ANNUAL MEETING 2014

INCORPORATING

Joint BSPGHAN and British Association of Paediatric Surgeons (BAPS) Meeting

on Friday 31st January 2014

Wednesday 29th - Friday 31st January 2014

Tower Hotel Conference Centre, St Katharine’s Way, London

Educational Grants provided by Principal Gold Sponsors:

and Platinum Sponsors:

Local Organisers: Dr Alastair Baker, Dr Neil Shah, Dr John Fell, Professor Ian Sanderson, Dr Sally Mitton, Dr Mark Furman, Ms Sarah Mancell, Dr Syrena Kyrana

E&OE and CPD Approved
Welcome address from Local Organising Committee

Dear Colleagues, Full, Trainee and Associate Members, Sponsors, Delegates and Friends.

May I extend my warmest welcome on behalf of the organising committee to this 28th meeting of BSPGHAN in London.

Each year our annual meeting sets the bar higher, with higher attendance, higher standards of speakers and presentations, greater numbers of high quality abstracts submitted, a consistently impressive venue, a more lively social programme and greater approval and participation from our colleagues in the Paediatric Nutrition and Pharma industries. A formula that you all know has developed comprising a daily programme structure, Society business meetings, entertainments including the Gala dinner, social and networking opportunities and a football match all arranged perennially. The friendly atmosphere that is so much part of the Society infuses all the events making each meeting memorable and enjoyable, attracting us all back year after year in increasing numbers.

This year, the organising committee have worked to make the meeting as representative of Paediatric Gastroenterology in London as possible, and give the flavour of London as an exciting city venue. We have tried to built on the successful formula, using all the tried and tested components but stretching the envelope just a bit. Firstly, recognising that we are all in career-long continued education and that the programme content continues to grow, we have changed the ‘Trainees’ day’ on the Wednesday to be a full programme day much like the other two with content aimed to be suitable for all members alike. Secondly, we have joined with BAPS to hold their Winter meeting on the third day of our meeting, so that comparative content can be presented and discussed ‘back to back’. We hope this will be a model for future meetings with other groups and societies. Thirdly, we have opted for a quiet evening of networking on the Wednesday night being the preference of feedback we received. Finally in recognition of these changes we have reaffirmed the recently adopted name of the event as ‘The BSPGHAN Annual Meeting’.

None of the above successes would be possible without the input of you, our colleagues in the Pharma and Nutrition industries. We value your input into the content through the symposia, your contribution to abstracts, your financial support and of course your attendance at all the various activities comprising the meeting. We recognise that the company representatives value and enjoy BSPGHAN meetings as much as we do and hope you will participate and enjoy this year as much as past meetings.

Best wishes

Academic Committee: Dr Alastair Baker, Dr Neil Shah, Dr John Fell, Prof Ian Sanderson, Dr Sally Mitton, Dr Mark Furman, Ms Sarah Mancell, Dr Syrena Kyrana
Organising Team: Dr Rafeeq Muhammed, Mrs Carla Lloyd, Ms Pamela Ward

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The organising committee have worked to make the meeting as representative of Paediatric Gastroenterology in London as possible, and give the flavour London as an exciting city venue. We hope you will participate and enjoy this year as much as past meetings.

Best wishes

Organising Committee:
Dr Alastair Baker - Consultant Paediatric Hepatologist, King’s College Hospital London
Dr Neil Shah, Consultant Paediatric Gastroenterologist, Great Ormond Street Hospital, London
Dr John Fell, Consultant Paediatric Gastroenterologist, Chelsea and Westminster, London
Professor Ian Sanderson, Consultant Paediatric Gastroenterologist, Barts and The London, London
Dr Sally Mitton, Consultant Paediatric Gastroenterologist, St George’s Hospital, London
Dr Mark Furman, Consultant Paediatric Gastroenterologist, Royal London and Great Ormond Street Hospital, London
Ms Sarah Mancell, Lead Children’s Dietitian, King’s College Hospital, London
Dr Syrena Kyrana, Specialist Registrar, King’s College Hospital, London

Organising Team:
Dr Rafeeq Muhammed, Mrs Carla Lloyd, Ms Pamela Ward

Master of Ceremonies:
Wednesday: Dr Rafeeq Muhammed
Thursday: Professor Dino Hadzic
Friday: Dr Neil Shah

Abstract selection committee:
Dr Rafeeq Muhammed, Chair Education Group; Dr Julian Thomas, Chair Research Group; Dr Fiona Cameron, Chair Trainees’ Group; Mr Mick Cullen, Chair Associate Member’s Group; Dr Alastair Baker, Chair Local Organising Committee
Wednesday 29th January 2014

Tower Hotel, London

8.00
Registration Opens - Tower Hotel, Foyer

9.20
BSPGHAN Annual Meeting Opens - Tower Suite, Tower Hotel

9.20 – 9.30
Welcome and Introduction
Dr Alastair Baker - Consultant Hepatologist, London President BSPGHAN

9.30 – 10.45
Theme - By popular request

Chairs:
Dr John Puntis - Consultant Gastroenterologist
Leeds General Infirmary, Leeds
Dr Neil Shah - Consultant Gastroenterologist
Great Ormond Street Hospital, London

9.30 – 9.55
Chronic Diarrhoea – a practical approach
Professor Simon Murch
Consultant Paediatric Gastroenterologist
Clinical Sciences Research Institute
Clifford Bridge Road
Coventry CV2 2DX

9.55 – 10.20
Gluten intolerance v Coeliac Disease
Dr Fevronia Kiparissi
Consultant Paediatric Gastroenterologist
Great Ormond Street Hospital for Children
Dept of Paediatric Gastroenterology
London WC1N 3JH

10.20 – 10.45
The Impossible IBD Case – presentation Q&A
Dr Richard Russell
Consultant Paediatric Gastroenterologist
Yorkhill Hospital
Dalnair Street
Glasgow G3 8SJ
10.45 – 11.10
COFFEE AND POSTER VIEWING

11.10 – 13.00
Theme - ‘Predicting the future’

Chairs:
Professor Deirdre Kelly - Consultant Paediatric Hepatologist, Liver Unit, Birmingham Children's Hospital, Birmingham and
Dr Huw Jenkins - Consultant Paediatric Gastroenterologist University of Wales, Heath Park, Cardiff

11.10 – 11.35
New Investigations and therapies in liver disease
Dr Richard Thompson
Consultant Paediatric Hepatologist
Senior Lecturer in Paediatric Hepatology
The Institute of Liver Studies
King's College Hospital
London SE5 9RS

11.35 – 12.00
New GI therapies – Faecal transplantation
Dr James Lindsay
Senior Lecturer and Consultant Paediatric Gastroenterologist
Barts and The London
Turner Street
London E1 2AD

12.00 – 12.30
Acute Liver Failure: Transport and PICU Care
Dr Akash Deep
Consultant Paediatric Intensivist
King's College Hospital
Denmark Hill
London SE5 9RS

12.30- 12.40
Constitutive Type I Interferon, via STAT1 activation, promotes regulatory T cell function in the healthy human intestine, but not in inflammatory bowel disease
Giles, Edward: Pathak, Mohini: Sanders, Theo: McCarthy, Neil: Sanderson, Ian: MacDonald, Tom: Lindsay, James: Stagg, Andrew:
Centre for Immunology and Infectious Disease, Barts and the London School of Medicine and Dentistry, London

12.40 – 12.50
Anti TNF dependency in Paediatric IBD: the Scottish Experience
1Child Life and Health, University of Edinburgh, Scotland, UK, 2Raigmore Hospital, Inverness; 3Royal Aberdeen Children's Hospital, Aberdeen; 4Royal Hospital for Sick Children, Glasgow

