BSPGHAN
working for children with digestive and liver disorders

British Society of Paediatric Gastroenterology Hepatology and Nutrition

WINTER MEETING

28-30th January 2009

Sheffield Children's Hospital
and St Paul's Mercure Hotel

Educational Grants: We wish to thank the following sponsors for their generous support

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Local Organising Committee:

Dr David Campbell

Dr Sally Connolly

Professor Chris Taylor

Dr Mike Thomson

Abstract Selection Committee and Prize Selection Judges

Dr Sue Protheroe - Chair

Dr David Campbell

Dr Sally Connolly

Dr Ronald Bremner

Brochure produced by

Louise Chorley

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Thanks to our Principal Sponsors

As in previous years our relationship and support by Mead Johnson, Children's Liver Disease Foundation and Nutricia has continued to thrive. As we all know and acknowledge such gatherings of like minded individuals to learn from each other, support trainees, to network, and to drive forward the three spines of our group -gastroenterology, nutrition, and hepatology- would not be feasible without the extremely generous and committed grants from these organisations. Many of us know and have known the individuals involved, but it the legacy of these people and of successive BSPGHAN Councils in forging, which is an appropriate verb for a meeting being held in Sheffield, strong and future-proofed relationships with common goals of education and excellence within the disciplines that is more important. Thank you.

David Campbell
Sally Connolly
Chris Taylor
Mike Thomson

Thanks to our Major Sponsors

We are absolutely delighted that many of our old friends have provided us with Educational Grants to support this Annual Meeting again, and we extend our warm thanks to them. We are equally excited and grateful that many more Companies, including for the first time many of those involved in endoscopy, have come forward and helped us this year. We know that without your generous support such an important endeavour year on year would grind to a halt. We are looking forward to an ongoing and fruitful collaboration. Thank you.

David Campbell
Sally Connolly
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Mike Thomson

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Welcome address from Local Organising Committee

We are all tickled pink that you have all taken the time and made the effort to come to our proud and wonderful Yorkshire for this the 23rd Annual BSPGHAN Meeting.
We would like to extend a warm and generous Northern/cuddly and soft Southern (delete as appropriate dependent on your geographical origin) welcome.

Each year the 'Gathering' seems to expand and the calibre of the abstracts was superlative this year. Particularly pleasing was the proportion emanating from Associate Members. We are unique amongst our paediatric colleagues for two reasons.
One, the close and important relationship that we have in working within teams of multi-disciplinary individuals (all of whom are, of course, gorgeous in their own way), and two, the fact that we are a procedural specialty. We have deliberately invited a number of lectures from Associate members to reflect the former. We hope that the Wednesday of this Meeting reflects the latter, and will provide a platform for discussion on where lies the future of children's endoscopy, and in particular training, in the next decade or so.

A sincere thanks here is absolutely necessary for the fantastic support, encouragement, cajoling, and efficiency of Carla Lloyd who has been a tower of help over the past 12 months. Also to all our own team who have all helped out in various invaluable ways.

This year we have tried to spice things up even more than in previous meetings with a good dose of plain speaking in the great Yorkshire tradition with pro-and con- debate sessions. A free and frank discussion on a number of topical,yet unresolved, issues are expected, which may be continued into the evenings with our equally-to-be-expected hospitality. We are honoured to be the first National Meeting to receive a full Master Cutler Reception at the Cutler's Hall. He is, in Sheffield, the equivalent of the Mayor of London, except much more important. Fabulous music and dancing will follow and, we hope in part at least, make up for Dr Jenkins' after dinner speech.

So it is with a modest and optimistic hope that we invite you to enjoy, learn, opine, wine and dine with us in sunny Sheffield, the home of climbing, steel, humour, and Henderson's relish – oh, and the Arctic Monkeys. Welcome.

David Campbell
Sally Connolly
Chris Taylor
Mike Thomson

Wednesday 28th January 2009

BEST PRACTICE, TOP TIPS, AND RECENT ADVANCES IN PAEDIATRIC ENDOSCOPY

Venue: Endoscopy Unit and Lecture Theatre,
Sheffield Children's Hospital

9.00 – 10.00
Registration and Coffee

10.00 – 11.30
Chair: Dr H Jenkins and Dr F Torrente

Introduction by Dr Mike Thomson

Lectures

10.05 – 10.25

A possible paediatric endoscopy curriculum and syllabus
Dr Mike Thomson,
Consultant Paediatric Gastroenterologist
Sheffield Children's Hospital
Western Bank
Sheffield S10 2TH

Dr Ronald Bremner
Chair of Trainees
Birmingham Children's Hospital
Steelhouse Lane
Birmingham B4 6NH

10.25 – 10.45

eLearning for Health (ELfH) endoscopy project
Dr Neil Hawkes
Consultant Gastroenterologist at the Royal Glamorgan Hospital
Llantrisant, South Wales;
National Endoscopy Training Lead for Wales & Joint Clinical Lead for the e-Learning for Healthcare (e-endoscopy) project

10.45 – 11.00

Nurses in Endoscopy
Mick Cullen
Paediatric Gastro Nurse Specialist
Child Health
Southampton General Hospital
Tremona Road, Southampton, SO16 6YD

11.00 – 11.45

What would you do next? Difficult and interesting cases from delegates.

Dr Nick Croft
Consultant Paediatric Gastroenterologist
Royal London Hospital
Wingate Institute
London

Dr Stephen Murphy
Consultant Paediatric Gastroenterologist
Birmingham Children's Hospital
Birmingham

Simultaneous Clinical Session

10.05 – 11.30

Endoscopic oesophageal dilation from Sheffield Children's Hospital Western Bank Sheffield

Dr David Campbell
Consultant Paediatric Gastroenterologist
Sheffield Children's Hospital
and

Mr Sean Marven
Consultant Paediatric Surgeon

PEG/PEJ insertion - LIVE LINK from Royal Hospital for Sick Children 20 Sylan Place Edinburgh

Mr Gordon McGinlay
Consultant Paediatric Surgeon

And

Dr Peter Gillett
Consultant Paediatric Gastroenterologist

11.30 – 12.00
Coffee

12.00 – 13.00
Chair: Dr Camilla Salvestrini

Lectures

The theory of loops in colonoscopy, and practical solutions
Dr Stuart Riley
Consultant Paediatric Gastroenterologist
Northern General Hospital
Sheffield

Simultaneous Clinical Session

Colonoscopy with 3-D imager
Dr Mark Donnelly
Consultant Paediatric Gastroenterologist
Northern General Hospital
Sheffield

Dr Prithvi Rao
Specialist Registrar
St James' University Hospital, Leeds

13.00 - 13.45
LUNCH

13.45 – 16.00
Chair: Dr D Wilson and Dr J Bishop

Lectures

13.45 – 14.15

Double and single balloon enteroscopy

Dr Chris Fraser
Consultant Paediatric Gastroenterologist
Wolfson Unit for Endoscopy
St Mark's Hospital
Watford Road, Harrow
Middlesex HA1 3UJ

14.15 – 15.15

Variceal and non-variceal endo-haemostasis
Dr Patrick McKiernan
Consultant Paediatric Hepatologist
Birmingham Children's Hospital
Birmingham B 4 6NH

Simultaneous Clinical Session

13.45 – 16.00

Diagnostic and therapeutic colonoscopies including confocal endomicroscopy and magnification colonoscopy

Dr Mike Thomson
Consultant Paediatric Gastroenterologist
Sheffield Children's Hospital
Western Bank
Sheffield, S10 2TH

and

Live Link from Nice, France

15.15 – 15.30
TEA

Lectures

15.30 – 16.00

NOTES: is there a place for this

Dr John Morris
Consultant Paediatric Gastroenterologist
Royal Infirmary
84 Castle Street
Glasgow
G4 0SF

16.00 – 17.30

16.00 – 17.30

Endoscopy Steering Group, Open Meeting Training 2009 and beyond

Dr Mike Thomson
BSPGHAN Endosocopy Steering Group Chairman
Consultant Paediatric Gastroenterologist
Sheffield Children's Hospital
Western Bank,
Sheffield, Sl0 2TH

And

Professor Roger Barton,
Consultant Paediatric Gastroenterologist
North Tyneside General Hospital Department of Medicine
Rake Lane
North Shields
Tyne and Wear
NE29 8NH

17.30
Close

Coaches to St Paul's Mercure Spa and Hotel, Sheffield

Annual Trainees v Consultants football match at Sheffield University at 6.00 p.m.
Leave from Sheffield Children's Hospital at 5.30 p.m.

Please note participants to take kit to the endoscopy meeting and go straight from the meeting to the football venue. Minibus transport from Sheffield University to hotel at 19.15

19.30
Ice breaker session – Meet the sponsors and Dinner
Wednesday 28th January 2009, City Suite, St Paul's Mercure Hotel, Sheffield

Thursday 29th January 2009

NEW HORIZONS AND OLD CONTROVERSIES IN PAEDIATRIC GI

Venue: St Paul's Mercure Hotel and Spa
119 Norfolk Street, Sheffield

9.30 – 12.00 **Registration**

10.00
Sponsors Exhibition Open

Poster session I viewing from 10.30

23rd BSPGHAN Meeting

12.00 – 12.45
Buffet Lunch

Poster session I and judging

12.45 – 12.50 **Opening and welcome on behalf of the organising committee**
Dr Mike Thomson
Consultant Paediatric Gastroenterologist
Sheffield

Session I Current Controversies in CF
Or "how we ignore a lack of evidence-base in personal practice"

Chair: Dr David Campbell and Dr Indra van Mourik

12.50 – 13.10 **Paediatric Gastroenterologists should look after children and young people with Cystic Fibrosis**
Professor Chris Taylor
Consultant Paediatric Gastroenterologist
Sheffield Children's Hospital
Sheffield S10 2TH

13.10 – 13.30 **Respiratory paediatricians should look after children and young people with Cystic Fibrosis**
Dr Mark Everard
Consultant Respiratory Paediatrician
Sheffield Children's Hospital
Western Bank
Sheffield S10 2TH

13.30 – 13.45 **Debate and vote**

13.45 – 15.20 **Plenary session I with abstracts from Gastroenterology, Hepatology and Nutrition**

13.45 – 13.58 **A BSPGHAN audit of the effectiveness and safety of adalimumab in children and young people with Crohn's disease**
Presenter: Dr David Wilson
RK Russell[1,2], ML Wilson[3],N Shah[2],G Mahdi[2], W Hyer[2], C Spray[2], C Daman[2], R Heuschkel[2], N Afzal[2], M Elawad[2], F Torrente[2],N Ayub[2], J Fell[2],GT Ho[2], S Naik[2], DC Wilson[2,3].
1. Department of Paediatric Gastroenterology, Yorkhill Hospital, Glasgow. 2.BSPGHAN Adalimumab study group. 3.Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh and Child Life and Health, University of Edinburgh.

13:58 – 14:11 **A 20 year single centre experience of Home Parenteral Nutrition**
Presenter: Ms Tracey Johnson
Tracey Johnson[1], Chris Holden[2], Elaine Sexton[2], Sue Protheroe[3], Ian Booth[3], Stephen Murphy[3]
1 Department of Dietetics, 2 Department of Nutritional Care, 3 Department of Gastroenterology and Nutrition, Birmingham Children's Hospital

14:11 – 14:24 **Haematopoeitic Stem Cell Transplant for Severe Inflammatory Gut Disorders with Immune Dysregulation**
Presenter: Dr Amel Hassan
Hassan A, Veys P, Shah N, Lindley KJ, Elawad M. Great Ormond Street Hospital, London WC1N 3JH

14:24 – 14:37 **Faecal calprotectin remains high during enteral feed and corticosteroid therapy in newly diagnosed paediatric Crohn's disease.**
Presenter: Ms Jo Grogan
Grogan J L BSc (Hons) SRD*, Terry A BSc (Hons) SRD*, Casson D BA (Oxon) MRCPI†, Dalzell AM BSc MBBS FRCPCH †
*Advanced Paediatric Dietitian, Royal Liverpool Children's NHS Trust, Liverpool
†Consultant Paediatric Gastroenterologist, Royal Liverpool Children's NHS Trust, Liverpool

14:37 – 14:50 **How to develop your MCN using Telemedicine! The Scottish Paediatric Gastroenterology, Hepatology and Nutrition Group (SPGHANG) experience**
Presenter: Dr Andrew Barclay
A R Barclay[1], P M Gillett[2], D Goudie[3], W M Bissett[4],P McGrogan[5]
1. Division of Developmental Medicine, University of Glasgow.
2. Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh. 3. Department of Paediatrics, Raigmore Hospital, Inverness
4. Department of Paediatric Gastroenterology, Royal Aberdeen Children's Hospital.
5. Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Yorkhill, Glasgow.

14:50 – 15:03 **RCT of antibiotic prophylaxis to reduce infection rates at paediatric percutaneous endoscopic gastrostomy (PEG) tube insertion**
Presenter: Ms Catherine Paxton
[1]CE Paxton, [1,2]ML Wilson, [3]DM Hoole, [4]FD Munro, [1]PM Gillett and [1,2]DC Wilson. Departments of [1]Paediatric Gastroenterology and Nutrition, [3]Pharmacy and [4]Paediatric Surgery, Royal Hospital for Sick Children, Edinburgh and [2]Child Life and Health, University of Edinburgh.

15:03 – 15:16 **Development and Performance of a New Paediatric Nutritional Screening Tool in a Tertiary and District General Hospital. The PYMS Project**
Presenter: Mr Konstantinos Gerasimidis
Gerasimidis K[1,2], Macleod I[1], McGrogan P[1], Maclean A[2], Buchanan E[1,2], McAuley M[3], Stewart G[5], Wright CM[4], Flynn DF[1]
[1]Department of Paediatric Gastroenterology, Hepatology and Nutrition, Royal Hospital for Sick Children, Glasgow, UK [2]Department of Dietetics, Royal Hospital for Sick Children, Glasgow, UK [3]Medical Paediatrics, Royal Hospital for Sick Children, Glasgow, UK [4]Division of Developmental Medicine, University of Glasgow, Royal Hospital for Sick Children, Glasgow, UK [5]Department of Paediatrics, Royal Alexandra Hospital, Paisley, UK

15.20 – 15.45 **Tea**

Session II "When the brown stuff hits the revolving thing"

Chair: Professor Ian Booth

15.45 – 16.15 **New developments and drugs in the treatment of acute diarrhoea**
Dr Stephen Murphy
Consultant Paediatric Gastroenterologist
Birmingham Children's Hospital
Whittall Street
Birmingham, B4 6NH

16.15 – 16.45 **Resource implications and budget impact of managing cow milk allergy in the UK**
Professor Julian Guest
Director
CATALYST Health Economics
Northwood
Middlesex
HA6 1BN
Sussex University

17.00 – 18.30 **Annual General Meeting**

19.30
Master Martin Howell
Master Cutler's Official Civic Reception, Cutler's Hall

20.30 – late.
Conference dinner and entertainment

Friday 30th January 2009

NUTRITION AND HEPATOLOGY

Venue: St Paul's Mercure Hotel

Session III: Current controversies in Coeliac Disease
"There is no body cavity that cannot be reached by the application of a strong arm and a 6-FG needle"
"House of God" by Samuel Shem

Chair: Dr Rajeev Gupta and Dr Neil Shah

9.00 - 9.20 **There remains a need for biopsy in coeliac disease**
Dr Muftah Eltumi
Consultant Paediatric Gastroenterologist
Watford Hospital
Herts

9.20 – 9.40 **There is no longer a need for biopsy in coeliac disease**
Professor Paul Ciclitira
Professor of Gastroenterology
The Rayne Institute
4th Floor Lambeth Wing
St Thomas' Hospital
London SE1 7EH

9.40 – 9.55 **Debate and vote**

Session IV: Sponsored by Children's Liver Disease Foundation

Session IV: Current controversies in Liver Disease
Or 'the definition of an alcoholic is someone who drinks more than his (her) doctor'

Chair: Dr Patricia McClean and Dr Sally Connolly

Intestinal Failure, Short Bowel Syndrome and Associated Liver Disease

9.55 – 10.15 **Dr John Puntis**
Consultant Paediatric Gastroenterologist
Room 142, B Floor
Clarendon Wing
The General Infirmary at Leeds
Belmont Grove
Leeds
LS2 9N,
West Yorks

10.15 – 10.35 **Miss Jenny Walker**
Consultant Paediatric Surgeon
Sheffield Children's Hospital
Western Bank
Sheffield
S10 2TH

10.35 – 10.45 **Debate and vote**

10.45 – 11.15 **COFFEE**

10.45 Poster Session II Viewing

Session V:

Chair: Dr David Rawat and Dr Paraic McGrogan
"To boldly go where no man has gone before" Captain James T Kirk, USS Enterprise 2358

11.15 – 11.35 **The FOX amongst the chickens**
Dr David Campbell
Consultant Paediatric Gastroenterologist
Sheffield Children's Hospital
Western Bank
Sheffield S10 2TH

11.40 – 12.00 **Paediatric GORD management: where's the evidence? Update on the ESPGHAN and NASPGHAN Conjoint Reflux Working Group 2007 - 2009**
Dr Mike Thomson
Consultant Paediatric Gastroenterologist
Sheffield Children's Hospital
Western Bank
Sheffield S10 2TH

12.05 – 12.25 **Quality of Life in GI disorders**
Miss Kate Blakeley
Paediatric Liaison Team
2nd Floor Fielden House
Royal London Hospital
Whitechapel
London
E1 1BB

12.30 – 13.30
BUFFET LUNCH

Poster Session II and judging

Session VI

Chair: Dr David Wilson and Professor Billy Bourke
or "What's the collective noun for Professors? – An absence of…"

13.30 – 15.00 **Plenary abstract session II**

13:30 – 13:43 **Confocal Endomicroscopy: A New Tool in the In Vivo Diagnosis of Coeliac Disease**
Presenter: Dr Krishnappa Venkatesh
Krishnappa Venkatesh[1], Ashraf Abou-Taleb[1] Marta Cohen[2], Clair Evans[2], Tracey Young[3] Christopher Taylor[1], Mike Thomson[1].
1 Centre for Paediatric Gastroenterology, Sheffield Children's NHS Foundation Trust, Sheffield, United Kingdom 2 Department of Histopathology, Sheffield Children's NHS Foundation Trust, Sheffield, United Kingdom. 3 Lecturer in Medical Statistics, University of Sheffield, Sheffield, United Kingdom

13:43 – 13:56 **The Health Economics of Gastrostomy Insertion and Fundoplication in Children with Neurological Impairment**
Presenter: Ms Angharad Vernon-Roberts
Angharad Vernon-Roberts[1] Jose Leal[2] Alastair Grey[2] Hugh Grant[3] Peter Sullivan[1]
1Oxford University Department of Paediatrics
2Oxford University Department of Health Economics
3Oxford Children's Hospital Department of Paediatric Surgery

13:56 – 14:09 **A sudden and marked reduction in PN cholestasis on changing from a conventional intravenous lipid source to SMOF lipid**
Presenter: Dr Muhammed Rafeeq
Rafeeq M, Bremner R, Davies P, Protheroe S, Holden C, Johnson T, Murphy MS.
Department of Paediatric Gastroenterology & Nutrition, Birmingham Children's Hospital NHS Foundation Trust

14:09 – 14:22 **Childhood-onset versus adult-onset inflammatory bowel disease: Phenotype**
Presenter: Dr Johan van Limbergen
Johan Van Limbergen[1,2,6] MD, MRCPCH; Richard K Russell[3] MRCPCH, PhD; Hazel E Drummond[1] BSc; Marian C Aldhous[1] BSc, PhD; Nicola K Round[1,2]; Elaine R Nimmo[1] BSc, MSc, PhD; Linda Smith[1] RMN, RGN; Peter M Gillett[2] MBChB, FRCPCH; Paraic McGrogan[3] MBChB, MRCP; Lawrence T Weaver[4] MD, FRCPCH; W Michael Bisset[5] MD, FRCPCH; Gamal Mahdi[5] FRCPCH; Ian D Arnott[1] MD; Jack Satsangi[1] DPhil, FRCP; David C Wilson[2,6] MD, FRCPCH
1. Gastrointestinal Unit, Molecular Medicine Centre, Western General Hospital, University of Edinburgh, Edinburgh, United Kingdom.
2. Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh, United Kingdom.
3. Department of Paediatric Gastroenterology, Yorkhill Hospital, Glasgow, UK

14:22 – 14:35 **Childhood-onset versus adult-onset inflammatory bowel disease: Genotype**
Presenter: Dr Johan van Limbergen
Johan Van Limbergen[1,2,4] MD, MRCPCH; Richard K Russell[3] MRCPCH, PhD; Jack Satsangi[1] DPhil, FRCP; David C Wilson[2,4] MD, FRCPCH on behalf of the International Pediatric IBD Genetics Consortium
1. Gastrointestinal Unit, Molecular Medicine Centre, Western General Hospital, University of Edinburgh, Edinburgh, United Kingdom.
2. Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh, United Kingdom.
3. Department of Paediatric Gastroenterology, Yorkhill Hospital, Glasgow, United Kingdom.
4. Child Life and Health, University of Edinburgh, Edinburgh, United Kingdom

14:35 – 14:48 **Is TauroLock® the answer to recurrent Central line infections?**
Presenter: Dr. Balaji Krishnamurthy
Dr. B Krishnamurthy[1], Dr. V Zamvar[2], Mrs. G Lazonby[3], Dr. JWL Puntis[1]
Department of Paediatric Gastroenterology[1] & Nutrition[3], The General Infirmary at Leeds, UK Department of Paediatrics[2], The Calderdale Royal Hospital, Halifax

14:48 – 15:01 **Food reintroduction following exclusive enteral feeding in Crohn's disease**
Presenter: Ms Jackie Falconer
Jackie Falconer[1], Tracey Johnson[2], Elaine Buchanan[3], Julia Smith[4], Karen Maxwell[5], Joanna Caines[6] on behalf of the Associate members BSPGHAN
1 Nutrition and Dietetics Dept, Chelsea and Westminster Hospital
2 Nutrition and Dietetic Dept, Birmingham Children's Hospital
3. Nutrition and Dietetic Dept, Royal Hospital for sick Children
4. Nutrition and Dietetic Dept, Addenbrookes Hospital
5. Nutrition and Dietetics Dept, Kings College Hospital
6. Nutrition and Dietetics Dept, Northwick Park Hospital

15.01 – 15.20 **A comprehensive young person's obesity service – can it exist and can it be effective**
Dr Jerry Wales
Consultant Paediatric Endocrinologist
Sheffield Children's' Hospital
Western Bank
Sheffield S10 2TH

15.20 – 15.40 **Feeding disorders in infants**
Professor Carlos Lifschitz
Associate Prof of Pediatrics
Baylor Coll of Med
Houston
TX and Med Dir U, Mead Johnson Nutritionals

15.40 – 16.00 **Feeding aversion and disorders in young children: a guide to management and provision of an MDT feeding clinic and service**
Lesley Cogher
Manager, Sheffield Speech & Language Therapy Service
Ryegate Children's Centre
Tapton Crescent Road
Sheffield
S10 5DD

16.00 **Presentation of prizes**

Sean Devane Memorial Prize – Presented by Stephanie Devane
Alex Mowat Memorial Prize
Best Allied Health Professional Prize

Close of Meeting
Dr Huw Jenkins
President BSPGHAN

24th BSPGHAN Winter Meeting
27th to 29th January 2010, Liverpool
Local organizer: Dr Mark Dalzell

ABSTRACTS FOR WEDNESDAY 28TH JANUARY 2009

A possible paediatric endoscopy curriculum and syllabus - UK Paediatric Endoscopy Curriculum BSPGHAN/BAPS 2008

Dr Mike Thomson, Consultant Paediatric Gastroenterologist, Sheffield
Dr Ronald Bremner, SpR, Chair of Trainees, Birmingham Children's Hospital

November 2008

Background

With the increased relevance and emphasis on excellence of skill base in procedural specialities, combined with the identified risk adherent to variable skill acquisition and maintenance, it is clear that paediatric endoscopy practitioners require not only a sound training base but an ongoing commitment to the highest levels of competency for the children undergoing these endo-diagnostic, and especially endo-therapeutic procedures.

