BSPGHAN
working for children with digestive and liver disorders

British Society of Paediatric Gastroenterology Hepatology and Nutrition

WINTER MEETING

27-29th January 2010, Liverpool

CPD approved

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

Mead Johnson Nutrition

Caring for young lives
Children's Liver Disease Foundation

E&OE

Thanks to our Principal Sponsors

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

Thanks to our Silver Sponsors

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

Brochure produced by

Louise Chorley

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British Society of Paediatric Gastroenterology Hepatology and Nutrition

Welcome address from Local Organising Committee

It is with the greatest pleasure and honour that we welcome you all to the 24th annual winter meeting of BSPGHAN and to the city of Liverpool which is a place of great ethnic and cultural diversity as a result of the port's trading legacy. We sincerely hope that you will enjoy the programme that has been brought together by the hard work of many people and especially the commitment of the contributors and support from our sponsors and that you will also enjoy some of what our city has to offer.

In conception, the meeting has been bitter-sweet to us as organisers given the sadness which has beset us following the death of our dear colleague and friend Dave Casson. Dave will be known to many of you as an ebullient, charming person and we all miss him greatly. Just to inform, that we celebrated Dave's life at Alder Hey in November by tribute with the permission and involvement of Dave's family. In addition, Dave wished to be well enough to participate in the meeting - sadly it was not to be, yet, neither he nor his family would wish his untimely death to adversely affect our enthusiasm to participate in this meeting which Dave helped to conceive. I think we owe it to him to grasp this opportunity with both hands as he would have done and both enjoy the scientific and medical learning opportunities as well as maintaining and strengthening our friendships.

It is not an exaggeration to say that the meeting would not be taking place but for the professional and unstinting administration and organisation skills of Carla Lloyd with her ever present optimism and enthusiasm. We are hugely grateful Carla - thank you so much.

The programme speaks for itself and will hopefully engage and stimulate you. It is a privilege to host the meeting and we are all made up as we say here - let us enjoy.

For "Guests, wives and partners"

Liverpool has much to offer and we would be delighted to assist those persons not attending the whole meeting to find alternative attractions. Some suggestions: Liverpool Bus tour & Mersey Ferry, Metropolitan & Anglican Cathedrals, Liverpool Philharmonic Hall and theatres, Liverpool & Everton football Clubs, Liverpool One shopping centre and Albert dock complex, Walker Art Gallery and Tate Gallery, Maritime and Liverpool Museums, Numerous pubs including the Philharmonic, Crack, Dr Duncan's, Enjoy.

For full details on Liverpool attractions visit the website http://www.liverpool.gov.uk/Leisure_and_culture/index.asp

Local organising Committee Liverpool 2010

Back row Left to Right
Marcus Auth, Gill Murphy, Emma Whittle, Maria Conaghan, Ramesh Srinivasan, Mark Dalzell

Front row Left to Right
Brenda Hill, Tracey Irvine, Sharon Irving, Kay Crook, Michelle Simon

Abstract Selection Committee

Dr Susan Protheroe, Education Representative for BSPGHAN, Birmingham Childrens Hospital
Dr Marcus Auth, Alder Hey, Liverpool
Dr Ramesh Srinivasan, Alder Hey, Liverpool
Dr Richard Hansen, Trainee Representative BSPGHAN, Aberdeen

Dave was born in Didsbury Manchester and attended Manchester Grammar School before obtaining a BA in Physiological Sciences at Oxford and MBBS in London in 1988. I first met Dave when he came to see me in 1995 with ambition to be a Paediatric Gastroenterologist. Following a period in London with John Walker Smith as Lecturer/Honorary Senior Registrar in Paediatric Gastroenterology at The Royal Free Hospital, he spent 6 months in Australia at The New Children's Hospital Sydney in early 1999 and was appointed as Consultant Paediatric Gastroenterologist at Alder Hey Children's NHS Foundation Trust in December 1999. Together we established a Paediatric Gastroenterology unit at Alder Hey over the next 10 years.

Dave worked long hours and his commitment to his patients and to his other professional interests including the National paediatric inflammatory bowel disease registry was unstinting. Dave had a hunger for knowledge and idiosyncratic phenomena fascinated him. He always wanted to know the reason why? If he had not studied medicine, I am sure he would have found a career in scientific enquiry. As a youngster Dave was a more than competent swimmer and butterfly was his best stroke. In recent years Dave found great pleasure in open water swimming with friends and colleagues Matthew, Colin and Steve and only months before his illness was revealed, completed a relay cross channel swim in the summer of 2008. One of many amusing episodes he recounted was in sampling the delights of a local lake and being chased from the water by a less than enamoured swan unappreciative of a paddling companion – he was unscathed – physically.

Despite his outgoing persona resulting in him being the focal point within a room, Dave was always a private person. He was a proud father and husband and said to me with some irony, that the most recent months were the happiest of his life, having the greatest opportunity to spend with his wife Penny, and children Ella and George. Dave's cheerful disposition uplifted colleagues and patients alike and he is one of those personalities once met, never forgotten.

AMD Sept 2009

Dr David Howard Casson, 8th December 1963 - 28th September 2009

Wednesday 27th January 2010

POSTGRADUATE DAY CONFERENCE

Venue: Concert Room, St George's Hall, Liverpool

"Putting theory into practice"

8.30 – 10.00
Registration and Coffee – North Hall

Exhibitor Stands in Great Hall

"Life is short, science is long; opportunity is elusive, experiment is dangerous, judgement is difficult. It is not enough for the physician to do what is necessary, but the patient and the attendants must do their part as well, and circumstances must be favourable"

Session 1

Sponsored by Children's Liver Disease Foundation

Chairs:
Dr Su Bunn, Consultant Paediatric Gastroenterologist,
Royal Victoria Infirmary, Newcastle upon Tyne,
and
Mrs Jaswant Sira, Viral Hepatitis Research Nurse,
Birmingham Children's Hospital, Birmingham

Introduction Dr Mark Dalzell

10.00 – 10.20

Neonatal liver failure due to inborn errors of metabolism.
Dr Andrew Morris
Consultant in Metabolic Diseases
Royal Manchester Children's Hospital
Oxford Road
Manchester, M13 9WL

10.20 - 10.40

Chronic Viral Hepatitis.
Dr Suzanne Davison
Consultant Paediatric Hepatologist
Children's Liver and GI Unit
Ward 11 Glenhow Wing
St James University Hospital
Beckett Street, Leeds
LS9 7TF

Session 2

Chairs:
Professor Billy Bourke, Consultant Paediatric Gastroenterologist, Dublin
and
Dr Jo Blair, Consultant Paediatric Endocrinologist, Alder Hey, Liverpool

10. 45 - 11. 05

Improving growth in children with inflammatory bowel disease
Dr Jarod Wong
SpR, Paediatric Endocrinology
Yorkhill Hospital
Dalnair Street
Glasgow, G3 8SJ

11.05 - 11.25

GI polyposis – Screening and Surveillance guidelines
Dr Warren Hyer
Consultant Paediatric Gastroenterologist
Northwick Park and St Mark's Hospital
Watford Road
Harrow
Middlesex, HA1 3UJ

Session 3

Chairs:
Mrs Pat Coldicutt, Clinical Nurse Specialist, Alder Hey, Liverpool
and
Dr Robert Heuschkel, Consultant Paediatric Gastroenterologist,
Addenbrookes Hospital, Cambridge

11.30 - 11.50

Stoma care in IBD. Dermatological perspectives.
Dr Calum Lyon
Consultant Dermatologist
Nuffield Health York Hospital
Haxby Road
York, YO31 8TA

11.50 - 12.10

Food allergy – an allergist's perspective
Dr Helen Cox
Consultant in Paediatric Allergy and Immunology
Imperial College London
South Kensington Campus
London, SW7 2AZ

12.10 – 13.00
BUFFET LUNCH
Great Hall

"Black excrement, like blood, appearing spontaneously has a serious significance whether it be accompanied by fever or not. The darker it is the more serious the condition. But when dark stools are due to drugs, however dark the colour, it is of little significance."

Session 4 – Concert Hall

Chairs:
Dr Marcus Auth, Consultant Paediatric Gastroenterologist, Alder Hey, Liverpool
and
Dr Sian Snelling, Consultant Paediatrician, Alder Hey, Liverpool

13.00 - 14:20: Fabricated illness in Paediatric Gastroenterology. Selection of cases (4 x 20min) submitted by members.

13.00 - 13.20

A serious case of Factitious Illness in an infant with Autosomal Dominant Polycystic Kidney Disease that lead to Child Protection Care Proceedings, a Criminal Conviction and a Custodial Sentence following Laxative poisoning.
Presenting Author: Dr Ieuan Davies, Consultant Paediatric Gastroenterologist,
University Hospital of Wales

13.20 - 13.40

Fabricated and factitious illness in 4 patients referred with intestinal failure to tertiary Paediatric Gastroenterology Centre
Presenting Author: Dr Fevronia Kiparissi, Staff Grade in Paediatric Gastroenterology
Kiparissi F, Elawad M, Hill S, Shah N, Lindley K
Institute of Child Health/Great Ormond Street Hospital, London

13.40 - 14.00

Review of children with medically unexplained symptoms referred to a tertiary paediatric gastroenterology centre
Presenting Author: Sarah Kapur, Merseyside Deanery, University of Liverpool.
Sarah Kapur, Merseyside Deanery, University of Liverpool.
Marcus KH Auth, Paediatric Gastroenterology, Alder Hey Children's NHS Foundation Trust

14.00 - 14.20

Fabricated Illness – Lessons from a gastroenterology and paediatric surgical service
Presenting Author: Sarah Wood, SpR, Paediatric Surgery, Royal Manchester Children's Hospital, Oxford Road, Manchester
S Conway[1,2], S Snelling[4,5], M Dalzell[3], PD Losty[1] Dept Paediatric Surgery – Alder Hey[1] and Manchester[2], Paediatric Gastroenterology, Alder Hey[3] and Community Paediatrics[4] and The University of Liverpool[5]

14.20 – 14.40
Coffee

Session 5

Chairs:
Professor Bhu Sandhu, Consultant Paediatric Gastroenterologist,
Royal Hospital for Children, Bristol
and
Ms Jo Grogan, Dietician, Royal Manchester Children's Hospital

14.40 - 15.00

Multi Disciplinary Feeding Clinic for Children with Complex Congenital Heart Conditions
Wendy Blumenow
Senior specialist speech and language therapist
Royal Liverpool Children's NHS Trust
Alder Hey Children's NHS Foundation Trust
Eaton Road
Liverpool, L12 2AP

15.00 - 15.20

New UK/WHO Growth charts
Dr Subra Mahadevan
Consultant Paediatrician
Dept of Paediatrics
Russell's Hall Hospital
Pensnett Road
Dudley
DY1 2HQ

15.20 - 15.40

Capsule endoscopy – Indications and diagnostic findings.
Dr David Campbell
Consultant Paediatric Gastroenterologist
Sheffield Children's Hospital
Western Road
Sheffield, S10 2TP

15.40 - 16.00

Protein losing gastropathy (Ménétrier's disease) associated with swine flu
Dr Sona Matthai
SpR Gastroenterology
Sheffield Children's Hospital
Western Bank
Sheffield, S10 2TH

16.00 - 17.00

Associate Members Group Meeting
Trainee Group Meeting
Education Committee Meeting

17.00 - 18.00

Endoscopy Steering Group Meeting
IBD Nurses Group Meeting

18.00 - 19.00
Football / Netball / Guided Tour of the Walker Art Gallery

19.30
Reception and Meet the Sponsors Dinner
at the 'Alma da Cuba'

Thursday 28th January 2010

BRITISH SOCIETY OF PAEDIATRIC GASTROENTEROLOGY, HEPATOLOGY AND NUTRITION 24TH WINTER MEETING

Venue: Concert Room, St George's Hall, Liverpool

"What we need to know"

08.00 - 9.45

PEGHAN Group Meeting

8.30
Registration - North Hall

8.30 - 9.45
Exhibition and Coffee Great Hall

09.45 - 10.00
Opening and Welcome

24th Annual Meeting
British Society of Paediatric Gastroenterology, Hepatology and Nutrition

Dr Mark Dalzell
Consultant Paediatric Gastroenterologist, Alder Hey, Liverpool

Session I

Chairs:
Dr Keith Leiper, Consultant Gastroenterologist, Royal Liverpool Hospital, Liverpool
and
Ms Kay Crook, Clinical Nurse Specialist, Alder Hey, Liverpool

10.00 - 10.20

Bacteria in the pathogenesis of Inflammatory Bowel Disease
Professor Jonathon Rhodes
Royal Liverpool University Hospital
Prescot Street
Liverpool
L7 8XP

10.20 - 10.40

Parenteral Iron replacement in Inflammatory Bowel Disease
Dr Stefanie Kulnigg
Medical University of Vienna
Department for Gastroenterology and Hepatology
AKH Wien, Klinik Innere Medizin 4
Wahringer Gurtel
A-1090
Vienna

10.40 - 11.00

Magnetic resonance imaging of the abdomen in Paediatric Inflammatory Bowel disease - Is it the end of the small bowel follow through?
Dr Gurdeep Mann
Consultant Paediatric Radiologist
Royal Liverpool Children's NHS Trust
Alder Hey Children's NHS Foundation Trust
Eaton Road
Liverpool
L12 2AP

11.00 – 11.25
Coffee and poster viewing – Great Hall

Session II

Chairs:
Professor Paul Losty, Consultant Paediatric Surgeon, Alder Hey, Liverpool
and
Dr Wael El-Matary, Consultant Paediatric Gastroenterologist, Alder Hey, Liverpool

11.25 - 11.45

Anti Reflux surgery – Indications, Procedures and Outcome.
Mr Matthew Jones
Consultant Paediatric Surgeon
Royal Liverpool Children's NHS Trust
Alder Hey Children's NHS Foundation Trust
Eaton Road
Liverpool, L12 2AP

11.45 - 12:05

Laryngopharyngeal reflux in Children. Diagnosis, Investigation and Management.
Dr Tobias Wenzl
Kinderklinik
Universitätsklinikum Aachen,
Pauwelsstrasse 30
D 52074,
Aachen
Germany

12.05 - 12.25

Eosinophilic Oesophagitis – Recent advances in Management.
Dr Mike Thomson
Consultant Paediatric Gastroenterologist
Sheffield Children's Hospital
Western Bank
Sheffield
S10 2TH

12.30 - 13.30
BUFFET LUNCH

Poster viewing and judging

"As for the daily changes in the weather: a north wind stimulates the body and makes it of good tone and agile, and makes for a good complexion and acuity of hearing: the bowels are constipated and the eyes sting. On the other hand, south winds relax the body, make the tissues moist, reduce acuity of hearing and produce headaches and vertigo. Movement both of the eyes and of the body generally is sluggish and the bowels relaxed"

Session III

Chairs:
Dr Peter Sullivan, Consultant Paediatric Gastroenterologist, Oxford
and
Dr Mark Beattie, Consultant Paediatric Gastroenterologist, Southampton

13.30 - 15.00: Plenary oral session I

13.30 - 13.40

Azathioprine use in children with inflammatory bowel disease: A survey of clinical practice in UK
Presenter: Himadri Chakraborty
Himadri Chakraborty, Ipswich Hospital NHS Trust; Mary-Anne Morris Norfolk and Norwich University Hospital

13.40 - 13.50

Restorative Proctocolectomy in Paediatric Ulcerative Colitis - A Single UK Centre Experience
Presenter: A. Anish
A.Anish[1], S.Bunn[1], B.McLain, P.Dryden[1], B.Jaffray[2]
[1]Department of Paediatric Gastroenterology, [2]Department of Paediatric Surgery, Royal Victoria Hospital, Queen Victoria Road, Newcastle upon Tyne, NE1 4LP

13.50 - 14.00

Wide variation in diagnostic criteria and management approach to Eosinophilic Oesophagitis in the UK: Results of a BSPGHAN survey
Presenter: Naresh P Shanmugam
Naresh P Shanmugam[1], Paraic McGrogan[2], John Fell[1]
[1] Chelsea and Westminster Hospital, London, [2] Royal Hospital for Sick Children, Glasgow, Royal Hospital for Sick Children, Glasgow

14.00 - 14.10

Rationalisation of services has failed to improve outcomes for biliary atresia in Scotland. A report on behalf of the Scottish Paediatric Gastroenterology Hepatology and Nutrition Group (SPGHANG)
Presenter: Rachel Taylor
Rachel Taylor[1], Andrew R Barclay[1], David Devadson[2] Pam Rogers[2], Mike Bisset[3], Karen McIntyre[4], Richard K Russell[1] Paraic McGrogan[1]
1. Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Glasgow
2. Department of Paediatric Gastrenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh
3. Department of Paediatric Gastroenterology and Nutrition, Royal Aberdeen Childrens' Hospital
4. Department of Paediatrics, Ninewells Hospital, Dundee

14.10 - 14.20

Non-invasive biomarkers and paediatric NAFLD: new methods to predict disease and stratify severity
Presenter: Emer Fitzpatrick
Emer Fitzpatrick[1], Ruth deBruyne[1], Alberto Quaglia[2], Ragai Mitry[2] and Anil Dhawan[1,2] Paediatric Liver, GI and Nutrition Centre[1] and Institute of Liver Studies[2], King's College Hospital

14.20 - 14.30

Trial of a micronutrient rich, high fibre, low energy density enteral feeding formula for gastrostomy feeding disabled children
Presenter: Peter B Sullivan
Angharad Vernon-Roberts[1], Dr Jonathan Wells[2], Dr Muftah Eltumi[3], Dr Peter B Sullivan[1], [1]Oxford University Department of Paediatrics, Oxford, [2]UCL Institute of Child Health, London, [3]Watford General Hospital

Plenary poster session I

(3 mins plus 2 mins discussion)

To be chosen on day

14.35 - 14.40

14.40 - 14.45

14.45 - 14.50

15.00 - 15.30
Coffee

Session IV

Chairs:
Dr John Puntis, Consultant Paediatric Gastroenterologist, General Infirmary, Leeds
and
Mr Naved Alizai, Consultant Paediatric Surgeon, St James, Leeds

15.30 - 16.00

Fibropolycystic diseases of the Liver - ***sponsored by*** Caring for young lives Children's Liver Disease Foundation
Dr Patrick McKiernan
Consultant Paediatric Hepatologist
Liver Unit , Birmingham Children's Hospital
Steelhouse Lane, Birmingham B4 6NH

16.00 - 16.45

Chronic Pancreatitis: Results from the EUROPAC study
Mr Bill Greenhalf
University of Liverpool, 5th Floor, UCD Block
Royal Liverpool University Hospital
Daulby Street, Liverpool, L69 3GA

Session V

Chairs:
Professor Ian Booth, Consultant Paediatric Gastroenterologist, Birmingham Children's Hospital, Birmingham
and
Ms Tracey Irvine, Clinical Nurse Specialist, Alder Hey, Liverpool

16.50 - 17.30

Quality indicators with relation to Paediatric Gastroenterology
Mr John Smith
Director of Consultancy
Civil Eyes Research Limited, PO Box 59509
Dulwich, London SE21 7WL

17.30 - 19.00

Annual General Meeting

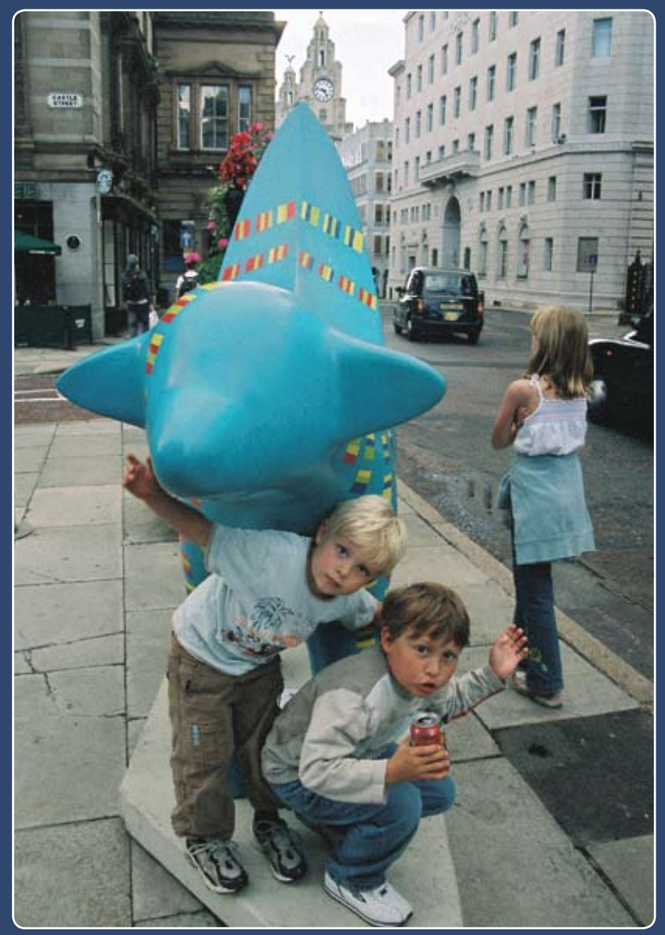
20.30
Reception: Marriott Hotel

21.00 - Late
Gala dinner 60's night entertainment with 'THE BEATLES', followed by disco

Friday 29th January 2010

"MORE OF THE SAME"

Venue: Concert Room, St George's Hall, Liverpool



08.00 - 09.00
Breakfast Symposium: GI Motility Disorders

Chairs:
Professor Peter Milla, Consultant Paediatric Gastroenterologist,
Great Ormond Street Hospital, London
and
Dr Ramesh Srinivasan, Consultant Paediatric Gastroenterologist,
Alder Hey, Liverpool

08.00 - 08.30

Overview of Paediatric GI motility disorders – Investigation and Management.
Dr Manu Sood
Paediatric Gastroenterologist
8701 West Watertown
Plank Road
Milwaukee
WI 53226

08.30 - 09.00

Advances in the investigation of functional abdominal Pain
Professor Qasim Aziz
Professor of Neurogastroenterology
Bart's and the London NHS Trust
Princess Grace Hospital
47 Nottingham Place
London
W1U 5LZ

Session VI

Chairs:
Dr Mark Dalzell, Consultant Paediatric Gastroenterologist, Alder Hey, Liverpool
and
Mr Khalid Sharif, Consultant Paediatric Transplant Surgeon,
Birmingham Children's Hospital, Birmingham

9.10 - 9.55 Key note address

Advances in the Management of Intestinal failure associated Liver Disease
Prof. Olivier Goulet, Paris.
Professor of Paediatrics
Service de Gastroentérologie pédiatrique
Hôpital Necker Enfants malades
149, rue de Sèvres
75743 PARIS Cedex 15

9.55 - 10.40

UK small bowel transplant experience
Dr Sue Beath
Consultant Paediatric Hepatologist
Birmingham Children's Hospital
Liver Unit
Steelhouse Lane
Birmingham
B4 6NH

"As a general rule, if those who are poorly take their food well at first, but fail to put on weight, they finish by refusing food. On the other hand, if they firmly refuse food at first but take it later on, they make a good recovery"

Session VII

Chairs:
Dr Sue Protheroe, Consultant Paediatric Gastroenterologist,
Birmingham Children's Hospital, Birmingham
and
Mr Adrian Bianchi, Consultant Paediatric Surgeon,
Royal Manchester Children's Hospital, Manchester

10.40 - 11.20 Debate

Bowel lengthening for short bowel syndrome – To STEP or not to STEP.

For

Mr Antonino Morabito
Consultant Paediatric Surgeon
Royal Manchester Children's Hospital
Oxford Road
Manchester
M13 9WL

Against

Mr Colin Baillie
Consultant Paediatric Surgeon
Alder Hey Children's NHS
Foundation Trust,
Eaton Road
Liverpool L12 2AP

11.20 - 11.40
Coffee and poster viewing

Session VIII

Chairs:
Professor Simon Murch, Consultant Paediatric Gastroenterologist, Warwick
and
Dr Naeem Ayub, Consultant Paediatric Gastroenterologist,
Royal Shrewsbury Hospital, Shrewsbury

11.40 - 12:00

Stem Cell transplantation in an animal model for Hirschsprung's Disease:
Mr Simon Kenny
Consultant Paediatric Surgeon/Urologist
Alder Hey Children's NHS Foundation Trust
Eaton Road
Liverpool
L12 2AP

12.00 - 12.20

Bone Marrow transplantation for paediatric gastrointestinal diseases.
Dr Mamoun Elawad
Consultant Paediatric Gastroenterologist
Great Ormond Street Hospital
Department of Gastroenterology
London

Session IX

Chairs:
Dr Adrian G Thomas, Consultant Paediatric Gastroenterologist,
Booth Hall, Manchester
and
Dr Balaji Krishnamurthy, Grid Trainee Gastroenterology, Alder Hey, Liverpool

12.20 - 13.00 Debate

Anti TNF therapy – Is it the Panacea?

For

Dr David Wilson
Consultant Paediatric Gastroenterologist
Child Life and Health
University of Edinburgh
20 Sylvan Place
Edinburgh
ED9 1UW

Against

Dr Huw Jenkins
Consultant Paediatric Gastroenterologist
Dept of Child Health
University Hospital of Wales
Heath Park
Cardiff
CF4 4XN

13:00 - 13:40
BUFFET LUNCH and Poster Judging

Session X

Chairs:
Dr Nick Croft, Consultant Paediatric Gastroenterologist,
Wingate Institute, London
And
Dr Alastair J Baker, Consultant Paediatric Hepatologist,
Kings College Hospital, London

13.40 - 15.10 Plenary Oral Session 2

13.40 - 13.50

Rapid increase in the incidence of paediatric inflammatory bowel disease in the Republic of Ireland.
Presenter: Raveen Shahdadpuri
Raveen Shahdadpuri, Marion Rowland, Shoana Quinn, Annemarie Broderick, Tim Bohane, Mary Hamzawi, Billy Bourke; Children's Research Centre, Our Lady's Hospital, Crumlin

13.50 - 14.00

Reduction of Sepsis rate after introduction of 2% Chlorhexidine wipes for the care of central line in children with intestinal failure on parenteral nutrition
Presenter: Judith Pichler
Judith Pichler, Venetia Horn, Sarah MacDonald, Dr. Susan Hill, Great Ormond Street Hospital, London

14.00 - 14.10

Comparison of quick point of care test for small bowel hypolactasia with biochemical lactase assay
Presenter: Prithvi Rao
P Rao, M Jordinson[1], Reed C[1], D Campbell.
Paediatric Gastroenterology and [1]Biochemistry Unit, Sheffield Children's Hospital NHS Foundation Trust, Sheffield S10 2TH, U.K.

14.10 - 14.20

Fatty Liver Disease – Is further investigation necessary?
Presenter: Balaji Krishnamurthy
Dr. B. Krishnamurthy[1], Dr. A. Urs[1], Dr. J. Stahlschmidt[2], Dr. P. McClean[1]. Department of Paediatric Hepatology[1] and Histopathology[2], St. James' University Hospital, Leeds

14.20 - 14.30

Clinical Outcome in Cystic Fibrosis Patients with or without Meconium Ileus: A Comparative Study
Presenter: Krishnappa Venkatesh
K Ventakesh, C Taylor; Sheffield Children's Hospital, Sheffield

14.30 - 14.40

The Practical Outcomes of Thiopurine Metabolite measurement: The experience of a tertiary PGHAN unit
Presenter: Lawrence Armstrong
Lawrence Armstrong[1], Peter Galloway[2], John Bishop[1], Paraic McGrogan[1], Richard K Russell[1].
[1]Dept of Gastroenterology, Royal Hospital for Sick Children, Yorkhill, Glasgow, G3 8SJ. [2]Dept of Biochemistry, Royal Hospital for Sick Children, Yorkhill, Glasgow, G3 8SJ.

Plenary poster session 2

(3 mins plus 2 mins discussion)

To be chosen on day

14.42 - 14.47

14.48 - 15.03

15.04 - 15.09

Session XI

Chairs:
Dr Marcus Auth, Consultant Paediatric Gastroenterologist, Alder Hey, Liverpool
and
Jo Grogan, Senior Paediatric Dietitian, Manchester

15.10 - 15.30

Fish oil based parenteral nutrition. Do we all need to go this way?
Dr Girish Gupte
Consultant Paediatric Hepatologist
Liver Unit, Steelhouse Lane
Birmingham B4 6NH

15.30 - 16.00

A Medical director's perspective on the current NHS reforms.
Dr Steve Ryan
Consultant Paediatrician
Royal Liverpool Children's NHS Trust
Alder Hey Hospital
Eaton Road, Liverpool L12 2AP

16.00

PRESENTATIONS OF PRIZES AND VOTE OF THANKS
Dr Mark Beattie
President BSPGHAN 2010 - 2013

CLOSE OF MEETING

25th BSPGHAN Winter Meeting
January 2011, Edinburgh
Local Organiser Dr David Wilson

ABSTRACTS FOR WEDNESDAY 27TH JANUARY 2010

Invited speakers' abstracts

Neonatal liver failure due to inborn errors of metabolism

Dr Andrew Morris, Consultant Metabolic Diseases, Manchester

Different inborn errors present at different ages and with different associated features. In the first few days, severe liver dysfunction can be due to urea cycle disorders, fatty acid oxidation disorders or organic acidaemias but the main clinical feature is encephalopathy, associated with hyperammonaemia, hypoglycaemia or acidosis. Mitochondrial disorders can cause liver failure at any age, including the first week of life. Galactosaemia typically presents in the first or second week, initially with poor feeding and jaundice. Tyrosinaemia type 1 presents with liver failure and prominent coagulopathy at any age from 2 weeks onwards. Niemann-Pick disease type C, a -1-antitrypsin deficiency and bile acid synthesis defects usually present after 3 weeks of age, predominantly with cholestasis.