12.50 – 13.00
Mucosal healing in children with Crohn’s disease on long term maintenance treatment
Dr Bhavsar Hemant: Dr Theodoric Wong: Dr Susan Protheroe: Dr Stephen Murphy: Dr Ronald Bremner: Dr Rafaq Muhammed:
Dept of Gastroenterology, Birmingham Children's Hospital, Birmingham

13.00 – 14.00
LUNCH AND POSTER WALKS

14.00 – 15.15
Theme - The Post Antibiotic Era

Chairs:
Dr Julian Thomas - Consultant Gastroenterologist
Royal Victoria Infirmary, Newcastle and
Dr Babu Vadmalayan - Consultant Paediatric Gastroenterologist
King's College Hospital, London

14.00 – 14.25
New antibiotic forms and effectiveness
Dr David Wareham
Senior Clinical Lecturer (Honorary Consultant) in Microbiology
Barts and The London Hospital
Turner Street
London E1 2AD

14.25 – 14.50
CRE – The Newest Superbug – learning from an outbreak
Anita Verma
King’s College Hospital
Denmark Hill
London SE5 9RS

Plenary Session One
12.30 – 13.00

Chairs:
Professor Simon Murch - Consultant Gastroenterologist
Clinical Sciences Research Institute, Coventry and
Dr Paul Henderson - Clinical Research Fellow
Queen’s Medical Research Institute, Edinburgh
14.50 – 15.15
Very early onset inflammatory bowel disease - the expanding spectrum of monogenic disorders
Dr Holm Uhlig
University Research Lecturer and Hon Consultant Gastroenterologist
Translational Gastroenterology Unit and Department of Paediatrics
John Radcliffe Hospital
Oxford

15.15 – 15.35
COFFEE

15.35 – 16.00
Theme - Pancreas
Chairs:
Dr Rob Heuschkel - Consultant Gastroenterologist
Addenbrooke’s Hospital, Cambridge
and
Dr Mark Furman - Consultant Gastroenterologist
Royal Free Hospital, London

Pancreatic diseases and their clinical management – Everything you need to know
Dr Tassos Grammatikopoulos
Locum Consultant in Paediatric Hepatology
Paediatric Liver, GI & Nutrition Centre
King’s College Hospital NHS Foundation Trust
Denmark Hill, London, SE5 9RS

16.05 – 16.15
The epidemiology and management of acute pancreatitis in children
Dr Farah Mustaq (Princess of Wales Hospital, Bridgend), Dr Mike Coagrove (Morriston Hospital Swansea), Mr Kim Hutton, Dr Huw Jenkins & *Dr Ieuan Davies (all University Hospital of Wales).

16.15 – 16.25
The effect of commonly used IBD drugs on autophagy induction using an in vitro cell culture system
Henderson, Paul1; Satsangi, Jack2; Wilson, David C1; 1University of Edinburgh, Edinburgh
Stevens, Craig2; 2Edinburgh Napier University, Edinburgh

16.25 – 16.35
Autoimmune pancreatitis in children; evolving diagnosis and management
Grammatikopoulos Tassos1; Zen Yoh2; Karani John1; Bogdanos Dimitrios1; Pavlidou Maria1; Harrison Philip1; Devlin John1; Mieli-Vergani Giorgina1; Dhawan Anil1; 1Paediatric Liver Centre, King’s College Hospital NHS Foundation Trust, London; 2Institute of Liver Studies, King’s College Hospital NHS Foundation Trust, London; 3Department of Radiology, King’s College Hospital NHS Foundation Trust

16.35 – 16.45
Cost effectiveness of single visit parenteral iron (SVPI) infusions in children with Inflammatory bowel disease (IBD)
Rhona Hubbard, Dr P Narula, Dr M Thomson, Dr P Rao,
Dept. of Paediatric Gastroenterology, Sheffield Children’s Hospital NHS Foundation Trust, UK

16.45 – 16.55
MRI assessment of body composition in paediatric Crohn’s disease; intra-abdominal adipose tissue association with disease severity
Thangarajah D1; Chappell KE1; Gale C1; Parkinson JRC1; Epstein J1; Hyer W1; Soondrum K1; Frost G1; Fall JME1; 1Section of Academic Neonatal Medicine, Imperial College, London; 2Department of Radiology, Chelsea and Westminster NHS foundation trust; 3Paediatric Gastroenterology department, Chelsea and Westminster NHS foundation trust; 4Nutrition and Dietetic Research Group, Faculty of Medicine, Imperial College Hammersmith Hospital

17.00 – 18.00
Mead Johnson Symposium
Chair:
Professor Ian Sanderson - Consultant Paediatric Gastroenterologist
Bart’s and the London, London

17.00 – 17.30
Evolving treatments for cow’s milk allergy
Dr Adam Fox
Consultant Paediatric Allergist
 Evelina London Children’s Hospital
St Thomas’ Hospital
Westminster Bridge Road
London SE1 7EH
**Thursday 30th January 2014**

**Tower Hotel, London**

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**17.30 – 18.00**

The role of probiotics in the management of infants with cow milk allergy

Dr Erika Isolauri

Department of Paediatrics

Turku University and the University of Turku

Turku

Finland

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**18.00 – 19.00**

Group Meetings - The Tower Hotel, London

Open to All delegates

Associate Members AGM

Trainee Members

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**Followed by**

Football: Consultants v Trainees

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**20.45 till late**

Ice Breaker

and Meet the Sponsors Supper at

The Tower Hotel

Followed by informal social evening
8.00
Registration Opens - Tower Hotel, Foyer

7.45 - 9.00
Open Working Group Meetings:
The Tower Hotel, London

Groups to be confirmed

9.00 – 9.30
Standards and Commissioning – What will that mean in paediatric Gastroenterology
Dr Hilary Cass
President
RCPCH
5-11 Theobalds Road
London
WC1X 8SH

9.30 – 10.45
Theme: Organisation

Chairs:
Dr Mark Beattie - Consultant Gastroenterologist
Southampton General Hospital, Southampton
and
Dr Patricia McClean - Consultant Hepatologist
Leeds General Infirmary, Leeds

9.30 – 10.45
Hepatitis B & C – Adult innovations and the future for children
Dr Ivana Carey
Clinical Lecturer in Viral Hepatitis
Institute of Liver Sciences
King's College Hospital
London

9.30 – 10.45
Strategy – what does that mean for healthcare professionals?
Dr JH Coakley MD FRCP
Medical Director, Caldicott Guardian and Responsible Officer
Homerton University Hospital NHS Foundation Trust
Homerton Row
London
E9 6SR

10.45 – 11.05
COFFEE AND POSTER VIEWING

11.05 – 12.05
MSD Symposium - 4 Perspectives; 2 Patients
Chair:
Dr Nick Croft - Consultant in Paediatric Gastroenterology
Senior Lecturer, Barts and The London School of Medicine and Dentistry,
Whitechapel, London

Dr Nick Croft
Consultant in Paediatric Gastroenterology
Senior Lecturer
Barts and The London School of Medicine and Dentistry
Whitechapel
London

Dr James Lindsay PhD FRCP
Consultant Gastroenterologist
Barts and the London NHS Trust
The Royal London Hospital
Whitechapel
London

Ms Vikki Garrick
Paediatric Inflammatory Bowel Disease Nurse Specialist
Royal Hospital for Sick Children
Glasgow

12.05 – 13.00
Plenary Session Three

Chairs:
Dr Susan Protheroe - Consultant Gastroenterologist
Birmingham Children's Hospital, Birmingham
and
Mr Mick Cullen - Paediatric Gastro Nurse Specialist
Southampton General Hospital, Southampton

12.05 – 12.15
Burden of care at night when living with a child on parenteral nutrition at home.
Hughes A, Koegelmeier J, Hill S.
Intestinal Failure Rehabilitation Unit, Dept Gastroenterology, Great Ormond Street Hospital,
Great Ormond Street, London
12.15 – 12.25
Long term outcome of children with intestinal failure in the UK
H. Gowen1, Lloyd, C2, JWL Puntis1
1Birmingham Children’s Hospital, Birmingham, UK, and 2The General Infirmary at Leeds UK.