As part of this initial and ongoing training process it is now widely recognised in adult GI training endoscopy circles that the components of such a landscape involve some or all of the following, which we believe should be actively adopted in paediatric endoscopy training to enable credibility of such a training process and allow a clinical governance process leading to excellence of clinical care which is therefore above reproach:

- An endoscopy curriculum of theory and knowledge base, but also practical skill exposure and competence. This would be monitored using a log book and trainer assessment of skill level on a contemporaneous basis throughout training.

- Formative and summative DOPS (Direct Observational Practical Skill) assessment with a trainer who has undergone a 'Training the Trainer' Course. (Suggestion for one trainer per training centre to have a lead for this).

- Strict observation of the STR (College assessment process) in respect of endoscopic skill and ability, with recourse to retraining as required by an independent trainer from the training centre primarily involved in the trainee's career in paediatric GI training.

- CCST to be at a number of prescribed 'levels' of differing skill complexity, and also relevant to the eventual career aim of the individual. Hence CCST may differ for paediatric surgeons and paediatric hepatologists, and 'gastroenterologists with an interest' from, for instance, tertiary gastroenterologists. (See Table)

- A commitment to all trained paediatric endoscopy practitioners to submit to an agreed re-validation and re-accreditation process for endoscopic skills.

- That such a training process and re-assessment process occur under the auspices of the UK recognised training umbrella of the Joint Advisory Group on Endoscopy Training for the UK (JAG) as occurs for other endoscopic practitioners in the UK.

- Web-based lesion recognition tools are developed – eLfH via DoH.

- Short Hands-On Courses are considered highly desirable, if not mandatory, in order to achieve level 1 ileo-colonoscopy knowledge and skill base.

- Global Rating Score (GRS) and Unit visits occur in order to approach an ideal training environment.

As perhaps the most important first step in this approach a robust and valid curriculum would seem to be the first cornerstone to be put in place.

Such a curriculum is proposed in this document. Patient and parent interaction, information, and appropriate doctor-patient relationships are assumed throughout.

1. Core knowledge and skill base

Knowledge base:

Endoscopy team members and unit management

Endoscope cleaning, disinfection and storage

Endoscope construction and design – problem solving for malfunction

Consent process specific to endoscopy and all endoscopic procedures with familiarity with complication rates

General anaesthesia and where necessary sedation: safety and applicability

Indications for diagnostic upper endoscopy and ileo-colonoscopy

Contra-indications for diagnostic upper endoscopy and ileo-colonoscopy

Patient bowel preparation for diagnostic and therapeutic endoscopy e.g. bowel preparation for ileo-colonoscopy

Management of clinical problems such as IDDM; coagulation abnormalities; and congenital heart disease

Process for dealing with potential vCJD

Knowledge of data base and recording of procedure with appropriate still and moving image capture

Ability to generate an appropriate report

Knowledge of importance of continual assessment with contemporaneous account with DOPS assessment of a procedure with a trainer, and log book

Knowledge of size of endoscopes appropriate for different age groups

Knowledge of endoscope controls for differing visualisation e.g. magnification; narrow band imaging; haemoglobin accentuation; and use of chromoendoscopy

Knowledge of diagnostic accessories

Knowledge of specimen handling e.g. for disaccharidase analysis. Appropriate biopsy orientation with dissecting microscope if available

Knowledge of therapeutic accessories. E.g. balloon dilators; haemostasis devices; grasp forceps; snares; Roth nets etc

Use of thermocoagulation unit and application of patient plates

Level 1: Basic upper GI and ileo-colonoscopy skill level

Diagnostic upper endoscopy

Pre-endoscopy patient and equipment check
Intubation of oesophagus, with and without direct vision
Examination of and identification of anatomical landmarks and abnormal anatomy
Negotiation of stomach
Pyloric intubation
D1 to D4 negotiation
J manoeuvre
Lesion recognition
Site and number of biopsies required
Aspiration of duodenal juice if required
Knowledge of 'quick bedside' tests available: H pylori; lactase etc

Diagnostic ileo-colonoscopy (not required for hepatology trainees, and surgical trainees not aiming to focus on colorectal)

Pre-endoscopy patient and equipment check
PR examination
Patient positioning
Appropriate use of gut paralysing agents such as buscopan and glucagon
Anal intubation
Recto-sigmoid negotiation
Torque-steer technique
Clock-face concept
Ability to negotiate tip away from faecal contamination of colon
Recognition of lumen and no advancement of colonoscope without vision of lumen
Appropriate use of abdominal pressure by assistant
Appropriate insufflation and use of CO2, where available
Recognition of loop formation and types of loops
Recognition of paradoxical movement and loss of one-to-one shaft-to-tip relationship
Appropriate use of 3D Scope-Guide if available
Ability to remove a loop if formed (use of stylised format in order to do so with clockwise or anti-clockwise rotation etc) and knowledge of when a loop can be managed without being resolved
Appropriate and frequent patient position change
Knowledge of landmarks and comprehension of high variability of position of splenic and hepatic trans-colonic blue colouration
Negotiation of splenic flexure
Negotiation of transverse colon
Use of variable stiffness facility of colonoscope if available
Recognition of deep transverse loop formation
Patient position change for successful hepatic flexure negotiation
Recognition of caecum and modes of confirmation
Recognition of ileo-caecal valve
Rotation of ileo-caecal valve to 6 o'clock position
Ability to intubate ileo-caecal valve with 'blind' technique and biopsy forceps-led 'Seldinger'-type technique
Knowledge of importance of patient position change on extubation and speed of extubation
Rectal retro-flexion
Lesion recognition
Appropriate biopsy site and technique

Level 2: Intermediate knowledge and skill base – (if required by trainee)

Foreign body removal (this may be in Level 1 for surgical trainees)

Placement of naso-gastric and naso-jejunal tubes by endoscopy

Haemostasis – variceal and non-variceal (variceal will be in Level 1 for hepatological trainees and would include staging of varices and possible use of endo-ultrasound in this procedure)

PEG insertion (this may be moved to Level 1 for surgical trainees)

Appropriate consent procedure with knowledge of all potential complications and incidence
Indications
Contra-indications
Familiarity with PEG, PEGJ, and PEJ and all varieties available in the market place
Knowledge of and design of protocol, if not locally available, of post-procedure care and use, including need for antibiotic cover
Use of marcaine
Need for first needle passage with aspiration and direct vision to ensure first viscous perforated is stomach and not colon
Use of trochar and acquisition of wire or thread by assistant
Placement by pull-through technique of PEG
Use of anchoring device and connections
Directions for use post-PEG
Knowledge of long-term complication rate and after care required
Knowledge of when and how to change to low-profile device

Polypectomy

Appropriate patient selection
Knowledge of need for additional procedures such as upper GI endoscope or wireless capsule endoscopy
Appropriate consent and knowledge of incidence of potential complications
Familiarity with electrocautery delivery device and settings plus application to patient of earthing patch
Check pre-introduction of handle, marking of handle, and assistant familiar with snare
Knowledge of different snares available
Ability to use effectively chromo-endoscopy with indigo-carmine or Lugol's iodine
Knowledge of use of adrenaline and saline injection for endomucosal resection is not required in this level.
Use of snare to avoid complications
Importance of performing polypectomy on insertion if polyp identified
Importance of changing patient position if bowel preparation poor
Knowledge of variable polyp retrieval techniques
Knowledge of types of polyps
Knowledge of use of Endoclips and Endosnares if required to prevent post-polypectomy bleeding
Knowledge of approach in the face of complications such as haemorrhage or perforation
Adequate documentation for follow up and if necessary surveillance

Stricture dilation

Appropriate consent procedure with knowledge of all potential complications and incidence
Indications
Contra-indications
Knowledge of use of pre-endoscopic radiological assessment of stricture anatomy and associated pathology
Familiarity with different techniques and superiority of balloon radial dilation above push bougie-type dilation
Knowledge of radiological screening and its use
Knowledge of 'Rule-of-3s' for diameter of proposed dilation
Ability to recognise when a perforation has occurred, immediate or late post-procedure and to institute appropriate course of action
Knowledge of various aetiologies of strictures and amenability to dilation alone
Knowledge of use of post-dilation anti-fibrotic topical agents
Post-procedure care protocol
Indications for use of stents

Level 3: Advanced skill base. This could include some or all of the following:

Knowledge of therapeutic ERCP
Endo-ultrasound
NOTES (Natural Orifice Trans-Endoluminal Surgery)
Endoscopic fundoplication
Wireless capsule endoscopy
Enteroscopy – laparoscopy-assisted; single and double balloon
Sigmoidostomy and Caecostomy
Use of advanced endo-diagnostic technologies:
Narrow band imaging
Magnification and zoom endoscopy
Chromo-endoscopy
Confocal endo-microscopy
Knowledge of use of narrow band imaging to detect dysplasia
Knowledge of piecemeal resection techniques for large polyp removal
Knowledge of use of adrenaline and saline injection for endomucosal resection

Appendix 1: Table: Skill mix of training suggested for differing career aims

	Gastroenterologist	Hepatologist	Surgeon (All)	Surgeon (Upper GI)
Level 1	OGD (This could be Level 1 OGD alone). Ileo-colonoscopy.	OGD. Variceal endo-management	OGD. PEG. FB removal.	OGD. All skills noted in Level 2 training except ileo-colonoscopy.
Level 2	See Level 2 details. (It is important that a surgical back up is available on site for complications of procedures such as PEGs and dilations.)	No.	No.	See above.
Level 3	See Level 3 details. (May involve some or all components)	No.	No.	No.

eLearning for Health (eLfH) endoscopy project
Dr Neil Hawkes

Overall project aims

- To provide a blended learning tool for health professionals involved in the delivery of endoscopy services
- To develop a curricular structure for knowledge-based competencies
- To align the project with existing recommendations for training in endoscopy in the UK

Project structure

The e-endoscopy project will comprise nine Modules;

- Foundation Module
- Basic Upper GI Endoscopy
- Basic Lower GI Endoscopy

- Advanced Upper GI Endoscopy
- Advanced Lower GI Endoscopy
- ERCP
- Endoscopic Ultrasound
- Trainers
- Advanced endoscopy assistant

Each module is divided into domains comprising specific learning sessions. Each session is designed to last approximately 20 minutes. A session starts with stated learning objectives and ends with formative assessment of these learning objectives. As an example the Foundation Module comprises six domains & 15 e-learning sessions.

Project timetable
Work has commenced on the Foundation & Basic Upper & Lower GI modules. This will comprise the first phase of the project (completion date March 2010). A demonstration e-session will be available by March 2009 and work will begin on the other modules in a parallel to the first phase developments.

Nurses in Endoscopy
Mr Mick Cullen
Paediatric Gastroenterology Nurse Specialist
Southampton

As a specialist nurse in paediatric gastroenterology in a busy regional unit one of my roles is to help facilitate the patient pathway through the diagnostic process including endoscopy. As part of the multidisciplinary team I am involved in selection for endoscopy, preparation, endoscopy and follow- up of children and their carers as they experience the service.

Over the last 2 years I have taken on the extended nurse role of diagnostic upper gastrointestinal endoscopy. With tutoring, supervision and mentoring from two consultants and support from paediatric anaesthetists I have now completed over 150 independent procedures and at present am seeking JAG accreditation.

The extended role was a response to service need in Southampton where more than 500 children have diagnostic endoscopy per year as part of the regional paediatric gastroenterology service.

Learning the technical skills needed to fulfil the role was only part of the process. I have undertaken an independent history taking and clerking course, education sessions on consent, attend histology meetings and run a weekly nurse led clinic. I have also spent 5 years working in paediatric gastroenterology and been at endoscopy lists during that period observing pathology and helping plan management.

Nurse endoscopists in adult practice have been established since the mid 1990's. The nurse endoscopist in paediatrics is not a new concept (Irvine 2007) but it is a resource that is rarely utilised in practice. It could be used to help facilitate the diagnostic process in children with suspected gastrointestinal pathology as part of their care pathway particularly with the increased emphasis on waiting times.

Reference

Irvine T, Dalzell M. The developing nurse role: Paediatric Upper GI Endoscopy. Gastrointestinal Nursing 2007;10(5):16-9

www.thejag.org.uk

NOTES PAGE

What would you do next? Difficult and interesting cases from delegates.

Dr Nick Croft, Consultant Paediatric Gastroenterologist, Royal London Hospital, London
Dr Stephen Murphy, Consultant Paediatric Gastroenterologist, Birmingham Children's Hospital, Birmingham

The theory of loops in colonoscopy and practical solutions
Dr Stuart Riley, Consultant Paediatric Gastroenterologist, Northern General Hospital, Sheffield

Double and single balloon enteroscopy
Dr Chris Fraser, Consultant Paediatric Gastroenterologist, St Mark's Hospital, Middlesex

Variceal and non-variceal endo-haemostasis
Dr Patrick McKiernan, Consultant Paediatric Hepatologist, Birmingham Children's Hospital, Birmingham

NOTES: Is there a place for this?
Dr John Morris, Consultant Paediatric Gastroenterologist, Royal Infirmary, Glasgow

Endoscopy Steering Group: Training 2009 and Beyond

Dr Mike Thomson, Consultant Paediatric Gastroenterologist, Sheffield
Professor Roger Barton, Consultant Paediatric Gastroenterologist,North Tyneside General Hospital
Department of Medicine, Rake Lane, North Shields, Tyne and Wear,NE29 8NH

The Medical and Surgical Royal Colleges and their Specialty Advisory Committees (SACs) tasked the JAG to agree uniform standards for endoscopy no matter the professional background of the endoscopist. The JAG also agrees appropriate evidence of meeting those standards. It assesses if the portfolio presented by a trainee endoscopist evidences that the individual meets the criteria. If the criteria are met, the JAG issues a certificate, thus making a recommendation to the SAC. The SACs take this into account when accrediting individuals.

To improve the quantity and quality of training, JAG aims to ensure that all trainees should have one protected potential training list weekly in endoscopy, that at least 50% of their trainers should have undertaken a "Training the Trainers" course, specific for endoscopy, and that trainees are actively trained.

Currently, the JAG is working on refining the accreditation process, and is soon to publish a validation and reliability of a component of the accreditation – the DOPS assessment. The other aspect of improving quality is in improving trainers' skills, and this presentation will show data to underpin these two themes.

Professor Roger Barton

Paediatric Gastroenterologists should look after children and young people with Cystic Fibrosis

Professor Chris Taylor, Consultant Paediatric Gastroenterologist, Sheffield Children's Hospital
Sheffield, S10 2TH

Cystic Fibrosis results from defective function of the cystic fibrosis trans membrane conductance regulator (CFTR) an anion channel that sits in the apical membrane of secretory epithelia. Much of the current mortality is related to recurrent endobronchial infection however, most patients are now ascertained through post-natal screening programmes and are essentially asymptomatic in terms of respiratory disease and diagnosis. However 50% will show evidence of pancreatic insufficiency at diagnosis and the majority of the remainder will become pancreatic insufficient before their first birthday. Moreover, study of the pathogenesis of cystic fibrosis using animal models has shown that defective chloride secretion is not the primary event in the CF lung, indeed in the CF mouse, lung disease is sub-clinical and it is indeed the ENaC mouse model that most closely resembles the CF phenotype.

Although the majority of children with cystic fibrosis now have excellent lung function nutritional issues, remain an issue for a substantial minority. It is now clear that pancreatic insufficiency is only part of the pathological process that leads to non-absorption and suboptimal nutrition; this in turn correlates strongly with survival. The US CF Foundation currently sees improving nutrition as its principal aim in clinical management. It is, therefore, vital that paediatric gastroenterologists take an increasing role in the management of children and young people with cystic fibrosis, not only to support nutritional deficiencies, but also to investigate and manage the increasing number of dis-mortility related disorders associated with cystic fibrosis and also to understand and treat the increasing burden of end stage liver disease which has become apparent as overall survival has increased.

Respiratory paediatricians should look after children and young people with Cystic Fibrosis

Dr Mark Everard, Consultant Respiratory Paediatrician, Sheffield Children's Hospital, Sheffield, S10 2TH

ABSTRACTS PLENARY SESSION THURSDAY 29TH JANUARY 2009

A BSPGHAN audit of the effectiveness and safety of adalimumab in children and young people with Crohn's disease

RK Russell[1,2], ML Wilson[3],N Shah[2],G Mahdi[2], W Hyer[2], C Spray[2], C Daman[2], R Heuschkel[2], N Afzal[2], M Elawad[2], F Torrente[2],N Ayub[2], J Fell[2],GT Ho[2], S Naik[2], DC Wilson[2,3].
1. Department of Paediatric Gastroenterology, Yorkhill Hospital, Glasgow.
2. BSPGHAN Adalimumab study group.
3. Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh and Child Life and Health, University of Edinburgh

Introduction: Adalimumab (Humira®, Abbott UK) is a humanised anti-TNF therapy that has been shown to be efficacious for induction and maintenance of disease remission for adults with Crohn's disease (CD). Experience in children however is extremely limited, with only two case reports published to date.

Aim: We aimed to summarise the UK experience in children with CD in terms of effectiveness and safety.

Subjects and methods: All members of BSPGHAN were approached by e-mail and invited to take part in the study during the summer of 2008. Members were asked to identify any patients with CD, aged <18 years at the time of commencing adalimumab, and with at least 4 weeks of follow up. Patient details collected included prior infliximab therapy and surgery. Adalimumab dosing schedule, dose escalation and side effects were collected. Adalimumab effect was assessed using the PCDAI or Physicians Global Assessment (PGA). PGA was classified as remission, steroid-free remission, no response or reaction leading to withdrawal. Response to adalimumab was recorded at 1, 6 and 12 months.

Results: 26 children and young people received at a median (range) of 14.5 (3.3-17.8) years; 18 (69%) were male, CD had been diagnosed at a median (range) age of 10.5 (2.1-13.7) years, and 10 (38%) and 24 (92%) had required surgery and treatment with infliximab respectively prior to trial of adalimumab. In terms of induction of remission, 8 (31%) entered remission, 9 (35%) had a response and 9 (35%) had no response respectively at week 4. However, 25 (96%) proceeded to maintenance therapy with adalimumab, and 7 (28%) of these required escalation of therapy (shortening of frequency or increase of dosage). Eighteen were followed up to 6 months and twelve to 12 months of adalimumab maintenance therapy; of these, 14 (78%) and 10 (83%) were in remission at 6 and 12 months respectively and 1 (6%) and 1 (8%) had a response compared to baseline status at 6 and 12 months respectively. Seventeen (65%) reported pain at the injection site and 6 (23%) reported serious side effects. Of these, there was 1 report each of serious bacterial infection, serious viral infection, transient visual loss, severe nausea and pain, transient leucopaenia, and a stomal bleed requiring surgical revision.

Summary and Conclusions: In UK clinical paediatric IBD practice, adalimumab is generally given to patients with disease refractory to medical and surgical therapy. The remission rates of 31%, 78% and 83% at 1, 6 and 12 months respectively show adalimumab has a place in these treatment-refractory patients. A significant minority require escalation of therapy (28%) or have serious adverse reactions (23%). With only 12 (46%) being followed up past 12 months of adalimumab therapy, there is a paucity of long-term follow up in terms of safety. These factors must all be borne in mind and support the role of exit strategies for biological therapies in paediatric Crohn's disease.

A 20 year single centre experience of Home Parenteral Nutrition

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Aim

Children with prolonged intestinal failure (IF) are frequently discharged home to continue parenteral nutrition. Our aim was to review outcomes of 68 children discharged home over 20 years. This has been a period of advances in medical, surgical and nutritional management and two 10 year cohorts were identified for comparison.

Subjects and Methods

Children discharged for home parenteral nutrition (HPN) between December 1987 and 2008 were identified. Details of diagnosis, age at discharge, duration of HPN and outcome were obtained from medical, nursing and dietetic records.

