (60.0%), 9 (45.0%) had loose stool, per rectal bleeding with mucus stool in 5(25.0%), poor weight gain in 4 (20.0%), paleness 4 (20.0%), ascites 3 (15.0%), jaundice 2 (10.0%), fever 2 (10.0%) arthritis 2 (10.0%), fast breathing 2 (10.0%) and miscellaneous 3 (15.0%).The exact aetiology was not found in 7 (35%) cases, among them 3 (43.0%) had Crohn's disease, 2 (28.0%) had ulcerative colitis and 2 (27.0%) had drug induced pancreatitis. 3 (15.0%) patients had gall bladder/ common bile duct stones, 2 (10.0%) Azathioprine induced pancreatitis. Other aetiological agents were parenteral nutrition, pancreatic pseudocyst, pancreas divisum, pancreatic tumour, diabetes mellitus and Varicella zoster virus. Genetic analysis was carried out in 5 cases, 1 patient was diagnosed with Hereditary Pancreatitis (PRSS1 gene positive) another patient had sequence variations of the PRSS1 gene. Imaging studies (Abdominal Ultrasonography (US), Computer tomography (CT), and Magnetic Resonance Image (MRI)) were performed. All patients had US, with positive findings seen in 11 (55.0%) cases (gall bladder stones (3), gall bladder sludge (2), cholecystitis (1), dilated common bile duct (3), choledochal cyst (1). Pancreatic abnormality found in 4 (20.0%) cases with findings of dilated pancreatic duct (3), pancreatic cyst (2), pancreatic (Frantz) tumour (1), pancreatic calcification (1)and irregular pancreatic duct (1). 5 children had a CT scan with abnormal finding seen in 4 cases: Pancreatic calcification (2), mass in the pancreatic tail (1), choledochal cyst (1), oedematous pancreas (1). 15 patients had MRIs with positive findings seen in 10 (66.6%): gall bladder stones (2), choledochal cyst (1), and gall bladder sludge (1). Pancreatic abnormalities in the form of pancreatic cysts (2), beaded irregular pancreatic duct with atrophic changes (2), pancreatic calcification (1), pancreatic tumour (1) and pancreas divisum (1).Various pharmacological therapies were tried, 7 were treated conservatively (intravenous fluids, analgesic and proton pump inhibitors), 9 children received parenteral nutrition, which was later stopped. 3 patients had surgery. 4 children received antibiotics, 2 were treated with antifungals and 1 child was given Acyclovir. The mean duration of hospital stay was 27 days, range 1 to 372 days. The mortality rate was zero %.

Conclusion:
Pancreatitis in children is rare, has multiple aetiologies and needs a low threshold in suspicion. Inflammatory bowel disease seems to be one of the major causes in our group. We recommend an abdominal US initially and later on an abdominal MRI/MRCP. Patients with recurrent pancreatitis should have an extended CFTR screen and screening for PRSS1 and SPINK1 gene mutations. Treatment should be supportive and deal with the underlying cause.

Suboptimal Vitamin D Status in Treated Coeliac Disease: Is Current Practice for Monitoring Bone Health Adequate?

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Introduction:
Vitamin D sufficiency has a well established role in bone health and is an area of active research. Abnormalities in bone mineral density, consequent osteopenia and fracture risk are well recognised complications of untreated coeliac disease (CD). Early diagnosis, intervention with gluten free diet (GFD) and careful monitoring of dietary compliance and bone health are thought to protect children from future complications. Recent expert consensus statements (1) and reviews have made recommendations about treatment goals for Vitamin D in patient groups considered to be at risk including malabsorptive conditions. Serum concentrations of 25,hydroxy, Vitamin D [25(OH)D] is agreed to be the most robust marker for vitamin D status: levels of >75 nmol/l are optimal; <75 – 50 nmol/L, suboptimal and <50 nmol/L deficient associated with disease risk.

Aim:
To evaluate vitamin D status in relation to our current clinical monitoring practice in a cohort of treated CD children.