12.25 – 12.35
Long term growth outcome of children on home parenteral nutrition (HPN) in a tertiary centre
Cunningham, S;
Paediatric Nutrition Team, Great North Children’s Hospital, Newcastle upon Tyne, NE1 4LP

12.35 – 12.45
Fish-oil based intravenous lipid emulsion as a rescue in septic infants with intestinal failure
and with or at risk of developing liver disease
Huey Miin Lee1, Ann Hickey2, Helen Callaby1, Marie O’Meara2, Lucy Thompson1, Jonathan Hind3;
1Paediatric Hepatology, 2Paediatrics, 3Pharmacy, King’s College Hospital, London, United Kingdom

12.45 – 12.55
Five Year Experience of a joint multidisciplinary intestinal failure and small bowel
Transplantation Assessment Centre – the Great Ormond Street and King’s College Hospital
Referral Model
Eirini Serena1; Jain Vandana2; Hind Jonathan1; Heaton Nigel3; Vilaca-Melendez Hector1;
Hill Susan1; Lindley Keith1; Köglmeier Jutta1;
1King’s College Hospital London; 2Great Ormond Street Hospital London

14.00 – 14.05
Models of Cachexia and body composition
Dr Serena Kyrana
Specialist Registrar
King’s College Hospital
Denmark Hill
London, SE5 9RS

14.15 – 14.30
Obesity – what works
Martha Ford Adams
Consultant Paediatrician
Lead Paediatric for Diabetes/Obesity Named Doctor for Safeguarding
King’s College Hospital
London, SE5 9RS

14.30 – 15.00
LUNCH AND POSTER WALKS

14.00 – 15.15
Theme - Basic Science
Chair:
Dr Mike Cosgrove - Consultant Gastroenterologist
Singleton Hospital, Swansea
and
Dr Sally Mitton - Consultant Gastroenterologist
St George’s Hospital, London

14.00 – 14.25
Mechanisms of liver disease in non-alcoholic steatohepatitis
Bernadette Moore
Lecturer in Clinical Nutrition
University of Surrey
Duke of Kent Building
University of Surrey
Guildford
Surrey
GU2 7TE

14.25 – 14.50
Models of Cachexia and body composition
Dr Serena Kyrana
Specialist Registrar
King’s College Hospital
Denmark Hill
London, SE5 9RS

15.00 – 15.15
Obesity – what works
Martha Ford Adams
Consultant Paediatrician
Lead Paediatric for Diabetes/Obesity Named Doctor for Safeguarding
King’s College Hospital
London, SE5 9RS

15.20 – 15.40
TEA

15.40 – 17.00
Theme - President’s Choice
Chairs:
Dr Patrick McKiernan - Consultant Paediatric Hepatologist
Birmingham Children’s Hospital, Birmingham
and
Mr Chris Smith - Dietitian
Royal Alexandra Hospital, Brighton

15.40 – 16.05
Stem Cells – How far away is a liver off the shelf?
Ludovic Vallier
University of Cambridge
The Old Schools
Trinity Lane
Cambridge
CB2 1TN

16.05 – 17.00
Stem Cell Science – The GI Promise
Dr Nikhil Thapar
Consultant Paediatric Gastroenterologist
Gastroenterology Unit
Institute of Child Health
30 Guilford Street
London, WC1N 1EH
16.30 – 16.55
Achieving normal oral intake
Dr Gillian Harris
Birmingham Children’s Hospital
Steelhouse Lane
Birmingham B4 6NH

16.30 – 16.55
Achieving normal oral intake
Dr Gillian Harris
Birmingham Children’s Hospital
Steelhouse Lane
Birmingham B4 6NH

17.00 – 17.10
Evaluation of Sedation versus General Anaesthesia for endoscopy in children and adolescents
Ibrahim El busifi1, David Rawat1, Sandhia Naik2, Nigel Meadows2, Nicholas Croft2
1Centre for Digestive Diseases, Blizard Institute, Barts and the London Queen Mary's School of Medicine & Paediatric Gastroenterology 2Paediatric Gastroenterology, Barts Health NHS Trust

17.10 – 17.20
Paediatric Endoscopy Global Rating Score: Pilot data from 2 tertiary gastrointestinal centres
Protima Amon1, Ed Giles1, Ronald Bremner1, David Rawat1
1Department of Paediatric Gastroenterology, The Royal London Hospital; 2Department of Paediatric Gastroenterology, Birmingham Children’s Hospital

17.20 – 17.30
Evaluation of neonatal cholestasis in infants born pre-term
E Manea1, S Davison1, N Alizai1, T Humphrey1, H Woodley1, S Rajwal2, SV Karthik1, P McClean1
1Children’s Liver Unit and 2Department of Radiology, Leeds Teaching Hospitals NHS Trust

17.30 – 17.40
Growing up with biliary atresia without liver transplantation; a single centre experience.
Jain, Vandana1; Kolimarala1; Vinod1; Samyn, Marianne1; Davenport, Mark1; Heaton, Nigel1;
1Paediatric Hepatology, Gastroenterology and Nutrition centre, KCH, London, United Kingdom. 2Institute of Liver Studies, KCH, London, United Kingdom.

17.45 – 19.15
Annual General Meeting

20.00
Gala dinner at The Tower Hotel
Rocking to Queen Tribute Band and dancing till late
8.30
Registration Opens - Tower Hotel, Foyer

7.45 – 9.15
Open Professional Group Meetings
The Tower Hotel, London

9.15 – 11.00
Theme - Collaborative outcomes in GI surgery

Chairs:
Mr Mervyn Griffiths - Consultant Paediatric Surgeon
Southampton General Hospital, Southampton
and
Dr David Rawat - Consultant Gastroenterologist
Chelsea & Westminster, London

9.15 – 9.30
Launch of e-BANS
Dr Andy Barclay
Chair of BIFS and e-BANS Group
Consultant Paediatric Gastroenterologist
Royal Hospital for Sick Children
Dalnair Street
Glasgow

9.30 – 9.55
Endoscopy Training and Standards – The JAG approach
Dr Ieuan Davies
Consultant Paediatric Gastroenterologist
Dept of Child Health
University Hospital of Wales
Heath Park
Cardiff
CF14 4XW

9.55 – 10.20
Endoscopy Training and Standards – The BAPS approach
Mr Mark Powis
Consultant Paediatric Surgeon
Department of Paediatric Surgery
C Floor
Clarendon Wing
Leeds General Infirmary
Georges Mead
Leeds, LS2 9NS

10.20 – 10.45
Advances in the management of NEC
Mr Paolo di Ceggi
Senior Clinical Lecturer
ICU - Surgery Unit
Gower Street
London, W1E 6BT

10.45 – 11.05
COFFEE

11.05 – 12.45
Theme - Collaborative outcomes in GI surgery

Chair:
Mr Graham Haddock - Consultant Surgeon
Royal Hospital for Sick Children, Glasgow
and
Dr Anthony Akobeng - Consultant Gastroenterologist
Royal Manchester Children's Hospital, Manchester

11.05 – 11.30
To fundoplicate or not? Decision making based on high resolution manometry impedance
Professor Dr Nathalie Rommel
KU Leuven
Translational Research Center for Gastrointestinal Disorders (TARGID)
Neurosciences, ExpORL
University Hospital Leuven
Dept of Gastroenterology, Neurogastroenterology and Motility
Herestraat 49
3000 Leuven Belgium