Results

There were 15 children in cohort 1 and 53 in cohort 2. The predominant diagnosis in both cohorts was short bowel syndrome (53% in cohort 1 and 53% in cohort 2). Other diagnoses include microvillous inclusion disease, phenotypic diarrhoea, motility disorders, graft versus host disease, enteropathies and Crohn's disease.
Outcome data is shown in Table 1 and Table 2

Table 1 Patient Outcome data

		1987-1997	1998-2008
Number of patients discharged on HPN		15	53
Median age at discharge (months)		16.5 (6-197)	11.3 (4-167)
Median duration of HPN (months)		22.5 (1-228)	13 (0.5-122)
Outcome:	Discontinued HPN (Full EN)	6 (40%)	28 (53%)
	after liver transplant)	(0)	(5)
	Continues HPN	2 (13%)	12 (23%)
	Died	7 (47%)	13 (24%)
	IFALD	5	7
	Sepsis	0	3
	Other	2	3
Transition to adult services		4	1

Table 2 Patient Outcome data – short bowel syndrome

		1987-1997	1998-2008
Number of patients		8	28
Median bowel length (cm)		30 (10-50)	33 (18-80)
Median age at discharge (months)		10.0 (6-17)	11.2 (5.5-18)
Outcome:	Discontinued HPN (Full EN)	3 (37%)	21 (75%)
	(after liver transplant)	(0)	(5)
	Median time to full EN (months)	96 (84-228)	13.3 (2.0-84)
	Continues HPN	1 (13%)	3 (11%)
	Died	4 (50%)	4 (14%)
	IFALD	4	2
	Sepsis	0	2

Summary and conclusion

Comparing the two cohorts there has been more than a three-fold increase in the number of children receiving HPN, primarily due to increased referrals of surgical neonates. Earlier discharge was achieved in the second cohort and was helped by an improved nursing homecare service. Overall survival in cohort 1 was 53% compared to 76% in cohort 2. 40% children ultimately achieved full EN in cohort 1 and 53% in cohort 2. Looking specifically at the outcome of children with SBS there was an overall survival rate of 50% in cohort 1 compared to 86% in cohort 2. 37% achieved full EN in cohort 1compared to 75% in cohort 2 reflecting our increased use of specialised and aggressive EN in these children who continue to have the potential for gut adaptation after discharge.

Haematopoeitic Stem Cell Transplant for Severe Inflammatory Gut Disorders with Immune Dysregulation

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Introduction/Background;

Severe inflammatory gut disorders due to immune dysregulation present a challenge for clinicians and may fail to respond to conventional immunosuppressive therapy.

Therefore significant number of children ends up on long term parentral nutrition or suffers the sequences of long term immunosuppressive therapy.

Aim;

The aim of this study was to determine if the natural history of severe inflammatory gut disorsers can be altered by hematopoietic cell transplants from healthy allogeneic donors.

Subjects and Methods;

We report here seven children with severe inflammatory gut disorders who were refractory to conventional immunosuppressive therapy.

The seven cases consisted of two brothers with intractable ulcerating enterocolitis (IE) of infancy, four children with severe autoimmune enteropathy and intestinal failure, one child with IPEX and one child with severe panenteric fistulising disease. Five children presented within the first three months of life while the other two presented at the age of two and eight years. All patients failed to respond to immunosuppressive therapies including high dose steroids, Azathioprine , Tacrilomus, Thalidomide and Monoclonal antibodies.

After evaluation by a multidisciplinary team including gastroenterologist, immunologist and bone marrow transplant team, all children underwent allogeneic Haematopoeitic Stem Cell Transplantation (HSCT). Two children received matched family donors and five received matched unrelated donors. All patients were conditioned using reduced intensity. Median follow up post HSCT is 18 months (0.5 to 8 years).

Results:

All patients were successfully engrafted with significant clinical improvement. Five children manage to stop all immunosuppressive therapy within one year following HSCT. One patient is now five months post transplant and currently on cyclosporine for GVHD prophylaxis and one patient died of pre-existing lung disease. Five children are now on full enteral feeds and one is currently weaning off TPN. One patient was discharged home at the age of two years for the first time since birth. Complete mucosal healing was confirmed histologically in four and significant improvement was noted in two children.

Summary and Conclusion.

This is the first reported series of patients with severe inflammatory gut disorders who were successfully treated with HSCT. HSCT may be considered as a potential option of treatment for children with severe inflammatory gut disorders that is refractory to conventional immunosuppressive therapy.

Multidisciplinary approach is crucial for patient selection and appropriate type of transplant.

Faecal calprotectin remains high during enteral feed and corticosteroid therapy in newly diagnosed paediatric Crohn's disease.

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Background Calprotectin is a protein found abundantly in all body fluids and is raised in proportion to the degree of inflammation present; it is distributed evenly in faeces. Faecal calprotectin (FC) is increased in inflammation of the bowel and levels of >50ug/g have consistently demonstrated organic gastrointestinal disease. FC has been shown to be a useful tool for predicting relapse in children with Crohn's disease (CD). FC levels can also accurately predicted mucosal healing. In children with inflammatory bowel disease (IBD) FC has been shown to be higher than controls regardless of disease activity. Here we present further data which may help towards our understanding of the application of FC measurement in a clinical setting.

Subjects and methods As part of a larger study looking at the use of enteral feeds to treat newly diagnosed paediatric CD we measured FC as a marker of mucosal inflammation and compared the results with paediatric crohn's disease activity index (PCDAI) remission and future relapse rates.

Children aged 5-15yrs with suspected IBD were recruited to the study. Those with PCDAI ≥11 and endoscopic evidence of CD went on to receive enteral feeds for 6 weeks. Those with no clinical improvement during the six week treatment period were commenced on steroids. FC was assessed in all children at 0 wks. In those with CD, FC was measured and PCDAI was recalculated at 6 weeks. Those without CD were not followed up in this study.

Results Thirty nine children (M=21 F=18) aged 5-16yrs provided faecal samples at endoscopy. Thirty were diagnosed with CD, PCDAI: median (range) 35 (13-55), 2 children had ulcerative colitis, 1 ileal lymphoid hyperplasia and 1 lymphocytic colitis, 5 had no histological abnormality of the ileum or colon.

5/5 children with normal histology had a normal (<50ug/g) FC: 15 ug/g (5-33). Two(6%) of the 32 children with CD had a normal FC at 0 weeks. Both were male, child one had a duodenal stricture and severe gastritis but no histological abnormality of the ileum or colon. PCDAI at 0 wks: 37 and 6: wks 0. Child two had acute and chronic granulomatous inflammation (non casseating) of the ileo caecal valve, caecum and colon. PCDAI for this child was 32.5 at 0wks 0 at 6wks.

During the 6 weeks treatment period all 30 children with CD achieved remission according to their PCDAI: 3 (0-10) (PCDAI unavailable for one patient). In 3/30 (10%) feeds did not improve clinical observations and corticosteroids were commenced.

28/30 children with histological evidence of CD had raised FC at 0wks: 1892ug/g (35-2730). Only 17/30 stools were available for analysis of FC at 6 weeks and only 1/17 had a normal FC level (48ug/g). In the other 16 FC remained high: 1113ug/g (48-2690). However in these 17 children there was an overall mean reduction in FC of 27% over the six week treatment period.

Of the 30 children with CD, 21(70%) relapsed within two years the median length of time to first relapse was 170 days (53-301).

Summary These results suggest that in treated CD, FC falls in line with clinical improvement but does not normalise within the 6 week treatment period. This indicates that complete mucosal healing had not been achieved despite normal PCDAI and prolonged clinical remission. As has been previously hypothesised it is likely that there is a threshold of disease activity at which a patient becomes asymptomatic and this persistently raised FC has previously been demonstrated in children with chronic colitis treated with glucocorticoid therapy.

Conclusion Our results cannot support the use of FC as a marker of clinical remission and success of treatment in paediatric CD.

How to develop your MCN using Telemedicine! The Scottish Paediatric Gastroenterology, Hepatology and Nutrition Group (SPGHANG) experience

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4. Department of Paediatric Gastroenterology, Royal Aberdeen Children's Hospital. 5. Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Yorkhill, Glasgow.

Background: Emphasis has been placed on the development of Regional and National MCN working models for Paediatric Gastroenterology, Hepatology and nutrition (PGHAN) in the U.K. SPGHANG is a multidisciplinary society whose primary goals involve the development of close working relationships between health-professionals with the aim of improving paediatric gastroenterology services. Regular multi-agency CPD and the development of management guidelines are integral to these aims. The geographic distribution of paediatric services within Scotland limits the ability of such groups to develop effective working relationships for MCN development. In 2005 SPGHANG piloted the introduction of bi-monthly telemedicine education sessions to help address these issues.

Aims: We assess the development of the SPGHANG telemedicine sessions over the first three years, with reference to increasing numbers in the group, formalising educational value and expanding the remit of sessions.

Methods: Initial case discussions between physicians in the three lead centres (Glasgow, Edinburgh and Aberdeen) were redesigned in 2006 with the introduction of multidisciplinary presentations and encouragement for presentation from the district general hospital setting. Formal accreditation was introduced by harmonising sessions with RCPCH guidelines on CPD (http://www.rcpch.ac.uk/Education/CPD/CPD-Approval-for-Events) with virtual CPD registration forms, central storage of attendance and session details. Session format was expanded to include, journal club, original research, guideline development and discussion of national planning documentation. The quality of sessions was improved with the development of a code of conduct relating to formatting of presentations and communication etiquette. Appraisal of effectiveness of telemedicine sessions was measured by number of centres participating in sessions, a review of the session diary, SPGHANG membership, annual feedback forms and attendance at the SPGHANG annual winter meeting. Time expended was estimated from work diary review of booking sessions, speakers, advertising, handouts, emailing presentation, obtaining and collating feedback and amending CPD logbook. Costs were calculated on NHS Greater Glasgow SpR 5 pay-scale (£23.06/hr)

Results: SPGHANG telemedicine has expanded its network to include 11 participating paediatric centres with over 50 health professionals involved and now registered with the SPGHANG group. The majority of annual feedback rated sessions as either good or very good in terms of quality of presentations, relevance to current practice, CPD needs. It has been requested that sessions be increased from bimonthly to twelve times a year. Responders also suggested 'real-time' discussion of problem cases or the teaching of practical skills such as accessing central venous catheters or 'virtual' endoscopy. Sessions have successfully led to the implementation of local guidelines for the management of bleeding oesophageal varices and fulminant colitis as well as developing regional guidance for intestinal failure. This has improved disease understanding and clarified roles and responsibilities for shared care patients. A unified and robust response to the Scottish executive's future planning for tertiary services document has been facilitated by 'virtual' discussion. Attendance at the SPGHANG annual winter meeting has increased by greater than 100% from 2005 to 2007. An initial 4hr were required to organise annual meeting schedule with, on average, 2hr of administration per CPD session. Total time was 16hr annually at a cost of £368.96

Conclusions: The use of telemedicine has developed a high quality regular accreditable CPD for professionals who would otherwise have to travel excessive distances to receive this. Additional benefits of to service development include development of MCN practice and harmonisation of SPGHANG's aspirations for future service development. Telemedicine has helped to cultivate relationships between allied health professionals and forge a group identity for SPGHANG. Such processes may serve to enhance the political influence of groups when responding to national planning documentation and accessing additional resources for regional services and thus improve services for PGHAN patients. Future planners of PGHAN services should note the cost effectiveness of Tele-education, but should budget time and costs into proposed network developments.

RCT of antibiotic prophylaxis to reduce infection rates at paediatric percutaneous endoscopic gastrostomy (PEG) tube insertion

[1]CE Paxton, [1,2]ML Wilson, [3]DM Hoole, [4]FD Munro, [1]PM Gillett and [1,2]DC Wilson. Departments of [1]Paediatric Gastroenterology and Nutrition, [3]Pharmacy and [4]Paediatric Surgery, Royal Hospital for Sick Children, Edinburgh and [2]Child Life and Health, University of Edinburgh.

Background and aim: Infection is a common complication and cause of morbidity of gastrostomy tube insertion. Although meta-analysis has shown the value of antibiotic prophylaxis for adults having PEG tube insertion, there have been no controlled trials in children. We aimed to determine whether a pre-operative single dose of intravenous antibiotic is more effective than a normal saline placebo in the occurrence of peristomal infection at paediatric PEG tube insertion.

Methods: A double blind, randomised placebo-controlled trial of a single intravenous injection of Ceftriaxone or of 0.9 % saline at paediatric (<18 years) PEG tube insertion was performed in a regional paediatric teaching hospital. Children were excluded if they had received any antibiotics in the previous 14 days or required antibiotic prophylaxis for clinical reasons. Randomisation was performed by a pharmacist using consecutive numbered sealed opaque envelopes, previously devised by an uninvolved statistician using a computer random number generator. This pharmacist was the only person aware of treatment allocation. The antibiotic or placebo were packaged in brown opaque syringes, and administered to the subject by the anaesthetist at the induction of anaesthesia. Outcomes were measured by a research nurse, blinded to treatment assignment. Primary outcome, peristomal infection, was measured using the Jain et al (Ann Int Med 1987; 107: 824-8) score - presence and amount of erythema (score 0-4), induration (score 0-3) and exudate (score 0-4). A maximum combined score of 8 on the Jain infection score or the presence of pus (exudate score > 3) and positive microscopy swab was the clinical criterion for infection. The follow up period was 14 days per subject. A microscopy swab was routinely sent on all children at day 7 if still inpatient or at any assessment with a score of 8 on the Infection Score or at any presence of pus (exudate score >3). Primary analysis was performed using intention to treat analysis.

Results: Recruitment to the study was slow due to high prevalent community antibiotic usage, and stopped after 56 children had been enrolled. 7 had major violations (3 PEG tube insertion impossible, 3 study drug not prepared by pharmacy by time of procedure, 1 enrolled in error as already receiving prophylactic antibiotics for procedure) and were removed from the study. The 24 children in the intervention group (54% male, median age 2.3 yr) had a significantly lower peristomal infection rate than the 25 children in the control group (56% male, median age 4.8 yr), at 8% and 32% respectively (P=0.04), with a number needed to treat (NNT) of only 4.2.

Summary and conclusions: Antibiotic prophylaxis significantly reduces peristomal infection rates at PEG tube insertion in children, with a NNT of 4.2. The results of this study have led us to change practice in our institution.

Development and Performance of a New Paediatric Nutritional Screening Tool in a Tertiary and District General Hospital. The PYMS Project

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Background: Identification of children at risk of malnutrition is an important part of any program for providing optimal health. Recent national guidelines state that all patients should be screened for malnutrition on admission and periodically during hospital stay1,2. A number of nutritional screening tools are available for adults but there are no valid screening tools for use in children.

Aim: The aim of this study was to develop and assess the performance of a nutritional screening tool in paediatric medical and surgical wards of a tertiary and district general hospital.

Subjects & Methods: A preliminary tool was proposed using the European Society of Clinical Nutrition and Metabolism guidelines3, based on information regularly collected by nursing staff. The Paediatric Yorkhill Malnutrition Score (PYMS) assesses 4 elements: BMI, history of weight loss, dietary intake and predicted effect of the current hospital admission on nutritional status, with a score of 0-2 for each element. Patients with total score of 2 or more are referred for dietetic review. A four month pilot phase was launched in 4 medical and 1 surgical wards of a tertiary hospital and 1 general paediatric ward of a district general hospital. Prior to the pilot all nursing staff attended awareness sessions and received appropriate training.

Results: There were 1491 eligible patients admitted for care in the tertiary and 372 in the general hospital during the pilot, of whom 1086 (73%) and 242 (65%) respectively were screened on admission. A similar proportion in each unit scored ≥2 (10.6% tertiary, 9.2% general). The distribution of the PYMS individual steps in the patients at high nutritional risk is displayed in table 1.

Table 1: Distribution of the PYMS Individual Steps in Patients at High and Medium Nutritional Risk

	Low BMI	Recent Weight Loss	Decreased Dietary Intake	Acute Condition Effect
High Risk n;(%)	53;(45)	56;(41)	96;(71)	82;(60)
Medium Risk n;(%)	N/A	14;(12)	63;(52)	45;(37)

Sixty patients at high nutritional risk were referred for dietetic review. Thirty three of these patients (55%) were referred for first time with the remainder previously under dietetic care. Fifty one of these (85%) were judged to be at actual risk of malnutrition by the hospital dietitian.

Summary & Conclusion: The great majority of children with high PYMS scores were found to be truly at risk of malnutrition. Half of these patients were newly identified and might otherwise have gone undetected. The high compliance rate shows that its use is feasible in both a tertiary and general hospital setting. Further research is ongoing to assess the validity of PYMS and how this compares to other nutritional assessment/screening methods.

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New developments and drugs in the treatment of acute diarrhoea

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ABSTRACTS FOR FRIDAY 30TH JANUARY 2009

Resource implications and budget impact of managing cow milk allergy in the UK

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Objective: To estimate the current treatment patterns for infants with cow milk allergy (CMA) in the UK and the associated resource implications and budget impact, from the perspective of the National Health Service (NHS).

Methods: A computer-based model was constructed depicting the current management of newly-diagnosed infants with CMA over the first year following initial presentation to a GP. The model was based on the case records of 1,000 randomly selected infants with CMA from the Health Improvement Network (THIN) database, which comprises the longitudinal medical records of 5 million patients in the UK. Clinical outcomes and CMA-related healthcare resource use were extracted from the case notes. Using national unit resource costs at 2006/07 prices the model estimated the 12-monthly NHS cost of treating infants with CMA from the time they initially present to their GP.

Results: Patients presenting with a combination of gastrointestinal and atopic symptoms accounted for 59% of all patients whereas those with urticaria accounted for <10%. The age at the time of presentation was a mean 3.0 months. From the initial GP visit for CMA it took a mean 2.2 months to be put on a diet, a mean 2.9 months to achieve symptom resolution (although this varied according to diet) and a mean 3.6 months for a diagnosis to be made. Treatment patterns varied according to presenting symptoms with 60% of all infants being initially treated with soy, 18% with an extensively hydrolysed formula (eHF) and 3% with an amino acid formula (AAF). A mean 9% of patients were intolerant of soy and 29% were intolerant of an eHF. Patients had a mean 18.2 GP visits over the 12 months and a mean 42% of patients were referred to a specialist physician. The total NHS cost of managing CMA over the first 12 months following initial presentation to a GP was estimated to be £1,381 (95% CI: £1,115; £1,649) per patient and £25.6 million for an annual cohort of 18,350 infants.

Conclusion: CMA imposes a substantial burden on the NHS. Any strategy that can improve healthcare delivery and thereby shorten time to treatment, time to diagnosis and time to symptom resolution should potentially decrease the burden CMA imposes on the UK's NHS and release resources for alternative use

There remains a need for biopsy in celiac disease

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There is no longer a need for biopsy in coeliac disease

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Serological testing is routinely used to diagnose coeliac disease (CD). The current standard practice is to confirm the diagnosis of CD with a duodenal biopsy.

Hopper et al (2008) retrospectively analysed the medical records of patients who had had a gastroscopy and duodenal biopsy to diagnosed coeliac disease (CD). They investigated individuals on the basis of indication for gastroscopy as high or low risk of developing coeliac disease. The authors developed a clinical decision making algorithm for subject referrals over 26 months. They categorised patients as either high or low risk of developing CD. They evaluated blood samples for anti-IgA, anti-tTG status and duodenal biopsies according to the Marsh criteria. They proposed an algorithm from their results in patients referred for endoscopy.

The combined results revealed a sensitivity for detection of high risk individuals with both a positive anti-tTG and abnormal biopsy of 100%. 9.1% of patients with CD were anti tTG negative in the high risk group. No cases of coeliac disease were missed and 58% of patients could have avoided a biopsy if the algorithm had been used prospectively.

Several studies reveal that Marsh type 1 changes of coeliac disease can have up to 50% negative coeliac serology, as undetermined by anti-tTG and endomysial antibodies. There are a significant number of cases in which there are positive anti-tTG and anti-endomysial antibodies with normal duodenal biopsy morphology. This group has been termed latent coeliac disease. It is generally accepted that this latter group should not be treated with a gluten-free diet but should be followed up.

Thus, in conclusion, it can be argued that symptomatically high risk individuals with positive anti-tTG antibodies do not need an endoscopy whilst symptomatically low risk subjects do require an endoscopy to confirm the diagnosis.

Care for children with intestinal failure should be based in appropriately staffed and funded specialist centres.

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Short bowel syndrome, severe motility disorders and gastrointestinal mucosal disease account for the majority of patients with long term intestinal failure (IF). Many will be dependent upon PN for months or years before adapting to full enteral feeding while others will never become independent of PN. All are at risk from life threatening complications including catheter sepsis, intestinal failure associated liver disease and venous thrombosis. For an important minority, small bowel transplantation offers the only hope of survival. Expert opinion considers that there are clinical benefits from being managed in a specialist centre (1,2,3) and emphasises the crucial role of nutritional support teams (4).

Recent reports have highlighted the vulnerability of these patients (high morbidity and mortality) as well as raising important concerns about lack of equity with regard to access to specialist services (5). In fact, current provision has developed on an ad hoc basis, and centres that are not adequately organized or funded will find it difficult to meet rising demand. The National Commissioning Group for Highly Specialised Services has recently published a strategic framework for IF and home parenteral nutrition services for adults with specific recommendations for organization and commissioning (6). We need to lobby the Department of Health to ensure that a similar exercise is undertaken on behalf of the increasing number of children and young people with IF. There is a need for investment in specialist staff (e.g. nutrition nurses, pharmacists, dieticians, speech and language therapists, interventional radiologists, etc.) and multidisciplinary teams facilitating home parenteral nutrition and networking with non-specialist units. The challenge ahead is to convince commissioners that such services are worthwhile (7). The British Society of Paediatric Gastroenterology and Nutrition, and the British Association of Paediatric Surgeons have recommended that care be based in designated centres and have described the requirements of such units (8). Few would argue against the principle of centralisation of services, while the optimal number of centres remains uncertain. This will be clarified in time through a clearer understanding both of need (currently being investigated through the British Intestinal Failure Survey) and the level of investment required to support a properly staffed and supported unit.