Method:
20 unselected consecutive patients (mean age 10 years 3 months ± 3 years 5 months SD, 5 male and 15 female), requiring routine serological testing for tissue transglutaminase (tTG) to assess disease response or dietary compliance were identified between September 2010 – October 2011. Only patients with diagnosis of CD >1 year were included. 25(OH)D, bone chemistry markers (albumin, phosphate, calcium, alk phos), and parathyroid hormone (PTH) were measured. In addition, clinical assessments were made to assess bone health, diet and anthropometry, including patient/parent rated physical activity score (range 1-5), GFD compliance (Full +/- accidental exposure 3, partial 2, poor 1),calcium, iron and vitamin D intake, Ht, Wt and BMI Z scores.

Results:
25(OH)D levels were < 75nmol/L (sub-optimal) in 12/20 (60%) of patients tested including 4/20 (20%) who showed levels < 50nmol/L (deficient). Vitamin D levels showed no clear correlation with time elapsed from diagnosis (mean 37 months ± 19.5 SD) or age at testing (as above). None of the patients showed any clinically significant abnormality in bone chemistry. Only 1 patient showed marginally increased PTH (66 ng/L) in association with 'deficient' 25(OH)D levels (29.2 nmol/L) and normal bone chemistry indicating secondary hyperparathyroidism. GFD compliance by patient/parent reports was poor (score 1/3) in 1 patient only, associated with abnormal tTG (48.4 u/ml) and 'sub-optimal' 25(OH)D level (67 nmol/L). All others showed compliance that was good (score 3/3): of this group tTG was abnormal in 2/18 patients showing tTG and 25(OH)D levels respectively as follows; (a) 16.7 u/ml and 74.4 nmol/L (suboptimal) and (b) 35.7 u/ml and 160 nmol/L (optimal). Dietary Vitamin D intake was assessed as sufficient in all but 1 patient in whom 25(OH)D was 38.3 nmol/L (deficient): the assessment failed to identify 11/12 (91%) of patients who showed suboptimal 25(OH)D levels. Poor dietary intake of calcium was shown in only 2/19 patients associated with 25(OH)D levels of 19.9 and 74.4 nmol/L, deficient and suboptimal respectively. Physical activity score (mean 4 ± 0.75 SD) was similar in all patients. Anthropometric parameters were comparable in all groups including BMI Z score (mean 0.1± 0.82 SD).

Conclusion:
This preliminary study suggests that Vitamin D status in treated CD is suboptimal in the majority (60%) of patients tested and would not be identified by current routine clinical monitoring practice. Consideration should be given to include measurement of 25(OH)D levels as part of more comprehensive evaluation of bone health in patients on a GFD.

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Potential Impact of Revised ESPGHAN Guidelines for Diagnosis of Coeliac Disease

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Introduction:
Revised ESPGHAN criteria for diagnosis of coeliac disease are likely to recommend a change in management of symptomatic patients with suspected coeliac disease who have a tissue Transglutaminase (tTG) > 10 times upper limit of normal (ULN). In this group only it is suggested a definitive diagnosis can be reached without the need for intestinal biopsy if a second blood sample is both positive for Endomysial antibody (EMA) and the patient has an HLA type consistent with coeliac disease. Although BSPGHAN response to this is awaited we consider the implications for patients and our service if these proposals are adopted.

Method:
Patients attending the Paediatric Coeliac clinic were identified from the Paradox Gastroenterology database and cross checked with clinic records. A review of notes and hospital results system was undertaken for all patients diagnosed in an 8 year period between Sept 2003 and Sept 2011. Costs for EMA immunoflourescence and HLA typing were provided by laboratory services; costs of day case admission for OGD and biopsy are from our Finance department. Time to diagnosis was the average wait for endoscopy plus pathology turns round times as families are telephoned as soon as results are available.