11.30 – 11.55
Fundoplication – the surgical decision
Mr Bruce Jaffray
Consultant Paediatric Surgeon
Royal Victoria Infirmary
Newcastle upon Tyne, NE1 4LP

11.55 – 12.20
Surgical interventions in intestinal failure
Mr Mark Davenport
Consultant Paediatric Hepatobiliary Surgeon
King's College Hospital
Denmark Hill, London
12.20 – 12.45  
**BAPS-CASS Hirschsprung’s Study**  
Mr Gregor Walker  
Consultant Paediatric Surgeon  
Royal Hospital for Sick Children  
Dalmair Street  
Glasgow

12.45 – 13.45  
**LUNCH AND POSTER WALKS**

13.45 – 15.00  
**Theme - Collaborative outcomes in GI surgery**  
**Chairs:**  
Mr Simon Huddart - Consultant Surgeon  
University Hospital of Wales, Cardiff  
and  
Dr Nick Croft - Consultant Gastroenterologist  
Barts and The London, London

13.45 – 14.10  
**Colectomy – the medical decisions**  
Professor Ian Sanderson  
Consultant Paediatric Gastroenterologist  
Adult and Paediatric Gastroenterology  
ICMS  
Bart’s and the London  
Turner Street  
London, E1 2 AD

14.10 – 14.35  
**Colectomy – the surgical decisions**  
Mr Richard Cohen  
Consultant Surgeon  
University College Hospital  
London

14.35 – 15.00  
**Surgery in pseudo obstruction**  
Mr Joe Curry  
Consultant Paediatric Surgeon  
Great Ormond Street Hospital  
Great Ormond Street  
London

15.05 – 15.50  
**Key note lecture**  
**Chair:**  
Dr Alastair Baker - Consultant Paediatric Hepatologist  
King’s College Hospital, London

The Future Transplantation of Digestive Organs  
Mr Khalid Sharif  
Consultant Paediatric Hepatobiliary and Transplant Surgeon  
Birmingham Children’s Hospital  
Steelhouse Lane  
Birmingham  
B4 6NH

**PRIZE PRESENTATION AND CLOSE OF MEETING**  
Dr Alastair Baker  
and  
Dr Rafeeq Muhammed

**Previous Prize winners**

**2008 Southampton**  
Alex Mowat Prize – Dr Andrew Barclay  
Best Abstract Presentation – Ms Elaine Buchanan  
Best Presentation – Dr Sherina Ross

**2009 Sheffield**  
Alex Mowat Prize – Dr Johann van Limbergen  
Sean Devane Memorial – Dr Jenny Epstein  
Best Allied Health Professional – Ms Jackie Falconer

**2010 Liverpool**  
Alex Mowat Prize – Dr Emer Fitzpatrick  
Sean Devane Memorial – Dr Rachael Taylor  
Best Poster Presentation – Dr Paul Henderson

**2011 Edinburgh**  
Alex Mowat Prize – Dr Paul Henderson  
Sean Devane Memorial – Dr Emer Fitzpatrick  
Best Poster Prize – Ms Helen French
## Prize Presentation and Close of Meeting (cont...)

**Previous Prize winners**

**2012 Nottingham**
- Alex Mowat Prize – Mark Goddard
- Sean Devane Memorial – Anna Gregory
- Challenging Case – Lisa Whyte
- Best Poster – Ms Hannah Williamson

**2013 Manchester**
- Alex Mowat Prize – Dr Protima Amon
- Sean Devane Memorial – Dr Lisa Whyte
- Best Poster Prize – Dr Rana Bitar

## Future Meetings:

**2015**
- Professor Simon Murch, Warwick

**2016**
- Bids invited

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**BSPGHAN 2014 Annual Meeting**

Invited speaker biographies, abstracts and notes pages
Chronic Diarrhoea – a practical approach

Professor Simon Murch
University Hospital Coventry & Warwickshire

The important challenges posed by a child suffering chronic diarrhoea are to determine the diagnosis, maintain electrolyte balance and nutrition, and to deliver effective treatment to the underlying cause. The likely cause may vary by age. Clearly the first priority is to stabilise the child from the metabolic and fluid balance viewpoint, often requiring parenteral fluid administration.

Once the child has been stabilised, it is critical to determine whether the diarrhoea is a) osmotic, b) secretory or c) mixed osmotic and secretory. Clinical pointers towards osmotic diarrhoea notably include perianal erythema. Detection of reducing substances in stool is usual – providing that the fluid portion can be adequately captured. The critical clinical test is to determine whether faecal losses normalise while nil by mouth, which mandates iv fluid administration. If the history is chronic and the child emaciated, this may have to wait until parenteral nutrition is established.

Important initial investigations also include faecal electrolytes, faecal culture for pathogens, with specific culture for staphylococci and toxin secretion status if identified. Blood tests should include inflammatory markers. Any history of unexpected or prolonged infections should provoke in depth immunological assessment. If no cause emerges from initial screening, endoscopic assessment is required – the decision is whether both OGD and colonoscopy are required. If in a young infant, additional frozen and glutaraldehyde-fixed specimens may be needed for immunohistochemistry and electron microscopy – thus early liaison with a tertiary centre might spare the patient additional procedures.

1. **Presentation on first day of life - early diagnosis clinically vital.**
   a. Osmotic – Glucose Galactose malabsorption (n.b likely perianal erythema)
   b. Secretory – Congenital chloride rhoea, Congenital sodium losing enteropathy.

2. **Presentation in first week.**
   Mixed osmotic/secretory – microvillous inclusion disease, tufting enteropathy etc. Enteric infections – including staphylococcal enterocolitis.

3. **Presentation after first week.**
   Usually mixed osmotic/secretory – most common – undertreated food sensitive enteropathy. Most severe – autoimmune enteropathy. Multiple additional causes – thus extensive infectious/immunological work-up may be needed.
   Osmotic – after weaning – sucrase isomaltase deficiency, fructose malabsorption.
   Unexpected illogical pattern (osmotic, but continues while nil by mouth) – consider factitious/induced diarrhoea

4. **Presentation in later childhood.**
   A careful dietary history is needed. Coeliac disease and Inflammatory Bowel Diseases should be excluded, and an extensive search for enteric pathogens undertaken.
Dr Richard Thompson  
New investigations and therapies in liver disease  

Traditionally genetic testing has been used at a late stage in diagnostic pathways; generally to confirm a clinical, or phenotypic, diagnosis. In recent years our understanding of the genetic contribution to disease has increased enormously. However even this has been outstripped by quantum leaps in genetic testing technology. Genetics can now be used much earlier in a diagnostic pathway allowing early, accurate diagnoses to be made; saving considerable time and money. Next generation sequencing technology allows whole panels of genes to be sequenced simultaneously in several patients. The cost is often less than testing a single gene by conventional sequencing technology. The greatest savings are made where mutation of several genes may lead to a similar phenotype. Cholestasis, glycogen storage disease and immunodeficiency are good examples of early clinical application of this technology.

The speaker has been accused of bias, but it should now be obvious that many liver diseases, and a great many less obvious conditions, are either determined or modulated by bile acids. This understanding has led to renewed interest in manipulating bile salt pool size and flux. The tools to do this are becoming available.

Biography

Richard Thompson is a Senior Lecturer in Paediatric Hepatology at King’s College London, and Honorary Consultant in the Paediatric Liver, GI and Nutrition Service at King’s College Hospital. He attempts to combine research, teaching, diagnostics, clinical work and clinical trials.
James Lindsay – biography

Consultant Gastroenterologist Barts Health NHS Trust and Senior Lecturer at Barts and the London School of Medicine and Dentistry, Queen Mary University of London.