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A Fox amongst the Chickens

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Rarely has a single area of basic immunology so consistently suggested that it may be a true "master regulator" of inflammation as the transcription factor Foxp3.

What is Foxp3?
Answer: A protein (73 KDa, 431 amino acids) with DNA binding ability that migrates in to the nucleus and interacts with 11,000 gene promoters (to stimulate or repress mRNA transcription). Binding of Foxp3 results from the T-cell receptor binding to antigen / HLA class II on antigen presenting cells. Expression of Foxp3 is restricted to the subclass of T lymphocyte, Treg (or regulatory T-cell) with surface markers CD4+CD25+.
The net effect of Foxp3 activity is to switch off local inflammation (both adaptive and innate).

What is the importance to gut immunity?
Answer: With regards to onset of inflammation:

i) Total absence: This leads to IPEX syndrome with multi-organ autoimmunity (if there is a mutation in the Foxp3 gene). Selective depletion of Foxp3 producing cells leads to spontaneous intestinal inflammation1 either enteropathy or panenteritis.
ii) Deficiency state: Either via partial depletion of Treg cells or presence of mutation in non-coding intron 1 (<50% expression of Foxp3)2, leads to severe food allergy and intestinal inflammation (colitis).

With regards to resolution of inflammation:

i) Intestinal GvHD can be effectively prevented in mice by action of Foxp3 producing cells3.
ii) Both Crohn's and UC appear to regress in tissue populated Foxp3+cells4 5.

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Paediatric GORD management: where's the evidence? Update on the ESPGHAN and NASPGHAN conjoint reflux working group 2007- 2009

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Quality of Life in GI disorders

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Issues relating to quality of life and more specifically health related quality of life are more and more frequently cited and demanded as part of research projects indeed quality of life is regularly used as a secondary outcome in clinical trials and is integral to health economic models. This presentation will ask what we mean by quality of life, what specifically we can use to objectively assess quality of life, how children and their families experience a change in quality of life during periods of illness and adaptation to illness and how clinical assessment of quality of life plays a part in our day to day management of children who are diagnosed with GI disorders.

Confocal Endomicroscopy: A New Tool in the In Vivo Diagnosis of Coeliac Disease

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Background and aims
Confocal laser endomicroscopy (CLE) is a recent development which enables surface and subsurface imaging of living cells in vivo at x1000 magnification. The aims of the present study were to define confocal features of coeliac disease and to evaluate the usefulness of the CLE in the diagnosis of coeliac disease in children in comparison to histology.

Methods
9 patients (7 female) with a median age 8.1 years (range 2-10.5) and weight of 23 kg (range 10.5 -71) with positive coeliac serology and 10 controls with a median age 10.9 (1.8-15.5) and weight of 34.7 (12.6-63.3) underwent oesophago-gastro-duodenoscopy (OGD) using the confocal laser endomicroscope (EC3870CILK; Pentax, Tokyo, Japan). Intravenous sodium fluorescein and topical acriflavine were used as contrast agents. Histology of coeliac disease was graded according to Marsh classification. Confocal features of coeliac disease were defined prior to blinding. These included loss of surface villous architecture, presence of broad villi, infolding of villi, intervillous bridging ("sticky villi") and decreased goblet cells in Marsh type 3b (partial villous atrophy) and absence of villi, crypt hypertrophy and decreased goblet cells in Marsh type 3c (total villous atrophy). Histologic sections were compared with same site confocal images by 2 experienced paediatric histopathologists and endoscopists, who were blinded to the diagnosis, respectively.

Results
The median procedure time for OGD was 16.4 minutes (range 8-25). A total of 1273 confocal images from both patients and controls were compared with 44 same site duodenal biopsies. 6 patients with coeliac disease had crypt hypertrophy and total villous atrophy (Marsh type 3c) and 3 had crypt hypertrophy with partial villous atrophy (Marsh type 3b). Sensitivity and specificity for the diagnosis of coeliac disease were 100% and 89% with a kappa coefficient for inter-observer agreement between the paediatric gastroenterologists was 0.758. In addition 74% of the images were considered to be of good quality.

Conclusion
Confocal endomicroscopy offers the prospect of diagnosis of coeliac disease during ongoing endoscopy. It also enables targeting biopsies to abnormal mucosa and thereby increasing the diagnostic yield especially when villous atrophy is patchy in the duodenum.

The Health Economics of Gastrostomy Insertion and Fundoplication in Children with neurological Impairment

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Introduction
Gastrostomy tube (GT) feeding in children with neurological impairment (NI) has been shown to improve weight gain, reduce feeding times and improve Quality of Life. The development or exacerbation of gastro-oesophageal reflux (GOR) following GT insertion is associated with frequent concomitant use of fundoplication. Anti-reflux surgery in children with NI is associated with high recurrence rate and significant morbidity and mortality. The associated costs to the health service of surgery in children with NI for gastrostomy insertion with or without fundoplication, and subsequent follow up, have not been investigated. This comprehensive health economics study has used detailed information from hospital records to assign costs to all stages of gastrostomy and anti-reflux surgery in this patient group.

Aim
The study aim was to accurately assess all costs associated with gastrostomy insertion with or without fundoplication at a tertiary referral centre from the initial surgical referral through to the time of data collection. The primary outcome measure was the total cost to the health service of performing GT insertion with and without fundoplication, and a UK NHS economic perspective was adopted.

Subjects and methods
Only children with NI were included in the study, exclusion criteria being the presence of genetic, metabolic or neurodegenerative disease. Study patients were identified from the operating and endoscopy diaries of the Paediatric Surgeon and Paediatric Gastroenterologist in a tertiary referral dating from January 2000 until December 2005. This allowed a minimum follow up period of 1 year. Of the 167 gastrostomies inserted in that time 76 children were eligible to take part and of these 52 parents/guardians consented to the study.
Patient case notes for all children were retrieved and the cost areas investigated were: out-patient attendance, ward admissions, surgery/endoscopy, radiology, allied health professionals, and pathology. Costs were examined over a minimum period of 1 year per patient from the initial procedure to capture maintenance and replacement costs and subsequent admissions.
All costs are reported in GB pounds. Unit costs were assigned using Trust Financial Returns, NHS Reference Costs, Personal Social Services Research Unit data, manufacturers and the British National Formulary, and each data point was inflated to 2006 rates using the Pay and Prices Index. The total cost of each category per patient was calculated by multiplying the data on patient service use with the relevant unit cost.

Results
A total of 84 gastrostomy insertions (including revision to button device) were performed in the time period. Of these procedures 34/52 had concurrent fundoplication. The average cost of insertion of GT insertion was £408.42. The average cost of insertion of GT plus fundoplication was £1581.52. Significant differences (95%CI) in cost were associated with performing anti-reflux surgery (p=0.0008) including the cost of out-patient follow up (p=<0.0001) and allied health professional support (p=0.04). The cost of ward admissions for the anti-reflux surgery itself was not significantly different from those associated with insertion of the GT (p=0.059) but costs for subsequent admissions for related complications were significantly different (p=0.018) in those who had a fundoplication.

Conclusion
A recent Cochrane review highlighted the uncertainty regarding the optimal treatment in relation to the outcome of fundoplication surgery versus anti-reflux therapy for gastro-oesophageal-reflux in the NI child undergoing GT insertion.
The data presented here contributes to this debate by highlighting the significant additional costs associated with fundoplication in this patient group.

A sudden and marked reduction in PN cholestasis on changing from a conventional intravenous lipid source to SMOF lipid

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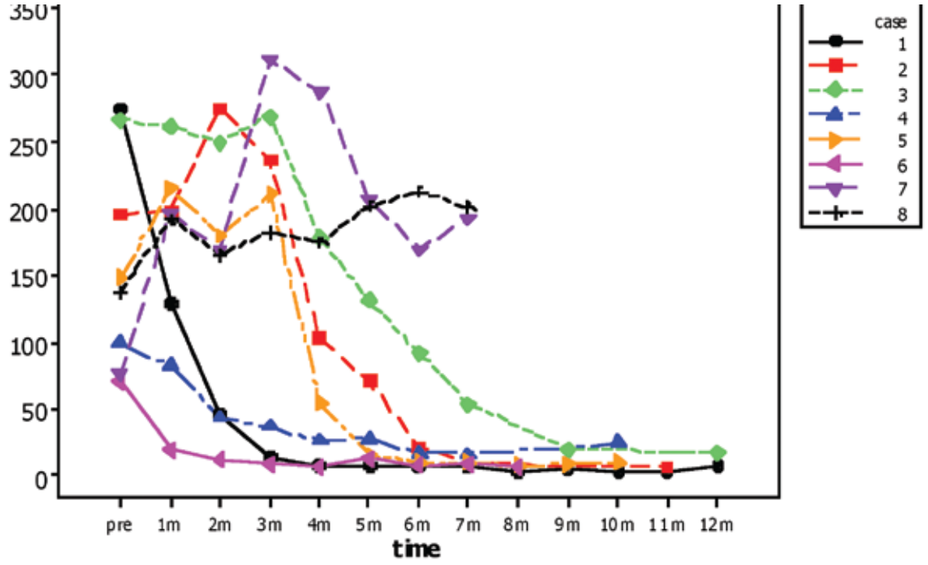
Background. Lipids used in PN are suspected of having detrimental effects on immunity leading to a risk of infection. Recently novel lipid formulations have been introduced aimed at positive modulation of immune function. Conventional lipid emulsions such as Intralipid (Fresenius-Kabi) are prepared from soybean oil. SMOF lipid (Fresenius-Kabi) is a complex mixed-type emulsion including soybean oil (30%), medium chain triglyceride (30%), olive oil (25%) and fish oil (15%).

Aim. We describe our experience with a series of infants and young children with PN cholestasis who were changed from Intralipid to SMOF. A comparison is made with a historical pre-SMOF cohort.
Subjects & Methods. We reviewed the records of all those changed to SMOF because of cholestasis over the year following its introduction in our unit, and compared them with cholestatic children receiving Intralipid during the previous year. Only those receiving PN for at least 6 months were included.

Results. In total 8 were changed to SMOF (short bowel syndrome=4, phenotypic=2, autoimmune enteropathy=1, idiopathic protracted diarrhoea=1). The comparison group also consisted of 8 children with PN cholestasis. In both the SMOF and comparison groups although there was a fall in the amount of PN energy and lipid being received after 6 months of follow-up, both groups were still receiving large and comparable amounts of PN. In the SMOF group one died of liver disease and another is awaiting liver transplantation. However in remaining six there was a sudden, often dramatic and sustained fall in bilirubin about 1-3 months after commencing SMOF



Repeated measures analysis of variance showed a significant fall in bilirubin during the period of follow up (p=0.03). Least Significant Difference methods to control for multiple comparison errors showed statistically significant falls in the first 2 months after starting SMOF. The comparison group (n=8) by contrast had a steady and significant increase in bilirubin during a 6 month follow-up period.

Conclusion. The sudden fall in bilirubin observed with SMOF was striking and unexpected. Lipids have potent biological effects, and these could be beneficial in preventing or treating cholestasis. A RCT should be undertaken to evaluate the efficacy of SMOF in preventing PN related liver disease in young children

Childhood-onset versus adult-onset inflammatory bowel disease: Genotype

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Background & Aims: Our current understanding of the genetic basis of the inflammatory bowel diseases (IBD), Crohn's disease (CD) and ulcerative colitis (UC), is incomplete. Even though 30 distinct loci have been identified in the genetic susceptibility to CD following a meta-analysis of the genome-wide association studies performed to date, the most recent estimate is that they explain not more than 20% of the genetic risk.(1) In UC only a handful of genome-wide significant loci have been identified.(2,3) These studies have mainly recruited adult-onset IBD patients. In Scotland, we have replicated the association of childhood-onset CD with germline variation of NOD2/CARD15 and IL23R. Based on its distinct phenotype, we hypothesised that childhood-onset IBD provides a unique patient cohort for uncovering novel IBD genes. We therefore set up an international collaboration with groups in the USA, Canada and Italy to enable us to study the genetics of childhood-onset IBD.

Subjects & Methods: DNA samples from 2414 patients with childhood-onset disease IBD and 6197 genetically matched controls were genotyped, using the Illumina Infinium™ II HumanHap550 BeadChip technology (Illumina, San Diego), at the Center for Applied Genomics at the Children's Hospital of Philadelphia. All patients were diagnosed under the age of 19 years and fulfilled standard IBD diagnostic criteria. Phenotype characterization was based on the Montreal classification. The mean (SD) age of diagnosis was 11.8 years +/- 4.01 for IBD, 12.0 +/- 3.84 years for CD, and 11.4 +/- 4.38 years for UC. The Research Ethics Boards of the respective Hospitals and other participating centers approved the study, and written informed consent was obtained from all subjects.

Results: Of the 30 CD loci implicated by meta-analysis, 19 showed evidence of replication ($P<0.05$, Bonferonni adjusted) and 11 loci were genome-wide significant in their own right. Further analysis is currently underway to identify childhood-onset specific loci. While the agreement of our results with published CD and UC reports shows that there are many commonalities in the genetic pathogenesis of adult and childhood-onset IBD, preliminary data suggest a number of additional loci influence the onset of IBD during childhood.

Conclusions: The distinct phenotype of childhood-onset IBD cannot be explained by a distinct genotype of the mainly adult-onset loci implicated thus far in IBD susceptibility. Further analysis of childhood-onset specific loci is currently underway and will be presented at the meeting.

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Childhood-onset versus adult-onset inflammatory bowel disease: Phenotype

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Background & Aims
Childhood-onset inflammatory bowel disease (IBD) might be etiologically different from adult-onset IBD. We analyzed disease phenotype and progression of childhood-onset disease and compared them with characteristics of adult-onset disease in patients in Scotland.

Methods:
Anatomical location and behaviour were assessed in 416 patients with childhood-onset (276 CD, 99 UC, 41 IBDU diagnosed before 17th birthday) and 1297 patients with adult-onset (596 CD, 701 UC) IBD using the Montreal classification.

Results:
At the time of diagnosis in children, Crohn's disease (CD) involved small bowel and colon (L3) in 51% (138/273), colon (L2) in 36%, and ileum (L1) in 6%; the upper GI tract (L4) was also affected in 51%. In 39%, the anatomical extent increased within 2 years. Behavioural characteristics progressed - 24% of children developed stricturing or penetrating complications within 4 years (versus 9% at diagnosis; $p<0.0001$, OR 3.32 [1.86-5.92]). Compared with adults, childhood-onset disease was characterized by a 'panenteric' phenotype (ileocolonic plus upper GI [L3+L4]) (43 % versus 3%; $p<0.0001$, OR 23.36 [13.45-40.59]) with less isolated ileal (L1) (2% versus 31%; $p<0.0001$, OR 0.06 [0.03-0.12]) or colonic disease (L2) (15% versus 36%; $p<0.0001$, OR 0.31 [0.21 -0.46]). Ulcerative colitis (UC) was extensive in 82% of the children at diagnosis, versus 48% of adults ($p<0.0001$, OR 5.08 [2.73-9.45]); 46% of the children progressed to develop extensive colitis during follow-up. 46% of children with CD and 35% with UC required immunomodulatory therapy within 12 months of diagnosis. The median time to first surgery was longer in childhood-onset than adult-onset patients with CD (13.7 versus 7.8 years, $p<0.001$); the reverse was true for UC.

Conclusions:
Childhood-onset IBD is characterised by extensive intestinal involvement and rapid early progression.

Is *TauroLock®* the answer to recurrent Central line infections?

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Introduction: Bacteraemia and thrombosis are the major complications of central vein catheters associated with long-term parenteral nutrition (PN). Catheter related bacteraemia (CRB) can be caused by organisms entering from the skin insertion site, from the hub of the device, or from a distant septic focus. Bacterial biofilm is the source of CRB. Instillation of catheter lock solution (CLS) containing antibiotic-anticoagulant may cure and prevent CRB. Most studies report an average CRB incidence of 4-6 episodes per 1000 catheter days.

Aim: The aim of this abstract is to report our experience with the use of CLS - TauroLock® (1.35% Taurolidine and 4% citrate) in the prevention of catheter related bacteraemia.

TauroLock® (Bio-Implant HealthCare, Winsen, Germany)
Taurolidine is a derivative of the amino acid taurine and its mode of action is thought to be due to irreversible binding of its methylol groups to the cell walls of organisms. It is reported to prevent biofilm formation in new catheters and reduce that in already sited catheters. It is suitable for short or long-term catheters and is made up of 1.35% Taurolidine as the antimicrobial component and 4% citrate to prevent thrombus formation. It has broad spectrum bactericidal activity within hours and is effective against a wide variety of gram positive and gram negative organisms as well as fungi. It is not an antibiotic and no antimicrobial resistance has been observed in vitro.
TauroLock® is being marketed for prophylaxis against CRB; the manufacturers claim that infection rate can be reduced by more than 90%, without risk of side effects. ***Taurolock®*** is an unlicensed preparation in the UK but is available from the medicine importers IDIS.

Our experience: We are reporting our experience at the Leeds General Infirmary, where ***TauroLock®*** was used in 2 patients on home PN.
One patient had 11 central line insertions since September 2005 with 24 positive blood cultures (CoNS n = 12; Gram negative sepsis n = 7; Candida n = 5) in 28 months till February 2008. ***TauroLock®*** line lock with 2ml once daily after flushing off PN was commenced on Jan 2008 and there have been no positive blood cultures since then. The central line has been replaced only once due to displacement since commencing ***TauroLock®.***
The second patient had commenced on ***TauroLock®*** in January 2008 after recurrent Fungal sepsis and liver dysfunction. Since commencement of ***TauroLock®*** line locks, there have been occasional positive central cultures for Candida on regular monitoring but no systemic illness and therefore not requiring intravenous antifungal agents. We feel this represents central venous catheter colonization with Candida but the systemic infection was prevented with ***TauroLock®.***
No significant side effects to ***TauroLock®*** were noted in either the patients.

Conclusion: The use of Taurolidine/Citrate (***TauroLock®***) reduced the number of primary catheter related bacteraemia due to CoNS, Gram negative bacteria and Candida. Prevention of septicaemia in presence of colonization was also noted and no significant side effects have been reported. Further studies in children are warranted.

Food reintroduction following exclusive enteral feeding in Crohn's disease

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Background: Elemental feeds were initially used for managing exacerbations of Crohn's disease with gradual food reintroduction.(1) Recent studies suggest that the efficacy of liquid diet therapy (LDT) is unrelated to specific food allergens.(2). Gradual food reintroduction remains common practice throughout the UK despite the majority of centres using a cows' milk based feed for exclusive enteral nutrition (EEN)

Aim: The aim of this audit was to establish current food reintroduction practices amongst dietitians and clinicians in the UK following EEN.

Method: All full BSPGHAN members were sent a questionnaire asking about type of feed used, duration of EEN, other foods allowed and food reintroduction practices and reasons for them. A literature search was also undertaken.
Results: 84 questionnaires (42%) were completed and analysed.

Type of Feed	Number (%)
Elemental	14 (17%)
Polymeric	56 (66%)
Both	14 (17%)

Duration	Number (%)
< 4 weeks	0
4-6 weeks	2 (2%)
6 weeks	56 (67%)
6-8 weeks	3 (4%)
8 weeks	23 (27%)

38% of respondents allowed no additional food or drink during EEN. The remaining respondents allowed a combination of the following drinks and food: water, squash, fizzy drinks, black tea, herbal tea, Bovril, boiled sweets, chewing gum, jelly sweets and ice lollies.
89% of respondents did not think that children with Crohn's disease had an increased incidence of food allergy. Food reintroduction practices varied widely. 18% of respondents introduced normal diet from day 1 and 50% by day 7. Single food reintroduction was favoured by 28% of respondents, the remainder practising empirical exclusion diets. 68% of respondents reported the reason for their practice was historical rather than evidence based. In a study of 102 children undergoing food reintroduction following EEN only 2 remained persistently intolerant (3).

Conclusion: Whilst the majority of centres use a polymeric feed for management of exacerbations of Crohn's disease, single food reintroduction and exclusions remain common practice. There is no evidence to exclude single or multiple foods whilst reintroducing diet after LDT and delaying the introduction of a normal diet prolongs a difficult treatment, may compromise nutritional intake and status and may make further courses of treatment less desirable for the patient. The Associate members of BSPGHAN propose unified practice throughout the UK.

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A comprehensive young person's obesity service – can it exist and can it be effective?

Dr Jerry Wales, Consultant Paediatric Endocrinologist, Sheffield Children's Hospital, Sheffield

In answer to the question posed, the short answer is "No, not currently in the UK".

Comprehensive intervention requires an underpinning of political will and legislation to halt the process at root cause, and has been effective in some nations. Local initiatives are likely to be ineffective for those most in need of the process.

There are behavioural, medical and surgical options that can be effective in certain highly selected cases. The evidence base for these will be reviewed. However there does need to be a national approach to commissioning linked to public health interventions - and there is no evidence of this currently.

There is scope for high quality research into innovative interventions which will be discussed.