Peroxisomal disorders generally present with dysmorphism or neurological problems rather than liver disease. Hereditary fructose intolerance only presents at this age if fructose is given. Wolman disease may present in the neonatal period with hepatomegaly but not liver failure. Phosphomannose isomerase deficiency (CDG 1b) & citrin deficiency seldom present within the first month. Other causes of liver failure may mimic inborn errors. Disseminated Herpes simplex can cause severe hyperammonaemia and there may have been an affected sibling in cases of neonatal haemochromatosis or haemophagocytic lymphohistiocytosis.

Mitochondrial liver disease in infants is often associated with depletion of mitochondrial DNA (mtDNA). This results from mutations in nuclear genes involved in mtDNA replication, including the genes for deoxyguanosine kinase and DNA polymerase gamma (POLG1). POLG1 defects also cause Alpers syndrome, which presents with seizures and regression in young children, followed by liver failure. Diagnosis can be difficult, as mitochondrial studies in muscle may be normal, and mutation analysis may be needed.

All these inborn errors have genetic implications and several are treatable. Nitisinone is effective treatment for tyrosinaemia but needs to be accompanied by dietary tyrosine restriction. Monitoring is needed for hepatocellular carcinoma, especially in patients who present at an older age. In galactosaemia, the liver disease responds to galactose restriction and does not recur even if the diet is normalised in older children. Unfortunately, the diet does not prevent the neurological and ovarian problems. Bile acid synthesis defects respond to treatment with cholic and chenodeoxycholic acids, and CDG 1b can be treated with mannose.

Chronic Viral Hepatitis

Dr Suzanne Davison, Consultant Paediatric Hepatologist, Leeds

Chronic infection due to Hepatitis B virus (HBV) or Hepatitis C virus (HCV) are a global health issue, leading to one million deaths annually. In Europe, 25,000 adults have undergone liver transplantation in the last 20 years as a consequence of HBV and HCV related cirrhosis or hepatocellular carcinoma (HCC). Following infection in childhood, there is a 20% lifetime risk of developing cirrhosis or HCC, though early prognostic factors are not well established.

HBV acquisition in infancy is the major risk factor for chronic infection, and transmission from an infected mother leads to perinatal infection in 95% of offspring unless successsfully immunised. Paediatricians have a role to play in prevention, early diagnosis and in assessing disease progression. Medical treatment, with interferon and nucleoside analogues, cannot eradicate HBV infection but is aimed at "damage limitation". Selecting those who are likely to benefit from treatment requires understanding of the natural history, and identifying transition through the phases of immunotolerance and immuno-clearance, to the inactive carrier state. The ability of HBV to mutate allows both reactivation and resistance to thwart attempts either of both spontaneous and treatment induced viral clearance. Fortunately, HBV immunisation is effective in preventing transmission, and is an essential strategy in reducing the morbidity and mortality of HBV related disease.

HCV infection is acquired in childhood predominantly by perinatal transmission from an infected mother. Intravenous drug use is the most frequent risk factor in the mother, and the risk of HCV transmission to offspring is 5%. Children are likely to remain asymptomatic until adulthood, though may have fluctuating ALT. Rare cases of rapid progression to cirrhosis are reported. Spontaneous clearance may occur in early childhood in up to 20%. In those who remain infected, treatment with interferon and ribavirin should be considered. Eradication is possible in 50 - 95% depending on genotype.

Families of children with HBV or HCV risk stigmatisation and social isolation. Education of the public, teachers and health care professionals is essential to avoid inappropriate management.

Improving growth in children with inflammatory bowel disease

Dr Jarod Wong, SpR Endocrinology, Glasgow

Growth retardation and pubertal delay is often encountered in children with inflammatory bowel disease (IBD), particularly those with crohn's disease(CD). The aetiology of growth retardation is multifactorial and is due to the combined effects of the inflammatory process itself mediated by pro-inflammatory cytokines, use of glucocorticoid and poor nutrition. These factors can affect growth and puberty by their effects on systemic hormonal factors leading to a degree of functional growth hormone (GH) and insulin like growth factor (IGF) insufficiency and resistance. Cyokines can also have a direct effect on local bone growth by affecting chondocyte dynamics and signalling pathway. Targeting the inflammatory process with anti-TNF therapy can improve growth in some children, although the précise mechanism of this is still unclear. In groups of children with delayed puberty and poor linear growth, pubertal induction with sex steroids may improve growth and lead to improvement in pubertal status but the response is variable and maybe less optimal compared to healthy children with constitutional delayed in puberty. Finally, in groups of children where delayed puberty has been addressed and disease is relatively well controlled, recombinant human GH or IGF-1, may be a therapeutic option. Further understanding of the basic mechanisms by which cytokines influence growth will facilitate the development of other therapeutic modalities to improve growth in children. Clinical management that address growth and puberty in these children with IBD should be in partnership between the IBD team and paediatric endocrinologist. Well designed trials of growth promoting hormonal treatment are needed to help answer questions regarding the efficacy and safety in the sub group of children with IBD who continue to grow poorly despite optimal management of their disease and nutrition.

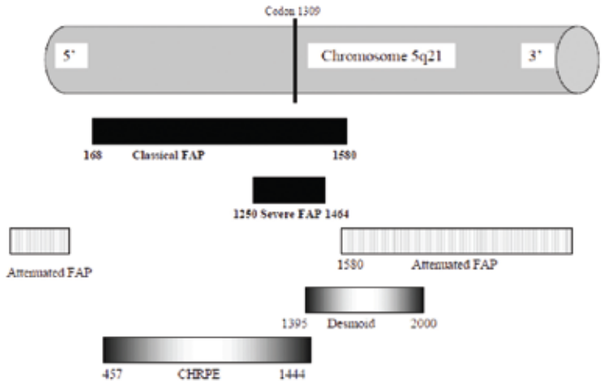
Paediatric Polyposis syndromes

Dr Warren Hyer MB ChB FRCPCH MRCP, Consultant Paediatric Gastroenterologist, Polyposis Registry, St Mark's Hospital, Middx, UK. www.polyposisregistry.org.uk warrenhyer@aol.com

Learning points:

1. Genotype – phenotype correlation in FAP.
2. IRA preferred surgical procedure in FAP unless unfavourable colon
3. Multiple juvenile polyps = JPS or syndromic JPS
4. PTEN mutations and juvenile polyposis
5. Risk of intussuception in PJS syndrome
6. Preferred imaging in PJS

Figure 1 APC protein domains showing FAP genotype - phenotype correlation with codon number



Assessment of a child with colonic or GI polyps

History

- Nature of bleeding and frequency
- Painful or painless rectal bleeding
- History of GI obstructive symptoms
- Detailed family history exploring early deaths or diagnosis of GI cancer
- Weight loss, anorexia (tumour)
- Learning difficulties (JPS or PTEN hamartoma)

Examination

- Mucosal pigmentation (PJS)
- Dysmorphic features (JPS)
- Oedema (hypoalbuminaemia in infantile JPS)
- Extra intestinal manifestations of FAP – see table 5 e.g. subcutaneous cysts, exostosis, congenital hypertrophy of the retinal pigment epithelium
- Hepatic mass (FAP)
- Thyroid mass (FAP or Cowden)

Adenomatous polyposis syndromes
Familial adenomatous polyposis
Turcots syndrome

Hamartomatous polyps
Solitary juvenile polyp
Juvenile polyposis syndrome
PTEN -hamartoma tumour syndrome e.g.
Bannayan - Riley - Ruvalcaba
Gorlin syndrome
Cowden syndrome
Peutz Jeghers syndrome

Inflammatory polyps
Mixed polyposis syndrome

Figure 2 Management protocol for screening children and adolescents at risk of FAP

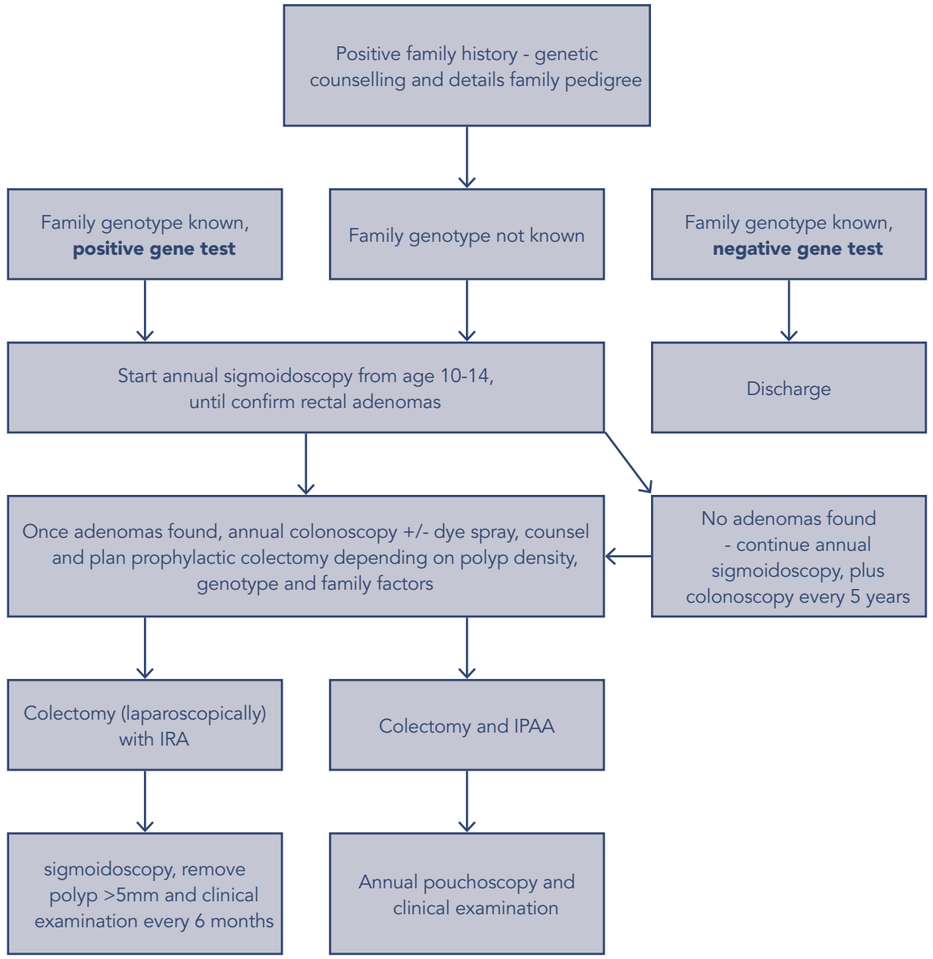


Figure 3 Management protocol of juvenile polyposis in childhood

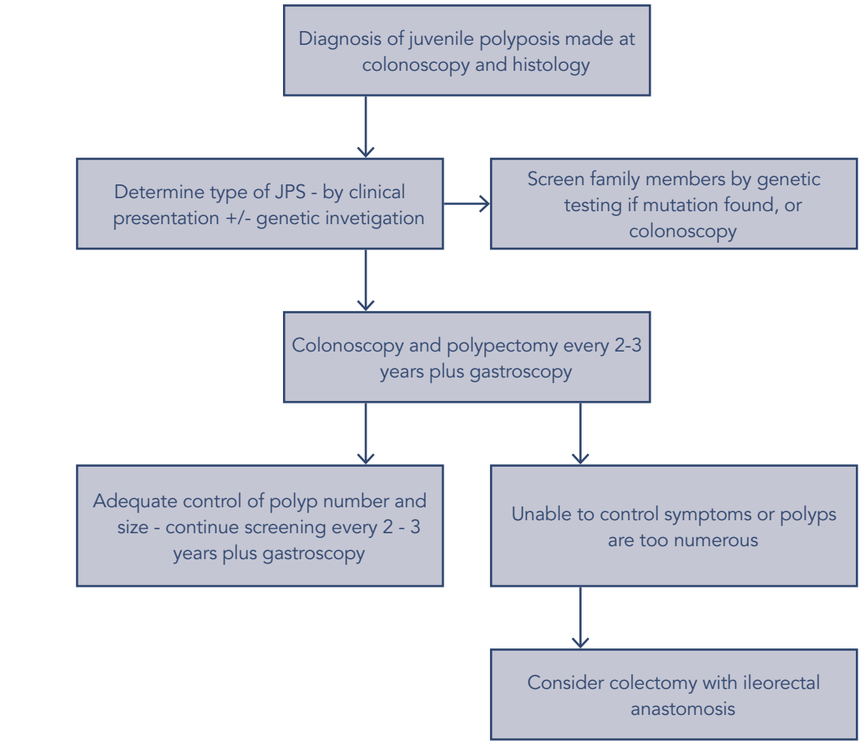
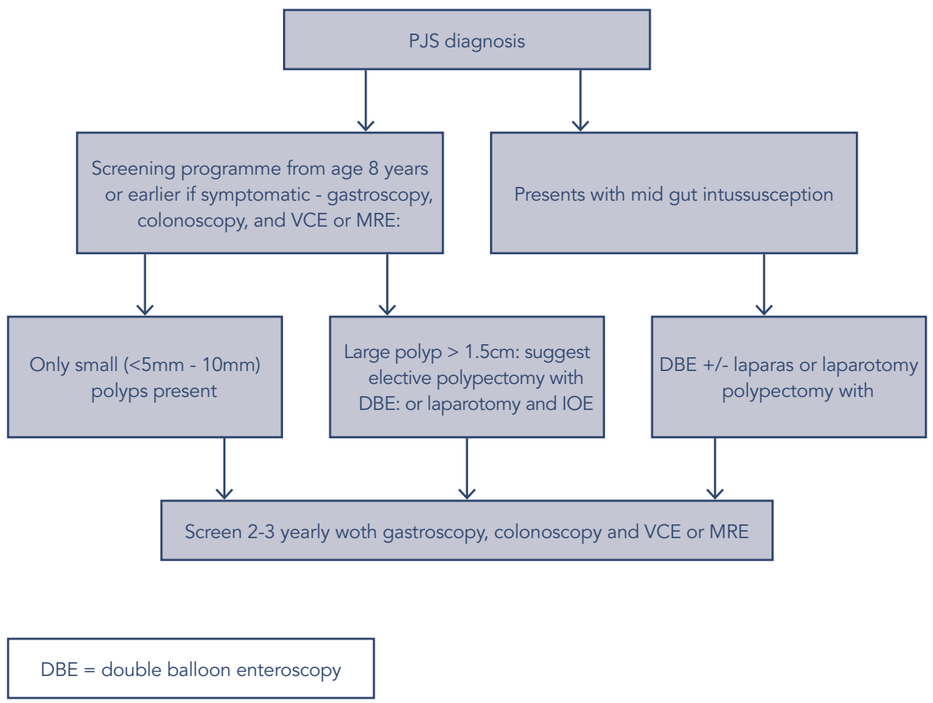


Figure 4 Management protocol for screening of children and adolescents at risk of PJS



DBE = double balloon enteroscopy

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The Skin Disorders of Abdominal Stomas

Dr Calum C Lyon, Consultant Dermatologist, York

Approximately one third of stomas are formed because of malignant disease of the bowel or renal tract, one third because of inflammatory bowel disease and one third for a range of other disorders. Surprisingly one in six patients with skin problems fails to seek professional advice usually because they mistakenly believe that peristomal skin disease is inevitable and untreatable. We set up a monthly dermatology/stoma-care clinic at Hope hospital Salford and at York hospital nearly 10 years ago initially to study these skin problems. The information presented is the result of experience gained in the\clinic.

The Stoma Patient
Common to all patients is the need to use a collecting device to contain the stoma effluent. This is usually a laminated plastic pouch attached to an adhesive barrier that sticks to the skin. The majority of skin barriers consist of a hydrocolloid gel similar to that used for wound dressings, many modern barriers also incorporate an adhesive tape component on the outer perimeter. Ileostomies and urostomies, in contrast to colostomies, produce frequent fluid effluent, so that these patients tend to have more skin problems and leaks than colostomists.

SKIN PROBLEMS
Table 1 lists the wide range of dermatoses that may affect peristomal skin.

Contact Dermatitis
Almost all patients who present with a peristomal rash believe that it is caused by an allergy to their stoma bag. In fact true allergic contact dermatitis is unusual and in our experience, when it does occur it is due to fragrance in deodorisers or components of medicaments rather than the bag materials. Irritant reactions are far commoner, accounting for over half of all the dermatoses seen. The prevalence of irritant reactions is not surprising when one considers the stresses placed upon peristomal skin.

Dermatitis due to contact with urine or faeces is the commonest single cause of peristomal skin disease and there are a number of potential causes of leakage onto the skin that should be considered (table 2). Whatever the primary cause of leakage, the resulting skin inflammation may impair bag adhesion so that a vicious cycle is set up. Management of faecal/urine irritant dermatitis involves firstly prevention advice as part of the pre and postoperative care provided by the stoma care specialist nurse. When problems do occur the bulk of them can be identified and treated by an experienced specialist nurse. The aim of treatment is to suppress inflammation so that appliance failures are minimised and further inflammation from leaks or frequent bag changes is prevented. In this situation, short term topical steroids or other anti-inflammatories are appropriate.
Two distinctive papular forms of irritant reaction can be seen in peristomal skin. The first, chronic papillomatous dermatitis is unique to urostomies and is the result of chronic leaks of urine onto the skin, especially if the urine is infected. The condition will resolve over a month or so if leaks are prevented by appliance modifications. The second papular irritant reaction affects ileostomies and colostomies and presents as red, friable papules usually referred to as "granulomas". They are normally restricted to the stoma itself but if there is recurrent faecal leakage they will begin to involve the peristomal skin. Most cases are limited and asymptomatic but they can be sore and bleed easily causing leaks. In these situations it is appropriate to ablate them. This can be achieved by cryotherapy, laser or Silver nitrate cautery.

INFECTIONS
Infections of the peristomal skin account for 7% of the skin problems seen in our clinic. Viral infections are rare. Colonization of the peristomal skin with Candida species is common although frank infection is less so. When it does occur it presents like a Candidal intertrigo with an itchy erythema surrounded by papular or pustular satellite lesions.

Bacterial folliculitis is relatively common and typically affects men who shave their abdomen to prevent discomfort on appliance removal, but it can affect any stoma patient.

Superficial bacterial infection with any of wide range of pathogens can produce rashes clinically indistinguishable from irritant contact dermatitis, it is therefore important to swab all peristomal rashes and treat potential pathogens with specific antibiotics.

Primary Skin Disease Affecting Stomas
Theoretically any of the multitude of skin diseases might affect peristomal skin. In practice the common conditions psoriasis seborrhoeic dermatitis and eczema account for almost all the disorders in this group.

We have seen over 1100 new patients ranging in age from 6 months to 91 years old. The older age group includes more colostomists and urostomists for cancer whereas the younger age groups has relatively greater numbers of ileostomists for IBD and congenital problems e.g. Hirchsprings.

In the talk I will obviously concentrate on IBD in the younger age group and discuss irritant problems Crohn's, Pyoderma and coincidental skin diseases

Table 1

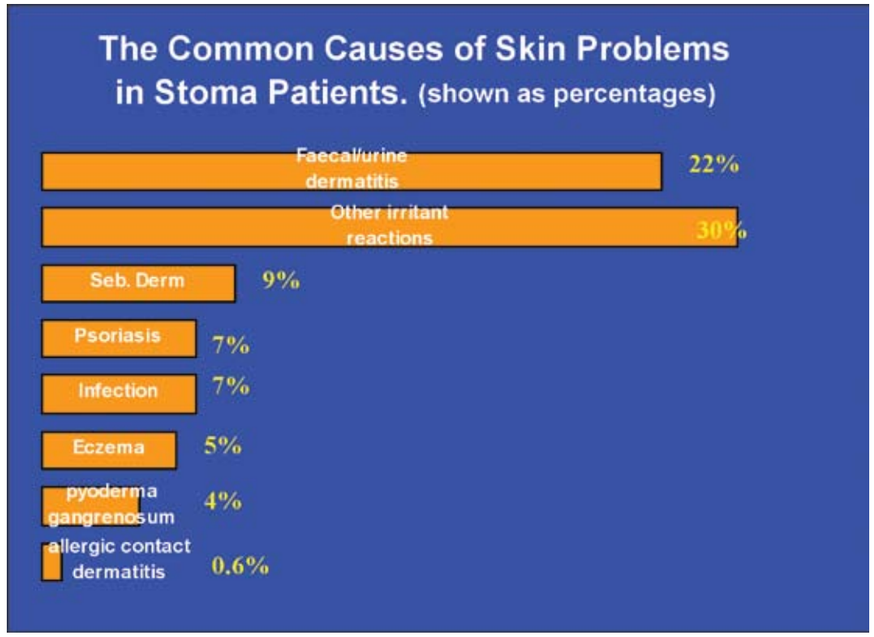

Table 2 Causes of Stoma Leakage
- Use of an appliance with too large an aperture allowing effluent to bathe the peristomal skin and cause inflammation
- Buried stoma. Weight gain after surgery may cause the stoma to be buried in abdominal fat preventing good appliance fitting
- Short or receding stoma (often related to postoperative infection)
- Inappropriately sited stoma e.g. near umbilicus, scar tissue or in body fold
- Very high output stoma especially jejunostomy where the corrosive effluent can damage the appliance barrier
- Skin disease that impairs bag adhesion especially psoriasis,eczema and local infection
- Surgical complications, particularly parastomal hernia and prolapsing stomas that cause the bag to lift
- Use of oily cream or ointment preparations (including medicaments). These cause the bag to lift because of poor adhesion.

Food allergy - an allergist's perspective

Dr Helen Cox, Consultant in Paediatric Allergy and Immunology, London

Atopic eczema has been proposed as the cutaneous manifestation of a systemic disorder that also gives rise to asthma, food allergy and allergic rhinitis. This observed pattern is frequently referred to as "the allergic march".[1] Sensitisation to food and inhalant allergens increases with increasing eczema disease severity, suggesting a role for the skin barrier in initiating allergic disease. The recent discovery of novel skin barrier genes for atopic eczema ie fillagrin, a key hygroscopic protein in the epidermis, offers a mechanistic understanding for the role of the epithelial barrier in allergic sensitization and eczema pathogenesis.[2]

Sensitisation to food and inhalant allergens in infancy predicts for subsequent allergic disease.[3] 1:5 infants with egg allergy develop nut allergy; Sensitisation to house dust mite and egg allergy in infancy predicts for the onset of asthma at 5 years; food allergen sensitization increases the risk of adult asthma (OR 12).[3] Published threshold levels for specific IgE to milk, egg, peanut and cod enable the clinician to predict the probability of clinical reactivity to specific foods prior to introduction.[4,5]

Despite sensitisation to numerous foods, clinical reactivity occurs to relatively few foods with cows milk, hens egg, soya, nuts, fish, shellfish, sesame and kiwi accounting for the majority of food induced allergic reactions. Delayed phase reactions are commonly seen, with up to 40% of patients developing eczematous reactions within 72 hours of challenge.[6] These observed patterns of food reactivity have challenged the traditional concepts of IgE and Non-IgE mediated food allergy with overlapping phenotypes occurring in patients with atopic eczema and eosinophilic gastroenteropathy.

At a cellular level there has been improved characterization of allergens with recognition of heat labile, conformational and heat stable, sequential epitope structures governing the properties of common allergens. Recent studies have eluded to the presence of epitope binding patterns conferring risk factors for disease persistence and severity eg multiple epitope binding to ara h1,2 and 3 predicts for peanut allergy persistence and anaphylaxis.[7] It also opens up avenues for understanding allergen co and cross reactivity allowing the clinician to tease out irrelevant specific IgE binding such as is frequently observed in patients with the oral allergy syndrome.

The observed patterns of allergic disease have raised questions as to whether there are opportunities to intervene and potentially halt this process. There are currently studies underway addressing models of primary and secondary prevention eg early oral tolerance to peanut (LEAP study), SLIT in inhalant naïve eczema patients. Immunotherapy offers scope for both prevention and treatment and novel anti-inflammatory agents ie FAHF2 and anti IgE are currently being explored.[8]

Recognition of the full spectrum of allergic gut disease in patients presenting to allergists is becoming an essential part of clinical practice. The sharing of expertise between allergists and gastroenterologists at a clinical, genetic and molecular level can only benefit advances in our understanding of the pathogenesis and management of food allergic disease.

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Multi Disciplinary Feeding Clinic for Children with Complex Congenital Heart Conditions

Wendy Blumenow, Senior Specialist Speech & Language Therapist, Alder Hey Children's NHS Foundation Trust

Alder Hey Children's Hospital, Liverpool runs a weekly feeding clinic for children with congenital heart conditions. It is staffed by a consultant gastroenterologist, cardiac liaison nurse, specialist speech and language therapist and specialist dietician. This presentation will highlight the benefits of multidisciplinary working and early diagnosis of children with feeding difficulties related to heart problems. Included in this will be the results of a recent Quality of Life audit carried out with those families who have had a Percutaneous Endoscopic Gastrostomy inserted.

UK WHO growth chart – same measurements but new Growth chart

Dr Subramanian BK Mahadevan, Russell's Hall Hospital, Dudley

UK-WHO chart: Resources and Materials for NHS Staff and Experienced Chart Users are available from www.rcpch.ac.uk/Research/UK-WHO growth charts.
Growth assessment is the single measurement that best defines the health and nutrition status of children and provides an indirect measurement of the quality of life of an entire population.

Historically, pediatric healthcare providers have used height, weight, and head circumference measurements to assess changes in growth and development. These anthropometric measurements, a basic component of healthcare services for children, have been used to screen individuals and populations for nutrition-related health problems.

Why did the WHO think we needed new charts?
All previous growth charts have been based on a mixture of breast and bottle fed infants and differences in weight gain were seen between breastfed infants and these charts.

UK 1990 growth charts
These were constructed using measurements from a large number of British children at different ages collected in the late 1980s and were the main charts in use. They were based on data from studies on breast - and formula-fed children so do not reflect normal weight fluctuations of breast-fed infants in the first few weeks.

There has been a growing feeling that 'when we have a chart based on breastfed babies, we won't get women being told to supplement because their babies are falsely diagnosed as not gaining enough weight'.

At the same time it was found that healthy breastfed infants showed very similar growth patterns around the world. The WHO therefore decided to produce charts that set breastfeeding as the norm and described optimal rather than average growth that could be used worldwide. The process of planning, data collection and analysis took 15 years and charts were finally published in 2006.

The new UK-WHO growth charts for children aged 0-4 years (combines WHO and UK90 data) were launched on 11th May 2009 and have been developed for the Department of Health by the Royal College of Paediatrics and Child Health will replace current UK 1990 charts for this age group, 0-4 years.

Why Were the Growth Charts Revised? UK-WHO growth charts
One of the most important factors in assessing a child's growth is having an appropriate reference population. The WHO Multicentre Growth Reference Study (MGRS) collected primary growth data and related information from approximately 8500 children from widely different ethnic backgrounds and cultural settings (Brazil, Ghana, India, Norway, Oman and the USA). They were exclusively breastfed for the first 4 months, and were living in a well-supported health environment.

The new growth curves are expected to provide a single international standard that represents the best description of physiological growth for all children from birth to five years of age and to establish the breastfed infant as the normative model for growth and development.

In consequence the WHO aims to provide for the first time a standard on **"how children should grow" (Optimal growth chart)**, rather than a traditional growth reference that describes **"how children are growing" (Reference growth chart)**. The process of planning, data collection and analysis took 15 years and charts were finally published in 2006.

This standard was adopted by the UK for children under 4 years and used to construct the UK-WHO charts. Because the WHO charts do not include preterm data, the UK 1990 data have been used to make the birth section of the UK-WHO charts, as well as the charts for use after the age of 4.

Research shows that breast-fed babies tend to gain weight at a healthier pace and are less likely to become obese in later life. The new charts will play an important role in establishing breastfeeding as the norm. They will help parents and healthcare professionals identify children at early risk of obesity and provide important reassurance for parents of breast-fed babies, who are likely to gain weight more slowly.

The decision to produce UK-WHO Charts
The Scientific Advisory Committee on Nutrition (SACN) studied the charts once they had been released and looked at how UK children compared to them and, in 2007, recommended that they be adopted in the UK. It is available for use in public since May 2009.

They decided to adopt from age 2 weeks as this would allow continued use of the UK 1990 preterm and term birth data. There will then be a switch back to UK 1990 charts at age 4 to allow all school entry measures to be plotted on the same charts.

The new charts consist of a new A4 chart and 6 PCHR charts on 3 pages both covering 32 weeks gestation to 4 years. In addition there is a new low birth weight chart (23 weeks gestation to 2 years) for very preterm (less than 32 weeks) and sick neonates. This chart has been designed for plotting growth measurements of preterm and low birth weight infants from 23 weeks gestation to the corrected age of two years, but is also suitable for term neonates or young infants requiring close monitoring.