Results:
122 patients were identified, with an average of 18 new diagnoses per annum over the last 3 years. 5 were excluded as they are IgA deficient, 7 had no quantitative result tTG , 1 was Marsh Grade 1 only and there insufficient data for 4 others. Of the remaining 104 a further 18 were excluded as they were asymptomatic at diagnosis. 85 patients were symptomatic at the time of diagnosis, 61 (73%) of whom had tTG >10 ULN representing 60% of total coeliac clinic population.
Approximate costs for EMA are £15 with a turn round time of 10 days. HLA DR3/DQ2 testing cost is £143 with a time from blood collection to results of 3 weeks giving a combined cost of £158 for second line blood tests. Average charge to PCT per day case for endoscopy and biopsy is £1163. Average time from listing to biopsy result 7 weeks.

Conclusion:
In our unit 11 procedures per annum would be avoided, ,shortening time to diagnosis by 4 weeks whilst making savings of £11055 to the health care economy. Impact on endoscopic training is likely to be small but reducing pressure for endoscopic procedure is likely to have a beneficial effect on the service overall.

Discussion:
Use of the revised guidelines would have resulted in a reduction of 4 weeks in time to diagnosis by replacing endoscopy with a second blood sampling, also avoiding procedure related and anaesthetic risks. One additional blood test would be required and the costs of this have not been calculated.
No additional outpatient appointments would be needed if the two stage testing is arranged at first appointment. Patients who have a tTG>10ULN at referral would be seen to be assessed for symptoms before second stage of testing is undertaken.
Current diagnosis rates are 18 per annum, as 60% of our patients were symptomatic and had tTG>10ULN this suggests that 11 patients per annum could be diagnosed without needing biopsy. The cost saving of £1005 per patient would give a £11055 saving per annum to the PCT. A reduction in income to the acute unit is unlikely to be significant as current endoscopic demand exceeds capacity and our prevalence figures suggest there remains a considerable number of undiagnosed patients. It could be surmised that increased education at the launch of new guidelines may result in an increase in referrals. There will not be significant training issues as patients who are asymptomatic or have a tTG which is less than >10ULN will still require biopsy.
Exact costs for tests, turn round time for results, logistical arrangements and numbers of relevant cases per annum will vary between units although similar benefits are likely.

Can anti-tissue transglutaminase antibody (tTG) levels predict mucosal inflammation in children with Coeliac Disease thus avoiding endoscopy? Experience in a single Paediatric Gastroenterology Centre

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Background:
The diagnostic process for Coeliac disease has evolved over the decades with advancing scientific development. One of the aims in children is for less invasive investigations. Antibody screening is commonly performed, triggering the need for intestinal biopsies if positive. ESPGHAN is proposing a diagnostic level of TTG of >10 times of upper limit of normal (ULN) for the diagnosis of Coeliac disease, minimising the need for intestinal biopsies.

Aim:
Our aim was to assess the correlation between tTG and histology of intestinal biopsies in our population, and to understand the reasons for possible false positives and negatives.

Method:
We identified all patients with tTG levels above 8 presenting to our unit. These levels were matched with histology of intestinal biopsies taken at the same time or soon after the levels were done. Clinical details were obtained from previous correspondences and personal details from the hospital data system.

Results:
We retrospectively reviewed all positive tTG levels over an 8.5 year period, normal range at our institution is 0-7.99 U/ml. There were 188 tTG levels over 8 U/ml, range of 8-699 U/ml; female 116, male 72; age range of 1-16 years, median age 7 years. 33 patients were excluded as the tTG levels were done after biopsies had been taken.
The patient ethnicity (total n=178) were Caucasian n= 104 (58 %), Asian n=24 (13 %), others n=5 (3 %), and non-specified n=45 (25 %).
There were 19 (10%) children under the age of 2 years, with tTG level range of 10-341 U/ml.
In the 1-3 x ULN group (n=53), 8 (15%) patients had normal histology, 7 (13%) Marsh 1 changes, 21 (40 %) Marsh 2 and 3, giving an overall correlation of 53% (n=28). 17 (32%) patients had non-specific histological changes, e.g. eosinophilic enteropathy, post-infection and gluten-free diet (GFD) started pre-biopsies.
In the 3-5 x ULN group (n=24), 3 (12%) patients had normal histology, 0 (0%) Marsh 1 changes, 17 (70%) Marsh 2 and 3, giving an overall correlation of 71% (n=17). 4 (17%) patients had non-specific histological changes, similar to those stated above.
In the 5-10 x ULN group (n=23), 3 (13%) patients had normal histology, 1 (4%) Marsh 1 changes, 16 (70%) Marsh 2 and 3, giving an overall correlation of 74% (n=17). 3 (13%) patients had non-specific histological changes, similar to those stated above.
In the >10 x ULN group (n=55), 1 (2%) patient had normal histology, 5 (9%) Marsh 1 changes, 47 (85%) Marsh 2 and 3, giving an overall correlation of 96% (n=53). 2 (4%) patients had non-specific histological changes, similar to those stated above.
In the under 2 years of age group, only levels of >5 x ULN showed some poor correlation, with 8 (42%) patients having Marsh 1-3 changes and 3 (16%) having non-specific changes.