Feeding disorders in infants

Professor Carlos Lifschitz, Associate Prof of Paediatrics, Baylor College of Medicine, Houston, USA

Although it is difficult to provide accurate data, it would seem that the incidence of feeding disorders in children is increasing: 25% of children are reported to present with some form of feeding disorder. Increased survival of prematures, infants with complex heart diseases and other previously life threatening conditions may explain some of the organic based problems. Changes in family dynamics, lack of social or family support may be responsible for the behavioral problems. Successful treatment of clinically significant feeding problems involves careful assessment of the full range of influences on the feeding relationship and integrated treatment approaches. However, current diagnostic approaches to feeding disorders tend to be exclusively focused on the child as an individual, and overly concerned with exclusionary criteria. Depending of the clinical condition, a multiaxial diagnosis that describes the child (including medical, developmental, and behavioral characteristics); the parent; the parent-child relationship; and the social and nutritional context of feeding will more accurately

ABSTRACTS POSTER SESSION I
THURSDAY 29TH JANUARY 2009

Feeding aversion and disorders in young children: a guide to management and provision of an MDT feeding clinic and service

Lesley Cogher, Manager, Sheffield Speech and Language Therapy Service, Rye Children's Centre, Tapton Crescent Road, Sheffield

The initial aversive event, or sequence of events, which results in food refusal may be compounded by factors which are internal to the child, implicit in the child's interactions with others, or present in the environment and may extend significantly the amount of time a child takes to achieve a balanced dietary intake. Unless the impact of these factors is evaluated and modified, even successful treatment of the primary condition may not lead to the child's being a willing eater.

This paper will briefly consider the impact of appetite and how it may develop under the influence of the child's experiences, the physical features of food, and how they influence the child's eating pattern. In addition, familial factors such as attachment, parental attitude to food and the mealtime environment will inevitably interact with the features above to create a complex reaction to food which is likely to be unique to each child. This complexity is likely to require multi-disciplinary assessment and therapeutic approaches.

In light of this it will be helpful to consider the role and design of a feeding clinic as part of a pathway which involves access to a range of professionals who can act together in the optimum combination to support children with food aversion.

A single centre experience of adalimumab for the induction of remission in refractory paediatric Crohn's disease

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Background: Adalimumab is a fully human monoclonal antibody to tumour necrosis factor – alpha (TNF – a), currently being used in the management of subjects with refractory Crohn's disease. Indications for its use include an attenuated response to infliximab after initial good response or allergic reactions to infliximab. Paediatric experience with this biological in Crohn's disease is limited.

Aim: To describe our single, tertiary centre experience of using adalimumab in children with the above indications.

Methods: This is a retrospective observational study. Case note analysis of patients treated with adalimumab at our centre was undertaken. All subjects received a loading dose of adalimumab at 24 mg/m^2 followed 2 weeks later with maintenance dose, which was then continued 2 weekly. The Paediatric Crohn's disease activity index (PCDAI) was used to define
1. Remission (PCDAI < 10 at 12 weeks of treatment),
2. Response (PCDAI</ =30 or a reduction of 15 points from baseline at 12 weeks) or
3. Lack of response (PCDAI > 30, increase in PCDAI > /= 15 after starting treatment).

One tailed 'p' values for reduction of PCDAI scores were done by comparing medians of distribution of PCDAI at baseline, 6 weeks and 12 weeks using the Wilocxon matched pairs- signed rank test (non parametric).

Results: Case notes of 15 patients (9 male: 6 female) treated with adalimumab at our centre since march 2007 were reviewed. The median age at diagnosis was 10.3 yr (3.84 – 15.59). The distribution of Crohn's disease was (not mutually exclusive): Ileocolonic -8, colonic -5, perianal – 9, upper GI – 4. Adalimumab was started at a median age of 14.74 yr (5.53 – 16.89). Median duration of infliximab therapy before adalimumab was 10 mo (1- 35). The last infliximab infusion before commencement of adalimumab was 4 wk (2- 156) – mode. The reason for starting adalimumab was allergy to infliximab in 5 subjects and lack of response to it in the remaining 10 respectively. Concomitant treatment included (not mutually exclusive) - mesalazine in 8, azathioprine in 7, high dose steroids in 3 and methotrexate in 2 subjects. Two patients had a diversion procedure done previously. Data including clinical and laboratory parameters for the PCDAI were available on 10 and 7 subjects respectively at 6 and 12 weeks. PCDAI at baseline (median) was 32.5 (15-60), at 6 weeks: 11.5 (2.5 – 35); n = 10, and at 12 weeks: 12 (2.5 - 15); n = 7. The reduction in PCDAI between baseline and 6 weeks was significant (p = 0.002) as was the reduction between baseline and 12 weeks (p=0.007). At 12 weeks of treatment, data was available on 7 patients – 3 were in remission, 4 were showing response. No adverse effects related to adalimumab use were recognised or reported in the studied subjects so far.

Conclusion: Adalimumab seems to be useful in inducing remission and disease response in children with refractory Crohn's disease. Significant reductions in PCDAI occur from baseline within 6 weeks of treatment, maintained at 12 weeks.

A study to investigate the complications of obesity in a group of children participating in the "Balance-It, getting the Balance right" scheme in Gateshead, Newcastle

Anna Hoad and Eleanor Morris – Newcastle Medical School.
Dr Anne Dale - Queen Elizabeth Hospital, Gateshead Health NHS foundation Trust

Introduction/Background
Obesity remains a huge public health issue amongst society. Childhood obesity has increased significantly since 1995 and continues to do so. The North East has one of the highest proportions of obese children in the UK. To combat this, the 'Balance It!' scheme was established in Gateshead, Newcastle which aims to promote weight loss in overweight and obese children. A significant number of mental and physical health risks are associated with obesity. These include high levels of blood cholesterol, blood glucose and blood pressure, which can be early signs of metabolic syndrome. This is known to increase risks of developing diabetes and cardiovascular disease. Obesity is also a cause of psychological ill health, and increases the risk of childhood asthma, liver disease, sleep apnoea and orthopaedic problems.

Aims and objectives
To investigate the presence and prevalence of complications of obesity in children, in particular:

- To identify the pathological signs caused by obesity.
- To identify the frequency of abnormal blood tests in obese children.
- To note the frequency of USS which show fatty livers in obese children.

Subjects and methods
Over a period of 2 weeks in February 2008 we analysed the notes of a population of 39 subjects: 23 girls and 16 boys. All children were included on the basis that they were obese, (classified as being >95th centile) 16 or younger when they first presented in clinic, and at some point had been participants of the 'Balance It!' scheme. We collected the following data: patient characteristics, relevant family history, clinical symptoms and signs and investigations done.

Results
The results showed many children from our cohort had pathological signs caused by obesity. 14% had increased blood pressure, 10% had obstructive sleep apnoea, 7% had fatty liver on ultrasound and 5% had acanthosis nigricans. The largest problem these children faced was psychological. Investigation results showed that 17.95% of children have abnormal LFTs, 15.38% abnormal cholesterol levels, 2.5% low HDL levels, 13% abnormal triglyceride levels, 2% abnormal glucose tolerance tests, 5% abnormal fasting blood sugar and 3% abnormal HbA1c levels.

Summary and Conclusion
This study demonstrates that even within a small cohort a significant proportion of obese children manifest pathological complications. From a national perspective if these findings were extrapolated up, the figures could highlight an astoundingly high incidence of complications in childhood obesity. Further monitoring of this population sample in the form of a longitudinal study would be of benefit to identify the long term implications associated with childhood obesity. National studies are also required to identify the scale of the problem.

Are non-pylori Helicobacter Organisms Associated with Paediatric Ulcerative Colitis? Retrospective observational study.

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Introduction- Non-pylori Helicobacter organisms were first identified as potential inflammatory bowel disease (IBD) pathogens in an animal model of ulcerative colitis (UC). Recent animal studies have shown that infection with non-pylori Helicobacter organisms can reduce commensal bacteria and that an IgG immune response to commensal bacteria precedes the development of colitis after infection with Helicobacter bilis3. These observations together with the reduced bacterial diversity seen in human IBD suggest a role for non-pylori Helicobacter in disease initiation.

Aim- To examine the prevalence of non-pylori Helicobacter organisms in paediatric ulcerative colitis and an appropriate paediatric control group.

Subjects and Methods- 5 years of paediatric colonoscopies in subjects with and without clinical or pathological suspicion of inflammation. 23 UC patients were represented 22 with rectal and 1 with a sigmoid biopsy. 17 (74%) were de-novo presentations. 15 control patients with a heterogeneous mix of clinical suspicions/diagnoses had macroscopically and microscopically normal colons, and all patients were free from antibiotics and systemic steroids in the 3 months prior to biopsy. 52% of the UC group and 60% of the control group were male with respective mean ages of 10.3 years (1.5-14.1) and 9.4 years (1.3-14.3).
Slides from each patient were de-waxed with xylene and ethanol before being hybridised with one of two sets of fluorescent probes. The first set utilised Helicobacter genus probes which fluoresce with the presence of any Helicobacter. The second set utilised both an eubacterial probe which fluoresced with the presence of any bacteria (to exclude false negatives because of destruction of the mucosal layer during paraffin embedding) and a Helicobacter pylori specific probe to exclude false positives because of H. pylori being transited from the stomach. Each set was examined in triplicate and the slide identities were blinded prior to microscopy. Any positive was interpreted as significant for the purposes of analysis.

Results-

		Patient Group		
		UC	Control	Total
Non-pylori Helicobacter	+	20*	6*	26
	-	3	9	12
	Total	23	15	38

*p=0.004

Of 23 UC biopsies, 23 were eubacteria positive, 21 were H. genus positive and 1 was H. pylori positive. Therefore 20/23 (87%) were non-pylori Helicobacter positive. Of 15 control biopsies, 15 were eubacteria positive, 10 were H. genus positive and 4 were H. pylori positive. Therefore 6/15 (40%) were non-pylori Helicobacter positive. Comparing the non-pylori Helicobacter positive results with Pearson's exact test reveals a highly significant p value of 0.004.

Summary- The prevalence of non-pylori Helicobacter species in the distal colon of children with ulcerative colitis is significantly higher than in control cases selected for a macroscopically and microscopically normal colon.

"Can Paediatric professionals reliably identify pale stool?"

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Introduction:
Biliary atresia (BA) is an important surgically remediable cause of neonatal cholestasis. Early recognition by identification of abnormal (pale) stool colours allows earlier surgery and better results. Unfortunately there are no objective means of identifying abnormal stools. Prior to designing a measurement device, we wished to know how reliably trained professionals recognise pale stools.

Methods:
Following ethical approval, stools were collected from normal and cholestatic infants and photographed against a white background with a colour calibration card. The colours were standardised by computers to allow for ambient light. Photographs of 5 normal, 3 indeterminate and 4 acholic stools were chosen and shown to professionals with a questionnaire asking them to classify each stool as "healthy" or "suspect".

Results:
A total of 81 questionnaires were completed by 36 paediatric doctors and 45 paediatric nurses in 3 London teaching hospitals, one of them is a National Referral Centre for paediatric liver disease. Doctors and nurses correctly identified suspect stools at 62.7% and 62.9% respectively.

1. Hospital "A "(Referral centre)

Stool	**Normal**	**Suspect**
Correct	69/100 (69%)	93/139 (66.9%)
Incorrect	31/100 (31%)	46/139 (33.1%)

2. Hospital "B"

Stool	**Normal**	**Suspect**
Correct	121/205 (59.0%)	186/287 (64.8%)
Incorrect	84/205 (41%)	101/287(35.2%)

3. Hospital "C"

Stool	**Normal**	**Suspect**
Correct	70/95 (73.7%)	72/133 (54.1%)
Incorrect	25/95 (26.3%)	61/133 (33.1%)

Conclusions:
There were no significant differences between the 3 institutions or between doctors and nurses in identifying pale stools but doctors were better than nurses at identifying normal stool (71.5% vs. 58.5%). Professionals can not recognise suspect stools to the level of reliability required for identification of biliary atresia. There is a need for objective methods of identifying stool colour.

Colonic strictures in Paediatric Crohn's disease : A Single tertiary centre experience.

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The incidence of Crohn's disease in Scotland is about 3 per 100,000 with a rising trend and prevalence is estimated to be 13.7 per 100,000. About 24% of these children develop stricturing or penetrating complications. A smaller percentage of these develop stricturing large bowel disease presenting with obstructive symptoms. Most of them require elective or even emergency surgery. Despite recent advances, there has been little change in the surgical resection rates. The knowledge of stricture pathogenesis remains relatively limited. There occurs disorganised expansion of the muscularis propria with deposition of abnormal collagen septa and increased activity of Fibroblasts, Myofibroblasts and Smooth muscle phenotypes. The signalling pathways are complex and not fully understood. TGF Beta-1 is the main receptor protein thought to influence the process through cascade involving transcription factor Smad3. Controversy exists on the use of Anti TNF-alpha treatment which may influence the fibrogenesis by blocking the regulatory effect of TNF-alpha on the TGF- Beta signalling. The economic burden of Crohn's disease in UK is as high as £300 million per year and 49% of this is inpatient costs. It is estimated that 40% of this is for surgical intervention and 50% of this is for strictures.

We conducted an observational study of all patients requiring surgery over a 15yr period(1991-2006) in a large tertiary Paediatric gastroenterology referral centre in South West of Scotland. This was by retrospective case notes review of patients who underwent Surgery for Crohn's disease. We studied the disease characteristics and management approaches. There were 31 cases identified of which 9(30%) had Colonic strictures and 19(61%) Terminal ileal strictures. All Colonic strictures were left sided; 3(33%) in descending colon, 4(44%) Sigmoid and 2(22%) in Rectum. 6 had Limited resection and 3 Total Colectomy. The average age at diagnosis was 11.4 years and the strictures developed 36.8 months later. The induction treatments required were 1.5 per year and 6(67%) were steroid dependent. All were on 5-ASAs. 7 had Azathioprine, 3 Methotrexate and 1 had Infliximab infusions prior to developing the stricture. Majority (88%) had poor growth despite optimising the treatment. 88% also had persistent ulcers on repeat Colonoscopy in the recto-sigmoid region. The diagnosis was mainly on Colonoscopy and only 55% tolerated Barium enema. The exacerbation of symptoms was on average for 6.9 months prior to the diagnosis of the stricture with abdominal pain and loose stools being the commonest symptoms.

In summary, Colonic stricture in Children with Crohn's disease in this population is a significant complication with probably higher economic burden. They occur in the left side of the Colon and develop just over 3 years after the diagnosis is made. Majority are steroid dependent and have poor growth. Just over half tolerated Barium enema and hence the diagnosis is mainly based on Colonoscopy. A high index of suspicion is required for the diagnosis of the complication. A better understanding of the patho-physiology and disease characteristics is required to prevent and manage the complication.

D-Lactic acidosis in children, a Case Report and Investigative Protocol

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Background

DLA is a rare complication of short bowel syndrome caused by fermentation of high carbohydrate load in the gut and presents with encephalopathy and metabolic acidosis. It can occur anytime from 2 months to ten years post-intestinal surgery. Only 22 cases have been reported worldwide (Ku et al HK J Paediatr 2006; 11:246-254).

Case report

A 3 year old boy born with type IV bowel atresia had intestinal failure (IF), isolated liver transplant for IF-associated liver disease and was parenteral nutrition-dependent for 19 months. He presented at age three years with ataxia following a respiratory infection. He had 3 separate episodes of presumed DLA during his 2 week admission with recovery between episodes. Paired blood and urine biochemistry on all 3 occasions showed significant metabolic acidosis, increased anion gap, normal hydroxybutyrate, normal L-lactate (Table) but a large D/L lactate peak on urinary organic acid analysis. Subsequent specific quantitative measurement of D lactate in urine (Herrera et al Ann Clin Biochem 2008; 45:177-183) gave extremely high results. He was managed successfully by stopping feeds, administering IV fluids with bicarbonate, antibiotics and dietary modification on reintroduction of feeds.

Episode	**Blood/plasma**					**Urine**
	pH	Std. Bicarb mmol/L	Base Excess mmol/L	Anion Gap mmol/L	L-lactate mmol/L	D-lactate µ mol/mmol creatinine
1	7.16	14.1	-13.6	30.2	1.91	13340
2	7.17	11.9	-16.7	23.7	1.15	15430
3	7.16	9.4	-20.0	27.4	0.8	25790

*Defined as sodium + potassium – (chloride + bicarbonate), reference range 9-23mmol/L

Discussion

Clinical suspicion of DLA in at risk children is key. Our investigative protocol for clinically suspected DLA is paired blood (for gases with anion gap, L-lactate and 3-hydroxybutyrate) and urine (for organic acid profile). Obtaining a quantitative measurement of plasma or urine D-lactate with the above clinical and biochemical findings confirms the diagnosis of DLA and guides management.

Gastroenterology & Psychology: Joining the body and mind!

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Introduction: There are recommendations for clinical psychology attachment to medically specialist teams such as Cystic Fibrosis and Diabetes with the aim of having a multi-disciplinary approach to physical health problems including the psychological effect of those health problems and any more specific mental health issues that arise as a result of physical health problems. This project looks at the level of demand for psychological input and type of intervention delivered in a paediatric gastroenterology team with a view to supporting similar recommendations being made for this speciality.

Aim: To look at the level of involvement from the team's clinical psychologist in a medical in-patient and out-patient setting with a view to assessing the demand for psychological input to paediatric gastroenterology patients.

Method: The team's clinical psychologist receives referrals from the multi-disciplinary team for both in-patients and out-patients across the range of gastroenterological presentations. 52 sets of clinical notes were reviewed retrospectively for the previous 1 year's referrals and information regarding reason for referral and psychological intervention summarised.

Summary: Of the 52 sets of notes 16 referrals were for problems related to Inflammatory Bowel Disease, 15 related to feeding problems, 13 related to constipation; 4 related to pseudo-obstruction, 2 related to TPN, 1 related to abdominal pain and 1 for a suspected eating disorder. Clinical contact with these children and young people was made on an in-patient and out-patient basis and involved co-working with other child health professionals as well as separate appointments with the clinical psychologist. Intervention included behaviour therapy, family work, psycho-education and assessment for more specific mental health disorders. Cases requiring only consultation to other health professionals were not included.

Conclusion: Gastroenterology teams regularly refer to psychological services for a range of GI presentations which require multi-disciplinary assessment and intervention as well as more specific mental health intervention. The paediatric gastroenterology team has one of has the highest referral rates to psychological services of any specialist team in the hospital and yet there are no recommendations for psychological input to gastroenterology teams nationally. A further project looking at the effect of psychological input on number of OP appointments and length of IP admissions would lend support to obtaining funding for sessions dedicated to gastroenterology.

Gastrointestinal disorders in DiGeorge syndrome

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Introduction

DiGeorge syndrome has various features including heart defects, hypocalcemia, velo-pharyngeal insufficiency leading to feeding difficulties and altered T cell response. Gastrointestinal (GI) disorders like celiac disease, necrotizing enterocolitis, Hirschsprung disease have been reported in children with this syndrome but no consistent pathology has been described in literature.

Aim

To assess association of gastrointestinal disorders with DiGeorge syndrome in a cohort of children with DiGeorge syndrome.

Subjects and Methods.

12 children (5 male, 7 female) with DiGeorge syndrome who presented and investigated with GI symptoms were assessed. Symptoms and intestinal biopsies were then reviewed. Age when first biopsies were taken ranged from 5 days to 8 years. Histological findings were then analysed.

Results

9 (75%) had abnormal histological findings.
Five children had upper oesophagealgastroduodenoscopy (OGD) while three had both OGD and colonoscopy. Rectal suction biopsies and surgical resection of a part of small bowel were done in two patients each.
The indications for only OGD were failure to thrive (2), gastroesopaheal reflux (2) and recurrent abdominal pain while for OGD and colonoscopy were dilated duodenum on barium meal, failure to thrive and chronic diarrhoea.
The histology from upper gastrointestinal tract showed duodenitis in 3(38%),esophagitis in 2(25%),mild gastritis in 1(12%) and was normal in 2.The colonic biopsies showed mild colitis in 2 while 1 showed pancolitis.
2 children had rectal suction biopsies,1 had Hirschsprung disease while the other was normal. Indication in former was intermittent abdominal distension while for the latter was constipation.
Two had surgical resection one for ischemic necrosis- on day 5 had necrotizing enterocolitis while inflammatory stricture was the indication in the other.

Conclusion

This is the first study in children to describe different GI pathology in DiGeorge syndrome. In this cohort of symptomatic patients there is a high incidence of GI pathology which might partly be explained by underlying immunodeficiency. We put forward a hypothesis that GI disorders are common in DiGeorge syndrome but a larger study is needed to address this issue.

Haemophagocytic lymphohistiocytosis in an adolescent with crohn's disease on azathioprine

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Case: A 16 year old boy diagnosed with crohn's disease presented 8 months after starting azathioprine as maintenance immunosuppressive therapy with persistent fever and lymphadenopathy. His fever was unresponsive to broad spectrum antibiotics and during his hospital stay he developed hepatosplenomegaly. His blood tests revealed pancytopenia, raised bilirubin and liver enzymes, hypoalbuminemia, deranged clotting and low fibrinogen, high LDH and markedly elevated ferritin levels, raised CSF protein and lymphocytic pleocytosis. His EBV serology was positive and EBV DNA was 42,465 copies/ml. A lymph node biopsy showed reactive changes and bone marrow showed increased macrophages with evidence of haemophagocytosis. A diagnosis of haemophagocytic lymphohistiocytosis was made and he was treated with etoposide, cyclosporine and dexamethasone (HLH4 protocol) which halted the hyperinflammatory state. His fever settled and bloods normalised and he is now clinically well. No abnormal mutations associated with haemophagocytic lymphohistiocytosis were detected.

Discussion: This is an unusual case of EBV triggered haemophagocytic lymphohistiocytosis in an adolescent with crohn's disease on azathioprine. The immunosuppressive therapy may have contributed to the development of this serious complication of EBV infection. Haemophagocytic lymphohistiocytosis is a life threatening condition of severe hyperinflammation caused by uncontrolled proliferation of activated lymphocytes and histiocytes secreting cytokines. Cardinal signs and symptoms include fever unresponsive to antibiotics, hepatosplenomegaly and pancytopenia. Biochemical markers include elevated triglyceride, ferritin and low fibrinogen. It can be genetic or acquired.
Azathioprine is commonly used as maintenance immunosuppressive therapy in patients with inflammatory bowel disease. This case highlights the potentially serious complications of immunosuppression. Awareness of HLH is crucial to diagnose this condition early and start life saving treatment.