Key new features of the new WHO/UK Growth Charts include:

- Separate preterm birth section
- No lines for 0 - 2 weeks of age
- Term births plotted at age 0
- De-emphasised 50th centile
- Length / height discontinuity at 2 years
- No 4-18 years section
- A new adult height predictor that allows approximate prediction of adult height (within 6 cm above or below)
- BMI lookup
- The head circumference curves are extended to two years

Children's weight gain patterns will appear different

- No 'dip' on chart at 2 weeks
- Low weight and centile falls much less

Availability and Use of the UK-WHO Growth Charts
UK-WHO chart Resources and Materials for NHS Staff and Experienced Chart Users are available from www.rcpch.ac.uk/Research/UK-WHO growth charts.

Capsule Endoscopy: Indications And Diagnostic Findings

David I Campbell, Consultant Paediatric Gastroenterologist, Sheffield Childrens Hospital

There is no doubt that the arrival of wireless capsule endoscopy (WCE) has provided a powerful diagnostic tool for the paediatric gastroenterologist.

Principal use of WCE to investigate occult gastrointestinal bleeding (OGB) has expanded to other conditions as software and hardware have pushed the picture resolution to new levels of detail.
This paper will consider where the limits lie with regards to the following:

1) Patient size (currently successful with a SB1 Given capsule at 12.5 kg or 2.5 years of age. Capsule retention is reported at patients below 10kg). Advent of MiroCam devices has lead to a 2mm reduction in capsule length (10%).
2) When to perform WCE for OGB (considering BSG guidance and how this dovetails with conventional endoscopic investigations).
3) More controversially when to use WCE for investigation of small bowel Crohn's disease.
4) Complications of WCE and use of patency devices.
5) Colon and ESO capsules.
6) Unique paediatric scenarios where WCE comes powerfully to the aid.

It is a personal view that the field of WCE is being lead by 2 main competitors (GIVEN Imaging and MiroCam from Intromedix), rapidly matching each other for specifications, yet not offering the same product. The 2 systems will be discussed.

New Developments
Exciting new developments of DCT domain software to analyse 3 D texture in 2 D using unique digital recognition, is opening up the way for ultra rapid detection of small bowel tumors, ulcers and polyposis syndromes.

Faster, smaller, clearer software linked to more sophisticated hardware (steerable devices, biopsy capable devices).

Protein losing gastropathy (Ménétrier's disease) associated with swine flu

Dr Sona Matthai, SpR, Sheffield

A previously healthy 10 year old boy presented with vomiting for three days with post prandial abdominal pain and no diarrhoea. He was moderately dehydrated (urea of 14.4 mmol/l) and was admitted for IV rehydration. Normal full blood count and inflammatory markers and a sodium of 132 mmol/l were noted

Post rehydration, periorbital oedema was noted and ascites was present confirmed on abdominal ultrasound. Albumin was 17g/l with normal coagulation and no proteinuria (protein creatinine ratio was normal at 12).A subsequent transaminitis was seen (ALT 92 and AST 105). Nose and throat swabs were positive for swine flu virus (H1N1 PCR).Albumin infusion resulted in diuresis and resolution of ascites but only a transient rise in albumin to 24g/l.In view of decreasing albumin an upper endoscopy was performed.

The appearances of stomach were typical of Ménétrier's disease (fig 1) and revealed hemorrhagic, oedematous macro rugae with relative antral sparing and was confirmed histologically (fig 2).H pylori were not found histologically. No viral inclusions were seen.Viral serology was negative for CMV, EBV and hepatitis panel.

He was commenced on omeprazole 20 mg twice daily resulting in an increase in albumin to 28-29 g/l.
He was discharged home and follow up endoscopy five weeks later showed regression of histological changes.

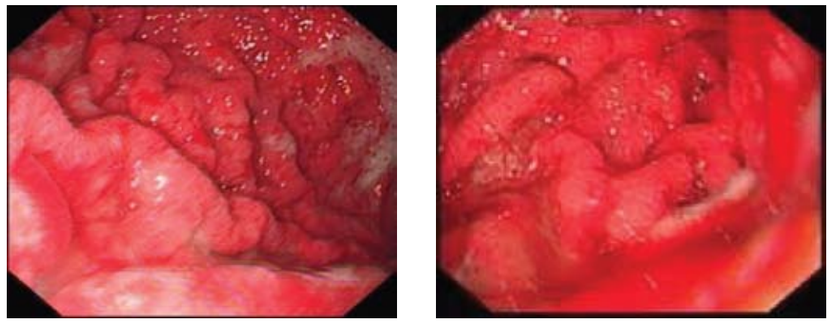

fig. 1

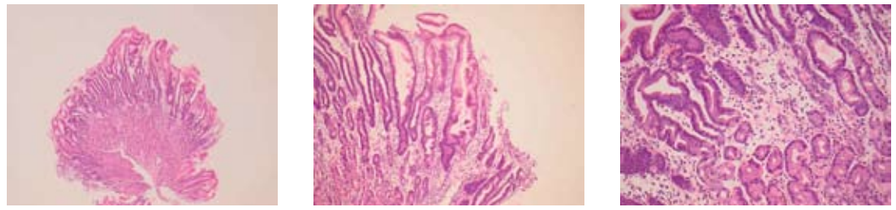

fig. 2
Gastric mucosa showing hyperplastic crypts with cyst formation. The lamina propria is oedematous with increased inflammatory cells.

Ménétrier's disease

This is a rare form of protein losing gastropathy in children.Association with CMV and H pylori has been previously reported (1).This is the first report of Ménétrier's disease associated with swine flu.

It is characterised by hypertrophy of the gastric mucosa resulting in increased mucin production, hypochlorhydria and loss of proteins across gastric mucosa with resultant hypoproteinemia.Unlike in adults who have chronic disease with predisposition to gastric carcinoma, it is a self limiting condition in most children.

Increased signalling of the epidermal growth factor receptor (EGFR) by transforming growth factor alpha (TGF a) has been implicated in the pathogenesis. Murine models have shown Ménétrier's changes when transforming growth factor alpha is over-expressed and immunostaining in children with Ménétrier's has demonstrated increased TGF a in gastric mucosa (2).

The most common symptoms are vomiting, abdominal pain and oedema. Diagnosis is by gastroscopy and histology. Most cases in children need only supportive treatment. However PPI therapy can expedite recovery and octreotide has also been effective in PPI refractory cases (3). Eradication of H Pylori and treatment of CMV with gancyclovir has also resulted in resolution of long standing disease in children (4). Recently monoclonal antibody to EGFR has been tried successfully in adults (5).

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ABSTRACTS FOR THURSDAY 28TH JANUARY 2010

Invited speakers' abstracts

Bacteria in the Pathogenesis of Inflammatory Bowel Disease

Jonathan Rhodes. University of Liverpool

There has been a longstanding suspicion that bacteria are involved in the pathogenesis of IBD. This has been strengthened by the recognition that all the genetic animal models of IBD require the presence of intestinal bacteria. Until recently there has been little evidence to implicate any specific bacteria. The field however has moved rapidly in the past few years. In Crohn's disease there are three areas where reproducible data have implicated bacteria:

(i) mucosa-associated E. coli – have now been shown by at least ten independent groups to be increased in Crohn's disease tissue. They have a characteristic "adherent and invasive" phenotype and are able to replicate inside macrophages – a process that is potentially linked to the defects in autophagy-related genes that have also been recently identified.
(ii) Reduction in the probiotic bacterium Faecalibacterium prausnitzii has been reproducibly reported in Crohn's and associated with increased risk of relapse after surgery.
(iii) Mycobacterium paratuberculosis has been found in low count numbers in a significant minority of Crohn's disease mucosal samples. Although unlikely to be acting as a direct pathogen, we have shown (a) that it carries the ASCA epitope and (b) that it releases a glycoconjugate (which expresses the ASCA epitope) that suppresses the ability of macrophages to kill bacteria, including intracellular E. coli. The recent linkage of NOD2 defects with lepromatous leprosy also adds weight to the plausibility of a pathogenic role for M paratb even though the beneficial effects of anti-TNFalpha treatments are strong evidence against a direct effect.

In ulcerative colitis there is no strong direct evidence for involvement of specific bacteria although we probably underestimate the extent to which relapse may be induced by conventional pathogens. Increased permeability of the mucosal barrier to bacterial components such as flagellar antigens seems likely to play a role in pathogenesis.

We have now reached the point where several bacteria-related hypotheses can be directly tested by targeted therapeutic interventions.

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Parenteral Iron replacement in Inflammatory Bowel Disease

Dr Stefanie Kulnigg, Vienna

Anemia is a common complication in IBD. At any given time point about 30% of patients suffer from anemia, most commonly iron deficiency anemia and anemia of chronic disease. Not only the continuous blood loss, but also low iron intake and iron uptake lead to a negative iron balance and therefore iron deficiency anemia. There are two routs of iron replacement, oral and intravenous. Oral iron therapy is cheap and application is comfortable for patients, however, the side effect rate is high and in animal models it has been shown, that there is an increase in disease activity and in colorectal cancer incidence. Intravenous iron is now used for more than 30 years in IBD. Different iron preparations are available. One of the oldest compounds is iron dextran. This dextran complex shows a slow release of iron, therefore high single doses, so called total-dose infusions, can be given. Iron gluconate is a labile and weak iron complex and directly releases iron into the plasma, therefore only low doses (max 125mg) can be given. It is mainly used in dialysis patients. In IBD patients most studies are available on iron sucrose. This iron complex is semi-labile, doses of 200-300mg can be used. A new intravenous iron preparation is ferric carboxymaltose, which is stable as iron dextran, but has a lower risk of anaphylactic reactions. Doses up to 1000mg can be infused within 15 minutes. In pediatric IBD, only two studies, both with iron dextran, are available, the most recent from 2002. In conclusion, iron deficiency anemia is very common and should be diagnosed and treated early, as it has great impact on quality of life and growth. Iron deficiency can be treated fast with parenteral iron preparations, which are considered safe and effective.

Magnetic resonance imaging of the abdomen in Paediatric Inflammatory Bowel disease - Is it the end of the small bowel follow through?

Dr Gurdeep Mann, Consultant Paediatric Radiologist, Liverpool

The barium follow through has long been the diagnostic imaging modality of choice to depict and monitor small bowel involvement in paediatric inflammatory bowel disease. There has been recent interest in pursuing non-ionising diagnostic imaging techniques such as MRI to avoid radiation exposure and in order to provide complete luminal, mural and extramural evaluation of the bowel. Paediatric practice eventually mimics adult practice where such a shift in emphasis is already well under way. This lecture will try to answer some of the many questions we must ask before we can even begin to think about jettisoning time honoured fluoroscopic techniques.

Anti Reflux surgery – Indications, procedures and Outcome.

Mr Matthew Jones, Consultant Paediatric Surgeon, Liverpool

This talk is a brief overview of the surgical treatment of reflux disease at a tertiary referral centre serving a population of roughly 4.5 million inhabitants. The talk covers various aspects of the surgical treatment of reflux disease, with particular emphasis on patient selection, case mix, rationale of treatment, surgical options and medium-term outcome. To the best of our knowledge we are the only centre within this sub-region offering such a service.

Laryngopharyngeal reflux in Children,Diagnosis, investigation and Management.

Dr Tobias Wenzl, Aachen, Germany

Gastroesophageal reflux (GER) may be a symptom of GER disease (GERD). GER-associated symptoms can be esophageal or supraesophageal (laryngoesophageal), e.g. affecting the airways.

The diagnostic tools available for GER, GERD and their associated esophageal and supraesophageal symptoms include esophageal pH monitoring, combined multiple intraluminal impedance (MII) and pH measurement (MII-pH), (high resolution) manometry and endoscopy with biopsies. Barium contrast studies, nuclear scintigraphy and esophagogastric ultrasound are not recommended for the routine evaluation of GERD in children. Test on ear, lung and esophageal fluids and bile are available, but no controlled studies have yet proven the benefit of these test in clinical routine.

GER treatment initially consists of parental advice and reassurance, and "lifestyle changes" (e.g. thickened formula). The pharmacologic therapies available include gastric acid buffering agents, mucosal surface barriers, alginate, gastric anti-secretory agents, i.e. histamine-2 receptor antagonists (H2RAs) and proton pump inhibitors (PPIs), prokinetic therapy, and surgical intervention.

Among the laryngopharyngeal symptoms in children with suspected GERD are predominantly respiratory / pulmonary / ENT symptoms such as infant apneas or apparent life threatening events (ALTE), upper airway or reactive airway disease, recurrent pneumonia and dental erosions, or behavioral / neurological symptoms such as unexplained crying or distressed behavior and dystonic (head) movements (Sandifer´s syndrom and BIRDY = bolus induced reflux dystonia).

Especially in extra-esophageal symptoms of GERD the concept of acid (pH < 4) reflux as the main cause and its detection with pH monitoring has been shown to be insufficient, as patients with normal pH measurements still may have weakly acidic (pH 4 - 7) or non-acid (pH > 7) reflux causing their symptoms.

Multiple intraluminal impedance studies (MII) combined with pH measurement (MII-pH) have replaced simple pH-metry as a diagnostic tool to evaluate retrograde bolus movements in the esophagus of children of all age groups. With the advent of MII-pH many pediatric intervention studies have be newly performed using the combination of intraesophageal impedance and pH and have delivered new and partly totally different results and insights into the influence of GER on laryngopharyngeal symptoms.

One of the most important issues when dealing with supraesophageal / laryngopharyngeal symptoms of GER (in children) is the detection and verification of an at least temporal association between the symptoms and the reflux. This may be difficult in rare (not occuring at least a few times in 24 hours) or ongoing, i.e. continuous symptoms or conditions, e.g. asthma refractory to standard asthma treatment. In discontinuous symptoms, e.g. apneas or oxygen desaturations, the symptom under investigation should be recorded automatically and time-synchronized with the MII-pH recording, as opposed to relying on a diary taken by the parents or the patient. Prototypes of new recording systems, combining synchronous registration of GER and symptoms, have been validated in clinical studies and will be available soon.

Eosinophilic oesophagitis - recent advances in management

Mike Thomson

FIBROPOLYCYSTIC DISEASES OF THE LIVER
Dr. P.J. McKiernan

Fibropolycystic diseases of the liver are hereditary disorders characterised by some degree of intrahepatic bile duct dilatation, cysts lined by biliary epithelium and peri-portal fibrosis. It is most commonly associated with renal cystic disease and the basic histological lesion is a ductal plate malformation. The varying clinical phenotype depends subsequently on genotype, the size of the portal tracts affected by the ductal plate malformation and the relevant balance of associated hepatic fibrosis and bile duct dilatation.

Three hepatic phenotypes are:

(i) Congenital hepatic fibrosis
(ii) Caroli's Syndrome
(iii) Polycystic liver disease

These diseases usually occur in the setting of hepato-renal fibropolycystic disease. The pathogenesis of these disorders is related to primary ciliary dysfunction, hence the term ciliopathies. Primary cilia are widely expressed in developing cells. Stimulation of these regulates cell growth and in the liver they are only expressed in cholangiocytes. The primary genetic defects are due to mutations in cilial expressed proteins which result in primary ciliary dysfunction.

Congenital hepatic fibrosis and Caroli's Syndrome are most commonly associated with auto-somal recessive polycystic kidney disease (ARPKD). ARPKD has an incidence of 1:20000 and is due to mutations in the PKHD1 gene. The gene product of this is polycystin which is expressed in primary cilia in kidney, bile ducts and other tissues. The most common clinical presentation is as a result of the renal involvement. Hepatic presentation can be an incidental finding, associated with abdominal pain but most commonly with signs of portal hypertension (25%) or cholangitis (4-10%). Clinical examination usually reveals firm hepatomegaly, hepatosplenomegaly and nephromegaly. Laboratory investigations may show evidence of hypersplenism and impairment of renal function but liver biochemistry is usually normal in the absence of cholangitis. Abdominal ultrasound is usually diagnostic showing a hyperechoic liver appearance and evidence of portal hypertension.

Caroli's Syndrome is recognised by irregular biliary dilatation (this may be best demonstrated with MRI). Renal ultrasound reveals symmetrical, enlarged, hyperechoic kidneys with loss of corticomedullary distinction. Macroscopic cysts may be visible. Liver histology will demonstrate the classical ductal plate malformation with varying degrees of fibrosis. Liver biopsy is, however, rarely necessary for diagnostic purposes.

Management: Initial management is supportive and educational making sure the family receive genetic advice. Cholangitis should be managed aggressively with long term prophylactic antibiotics if this is recurrent. Portal hypertension should be managed along conventional lines. If hypertension is present beta blocker treatment may have a dual role in reducing portal pressure. If variceal bleeding occurs endoscopic secondary prophylaxis with variceal banding is sufficient for the great majority. A porto-systemic shunt should be considered for those failing endoscopic treatment. Hepatic reserve is usually good and isolated liver transplantation is rarely necessary.

GLIS3 mutations
Management of the liver disease will also be dependent on the degree of renal dysfunction. Survival following isolated renal transplantation for ARPKD is worse than for other indications. A major source of morbidity and mortality in this group is related to hepatobiliary disease and sepsis. Where renal function is preserved hepatic management is unchanged. If end stage renal failure develops haemodialysis is preferred to peritoneal dialysis. Where transplantation is being considered the presence of cholangitis or severe bile duct abnormalities are indications for combined liver/kidney transplantation. Uncomplicated portal hypertension will not in itself be a contra-indication to isolated renal transplant.

Congenital hepatic fibrosis is also associated with a wide range of renal cystic disease (Table 1). In fact it is probable that a truly isolated congenital hepatic fibrosis/Caroli's Syndrome does not exist as a primary entity.

Polycystic liver disease (PLD) is overall commoner than ARPKD but is rarely a clinical problem in childhood. PLD usually occurs in the setting of autosomal dominant polycystic renal disease but a genetically a distinct subtype of isolated PLD exists.

Symptoms are rare before puberty but up to 20% will be hypertensive in childhood. Symptoms relate to abdominal mass effects and cyst complications. Liver function tests are usually normal. Diagnosis is by imaging showing multiple hepatic cysts, usually combined with evidence of autosomal dominant polycystic kidney disease.

Management should be individualised depending on the clinical features

Summary:
Fibropolycystic diseases of the liver are a complex group of genetic conditions which are usually associated with renal cystic disease. These will require lifelong supervision and management should be titrated to the individual clinical features.

Table 1.

Disorders associated with congenital hepatic fibrosis / Caroli's syndrome
Autosomal recessive polycystic kidney disease
Autosomal dominant polycystic kidney disease
Nephronophthisis
Medullary cystic kidney disease
Joubert syndrome (COACH syndrome),
Ivemark syndrome,
Jeune syndrome,
Bardet-Biedl syndrome ,
Ellis- Van Creveld syndrome,
Meckel-Gruber syndrome,
Carbohydrate deficient glycoprotein syndrome type 1
GLIS3 mutations

Chronic Pancreatitis: Results from the EUROPAC study:

Mr Bill Greenhalf, Liverpool

Hereditary pancreatitis (HP) is a rare autosomal dominant condition leading to acute pancreatitis in childhood, which frequently progresses to chronic pancreatitis and has a 40% lifetime risk (to 70 years) for the development of pancreatic cancer. The Royal Liverpool University Hospital is the host for the European Registry of Hereditary Pancreatitis and Familial Pancreatic Cancer (EUROPAC), which is the largest registry of patients with HP in Europe. Approximately 80% of patients on the registry have a mutation at either codon 122 or codon 29 in the PRSS1 gene; PRSS1 encodes the cationic trypsinogen enzyme. Age of onset, complications (endocrine/exocrine failure) and pancreatic cancer risk are variable even in patients with known mutations. Indeed, a substantial number of patients (>20%) carrying the mutation do not develop pancreatitis. In addition we have identified many patients with apparently sporadic chronic pancreatitis which is otherwise idiopathic who have genetic variants in the SPINK1 and a small number of patients with apparently sporadic disease who carry the p.A16V PRSS1 mutation.

In this presentation I will discuss the current understanding of the factors (genetic and environmental) that influence the penetrance of HP, which should in turn influence patient management. The significance of this to the pathology of chronic pancreatitis of other aetiologies will be considered. The EUROPAC experience of cancer screening will also be discussed, including a discussion of which subsets of patients with hereditary pancreatitis are most at risk focussing on the influence of diabetes and smoking in this group of patients.

Quality Indicators with relation to Paediatric Gastroenterology

John Smith of Civil Eyes Research will discuss some possible indicators based upon performance data drawn from fourteen specialist paediatric hospitals across the UK using published data drawn from Hospital Episode Statistics (HES) for 2008/09.

The analysis will be made using OPCS procedure based information and will refer to the impacts of HRG version 4. In particular John will discuss colonoscopy, oesophago-gastroduodenoscopy and percutaneous endoscopic gastrostomy as sentinel procedures.

John will provide a backcloth of performance in a variety of settings. He will review the mode of attendance, age of patients, length of stay and the local access to services.

ABSTRACTS FOR THURSDAY 28TH JANUARY 2010

Plenary abstracts

Azathioprine use in children with inflammatory bowel disease: A survey of clinical practice in UK
Himadri Chakraborty, Ipswich Hospital NHS Trust, Mary-Anne Morris, Norfolk and Norwich University Hospital

Background and Aims:
UK data is lacking on the existing practice of use of azathioprine as Immunosuppressant in children with IBD, British Society of Paediatric Gastroenterology, Hepatology and Nutrition (BSPGHAN) published guidelines for IBD patients in October 2008. This survey determined whether contemporaneous UK practice differ significantly from the guideline.

Methods:
Questionnaires were distributed (e-mail and telephone) to 28 UK Paediatric Gastroenterology Units between November to December 2008 . We enquired about azathioprine starting dose, presence of any local guideline, testing of thiopurine methyltransferase enzyme (TPMT), ranges deemed acceptable for lymphocyte and neutrophil counts, local blood monitoring policies and responsibility for patient monitoring.

Results:
24/28 (86%) hospitals responded of which 54%(13) of the units use a lower than recommended starting dose (1-<2mg/kg). 54% follow a specific guideline. Target lymphocyte count to assess treatment efficacy was 0.5-1×109/L in 54% of centres, lower than recommended 1-2×109/L. 79% of Units accept a median lower limit of 1×109/L neutrophil to monitor myelosuppression . BSPGHAN makes no recommendations. Contemporary blood monitoring schedule differed from the BSPGHN recommendations in 79%,67% and 50% of units for the first,second and ≥ third month respectively. Shared care pathways, though recommended by BSPGHN, is only followed in 50% of units. 91% of units were consistent with BSPGHAN regarding TPMT assay before commencement.

Conclusion:
Current practice in majority of units in UK differ from BSPGHAN recommendations regarding starting dose, treatment efficacy and safety assessment, blood monitoring strategy and responsibility sharing.

Restorative Proctocolectomy in Paediatric Ulcerative Colitis - A Single UK Centre Experience

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Background:
The majority of children with ulcerative colitis (UC) have pan-colonic distribution and 25% undergo colectomy in the paediatric age range. Restorative proctocolectomy (RP) is the gold standard surgical therapy but concerns about the magnitude of the procedure mean that children are often offered only colectomy and ileostomy.

Aim:
To describe the disease characteristics, prior medical therapy and surgical outcomes of children with UC who received RP over a 10 year period, 1999 – 2009.

Methods:
Retrospective case note and database review of the clinical features, medical management prior to RP and operative complications and pouch function after RP. Based on clinical features, the indications for surgery were categorised as either acute severe colitis/failure to induce remission or a failure to maintain remission. Data are expressed as median (range). Statistical comparison used Wilcoxon signed rank test, χ^2 test and binary logistic regression.

Results:
36 children, 19 boys were diagnosed at age 11.0 yr (2 to 16) and underwent RP at age 13.6yr (5 to 19). 23 had a 2 stage and 13 a 3 stage procedure. Follow up post RP is 5.6yr (0.7 to 10). Between diagnosis and colectomy haemoglobin concentration improved (10.7g/dl vs. 11.5, p = 0.005), while CRP and pediatric ulcerative colitis activity index (PUCAI) deteriorated (CRP: 19 vs. 34 p = 0.03. PUCAI: 60 (7) vs. 67 (6) p <0.001). Platelet count, albumin, stool frequency, night stooling and height and weight z-scores did not change. Indications for surgery were: failure to maintain remission 27, inability to induce remission 6 and toxic dilatation 3.

	Acute severe colitis/Failure to induce remission n=9	Failure to maintain remission n=27
Time to colectomy (yrs)	0.6 (0.1 to 5)	2.3 (1 to 12)
Number(%) pancolitis	100%	80%
Number(%) iv c/s	87%	48%
Steroid free time period (months)	0 (0-22)	6.2 (1.1-36)
Number (%) AZA / 6-MP	78%	100%
Number (%) Cyclo / IFX	28%	20%

At RP the median steroid dose was 1.2mg/kg and azathioprine was 2.2mg/kg.

15 patients experienced a complication following surgery and 3 have a permanent ileostomy. The probability of any complication or permanent ileostomy was not related to either age at diagnosis, time to colectomy, age at colectomy or PUCAI. Following RP median stool frequency is 5; 35% have nocturnal stool and 21% have experienced pouchitis.

Conclusions:
Children treated with RP show moderate PUCAI (60) at diagnosis which deteriorated to severe (67) by the time of colectomy 2.7 years later. In the failure to maintain remission group only 20 % of a median time of 2.3 years from diagnosis to RP was corticosteroid free despite the use of steroid sparing agents. The probability of any complication or permanent morbidity following RP in childhood is lower than in published series of adults and is not related to the age at surgery. Children with UC who fail to achieve sustained remission should be offered RP as the procedure of choice.

Wide variation in diagnostic criteria and management approach to Eosinophilic Oesophagitis in the UK: Results of a BSPGHAN survey

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Background:
Eosinophilic Oesophagitis (EO) is a recognised cause of upper gastrointestinal morbidity. It is a histologically defined condition with a strong association with food allergy. Diagnostic criteria and management protocols have been published (mostly from North America) but have not yet been widely adopted in the UK. This could lead to wide variation in diagnostic and management approaches. To highlight this problem in a paediatric population, a survey was undertaken looking at the variation in diagnostic criteria and management of EO across paediatric gastroenterology (GI) centres in UK.

Aim:
To document variation in practice across various paediatric GI centres with regards to diagnostic criteria, investigation and management of EO in children in the UK.

Methodology:
A questionnaire was designed focusing on various aspects of EO such as incidence, diagnostic criteria used, how the biopsy was performed, types of allergy testing conducted and treatment. This questionnaire was emailed to all BSPGHAN members in September 2009.

Results:
Incidence: 12 paediatric GI centres returned the survey questionnaire. The self reported median (range) populations served by these centres were 2.25 (0.3 to 5.2) million. They performed a median of 250 (70 to 400) upper GI diagnostic endoscopies/ yr. The number of new EO cases diagnosed varied between 0.38 to 6.6/ million/yr (median of 3.2/million/yr).
Diagnostic criteria: No centre always performed upper GI endoscopies while on proton pump inhibitors (PPI), (2 centres usually on PPI). With regards the exclusion of gastro-oesophageal reflux only 2(16%) always performed pH/ impedance study, while the rest either occasionally or never. 7(58%) centres use a cut-off of 20 or more eosinophils per high power field as diagnostic of EO while 3(26%) used ≥ 15, and 16% (n=2) used ≥ 10. There is wide variation in practice with regards to site and number of oesophageal biopsies. 16 %(n=2) of centres performed biopsies only at the lower oesophagus, 16%(n=2) lower & mid oesophagus, and 6%(n=2) lower, mid & proximal oesophagus (50%(n=6) did not comment). The number of biopsies varied from 1 to 5. Only one centre reported that 10 high power fields would be analysed (the rest of centres did not respond / were not sure). 10/12 undertook investigation of allergy; RAST (75%) skin prick test (25%).

Treatment:
Topical steroids are used as first line in management of EO by most centres (7, 58%), 4(33%) used dietary restriction, 2(16%) used Leukotriene receptor antagonist and 2(16%) antihistamine as first line (some centres use combination). One centre just uses PPI. The choice of topical steroids were inhaled fluticasone which would be swallowed in 7(58%) centres, 2(16%) used inhaled budesonide and 2(16%) used budesonide respules to be swallowed. In severe cases 2 centres would consider oral prednisolone.
Conclusion: The variation in incidence of EO across the UK may well be due in part to differences in diagnostic criteria used. This survey high lights the wide variation in practice with regards to investigation, diagnosis and management of EO. A consensus process needs to be undertaken to develop unified diagnostic and management protocols for EO.

Rationalisation of services has failed to improve outcomes for biliary atresia in Scotland. A report on behalf of the Scottish Paediatric Gastroenterology Hepatology and Nutrition Group (SPGHANG)

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4.Department of Paediatrics, Ninewells Hospital Dundee

Background:
Extra hepatic biliary atresia (EHBA) is a rare, ultimately fatal disease of the newborn for which outcome can be improved by early diagnosis and subsequent Kasai portoenterostomy. Previously Kasai procedures were routinely performed at Childrens Hospitals in Glasgow, Edinburgh and Aberdeen. Following publication of data suggesting that outcomes were superior in those centres performing >5 Kasai procedures a year1, services were rationalised to three supraregional centres in England and Wales. No such directive exists for Scotland; however in 2002 an informal agreement was made within the SPGHANG centres to aim to refer all new cases of BA to one of the three English centres.