James Lindsay is a Senior Lecturer and Consultant in Gastroenterology at Barts and the London School of Medicine. Along with a full multidisciplinary team he runs the adolescent IBD service at The Royal London Hospital and the Adult IBD service at Barts Hospital. He is Chair of the ECCO education committee and organises the ECCO Advanced Course in IBD. He has chaired working groups for the ECCO consensus on the management of Crohn’s disease and ulcerative colitis.

He serves on the British Society of Gastroenterology IBD clinical trials steering group and is the chief investigator for a series of investigator led and commercial clinical trials in IBD. In addition, he leads two translational research programmes. One focuses on the interaction of the intestinal microbiota with the immune system with a particular interest the role of dendritic cells. The other studies the potential of using miRNA profiles to direct phenotype in IBD.

Dr. James Lindsay

New GI therapiess – Faecal transplantation

Dr Akash Deep – Biography

Having trained in Mumbai and PGI, Chandigarh in India I went to pursue paediatric intensive care training in the UK in 2004. After training in St Mary’s Hospital, Royal Brompton Hospital and Great Ormond Street Hospital London, I took up the Consultant position at King’s College Hospital in 2008

Currently I am the Director of Paediatric Intensive Care at King’s College Hospital, Research and CRRT Lead for PICU. I am the Chair of Liver Failure-Research Group in the Paediatric Intensive Care Society UK. I am the Chair for the Renal/CRRT Section of European Society of Paediatric and Neonatal Intensive Care Society (ESPNIC).

I am the co-Chair for the Science and Education Committee of Paediatric Intensive Care Society (PICS) of UK. I have been the Organising chairperson of the National CRRT workshops across UK and will be hosting the 8th International pCRRT Conference in 2015 in London

Special interests include acute liver failure in ICU, anticoagulation in CRRT especially use of prostacyclin, CRRT in liver failure and haemodynamics in septic shock.

Dr Akash Deep

Acute Liver Failure: Transport and PICU Care
Dr David Wareham
New antibiotic forms and effectiveness

Dr Anita Verma
CRE - The Newest Superbug - Learning From an Outbreak

The emerging superbug CRE means carbapenem resistant Enterobacteriaceae (CRE) and are major cause of health care-associated infections worldwide particularly, immunosuppressed patients or in patients with chronic illnesses. The Enterobacteriaceae group of bacteria i.e E.coli, Klebsiella spp which are normal gut flora have become resistant not only to last resort antibiotics carbapenem that is meropenem, ertapenem or imipenem but also to most other antibiotics. We are left with very limited therapeutic options for treating infections with these superbugs. The available antibiotic choice is colistin which is an old drug used in 1960s when many antibiotics were not available. The drug was not used widely because of its toxicity and poor therapeutic efficacy. Mortality associated with infections due to CREs are now surpassing the other alert organisms C.difficile or MRSA. Among different resistant mechanism for carbapenem, production of most versatile enzymes called carbapenemases is of concern. These superbugs are also called carbapenemase producing enterobacteriaceae (CPE). This resistance is carried out on a mobile genetic element (plasmid) as a result spreading the resistance not only to the same species of bacteria but to another genera of bacteria such as Pseudomonas, Citrobacter, Enterobacter etc. We can call them as carbapenem resistant organisms (CRO). These carbapenemases are classified into four classes A,B , C & D based on their active sites serine or zinc. The most widely prevalent carbapenemases worldwide are: KPC, Klebsiella pneumoniae carbapenemase (belong to class A); NDM, New Delhi metallo-ß-lactamase & VIM, Verona integron–encoded metallo-ß-lactamase (both belong to class B); OXA, oxacillinase (class D).

We describe the challenges and lesson learnt in preventing the spread of first outbreak of bla VIM-4 Metallo-ß-Lactamase (MBL) & OXA-48 producing Enterobacteriaceae in a pediatric unit. Two outbreaks due to bla VIM-4 MBL - producing Klebsiella & bla-OXA-48 Klebsiella pneumoniae were detected between September 2012 – March 2013. Robust and bundled infection control (IC) measures were implemented including CPE care plan, enhanced environmental cleaning, awareness, education & training, transfer guidance to other hospitals and discharge summary. The barriers to control the outbreak: poor adherence to IC precautions by caregivers, staff & visitors, persistence of the organisms in the environment, lack of awareness and parent /caregivers not trained for nappies care professionally which was the main reservoir for CREs. Most of patients were very young, with chronic illness, on immunosuppression, multiple admissions to local or referral hospital. The outbreak due to bla VIM-4 was contained with last positive case in December 2012 and due to OXA-48 producing Enterobacteriaceae in March 2013 without any significant morbidity or mortality. The bla VIM-4 MBL producing bacteria spread rapidly in immunosuppressed patients in comparison to OXA-48. Implementation of active screening for CRE helped us to detect the spread of these organisms by implementing early and robust IC practices particularly hand hygiene practices of health care providers and parents of babies on nappies is recommended. However with limited therapeutic options CRE/CPE are of serious concerns to clinicians, microbiologists and ID consultants. These emerging superbugs are ticking time bomb for the world.
Very early onset inflammatory bowel disease - the expanding spectrum of monogenic disorders

Inflammatory bowel disease (IBD), encompassing Crohn's disease and ulcerative colitis, has multifactorial aetiology with complex interactions between genetic and environmental factors. Over 150 genetic loci are associated with IBD. Recent studies have reported over 45 human monogenic diseases that can present with IBD-like intestinal inflammation. A substantial proportion of patients with those genetic defects present with very early onset of intestinal inflammation. Those genetic defects can be grouped into defects in intestinal epithelial barrier and stress response, immunodeficiencies affecting granulocyte and phagocyte activity, hyper- and autoinflammatory disorders as well as defects with disturbed T and B lymphocyte selection and activation.

In addition, there are defects in immune regulation affecting regulatory T cell activity and interleukin (IL)-10 signalling. Due to the broad spectrum of extremely rare diseases, establishing the correct diagnosis is frequently challenging and often delayed.

In many cases, categorization by means of established IBD histologic nomenclature is problematic and immunological analyses do not provide clear insight into the underlying genetic defect. However, a genetic diagnosis is key to allocating appropriate treatment options including medical therapy, surgery, or hematopoietic stem cell transplantation. Understanding of monogenic 'orphan' diseases cannot only provide treatment options for the affected patients but also inform on immunological mechanisms and complement the functional understanding of the pathogenesis of IBD.

Pancreatic diseases and their clinical management – Everything you need to know

Dr Tassos Grammatikopoulos

Is a Consultant in Paediatric Hepatology in the Paediatric Liver, GI & Nutrition Centre at King's College Hospital, London.

He has graduated from the Aristotile University Medical School in Thessaloniki, Greece and continued his general paediatric and paediatric hepatology training in the UK. He has been awarded the Alex Mowat Prize (ESPGHAN 2006) and the Rising Star Award (ILTS 2008) for his research work. He is a member of the British Association for the Study of the Liver, Royal College of Paediatrics and Child Health, American Association for the Study of Liver Disease, British Society of Paediatric Gastroenterology, Hepatology and Nutrition and European Society for Paediatric Gastroenterology, Hepatology and Nutrition.

He has a special interest in genetics of cholestasis, autoimmune liver and pancreatic disease and portal hypertension, subjects he has published work on. He is currently involved in clinical trials and research projects in the above topics. As part of the clinical network service with Scotland he is conducting the paediatric hepatology outreach clinics in Aberdeen and Dundee.

e-mail: t.grammatikopoulos@nhs.net
Dr Hilary Cass
Standards and Commissioning – what will that mean in paediatric gastroenterology?