How compliant are patients with quiescent IBD to 5-aminosalicylic acid?

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Introduction: 5-aminosalicylic acid (5ASA) has remained one of the mainstay maintenance therapy of inflammatory bowel disease (IBD) for the last 50 years. Recent studies have led to the important considerations that it has a chemoprotective effect against IBD related carcinogenesis1, and that it can interfere with thiopurine metabolism, causing an enhancement of the response to azathioprine and 6-MP2. In view if this, long term compliance to 5ASA is clearly auspicated, although paediatric patients, and especially adolescents, might not fully appreciate its importance.

Aim: To evaluate the adherence to treatment with 5ASA, and the factors influencing it, in a cohort of IBD paediatric patients with quiescent disease.

Subjects and methods: From our database of 150 IBD patients, we identified 45 children who had been in remission for more than 3 months. Out of these, during the study period from March to June 2008, we were able to contact 30 patients. All of them had 5-ASA prescribed either as monotherapy or in combination with azathioprine as maintenance treatment. To minimize variability, both the patients and their parents were asked by the same investigator a series of questions from a standardized questionnaire.

Results: Of the 30 patients surveyed, 24 had Crohn's disease, 5 had ulcerative colitis and 1 had indeterminate colitis. The frequency of treatment with 5ASA varied between 2 to 3 times per day according to standard formulation. 60% (18/30) of patients stated to have omitted at least 1 dose in the previous 2 weeks (noncompliant group, NCG). In this group, 45% (8/18) forgot their medication at least 3-4 times per week. Reasons given for non-compliance were: forgetfulness (10/18), multiple dose frequency (14/18), worries about side-effects (2/18), lack of perceived benefit of medication (8/18). In the NCG, the person in charge of administering the medication was identified as the child him/herself in 72% (12/18) of the cases, whereas 75% (9/12) of subjects in the compliant group (CG) had the parents/carers in charge. There was no significant difference in the age between the NCG (10-17 years, mean 14 years) and CG (11-18 years, mean 15years) groups. However, there was a significant difference in the length of time the patients had been on the medication between the NCG (mean 5 years) and the CG (mean 3 years), p= 0.07.

Conclusions: Our study showed that more than half of the patients with quiescent IBD were not adhering to their medical regime. This may be due to the low acceptability of the currently available 5ASA formulations, which often require multiple daily dosing and a large number of tablets. Compliance was much more an issue when the child was put in charge of their medication. It also appeared that the longer the patient had been on the 5ASA, the less compliant s/he became. When prescribing 5ASA as maintenance therapy, switching to formulations offered in a once daily regime may improve the long term adherence in a selected group of patients.

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2 De Boer NK Am J Gastroenterol 2007 Dec;102(12):2747-53. Epub 2007 Aug 31

"I choked on my azathioprine"- an unusual case of Crohn's Oesophageal Stricture

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Background

Upper GI Crohn's Disease is being increasingly reported and the prevalence of oesophageal (CD) ranges from 6.5% to 43% in children1. Oesophageal Crohn's structuring in adults is unusual and often resistant to treatment. Reports in children are few2.

Aim

To describe a unique paediatric case of an upper oesophageal Crohn's stricture-presentation and response to treatment

Methods

Case note review and prospective follow up

Results

A 16 year old female with pan-enteric CD diagnosed 3 years previously, casually mentioned in outpatients that she had choked a couple of times on her Azathioprine. Her concomitant maintenance treatment was 8 weekly infliximab, Mesalazine and Omeprazole. At diagnosis oesophageal granulomas were seen on histology but no macroscopic CD.

Time 0 Barium swallow- suggestive of short stricture in the upper third of the oesophagus.

T + 3 weeks Endoscopy -Crohn's stricture 16cm from the incisors with active ulceration and inflammation- unable to pass paediatric scope

Patient initially declined treatment/intervention but dysphagia progressed

T+ 5 months. Consent for topical treatment but no dilation.

Endoscopy showed worsening of ulceration with obviously cobblestoning.

Triamcinolone 50mg/5 ml was injected circumferentially in the stricture in 0.5 ml aliquots

T + 6 months Clinical improvement but endoscopy- no change.

1st balloon dilation up to 10mm

T +7 months Persistent stricture on Barium –less mucosal irregularity.

2nd balloon dilation 12 mm – macroscopic improvement inflammation

T+13 months No dysphagia,no stricture radiologically

Conclusion Upper Oesophageal strictures are uncommon but dysphagia in children with CD requires full investigation. Combination of dilation and local topical Triamcinolone leads to resolution

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Improved serum bilirubin with SMOF lipid in children with intestinal failure on long term parenteral nutrition

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Background: Intestinal failure associated liver disease (IFALD) is a well known complication of long term parenteral nutrition (PN) and occurs in up to 60 % of children who require prolonged intravenous feeding. Although the underlying pathophysiology is still not entirely understood, recurrent episodes of sepsis, bacterial overgrowth, hyperinsulism due to excessive or prolonged glucose load and intravenous lipids are known causative factors. SMOF, a new lipid emulsion based on soybean oil, medium chain triglycerides, olive oil and fish oil, is presumed to be better tolerated than traditional lipid preparations with a lower incidence of IFALD (serum bilirubin > 100 µmol/L).

Aim: The aim of our study was to demonstrate the effect of SMOF on serum bilirubin levels in children with established IFALD.

Methods: All children with intestinal failure (IF) admitted to Chelsea and Westminster Hospital receive Intralipid as standard lipid emulsion when commenced on PN. Liver function is monitored on a regular basis. Since the beginning of 2008 patients whose serum bilirubin rose above 100 µmol/L were changed to SMOF lipid and the effect on liver function observed. A total of nine children were identified.

Results: Five patients were female and 4 male. Seven infants were born prematurely. All nine children received prolonged PN (> 28 days). 4/9 had short bowel syndrome secondary to necrotising enterocolitis, 2/9 were born with gastroschisis, 2/9 had congenital atresias of the small bowel and one child was unfeedable due to an undefined enteropathy. Median bilirubin levels at the start of SMOF was 138 µmol/L with a median time on Intralipid prior to commencing SMOF of 53 days. SMOF was well tolerated and no adverse effents occurred. In 3/9 children serum bilirubin levels went back to normal (median 26 µmol/L) and in 3/9 serum bilirubin levels improved (> 10% fall). In 3/9 children serum bilirubin continued to raise. All three patients had recurrent episodes of severe sepsis, which could account for deteriorating liver function.

Conclusion: In our cohort of children with established IFALD SMOF lipid had a positive effect on serum bilirubin level in two thirds of cases. Larger studies should follow to confirm our results.

Conclusion: In our cohort of children with established IFALD SMOF lipid had a positive effect on serum bilirubin level in two thirds of cases. Larger studies should follow to confirm our results.

Incidence of intestinal failure associated liver disease and outcome in infants treated with intravenous nutrition for more than 28 days.

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Introduction/Background

Children requiring intravenous nutrition from the neonatal period or infancy are highly susceptible to developing intestinal failure associated liver disease (IFALD) and are more likely than older children or adults to have a poor long-term outcome.

Aim

The aim of this study was to review the incidence and severity of IFALD and outcome in children with a primary gastrointestinal disorder (medical and surgical) commencing intravenous nutrition (IVN) in the neonatal period and infancy over a two year period from January 2006.

Patients

41 neonates and infants (22 male, 19 female) with a primary gastrointestinal disorder were given intravenous nutrition for more than 28 days.
Diagnoses were 21 short gut, 10 enteropathy, 5 gastroschisis, 3 exomphalos, 2 dysmotility. 18 children were born prematurely.

Methods

Results of liver function tests (bilirubin and ALT) were reviewed at time of starting intravenous nutrition, at 28 days and after stopping treatment or at 3 and 6 months in those still on treatment. The number of deaths and the cause were recorded.

Results

Bilirubin was normal on starting IVN. By 28 days bilirubin was > 100 mmol/l in three infants and 50-100 mmol/l in 8, a total of 27%. At 6 months of age it was still raised in 3 infants (120, 97, 91mmol/l). Nine children or 22% have died from 1-17 months of age (mean and median 5 months). Cause of death was sepsis in all cases. Two of these infants born prematurely had coexisting cholestasis and multi-system disease, two complex congenital heart disease and two bladder or cloacal extrophy.

Bilirubin level has since returned to the normal range in the one surviving patient in whom it was still raised (126mmol/l) at 6 months. Eight children still had raised ALT levels at 6 months up to 177 IU/l.

Summary

Children on long-term IVN from the neonatal period had a high mortality rate of 22%. Liver disease was common affecting 27%. Bilirubin levels fell to below 50mmol/l in virtually all surviving children, but at least 8 children still had raised liver enzymes. Routine treatment to aim to improve liver function included cycling the IVN, reducing lipid infusions to alternate days, early enteral feeding and in 3 children changing from intralipid to an alternative lipid source.

Conclusion

Liver disease is usually reversible in children treated with IVN from the neonatal period who survive beyond the first few weeks of life.

Incidental Iron Deficiency Anaemia in Hospitalised Children – A Missed Opportunity for i ntervention

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INTRODUCTION

Iron deficiency anaemia is the most common and wide-spread nutritional disorder in the world that afflicts not only developing countries but also the developed world. Iron deficiency reduces the potential of affected children and appropriate treatment is known to increase concentration, attitude and school attendance. It is most important therefore that any iron deficiency detected incidentally is recognised and treated appropriately.

AIM

To determine the incidence and management of iron deficiency anaemia in hospitalised children who had a full blood count performed as part of their investigations for their presenting illness.

METHODS

This study was conducted over a one year period at the Shrewsbury and Telford Hospitals NHS Trust and subsequently repeated over a 6 month period at the Worthing and Southlands Hospital NHS Trust.
A retrospective case note review was undertaken of all hospitalised children between the ages of 9 months and 16 years whose blood tests were consistent with the WHO criteria for the diagnosis of iron deficiency (Haemoglobin < 11g/dl and/or MCV <75 fl). Children with a known cause for their anaemia such as coeliac disease and hereditary spherocytosis were excluded from the study. Relevant data was extracted from the case notes to determine the management of these children against the hospital guideline.

RESULTS

63% (4045) of the paediatric inpatients had a full blood count performed during the study period. 171 children were eligible for the study but case notes were retrievable for only 131 and after applying the exclusion criteria, the study group was 86 children. 3.3% had incidental iron deficiency anaemia. 80% of the study group had a haemoglobin between 8 and 11 g and only 4% had a haemoglobin below 8 g/dl. An MCV of 65 -75fl was found in 56% of the children with a haemoglobin of 8 – 11g/dl. 84% of the children were under 6 years old with half of them under 2 years of age.
Guideline standards for iron supplements were met in only 22%, a dietican was involved in only 7% and follow up for both repeat blood tests and clinical review was arranged in a fifth of the eligible children.
All children with a haemoglobin less then 8g/dl were appropriately managed. There was no significant difference in all these parameters between the two different Hospital Trusts.

CONCLUSION

Incidental iron deficiency in hospitalised children is not uncommon but remains unrecognised and the opportunity to treat it appropriately is often missed. Greater awareness of this may lead to less missed opportunities for treatment.

MRI-the best evaluation tool for Paediatric Crohn's?

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Background

The current gold standard for assessing children with Crohn's Disease (CD) is a careful history and examination followed by baseline blood tests, upper endoscopy, ileocolonoscopy and Barium Follow through. Significant doses of radiation, unpalatability and volume intolerance are involved with Barium studies and repeated studies for re evaluating disease are thus not ideal. Adult practice in Gastroenterology, in particular with CD has changed with increasing use of Magnetic Resonance Imaging (MRI)

Aims

A prospective pilot study for service evaluation for radiological assessment of Paediatric Crohn's comparing Barium Follow throughs (BaFT) and abdominal MRI following endoscopic investigation

Methods

All consecutive patients with newly diagnosed IBD or previous diagnosis requiring reassessment over a seven week period aug-oct 2008 were requested to have both MRI abdomen and BaFT. Ethics was not required for this service evaluation but verbal consent was obtained in all cases. Two GI radiologists-adult and paediatric examined all films and compared findings on extent of disease and reported any imaging problems. Additional information regarding endoscopy findings were discussed at the regular departmental radiology meetings

Results

12 children fit criteria over the specified time period.
Age 9-18(15)years range(median).
8/12 male
4/12 newly diagnosed IBD.
2 patients with known CD refused Barium.
One newly diagnosed UC (ulcerative colitis) age 9 was reluctant for more tests so was withdrawn from study
MRI and Barium films were compared in 9 children, 3 new IBD, 7 CD.
MRI and Barium correlated well for Ileal disease particularly terminal Ileum.
Barium significantly underestimated extent of jejunal disease compared to MRI
MRI demonstrated colonic disease well and correlated with colonoscopy
Movement artefact with MRI was an issue with poor quality images in some cases
Patient feedback to date showed preference for MRI for tolerance and palatability of solution BaFT- 750-1000 mls flavoured contrast, MRI -100 mls lemon barley lactulose and water as tolerated

Conclusion

Pilot data suggests MRI is superior to BaFT for assessing IBD in children.
Artefactual problems with MRI may be improved with respiratory gating and requires further evaluation

Painful central venous catheter

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Introduction: Central venous catheters (CVC) are used for patients who require long-term chemotherapy or parenteral nutrition (PN). Commonly recognised mechanical complications include misplacement, occlusion, migration and fracture. We report two cases of children with intestinal failure and indwelling CVC who complained of severe pain under the arm or in the neck on flushing the catheter. Radiological investigation revealed fluid leakage into the skin tunnel in one, and extravasation due to backflow between the fibrin sheath and CVC in the other.

Case 1: An eight year old boy with chronic intestinal pseudo-obstruction needed eight CVC over six years for PN. Difficulties with venous access meant that the ninth CVC was inserted via the left axillary vein; three months after its placement he complained intermittently of sharp pain under his arm each time the catheter was flushed. Blood could be aspirated without difficulty and PN infused well; examination revealed no subcutaneous swelling. This symptom had initially developed after playing on a 'fireman's pole'. Initial chest X-ray (CXR) following through CVC injection of contrast showed no abnormality, but when symptoms persisted, a second CXR showed that contrast leaked into the subcutaneous tunnel. The CVC was removed and a fracture was found just distal to the cuff, where the external diameter of the CVC tapered. It is likely that the CVC was weakened by chronic repetitive kinking at this point, finally being ruptured during vigorous play.

Case 2: A six year old girl with short bowel syndrome secondary to gastroschisis had multiple replacements of CVC used for PN. Her 13th Broviac CVC was inserted into the left subclavian vein with the line tip in the right atrium. Two weeks later it was difficult to withdraw blood but flushed well; three weeks later she complained of neck pain each time the CVC was flushed. Chest X-ray following injection of the CVC showed contrast passing around the catheter and flowing retrogradely, with some extravasation at the entry point in the subclavian vein. We speculate that fluid flowed between the fibrin sheath around the CVC and the catheter surface, exiting where fibrin sheath joined the vein wall.

Conclusion: Pain on flushing a CVC indicates a mechanical problem with extravasation or leakage of fluid. Different mechanisms may be involved, but CVC replacement is required. Initial CXR should involve contrast injection and visualisation of the entire CVC from the skin exit point to the catheter tip.

Parental perception of Percutaneous Endoscopic Gastrostomy in children with Congenital Heart Disease

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Introduction: Over a 5 year period between 2002- 2006, we inserted 384 Percutaneous Endoscopic Gastrostomies (PEG's) in our centre. Neurodisability and Congenital Heart Disease (CHD) have been the main indications. While the usefulness of PEG is clearly established in the nutritional support of children with neurodisability, its role in substituting prolonged nasogastric feeding in children with CHD is only now being recognised. We aimed to study parental perceptions of PEG in children with CHD. There are no previous published experiences about the same.

Methods: Descriptive qualitative survey of parental perceptions using a semi structured questionnaire.

Results: 38 questionnaires were returned to the department over the study duration. Parental perceptions on the effects PEG insertion on preoperative problems and quality of life issues are depicted in tables 1 & 2 respectively.

Table 1

Pre operative problems	(n)	No of subjects who improved	No of subjects who worsened	No of subjects with no change	'p' values Sign (binomial) test – one tailed
Difficulty feeding (non specific)	12	11	1	0	0.003
Coughing, chocking	21	20	1	0	<0.0001
Vomiting	29	26	0	3	<0.0001
Fatigue while feeding	15	13	1	1	0.003

Table 2

Criterion	(n)	No of subjects who improved	No of subjects who worsened	No of subjects with no change	'p' values Sign (binomial) test – one tailed
Child's happiness & wellbeing	38	34	0	4	<0.0001
Ease of giving medications	22	17	1	4	0.0085
Time to devote to other children	31	19	1	11	0.140
Time to devote to self	38	14	10	14	0.07

Fifteen of the 38 (39%) parents wished the PEG had been done earlier, while 22 (57 %) felt that the PEG was done at the appropriate time. One (2.8%) parent felt that the PEG shouldn't have been done.

Conclusions: Majority of parents of children with CHD perceive an improvement in their child's symptoms, wellbeing and ease with administering medications after PEG insertion. Greater time to devote to their other children and themselves were additional benefits from PEG insertion though not reaching statistical significance. 97 % of parents felt that having a PEG was the right decision for their child's nutritional support.

The spectrum of presentation of coeliac disease

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Background:
Widespread use of serological testing has facilitated the earlier diagnosis of coeliac disease and altered the demographics of the disease presentation within the paediatric age range.

Aim:
To review the age and mode of presentation of paediatric patients diagnosed between 2003 and 2007 with coeliac disease in the Teeside area, UK.

Method:
Retrospective case note review of all biopsy proven coeliac disease in North Tees, Hartlepool, and Middlesbrough Hospitals between 2003 and 2007. Data collected included age, haemoglobin concentration and predominant clinical features at presentation.

Results:
36 children (33 % male) were identified. Median age was 4.2 years (range 0.8-15.0).
25/36 presented with classical symptoms; 9/36 (25%) with diarrhoea, 8/36 (22%) with weight loss/failure to thrive, 6/36 (17%) with abdominal pain, 2/36 (6%) with refractory anaemia. 7/36 (19%) were diagnosed following screening for coeliac disease. Screening was performed in cases of family history and mild gastrointestinal symptoms in 4, type II diabetes mellitus in 2 and Down's syndrome in 1. Four patients (11%) had non-classical symptoms including poor appetite, hair loss, food refusal, chronic oral ulcers, and bloating and muscle weakness.

Presentation	Mean haemoglobin at presentation (g/dl)
Classical symptoms	10.8
Non-classical symptoms	11.5
Screening	12.7

Conclusion:
The above data can present a rough guide to recent demographics for patients presenting with coeliac disease. A considerable proportion can present with uncommon symptoms. Screening in high risk patients can be of significant value.

Type I Refractory Coeliac disease (RCD) in childhood.

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Introduction:

Refractory coeliac disease (RCD) is rare and the true prevalence of RCD is unknown (assumed to be 0-5% in adults). RCD is defined as persistence of small intestinal biopsy changes despite strict adherence to gluten free diet (GFD) for at least 6 months1. RCD can be divided into type I (with out aberrant T cells), which responds to nutritional therapy/ immunosuppressive therapy and type II (with aberrant T cells)2 which carries a poor prognosis, which is also difficult to treat. We report a child with type I refractory celiac disease, which was resistant to conventional treatment and needed immunosuppressive therapy to control the disease.

Case report:

A 4 year old girl was diagnosed to have celiac disease following positive anti tissue transglutaminase antibody (tTG) and crypt hyperplasic sub-total villous atrophy (VA) and increased intraepithelial lymphocytes (IEL) on intestinal biopsy. Her symptoms improved (diarrhoea, lethargy, poor weight gain, severe dry skin and irritability) and had negative tTG on gluten free diet. Approximately 3 months later she started to have diarrhoea again. She was tried on pauciantigenic diet with little effect and eventually elemental feeds with a degree of clinical response. She also had normal total IgE, RAST, ESR, CRP, auto antibody screening, immunoglobulin levels and colonoscopy. Her repeat anti tTG was negative but OGD showed villous blunting, increased IEL and crypt hyperplasia. Immunohistochemisrty showed mixture of CD4+, CD8+ lymphocytes in lamina propria but exclusively increased infiltration of CD8+ intra epithelial lymphocytes (45 lymphocytes/100 enterocytes). Subsequently she was started on anti inflammatory & immunosuppressant medications including azathioprine with good response. Currently, She is 7 years old and doing very well on the above therapy.

Conclusion:

Loss of response to GFD needs to be investigated and trial of Pauciantigenic diet, elemental feeds and immunosuppressive therapy should be considered in addition to gluten free diet in RCD. Further studies are needed to assess the prevalence of RCD among children.

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Use of sirolimus to treat refractory Paediatric Inflammatory Bowel Disease -Case report.

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Introduction:

Sirlomius (rapamycin) and Mycophenolate mofetil (MMF) have been used extensively as immunosuppressant after organ transplantation but this has not been used in children with Inflammatory Bowel Disease. A case report of remission induced by sirolimus in refractory adult Crohn's disease has been found on literature search1. We report a child with resistant inflammatory bowel disease (IBD) who responded well to this combination treatment.

Case report:

An 8 year old boy was diagnosed to have indeterminate colitis at the age of four following extensive investigations after having chronic diarrhoea for 2 years. He was initially treated with mesalazine, sulfasalazine, azathioprine and predinsolone. Unfortunately he developed allergy reaction (rash) to sulfasalzine and also developed pancreatitis with azathioprine. He was then started on infliximab infusions in view of recurrent symptoms (ESR 80mm/hr, Platelets 683 x109/L, Hb 11.4g/dl and biopsy showed severe chronic active pan colitis with mild gastritis but with out granuloma). His symptoms initially improved but diarrhoea worsened again needing further treatment. A year later, in view of recurrent symptoms he was started (weight 22kg) on adalimumab at 20mg (subcutaneous injection) every 2 weeks and also started on sirolimus 2 mg (orally) once a day and MMF 250mg (orally) twice a day at the same time. Sirolimus level was low (3.6 ng/ml) initially and the dose was increased to 3 mg subsequently (aimed to keep between 10-12 ng/ml). His symptoms improved and he became asymptomatic on this combination treatment. Adalimumab was stopped a year later and he is currently on sirolimus, MMF and mesalazine without any symptoms. He is gaining weight and inflammatory markers were all normal (Hb 12.2 g/dl, WCC 6.9 x109/L, Plats 343 x109/L, ESR 9 mm/hr, albumin 47 g/l). His recent endoscopy was 15 months after he was started on sirolimus and this was reported as showing very mild chronic inflammatory changes in colon.