Aim:
To examine outcomes for patients with EHBA in Scotland since 2002 and compare this with the historical Royal Hospital for Sick Children Glasgow (RHSC)2 data from the orginal paper describing outcome from centres perfoming >5 Kasai procedures a year (Group A)1.

Subjects and methods:
A review of case-notes of all children diagnosed with BA in Scotland Jan 2002-Oct 2009 was conducted from databases of SPGHANG units. Patient demographics and data regarding presentation, investigation, co-morbidity timing of Kasai, and Kasai centre. In addition outcomes in terms of clearance of jaundice, transplant free survival (TFS) and survival were recorded for 6 months, 1 year and 2 years post Kasai. Clearance of jaundice was defined as conjugated bilirubin <20umol/l. Chi-squared test were performed on Minitab (Version 13) and p≤0.05 was taken as significant

Results:
26 children with BA were identified. A Kasai operation was performed in 24 (92%) infants and 2 (8%) had primary transplantation. 3 children had Kasai procedures in Edinburgh, and 1 patient moved to Scotland post-Kasai so were excluded from further analysis. 23 were referred to supraregional service and 21 had a Kasai procedure performed. Of these 22 children, 18 have reached 2 year follow up with 16 (89%) children had survived, 11 following liver transplantation and 6 with their native liver. 1 child died following transplantation. Although time to presentation has risen between RHSC 1987-2000 and SPGHANG 2009 (40 vs 49days) this was not statistically significant (Mann-Whitney p=0.286). Outcomes from the three groups are compared in table 1.

Group	Cases	Median Age presentation to GI (range)	Median Age Kasai (range)	Clearance jaundice (%)	1yr TFS (%)	2y TFS (%)
RHSC 1987-2000	20	40	58	13/20 (65)	16/20 (80)	13/20 (65)
Group A	57	44	53	33/53 (62)	44/57 (77)	35/57 (61)*
SPGHANG 2002-2009	22	49	60	9/20 (45)	10/20 (50)	6/18 (33)*

*Table 1: Outcome for EHBA in RHSC, Group A and SPGHANG 2002-2009 * p=0.03*

Summary and Conclusion:
Centralisation of Kasai services within the SPGHANG group has failed to significantly improve outcomes for Scottish children presenting with EHBA. Despite now having surgery performed at Group A centres, 2yr TFS is significantly lower in the Scottish population than previously published UK data and historical unpublished Scottish data. Delay in presentation appears to be the key factor but additional delay from tertiary centre to surgery needs minimised to facilitate optimal outcome. Improved communication between primary care, general paediatricians and gastroenterology centres through regional and managed clinical networks (MCN) may improve future outcomes for Scottish children with BA.

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2. Ling S, RHSC Glasgow personal communication 2002
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Non-invasive biomarkers and paediatric NAFLD: new methods to predict disease and stratify severity

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Introduction:
With the alarming growth in prevalence of paediatric non-alcoholic fatty liver disease (NAFLD), there is a real need for non-invasive methods of stratifying disease severity and following disease progression / response to treatment. The aim of this study was to evaluate a combination of serum biomarkers as measures of disease activity and severity in children with NAFLD.

Methods:
Forty five children who were biopsy proven to have NAFLD were enrolled in the study. Anthropometric, biochemical and radiological data were collected and serum was stored at -80˚C. CK18 M30 fragments, hyaluronic acid, leptin and adiponectin were measured in serum using specific ELISAs and high sensitivity C-reactive protein (hsCRP) with an automated colorimetric assay. Liver biopsies were scored by a single pathologist. Data was analysed using SPSS v17.0.

Results:
Median age was 12.7 years (IQR 9.8–14.2), 25 (55%) were male. Median BMI z-score was 1.66 (1.22-2.07). CK18 M30 levels were significantly higher in patients with NAFLD as compared to age matched healthy controls, median; 288 IU/L (202-494) versus 172 IU/L (146-205) respectively (p<0.001). CK18 M30 levels could also distinguish between significant steatohepatitis (NAS≥5), median; 347 IU/l (IQR 258 – 509) and simple steatosis (NAS<3), median; 191 IU/l (IQR 167 – 197), (p=0.006). Significant fibrosis (≥F2) could be differentiated from no / minimal fibrosis (<F2) using CK18 M30, median; 393 IU/l (225-533) vs. 243 IU/l (190-317), (p=0.03). Leptin was useful in distinguishing fibrosis <F2 from ≥F2 (21.9 ng/ml (16.6 – 83.6) versus 55.9 ng/ml (32.6-77)) (p=0.03). Neither adiponectin, hyaluronic acid nor hsCRP levels reached statistical significance in predicting NAFLD versus NASH, nor significant fibrosis (≥F2) from no fibrosis (<F2).

Conclusion:
This study combines the use of markers for different processes in the development of NASH. Serum biomarkers, especially CK18 M30 fragments, are of potential use in stratifying disease severity and thus in longitudinal monitoring of disease progression in paediatric NAFLD.

Trial of a micronutrient rich, high fibre, low energy density enteral feeding formula for gastrostomy feeding disabled children

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Introduction:
Gastrostomy feeding in the severely disabled child with oral-motor dysfunction (e.g. cerebral palsy) improves overall weight gain but is often associated with excess deposition of body fat. Most feeds used for enteral nutrition via gastrostomy are high energy proprietary feeds which are not formulated to meet the nutritional needs of immobile children with severe motor deficits who have low energy expenditure. Enteral feeds with a more appropriate energy composition are needed for gastrostomy feeding in children with cerebral palsy but no studies have been undertaken on this is topic to date. Our aim was assess the effect on weight gain and body composition of a low-energy micronutrient replete enteral feed.

Aim:
We prospectively studied the effects of a low energy density (0.75kcals/ml), micronutrient rich, high fibre feed on the growth, body composition and micronutrient status of disabled children fed via gastrostomy.

Subjects and Methods:
We studied 14 children (7 boys) with severe Cerebral Palsy (GMFCS IV or V) who were gastrostomy fed. Median decimal age 2.088 years, 13 had spastic quadriplegic cerebral palsy and 1 left hemiplegia. Subjects had all study assessments prior to commencing the low energy feed (baseline) and again after 6 months G-tube feeding of the low energy formula. Body composition and energy expenditure were measured using Doubly Labelled Water (DLW) O18 dilution. Anthropometry was used to measure growth, and a general health questionnaire administered for information on GI symptoms and medication use. Micronutrient status was also measured before commencing the low energy feed and was repeated 6 months later.
The feed was given at energy levels no greater than 75% of the EAR for age and was adjusted where appropriate following the results of their energy expenditure dilution tests.

Results:
Continuous data are presented as median and interquartile range as data is non-parametric (Shapiro Wilk test for weight p=0.0118).

Growth and body composition data is shown in table 1.There was a significant increase in weight, mid upper arm circumference (MUAC) and lower leg length (LLL) among the subjects across the course of the 6 month study. There was no significant corresponding increase in fat mass or fat percentage as had been observed by us in a previous study.

	Number (N)	Baseline median (IQ range)	N	6 months median (IQ range)	Wilcoxon p value N=6
Weight (kg)	14	12.09 (7.04)	8	14.3 (7.06)	0.012
MUAC (cm)	14	15.0 (3.84)	8	16.09 (4.68)	0.043
LLL (cm)	14	15.73 (9.12)	8	17.89 (9.5)	0.012
Fat mass DLW (kg)	11	3.11 (4.67)	6	4.71 (2.53)	0.347
Fat % DLW	11	28.27 (26.69)	6	31.85 (13.84)	0.173

Table 1

The median percentage intake of the estimated average requirements (EAR) for energy (kilocalories) was 43% (23.0) at the beginning of the study and 48.8 (8.5) after 6 months on the low energy feed.

Summary and Conclusion:
We have shown that severely disabled children who are fed a low energy, micronutrient complete, high fibre feed continue to grow at energy intakes below 75% of the EAR. Weight gain was not associated with a disproportionate rise in fat mass or fat percentage. There was also continued significant MUAC and linear growth. The low-energy feed is particularly suited to this group of children with cerebral palsy as their low energy requirements can be delivered in a higher volume thus maintaining satiety while ensuring adequate growth and nutrition.

ABSTRACTS FOR THURSDAY 28TH JANUARY 2010

Poster abstracts

A rare case of a blue rubber bleb naevus syndrome with a supraglottic haemangioma: Delayed diagnosis of gastrointestinal bleeding and treatment experience with Thalidomide

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Introduction/Background:
The classification and nomenclature of angiodysplasia including vascular anomalies of the gut have historically been inconsistent and confusing. Blue rubber bleb naevus syndrome (BRBNS) is a rare congenital disorder presenting with multifocal vascular malformations most prominent in the skin, soft tissues, and gastrointestinal tract (GI), however may occur in any tissue. Patients with BRBNS can develop severe anaemia due to chronic GI bleeding, and may require life-long supportive treatment including iron and blood transfusions. Currently, there is no consensus on the definitive medical or surgical management of these difficult cases from the limited clinical literature.

Aim:
We report a case of BRBNS who presented initially at birth with a supraglottic haemangioma(SG-H), in whom there was delayed recognition of refractory small bowel GI bleeding from multiple vascular lesions. We also describe our experience with use of thalidomide in the management of this case.

Subjects and methods:
Our patient was born floppy and had a stridor at birth, needing intubation and ventilation. He had a large SG-H causing obstructive symptoms for which he required tracheostomy in the neonatal period. He was discharged home with a tracheostomy in situ and over several years he underwent numerous sclerotherapy treatments for the SG-H by ENT/interventional radiology and maintained a relatively stable haemoglobin. However, subsequently from 7 years of age, there was a period of instability with recurrent anaemia (Hb as low as 4.4 g/dl),initially attributed to intermittent blood loss from his SG-H which was only partially responsive to local sclerotherapy. During this period he required a variable frequency (up to 3 weekly) infusions of iron or blood in addition to oral iron therapy. At 11 years of age he was referred to the Paediatric Gastroenterology service to investigate the possibility of gastrointestinal source of bleeding, as anaemia persisted despite sclerotherapy appearing to show control of bleeding from the SG-H lesions. Several investigations were done including imaging (barium meal and follow through, abdominal MRI),oesophago-gastro-duodenoscopy, colonoscopy, and capsule endoscopy. Capsule endoscopy identified numerous vascular lesions with areas of active bleeding throughout the small intestine. The capsule endoscopic appearances were suggestive of blue rubber bleb type naevi. In addition to the GI lesions and SG-H, our patient also had two small penile vascular lesions, however unusually there were no generalised cutaneous lesions. At that time, given the difficulties in accessibility of the small bowel, extent and multitude of vascular lesions and the potential for recurrence of bleeding, invasive and higher risk options such as, bowel resection or push enteroscopy with use of luminal sclerotherapy were not considered feasible. Consequently, medical options were carefully considered and decision made to treat with thalidomide on the basis of its anti-angiogenic properties and continue supportive care.

Results:
Prior to the thalidomide therapy, our patient required blood transfusions and iron infusions every 3-4 weeks. After starting thalidomide, no transfusion was needed for a period of 8 months suggesting a strong therapeutic effect. Thalidomide was however discontinued thereafter following abnormal nerve conduction studies and concerns about side effects. Four months later, as the nerve conduction studies improved, thalidomide was restarted at half the initial dose. However, on restarting thalidomide on this occasion and even after increasing to the original effective dose his haemoglobin continued to remain low and there was no improvement in terms of transfusion requirements. We also noted possible side effects of thalidomide including muscle cramps, paresthesias, bradycardia, mood disturbances, abdominal pain and constipation. Thalidomide was eventually stopped in view of lack of therapeutic response and the possible side effects.

Summary and Conclusion:
BRBNS is a very rare condition with only about 150 cases reported world wide. The vascular malformations in BRBNS can occur in any tissue; however associated supraglottic lesions are not described in the literature The atypical presentation in our patient resulted in delay in considering the possibility of GI bleeding as a cause of his persistent anaemia. To our knowledge, this is the first reported case of BRBNS in association with a SG-H. Effective treatment options are limited in tackling the very challenging problem of recurrent GI bleeding from small bowel lesions in patients with BRBNS. Treatment with thalidomide for its anti-angiogenic effect should be considered on an individual patient basis. In our patient, very good initial therapeutic response to thalidomide was not sustained and also, use was constrained by adverse effects.

A nutritional review of gastrostomy insertion in an Irish Paediatric Hospital

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Introduction:
Good nutrition in childhood is important to ensure children grow and develop to their full potential. Children need gastrostomy tubes for a number of reasons. These include children with swallowing problems and those who are unable to achieve adequate oral intake to grow properly and meet their nutritional needs.

Aim:
The aim of this study was to review gastrostomy tube insertion at the Children's University Hospital, Dublin, Ireland with a view to creating guidelines for the management of such patients for the future.

Methods:
84 children were identified as having a primary gastrostomy inserted between January 2003 and December 2007 in the Children's University Hospital. Patients' medical, dietetic and nursing notes were retrospectively reviewed. Relevant data was recorded on patients prior to tube insertion and for 1 year post insertion. Data was entered into a Microsoft EXCEL database and converted to SPSS 15.0 for statistical analysis.

Results:
The mean age at insertion was 5.1 years (Range: Day 1-19.3 years). The most common underlying diagnoses were cerebral palsy (32 %), other neurological disorders (15 %), cystic fibrosis (14 %), metabolic disorders (8 %) and chromosomal abnormalities (12 %).

61 % of patients were tube fed via a nasogastric tube for more than 2 months prior to gastrostomy insertion. A further 11 % of children were nasogastrically fed for less than 2 months prior to insertion. In those that were tube fed prior to insertion the mean duration of tube feeding was 59 weeks.

Weight z scores were significantly higher in those that had been tube fed for > 2 months prior to insertion versus the group that were not tube fed/tube fed for less than 2 months prior to insertion (p = 0.022).

45 % of patients had their gastrostomies placed endoscopically, while the remaining 55 % of patients were placed via open surgery. Nissen's fundoplication procedure was performed in 27 patients (32 %) at the time of insertion.

No feeding protocol is in place in the hospital at present. There are no definite practices on feeding post gastrostomy insertion. Fasting times post insertion, initial rates and time until full feeding is reached varies greatly in patients post gastrostomy insertion.

Weight z scores were significantly higher at follow up (p=0.000).

Assessment Of Support Services For Paediatric Home Enteral Tube Feeding

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Background:
There has been a steady rise in Paediatric Home Enteral Tube Feeding (HETF). There is a lack of information about how tube feeding would affect the child and family. With prolonged survival, there is an increasing demand on healthcare resources and a concern of lack of support for families to deal with problems arising from HETF.

Aim:
To monitor the quality of HETF services in a large regional district and determine patient satisfaction of service delivery.

Subjects & Methods:
Survey questionnaire consisting of open and closed ended questions was sent to 64 patients on HETF with a reminder sent after 1 month. Questions were based on the Scottish NHS- best practice statement 2008. Case notes were reviewed to document and compare demographic data with 2007 UK data.

Results:
Sixty-four patients (1.2% of UK HETF patients) were managed on HETF in Basildon and Thurrock region in Eastern UK. The age range was 1.2-17.2years (mean 7.0 years) with 8% in the adolescent group. Forty-seven percent had a neurological diagnosis. Seventy-five percent of children on HETF in Basildon had gastrostomy whereas only 50% of the UK had a gastrostomy.

The response rate on the survey questionnaire was 50%.The planning and coordination of care at the time of initial hospital discharge was rated to be satisfactory by 75% of responders (Figure1). Ninety percent were satisfied with services for home delivery of equipment, feeds and information given. Fifty-seven percent were not satisfied with the support and training at school. Only 28% of parents were given advice regarding dental care. All had follow-up by the community team and 90% had a yearly follow up by the dietician and the local hospital. Overall 78% reported their experiences were better or equal to expectations and only 9% found the quality of services poor (Figure 2). The common suggestions from parents were to improve communication between tertiary and district general hospitals, provide more spare equipment like gastrostomy tubes, provide information regarding support groups and improve training of hospital and school staff dealing with problems arising from HETF.

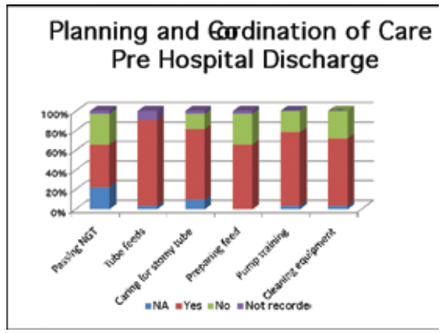

Figure 1

Figure 2

Summary:
There are a higher proportion of patients receiving HETF through gastrostomy when compared to the UK data which would relate to better access to surgical facilities. There is also a high proportion of adolescent patients emphasizing the need to consider transition to adult services. Majority of parents are satisfied with pre discharge and home delivery services, however training and support at schools needs to improve. Overall, most parents were satisfied with the quality of services and their experience with the team. Conclusion: This study is one of the first efforts to systematically audit paediatric HETF services in a large regional area and will help managers and policymakers to reflect on improving the paediatric HETF services in the future.

Children presenting with Minor trauma Fractures - do they need screening for Coeliac disease and associated abnormalities in bone mineralisation?

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Introduction:
Abnormalities in bone mineralisation and potential consequences of osteopenia with increased fracture risk are a well recognised complication of untreated Coeliac disease (CD) presenting in adults. However, in children, whilst abnormalities in bone mineralisation have been described, clinical presentation with fractures from minor trauma in isolation, have not been reported to our knowledge.

Aim:
We report on the presentation, investigation and outcome of a series of CD cases who presented with fractures in the absence of any major gastrointestinal symptoms (GI).

Subjects and Methods:
We reviewed the cases of four patients (age range - 4-19 y, two male and two female), identified from those attending the CD clinic (total number patients 139 from August 2002 –September 2009), who had presented with fractures from relatively minor trauma. Our index case presented with three out of four upper limb fractures associated with minor trauma over a four-year period. She was subsequently diagnosed to have CD. Gluten free diet and physiological vitamin D supplementation was introduced. Lumbar spine DEXA scans were used to assess bone mineralisation soon after diagnosis and at 1 year follow up. Second patient was an insulin dependent diabetic with known CD but was non-compliant with a gluten free diet. He sustained one fracture associated with minor trauma. Third patient had two previous upper limb fractures associated with minor trauma. He had minor abdominal cramps and a family history of CD. Fourth patient had one upper limb fracture following a minor fall. She had minor abdominal symptoms and a family history of CD. In all patients CD was confirmed with serology and intestinal mucosal biopsies. Patients underwent a range of blood tests to assess bone chemistry.

Results:
A relatively high proportion (4 out 139) of CD patients from our clinic presented with fractures. All 4 cases we report, presented with only a minor history of trauma. GI symptoms were absent in 2 out of 4 cases and only minor symptoms present in the remainder.

Our index patient had normal calcium, phosphate, alkaline phosphatase and vitamin D levels but a very high Parathormone level, which subsequently normalised after introduction of gluten free diet and physiological vitamin D supplementation. DEXA scan shortly after presentation was borderline abnormal; Bone mineral density (BMD) 0.58g/cm^2, Z score -2.0 and at 1 year follow up (BMD) was improved to 0.78g/cm^2, Z score -1.5. Second patient, had normal calcium and phosphate level but a low vitamin D level. Physiological vitamin D supplementation and advice to improve gluten free diet compliance was given. Third patient, had a low normal calcium level, but no active treatment was instituted apart from gluten free diet. Fourth patient, had a normal Parathormone, Vitamin D, Calcium, Alkaline phosphatase level but a slightly higher phosphate level, but no active treatment was instituted apart from gluten free diet. In all patients after establishing appropriate treatment, no further fractures were reported.

Summary:
Our case series suggests a higher than expected prevalence of untreated childhood coeliac patients presenting with fractures in association with minor trauma. Furthermore, GI symptoms were either absent or minor in all reported cases. Consequently, we suggest that a higher index of suspicion is necessary to consider a diagnosis of CD disease in patients presenting in this way.

A variable pattern of findings were noted on bone chemistry. In the index case, evidence of hyperparathyroidism, abnormal bone mineralisation and resolution of both without recurrence of fractures is supportive of a causal relationship. Relative Vitamin D deficiency identified in one case would influence bone mineralisation and may be a consequence of malabsorption in CD disease or due to other confounding causes.

Conclusion:
We suggest that undiagnosed CD in children can present with isolated fractures from relatively minor trauma and in the absence of GI symptoms. Consequently, a high index of suspicion is required in such children. We propose that all such patients should be investigated for CD disease and any possible associated abnormalities of bone mineralisation to aid in earlier diagnosis and help optimise treatment for both CD and bone health.

Clinical, pathological & epidemiological features of paediatric eosinophilic oesophagitis (EE) in a single tertiary centre

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Aims:-*To identify*
Primary- The clinical, endoscopic & histopathological features of paediatric EE presenting at our institution.
Secondary-The incidence of paediatric EE in the Humberside region & characteristics of a subgroup of patients with signs of both GERD & EE.

Methods:
The oesophageal biopsies with ≥ 15 eosinophils(e)/ high power field (HPF) between January 1st 2007 & 31st December 2008 identified 24 patients whose notes were then retrospectively reviewed.

Results:
1046 children had an oesophagoscopy of which 15% had features of oesophagitis on histology & 2.1% (24/1046) had EE (13male). Median age was 6 years (range: 0.5-15). The presenting symptoms were: feeding/swallowing problems (50%), other gastrointestinal symptoms (42%), & dietary allergy (33%). 6 patients had eczema +/-asthma. 4 children had refractory asthma & EE was diagnosed after a combined bronchoscopy & oesophagoscopy. Dietary elimination of proteins improved symptoms in 5 children.

In 17% (4/24) EE was associated with GERD, confirmed on pH study.

The commonest macroscopic findings were:
- Normal in 38%(9/24),
- Furrowing or trachealisation in 42%(10/24),
- ? Candida in 8%(2/24),
- 'oesophagitis' in 8% (2/24) and
- No record/unknown in 4%(1/24)

The median number of e/HPF was 32 (range:16-57).

Site	Median number & range of e/HPF per biopsy site was
Proximal oesophageal biopsies	24.5 (4-55)
Mid-oesophageal biopsies	37.5 (22-55)
Distal oesophageal biopsies	38 (20-57)

An otherwise rare presence of an 'increased' number of so called "squiggle" cells (> 6/HPF) was also seen in this cohort.

Conclusion:
EE should be suspected in any child with atopy &/or difficulty in feeding or swallowing. Suggestive macroscopy should prompt the endoscopist to obtain proximal oesophageal biopsies in addition to distal oesophageal biopsies as a high yield of e/HPF may help distinguish EE from GERD. pH study proved GERD & an increased presence of 'squiggle cells' in patients with EE suggests the possibility of an 'overlap' syndrome. The extrapolated population incidence of paediatric EE was 8 /100,000 in our region. Severity of symptoms could not be correlated with e/HPF

Cystic Fibrosis Liver Disease – a more severe phenotype

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Background
Evidence suggests that cystic fibrosis liver disease (CFLD) does not impact on mortality or morbidity in CF. However most studies to date have not used a comparison group of cystic fibrosis patients without liver disease. The aim of this study was to review 42 children with CFLD and their age and sex matched controls 7 years after they were enrolled in a baseline study of CFLD.

Methods
Participants were reviewed clinically, biochemically and radiologically in so far as possible during a routine hospital visit. 3 participants (2 cases, 1 control) were lost to follow-up, 1 control refused to participate, and 8 participants (5 cases) were excluded from follow-up. Ethical approval was obtained from all institutions providing care for participants.

Results
There was no difference in age, (mean 19.9±3.2yrs), gender (63% male) between cases and controls at follow-up. 7 participants with CFLD died (3 liver failure) 1 received a liver transplant, and 3 controls died. Patients with liver disease had 2.66 times the risk of death compared to controls. Patients with liver disease died younger mean age of death 17.38 ± 2.94 yrs compared to 20.66 ± 1.44 years (p=0.06) in controls. 5 of 7 patients with liver disease who died or received a transplant were female compared to 1 of 3 in the control group. There was no difference in height, weight or BMI between the groups. Nutritional parameters (sum skin fold thickness 30.1±17.4 V 38.1±21.2 p=0.03, upper arm fat area (1508.4±573.6 V 1508.9 ±662.3 p = 0.001), Shwachman score (49.1±11.4 V 55.4±12.5 p=0.02) were poorer in patients with CFLD. Participants with CFLD had a higher proportion of CF Diabetes Mellitus (40.7% V 15.2% p=0.02) at follow-up. There was no statistically significant difference in pulmonary function between cases and controls at follow-up. Nine children with evidence (clinical/radiological) of liver disease at baseline, had no evidence of portal hypertension as adults.

Conclusion
Patients with CFLD may have a higher mortality risk and die younger than CF patients without liver disease. However some children with CFLD will not manifest clinically significant liver disease as adults.

Development and Validation of a Disease-specific Quality of Life Measure for Children with Achalasia

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Background:
Achalasia is an uncommon oesophageal motility disorder occasionally affecting children. Children with achalasia can experience significant morbidity and quality of life (QoL) has never been studied in this population.

Aim:
To develop and validate a disease-specific quality of life (DS-QoL) measure for children with achalasia.

Methods:
Item response theory methods were used to develop the DS-QoL measure. All children with a diagnosis of achalasia at one hospital were prospectively asked to complete this DS-QoL measure as well as the well validated PedsQL™ generic quality of life questionnaire, either in the clinic setting or in their own home. The construct validity of the DS-QoL measure was assessed by comparing items and domains with the PedsQL™ questionnaire. Reliability was assessed using Cronbach's alpha coefficient for internal consistency.

Results:
17 children completed the final DS-QoL measure, which consisted of 20 items in three domains. The completion rate for items was 99%. "Floor and ceiling effects" ranged from 0-19%. Construct validity was good with significant correlation between 2 domains and 2 items on the PedsQL™. Reliability was excellent, with Cronbach's alpha coefficient ranging from 0.78-0.93.

Conclusions:
This DS-QoL measure is appropriate for use in children with achalasia and has shown good results in this validation study. Further work in higher numbers is necessary to determine discriminant validity and test-retest reliability.

Does the presence of eosinophilic esophagitis in children with asthma suggest food allergy aetiology?

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Dr G Vance, Consultant Paediatric Allergy
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Introduction:
Two thirds of the children who have eosinophilic esophagitis (EE) have a history of asthma, eczema, food allergies, environmental allergies or chronic rhinitis. We describe two children with asthma and eczema who also had EE. A trial of non allergenic diet significantly improved all their symptoms and allowed reduction in asthma medications.

Case 1:
4 year old girl had been under the respiratory team since the age of 30 months with recurrent wheezing since infancy. She required frequent courses of prednisolone, hospitalisation including HDU/PICU admissions. Her asthma management was stepped up to long term prednisolone 10 mg daily, theophylline and home nebulisations.

She was referred to the gastroenterology team because of reflux like symptoms not responding to acid suppression. On endoscopy typical macroscopic features of EE was noted and confirmed on histology. She was commenced on exclusive elemental feeding with neocate advance and four weeks later repeat endoscopy showed complete resolution of EE. Whilst on the exclusive diet her asthma control improved, prednisolone dose was weaned to 5mg alternate days and she did not have any hospitalisation. After four weeks few foods were reintroduced successfully without any clinical deterioration. Three months later some of her symptoms returned and this was due to non adherence to the recommended diet.

Case 2:
36 month old girl initially presented to allergy clinic at 1 year of age with severe food allergies and extensive eczema. At two years symptoms of asthma were also noted. There was progressive worsening of her asthma with disturbing nocturnal wheezing. She required stepwise increases in asthma treatment from high dose inhaled beclomethasone and monteleukast to regular prednisolone and high dose theophylline.

She was referred to gastroenterology to look for evidence of EE. On endoscopy, typical macroscopic features of EE was noted and confirmed on histology. She was commenced on exclusive non allergenic diet with neocate advance. Within 2 weeks of starting elemental feeds her asthma control improved and eczema was remarkably better. She had complete resolution of nocturnal symptoms. By 4 weeks, prednisolone dose was weaned down from 10 mg daily to 5 mg alternate day.

Discussion:
In 1995, Kelly and colleagues published a sentinel paper on EE with a case series of 10 paediatric patients who showed symptomatic and histologic improvement on an elemental diet. Case series data from several centres in both adults and children report a 92% to 98% patient response to amino acid–based formula. Only very few cases of environmental allergens causing EE have been reported. Thus strongly suggesting dietary allergens as the main trigger for EE.

In children with multiple allergies especially brittle asthma and eczema often environmental allergens are the major offending agents. In our cases asthma and eczema improved on exclusive elemental diet indicating that the major allergen was dietary and not environmental. It should be noted both our cases had significant EE whilst on steroids, which is a well known treatment for EE.

The above two cases support our hypothesis that the presence of EE in the multiple allergic child is suggestive of food allergy aetiology. Larger studies will be required to answer this question more convincingly.

Conclusions:
Clinician must be alert to the possibility of EE, particularly when the symptoms of gastro oesophageal reflux fail to respond to anti-reflux therapy, or when the gut symptoms are associated with eczema, overt food allergies or reactive airways.