Conclusion:
Our data suggest that in patients over the age of 2 years with tTG levels of >10 x ULN, the diagnosis of Coeliac disease can be safely made on laboratory results only. Our data also suggest that lower tTG levels do not correlate well, thus still warrants an OGD for the diagnosis Coeliac disease, as do patients under the age of 2 years.
We suggest that an UK-wide audit is performed to collate results from all GI-Units, to suggest UK-wide recommendations on the diagnosis of Coeliac disease.

A STAT change in Inflammatory Bowel Disease? Altered signalling in intestinal T cells

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Introduction:
The immunological basis of disease heterogeneity within Inflammatory Bowel Disease (IBD) is poorly understood. In part this is because the plasticity of T cell function in the mucosa means responses are dynamic and not adequately described by categorisation based on cytokine production potential. Analysis of the phosphorylation status of key proteins in immune signalling pathways, such as Signal Transduction and Activator of Transcription (STAT) proteins, has the potential to offer a more dynamic picture. Traditional analysis of phospho(p)STATs at the whole tissue level has shown IBD associated differences but is a rather blunt tool. "Phosflow" is a novel technique using flow cytometry and phospho-specific monoclonal antibodies to analyse signalling pathways at the single cell level and has yet to be applied to intestinal tissue.

Aim:
To evaluate Phosflow of intestinal T cells for analysis of immune heterogeneity in IBD.

Method:
Endoscopy biopsies were obtained from IBD patients and controls who had no inflammatory disease. Cell suspensions were obtained either by enzymatic digestion or by allowing cells to spontaneously migrate out of cultured tissue ('walk-out cells'). The cells were stimulated for 15 minutes with cytokines (eg interferon∞) to induce STAT phosphorylation or left unstimulated. After fixation and permeabilisation, the cells are labelled with different phospho-specific antibodies (pSTAT1, 3, 4, 5, 6), cell surface markers (CD3, CD4) and transcription factors (T-Bet) for flow cytometry.

Results:
Superior staining of phospho-antigens was obtained using walk-out cells compared with those obtained by enzymatic digestion. Both with and without stimulation, staining of intestinal CD4+ T cells for pSTAT1 (a T_H1 transcription factor), pSTAT3 (a T_H17 transcription factor) and pSTAT5 (a Treg transcription factor) was obtained. Expression of pSTAT1 was increased in CD4+ve lymphocytes of IBD patients compared with similar cells from controls ($p=0.025$). In paired samples from Crohn's patients, levels of pSTAT1 were higher in these cells from uninflamed than inflamed areas ($n=18$, $p=0.01$). Expression of the T_H1 transcription factor T-Bet was also observed in a proportion of intestinal CD4+ T cells. However, no relationship was observed between pSTAT1 and T-Bet expression suggesting that pSTAT1 expression is not confined to a conventional T_H1 population. The pSTAT1:pSTAT3 ratio of expression was different between controls and IBD ($p=0.001$), suggesting a change in T_H1:T_H17 balance of signals.