Dr Hilary Cass is a Consultant in Paediatric Disability at Evelina Children’s Hospital, Guy’s & St Thomas’s NHS Foundation Trust and President of the Royal College of Paediatrics and Child Health.

She runs a national service for children with Rett syndrome and has published widely in this area. In addition to her neurodisability practice, she has been closely involved in the development of paediatric palliative care services at Evelina Children’s Hospital. She is also co-leading a programme to develop an integrated primary-secondary service for children in Lambeth and Southwark.

Dr Cass has held senior education and management roles in hospital Trusts, and was previously Head of School of Paediatrics in London. At a national level she has had a longstanding involvement in policy development for children’s healthcare, as well as advancing new models of paediatric service delivery.

Dr John Coakley
Strategy – what does that mean for healthcare professionals?
Dr Bernadette Moore Biography

Dr. J. Bernadette Moore obtained her Ph.D. in Nutritional Sciences at the University of Florida in 2002. Subsequently, she earned a Christine Mirzayan Science Policy Fellowship and worked for the Food & Nutrition Board of the National Academies’ of Science in Washington DC. There, working on the seminal report “Preventing Childhood Obesity: Health in the Balance”, she developed her research interest in the health consequences of obesity. After completing an intramural post-doctoral fellowship at the National Institute of Diabetes, Digestive and Kidney Diseases (NIH), Dr. Moore earned a Marie Curie Transfer of Knowledge Fellowship that permitted her to return to her home country, Ireland, in 2005. She joined the University of Surrey in November 2007. In less than six years she has established a young vibrant laboratory and raised over £800,000 in research funding. The research in her laboratory is focused on understanding the molecular basis of non-alcoholic fatty liver disease (NAFLD), the most common cause of liver disease affecting an estimated 30% of adults and 10% of children worldwide. There are currently no effective pharmaceutical therapeutics for NAFLD but nutrition is a key modifiable risk factor. The long-term vision of her work is clinical and public health translation with the overarching goals of addressing unmet needs for discovery of diagnostic/prognostic biomarkers (stratified medicine) and development of novel therapeutics. Specifically, current approaches include: (i) utilizing proteomics for the identification of new biomarkers and (ii) developing and applying new systems biology tools to dissect the molecular etiology and pathogenesis of NAFLD.

Dr Bernadette Moore
School of Biosciences and Medicine, Faculty of Health and Medical Sciences, University of Surrey
GU2 7XH

Metabolic evidence for the mechanisms of liver disease in NASH

Non-alcoholic fatty liver disease (NAFLD) is now the most common cause of chronic liver disease in developed countries, affecting 20-45% of adults and an alarming 10% of all children. The term NAFLD encompasses a range of histopathology including simple steatosis, considered relatively benign, and non-alcoholic steatohepatitis (NASH), which can progress to advanced fibrosis/cirrhosis and end-stage liver disease. Patients with NASH have an increased risk of mortality from both liver- and cardiovascular-related causes. Progressive disease is strongly linked to obesity, insulin resistance and type 2 diabetes; however, the molecular mechanisms involved remain only partially understood.

While the initial (two-hit) hypothesis of NAFLD pathogenesis highlighted the role of excess reactive oxygen species in driving inflammatory processes, more recently the roles of lipotoxic intermediates and hepatic fatty acid trafficking have come to be appreciated. The dynamics of lipid droplet formation and the role of autophagy in fat mobilization is a very active area of research, although our understanding of how these processes may influence NAFLD pathogenesis remains rudimentary. However, it is clear that lipid droplets are not inert; lipolysis of triacylglycerol from lipid droplets is more dynamic and complex than previously envisioned and lipid droplet associated proteins, such as the PNPLA3 triacylglycerol lipase, play a role in NAFLD pathogenesis.

A handful of genetic variants have been identified as contributing to NAFLD risk and it is certain that additional genetic and epigenetic factors modifying NAFLD susceptibility remain to be identified. However, as the heritability of liver fat has been estimated to be only 39%, it is clear that environmental factors play a large role in NAFLD development. This talk will cover work from our laboratory and others that has expanded our understanding of the complexity of the molecular mechanisms underlying liver disease in NASH. We argue that full interpretation of NAFLD pathogenesis will require the application of computational and systems biology approaches currently the focus of our group.
Dr Martha Ford-Adams  
Obesity – what works

Dr Martha Ford-Adams is a consultant paediatrician at Kings College Hospital, with extensive experience in treating diabetes. Since her appointment in 2004 she noticed a steady increase in Type 2 diabetes in her diabetes clinic and the high prevalence of obesity in the local area. It became apparent to her that there was a need for an obesity service to compliment the busy adult service so with the help of colleagues in dietetics and psychiatry she established a service. The appointment of Mr Desai in 2006 with an interest in bariatric surgery led her to consider bariatric surgery for her patients and in 2011 their 1st patient was operated on successfully. With now a multidisciplinary team further surgeries have been carried out with good results. There is also collaboration with Institute of psychiatry looking at behavioural modification to treat obesity. Dr Ford-Adams and Mr Desai have also collaborated with Prof Thomas Inge in Cincinnati Ohio USA lead for “surgical weight loss programme for teens” and visited US early in 2013. Dr Ford-Adams is pleased to share the results of the pilot bariatric surgery programme and also explore what else “works” for obesity.

Dr E Kyrana, MD, MSc, MRCPCH, Dip Paed Nutrition

Dr Eirini Kyrana has completed training in paediatric hepatology at King’s College Hospital and is currently undertaking research in cachexia in children with end stage liver disease.

Dr Serena Kyrana
Models of Cachexia and body compositions

Cachexia is an emerging active area of clinical and experimental research in the fields of cancer, chronic heart failure and other chronic illnesses including chronic liver disease and inflammatory bowel disease. It is defined as “a complex metabolic syndrome associated with underlying illness and characterized by loss of muscle with or without loss of fat mass”. Cachexia may co-exist with, but is distinct from anorexia and malnutrition. Its presence increases mortality independent of disease severity. Patients with chronic liver disease frequently have muscle wasting, muscle weakness, fat loss as well as endocrine abnormalities like insulin resistance, all characteristic of cachexia. These patients have been shown to have worse outcomes whilst waiting for a liver transplant, but also after transplant. Research is underway to further define and describe the molecular mechanisms underlying this syndrome and treatment options are currently being tested in clinical trials.

Dr E Kyrana, MD, MSc, MRCPCH, Dip Paed Nutrition

Dr Eirini Kyrana has completed training in paediatric hepatology at King’s College Hospital and is currently undertaking research in cachexia in children with end stage liver disease.
Professor Ludovic Vallier

Stem Cells – How far away is a liver off the shelf?

End stage liver diseases target critical liver functions and must be dealt with orthotopic liver transplantation (OLT) since only a healthy donor liver can restore the missing metabolic functions. However, this ultimate solution does not represent a cure since it implies high risk of surgical complications, indefinite immunosuppression associated with severe side effects, and eventual rejection. Furthermore, the number of organ donors has remained constant the past 10 years while the demand for liver transplantation has more than doubled in the mean time. This situation will continue to worsen in the foreseeable future due to Hepatitis C pandemic and increase in cirrhosis associated with obesity. For all these reasons, the development of alternative therapies to OLT has become a major objective in the field of regenerative medicine.

Cell based therapies approach involving transplantation of healthy hepatocytes into the livers of affected patients may facilitate a complete correction of all aspects of the clinical syndrome especially with inherited metabolic disorders (IMDs). Unfortunately, cell based therapy in the liver is limited by the scarce number and quality of available donor cells. Indeed, hepatocytes are fully differentiated cells that are impossible to expand in vitro. As a consequence, cell based therapy of the liver also relies on organ donation and often only livers of poor quality are available for cell purification.