In all children with difficult to control multiple allergies the presence of EE should be sought for and if present a trial of non allergenic diet should be considered.

Evaluation Of Aggressive Nutritional Intervention In Very Low Birth Weight Infants (VLBW) During The First 28 Days Of Life.

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Introduction:
In 2005-06, we piloted a study to assess our nutritional practice at Leicester Royal Infirmary (LRI) and changes were made to optimise the energy and protein intake. Following this change in practice, a retrospective observational study was conducted to evaluate if aggressive nutritional intervention could correct the nutrient deficiencies and to determine the average daily weight gain during the first 28 days.

Methods:
Daily nutrient intakes were analysed in very low birth weight infants (Birth wt <1250gms) admitted to our unit between July 2007 - June 2008. Infants were excluded if they had major congenital anomalies or abdominal surgical problems. Actual intake of calories and protein (both enteral & parenteral) was subtracted from recommended dietary intake (120kcal/kg & 3.8g/kg) respectively and nutrient deficiencies calculated1, 2. The data were analysed using SPSS Version 16 software.

Results:
During the current (2007-08) study period, complete data were available for 42 out of 54 eligible infants. There was no statistical difference when matched with previous study in terms of mean gestational age, sex distribution, birth weight (p>0.152) nor weight gain at 28 days (p>0.05). The average daily weight gain was 9.4g/kg/d when compared to 8.4g/kg/d in previous study. The weekly and cumulative energy and protein intake and deficits are calculated and depicted in the table. There was significant statistical difference (p<0.001) in both energy and protein intake in the first week. Although there was significant nutrient deficits accrued by 4 weeks, there was no statistical significance between the 2 study group (p>0.112).

Calories	Group	Mean calorie intake (Kcal/kg/d) + SD	Mean calorie deficit (Kcal/kg/wk)
Week 1	2005 2008	59.66±13.00 70.55±10.67	-422.38 -346.15
Week 2	2005 2008	105.38±22.12 111.13±18.42	-102.34 -62.09
Week 3	2005 2008	120.52±24.52 122.87±21.52	3.64 20.09
Week 4	2005 2008	128.67±23.61 130.01±18.92	60.69 70.07
Cumulative deficit over 28 days	2005 2008		-460.39 -318.08

Protein	Group	Mean protein intake (g/kg/d) +SD	Mean protein deficit (g/kg/wk)
Week 1	2005 2008	1.26±0.42 1.86±0.37	-17.78 -13.58
Week 2	2005 2008	2.94±0.72 3.10±0.63	-6.02 -4.90
Week 3	2005 2008	3.37±0.86 3.47±0.82	-3.01 -2.31
Week 4	2005 2008	3.57±0.83 3.80±0.68	-1.61 0.00
Cumulative deficit over 28 days	2005 2008		-28.42 -20.79

Conclusion:
The aggressive nutritional intervention resulted in significant improvement in both energy and protein intake during the early postnatal period. Despite these efforts cumulative nutrient deficits were not recouped which further raises the question of whether provision for catch up growth should be made as current RDI's make up for maintenance and normal growth3.

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Idiopathic constipation in infants and young children – a proactive approach to treatment

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Aim
This paper aims to give an overview of a project being undertaken to raise awareness regarding the appropriate treatment of idiopathic constipation in infants and young children, and enable health care professionals to identify and manage the problem of constipation in this age group more effectively. This included working with both local pharmacists and health care professionals to signpost and advise families appropriately.

Background
Idiopathic constipation in children is not uncommon however the management of constipation in infants is not well studied and often appears to focus primarily on dietary manipulation. A study by Loening- Baucke (2005) showed that dietary changes alone resolved constipation in only 25% of children with constipation whilst laxatives, including polyethylene glycol resolved constipation in 92% and was found to be efficient and safe in infants and toddlers, yet the use of laxative treatment in this age group is poorly documented.

Subject and methods
In 2008 there were 96 new referrals to the paediatric continence promotion service for children with idiopathic constipation and although over 85% of the children gave a history of the problem starting before the age of 2 years, the majority had not been referred until the problem had become chronic – some 12mths to 2 years later.

There appeared to be a general lack of understanding regarding idiopathic constipation in infants and young toddlers resulting in inappropriate or inadequate treatment and it was only when the problem started to affect potty training or continued into school age for example was the problem taken seriously. A study by Blum et al (2004) which looked at 380 children between 17-19 months reported that of the children who demonstrated stool withholding 93.4% were constipated.

There was a belief by some healthcare professionals that laxative treatment in this age group was not appropriate, yet clinical experience has shown that treatment of constipation in infants and young children is safe and effective (Loening-Baucke 2005). There had also been a number of incidences when local pharmacists had been reluctant to dispense laxatives prescribed to young children as the dosages requested were outside what was suggested in the BNFc – this obviously caused the families great anxiety. Also we know that once constipation becomes chronic it is more difficult to treat and is accompanied by associated problems such as pain, stool withholding and the risk of acquired mega rectum/colon. This is not only distressing for the child and family but has potential cost implications for the NHS (Liem et al 2009).

A number of studies have also reported that early management of constipation is important in preventing long term defecation problems (Candy & Belsey 2009, van den Berg et al 2005) and early intervention with oral laxatives will improve outcome of functional constipation (Michail et al 2004, Pijpers et al 2009). Waiting until constipation becomes chronic is clearly not appropriate

Awareness raising sessions were initially undertaken with community nursing staff including HV's and Child Health Practitioners then links were made with both community and primary care pharmacists and a care pathway mapped out for the GP's to signpost early and timely referrals to the Paediatric Continence Promotion Service

Results and conclusion
Early indications are that the project is having a positive effect with a significant increase in number of children under aged 2 years, with constipation, being referred to us, compared with the same time period in 2008. The early effective treatment of constipation in infants and young toddlers has resulted, in the majority of cases, to complete resolution of the problem within weeks to the obvious benefit to the child and family. Particularly as, as a result, the learned behaviour of 'holding on' does not then develop which often self perpetuates the cycle of delayed passage of hard painful stools with all the associated problems. The introduction of laxatives in this age group, when initial diet and fluid adjustment alone has failed, is vital to ensure early resolution and prevent the problem of idiopathic constipation becoming chronic.

Is rejection less common in children undergoing liver transplantation for hepatoblastoma?

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Introduction:
Hepatoblastoma is a rare malignant tumour, almost exclusive to childhood. Treatment consists of chemotherapy followed by either hepatic resection, where feasible, or liver transplantation. As this chemotherapy is immunosuppressive the incidence of rejection post transplantation may be reduced.

Aim:
To compare the incidence of histological rejection in children undergoing liver transplantation for hepatoblastoma with those transplanted for biliary atresia in a single unit.

Subjects and methods:
All 20 patients who underwent transplantation for hepatoblastoma were identified retrospectively. These were matched for age, sex, year of transplant and type of immunosuppression to the control group transplanted for biliary atresia (n=60). Exclusions were patients transplanted for other types of liver tumour and patients with polysplenia syndrome.

Results:
Mean age at transplant was 3.25 years in the tumour group and 3.28 years in the biliary atresia group. Overall survival was 75% of tumour patients currently well compared with 88.3% of biliary atresia patients. Acute histological rejection was less common in the hepatoblastoma group (50% vs 75% respectively, $p<0.04$). In the hepatoblastoma group, there appeared to be 2 peak periods for the development of acute rejection, at 2 weeks at 4 weeks. The control group had a single peak in rejection at 4 weeks. Episodes of chronic rejection were double in the biliary atresia group compared with the tumour group (10% vs 5% respectively). Equal levels of immunosuppression were achieved in both groups. Renal function was reduced 1 year post transplant for both groups (GFR (ml/min/1.73m2) was 103.5 pre-transplant and 78.88 post transplant in the hepatoblastoma group compared to 127.1 pre-transplant and 93.24 post transplant in the biliary atresia group).

Summary:
Rejection is less common in children undergoing liver transplantation for hepatoblastoma.

Conclusion:
Children undergoing liver transplantation for hepatoblastoma could benefit from less intensive immunosuppression regimens.

Joint Feeding clinic The way Forward

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Objectives
To assess the effectiveness of a joint feeding clinic run by a paediatric surgeon and a community paediatrician in addressing the nutritional needs and improving the feeding difficulties of children with neurological impairment and severe gastro oesophageal reflux disease (GORD)

Material and methods
Clinic data between August 2005 and June 2009 was audited using a specifically designed Proforma. Type of referral, reason for referral, number of visits paid by each patient, presenting symptoms, investigations ordered and outcomes were recorded.

Results
Over 4 years an average of one clinic per month was held resulting in 44 clinics in total. A total of 101 children were seen. Most referrals to clinic came from hospital paediatricians, followed by community paediatricians. Main reasons for referral were GORD and its associated sequelae, request for Gastrostomy insertion, and poor feeding. Vomiting, poor weight gain and retching were the main presenting symptoms. Most patients had attended clinic only once (fewer than 10% attended more than 3 appointments). PH study was the commonest investigation requested followed by videofluroscopy and upper GI contrast study. The majority of children attending the clinic were already on various medications, and 25 received new prescriptions at the clinic. Symptoms improved in the majority of children. Surgery was discussed in 52 appointments, 33 children had a gastrostomy, and total oesophago-gastric dissociation was performed in 23. 4 children had Nissan fundoplication. Dietary advice was offered to 11 children, 25 required no intervention.

Conclusion
The clinic appears to have succeeded in providing a useful service for neurologically impaired children with feeding problems. Whilst the clinic is predominately surgical, surgery is not the outcome for every patient, and other interventions and treatments are also offered specifically tailored to each patient's unique needs.

Low Gamma GT Cholestasis in Microvillous Inclusion Disease

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Introduction/Background

Microvillous Inclusion Disease (MVID) is a congenital disorder characterised by ultrastructural abnormalities of the epithelial brush border which causes intractable watery diarrhoea, electrolyte imbalance and failure to thrive. Most affected individuals require lifelong parenteral nutrition (PN) or progress to intestinal transplantation. Although liver disease is reported in this patient group, this has primarily been described as a complication of intestinal failure and long-term parenteral nutrition. We describe a case of MVID who was successfully weaned from PN in early childhood, but went on to develop recurrent episodes of cholestasis, clinically resembling low Gamma GT familial intrahepatic cholestasis.

Aim

To describe the clinical presentation of liver disease in a child with MVID and discuss the potential aetiology, including comparison with other forms of low Gamma GT cholestasis.

Subjects and methods

A 10 year old girl presented to our unit with a three day history of pruritus and jaundice. She was well known to the team, having presented in early infancy with severe, intractable diarrhoea and weight loss. A diagnosis of MVID was made on the basis of histology and electron microscopy (with subsequent finding of a genetic mutation in keeping with the diagnosis). She was PN dependent from diagnosis and tolerated PN well, with no evidence of Intestinal Failure Associated Liver Disease. Contrary to the usual disease course, was weaned from PN at four years of age and subsequently thrived on a normal diet.

On re-presentation, she was noted to be mildly jaundiced but with no hepatosplenomegaly or stigmata of chronic liver disease. Extensive investigation failed to reveal a cause for her jaundice and liver biopsy revealed a mild inflammation in portal tracts (predominantly eosinophils and lymphocytes), low grade bile duct injury and associated biliary recruitment. No significant portal tract expansion or fibrosis was demonstrated. She received symptomatic treatment for her pruritus and over the next 6 months her jaundice and itch gradually resolved.

The girl presented again at 15 years of age with a similar short history of jaundice and itch.

Results

Liver function tests at presentation revealed a conjugated hyperbilirubinaemia and raised liver enzymes (Bilirubin 93µmol/l, ALT 410u/l, AST 135u/l). Gamma GT was normal at 12 iU/l. She underwent a repeat diagnostic work-up (including extended virology, immunoglobulins and autoantibody screen, alpha-1-antitrypsin level and phenotype and copper and caeruloplasmin), which did not reveal a cause for her liver dysfunction. Repeat liver biopsy revealed prominent features of cholestasis with canalicular bile plugs, but no portal inflammation or biliary pathology. Once again there was no significant evidence of fibrosis.

The biochemical and histological features in this case share similarities with the various types of familial intrahepatic cholestasis. A form of intrahepatic cholestasis has previously been described after small bowel transplantation in a small case series of patients with MVID. Although the pathophysiology of cholestasis in these cases has not been elucidated at a molecular level, the pattern of liver disease would suggest a defect of bile canalicular transport. The gut manifestations of MVID are thought to arise due to a major defect in membrane trafficking in enterocytes secondary to altered structure of the cytoskeleton. The presentation of liver disease in these patients suggests that the effects of MVID are not limited to enterocytes. It may be that an analogous failure of membrane trafficking occurs at the biliary level, giving rise to cholestasis.

The episodic nature of cholestasis in this case and the lack of progression echo the clinical features of Benign Recurrent Intrahepatic Cholestasis. However, prognosis must remain guarded for our patient at this stage.

Summary and Conclusion

The development of intrahepatic cholestasis in MVID suggests that the pathophysiology of MVID is not limited to the gut. It should not be assumed that jaundice occurring in these patients always represents Intestinal Failure Associated Liver Disease.

"My tummy hurts."... Or rather ..."Her tummy hurts"

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Abstract not published due to sensitive nature of content (Fabricated illness submission)

Outcome and Complications such as IFALD of Parenteral Nutrition (PN) in Hospitalised Children under one year of age

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Introduction:
The development of PN has altered the outcome for children under one year of age with intestinal failure (IF). The aim of this study was to review type of diagnosis that predispose these children to IF and dependency on PN and review their complications and outcome.

Methods:
63 children under one year received PN for > 28 days at Great Ormond Street NHS Trust over a 2- year period. Age, sex, underlying disease and survival were recorded and analysed. Complications such as intestinal failure associated liver disease (IFALD) were recorded during the duration of PN (definition of BSPGHAN Intestinal Failure Working Group). Outcome was assessed according to underlying disease.

Results:
63 children received PN for > 28 days. This group composed of 35 (56%) males with a mean age at start of PN of 0,3 year (birth-0.95yr). The mean duration of PN was 80days (28-247 days). 18 (28.6%) were born premature with gestational age of 28 weeks (24-33).
40 (63.5%) had a primary non- digestive diagnosis and 23 (36.5%) a primary digestive diagnosis (PDD). 16 (26.7%) developed IFALD as a complication of PN. 9 (15%) developed IFALD type I, 5 (8.3%) type 2 and 2 (3.3%) developed IFALD type 3 as end stage liver disease. IFALD was more prevalent in children with PDD. 40 (63.5%) children could be weaned off PN and start on enteral feeds, 8 (12.7%) patients had irreversible IF and were discharged on home PN (HPN), 14 (22.2%) children died. Deaths in 6 (9.5%) were related to PN and in 7 (11.1%) to the underlying disease.

Conclusion:
Infants under one year with severe intestinal failure for > 28 days have a survival rate of more than 75%. 64% of infants could be successfully weaned off PN, 12% developed irreversible IF and remained dependent on PN. In our cohort is a mortality rate of 22%. 27% developed IFALD as a complication of PN. Children with a primary digestive disorder have the highest risk for developing liver damage associated with PN.

Safety and efficacy of Heparon Junior in infants with cholestatic liver disease

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Background:
Children with cholestasis benefit from feeds that are rich in medium chain triglycerides (MCTs), branched chain amino acids and preferably low in sodium. Of the currently available specialist products Heparon Junior (SHS International) appears to have the best nutrient profile to meet the needs of these infants however the product has not been evaluated systematically in the UK. The composition of Heparon Junior is high energy/high protein, nutritionally complete, with branched chain amino acids (30%), low sodium, and high MCT (49%).

Aim:
To evaluate the safety, tolerability, and efficacy of Heparon Junior in infants with cholestatic liver disease.

Patients and methods:
Seven (4 female) infants with cholestasis (median age 9 weeks (range 5-28 weeks)) were given Heparon Junior (minimum of 60% of energy intake) orally or via NG tube for a period of 24 weeks. Gastrointestinal symptoms (vomiting, diarrhoea, discomfort) were recorded daily by parents along with any additional observed adverse effects.

Anthropometrics (calculated z scores for weight, length, BMI, head circumference (HC), mid upper arm circumference MUAC), liver function tests (total bilirubin, GGT, AST), serum albumin and sodium were assessed at baseline, 4, 12 and 24 weeks.

Results:
The patients' diagnoses were biliary atresia (4), idiopathic cholestasis (2) and alpha-1-anti-trypsin deficiency (1). Of these, 1 withdrew from the study after 12 weeks as was no longer cholestatic, and 3 received liver transplants at weeks 18, 20 and 21 respectively. In these patients end data was analysed at 12 weeks rather than at 24 weeks. Median baseline serum markers were: total bilirubin 138 (range 12-164), GGT 832 (range 105-2080), AST 185 (range 61-294), albumin 39 (range 30-58), sodium 138 (135-139). At completion median serum markers were: total bilirubin 62 (range 2-279), GGT 196 (range 15-381), AST 147 (range 61-306), albumin 35 (range 30-45), sodium 140 (135-140).
No adverse effects from Heparon Junior were reported to the investigators by parents or medical staff.
No persistent gastrointestinal symptoms attributable to Heparon Junior occurred.
Median z score change values from baseline to completion for anthropometrics were: weight 1.09 (range 0.53-2.66), length 0.47 (range 0.15-4.55), BMI 1.11 (range 0.15- 1.62), HC 0.62 (range -0.40- 2.15). MUAC z scores were calculated for 4/7 subjects (no WHO centile charts for infants <3 months) 1.10 (-0.01- 3.0).

Conclusion:
These results show that Heparon Junior is safe, well tolerated and that cholestatic infants can show increased growth parameters when given Heparon Junior as their primary energy source.

SMOF lipid in intestinal failure associated liver disease – experience in a regional centre

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Background:
Parenteral nutrition (PN) is life saving in patients with intestinal failure (IF).IF associated liver disease (IFALD) results in high morbidity and mortality. Aetiology, prevention and treatment of IFALD are poorly understood. Soybean-oil emulsions (rich in omega-6) used in standard PN may contribute to the development of IFALD(1).Recent studies have shown that fish oil based fat emulsions (rich in omega-3) may influence IFALD by improving bile flow, inhibiting steatosis and through immunomodulatory/anti-inflammatory effects(2).

Aims
To review the effect of SMOF lipid on IFALD as rescue treatment

Methods
Retrospective study 2007 -2009 on patients with intestinal failure & persistently elevated conjugated serum bilirubin (>50 µmol/L) .SMOF (lipid emulsion based on soybean oil, medium chain triglycerides, olive oil and fish oil) used as lipid source .

Results
8 patients were identified over the specified period who developed IFALD. in 2 patients, bilirubin >200µmol/L in 6 patients , bilirubin 50 - 200µmol/L Indications for PN were short bowel syndrome(SBS) secondary to gastroschisis repair or necrotising enterocolitis (n=7),& difficulty establishing feeds secondary to gastroschisis repair (n=1).The median age at start of SMOF lipid was 63(15-252) days and at IFALD resolution was 148.5(46-420).The mean SMOF lipid use at the start of rescue treatment and at resolution of IFALD were 2.5g/kg/day and 2.3g/kg /day respectively. The feed tolerance at start of SMOF lipid commencement and resolution of IFALD were 30 ml/kg/day(0-125) and 100 ml/kg/day(55-132) respectively. All patients except two showed marked improvement and resolution of IFALD following SMOF The median time for resolution of bilirubin, alkaline phosphatase & alanine transferase to normal levels were 9 (4-22) weeks,13 (4-22) weeks, 8 (2-28) weeks respectively. One patient has hepatic fibrosis and One patient has signs of portal hypertension and recurrent line infections which necessitated referral for consideration of combined small bowel and liver transplant. 4 patients now off PN& 4 children still on PN and tolerating feeds There were no adverse side effects observed with the use of SMOF.

Conclusions
Our study demonstrates that the use of fish-oil–based emulsion in infants who depend on PN as a life-sustaining measure may reverse cholestasis and fatal liver disease.Parenteral fish oil emulsion appears to be safe and effective in the treatment of IFALD in infants. As noted in earlier studies,SMOF lipid may reduce mortality and organ transplantation rates in children with SBS.Large multicentre randomised controlled trials are needed.

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Technetium Isotope Scans in the Evaluation of Gastrointestinal Haemorrhage in Childhood and their Accuracy in Detecting a bleeding Meckel's Diverticulum.

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Background
Radionucleotide Technetium scans have been used as an aid to the diagnosis of symptomatic Meckel's Diverticulum (MD) for over 30 years and are regarded as the 'Gold Standard' test for MD detection. Anecdotal evidence suggests that scans are frequently requested but are rarely found to be useful.

Objectives
To document the frequency of diagnosis of MD and to suggest clear indications for requesting a Meckel's Scan in children under 16 years of age.

Methods
This was a retrospective single centre study with both a radiological and pathological arm. The reports for all Meckel's scans carried out on children under the age of 16 years between 1998 and 2008 were identified and evaluated.

Simultaneously, the children having a MD removed surgically were identified via the histopathology database. Notes were retrieved and data collected for cases with a positive scan and all cases where MD histology was available.

Results
Five of 106 children had a positive scan, all 5 proceeded to surgery. Of these only 3 were found to have a MD at operation. All 3 had rectal bleeding and anaemia with an average Hb of 7.9g/dl.
Thirty-six children were identified as having a MD from the pathology database. Three had a positive Meckel's scan at our centre (see cases above) and 3 other cases had had a positive scan performed at the local District General Hospital. These additional cases also presented with rectal bleeding and anaemia.

Conclusion
Meckel's scans are highly sensitive (100%) and specific (98%) but are probably ordered inappropriately (95% negative). Children with a positive Meckel's scan as well as rectal bleeding and anaemia are more likely to have a MD confirmed at surgery.

The Effect of Biologics on growth in children with Inflammatory Bowel Disease (IBD): significant increase in responders that can be independent of steroids and puberty

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Introduction
Treatment with biologics [Infliximab (IFX), Adalimumab (ADA)] in children have been used to treat IBD unresponsive to other therapies. The data studying their effect on growth, however, is limited.

Aim
A retrospective study of growth, puberty and disease activity over the 6 months prior (T-6) to starting biologics, baseline (T0) and for the following 6 months (T+6) after in children with IBD

Subjects & Methods
The growth and treatment details of 42 children were reviewed of whom 32 (CD 28, UC4) were eligible for the study. The median (10th, 90th) age at IFX treatment was13.2yr (8.0, 16.7). 5 patients all who had previously been treated with IFX subsequently received ADA after a median period of 1.7yrs (2.0, 3.1). Data on disease markers (CRP, ESR, and Albumin), total Alkaline Phosphates (ALP) and a physician global assessment were also collected.

Results
In the 28 children with CD, height velocity (HV) increased from 3.6cm/yr (0.4, 7.8) at T0 to 5.5cm/yr (2.1, 9.2) at T+6(p=0.003). 21(75%) demonstrated a clinical response to IFX in whom, HV increased from 2cm/yr(0.3.7.1) to 6cm/yr(3.3,9.1)(p=0.003);in non-responders, HV was 4.3cm/yr(2.5,8.6) at T0 and 3.0cm/yr(2.0,11.3)(0.110) at T+6. In 13 children who had remained prepubertal, during the study period HV increased from 4.5cm/yr(0.4,8.0) to 5.5cm/yr(3.3,8.4)(p=0.05).In the subgroup of 11 children who had a reduction(n,2) or cessation in GC (n,9), HV increased from 1.8cm/yr (0.3,8.3) at T0 to 5.6cm/yr(2.2,9.2) at T+6 (p=0.110), whereas those children who did not receive any GC over the12 months an increase from 3.7cm/yr(0.6,6.5) to 6.4cm/yr(2.9,9.0)(p<0.05). HV at T0 and T+6 showed a significant association with the average ALP over the prior 6 months(r, 0.39, p<0.05).

Of 28 children with CD, 5 switched over to ADA therapy (4 had lost response and 1 was intolerant to IFX). Each child received a median of 9 injections (5, 14). Median HV increased from 2.2cm/yr (0.4, 10.8) at T0 to 5.5cm/yr (4.7, 10.0) (p=0.14) at T6. Of 4 children with UC, 3 demonstrated a clinical response to IFX therapy. None of these patients showed an improvement in growth from T0 to T6 2.6 cm/yr (0.5, 3.2) and 2.4 cm/yr (0.83, 3.8).

Conclusion:
Clinical response to biologic therapy is associated with an improvement in linear growth in children with CD. This increase is also seen in prepubertal and GC naïve children and cannot solely be attributed to a change in these factors.

The intestinal expression of toll-like receptor genes is dysregulated on endoscopic biopsies from patients with inflammatory bowel disease

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Introduction
The Toll-like receptors (TLRs) are a family of pattern recognition receptors which have been classically associated with macrophage activation. However, the discovery that enterocytes also express TLRs, and that germline mutations in certain TLRs confer susceptibility to inflammatory bowel disease (IBD), has led to further research into this group of molecules. With regard to IBD the most studied in this group is TLR4 which is associated with the activation NF-κB and the release of pro-inflammatory cytokines. Mechanisms such as intestinal restitution and apoptosis are also influenced by TLR4 signalling, with TLR2 and TLR9 having been shown to be important in intestinal inflammation.

Aims and Methods
To compare intestinal TLR gene expression in adult patients with Crohn's disease (CD) (n=53), ulcerative colitis (UC) (n=67) and controls (n=31; 23 normal and 8 inflamed non-inflammatory bowel disease patients). Paired endoscopic biopsies were taken from four specific anatomical locations for RNA extraction and histology. Amplified cRNA was then analysed for ten human TLR genes using the Agilent platform. Statistical analysis was carried out using Mann-Whitney U and Kruskal-Wallis testing.

Results
Comparing all inflamed and non-inflamed biopsies from disease and control groups, similar patterns were seen in both CD and UC. In both disease-group biopsies TLRs 1-5 were upregulated, with TLR9 additionally upregulated in UC. Controlling for anatomical location and inflammation status, TLR3 was significantly upregulated in non-inflamed sigmoid CD and UC biopsies compared to non-inflamed sigmoid control biopsies (p=0.0036 and p=0.0294 respectively). In non-inflamed control biopsies, four genes (TLR1, TLR4, TLR6 and TLR9) showed significant variation throughout the three areas of the large intestine. All four of these genes showed loss of this 'normal' variation in non-inflamed CD biopsies and besides TLR6 a similar loss of variation was seen in UC.

Conclusions
Although toll-like receptor gene expression on endoscopic biopsies does not account for varying expression on different cell types, the variation observed between inflammatory bowel disease patients and controls is notable. The similarities between both Crohn's and ulcerative colitis are also of interest, suggesting that dysregulation of TLRs may be closely involved in the propagation of inflammation. It is unclear if these differences are a result of significant dysregulation on certain cell lineages and whether genetic mutations or abnormal cell responses play a role. Further work is required to determine the function of these receptor molecules within inflammatory bowel disease to increase the understanding of pathogen-recognition mechanisms and their possible dysfunction in intestinal disease.

Conflict of Interest: None declared

The role of extra-intestinal viral infections in exacerbations of inflammatory bowel disease: a systematic review.

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Introduction

Inflammatory bowel disease (IBD) is a chronic disease characterised by acute exacerbations, often leading to hospital admission and significant morbidity. Studies have so far not provided strong evidence for the causes of these exacerbations. However, recent research detailing the role of a dysregulated immune response in those affected and previous work raising the possibility of seasonal variation in exacerbation frequency both suggest an infectious aetiology.

Aim and Methods

To study the current evidence for the role of extra-intestinal viral infections in the exacerbation of established IBD at any age by formal systematic review. An electronic database search of the Cochrane Library, Medline, Embase and the British Nursing Index and Archive was performed with keywords related to IBD, exacerbation and viral infections; PubMed was also searched. A hand search of reference lists of articles was also performed, along with reviews of major gastroenterology journals and meeting abstracts. The papers identified were then reviewed and the level of evidence (EL) was assessed using the Scottish Intercollegiate Guidelines Network (SIGN) methodology.

Results

From the initial 1095 hits 114 papers were reviewed in full text. Of these, four were identified that presented data on the association of extra-intestinal viral infections with exacerbations of IBD. Two papers were case series (EL 3), with one combined case series and cohort (EL 3) and one prospective cohort (EL 2-). Two studies had poor definitions of IBD relapse and all four relied on patient-reported symptoms at each outpatient visit regarding symptoms of viral upper respiratory tract infection. In one case series viral serology was also taken in an unplanned manner. As a result of this poor methodology there were no papers with both clearly defined exposure and outcome measures. Across two of the case series and the pure cohort a total of 456 attendances were recorded; 71 presented with relapse, 25 (35%) of whom had recent symptoms of viral URTI; of the non-relapsers 118 (31%) had recent URTI symptoms (χ^2=0.580; p=0.4465). Overall, two papers concluded that extra-intestinal viral infections were associated with IBD relapse with two unable to confirm this association.

Conclusion

The current evidence does not suggest a clear association between extra-intestinal viral infections and the exacerbation of IBD. However the evidence is very limited and is based on subjective reporting and serology. Further prospective cohort studies, with a combination of patient reporting and more robust laboratory investigation at the time of exacerbation, need to be performed to determine if such a relationship exists.