Conclusion:

This is the first case report of Sirolimus induced remission in Paediatric Inflammatory Bowel Disease (IBD). Further research is needed to assess the role of sirolimus in managing children with refractory IBD.

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POSTERS
FRIDAY 30TH JANUARY 2009

A single centre Videofluoroscopy swallow study: A Neonatal cohort

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Background: Clinical assessment of feeding by subspecialist Speech and Language Therapists (SALT) is integral to the detection of swallow dysfunction in the paediatric and even more so the neonatal population. The results of the assessment ultimately determine the need and type of instrumental or objective assessments that may be required to further investigate the nature or cause of the dysphagia.

Videofluoroscopy swallow study is the 'gold standard' for the assessment of oropharyngeal and oesophageal stage function during swallowing because of its ability to capture the rapid and dynamic movements during swallowing. VFSS involves the use of ionising radiation. Information obtained from the VFSS must result in diagnostic clarity or impact on current management recommendations. With a strong emphasis on evidence based practice and clinical accountability clinicians need to examine the evidence for the use and efficacy of VFSS. Limited data is currently available on normative values for VFSS in the neonatal and pre-term population. In this retrospective study we analyse our data of patients on our neonatal unit who underwent VFSS.

Objectives: The aim of this study was to examine the outcome of VFSS as an assessment tool in the diagnosis of swallowing dysfunction in neonates and moreover to determine the ultimate impact of VFSS results on the management of oral feeding difficulties.

Methods: Data of VFSS from 17 neonates from a single tertiary (supra-regional surgical) neonatal unit referred to the feeding team over the duration from January 2007 to September 2008 was analysed. Median gestation of prematurity was 27 weeks (range: 24+4 - 41), 8 males and 9 females. All assessments were undertaken by the same SALT (experienced in paediatric and neonatal VFSS). VFSS were undertaken under institutional guidelines.

Results:A total of 22 VFSS were analysed. Four patients underwent multiple studies. All but three patients were found to be aspirating and therefore had their feeding regimens altered because of abnormal results. Four patients were diagnosed with gastro-oesophageal reflux (GOR) on the basis of the intraoesophageal reflux demonstrated on videofluroscopy swallow studies. In one of these subjects acid gastroesophageal reflux had been previously excluded by pH study. The most common outcomes were the discontinuation of oral feeds (6) and the thickening of milk feeds (12) respectively.

Conclusions: VFSS is an effective and very instructive diagnostic tool in the management of neonatal feeding difficulties. These findings suggest that VFSS, is a sensitive assessment tool not only in the detection of aspiration but also in the identification of the 'at risk' neonate with immature or abnormal co-ordination of the oropharyngeal stage of swallowing. Moreover VFSS can be a useful diagnostic modality in the detection of silent or indeed non-acid or alkaline GORD in neonates.

Alt167Val- a missense mutation in the AAAS gene- detected in a Greek-Cypriot family: A case series

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Background: Triple A syndrome- first described in 1978 by Allgrove et al- is a rare autosomal recessive disorder characterised by the classical symptom triad of achalasia, alacrima and adrenal insufficiency. The responsible triple A syndrome gene (AAAS) has been mapped to chromosome 12q13 and its gene product is a protein called ALADIN (alacrima, achalasia, adrenal insufficiency neurologic disorder).

Case series: We describe three children of a non- consanguineous Greek- Cypriot family, who were referred to our paediatric gastroenterology unit for investigation of achalasia.

Patient A, a 16 year old girl, initially presented at the age of 3 yrs with failure to thrive and dysphagia. She was ultimately diagnosed at the age of 6 yrs with achalasia. Oesophageal manometry (OM) undertaken in Greece at time of diagnosis revealed a hypertensive non- relaxing lower oesophageal sphincter (LOS) with notable preservation of oesophageal body function. She subsequently underwent an open Heller's cardio-myotomy a year later. She was referred to our department because of a recent relapse of symptoms with progressive dysphagia unresponsive to serial pneumatic oesophageal dilatations. A further detailed history disclosed alacrima since birth. She also described symptoms of a dry mouth and dizzy spells. She was known to have mild learning difficulties. Two paternal uncles have alacrima without any characteristic symptoms of achalasia and two siblings are asymptomatic.
Patient B, 10 years and brother of Patient A, presented with failure to thrive and evolving dysphagia (not previously investigated). He also had alacrima since birth and mild learning difficulties.
Patient C, a 10 yr old girl, the cousin (paternal side) of patient A and B was diagnosed at the age of 6 yrs with achalasia. OM performed in Greece at that time was also pathognomonic of achalasia. Unlike patient A, she also had significant dysfunction of the oesophageal body at diagnosis. She underwent a laparoscopic Heller's procedure, however because of relapse of symptoms within a year of surgery subsequently required serial oesophageal dilatation. Detailed history again revealed alacrima since birth. She also complained of dizziness and reduced energy levels.

All three patients underwent investigation including upper GI contrast studies, upper GI endoscopy and OM at our unit. Diagnostic OM performed in Patient B confirmed a non- relaxing hypertensive LOS. As with patient C at diagnosis (compared to patient A) he was also noted to have low amplitude isovolumetric contractions of the oesophageal body. Interestingly of the three patients only patient B and C had typical phenotypic facial features including a thin face, long philtrum and thin upper lip. The oesophageal dysfunction in both of these patients was more severe compared to patient A, who lacked phenotypic facial features. None of the patients showed signs of adrenal insufficiency. Sequencing of the AAAS gene confirmed the diagnosis of triple A syndrome. All three patients had a homozygous missense mutation in exon 6 at nucleotide position 500 (C>T), resulting in a change of alanine into valine at amino acid position 167 (Ala167Val).

Summary/ Conclusion: Triple A syndrome is a complex multisystemic autosomal recessive disorder. It is very rare and the presentation is variable. Patients often present with dysphagia and are subsequently diagnosed with achalasia. At the time of presentation most patients have previously unrecognized alacrima. Adrenal insufficiency does not have to be present when the diagnosis is made but usually develops during the first two decades of life.
The triple A syndrome is caused by mutations in the AAAS gene. The gene product- a protein known as ALADIN- localises near to the nuclear pore complex (NPC). Even though the exact function remains unclear it is supposed to play an important role in RNA and/ or protein trafficking between the nucleus and cytoplasm. Ala167Val is only one of a wide variety of AAAS mutations and an exact phenotype has not yet been described. A genotype/ phenotype meta- analysis of over 140 cases including our family is currently taking place.

Assessment of modified strobel formula in pH catheter placement: A Prospective Study

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Introduction
Gastro-oesophageal reflux is a common disorder seen in infancy and childhood. Early diagnosis and intervention is essential for prevention of complications and management. Oesophageal pH monitoring is the gold standard technique for detection of acid gastro-oesophageal reflux episodes and correct placement of catheter is crucial. The Strobel formula (0.252 X Height+5) is frequently used as a guide to determine distance from the nostrils to the lower oesophageal sphincter in term infants. Our experience showed that the pH catheter placement was overestimated using Strobel formula. A study using modified Strobel formula was carried out to assess accuracy.
Modified strobel formula was calculated as follows-
1. Infants<12 months=Height X 0.252+2 cms
2. Older children >12 months= (Height X 0.226 + 4.6)X 0.87 cms

Aim:
To compare accuracy of Strobel formula and modified Strobel formula measurements in the correct placement of catheter.

Subjects and Methods:
Prospective data of 15 patients was collected between November 2006 and July 2008. The total number of investigations studied was 16 as 1 patient had two pH studies done (aged 44 days and 273 days). The double sensor and single sensor probe was used in 13 and 3 studies respectively. The double sensor probe was changed to single sensor during the course of the study as the manufacturing company had changed double sensor probe production to single sensor probe. Length of catheter was calculated using Strobel and modified Strobel formula. Catheter placement was done using modified Strobel formula in 15 patients and Strobel formula in 1 patient. The standard was radiological check of catheter tip position between T8-T10. The probe position was checked by X ray in each case by an experienced radiologist. Comparison was made between the 2 calculated lengths for each patient and the most accurate length was identified. The probe was adjusted if the tip was outside T8-T10 to T9.

Results:
The 15 patients included in this study ranged in age from 3.5 weeks to 64 weeks. Male n=7, Female n=8. Mean age 19.3 weeks and median age 13.4 weeks. Two subjects (32+2 weeks) had the study at corrected gestational age of 35+6 weeks. 10/16 (62.5%) catheter placements had X ray confirmation between T8-T10. 7 catheters needed no repositioning and 3 catheters although placed between T8-T10 were slightly readjusted. 6 catheters (37.5%) needed repositioning. Modified Strobel formula (12/16) was identified as more accurate (75% of cases) than Strobel formula (4/16) in obtaining correct placement of catheter.

Summary and Conclusion:
1) This study showed that the modified Strobel formula was more accurate than the Strobel formula in the probe placement.
2) The data is inadequate to show accuracy in children aged more than 12 months.
3) Radiographic confirmation of catheter position must be considered when using any formula.

Coeliac Disease Occurs In Asian Children Living In Wales

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Introduction:
Coeliac disease (CD) is often thought of as a disease of Mediterranean and Irish aetiology and not occurring in the Asian community. However previous studies have highlighted areas in Northern India where coeliac disease is prevalent. In the U.K. Coeliac disease affects around 1% of the population and is frequently underdiagnosed throughout all population groups.

Aim:
To determine the detection of coeliac disease amongst the Asian childhood population residing in Mid and South East Wales.

Methods:
All cases of children diagnosed having coeliac disease between 1990 to 2006 were assessed. Cases from the whole of Mid and Southeast Wales were identified from histological and serological records and clinic letters. All cases are endoscopically diagnosed at the Children's Hospital for Wales. The Registrar General's Census data was used to estimate the childhood population**.**

Results:
A total of 99 children were diagnosed as having coeliac disease, 94 Caucasian and 5 Asian. According to 2001 Census data 358,846 children aged <17 years resided within Mid and Southeast Wales (Caucasian 342,579 Asian children 6664).

	Prevalence of diagnosed coeliac disease
All children	0.03%
Caucasian children	0.03%
Asian children	0.08%

Chi Squared analysis with Yates correction show a borderline statistically significant increased frequency of coeliac disease in the Asian population (p=0.055).

Conclusions:
Most cases of coeliac disease still remain undiagnosed. We wish to draw attention to the existence of coeliac disease in the U.K. Asian population.

Curcumin suppresses p38 MAPK activation, reduces IL-1βand MMP-3, and enhances IL-10 expression in inflammatory bowel disease

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Introduction/Background
In inflammatory bowel disease (IBD), both chronic pro-inflammatory pathways and a failure of anti-inflammatory (healing) mechanisms sustain disease. p38 mitogen-activated protein kinase (MAPK) is central to the coordination of inflammatory responses and is consistently raised in IBD, suggesting a critical role in pathogenesis and presenting a potential target for therapy. In IBD there is excess production of pro-inflammatory cytokines including interleukin (IL)-1βand under-expression of the major anti-inflammatory gut cytokine IL-10. Fibroblasts over-produce matrix metalloproteinases (MMP) in IBD, mediating tissue destruction. Curcumin, a component of the spice turmeric, an anti-inflammatory agent and a known inhibitor of acetylation, shows clinical benefit in IBD.

Aim
To assess the effect of curcumin on p38 MAPK activation and on the downstream expression of cytokines and MMP-3 in the gut of children, adolescents and adults with IBD.

Subjects and methods
Fresh intestinal mucosal biopsies from children with active IBD and colonic myofibroblasts (CMF) from children, adolescents and adults with active IBD were cultured ex vivo in the presence or absence of curcumin. p38 MAPK and NF-βB activation were measured in biopsy and CMF lysates and MMP-3 in CMF supernatants by immunoblotting. IL-1βand IL-10 were measured in biopsy supernatants by enzyme-linked immunosorbent assay (ELISA). Some experiments were repeated with anacardic acid, a component of cashew nut shell liquid and one of the only other known naturally occurring inhibitors of acetylation.

Results
We show a marked reduction in p38 MAPK activation in curcumin-treated mucosal biopsies and favourable modulation of the cytokine profile with enhanced IL-10 expression and reduced IL-1β We also demonstrate dose-dependent suppression of MMP-3 expression in CMF with curcumin, by a mechanism which appears to be p38-independent and may relate to acetylation.

Summary and Conclusion
Curcumin, a naturally occurring food substance with no known human toxicity even at very high dose, holds promise as a potential new therapy in IBD.

Focal nodular hyperplasia in two children with McCune-Albright syndrome

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Introduction

McCune-Albright syndrome (MAS) is a sporadic and phenotypically variable disease defined by the clinical triad of café-au-lait pigmented skin lesions, polyostotic fibrous dysplasia (FD) and precocious puberty. Beyond these classical symptoms other hyperfunctional endocrinopathies as well as non-endocrine manifestations have been previously reported. We describe two children with MAS and focal nodular hyperplasia (FNH) detected during follow-up for neonatal cholestasis and liver dysfunction.

Methods

The necessary data were collected by reviewing the patient's medical notes.

Results

Case 1. A male term newborn was referred at the age of three weeks because of conjugated hyperbilirubinemia, pale stools and failure to thrive. An extensive workup was negative and he was subsequently followed up for some residual transaminitis. At the age of 7 years he presented with pain and weakness of his right leg and radiological investigations confirmed fibrous dysplasia. Clinical examination revealed café-au-lait spots of his back and investigations for precocious puberty were consistent with a diagnosis of MAS. The left liver lobe appeared enlarged and subsequent imaging (abdominal ultrasound, MR scan) was in accordance with FNH. The management is conservative up to now.

Case 2. A female term newborn was referred at the age of two weeks because of conjugated hyperbilirubinemia, pale stools and failure to thrive. Liver histology confirmed neonatal hepatitis. An ERCP showed biliary hypoplasia. The further workup was negative. At the age of 3 months there were features consistent with MAS (café-au-lait spots, clitoral and breast enlargement). Adrenalectomy was performed at the age of 9 months and hormonal substitution was instituted as well as treatment with aromatase inhibitors. Her course was complicated by repeated fractures due to fibrous dysplasia. At 7 years of age clinical examination revealed a slightly enlarged liver with a liver mass in the left liver lobe on abdominal ultrasound. An abdominal MR scan confirmed the diagnosis of FNH. The management is conservative to this day.

Summary

There is no previous report of FNH in children with McCune-Albright syndrome: in our patient cohort 2 out of 2 patients with MAS developed focal nodular hyperplasia during their follow-up for neonatal cholestasis and ongoing transaminitis.

Conclusion

FNH is an important non-endocrine manifestation of MAS in childhood and should be considered when presenting with a liver mass. The underlying pathophysiology of FNH in this patient cohort remains unclear.

Growth and pubertal status following pubertal induction in boys with inflammatory bowel disease

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Background: Poor linear growth associated with delayed puberty is common in children with inflammatory bowel disease(IBD). Anecdotal reports suggest that pubertal induction with sex steroid is effective in promoting linear growth but there is currently no published study available in this group of children.

Objective: To assess pubertal status and growth in a group of boys with IBD 6 months before and after testosterone therapy.

Methods: Retrospective study of 8 boys with IBD who underwent pubertal induction with testosterone. Height(Ht), weight(wt) and pubertal status were obtained before and after testosterone. Markers of disease activity and data on concomitant medication were also collected. Response to testosterone was based on advance of pubertal status and a greater than 50% increase in height velocity (HV). Results were expressed as median(range).

Results: Eight boys with IBD(7 with crohn's disease and 1 with indeterminate colitis),median age of 14.8 yrs(range,13.6,15.6), median TS of 1(1,2) and a median bone age delay of 2.9yrs(0.8, 3.5) had pubertal induction using either sustanon 50mg IM. monthly or andropatch 2.5mg or 5mg daily applied for 12hrs each day. Seven boys showed an advance of pubertal status to a median TS of 2/3(2,4); six boys showed progression of testicular volumes after testosterone therapy, suggesting activation of the gonadotrophin axis. Six boys had a greater than 50% increase in HV following testosterone. Median HV at T+0 and T+6 was 1.6 and 6.9 cm/year, respectively (p=0.005). Median HtSDS at T-6, T+0 and T+6 was -1.4(-3.0,-0.8), -1.6(-3.4, -1.1) and -1.4(-3.3, -1.1), respectively. Median height velocity(HV) SDS at T+0 and T+6 was -4.2(-7.6, +2) and +1.2 (-2.7, +8.5) respectively. HVSDS was significantly different following treatment when corrected for both age and pubertal stage (p=0.005 and p=0.01). Median albumin, CRP, ESR and platelets were similar at T+0 and T+6. Median CRP showed a significant negative association with actual HV (cm/yr) at T+6 (r= -0.8; p=0.02).

Conclusion: Our study showed for the first time that testosterone therapy improves linear growth and pubertal status in children with inflammatory bowel disease and delayed puberty. The improvement in linear growth following testosterone is variable and may be related to the degree of inflammation. The effects of pubertal induction in boys with IBD on linear growth, puberty, bone health and body composition require further systematic study.

Growth retardation in inflammatory bowel disease is not universally associated with growth Hormone resistance

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Background and aim: It is often proposed that growth retardation in IBD is associated with a state of growth hormone resistance but published data on the growth hormone (GH)-insulin like growth factor1 (IGF1) axis are scarce.We describe the results of routine clinical assessment of this axis in a group of such children.

Methods:A retrospective study of 28 children(23M)with IBD(22 CD, 3 UC)with growth retardation and/or pubertal delay who underwent insulin tolerance test(ITT)following referral to the endocrine clinic. Height velocity(HV)and IGF1 were converted to SDS;to account for delayed puberty for girls>11yrs and boys>12yrs,HV and IGF1 were adjusted for bone age. Results expressed as median(range)

Results: Eight children(29%)were on oral Prednisolone at point of ITT. Median age at diagnosis and at time of ITT was 10.9yrs(6.0,13.9) and 14.3yrs(7.7,17.0)respectively. Median HtSDS at diagnosis and at time of ITT was -1.4(-3.4,0.4) and -2.0(-3.6,-0.9)respectively. Median HVSDS over 12 months was -2.2(-7.7,2.8). Median midparental HtSDS was -0.5(-1.6,0.7). Median BMISDS was -0.6(-2.2,2.3).Fourteen out of 28(50%) were prepubertal at the point of ITT; seven out of the 20 children > 12 yrs (35%)were prepubertal. Median peak GH was 19.3mU/L(4.2,80.0) and median IGFSDs was -0.9(-3.4,0.1). Five out of 28 (18%)had exaggerated GH response to ITT (peak GH>40mU/L). Only four out of 28 (14%)had IGF1SDS<-2.0.

	Peak GH< 10 mU/L	Peak GH ≥ 10,<20 mU/L	Peak GH ≥ 20mU/L
IGF1SDS< 0	4/28	11/28	11/28
IGF1SDS≥ 0	0/28	0/24	2/28

In this cohort, 11 children(39%)were GH resistant(peak GH≥20mU/L,IGF1SDS<0), whilst 4(14%) were GH insufficient(peak GH<10mU/L,IGF1SDS<0).Two children, in fact, had combined GH-IGF1 resistant(peak GH≥20mU/L,IGF1SDS≥0). Only two children had peak GH ≥20mU/L and IGF1SDS<-2.0(severe GH resistant).Peak GH showed no association with IGF1SDS(r=0.1,p=0.7). HVSDS showed no association with peak GH(r=-0.3,p=0.4)and IGF1SDS(r=0.3,p=0.1).
Conclusion: Our study,which is the largest cohort of children with IBD who had evaluation of their GH axis,demonstrate that poor growth is associated with a range of abnormalities in the GH-IGF1 axis. Biochemical evidence of systemic GH resistance exists but is not universal; severe GH resistant is in fact not common in IBD.

Investigating the potential of mesenchymal stem cells for cellular therapy of liver disease

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Background: Cellular therapy in the form of hepatocyte transplantation is an alternative to whole organ transplantation for certain liver diseases and may serve as a 'bridge' in acute liver failure. As the supply of good quality hepatocytes is limited, a sustainable alternative source of cell is needed. Mesenchymal stem cell (MSC) transplantation has been shown to improve survival in animal models of liver disease and in small clinical studies of liver failure. It is not known whether this therapeutic effect is due to the potential of MSC to differentiate into hepatocytes in the appropriate microenvironment or to their immunomodulatory effects. The primary aim of this project is to determine the effect of MSC on human hepatocyte survival and function in vitro and to investigate the mechanism of the effect.

Methods: Human hepatocytes were isolated from livers donated but unsuitable for transplantation and from partial hepatectomy specimens using a collagenase perfusion technique. Hepatocytes were cultured under standard conditions on collagen coated plates. A human MSC line derived from adipose tissue was used for experiments. Co-culture was undertaken both with direct cell-cell contact (ratio from 1:1 to 10:1 MSC: hepatocytes) and indirectly through a semi-permeable membrane. Hepatocytes were also cultured in 50% and 100% MSC-conditioned medium. Survival of hepatocytes in different experimental conditions was analysed with trypan blue, annexin V / propidium iodide staining using flow cytometry and detection of caspase-3 using real time PCR. Albumin production was measured with ELISA and urea by a specific colormetric assay. PCR was used to detect the expression of liver specific genes in MSC following co-culture (to identify trans-differentiation).