Conclusions:
Phosflow is an effective tool for detecting activated intracellular proteins in human intestinal CD4+ T cells. There are higher levels of pSTAT1 in IBD than controls, and this is not simply a measure of inflammation or T_H1 cell phenotype. Different ratios of pSTAT1:pSTAT3 in IBD patients suggest that these cells have different responsiveness to local signals. Further experiments are ongoing to determine the mechanisms of these observed differences and their heterogeneity, and to link pSTAT expression to different patients' phenotype and disease course.

Paediatric Helicobacter pylori practice in the United Kingdom: A BSPGHAN Survey

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Introduction:
A joint evidence-based, Delphi consensus guideline has recently been published on the management of children with suspected Helicobacter pylori infection by the European and North American Societies of Pediatric Gastroenterology, Hepatology and Nutrition1. Although many of the recommendations are likely in keeping with current UK practice, some areas are potentially controversial.

Aim:
To "benchmark" the guideline against current UK practice by a survey of the members of the British Society of Paediatric Gastroenterology, Hepatology and Nutrition (BSPGHAN).

Method:
A structured multiple-choice survey was constructed around the fictitious case of a "10 year old boy with dyspepsia and a positive family history of *H. pylori* associated peptic ulcers." Questions were based on the guideline with "correct" answers being defined where possible. The survey was e-mailed on two occasions to the full membership of BSPGHAN in July and September 2011. Completed surveys were returned by e-mail or anonymously in the post.

Results:
Of 38 replies, 35 were from consultants and 3 were from trainees. 37 (97%) would test for *H. pylori* in the case, in accordance with the new guidelines.
The guideline recommends invasive tests (rapid urease (CLO) test or upper gastrointestinal endoscopy plus biopsy) for detection of *H. pylori* but only 23 of 38 (60%) respondents agree. 37 (97%) respondents chose appropriate triple therapy first- line treatment. All respondents would treat for the suggested 7-14 days.
The guideline recommends confirmation of eradication but just over half (20 of 38, 53%) would do this with others only testing if symptoms did not improve (17 of 37, 46%). One respondent would not confirm eradication. 27 of 36 (75%) would confirm eradication within the recommended 4-8 weeks. All others bar one would have waited longer. Non-invasive tests (stool antigen ELISA or urease breath test) are recommended to confirm eradication, 33 of 38 (87%) of responses concur.
Perhaps the most controversial area from the guideline concerns when to treat *H. pylori*. Surprisingly 9 of 36 (25%) answered that they would not treat H. pylori assuming a positive test and a gastric/duodenal ulcer. 33 of 36 (92%) respondents were in favour of treating a positive test assuming gastritis with no ulcers despite the guideline being ambiguous as to the correct course of action. Responses were split as to the correct course of action given a positive test result and a normal endoscopy (62% yes, 33% no, 5% unsure). This is also reflected by ambiguity in the guidelines.

Conclusion:
Overall there are many areas for optimism where the guideline accurately reflects current UK clinical practice; however there are areas where they do not correlate. Clearly this warrants reflection and discussion as to whether practice should change to accommodate these recommendations.

New arrangements for National Speciality Commissioning for (Paediatric) Gastroenterology, Hepatology and Nutrition

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Subject to Parliamentary approval, new arrangements will come into force in April 2013 for commissioning of all specialised service in England. (These changes do not affect Scotland, Wales or Northern Ireland.)

Under the current system 10 Specialised Commissioning Groups commission a variety of different services to different specification at different costs.

From April 2013, all specialised services will be commissioned by the NHS commissioning board (NHC CB). The NHS CB is likely to have some regional offices, but the planning, specification and monitoring of services will be national. The strict test of whether a service is commissioned by local groups or nationally by the CB will be laid down in the Act of Parliament: but a working assumption is that services specified in the Specialised Services National Definition Set (SSNDS) will be commissioned by the CB.

Definition number 23 covers children' services and includes (section 23.7) paediatric gastroenterology, hepatology and nutrition. It is available at http://www.specialisedservices.nhs.uk/library/21/Specialised_Services_for_Children.pdf