Conflict of Interest: None declared

The Spectrum of Eosinophilic Oesophagitis in a district General Hospital

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Background

Eosinophilic Oesophagitis is a separate clinical and pathological disorder of the oesophagus. Although symptoms may be similar with Gastro-oesophageal reflux disorder, yet it is well recognized to has a different management.

There have been reports in the literature of some rare association with eosinophilic oesophagitis in rare syndromes such as Rubenstein-taybi syndrome. Other conditions presenting with eosinophilic oesophagitis although there has been no clear association.

Aim

We have studied the epidemiology, associated medical condition, clinical presentation, and investigations including endoscopic findings, clinical and histologic response to treatment.

Methods

The clinical symptoms, presentation, diagnosis, management were reviewed. We a We noted other associated medical conditions.

Their investigations and clinical progress before and after treatment were studied.

Results

From 2004-2007, 15 patients diagnosed with eosinophilic oesophagitis in our GI clinic were selected.

The youngest was a male diagnosed at 16 month of age, and the oldest was 15 years old male. Only 2 were non-Caucasian

The gender ratio of the 15 patients was 9 males to 6 female.

One patient has Asperger's syndrome and also had celiac disease confirmed on endoscopy. Another patient has ADHD, and two patients have global developmental delay with chromosomal abnormalities.

Two patients with asthma, and one patient with multiple food allergies. One patient with mastocytosis which was shown on biopsy of a small skin lesion removed surgically from his anterior chest wall. Another patient had eosinophilic colitis as well as eosinophilic oesophagitis. Two patients developed eating disorders subsequently.

Nine patients have high IgE level, and nine had allergen specific RAST. In 5 patients, the eosinophil count was mildly elevated in the peripheral blood.

Transoral incisionless fundoplication for treatment of paediatric gastroesophageal reflux disease: a feasibility study

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Aim:
A new transoral incisionless fundoplication (TIF, EsophyX,TM) technique was evaluated for the treatment of paediatric gastroesophageal reflux disease (GERD) in a prospective feasibility clinical trial in the U.K.

Methods:
Inclusion criteria: Chronic & symptomatic GERD, refractory to, or dependent, on high dose proton pump inhibitor therapy.

Exclusion criteria: >18 years, or with dysphagia, obesity (BMI) > 99th centile, previous upper intestinal surgery, or hiatus hernia > 2cms.

Period: December 2008-May 2009.

Pre-procedure assessment included oesophagoscopy, 24 hour oesophageal pH, & validated reflux quality of life score. The TIF was designed to partially reconstruct the antireflux barrier through augmentation of the gastroesophageal junction.

Results:
12 patients (8 male) underwent the TIF procedure, of median age 12.25yrs (8-18) & weight 38.2kg (26-91)

The median duration of GERD symptoms in the patients was 45 months (24-70).The median pre-TIF % reflux index on pH study off treatment was 11.4% (6-48). Hiatus hernia was present in 17% (2/12)

Median operative time was 42 minutes (25-94). In all patients a greater curvature of 3cm, lesser curvature of 1cm and a 270 degree wrap was achieved.

Adverse events were limited to 2 patients that had retrosternal chest pain & were subsequently found to have pneumomediastinum on CT chest but no leak on barium swallow. 1 of them developed pyrexia with the chest pain & was treated for possible mediastinitis. He was discharged home after 5 days of intravenous antibiotics. Subsequently CO2 insufflation was employed & more rapid absorption resulted in no further mediastinal gas leak.

Conclusion:
This is the first report of paediatric experience with a full thickness transoral endoscopic anti-reflux procedure, & this shows that the TIF procedure using the EsophyX,TM is feasible, & safe with CO2 insufflation in children.

ABSTRACTS FOR FRIDAY 29TH JANUARY 2010

Invited speakers' abstracts

Gastrointestinal Motility Disorders in Children

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Gastrointestinal motility disorders in children include a wide spectrum of disorders of varying severity involving different segments of the bowel. Esophageal disorders often present with symptoms of feeding refusal, dysphagia, odynophagia, or vomiting. Gastric and small bowel disorders may also present with nausea or vomiting but symptoms of abdominal fullness, distention or pain are often described. Disorders of colonic motility or defecation may cause distention and pain but often are associated with constipation and/or incontinence. Severe small bowel and colon motility abnormalities result in bowel obstruction. Less severe motility disorders may overlap with behavioral disorders making diagnosis and treatment challenging.

The objectives of this presentation are:
1. To briefly review the types of disorders of gastrointestinal motility that occur in children.
2. To discuss the approach to the evaluation and treatment of severe disorders of gastrointestinal motility.

Esophageal motility disorders
Esophageal Motility disorders are rare in children except for gastroesophageal reflux disease. The most common esophageal motility disorder is associated with congenital tracheo-esophageal fistula which occurs in 1 of 5000 births. Following surgical repair the distal esophagus uniformly has poor or absent peristalsis and, feeding and swallowing problems usually persists into adult life.

Achalasia is a rare disorder in children and symptoms include dysphagia, regurgitation, recurrent pneumonia, weight loss and chest pain. The overall incidence of achalasia is 0.4 to 0.6 per 100,000 with only about 5% of cases occurring in children. Most commonly, diagnosis of achalasia is suspected after radiographic contrast studies are performed for suspect symptoms. Esophageal manometry is useful to confirm the diagnosis. Introduction of high resolution manometry has enabled visualization of esophageal motility as a spatial continuum along a given length of the esophagus and using closely spaced manometry pressure sensors. This has eliminated the problem of movement related artifact and made esophageal manometry easier to evaluate and less uncomfortable for children. Prior to definitive therapy, obstructive lesions should be ruled out by performing upper endoscopy. Medical treatment of achalasia has been shown to be somewhat effective using Isosorbide dinitrate (5 mg) or nifedipine (20mg) and terbulatine (3mg). Long term management utilizing these medical approaches has not proven to be tolerable due to problems of compliance and/or complications of therapy.

Botulinum toxin has also proven effective but provides only transient relief and has little role for the treatment of achalasia in children. Pneumatic dilatation is a reasonable first choice of therapy, with a success rate of close to 50% for prolonged relief in children. The risk of pneumatic dilation is not well documented in children since there are inadequate case series. Selection of balloon size is particularly challenging in infants and young children. Laparoscopic myotomy may be reasonably considered as an initial procedure. Using intra-operative manometry can help guide surgery and has been associated with improved outcomes in adults.

Other esophageal motility disorders in children are rare and poorly documented.

Disorders of Gastric Emptying and Small Bowel Motility

Gastric emptying disorders are more common in infants and children. Premature infants have relatively poor peristalsis with poorly coordinated antroduodenal activity until 30 weeks gestation. Mature MMC patterns are not observed until after 34 to 36 weeks gestation. Anatomic obstruction due to congenital disorders such as antral or duodenal webs, intestinal stenosis or infantile hypertrophic pyloric stenosis, are far more common than primary motility disorders and must be excluded before considering primary motility disorders. Pseudo-obstructive disorders due to congenital gastrointestinal neuropathies or myopathies often present in infancy. Gastroparesis in children and adolescents is most often associated with a recent viral infection. It may be the presenting finding in children with evolving pseudo-obstructive disorders.

Accelerated gastric emptying
Rarely occurs in children, but it may be responsible for dumping syndrome. Most instances have followed surgery. Increased liquid emptying rates are seen after both vagotomy and proximal as well as distal gastric resection. After fundoplication for the treatment of gastroesophageal reflux, the reservoir capacity of the fundus is lost and gastric contents empty faster. Symptoms include abdominal discomfort, diaphoresis, pallor, lethargy, and diarrhea. Diagnosis is based mainly on symptoms and an abnormal glucose tolerance test following bolus feeding. Symptoms can be controlled by a low-carbohydrate diet, frequent small thickened feedings, and anticholinergic drugs. Often, symptoms in infants improve within 1 year of surgery, possibly corresponding with dietary introduction of solids.

Chronic intestinal pseudo-obstruction
Is a rare, severe, disabling disorder characterized by repetitive episodes or continuous symptoms and signs of bowel obstruction, including radiographic documentation of dilated bowel with air-fluid levels, in the absence of a fixed, lumen-occluding lesion. Chronic intestinal pseudo-obstruction (CIP) can be classified as primary or secondary. In primary CIP, the disease usually is limited to the hollow viscera, whereas in secondary forms, CIP is associated with an existing systemic disorder. Some conditions classified as primary CIP are associated with extra-gastrointestinal manifestations, involving the urinary tract or the autonomic, peripheral, and central nervous systems. As our understanding of gastrointestinal ontogeny improves, it is likely that more sophisticated histopathologic evaluation will delineate a spectrum of developmental abnormalities of gastrointestinal nerve and muscle that may result in variable degrees of gastrointestinal propulsive functions. For example, cases of pseudo-obstruction associated with deficient Interstitial Cells of Cajal have been described. Similarly, abnormalities of smooth muscle actins and defects in specific neuronal populations have been associated with bowel dysfunction.

Further sub classification as congenital or acquired CIP is based on the presence or absence of symptoms at birth. In most cases of congenital CIP, disease severity is greatest at birth, and tends to improve or plateau over the first months of life. In contrast, forms of non-congenital CIP present later in childhood or adulthood. Other non-congenital disorders may be acquired through infection or immune-mediated inflammation within the myenteric plexus. Acquired forms of CIP may worsen or improve over time. In most kindreds presenting with CIP, those affected develop symptoms requiring medical care after childhood, and tend to worsen with time.

Symptoms include nausea, anorexia, dysphagia, regurgitation, vomiting, abdominal distension and pain, and constipation, depending on the regions of the gastrointestinal tract involved. Other symptoms may arise from secondary complications such as diarrhea from bacterial overgrowth or from involvement of other organ systems such as the urinary tract. Symptoms may be intermittent or persistent. In some kindreds, asymptomatic individuals may have clear radiographic signs of bowel dilation prior to onset of symptoms, indicating a discordance of symptoms and clinical findings.

In a patient with symptoms of gastrointestinal obstruction the evaluation begins with supine and upright plain radiographs of the abdomen. Documentation of intermittent or persistent bowel obstruction with characteristic radiographic findings of air fluid levels and bowel distention is required for the diagnosis of idiopathic CIP. In symptomatic patients without these radiographic findings, other diagnostic possibilities including functional abdominal pain, gastroparesis, aerophagia, functional constipation, and non ulcer dyspepsia should be considered. However, less severe disorders of motility may progress with time to a clear pseudo-obstructive process.

In chronic cases and after excluding mechanical obstruction, the secondary causes of pseudo obstruction should be considered. In immunodeficiency states, including those induced by medication following organ transplantation, symptoms of CIP prompt an evaluation for myenteric plexus neuritis due to cytomegalovirus or Epstein-Barr virus. Screening blood tests for, connective tissue and skeletal muscle disorders (antinuclear antibody panel, creatine phosphokinase, aldolase), and hypothyroidism should be performed. Other tests to consider include serology for Chagas disease, urinary catecholamines to rule out pheochromocytoma, urinary porphyrins and enteric neuronal autoantibodies, and mitochondrial DNA testing for possible mitochondrial neurogastrointestinal encephalomyopathy (MNGIE).

Antroduodenal manometry and full thickness bowel biopsies may aid in clarifying the pathophysiology and histology, respectively. These tests are most useful when the diagnsosis is uncertain. If symptoms and signs of pseudo-obstruction persist for more than 2 months after birth or for more than 6 months in a child presenting after birth, a firm diagnosis of idiopathic CIP is warranted.

Treatment of pediatric pseudo-obstructive disorders must focus on providing optimal nutrition by enteral or parenteral routes. When gastroparesis prevents oral or intragastric tube feedings, a trial of post-pyloric feedings is occasionally successful. Even when parenteral feeding is required, at least small volumes of enteral feeding is desirable to prevent cholestatic liver disease and to maintain bowel mucosal integrity. Bacterial overgrowth, a frequent complication of pseudo-obstruction, can cause malabsorption. It may also be associated with an increased mucosal permeability to macromolecules and with bacterial translocation across the bowel. Intermittent treatment of bacterial overgrowth provides symptomatic relief and may decrease the risk of hepatobiliary injury and sepsis. Medical approaches to therapy including cisapride, metoclopramide, bethanechol, domperidone, erythromycin, octreotide and pyrodostigmine have all been reported to have some success. Bowel decompression by nasogastric suction or a gastrostomy is useful during acute exacerbations. Patients with frequent exacerbations or chronic distension often benefit from placement of an ileostomy. Small intestinal transplant may offer a potential for cure in children with life-threatening disease due to bowel dilation and recurrent sepsis, or in those with serious complications of parenteral nutrition including liver failure or limited central venous access.

Chronic Intractable Constipation and Disorders of Defecation
Constipation is the chief complaint in 3% of all general pediatric outpatient visits. Up to 25% of children referred to pediatric gastroenterologists have a disorder of defecation. Constipation can be defined on the basis of the frequency of defecation (< 3 stools/week); size of stool; consistency of stool or discomfort/pain in passage of bowel movements. Fecal incontinence can result from overflow around a rectal fecal mass or passage of bowel movements without awareness or the inability to prevent passage of a bowel movement. In pre-pubertal children, constipation is more frequent in young boys than young girls (3:1 ratio) but after puberty constipation is about 3 times more frequent in females than males. The incidence increases when a parent, a sibling or a twin is constipated. Constipation in both twins is four times more likely in monozygotic than in dizygotic twins, suggesting a genetic predisposition. Fecal incontinence occurs in about 3% of 4 year old and 1.5% of 10 year olds children.

The physical examination in a child presenting with constipation should include careful evaluation of the position of the anus (anterior location), examination for hints of sacral dysraphism (pigmented or hairy patch over the lumbosacral spine), and a careful examination of sacral dermatome sensation (light touch with cotton wisp) and reflexes (cremasteric and anosphincteric reflexes) should be performed. Digital rectal examination is essential to determine if there is anal stenosis, a mass or a fecal impaction. In patients with painful defecation, examination should focus on causes such as dermatitis, anal fissure or a patulous anus (suggestive of sexual abuse).

In the majority of children with constipation the cause is a functional or behavioral problem. A majority of these patients improve with dietary changes, stool softeners and/or stimulant laxatives, behavioral modification therapy.

Hirschsprung's Disease:
In contrast to the high incidence of constipation in the pediatric population (3 to 15 per 100), the incidence of Hirschsprung's disease is only 1 in 6,000 births and Hirschsprung's disease is found in less than 1% of children with constipation. The incidence of anorectal malformations is 1 in 5,000. Other organic causes of constipation in children are even less common. Certain features shown in table 1 are useful to discriminate children with possible Hirschsprung's disease from functional constipation and fecal retention. Anorectal manometry and suction or full thickness rectal biopsy can be used to diagnose Hirschsprung's disease.

Symptom	Functional Constipation	Hirschsprung's Disease
Delayed meconium passage	rarely	60%
Constipation as newborn	rarely	almost always
Onset after age 2 years	common	sometimes
Fecal incontinence	common	almost never
Withholding behavior	common	rare
Stool in rectal ampulla	common	rare
Obstructive symptoms	rarely	common

Defecation problems following pull through surgery for Hirschsprung's disease have been reported in up to 60% of children. Persistent constipation can result from retentive behavior due to painful defecation following surgery, anal sphincter abnormalities which persist despite surgery and dysmotility in the histological normal appearing bowel. Fecal incontinence is also a common symptom after pull through surgery. It can result from the loss of recto sigmoid region, the normal storage area for stool. The high amplitude colon contractions which are normal children are associated with a bowel movement, are more frequent and propagate right up to the anal canal after pull through surgery. This causes rapid colon transit, fecal urgency and frequent bowel movements. The treatment includes a low residue diet and loperamide to slow the bowel transit.

Internal anal sphincter achalasia:
This condition is associated with chronic constipation and absent recto-anal inhibitory reflex during anorectal manometry evaluation and a normal rectal biopsy. Most patients require laxative treatment and Botox injection into the anal sphincter can help. The effect of Botox injection can wear off in 6-8 weeks and if symptoms keep relapsing after anal sphincterotomy can be considered.

Severe, idiopathic constipation:
In children without an underlying etiology for constipation and with debilitating symptoms it is important to more aggressively rule out anatomic obstruction with a barium enema. If Hirschsprung's disease is suspected, rectal biopsy or anorectal manometry is useful to rule out the disorder. Colonic transit studies using Sitz markers may provide some indication regarding whether there is a primary defecation disorder or a more generalized "slow transit" disorder of the colon. Manometric studies of colon function may provide an explanation for symptoms. Characteristic High Amplitude Propagating Contractions (HAPCs) are easily recognized after meals and following the administration of bisacodyl. If surgery is being considered (ileostomy, partial colectomy, ileoanal pull through), it is useful to rule out a more extensive gastrointestinal motility disorder which portends poor outcome after surgery. Trials of therapy with a PEG-balanced electrolyte solution, aggressive use of laxatives, bisacodyl, and other prokinetic agents may be indicated. In some children, the placement of a cecostomy for irrigation may be effective.

As for pseudo-obstruction, specific defects in the development of neuronal cell populations (Substance P neurons) or Interstitial Cells of Cajal have been associated with cases of intractable constipation in children. Similarly, myopathies may present with severe constipation in children.

Advances in the investigation of functional abdominal pain

Qasim Aziz, Professor of Neurogastroenterology, London

Neurogastroenterology Group, Centre for Digestive Diseases, Blizard Institute of Cell and Molecular Science, Barts and the London School of Medicine and Dentistry, Queen Mary, University of London, UK.

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

Due to recent advances in neuroscience, it is now possible to study the neurophysiological response to visceral pain in health and patients with FGID. This can be achieved by using brain imaging techniques such as functional Magnetic Resonance Imaging [fMRI], Positron Emission Tomography [PET], Cortical Evoked Potentials [CEP] and Magnetoencephalography [MEG]. Using these techniques it has been possible to identify the spatio-temporal distribution of the visceral pain neuromatrix in health. Furthermore, it has been demonstrated that this neuromatrix is modulated by psychological factors such as attention, anticipation and emotions. In FGID patients, these brain imaging techniques have made it is possible to differentiate between patients with visceral hypersenstivity due to sensitised afferent pathways and those with heightened cognitive or emotional response to the stimulus.

As our understanding of the mechanisms of visceral hypersensitivity grows in patients with functional abdominal pain, we will be better able to tailor treatments according to the pathophysiology of symptoms.

Advances in the management of intestinal failure associated with Liver Disease

Professor Olivier Goulet, Professor of Paediatrics, Paris

UK small bowel transplant experience

Sue Beath, Consultant Paediatric Hepatologist, Birmingham

Birmingham Children's Hospital started the first small bowel transplantation programme in the UK in 1993. Initially around 10 children per year were assessed and two thirds fulfilled the criteria for intestinal transplantation, but only 1-2 children per year were transplanted because of a shortage of size matched donors. This shortage of small size appropriate donors led to a high mortality of (65%) on the transplant waiting list from intestinal failure associated liver disease, but did provide data on risk factors and led to clearer criteria for referral and timing for small bowel transplantation.

In 1998, the surgical team was augmented by Prof Jean de Ville de Goyet, who developed a new technique for successfully reducing the size of donor organs. This technique increased the size of the donor pool, by making it possible to consider donors up to 3 times the size of the patient. This had the effect of reducing the waiting time and mortality on the transplant list, and also led to an increase in the number of referrals and increased activity for the programme in all areas.

In the 1990s, the outcome for small bowel transplantation, which was usually combined with liver transplantation, was still relatively poor with just 50% of recipients surviving for 2 years or more after transplantation (compared with 90% 5 year survival for liver transplantation). The main reason for this is the need to use approximately twice the amount of immune suppression to suppress rejection in the small bowel allograft than would be needed in a liver graft alone. This meant that the risk of opportunistic infections such as EBV and graft rejection were much higher in small bowel transplant recipients. Another important factor was the robustness of the candidates since the majority had advanced liver disease at the time of listing, and deterioration whilst waiting for transplantation was the norm.

To manage these higher complication rates we have developed a rigorous and systematic monitoring protocol for out-patients in which the results of immune suppression levels, biochemical profiles and intestinal graft function were reviewed every week by a specialist nursing team, dietician and doctor. We also set up study days to support and educate colleagues who would be managing the child once she/he was discharged home. This allowed the team to be more pro-active in reducing immune suppression or being in a position to arrange earlier clinic review. Although labour intensive, this approach, combined with the advent of monoclonal antibodies active against interleukin-2 (Basiliximab and Daclizumab), and continuing improvements in surgical techniques and anaesthesia have improved the 2 year survival to 80% and internationally there are now many children and young people who have survived 15 years or more. Currently the survival after primary small bowel transplantation (without a liver graft) is 100% in the past 2 years. The longest survivors in the Birmingham programme are now approaching their 12th year since transplantation and several are preparing to move over to adult services.

Since 1993, we have assessed over 300 children for possible small bowel transplantation and performed 69 small bowel transplant operations. The current challenges for us remain a shortage of size appropriate organs, the vulnerability of children with chronic intestinal failure to sudden deterioration, the task of identifying rejection in patients once they leave hospital before they become unwell again and the goal of launching our young patients in productive and independent adult lives. The learning curve remains steep but is not perhaps as vertical as it once seemed.

Debate:

Bowel lengthening for short bowel syndrome – To STEP or not to STEP

Mr Antonino Morabito - For

Patients with short bowel syndrome surviving on parenteral nutrition but who retain even a relatively small amount of residual autologous bowel, can realistically hope for enteral autonomy following a structured program of Autologous Gastro-intestinal Reconstruction (AGIR).

Initial bowel surgery should be as conservative as possible. Once the patient is stable with a healthy liver, structured bowel reconstruction commences with Bowel Expansion over several months to be followed by

Autologous Intestinal Reconstruction
It is now possible through a Combined-AGIR program, to offer most patients with short bowel the prospect of a quality-life and total or partial enteral autonomy on their own bowel. In the event of failure and if life on total or partial TPN is unacceptable, then bowel transplantation is a reasonable but high risk option.

Mr Colin Baillie – Against

The success of longitudinal intestinal lengthening and tapering (LILT) in selected patients is beyond dispute. However, the devil is in the detail and in the case of LILT the detail is patient selection. Intestinal adaptation occurs by lengthening, dilatation, changes in mucosal morphology and function, and quantitative and qualitative changes in colonic fermentation. Full weaning from parenteral nutrition has been described with only 25 cm of small bowel, and becomes increasingly likely with >50cm residual small bowel. Adaptive capability is positively correlated with the presence of the ileocaecal valve and with residual colonic integrity. Other factors such as the underlying disease process and quality of residual small bowel influence adaptive potential.

Full adaptation may take up to eighteen months. During this period the critical management issues relate to preserving central venous access and preventing irreversible changes in liver function. Developments in the formulation and administration of TPN regimens over time, and improved enteral feeding strategies have greatly reduced the incidence and severity of TPNAC. Despite this, a small percentage of children with short bowel syndrome (SBS) will develop liver failure or loose vascular access before being able to benefit from the adaptive potential of their residual bowel. These children can only be salvaged by transplantation.

Identification of this sub-group of children before the development of these life-threatening complications might enable LILT or other surgical interventions to speed the adaptive process. These surgical options are however not without risk and have an appreciable complication rate. The issue for debate is when and how to apply a "bowel expansion programme" in the setting of SBS, given the considerable heterogeneity of the patient population, and lack of widespread surgical familiarity with these techniques. Under use of this type of surgery or its late application will undoubtedly prevent the salvage of some children destined for transplantation or death. However, it is likely that some children undergoing LILT could achieve full adaptation without surgical intervention.

Stem Cell transplantation in an animal model for Hirschsprung's Disease:

Mr Simon Kenny, Consultant Paediatric Surgeon/Urologist, Liverpool

Hirschsprung'sdisease affects 1 in 5000 newborns and is caused by an absence of ganglion cells in a variable length of the distal gut. It commonly presents in the newborn period with life-threatening bowel obstruction requiring surgery. Despite apparently successful surgery the long-term outcomes are often unsatisfactory with some children facing a lifetime of continence issues or debilitating constipation. In this presentation the reason for this are examined and advances that have occurred in the surgical management of the disease are described. In the last two decades rapid progress has been made in understanding the genetics and molecular pathology of Hirschsprung's disease. The potential for harnessing this knowledge to develop a stem cell based therapy for Hirschsprung's is described.

Bone Marrow transplatation for paediatric gastrointestinal diseases

Dr Mamoun Elawad

Debate:

Anto TNF therapy - Is it the Panacea?

Dr David Wilson - For

Dr Huw Jenkins – Against

Fish oil based parenteral nutrition. Do we need to go this way?

Dr Girish Gupte, Birmingham

A Medical Director's perspective on the current NHS reforms

Dr Steve Ryan

Recent NHS History can be defined from the incoming Labour government in 1997. This was around the time of the 50th anniversary of the NHS. Labour established the first 10 Ten Year Plan for the NHS which looked to increase capacity and capability by investment. They looked to increase the amount of GDP expenditure on health significantly and by enough to meet international comparators. Targets were introduced (98% within 4 hours in Accident and Emergency, all patients seen and treated within 18 weeks, and 2 week referral times for cancer. Many medics were initially sceptical about the benefits of this – but not many would put up with those waits for themselves or their families. There have been very significant improvements as a result. It was acknowledged that many clinicians felt disenfranchised and disempowered in the new NHS. Calls began to emerge for clinical leadership and engagement in senior decision making and consequently leadership frameworks, networks and support emerged. The Clinical Leader's Network in the North West was one of the first.

Then came the 60th Anniversary and the opportunity to develop a new 10 year vision. The Next Stage Review led to the development of the national policy document – High Quality Care for All – which supported regional strategy documents such as Healthier Horizons in the North West. This was a clinically-led, clinically engaged process, led by a surgeon Lord Darzi. It focussed on what clinicians said they wanted to see; that was not lots of new hospitals – it was clinical quality and high standards. Lord Darzi divided quality into safety, effectiveness and experience. His review put in place the enablers to help clinicians and clinical teams to deliver the vision. He then placed and emphasis on innovation – it is now mandated. He also defined what it is to be a professional in this era – a professional, a partner and a leader.

Then came…. Lehamn Brothers and the Royal bank of Scotland! And suddenly there was a need to deliver all of this – for less money. And it was quickly realised that clinicians held the key. After all, whatever governments and health care managers do – it is ultimately clinicians at the front line who direct the resources. The came the newest campaign: QIPP – Quality, Innovation, Productivity and Prevention.

So clinical teams are at the front line clinically – but will need to be at the front line for business too. They will be increasingly given freedom and responsibility to deliver the objectives of the NHS. Members of BSPGHN will be at the forefront of this for their services, building on the quality systems they have already established and utilising systems more familiar to managers to deliver their aims. Are you up for it?

ABSTRACTS FOR FRIDAY 29TH JANUARY 2010

Plenary abstracts

Rapid increase in the incidence of paediatric inflammatory bowel disease in the Republic of Ireland.

Raveen Shahdadpuri, Marion Rowland, Shoana Quinn, Annemarie Broderick, Tim Bohane, Mary Hamzawi, Billy Bourke; Children's Research Centre, Our Lady's Hospital, Crumlin

Background:
Recent reports suggest a worldwide increase in the incidence of paediatric inflammatory bowel disease (IBD), especially in industrialized nations.

Aims:
To describe the incidence of paediatric IBD in the Republic of Ireland between 2000 and the first half of 2009, and to compare it with regional and worldwide trends.

Methods:
The Department of Paediatric Gastroenterology at Our Lady's Children's Hospital Crumlin and the National Children's Hospital Tallaght provide the national referral service for paediatric IBD in the Republic of Ireland. Newly diagnosed cases of paediatric IBD in children less than 16 years of age were included. Population data for the Republic of Ireland was obtained from the latest national census in 2006 conducted by the Central Statistics Office Ireland.

Results:
The Irish population aged less than 16 years was 922 767. During the 9 ½ year period from 1st January 2000 to 30th June 2009, a total of 316 new cases of paediatric IBD were diagnosed. There were 187 incident cases of CD, 88 incident cases of UC and 41 incident cases of IC. The incidence of all forms of paediatric IBD rose markedly across the period studied. In 2000, the incidence rates for paediatric IBD, CD, UC and IC were 3.1, 2.3, 0.7 and 0.1 per 100 000 population per year, respectively. By 2008, these figures had risen to 6.3, 3.6, 1.3 and 1.4 per 100 000 population per year, respectively. 2009 incidence levels are on track to at least equal 2008 levels.

Conclusion:
The overall incidence of paediatric IBD in Ireland has increased significantly over the course of the study period. In fact, total paediatric IBD incidence rates more than doubled from 3.1 in 2000 to 6.3 in 2008, and corresponded to increases in all sub-types of IBD. This is comparable to similar trends seen elsewhere in the British Isles, Sweden, North America and Australasia. However, the speed with which this increase has occurred in Ireland is unique.

Reduction of Sepsis rate after introduction of 2% Chlorhexidine wipes for the care of central line in children with intestinal failure on parenteral nutrition

Judith Pichler, Venetia Horn, Sarah MacDonald, Dr. Susan Hill
Great Ormond Street Hospital, London

Introduction:
Line sepsis is a common and serious life- threating complication of parrenteral nutrition (PN) therapy in children. Different aseptic techniques for accessing the line is an essential part of the care of the child. The aim our study was to investigate the outcome of sepsis after introduction of 2% Chlorhexidine in 70% isopropanol wipes.