Results: There was a 20% improvement in the viability of hepatocytes which were co-cultured with MSC through a semi-permeable membrane compared to control hepatocyte cultures. Hepatocyte albumin production was significantly improved in direct co-culture with MSC at a ratio of 2:1 on day 2 ($p<0.05$) and at a ratio of 10:1 on day 4 ($p<0.01$). This effect was not seen in cells which were indirectly co-cultured or cultured in MSC-conditioned medium. Urea production in co-culture was no different to monoculture. Partial trans-differentiation of MSC into hepatocytes was suspected in indirect co-culture as PCR revealed albumin expression in these cells in addition to MSC markers, however albumin production by MSC in indirect co-culture was not detected by ELISA.

Conclusion: Though an improvement in viability was seen in hepatocytes co-cultured with MSC through a semi-permeable membrane, direct contact was needed between hepatocytes and MSC to demonstrate an improvement in albumin production. It is not clear whether the pro-survival, anti-apoptotic, pro-function effect of MSC on hepatocytes is due to partial trans-differentiation of MSC into hepatocytes or to another mechanism involving soluble mediators.
The ability of MSC to improve the function and viability of hepatocytes may allow clinical translation to the cellular therapy of liver disease.

Paediatric Inflammatory Bowel Disease Associated Hepatobiliary Disease

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Introduction: The link between inflammatory bowel disease (IBD) and associated hepato-biliary involvement is very well-documented in the adult population. Several papers in the literature quote the incidence of patients with ulcerative colitis (UC) developing primary sclerosing cholangitis (PSC) between 3-5%. Malignancy rates are much higher in this subgroup of IBD patients. Other figures quote incidence for patients with PSC also having ulcerative colitis, between 70-90%. There are similar figures for autoimmune hepatitis (AIH), although not as well-documented, as both these entities are rare in their own right. What is currently lacking in the literature is whether this holds true for the paediatric population with IBD, or whether there are major differences.

Aim: We chose to analyse a cohort of paediatric patients with IBD to produce an incidence for PSC/AIH in children with IBD, and to identify any differences in demographics, investigations, treatment modalities and outcomes compared to the adult literature.

Methods: All patients with PSC, AIH and overlap syndrome were identified from The Royal London Hospital Paediatric IBD database and analysed in terms of gender, age at diagnosis, type of co-existing IBD, time of onset of abnormal liver function tests (LFTs), investigations performed, treatment modalities and outcomes.

Results: At time of analysis, there were 360 patients on the IBD database based at The Royal London Hospital (227 CD, 84 UC, 50 IC). Of these, 11 had PSC, AIH or overlap syndrome (3.3%). 9/11 were male. 6 patients had PSC, 4 AIH and 1 with overlap syndrome. 3 patients (27%) had liver disease diagnosed aged < 12 years. IBD diagnosis was UC in 10/11 and 1 with indeterminate colitis (IC). 5/11 patients had abnormal LFTs at the time of diagnosis of their IBD, 5/11 developed abnormal LFTs within 1 year and the youngest patient had 4 yrs before developing abnormal LFTs. All patients had an ultrasound, biopsy and cholangiogram as part of their liver work up. All 4 autoimmune patients responded to standard immunosuppression. 4 PSC patients are maintained in remission with ursodeoxycholic acid alone, one had aggressive liver disease and had a successful liver transplant within 2 yrs of diagnosis. The other has significant relapse 5 years post diagnosis and is awaiting transplant evaluation.

Summary: In conclusion, we have confirmed similar figures of Paediatric IBD associated Liver Disease incidence compared to the adult population. The findings highlight the importance of investigating patients with IBD with abnormal LFTs, even if asymptomatic.
The risk of malignancy in those diagnosed young is essentially unknown and close long term surveillance is essential in this subgroup for optimal outcome

PEG to Button: do you need access to upper GI endoscopy?

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Background and aim: Percutaneous endoscopic gastrostomy (PEG) tubes provide a secure and relatively simple means of providing nutrition support to children for the medium to long term. However, for practical and cosmetic reasons many children and their families prefer to have the PEG converted to a low profile gastrostomy device (button). Performing this blind without endoscopy could save resources. We aimed to review the need for access to upper GI endoscopy at the time of conversion of PEG tube to low profile device.

Methods: Retrospective audit by database and clinical note review of endoscopic use at time of conversion of PEG tube to low profile device in our regional paediatric centre for the 5 year period 01/07/03-30/06/08; the study population was children under 18 years of age living within SE Scotland. Children had their PEG tubes removed by traction and converted to low profile devices under general anaesthetic or deep sedation. Position of the low profile device was confirmed with gastric aspirate on pH paper (pH<5.5). If no aspirate or a non-acid aspirate was obtained, endoscopy was used to confirm position.

Results: 99 children had PEG tubes inserted over the review period; 6 of these were placed using laparoscopic assistance. 70 of the children had their PEG tubes converted to low profile devices, although in 3 this was electively performed at time of fundoplication. The timing of change was determined by family choice and theatre list availability with a median (range) of 224 (56-700) days. 25 of 67 (37%) of the children did not require endoscopic confirmation of position. Of the remaining 42 children, 20 (48%) had a planned endoscopy to re-evaluate their upper GI tract. 22 of 42 (52%) children had an unplanned endoscopy, and in 15 of these 22 (68%) the balloon of the low profile device was confirmed in the stomach. However in 4 of the 22 (18%) children the balloon was found not to be in the stomach and required manipulation under endoscopic guidance; 2 of these children were subsequently found to have a gastrocolic fistula, a rare but recognised complication of PEG tube placement. In 1 of 22 (4%) the internal retention disc separate from the external tube requiring it to be endoscopically retrieved. Following removal of 2 of the 22 (8%) PEG tubes the tract required to be dilated under endoscopic vision in order to insert a low profile device.

Conclusions: There is a significant risk of malplacement at the time of conversion of PEG tube to a low profile gastrostomy device. Access to upper GI endoscopy should be the standard of practice with 52% of children, in this regional 5year cohort study, requiring an unplanned upper GI endoscopy.

Profound anaemia with marked developmental delay in an infant secondary to Cobalamin (B12) deficiency; pitfalls in the diagnosis

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Background: Cobalamin is an essential vitamin in the maintenance of normal metabolism. Deficiency is uncommon in infants and is usually secondary to maternal B12 deficiency. Delay in the diagnosis or treatment may predispose infants to faltering growth, haematological and irreversible neurodevelopmental problems.

Aim: To describe an unusual presentation of B12 deficiency in an infant who was exclusively breast fed by a mother who was on a mixed diet and had negative intrinsic factor antibodies.

Methods/ result: A Caucasian male infant was born at 34 weeks gestation to a gravida 4 female (3 previous healthy males). He was fully breast fed and his mother had a normal mixed diet. He presented at six months of age with frequent upper respiratory tract infections, lethargy and poor feeding. He was noted to be anaemic (Hb 7.7 gm/dl), MCV (92 fl) and neutropenic (0.5*10^9). Other investigations included a normal B12 level (508 pg/ml (199-1222) and folate level (17.6 ug/l(3-20)). This presentation with anaemia was thought to be secondary to a viral infection. He was followed up and by 8 months of age became severely anaemic (Hb4.4), and neutropenic (0.5), with failure to thrive and marked developmental delay. He was referred to a tertiary haematology centre for further investigations which included repeat B12 levels. Bone marrow study showed all stages of myeloid maturation, down to mature neutrophils but was grossly megaloblastic in the red cell series. His repeat B12 level (4 weeks after the initial result) was well below the reference range for his age 62pg/ ml. Supporting this there was a large amount of methylmalonic acid and excess of homocysteine in his urine, compatible with severe vitamin B12 deficiency. He made a good recovery following treatment with intramuscular vitamin B12 and fortified milk and food products. His mother's B12 level was low at 115pg/ ml and her intrinsic factor antibodies were negative. There is no family history of anaemia or autoimmune disease and mother has remained well with no evidence of on going B12 deficiency after her initial treatment. Her practioners have not investigated her further.
The patients' subsequent B12 levels and urine metabolites were normal which excludes an inborn error of B12 absorption or metabolism. His development (mainly motor skills) remained markedly delayed for the first 3 – 4 years of age; he is now six years old and is in mainstream school with a statement of special educational needs.

Discussion/ Summary:
Many infants have a nadir of their of Hb levels at 4-6 months of age and it was assumed that this infant's Hb level had been further suppressed by an intercurrent viraemia, particularly as the initial MCV and B12 level were normal. The continued decline in Hb levels at 8 months of age prompted further investigations, confirming profound B12 deficiency. We concluded that the mother's mild B12 deficiency contributed to the anaemia and B12 deficiency in this infant.

Conclusion:
In addition to the known cases of cobalamin deficiency in exclusively breast fed infants of vegetarian mothers or those with pernicious anaemia our case illustrates the possibility of B12 deficiency in other infants. Delay in diagnosis can lead to severe neurological damage. B12 deficiency generally manifests between the ages of 4-8 months in infants who are at risk. The presenting signs and symptoms can be non specific, as can the blood picture in the early stages. Repeat cobalamin and metabolite levels may be indicated (as noted in our case).
Serum methylmalonic acid and total homocysteine levels are reliable indicators of B12 deficiency and should be checked in all doubtful cases to confirm the diagnosis.

Successful isolated liver transplantation in a child with atypical haemolytic uraemic syndrome due to a mutation in complement factor H

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Introduction
Mutations in complement factor H (CFH) are an important cause of recurrent atypical haemolytic uraemic syndrome (aHUS) in childhood. It often progresses to endstage renal disease (ESRD) despite regular plasma exchange therapy. Recurrence of disease following renal transplantation is usual. Combined liver-kidney transplantation has been successfully performed in patients with ESRD. We report a child with factor H deficiency who underwent successful isolated liver transplantation prior to the onset of ESRD.

Method
The necessary data were collected by reviewing the patient's medical notes.

Case report
A male child was diagnosed with aHUS due to a heterozygous mutation in CFH (R1215Q) at the age of 5months. He was managed by regular plasma exchange therapy decreasing from 4 times a week to twice weekly. His course was complicated by episodes of relapse triggered by sepsis. His renal function was impaired but stable and he did not require haemodialysis. He underwent cadaveric isolated liver transplantation aged 4 years. The transplantation protocol incorporated a 1.5 volume plasma exchange immediately prior to transplantation and 10ml/kg plasma infusion during the anhepatic phase. The transplant period itself was complicated by exacerbation of hypertension and complications related to preexisting vascular access difficulty. Plasma exchange was discontinued immediately post transplant and there have been no episodes of aHUS recurrence. His renal function is similar to pretransplant values after 4 months follow up.

Summary
Successful isolated liver transplantation has prevented relapse of aHUS without plasma exchange in the short term. Pretransplant plasma exchange avoided severe complications due to complement activation.

Conclusion
Isolated liver transplantation should be considered earlier in patients with aHUS due to mutations in CFH who are dependent on plasma exchange or who are at risk of developing ESRD.

The Increasing Diagnosis Of Coeliac Disease Since The Introduction Of Anti-TTG Testing

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Introduction: Coeliac disease (CD) is a lifelong intolerance to gluten that affects around 1% of the UK population. It remains vastly under diagnosed with only around 10% of all cases being diagnosed. Research carried out at the University Hospital of Wales between 1983 and 1998 showed an increase in the incidence of diagnosed childhood Coeliac Disease in South Glamorgan following the introduction of serological testing (Antigliadin and anti-endomysial) in 1990.

Aim: To determine the incidence of CD within South Glamorgan between 1999 and 2007, and the effect of the introduction of anti-tTG testing in 2004.

Methods: CD incidence rates were calculated for the area of South Glamorgan between 1999 and 2007. Mean incidence rates were calculated for each 4 year interval. Population figures were taken from the Registrar General's mid-year estimates of childhood population.

Results: A total of 50 children aged under 16 were identified as residing in the area of Cardiff and the Vale of Glamorgan - 36 were diagnosed between 1999-2006. The total childhood population decreased slightly from 96,553 in 1999 to 89,630 in 2006.

Period	Incidence per 100,000 children per year
1983 to 86	1.38
1987 to 90	2.21
1991 to 94	1.66
1995 to 98	4.70
1999 to 02	3.16
2003 to 06	6.88

Conclusions: The diagnosis of childhood CD in South Glamorgan generally rose throughout the study time. A significant increase in incidence of the disease has been identified since the year 2004. This coincides with the introduction of anti-tTG testing. Despite this most cases of coeliac disease remain undiagnosed.

Ulcerative colitis and Surgical Intervention in a UK paediatric population

K.P. Crook, D.H. Casson, On behalf of the Register of Paediatric IBD

Introduction: Surgical procedures for Inflammatory Bowel Diseases are referred to as the last resort. There are currently no data available to quantify the risk of surgery within the UK for the paediatric population.

Objective: To identify the risk of surgery for paediatric patients diagnosed with Ulcerative colitis

Methods: The Paediatric Register of Inflammatory Bowel database (PRIBD) collates data from 8 paediatric gastroenterology centres in the UK. All patients had consented to participate in this data collection; the details were entered via a secure data entry system. 604 children were registered with 372 patients having follow-up data available.

Results: Of the 372 patients with follow-up data, 128 children were diagnosed with Ulcerative Colitis with a median age at diagnosis of 12 years (range 2-16), 17 patients, 10 female and 7 male, were identified as having a surgical procedure. The median age at diagnosis of the patients who underwent surgery was 13 years (range 6-15), and the median age at surgery 14 years (range 8-16). 8 patients had their first surgical episode within a year of diagnosis (range 0-11 months), 6 within 2 years (range 13-22 months) while 2 had surgery 3 years after diagnosis and only 1 after 4 years. Subtotal colectomy was the most common initial procedure (14/17), followed by panproctocolectomy (2/17) and panproctocolectomy with ileal anal anastomosis (1/17), 1 patient initially had a diversionary stoma followed 17 months later by colectomy.

The number of surgical procedures ranged from 1 to 8. Majority of patients (7/17) required 3 surgical interventions 5 for reversal of ileostomy following pouch surgery, 1 pouch anal anastomosis and 1 needed reformation of stoma 31/2 years after having their ileostomy reversed. 6/17 patients only had 1 surgical episode and continue to have an ileostomy with no pouch surgery (length of follow-up 2-9 years). There were 3 patients who required 2 surgical interventions, 1 who had subtotal colectomy required further surgery for adhesions, 1 had a subtotal colectomy and 1 had panproctocolectomy with pouch-anal anastomosis. The patient that required 8 procedures initially had a single stage panproctocolectomy with ileoanal anastomosis and needed subsequent surgery for a variety of reasons – refashioning of stoma, adhesions x 3, drainage of abscess, closure of ileostomy, and reformation of ileostomy.

All patients had been on steroid treatment before surgery, 9/17 had also been on azathioprine. Azathioprine was started in 2 patients following surgery. 5ASA treatment was used in 7/17 with cyclosporine in 2/17.

Only one of patients was reported as having a relapse following surgery although he did not require further surgical intervention, however 2 patients did need to have their stomas reformed following ileal-anal anastomosis (12%). 2 patients had further surgery related to adhesions.
Of our 17 patients 12 patients continue to have an ileostomy, only 2 of these resulted from pouch complications. The 5 patients who have had successful pouches had the surgery over 3 stages.

Conclusion: 6% of patients will undergo colectomy within 1 years of diagnosis. This accords with other paediatric and adult studies which have indicated similar figures. The risk of surgical intervention decreased in this population after the first 2 years.

Video Capsule Endoscopy in Children

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Video Capsule Endoscopy (VCE) also known as wireless capsule endoscopy is an approved non invasive technique for assessing small bowel disease but data in children is limited. Currently the PillCam™ is licensed for use in children over the age of 10. Indications for performing VCE include obscure GI bleeding, iron deficiency anaemia, Crohn's disease, suspected small intestinal tumours, polyposis syndromes surveillance and suspected or refractory malabsorptive syndromes.

Aims
A retrospective audit to assess single centre experience of VCE in children over a 3 year period. The focus was indications for procedure, safety and tolerability and diagnostic yield compared to standard investigations.

Results
VCE was performed with the Given Imaging swallowable capsule.
Case notes and reports of 13 children, 6 female were examined.
Age range 11.7-17.5 years (median 15.1)
12/13 had a colonoscopy in preceding 18 months.
12/13 had a recent Barium FT or MRI abdomen to exclude strictures
8/13 pre existing conditions: 2 lymphangiectasia, 5 Crohn's, 1x Indeterminate colitis

Indications
Abdominal Pain - 4, Rectal Bleeding – 4, Assessing disease extent -4 , diagnostic -1

Safety and Tolerability

No Obstruction	
failed to leave stomach	x1
Placed endoscopically under General anaesthetic	x1
Small Bowel Passage Time	2h19 m – 7 h 15 m
3 failed to reach caecum:	delayed transit x2, battery died x1

Diagnostic Yield

Lymphangiectasia	Both cases extensive disease- surgery not possible
Bleeding Source	4/4 None found
Crohn's	2 cases pathology mild so treatment not escalated
Abdominal Pain	3/4 not IBD related
Diagnostic	CD excluded

Conclusion
VCE is safe and well tolerated in older children.
Results can influence long term management.
VCE is a useful investigative tool
The future? - adaptation for younger children?

Widespread Bullous Pyoderma Gangraenosum – a rare Complication of paediatric Inflammatory bowel disease

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Background: Pyoderma gangrenosum (PG) is a rare non-infectious neutrophilic dermatosis . In most cases it is associated with a systemic illness such as inflammatory bowel disease (IBD), rheumatoid arthritis or myeloproliferative disorders . Although patients of all ages can be affected only 3 to 4 % are children. Pathergy, the development of cutaneous lesions at the site of trauma is a frequent feature. Atypical or bullous PG is characterised by more superficial ulceration. The arms and face are more commonly affected than the legs.

Aim: Our aim was to present the case of a 15 year old girl with severe Ulcerative Colitis (UC) who developed widespread bullous Pyoderma Gangrenosum.

Subjects/Methods: She presented initially in September 2007 with abdominal pain, bleeding per rectum and raised inflammatory markers. Upper GI endoscopy and colonoscopy were suggestive of Ulcerative Colitis. Her symptoms responded to steroids and she was discharged on Asacol maintenance therapy. The girl remained in remission until February 2008 when she represented with a severe relapse following an episode of Norovirus enteritis. She required a period of parenteral nutrition and was commenced on Methylprednisolone (40mg). Several days into her treatment pustular lesions were noticed at cannula sites which were initially thought to be infectious in nature. Despite intravenous antibiotics the lesions continued to spread over all four limbs and neck. The patient also developed a painful lesion of the soft palate. She spiked high fevers and became systemically unwell.

Results/Treatment: Dermatological review and skin biopsy confirmed widespread bullous Pyoderma Gangrenosum. She received high dose Methylprednisolone (1g) and was started on iv Cyclosporine. Pyrexia resolved within 12 hours and no new lesions formed. The PG healed within a few days and the colitic symptoms resolved. She was weaned off steroids and Cyclosporine and is currently in remission on Azathioprine maintainance therapy. She is left with mild scarring on both forearms.

Conclusion: Bullous Pyoderma Gangrenosum is a rare complication of childhood IBD. Early recognition is important as lesions can spread rapidly. Our patient responded well to high dose immunosuppressive therapy.

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Wireless capsule endoscopy and enteroscopy – What's the evidence?

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Introduction/Background: Historically, the investigation of the small bowel beyond the ligament of Treitz and proximal to a short segment of the distal ileum has proven a major challenge for adult and paediatric gastroenterologists. For decades, radiological imaging with all its known disadvantages has been the mainstay of investigating the small bowel. Is this problem, with advances in enteroscopic procedures, the introduction of wireless capsule endoscopy and major improvements of MR and CT imaging, finally resolved?

Aim: To perform a systematic review of the literature for the paediatric age group: (1) How do wireless capsule endoscopy and balloon enteroscopy compare with small bowel enteroclysis (SBE), magnet resonance enteroclysis (MRE)/graphy and computed tomography enteroclysis (CTE)/graphy? (2) Are there specific considerations or investigations before the procedure? (3) What are the indications for wireless capsule endoscopy? (4) What are the indications for balloon enteroscopy?

Methods: A detailed literature search was performed using MEDLINE, EMBASE and CINAHL databases. Separate keyword searches for the different topics [pe(a)diatric* or child* and enteroscop*/capsule endoscop* as well as pe(a)diatric* or child* and Crohn* or (inflammat* and bowel) and follow-through/enteroclysis/CT/MR/MRI] yielded 749 hits. Publications in English language were subsequently screened for duplicates and relevance for the topic. Out of approx. 170 relevant articles, clinical studies on the topic "WCE in pediatrics", "Enteroscopy in pediatrics", "CT imaging in pediatric IBD", "Enteroclysis in pediatric IBD" and "MR imaging in pediatric IBD" were selected for further evaluation. Abstracts were reviewed and relevant full papers retrieved for scoring according to Oxford EBM criteria. Recommendations were then made for each of the above questions.

Results/Summary: If local expertise is available, WCE and MRE, novel and sophisticated methods of investigating the small bowel in children, should replace conventional small bowel contrast studies. WCE has established its usefulness mainly as part of the work up of patients with suspected IBD or in cases of occult gastrointestinal blood loss/iron deficiency anaemia. Balloon enteroscopy is not widely used in the paediatric age group. However it may be a safe technique in selected patients; and provides a non-surgical technique that allows tissue sampling and endoscopic therapy. The majority of evidence for the above topics was Level 4 Oxford Centre for Evidence-based Medicine Levels of Evidence and Grade C for Grades of Recommendation. There were no RCTS and the largest case series included 87 patients. Tables of evidence and recommendations will be listed.

Conclusion: There is an overall paucity of data and to our knowledge no prospective controlled studies in paediatrics on WCE or balloon enteroscopy are yet available. To date, clinical practice is based on adult data, small paediatric studies and individual experience. Prospective comparative studies are needed to give evidence based guidance on the use of these modalities in children.

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