Pluripotent stem cells generated from reprogrammed somatic cells (human Induced Pluripotent Stem Cells or hiPSCs) represent an advantageous solution since they can proliferate indefinitely in vitro while maintaining their capacity to differentiate into a broad number of cell types including hepatocytes. In addition, hiPSCs could enable the production of patient specific cell types which are fully immuno-compatible with the original donor thereby avoiding the need for immune suppressive treatment during cell based therapy. For all these reasons, hiPSCs represent a unique opportunity for regenerative medicine. However, several challenges need to be solved before in vivo applications. Indeed, Current protocols of differentiation only allow the generation of foetal hepatocytes which display different functional activities as compared to adult primary hepatocytes. While such embryonic cells are useful to model IMDs in vitro, their capacity to colonise a recipient adult liver could be limited. In addition, the development of a highly efficient method to edit the mammalian genome represents a key challenge to generate “personalized” hiPSCs useful for cell based therapy in the context of genetically inherited human disorders. Here, I will review the latest technological developments which could help to address these challenges and to deliver the clinical promises of hiPSCs.

Dr Nikhil Thapar

Stem Cell Science – The GI promise

Dr Thapar is a Senior Lecturer and Academic Lead for gastroenterology at University College London's Institute of Child Health and honorary consultant in Paediatric Gastroenterology at Great Ormond Street Hospital for Children. He runs a specialist multidisciplinary clinical service for children with gastrointestinal motility and functional disorders including a national service for children with intestinal pseudo-obstruction. Dr Thapar's research programme focuses on the pathogenesis and treatment of gut motility disorders including molecular mechanisms and regenerative medicine. Dr Thapar sits on committees of the European Society of Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN), United European Gastroenterology, UK’s Medicines for Children Research Network and chairs the British Society of Paediatric Gastroenterology Hepatology and Nutrition gut motility disorders working group. Dr Thapar is co-editor of the textbook of Paediatric Neurogastroenterology and co-director of the Academy of Paediatric Gastroenterology.
**Dr. Ieuan Davies**

**Endoscopy Training and Standards – The JAG approach**

Dr. Ieuan Davies
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I am a consultant in Cardiff and am fortunate to work in a clinical network covering South Wales. I chaired the BMA Wales Junior Doctors’ Committee, was a member of the BMA Medical Ethics Committee and later chaired the RCPCH Trainees Committee. Prior to taking up my consultant post I was on secondment to the Welsh Assembly Government advising on the extension of the Working Time Directive to training grade doctors.

I was elected to Chair the BSPGHAN Endoscopy Working Group two years ago and as a consequence am a member of Joint Advisory Group on GI Endoscopy (JAG). I am also chairing a NICE Guideline Development Group on Gastro-oesophageal reflux disease in children and young people.

In this talk I shall outline the progress being made by BSPGHAN / JAG with training, assessment and quality assurance in respect of Paediatric GI endoscopy.

I would welcome e-mailed comment and suggestions.

No conflict of interest to declare.

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**Dr. Gillian Harris**

**Achieving normal intake**

There are five main factors which contribute to the young child’s acceptance of a diet comprising a range of tastes, foods and textures.

- Family eating habits and environment i.e. what food is available, parental strategies
- Experience with tastes within the weaning window.
- Experience with textures during the sensitive period for introduction of solids.
- Sensory sensitivity to visual, tactile stimulation and tastes (innate or induced).
- Appetite (Plate clearers or “fussy”).

Each of these factors will be discussed in relation to ages and stages and how these might interact with periods of ill health.

**Gillian Harris; Short biography**

Dr. Gillian Harris is a Consultant Paediatric Psychologist, and runs a feeding clinic at The Children’s Hospital, Birmingham, where she works with infants and children who have complex medical conditions and who are food averse. Dr. Harris has also carried out research into infant and child feeding behaviour and appetite regulation at the University of Birmingham, School of Psychology for the past 28 years.

Her specific research and clinical interests are: the development of food acceptance and rejection in early infancy and the toddler period; the consequence to later food preference of early experience; and the development of avoidant eating behaviour in older children.

Dr. Harris has published extensively in peer reviewed journals, and written many popular factsheets and booklets for parents to help them with the infant and child feeding process.
Dr Mark Powis – Biography

My talk will be an overview of what we currently require, how we try to achieve that for trainees and comparison with JAG recommendations.

I am a Consultant Paediatric Surgeon at Leeds Teaching Hospitals NHS Trust. I have interests in GI Surgery, Minimally Invasive Surgery and Oncology. I am also interested in teaching and education. I am Training Programme Director for Paediatric Surgery in Yorkshire, Lead for National selection into Paediatric Surgery and a member of the Specialist Advisory Committee for Paediatric and Core Surgical Training.

Dr Mark Powis

Endoscopy Training and Standards – The BAPS approach

Talk will be an overview of what we currently require, how we try to achieve that for trainees and comparison with JAG recommendations.

Paolo De Coppi, MD, PhD

Reader and Consultant Paediatric Surgeon, Head of Surgery Unit at UCL Institute of Child Health and Great Ormond Street Hospital, London, UK

Mr. De Coppi is a Consultant Paediatric Surgeon at the Great Ormond Street Hospital for Children and Reader and Head of the Surgery Unit at the UCL Institute of Child Health both located in London, England. Concomitantly, from 2009 he has been an Adjunct Assistant Professor at the Wake Forest Institute for Regenerative Medicine, Wake Forest University, Winston-Salem, North Carolina and from 2005 he has been an Honorary Assistant Professor in pediatric surgery, University of Padua, Italy.

He has a special interest in congenital malformations and their treatment using minimally invasive techniques. He has focused his research interests on stem cells and tissue engineering by trying to find new modalities for the treatment of complex congenital anomalies. While working with Anthony Atala, M.D., at the Boston Children’s Hospital (Massachusetts), he identified the possibility of using stem cells from amniotic fluid for therapeutic applications. This finding generated an international patent and garnered the cover story of Nature Biotechnology January 2007. This finding has opened the door to discovery for novel approaches to correct congenital malformations. More recently, his team has demonstrated that these cells are able to differentiate into various tissues and to replace functional activity in animal model of diseases. He is now focused on developing reliable methods for stem cell isolation, expansion and differentiation at a clinical level (GMP-grade). Finally, in 2010 he was part of the team that performed the first successful transplantation of a tissue-engineered trachea on a child at the Great Ormond Street Hospital.

He has published more than 100 peer-reviewed articles in journals such as The Lancet, Nature Biotechnology, Blood and FASEB Journal; supervised more than 25 research fellow and Ph.D. students; and has been awarded various national and international grants. Since 2009, he has been on the editorial boards of Paediatric Surgery International, Stem Cell Development, and Fetal and Maternal Medicine Review. As of 2011 he has been associate editor for Stem Cell Translational Medicine.

Abstract

Necrotizing enterocolitis (NEC) is a devastating inflammatory disease of the gastrointestinal tract and a major cause of morbidity and mortality (20-30%) in premature neonates. The disease occurs in 0.1-0.3% of live births. As the overall survival rate of premature neonates improves, the relative risk of NEC increases, and it threatens to surpass respiratory disease as the major cause of deaths in this vulnerable population. Current management is surgical removal of affected intestine and supportive medical therapy and has not changed significantly in the last 20 years. Patients who survive may develop short bowel syndrome as a result of massive surgical resection, with subsequent dependence on parenteral (IV) nutrition or need for intestinal transplantation.