Methods:
143 children received PN for > 28 days at Great Ormond Street NHS Trust in 2006 and 2008. Age, sex, underlying disease and survival were recorded and analysed. Complications like sepsis, number of sepsis episodes, and aetiology of sepsis were analyzed before and one year after the introduction of 2% Chlorhexidine wipes and complication such as intestinal failure associated liver disease (IFALD) definition of BSPGHAN Intestinal Failure Working Group.

Results:
In 2006 66 children (44% males) with a mean age at start of PN of 2.9 year (birth-12.8yr) received PN for > 28 days. The mean duration of PN was 70 days (28-418 days). In 2008 78 children (49% males) with a mean age at start of PN of 3.3 year (birth - 17,1yr) received PN. The mean duration of PN was 84,6 days (29-247 days).

In 2006 49 (77.8%) patients had a sepsis with a mean of 2,2 sepsis episode in the time of PN (0-12), after the introduction of 2% Chlorhexidine wipes there was a significant decrease in the incidence of sepsis. In 2008 39 (51.3%) patients had a positive blood culture with a mean of 1 sepsis episode (0-7, $p<0.05$). There was no difference in the aetiology of sepsis, most common gram- positive pathogens were Staphylococcus spp. and Enterococcus spp. Gram- negative pathogens were Klebsiella spp. and Enterobacter spp. 22 (15.3%) children developed IFALD, 11 in each group (n.s.). In each group 7 with IFLAD type I, 3 with type 2 and 1 with IFALD type 3.

Conclusion:
After the introduction of a new aseptic accessing technique with 2% Chlorhexidine wipes the sepsis rate of children on long term PN could be significantly decreased.

No difference in the pathogens could be seen. The reduction of infection did not have any influence on the occurrence of IFALD or the severity of liver disease.

Comparison of quick point of care test for small bowel hypolactasia with biochemical lactase assay

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Background:
The usefulness of a new quick test for endoscopic diagnosis of paediatric-type hypolactasia was tested in duodenal biopsies. In this test, an endoscopic biopsy from the postbulbar duodenum is incubated with lactose on a test plate, and a colour reaction develops within 20 min as a result of hydrolyzed lactose (a positive result) in patients with normolactasia, whereas no reaction (a negative result) develops in patients with severe hypolactasia.

Aims:
The aim of this study was to compare the Biohit® Lactose Intolerance quick (BLIQ) Test to the "gold standard" biochemical duodenal lactase (DL) activity assay in the paediatric population.

Patients And Methods:
Two postbulbar duodenal biopsies were taken from 38 prospective children (0-16 years) who underwent upper GI endoscopy over a period of 1year [June 08-May 09] at a single tertiary paediatric gastroenterology unit. The biopsies were used for the Quick Lactase Test (Biohit® PLC, Helsinki, Finland) and in biochemical disaccharidase (lactase, trehalase, sucrase, and maltase) assays.

Results:
38 children (19 male) of median age 5.45years (0.3-14.8 years) had the combined testing. We further subdivided this group into those children that had their biopsies with a larger endoscope [XQ,n=26] and thus a bigger biopsy forcep and those children that had a smaller endoscope [XP, n=12] and thus a smaller biopsy forcep. The results are tabulated below.

USING SCOPE XQ, n=26 Mean weight of biopsies=0.003 gm	**DISSACHARIDASE ASSAY**	
	POSITIVE (normolactasia)	**NEGATIVE (hypolactasia)**
Biohit +ve (normolactasia)	4	3
Biohit –ve (hypolactasia)	0	19
Sensitivity 4/4=100%		Specificity 19/22=86%
Positive predictive value 4/7 =57.1%		Negative predictive value 19/19=100%

USING SCOPE XP, n=12 Mean weight of biopsies=0.002 gm	**DISSACHARIDASE ASSAY**	
	POSITIVE (normolactasia)	**NEGATIVE (hypolactasia)**
Biohit +ve (normolactasia)	2	2
Biohit –ve (hypolactasia)	0	8
Sensitivity 2/2=100%		Specificity 8/10=80%
Positive predictive value 2/4=50%		Negative predictive value 8/8=100%

Conclusions:
The Quick Lactase Test effectively identifies children with severe duodenal hypolactasia. These results are based on small numbers but tend to support findings in adult studies. The sensitivity and negative predictive value of the BLIQ was 100% on comparing it to DL. The specificity too appears to be high but variable (86% in XQ & 80% in XP groups). This would suggest a lower specificity perhaps, secondary to smaller size of the biopsies obtained and may warrant the need for 2 biopsies. In comparison with biochemical lactase assays, the sensitivity and specificity of BLIQ for indicating hypolactasia is very high and appears to be an effective point of care test for paediatric hypolactasia.

Fatty Liver Disease – Is further investigation necessary?

Dr. B. Krishnamurthy[1], Dr. A. Urs[1], Dr. J. Stahlschmidt[2], Dr. P. McClean[1].
Department of Paediatric Hepatology[1] and Histopathology[2], St. James' University Hospital, Leeds

Introduction:
Now-a-days non alcoholic fatty liver disease (NAFLD) is one of the most common causes of liver disease in adults and children. In most children it is asymptomatic and presents with abnormal liver function tests and/or ultrasound scan. However it can lead to cirrhosis even in childhood. The problem for paediatricians is how far to investigate these children as other metabolic conditions can lead to hepatic steatosis. The aim of this study was to examine the clinical and laboratory features of children with histologically proven steatosis in order to gather evidence for investigative guidelines.

Methods:
The casenotes of all children with histologically proven steatosis, diagnosed between January 1996 and August 2009, were reviewed. The study group was identified by an electronic word search of histology reports, and referencing the unit database. Mode of presentation, clinical features including height and weight, laboratory data and final diagnosis were recorded.

Results:
A total of 56 children (m=33) were identified. In 29 children the BMI was < 91st centile for age ("Normal"). Twenty three (79%) of these had a metabolic disorder (including 5 with Wilson's disease), steatosis was due to medication in 2 children, NAFLD in 1 child and 3 had other miscellaneous diagnoses.

Twenty seven children were identified to be overweight/obese (BMI ≥ 91st centile). Sixteen (59%) were diagnosed with NAFLD, 5 had steatosis secondary to medications, 3 had a metabolic disorder (including 1 with Wilson's disease) and 3 had other miscellaneous diagnoses.

Excluding the 14 children who presented with decompensated liver disease the table compares the laboratory and histological features of the children with NAFLD to those with other diagnoses.

Laboratory median (range)	**NAFLD (n=17)**	**Other diagnoses (n=25)**
Bilirubin µmol/l	7 (4 – 19)	9 (2 – 190)
ALT IU/l	90 (22 – 407)	113 (30 – 581)
GGT IU/l	42.5 (14 – 80)	60.5 (10 – 629)
Histology n (%)		
Steatosis only	6 (35%)	5 (20%)
Inflammation	5 (29%)	7 (28%)
Fibrosis	9 (53%)	16 (64%)

Conclusions:
1. 40% of children in the obese/overweight group had diagnoses other than NAFLD.
2. Children in the normal BMI range were more likely to have other causes for fatty liver disease.
3. While children with NAFLD only had raised ALT at presentation there was considerable overlap with the children with other diagnoses.

Children presenting as fatty liver disease can have a wide variety of possible underlying diagnoses and therefore thorough investigation is important even in overweight /obese children.

Clinical Outcome in Cystic Fibrosis Patients with or without Meconium Ileus: A Comparative Study

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Aims

Meconium ileus (MI) is a form of neonatal intestinal obstruction due to an abnormal viscid meconium within the terminal ileum. MI is the presenting symptom in 15-20% of patients with cystic fibrosis (CF). Approximately half of these patients present with complex MI. The aims of the present study were to assess the clinical outcomes in cohorts of patients with complex meconium ileus at 5, 10 and 15 years in comparison to CF patients without MI.

Methods

CF patients presenting to our centre with MI were reviewed. Data on gestational age, weight, type and extent of surgery, duration of parenteral nutrition were recorded. Age, sex and genotype-matched controls were used for comparisons. In both groups, clinical status at 5, 10, and 15 years records were recorded from annual review records.

Results

A total of 23 (9 females) infants with MI were identified. Overall survival for both simple and complex MI – 92%. Of these, 75% had complex MI; 80% were born at term; and 50% were homozygous for χF508. 11 patients received TPN for median of 24 days (10-120 days).

In patients with complex MI, the mean weight, height, BMI% for age (>50%), FEV1, FVC, abnormal live scan, Schwachman score at 5 years were 16.9 kg, 106.6 cm, 29.4%*, 81.6%, 78.6%, 27*, 80; at 10 years were 27.4 kg, 133 cm, 40%, 68%, 83%, 70%*, and 75; at 15 years were 43.3 kg, 155.2 cm, 33%*, 74%, 84%, and 60

Of the controls, the mean weight, height, BMI% for age (>50%), FEV1, FVC, abnormal live scan, Schwachman score, at 5 years were 18.1 kg, 107.8 cm, 55.3%, 87 %, 92%, 44% and 86 (75-92); at 10 years were 31.7 kg, 136 cm, 53%, 80%, 88%, 33% and 82; and at 15 years were 48.5 kg, 154 cm, 54%,82%, 89%*, 58%, and 70 respectively.

*p≤0.05

Conclusion

Compared with non-MI controls, children surviving complex disease tended to be smaller with lower BMI's at all age points. Lung function (FEV1) was also worse. A higher % showed abnormalities on liver scans. Conflicting results from earlier studies may reflect the % of simple MI in the data sets.

With this limited study clear trends are emerging showing that the outcome for infants with complex MI is poorer both in terms of growth and lung function

The Practical Outcomes of Thiopurine Metabolite measurement: The experience of a tertiary PGHAN unit

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Introduction:

The Thiopurines, 6-Mercaptopurine (6-MP) and Azathioprine (AZA) are potent immunomodulators, effective in maintaining remission in paediatric inflammatory bowel disease (IBD). The concentration of the active metabolite, 6-thioguanine nucleotide (6-TGN) is dependent on Thiopurine Methyl Transferase (TPMT) activity, an enzyme with common genetic polymorphisms. Meta analysis of data shows that clinical response to medication is significantly higher with 6-TGN levels above 230-260 pmol/8 x 108 red blood cells1. Because metabolite monitoring is not commonplace in the UK, some clinicians use proxy measures in routine blood monitoring to assess response to thiopurines.

Aims:

To present our experience of using thioguanine metabolite measurement in the management of patients attending a tertiary PGHAN service. To evaluate how use of these measurements affects clinical practice.

Methods:

6-TGN and 6-MMP was measured in children attending a PGHAN service who had been on thiopurine medication for at least 3 months. Our practice is to prescribe AZA at 2-2.5mg/kg, and 6-MP at 1-1.5 mg/kg. For each measurement data were collected on dosage, disease severity, concomitant use of 5-ASA, haematological and biochemical indices, and changes to management. Therapeutic 6-TGN levels were defined as 235-450 pmol/8 x 108 red blood cells. Toxicity is defined as WBC <4.0 x103mm, neutrophils <2.0 x 103mm, AST/ALT > 2 x upper limit of normal.

Subjects:

64 individuals (28 males) were included in study, median age 14 years (inter-quartile range 12-16). Underlying diagnoses were 'IBD' (54/64) and 'other' (10/64). 59 were treated with AZA, 5 with 6-MP. Median doses (inter-quartile range) for AZA and 6-MP were 2.1mg/kg (1.8-2.4) and 1.1mg/kg (1.0-1.3) respectively. A total of 95 separate measurements were made.

Results:

TPMT phenotype was measured in 51/64 individuals. 40/51 individuals had 'normal' phenotypes, and 11/51 were heterozygotes for TPMT gene mutations. Initial 6-TGN levels were significantly higher in heterozygotes (median levels 836 vs. 328 respectively, p=0.001) for comparable dosages (median 1.9 vs. 2.2 mg/kg, p=0.11), before dose adjustment had taken place.

Only 39% patients had 6-TGN levels within therapeutic levels on first measurement. 30% were sub-therapeutic (including 3 measurements of 0), and 31% were supra-therapeutic. 9% had 6-TGN levels > 800, all were heterozygotes for TPMT.

Toxicity occurred in 8 cases. In 7 of these cases, 6-TGN levels were supra-therapeutic (median 668, range 296-2172).

Concomitant use of 5-ASA did not significantly affect 6-TGN levels at comparable doses (median 6-TGN level 'on 5-ASA' 393 vs. 451 'not on 5-ASA', p=0.26).

In total, management was changed in 39 of 95 cases (41%). Medication dose was altered or stopped in 27 cases, and changed entirely in 8 cases. 6-TGN levels exposed 6 cases of non-compliance. Of the 39 cases of change in management, 33 were adjudged to be exclusively or predominantly influenced by knowledge of the 6-TGN level.

Conclusion:

Based on standard dosing regimens, clinicians can expect to achieve therapeutic levels of 6-TGN in a minority of cases. Measuring thiopurine metabolites aids therapeutic dose alteration, allows early detection of potential toxicity, and identifies issues of non compliance that cannot be detected on routine blood monitoring. Thiopurine metabolite measurement can be a vital adjunct in managing children on AZA or 6-MP.

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ABSTRACTS FOR FRIDAY 29TH JANUARY 2010

Poster abstracts

"All that glitters is not gold": A case of misdiagnosis of celiac disease "
Akshay Batra, Susan Protheroe, Dept of Gastroenterology, Birmingham Children's Hospital

Coeliac disease is an immune-mediated enteropathy caused by a permanent sensitivity to gluten in genetically susceptible individuals and its diagnosis is based on presence of typical histological changes in duodenal mucosa. We present a case of a girl, who following weaning presented with symptoms and histological changes consistent with coeliac disease, but did not respond to gluten free diet. She was diagnosed with autoimmune enteropathy and improved on treatment for it.
TH was born by a full term normal delivery following an uneventful pregnancy. At birth she was on 50th centile for both her weight as well as height. She was bottle fed since birth and weaning was started at the age of 4 months. She first presented to her local hospital at the age of 4 months with a short history of loose stools which was diagnosed as acute gastroenteritis. TH failed to gain weight since starting weaning and continued with loose stools. She represented at the age of 8 months with hypocalcemic tetany with seizures and was also noted to be severely malnourished on admission. Coeliac serology done at the time showed elevated TTg, at 32 U/ml. The symptoms of persistent diarrhoea, weight loss and positive TTg prompted an endoscopy which showed a near total villous atrophy of duodenal mucosa with marked glandular hyperplasia associated with numerous intraepithelial lymphocytes. A diagnosis of coeliac disease was made and TH was started on gluten free diet.

On the Gluten free diet TH did not establish any weight gain and there was only slight improvement in diarrhoea. Because of incomplete resolution of symptoms on gluten free diet further investigations were done which showed TTg which had gone up to 83.7U/ml. Other investigations included normal liver functions, DCT negative anaemia, low free thyroxine with normal TSH and normal immunoglobulin. There were no auto antibodies or anti erythrocyte antibodies detected. T and B cell subsets were normal with normal functional antibodies. Sweat test was negative. TH evidence of iron deficiency with haemoglobin dropping down to 5gm/dl. There was developed transient neutropenia which resolved spontaneously. Following confirmation of compliance with gluten free diet as an inpatient, a repeat endoscopy was performed. It showed complete villous atrophy despite 2 months of gluten free diet, with crypt hyperplasia and intraepithelial lymphocytosis. TH also was dependent on parenteral nutrition to achieve a satisfactory weight gain. In view of failure to respond to gluten free diet and histological changes a diagnosis of autoimmune enteropathy was made which was confirmed by repeat assessment at Newcastle General Hospital. Treatment was started with iv methylprednisolone and tacrolimus maintaining levels between 8 -12 microgms/L.

TH responded to treatment with complete resolution of symptoms. She was able to wean off parenteral nutrition completely and continues to gain weight on gluten and lactose free diet. An endoscopy was repeated after 3 weeks of being on treatment which showed a significant improvement in histology. She is currently on tacrolimus with weaning doses of oral steroids and continues to be symptom free.

Conclusion:
Autoimmune enteropathy is a rare cause of intractable diarrhoea associated with circulating gut autoantibodies. Histological features of autoimmune enteropathy can be variable and a small subgroup of these patients can present with features indistinguishable from celiac disease. Therefore in children where there is lack of response to gluten free diet alternative diagnosis such as autoimmune enteropathy must be considered.

A New Method in the Diagnosis of Oesophagitis: Confocal Endomicroscopy

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Background and aims
Confocal laser endomicroscopy (CLE) enables surface and subsurface imaging of living cells in vivo at x1000 magnification and up to 250 µm below the tissue surface. In the oesophagus, the distance between the surface to papillary (S-P) tip can be measured using CLE. Gastro-oesophageal reflux (GOR) related oesophagitis causes papillary elongation on histology leading to a decrease in the S-P distance. We hypothesise that measuring the S-P distance by CLE is a valid tool to differentiate the normal from the inflamed oesophagus.

Patients and methods
7 patients (5 females) with a median age of 7.6 years (range 1.8 to 15.5), median weight of 23 kilos (13.2 to 71) and 16 controls with a median age of 12.0 years (2.2 to 15.3) and median weight of 38.2 kilos (10.7 to 83) underwent oesophago-gastro-duodenoscopy (OGD) using the confocal laser endomicroscope. The S-P distance of the oesophagus was measured for both the patients and controls by CLE and histology and corrected for height.

Results
The median confocal and histologic measurements for S-P distance corrected for height for patients were 0.19 µm/cm (range 0.10-0.49) and 0.58 µm/cm (range 0.29-0.76) and for controls were 0.44 µm/cm (range 0.20–0.93) and 1.07 µm/cm (range 0.76–0.1.57) respectively. Both methods were statistically significant with a $p = 0.019$ and < 0.001 respectively.

Conclusion
Measurement of the S-P distance by CLE is a new tool in the real-time diagnosis of GOR related oesophagitis during ongoing endoscopy. This study has shown that the S-P distance is significantly reduced in GOR related oesophagitis. Further large studies are needed to standardise measurements in order to adopt this technique for the in-vivo diagnosis of oesophagitis.

Achieving Enteral Autonomy Following Surgery for Short Bowel Syndrome

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Background:
Paediatric short bowel syndrome (SBS) carries significant morbidity and mortality. Patients undergoing bowel reconstructive surgery for short bowel syndrome have poor enteral tolerance immediately post surgery. The nutritional status is often maintained solely by parenteral nutrition (PN). PN is associated with severe complications despite its importance in SBS management.

Enteral feeds are started as early as possible to stimulate intestinal adaptation, which is a vital phase in achieving enteral autonomy. Published data is limited for the nutritional progression of these patients.

Objective:
To assess the nutritional and clinical outcomes of infants with intestinal failure following autologous reconstruction surgery and identify factors associated with the resumption of a normal dietary intake.

Method:
This was a retrospective study of 12 patients (58% males, gestational age range 24 – 38 weeks) undergoing autologous reconstructive surgery at Royal Manchester Childrens Hospital from 2000 to 2008. Aetiologies of SBS were gastroschisis, small intestinal atresia, mid gut volvulus, malrotation and necrotizing enterocolitis, Fifty percent had more than one congenital gastrointestional condition.

Results:
Post-operative survival rate was 92%. PN was required post surgery (mean 536 ± 708 days). Enteral autonomy was achieved in 7 out of 12 patients (58%) (mean 627± 568 days) while 3 patients continue to progress. One patient developed intestinal and liver failure due to PN-related liver damage and required transplantation surgery. Another patient died perioperatively as a result of multi-organ failure secondary to sepsis. More than half of the patients were able to maintain or gain weight to higher percentiles after surgery. Enteral tolerances improved in majority of patients.

Conclusion:
PN is required post reconstruction surgery for extended periods. Oral intake is gradually increased as tolerated. Potential factors associated with successful resumption of normal dietary intake include early eating experience, short duration on PN prior to surgery, early surgical intervention and using hydrolysed formula as the initial feed.

Conservative Management of Multiple Button Battery ingestion – a Case Presentation

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Background:
Foreign body ingestion is a common problem in childhood. Swallowed button batteries can cause significant pathology due to electrical discharge, metal contents and size. Complications such as oesophageal impaction, mucosal erosion, Oesophageal stricture, tracheo-oesophageal fistula1, spondylodiscitis, mediastinitis2, metal toxicity, bilateral vocal cord paralysis3 have been reported. When oesophageal impaction is present, common practice is to remove the battery endoscopically or surgically4. No recommendations have been made for the management of multiple button battery ingestion in the absence of oesophageal involvement.

Aim:
To demonstrate successful conservative management in the event of multiple button battery ingestion in a child in the absence of oesophageal impaction.

Subjects and Methods:
A 21 month old girl presented to the emergency department five hours after accidental ingestion of approximately 30 button batteries. Her clinical examination was unremarkable and she remained well apart from one episode of bilious vomiting. Abdominal X-ray showed multiple batteries scattered in the stomach. A few were seen in the small and large bowel. The oesophagus was clear.
The child was conservatively managed with IV fluids, prokinetic agents (Domperidone 400 mcg/Kg QDS, Erythromycin 3mg/KG QDS) and Klean-Prep. Serial X- rays revealed gradual reduction of the number of batteries. She was discharged home after five days with only one button battery visible in the rectum.

She remained well in herself and follow up abdominal X-ray four weeks later did not reveal any residual pathology.

Conclusion:
In the case of button battery ingestion an initial X-ray and thorough clinical examination will guide further management. In the absence of oesophageal impaction invasive procedures should be avoided, even if a large number of batteries are detected, as they can be passed spontaneously.

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2. Korean J Radiol. 2008 Nov-Dec;9(6):555-8
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Cows Milk Allergy (CMA) and Intolerance (CMI) – a DGH experience

Sophie Smith – Final Year Medical Student, Keele University; Hazel Duncan, Paediatric Dietician, Naeem Ayub, Consultant Paediatric Gastroenterologist; Royal Shrewsbury Hospital

Introduction
Cow's milk allergy (CMA) and Intolerance (CMI) are common conditions suffered by 2-6% of infants. However, the range of symptoms is diverse with diagnosis based on clinical judgement and dietary exclusion trials. Long term outcome also remains unclear.

Aims
To determine the presenting symptoms, management and outcome of a group of patients with Cow's Milk Allergy and Intolerance presenting at the Royal Shrewsbury Hospital, UK.

Methods and Subjects:
A retrospective study of all children referred to the paediatric dietician with a suspected diagnosis of Cow's Milk Allergy or Intolerance between 1st January 2008 and 30th April 2009 at the Royal Shrewsbury Hospital, UK.

Relevant data was extracted from the case notes, dietician notes and hospital computer systems, recorded on a standard proforma and then input into a Microsoft Excel XP database by a single researcher. Simple analysis was performed on this data.

Results:
The 90 children identified were aged between two weeks and 9.13 years (mean age 1.36 years) with a male: female ratio of 3:2. Almost half (42%) were under 6 months old. Diarrhoea (62%) and vomiting (33%) were the commonest symptoms. Constipation was found in as many as 19%. Other symptoms were possible colic (28%) and irritability (18%). Although non-specific rashes (21%) were noted, eczema (7%) anaphylaxis (1%) and wheeze (4%) were rare as was blood (9%) or mucus (6%) in the stools.

69% of the patients showed a complete resolution of symptoms with dietary exclusion of cow's milk while 22% showed a partial response. 76% of the partial or non-responders responded fully to a second dietary intervention, 22% responded partially and only one of the original cohort of 90 children remained fully symptomatic.

Hospital follow up dropped successively at 3 months (90%), 6 months (60%) and 12 months (33%) making long term outcome difficult to assess.

Conclusions:
Although diarrhoea and vomiting remain common symptoms in cow's milk allergy and intolerance, early constipation should prompt consideration of this diagnosis. Anaphylaxis is rare. Clinical suspicion combined with appropriate dietary exclusion appears a valid way of making a diagnosis of CMA and CMI.

Current home parenteral nutrition (HPN) practices in London: an overview of 5 main centres

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Introduction:
HPN is one of an increasing number of 'hi-tech' treatments that is given at home by parents in replacement to treatment in hospital. The major advantages are improved quality of life, including holidays and sporting activities, decreased risk of catheter sepsis and cost savings to the health service. It is estimated that there are currently about 100 children at home on parenteral nutrition (PN) in the UK (BSPGHAN data 2009). We reviewed our experience of HPN in London.

Method:
5 main London centres with severe intestinal failure children on HPN were identified in September 2009 (Chelsea and Westminster, Kings College, St Georges, Royal London, Great Ormond Street Hospital) and sent a questionnaire. HPN patient numbers, ages, medical diagnosis and methods of service were analysed.

Results:
London had 59 children <18yrs on HPN (M:F 27:32).29 were <5 years old. Among these patients motility (19) was the most common cause of intestinal failure, followed by short gut (17) and enteropathy (14).

All centres had a dedicated multidisciplinary team (nutrition nurse, PN pharmacist, PN dietician and paediatric gastroenterologist) used portable pumps, and a variety of lipid sources. Features that varied between units included content of training and technique for connecting and disconnecting the nutrition. One unit used a sterile technique and four an aseptic non touch technique. Two units used a one bag system and three used a two bag system for overnight infusions. Sepsis protocols and annual review investigations varied between units.

Conclusion:
Although all centres complied with the recommendation to have a multidisciplinary nutritional care team there were significant variations in the way in which PN was organised at home. It is important that there is effective communication between the separate teams if all patients are to receive the highest possible standard of care available and that the best practices are used by all.

Differences in costimulatory molecule gene expression on endoscopic biopsies from patients with ulcerative colitis

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Introduction
There is strong evidence that dysregulated interactions between T cells, antigen-presenting cells and the mucosa are important in the pathogenesis of inflammatory bowel disease. Abnormalities in T cell differentiation due to aberrant costimulation is one proposed mechanism leading to this abnormal interaction.

Aims and Methods
To compare intestinal costimulatory molecule gene expression in adult patients with ulcerative colitis (n=67) and controls (n=31). Paired endoscopic biopsies were taken from four specific anatomical locations for RNA extraction and histology. Amplified cRNA was analysed for 25 genes involved in T cell costimulation using the Agilent platform.

Results
Comparing all inflamed and non-inflamed biopsies from disease and control groups ten genes were significantly upregulated in ulcerative colitis including CD40, OX40 and ICOS; only CD18 was significantly downregulated. Controlling for anatomical location and inflammation status, four genes (CD86, CD80, OX40 and 4-1BB) were significantly upregulated in non-inflamed sigmoid ulcerative colitis biopsies compared to non-inflamed sigmoid control biopsies. In healthy control biopsies, 11 genes showed significant variation throughout the large intestine with the majority showing a significant decrease in expression distally. Comparing variation in healthy controls and disease biopsies, three genes showed a highly significant ($p<0.0001$) variation in healthy controls with loss of this 'normal' variation in ulcerative colitis biopsies for CD80 and 4-1BB; a further four genes also showed significant loss of variation in ulcerative colitis biopsies compared to controls. A total of six genes, including HVEM, ICOS and OX40L, showed significant variation in expression gradient in ulcerative colitis biopsies with differences between the ascending and sigmoid colon biopsies primarily driving the separation.

Conclusions
Although costimulatory molecule gene expression on endoscopic biopsies does not account for varying expression on different cells types, the variation observed between ulcerative colitis patients and controls is notable. Many of these genes are upregulated in ulcerative colitis and the normal expression gradient seen in the healthy adult colon is lost in disease. It is unclear if these differences are a result of significant dysregulation on certain cell lineages. Further work is required to determine the expression of these costimulatory molecules in inflammatory bowel disease to help aid identification of possible targets for future therapeutic intervention.

Conflict of Interest: None declared

Isolated small bowel transplantation- long-term survival rates are comparable to home parenteral nutrition

Muhammed R, Sharif K, Lloyd C, Beath SV, Hartley J, Hoggs L, Mirza DF, Gupte GL.
Liver Unit (including small bowel transplantation).

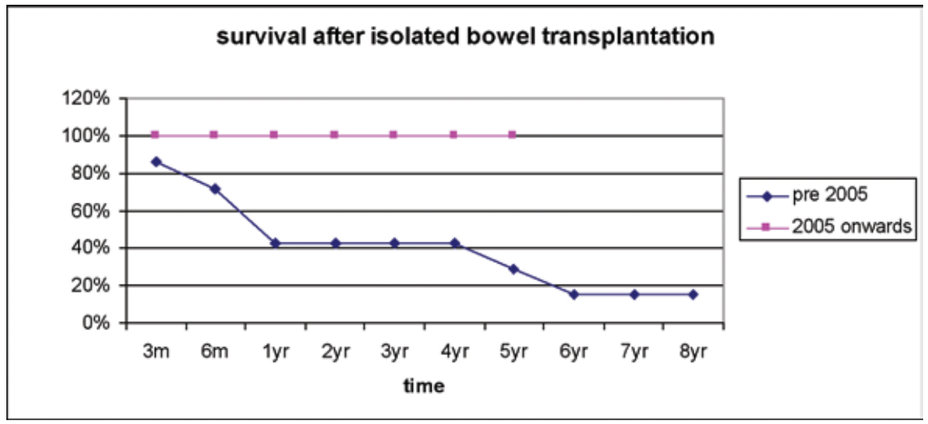
Background:
Five year survival rates of children with home PN are around 90%. Intestinal transplantation is used as a rescue therapy for children with life threatening complications related to long term parenteral nutrition (PN). Worldwide results of intestinal transplantation have improved with increased expertise in the surgical techniques and improved knowledge in the use of immunosuppression.

Aim:
We analysed the outcome of children who had undergone primary isolated small bowel transplantation in the last 5 years and compared these with our experience of intestinal transplantation prior to 2005.

Subjects and methods:
Retrospective review of the records of the children who had undergone primary isolated small bowel transplantation between 1993 and 2009. Children who had undergone re transplant of the small bowel were excluded from this study.

Results:
Since 2005, 10 children had undergone isolated small bowel transplantation, of which 8 were primary isolated small bowel transplants. The median age of these 8 children was 77 months (range 21-141). 3 children had short bowel syndrome, 3 had bowel dysmotility, and one each had microvillus inclusion disease and small bowel bacterial overgrowth refractory to medical treatment. Two children needed additional laparotomy before their discharge from hospital. All children were discharged home on full enteral feeds.

Prior to 2005, 7 children had isolated small bowel transplantation. 4 children had short bowel syndrome, 2 had microvillus inclusion disease and one had bowel dysmotility. The median age group in these children was 48 months (range 10-66). 5 children had died in this group and two children were listed for retransplantation of the bowel because of chronic rejection. The survival rate of both groups of the children is shown in Fig 1.



Conclusion:
The improved survival of the children with isolated small bowel transplantation is reflection of the increased experience and is comparable to survival rates following home PN. Intestinal transplantation may be offered as an alternative treatment to home PN in select patients in the future.

Percutaneous endoscopic gastrostromy in paediatric oncology: Experiences from specialists and patients.

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Introduction:
In paediatric oncology, malignancy and treatment can exert life-threatening effects on nutrition, metabolism, hydration, and immune response. Although weight loss has been reported to range from 8 to 60%, most treatment protocols do not include measures to improve enteral feeding. Insertion of percutaneous endoscopic gastrostomy (PEG) is a well established procedure in children, but there is little data about safety and benefits in paediatric oncology.

Aim:
To audit the changing practice and impact of PEG tube feeding and in paediatric oncology patients over a three year period in a tertiary centre (2006 - 2009).

Subjects and methods:
Retrospective analysis of all oncology patients who underwent PEG insertion, regarding indication of PEG insertion,usage of device , complications, blood count indices, and change in weight (feasible for assessment in 23/34 patients). In this cohort, infection was defined as growth of organisms from the PEG site. PEG insertion was performed either following weight loss for nutritional rehabilitation under chemotherapy, or prophylactically before start of the chemotherapy regime. Furthermore, a survey was performed by questionnaire to patients and parents.

Results:
Over 3 years, 34 patients had a PEG inserted at a mean age of 10.3 years, in 3 of the 34 patients this was electively done by laparotomy. Malignancy comprised of haematological (n=12), head and neck (n=12), or bone (n=10). With increasing experience over each year, nutritional rehabilitation (50%, 23%, 10%) was replaced by prophylactic (50%, 77%, 90%) for feeding prior to high dose chemotherapy. All patients used the PEG within 5 days of insertion. 17 patients (50%) had minor complications: local infection (n=13,), oesophagitis (n=2), local irritation (n=2), and incidential pulling out by the child (n=1), none resulted in major complications.

Of the (n=10), 8 patients gained weight with a mean increase of 4.75kg (95% CI: 2.95- 6.55kg; $p < 0.05$), mean 14.3% weight gain. Of the prophylactic group (n=13), 11 patients gained weight, with a mean increase of 2.7kg (95% C.I: 1.5kg- 3.85kg; $p < 0.05$), mean 12 % weight gain. None of the patients d blood transfusion as a result of the procedure. Anaemia (n=21), thrombocytopenia (n=6), or neutropenia (n=1) did not preclude the intervention.

All patients/parents (n=34) surveyed believed that PEG feeding had a positive impact on the condition of the patient.

Conclusion:
In our centre, the prophylactic use of PEG for nutritional support in paediatric oncology patients is increasing. In our experience, it is a safe procedure with the most common minor complication being local infection or erythema. For this cohort of patients, weight gain is substantial and has become an integral part of our therapy plans. In addition to these encouraging results, further potentially beneficial effects are under investigation and will be discussed.

Recto anal strictures in children with Crohn's disease

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Introduction:
Anal strictures are well recognized in adults with Crohn's disease (CD). Prevalence rates up to 7.5% have been reported, with average interval between onset of illness to development of stricture being 14 years. There is concern that treatment with Infliximab might promote stricture formation. Anal strictures are not commonly reported in paediatric Crohn's disease. A French group observed anal strictures in 5.7% (out of 42 children) of their patients aged between 4– 20 years old. We looked back over 5 years (from 2004-2009) and identified 7 children with Crohn's disease and anal strictures. We report our experience of presentation, diagnosis and management.

Results

Age at diagnosis of CD (years)	Sex	Distribution of CD at diagnosis	Treatment at the time of diagnosis of stricture	Perianal disease	Time to identification of stricture (years)	Presenting feature leading to diagnosis of stricture
6	F	Colon	Azathioprine	Present	6	Constipation
11	F	Colon, Terminal Ileum and stomach	Azathioprine	Present	0	Present at diagnosis
15	M	Rectum, Colon, Terminal Ileum, duodenum and esophagus	Azathioprine	Present	0	Present at diagnosis
8	M	Colon, Terminal Ileum, duodenum, stomach and esophagus	Methotrexate Azathioprine Infliximab	Present	2	bloody diarrhoea
4	M	Colon, Terminal Ileum, stomach and esophagus	Azathioprine Infliximab	Absent	4	Soiling
13	F	Oesophagus, stomach, terminal ileum and colon	Azathioprine Methotrexate	Absent	2	Bloody diarrhoea alternating with constipation
10	F	Colon, Terminal Ileum, duodenum, stomach	Azathioprine	Present	4	Perianal disease and abscess needing surgical drainage

Anal stricture was identified in 4 females and 3 males. The youngest was 8 years (mean age, 12.1y) and the mean duration for development of stricture following diagnosis of CD was 2.6 years (range of 0 – 6y). Two children, newly diagnosed with CD, were found to have strictures at first colonoscopy, following presentation of symptoms suggestive of CD. In those known to have CD, presenting symptoms leading to diagnosis of stricture were constipation with soiling in 2 cases and in 4 cases anal stricture was diagnosed at repeat colonoscopy assessment for disease activity because of persistent symptoms of bloody diarrhoea and diarrhoea alternating with constipation. 2 of these patients had severe active colonic inflammation but 2 had only mild left sided colonic inflammation and persistent symptoms were attributable to anal stricture rather than active colonic disease. 5/7 patients had associated active perianal disease at the time of diagnosis, with 3 having severe disease as per U.F.S (Ulcerating, fistulating, structuring/ abscess) classification. Association between anal strictures and presence of genital CD in both sexes has been reported. In our series 2 patients had genital CD, both girls. Only 2 patients had received treatment with infliximab prior to diagnosis of stricture. Following diagnosis of anal stricture all patients underwent anal dilatation under general anaesthetic (GA) with Hegars. All patients were instructed in self anal dilatation at home but only 1/7 patient was partially compliant and all needed repeat dilatation under GA. There were no complications following anal dilatation and none of the patients have undergone surgery.

Conclusion:
Anal strictures can present at any time during the course of CD. Perianal and genital CD appear to be commonly associated as reported. In patients with apparently poorly controlled symptoms, the possibility of anal stricture should be considered as management of the patient may be very different.

School in or out? School attendance in children on home parenteral nutrition (HPN)

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Introduction:
Surveys have shown the incidence of absenteeism in chronic diseases is approximately twice that of healthy children (Charlton, 1991). Continuing improvements in treatment have enabled children with chronic health problems to be discharged home on "hi tech" treatment and to attend full time school. This study investigates the absences of children with severe intestinal failure on long term HPN.

Aim:
To identify if improvements in medical treatment and co ordination between health professional and schools has enabled children with intestinal failure requiring long term PN at home to attend school and compare attendance with the national average.

Method:
Details of absences from the attendance registers in the child's school were obtained for all HPN children attending our unit and for all children in the United Kingdom and Ireland in full time education between 5-16 years old, for the academic years 2005-2006 and 2008-2009. Age, sex and underlying disease of our HPN children were recorded and analysed.

Results:
In 2005-2006, 21 children (female:male 12:9) at home on PN treatment were in full time education. 18 were at school and 3 were educated at home with a tutor (all 3 had gut dysmotility). The average absenteeism of these children during the year was 15.9% (range 3 – 34%) compared with national average 6.8%. No significant difference noted between secondary/primary school and sex. Significant difference ($p = 0.01$) was noted in the motility category with a higher absenteeism.

In comparison, in 2008-2009, 15 of the 21 children (female:male 8:7) were still on HPN. Eleven of the 15 on HPN were still at school and 1 was educated at home. Two had progressed to university education and 1 was in full time employment (all 3 had completed 'A' levels). Five children were no longer on treatment and had weaned onto enteral feeds, 1 had had an intestinal transplant. The average absenteeism of a pupil during this year was 18.8% (range 8 – 38%) compared with national average 6.2%.

Problems identified include unrecorded absence, and which absences were PN related to or not.
Conclusion: Children on HPN have higher absences than the national average although still have good attendance levels and an excellent level of achievement. This absenteeism level is in keeping with other chronic diseases. School attendance is an important indicator for qualify of life with children proving they can do academically well as demonstrated by children progressing to university education.

Children who suffer chronic disease should have the opportunity to have a future career. The importance of school should therefore be remembered in a child's medical management.

Charlton, A (1991). Absence from school related to cancer and other chronic conditions, *Archives of Disease in Childhood,* 66, 1217 – 1222.

The Role of MRI small bowel in the assessment of Inflammatory bowel disease: a single centre prospective study

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Background:
Radiological evaluation plays a critical role in the investigation of Inflammatory Bowel Disease (IBD). Primarily its role is in diagnosing lesions in the small bowel from the distal duodenum to the terminal ileum that often remains a challenge. Repeated exposure to ionising radiation is a concern particularly for those patients with early onset IBD who will require multiple diagnostic imaging procedures. MRI of the small bowel has been made possible with the development of rapid scanning techniques.

Aim:
To assess the reliability of MRI in assessing extent and activity of IBD.

Methods:
Over a one-year period all patients being investigated for suspected IBD or patients with existing IBD who required reassessment of small bowel disease, underwent MRI small bowel with enteral and IV contrast. T1 and T2 weighted images pre contrast with T1 volumetric images following intra-venous contrast were obtained within weeks of concurrent upper GI endoscopy and ileocolonoscopy. Comparison of MRI findings was made with endoscopic findings and histopathology of biopsies obtained.

Results:
MRI small bowel was carried out in 58 patients (27 female; 31 male) (mean age 13.4 years (6 – 17y)); 34 patients had an MRI as part of the primary evaluation for suspected IBD; 3/34 patients did not have features of IBD on MRI examination or on endoscopic evaluation; 17/31 patients had Crohn's Disease (CD), 9/31 had Ulcerative Colitis (UC) and 5/31 had IBD-Unclassified. Of 17 patients with CD, 13 had features of small bowel disease on MRI small bowel study (76%); 1 patient with CD had isolated MRI small bowel changes (Ileitis) without endoscopic or histology changes; 4 patients had a diagnosis of CD based on MRI small bowel changes where it was impossible to classify colitis detected on endoscopy/histopathology alone. MRI small bowel study was performed on 24 patients with pre-existing CD; 15/24 had MRI changes (63%). There was significant agreement between active ileal CD on MRI and histopathology/gross endoscopy.

Conclusion:
In the investigation of paediatric IBD, MRI Small Bowel may offer an alternative method of detecting small bowel disease. MRI ileal disease agrees closely with histopathology and/or gross endoscopy. The absence of radiation and the potential to identify subtle inflammation, active inflammation in strictured areas, extraintestinal manifestations, and colorectal disease offer distinct advantages over fluoroscopic studies.

The role of seasonality in exacerbations of inflammatory bowel disease: a systematic review

Dr Paul Henderson, Prof Jürgen Schwarze, Dr David C Wilson: Department of Child Life and Health, University of Edinburgh

Introduction:
Inflammatory bowel disease (IBD) is characterised by acute exacerbations, often leading to hospital admission and significant morbidity. Due to the likely role of a dysregulated immune response to environmental pathogens in IBD and the known seasonal variation in infectious diseases, confirmation of a seasonal pattern to IBD exacerbations would be significant.

Aim and Methods:
To evaluate the role of seasonality in the exacerbation of established IBD at any age by formal systematic review. An electronic database search of the Cochrane Library, Medline, Embase and the British Nursing Index and Archive was performed with keywords related to IBD, exacerbation and seasonality. A hand search of reference lists of articles was also performed, along with reviews of major gastroenterology journals and meeting abstracts. An English language restriction was implemented. The papers identified were then critically appraised and the level of evidence (EL) assigned using the Scottish Intercollegiate Guidelines Network (SIGN; www.sign.ac.uk) methodology.

Results:
From the initial 2111 hits 41 papers were reviewed in full text. Of these, 14 were identified that presented data on the association of seasonality with exacerbations of IBD. Ten of the papers were case series (EL 3), with two prospective cohorts (EL 2-) and two retrospective cohorts (2+, 2-). Six papers looked at ulcerative colitis (UC) exacerbation only with one examining Crohn's disease (CD) exacerbation exclusively. The retrospective cohort study gaining an EL 2+ grading used a cross-over design to reduce confounding significantly. Overall the two prospective cohorts demonstrated an increase in UC exacerbations in the autumn/winter months, with the retrospective cohort showing a significant increase in UC exacerbation in spring (March-May). The remaining case series suggested an increase in UC exacerbation from Sept-Feb, peaks of exacerbation in spring/summer with no specific months defined and a peak of UC exacerbations between Mar-Aug.

Conclusion:
The conclusions of published studies to establish a relationship between seasonality and IBD relapse are variable. The level of evidence is poor due to inadequately defined outcome measures and reliance on self-reporting. It is clear that there is a need for more robust, prospective studies with clearly defined outcome measures to determine if seasonality truly affects IBD relapse rates. If such an effect does exist, then this could have wider implications for disease prevention measures, health care service provision and current IBD pathogenic mechanisms.

Conflict of interest: None declared

Tracking paediatric cognitive outcomes following combined liver small bowel transplantation: a case study.

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Introduction:
Infants with intestinal failure (IFx) now survive and grow satisfactorily with parenteral nutrition (PN), but IQ may be affected. A potential reason for this maybe dependency on a single source of lipid based on soya oil (Intralipid), which lacks essential polyunsaturated fatty-acids (PUFAs) normally found in breast milk and diet.

Aim:
Our aim was to assess the long term cognitive and developmental effects of combined liver and small bowel transplantation and the potential detrimental effects of PN induced PUFA deficiency.

Methods:
Psychometric data collected over ten years were used to assess developmental outcome of an 11year old boy who received SBTx at age 9 months because of liver failure secondary to PN and IFx caused by Hirchsprung's disease. Current dietary intake was assessed with a comprehensive 5-day diet diary. PUFA status was assessed in red blood cell (RBC) membranes by conventional Gas Chromatography-Flame Ionised Detection. Neurochemistry was non-invasively assessed with proton-magnetic resonance spectroscopy (1H-MRS) in occipitoparietal and frontal cortex regions.

Results:
Pre-Tx, the patient demonstrated mild cognitive delay and normal motor development. Six months post-Tx the patient displayed significant motor and mental delay. Between three and five years post-Tx, IQ plateaued in the borderline/low average range (72 and 79 respectively). By ten years post-SBTx, IQ had risen to 97, well within average range. His PUFA intake was negligible until commencing Nutrini orally (662 mg/L PUFA), 6 months post-SBTx. The patient is now on a normal diet (300mg PUFA/day). Neurometabolite values recorded by MRS, such as N-acteyl aspartate and Choline, which provide markers of neuronal health, showed a normal profile for a healthy 11 year old child. At 10 years post-SBTx the patient's essential PUFA levels, specifically Docosahexaenoic acid and Eicosapentaenoic acid, were no different to healthy age-matched controls (4% vs 4.23% of total fatty acid content), which is consistent with his MRS and cognitive assessments.

Conclusion:
In contrast to a cohort of children maintained on PN for 5 years or more (S.Hill et al Arch Dis Child. 2005;90:A16), this boy, who commenced normal diet from 1 year, has a normal IQ and distribution of neurometabolites and essential blood lipids, demonstrating that early SBTx is consistent with good long-term cognitive outcome.

We are grateful to Dr JWL Puntis for referring his patient to us.

Type II and type III paediatric intestinal failure within the Scottish Home Parenteral Nutrition Managed Clinical Network 01/97-12/08

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Background/Aim:
Management of intestinal failure (IF) may require prolonged periods of parenteral nutrition (PN) in hospital; if so, home parenteral nutrition (HPN) is usually considered early in the course or when the likely prolonged duration of PN becomes evident. IF in children is more likely than IF in adults to progress to intestinal transplantation (ITx), usually as combined small bowel-liver transplant (CSBLT), but occasionally isolated small bowel transplant or isolated liver transplant (as a 'bridging' procedure or where portal hypertensive enteropathy is the major issue). This is mainly due to the high prevalence of IF-associated liver disease in children, especially infants, but vascular access issues are also important. The 4 tertiary centres in Scotland providing paediatric (<18 years of age) home parenteral nutrition (HPN) had informally collected audit data on IF and HPN from 01/97 onwards, and this was formalised within the Scottish HPN MCN in 12/00. Only these 4 HPN centres have the facilities and expertise to give prolonged PN beyond term. Scotland comprises 8.6% (5.2 million/61 million x 100) of the paediatric population of the UK. Our aim was to evaluate how many children had paediatric type II (prolonged PN in hospital) or type III (HPN) IF, how many required intestinal transplant assessment, and their subsequent course.

Methods:
An audit of the databases of all 4 tertiary paediatric HPN centres in Scotland (Aberdeen, Dundee, Edinburgh and Glasgow) has been ongoing from 01/97. We reviewed all type II and type III IF from 01/97 to 12/08, plus cases of HPN not primarily due to IF (complex Crohn's disease prior to biological medication introduction or severe GI dysmotility with refractory bilious vomiting). Outcomes of interest included type II IF where the paediatric complex nutrition support team (NST) initially or during the PN course considered HPN as a possibility; ITx assessment referrals to the single UK intestinal transplant unit in Birmingham Children's Hospital; ITx assessments; small bowel transplant, isolated liver transplant and CSBLT; need for HPN; and death.

Results:
There were 106 referrals where the paediatric complex NST initially or later considered HPN as a possibility, of which all but 2 due to type II IF. 38 received HPN, of which 10 were actively on HPN as of 31/12/08. There were 22 referrals for ITx assessment, of whom 5 received CSBLT, 2 isolated liver transplants, and 6 deaths whilst awaiting ITx/CSBLT.

Summary and conclusions:
We report nationwide data on the prevalence of type II IF in childhood in Scotland, managed in the 4 HPN centres. Over 35% progress to HPN, and over 20% are referred for ITx assessment. Due to lack of appropriately sized organ availability, more than 25% of those referred died on the waiting list for transplantation. Extrapolation of our Scottish nationwide data (collected within a national MCN) to the UK ((nx100/8.6)/12 years) suggests that there are annual paediatric prevalences of 101 cases of type II IF who may need HPN, 37 cases requiring HPN and 21 cases of type II or type III IF requiring ITx assessment. These data are important for counselling families and for planning regional and national paediatric type II and III IF specialist services in the UK.

Users' views on transition services for young people with Inflammatory Bowel Disease.

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Background:
The benefits of a well managed transition for adolescents with Inflammatory Bowel Disease (IBD) are being increasingly recognised. However, effective means of transition are currently supported by limited evidence. Patient feedback is important to establish the value of such services. This survey looks at how young people with IBD and their families perceived the effectiveness of the 'Alder Hey model' of transition and what improvements were desired.

Design:
Postal questionnaire survey.

Setting:
Transition between Paediatric services at Alder Hey Children's NHS Foundation Trust, Liverpool and the Royal Liverpool University hospital (adult centre). The process involves introducing the concept of transition by the age of 13- 14, a 'prehandover' meeting attended by both the adult and paediatric teams around the age of 16, and eventually handover at the transition clinic. Strong emphasis is placed on 'individual' cases to assess readiness for transition.

Subjects:
Young people (adolescents) who have been through the transition process between the above centres over the years 2003- 2008

Interventions:
Semi structured questionnaire survey designed to obtain patient feedback on transition care arrangements and provision of service.

Main outcome measures:
Patient information, satisfaction and suggestions.

Results:
35 of 66 families returned the questionnaire. High rates of satisfaction were reported (33/35; $p < 0.001$ – sign test). Feed back included provision of adequate information, opportunity to discuss change ($p< 0.001$) and reduction in perceived anxiety about the transfer ($p = 0.08$).

Conclusions:
High rates of satisfaction from the 'Alder Hey model' of transition were reported.

Varying predictive values dictate the need to standardize commercialkits for tTG testing

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Background:
Over the previous several months we have noticed an increased false positive coeliac serology rate in patients referred from a particular catchment area of our centre. We investigated serologic methods employed and compared it to methods used at our centre and other hospitals in our catchment.

Methods:
We investigated serological methods employed to screen 67 patients referred for histological conformation of coeliac disease to our service in the previous year (2008). Pathology departments in referring hospitals were contacted to ascertain serological methods used. Coeliac disease was diagnosed based on biopsy evidence (gold standard). Predictive values for the screening serological tests were calculated.

Results
are summarised in the following table.

Hospital	tTg ag / preparation	tTg positive cut off	Positive serology	Positive biopsy	Positive predictive value (%)
A	Phadia (EIIA Celikey)	>7 u/L	47	46	97.8
B	Euroimmun re Hu /baculovirus	>20 u/L	15	3	16.66
C	Phadia ELISA	>4u/L	4	2	50
D	Flourometric testing	>8	1	1	100
E	Dia Sorin (re Hu tTG chemo luminescence)	>8	2	1	50

Conclusions:
Positive predictive values vary significantly between commercially available tTG kits. When faced with a high false positive trend it is worthwhile investigating laboratory methods used, bench marking against reference laboratories and standardising serological testing.

What is the evidence for the transmission of infection from breast milk to preterm infants? A systematic review

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Introduction:
Human breast milk is a source of probiotic and potentially pathogenic bacteria. Pasteurisation eliminates these pathogens, but may destroy nutritional, immunological and probiotic elements vital to the health of preterm infants, and is by no means universal. Notably, in the United Kingdom although guidelines exist for the routine pasteurisation of DEBM, these do not apply to MEBM. We examined the evidence base for infection transmitted to preterm infants from breast milk.

Methodology:
Electronic searches were performed through: Medline, Cochrane, and Pubmed databases (1950-September 27th 2009). MeSH Keywords included: infant; preterm; human/maternal/breast milk/donor milk; sterilisation/pasteurisation/disinfection; milk banks; storage/freezing/refrigeration; and infection/sepsis/septicaemia. Review articles on breast milk for preterm infants were cross referenced. Selection criteria comprised articles concerning preterm infants (<37 weeks) up to 28 days corrected gestational age, with microbiology and epidemiology confirming breast milk as the source of infection. The SIGN guidelines for appraisal were employed (www.sign.ac.uk), ranking evidence in descending order of merit according to study type (1: RCT, to 4: expert opinion) and quality (++, + and -).

Results:
105196 articles were cited, yielding 1081 abstracts for review. 15 articles were relevant: two case-control studies were assigned EL 2-, and the remainder were lower EL case series' 3 (13). In total 45 infected 'cases' and 29 uninfected 'controls' were identified. 12 infants were infected by single donor expressed breast milk (DEBM), and 14 were infected by pooled DEBM. Maternal EBM (MEBM) was implicated for 15 infants, a mix of MEBM and breast feeding in three, and a single infant was exclusively breast fed. Milk was pasteurised in only one article (n=14, all fed DEBM). Mean gestation: 28.7 weeks ± 2.75 (Not specified [NS]: n=28). Mean day of life on which milk-related sepsis first diagnosed: 21.9 ± 12.19 (NS: n=26). Milk storage methods included: refrigeration (n=12), freezing (n=22), or NS (n=11). Mothers were twice as likely to be asymptomatic (n=12) as symptomatic (mastitis (n=4), mastitis and wound infection (n=1), or known positive vaginal swab (n=1)). Only 1 report tested expressing equipment for colonisation (n=5 infants). Organisms included: Salmonella (n=10), Klebsiella (n=7), E. Coli 0125 (n=14), Group B Streptococcus (n=12) and MRSA (n=2). Methods of detection included molecular techniques and conventional culture. Three infants died, and one incurred significant long term morbidity.

Conclusion:
The current evidence base for the transmission of infection to preterm infants from human milk is limited and the role of pasteurisation unclear. Future studies identifying milk as a source of infection could consider expressing and handling equipment as a potential cause. Only one study considered contamination from this source. The need for pasteurisation could be obviated by employing strict hygiene protocols, routine milk culture and maternal screening questionnaires. Breast milk and handling equipment should be considered as a focus of late-onset, unusual or recurrent infection in preterm infants. Further studies are required to explore the effects of alternative methods of sterilisation.

Notes:

Exhibitors

Danone Baby Division

Ferring Pharmaceuticals

Pentax

Abbott

Orphan Europe

Vygon

NACC

CICRA

CIMS

Biohit

CME McKinley UK Limited

Dr Falk Pharma

BUPA

Calea

GBUK

Norgine

Nestle

Previous winners of prizes

2008 Southampton

Alex Mowat Prize - Dr Andrew Barclay

Best Abstract Presentation - Ms Elaine Buchanan

Best Presentation - Dr Sherina Ross

2009 Sheffield

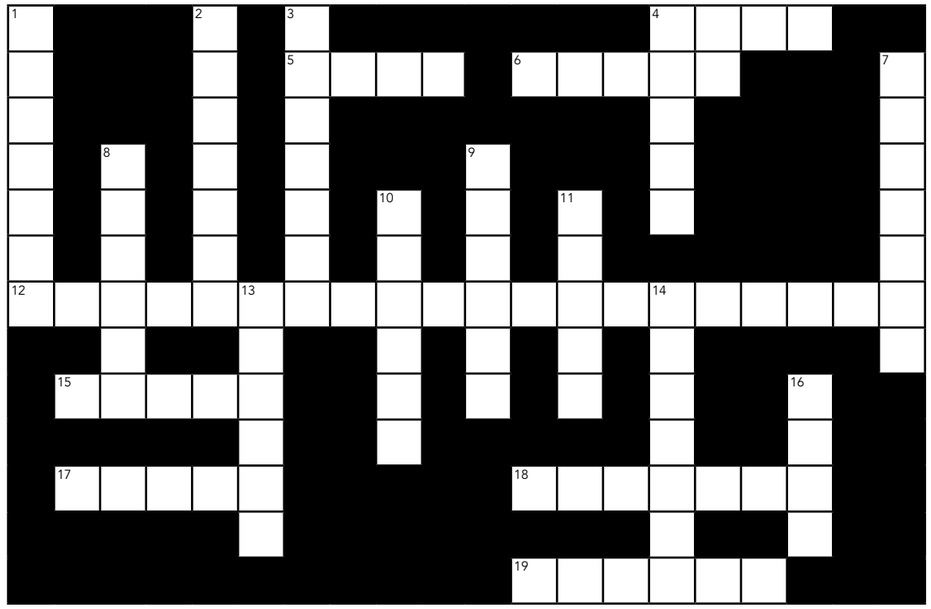
Alex Mowat Prize - Dr Johann van Limbergen

Sean Devane Memorial - Dr Jenny Epstein

Best Allied Health Professional - Ms Jackie Falconer

Quotations from Hippocratic oaths

Liverpool crossword



Clues Across

4 Fab (4)
5 Baby faced member of the Beatles (first name) (4)
6 Is is a bird? (5)
12 Global football anthem (5,5,4,5)
15 Hundred plus X consecutive years in top division by blues (5)
17 Drummer (first name) (5)
18 Band of northerners rejected by record lable as 'Have no Future' (7)
19 River in Liverpool (5)

Clues Down

1 Successful manager of the Reds (7)
2 Successful manager of the Blues (7)
3 Successful manager of the Fab Four (7)
4 How to get across No 19 across (5)
7 Supporters of the Blues (7)
8 Highly distinctive accent (5)
9 My Sweet Lord (first name) (6)
10 Club made famous by Fab Four (6)
11 National record goalscorer per season (Blues) (7)
13 Returned his MBE to the queen by signing "with love" (surname) (6)
14 Supporters of the Reds (7)
16 All leading Reds goalscorer (4)

LIVERPOOL MAP

Liverpool crossword (Answers)

Across

4 Four
5 Paul
6 Liver
12 You'll never walk alone
14 Seven
16 Ringo
17 Beatles
18 Mersey

Down

1 Shankly
2 Kendall
3 Epstein
4 Ferry
7 Toffees
8 Scouse
9 George
10 Cavern
11 Sharp
13 Koppites
15 Rush