Insertions of primary peritoneal drainage in comparison with laparotomy remain controversial, and this uncertainty stimulated the development of two randomized controlled trials. The mortality of the disease remains high, and new therapeutic interventions are needed. Novel forms of treatment that can improve the outcome of this disease are currently under investigation. These include whole-body moderately controlled hypothermia and administration of amniotic fluid stem (AFS) cells.

AFS cells have intermediate characteristics between embryonic and adult stem cells. They can be easily reprogrammed to a pluripotent status. AFS cells injected intraperitoneally in a HSA-Cre, Smn(F7/F7) Mouse Model, Stem Cells, 2012 May 29; doi: 10.1002/stem.1134.

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Mr Bruce Jaffray

Fundoplication - the surgical decision

Surgery for complex inflammatory bowel disease

The increasing incidence and severity of inflammatory bowel disease (IBD), among British children has led to more children undergoing surgery for their condition. The presentation, surgery, complications and outcomes of 100 consecutive intestinal resections for Crohn’s disease, and 56 children receiving an ileo-anal pouch procedure for colitis by one surgeon are discussed.

Particular emphasis is placed on the development of techniques specific for children to allow any age of child to receive an ileal pouch. The use of laparoscopic techniques for both Crohn’s disease and colitis is discussed.
Mr Mark Davenport
Surgical interventions in intestinal failure

Mr Gregor Walker
BAPS – CASS Hirschsprung’s Study
The main condition requiring colectomy in children beyond infancy is inflammatory bowel disease. Rarely, aggressive polyposis syndromes merit colectomy in children; and the indications for colectomy in pseudo-obstruction will be covered in another presentation.

In ulcerative colitis, the need for colectomy has lessened with the advent of anti-TNF medication in the management of children who do not enter remission with a combination of 5-ASA and intravenous corticosteroids. (It is not the purpose of the presentation to compare the merits of ciclosporine and infliximab.) Nevertheless, the indications for surgery remain as they have done for decades: (i) failure to enter remission on medical therapy; (ii) toxic megacolon, which does not settle promptly on conservative management; and (iii) corticosteroid dependence that cannot be ameliorated with other immunosuppressants. In each case, it is important to involve the surgical team before the decision to undertake an operation. The aim is for the child to undergo surgery in an orderly manner, rather than as an out-of-hours emergency. Because of the extensive nature of the disease when it comes to surgery, subtotal colectomy is the operation of choice in almost every case of ulcerative colitis. Laparoscopic surgery is increasingly used, but the decision on whether open surgery may be better should be carefully considered. Ultimately the creation of a pouch will be necessary as a later procedure and the need for later operations should be discussed with children and their families at the outset. The number of operations that a child will need is debated among paediatric surgeons; but the need for a competent surgeon to annually undertake a large number of cases in reconstructive surgery is universally acknowledged.

In Crohn’s disease, the theoretical indications for colectomy are the same as for ulcerative colitis. However, in practice, toxic megacolon is rare, and severe uncontrollable colitis requiring urgent colectomy is very uncommon. Most cases are undertaken for failure of medical treatment. These can be categorised into operations that are needed to control the colitic features, and those necessary to accelerate linear growth. Surgery is therefore sometimes needed in the child with growth retardation, where it would not be performed in the fully grown child. Because of the discontinuous nature of the lesions in Crohn’s disease, total colectomy is less often required than a right hemicolectomy, and a restorative proctocolectomy is not possible because of the transmural inflammation that can occur. Because inflammation directly affects linear growth, all resected tissue should be removed and inflamed intestine should not be left in the child. (This is not to say that occasionally a diversion may not be necessary for severe perianal disease). Anecdotal evidence suggests that while right sided colectomy is very successful for proximal disease in the colon; for extensive distal disease, left hemicolectomy has a poor efficacy and a total colectomy is usually needed.

The long term management of children after colectomy returns to the paediatric gastroenterologist after the post-operative phase.
Mr Khalid Sharif
The Future Transplantation of Digestive Organs

Mr Khalid Sharif is a Consultant Paediatric Hepatobiliary and Transplant Surgeon working at Birmingham Children's Hospital.

He trained in Pakistan and moved to the UK in the late 1990s. He then trained under Mr Jean de Ville de Goyet at Birmingham Children's Hospital.

Mr Sharif now leads the paediatric hepatobiliary and transplant programme at Birmingham Children's hospital undertaking 30-40 transplants each year including isolated liver, combined liver and kidney, isolated small bowel and combined liver and small bowel.

His hepatobiliary works focuses on Kasai procedures for babies with Extra Hepatic Biliary Atresia and tumours.

He is now an internationally recognised leader for hepatobiliary and paediatric transplant training.

Mr Joe Curry
Surgery in pseudo obstruction
Constitutive Type I Interferon, via STAT1 activation, promotes regulatory T cell function in the healthy human intestine, but not in inflammatory bowel disease

Giles, Edward; Pathak, Mohini; Sanders, Theo; McCarthy, Neil; Sanderson, Ian; MacDonald, Tom; Lindsay, James; Stagg, Andrew. Centre for Immunology and Infectious Disease, Blizard Institute, Barts and the London School of Medicine and Dentistry, London

Background

Control of T-cell reactivity with the human intestinal mucosa is poorly understood. Type I Interferon (T1IFN) signals via the JAK/STAT pathway, particularly STAT1, and has only recently been thought to be active outside of viral infections. Specifically, T1IFN has been shown to constitutively support Treg function in mouse models of colitis. T1IFN has been used as a treatment in Inflammatory Bowel Disease (IBD) with some success. We therefore hypothesised that constitutive T1IFN had a regulatory role in human intestinal T cells.

Methods

Endoscopic biopsies or resection specimens were frozen for immunohistochemistry (IHC) or cultured in the presence of neutralising anti-IFNß or isotype-matched control antibody. Cells were harvested, stimulated with anti-CD3/CD28 antibodies and analysed for cytokine production by intracellular staining and by multiplex ELISA of culture supernatants. Phosphorylated STAT1 was measured by flow cytometry with or without prior T1IFN stimulation. Frozen sections of colonic mucosa were stained with anti-IFNß and analysed using fluorescent IHC. Finally, CD3+ T-cells were FACS sorted and expression of Interferon Stimulated Genes (ISGs) determined by quantitative real-time PCR.

Results

IFNß was detected in the lamina propria of both control and IBD tissue and ISGs (MXA and 250AS) were expressed by intestinal T cells sorted from tissue. In vitro, IFNß neutralisation reduced the frequency of pSTAT1+ intestinal T cells (n=6, p=0.05) and, in healthy controls, decreased the proportion of IL10-producing intestinal T cells (n=8, p=0.01). Blocking IFNß also led to a trend for more IFNß-producers (p=0.059) and IFNß concentrations in supernatants were significantly increased (n=10, p=0.016). In IBD, intestinal T cells were more responsive to IFNß in vitro, as assessed by ISG induction, (n=10 for patients and controls, p=0.03) and constitutive pSTAT1 was increased in T-cells isolated from non-inflamed mucosa of IBD patients compared with controls (n=30 IBD, 16 control, p=0.03). In contrast to control tissue, neutralisation of IFNß in non-inflamed IBD samples led to a generalised increase in cytokine production, with a significant increase in the frequency of T cells producing all cytokines examined (IL-10, IFNß, IL-17 and TNFß, n=10).

Discussion

T1IFN is present in the mucosa of the human intestine and in health may have a regulatory role by selectively supporting T cell production of IL-10 (Tregs). There is increased responsiveness of the T1IFN pathway in T cells from non-inflamed tissue from IBD patients compared with controls associated with a generalised suppression of cytokine production. Thus, immunoregulatory effects of intestinal T1IFN are context dependent and may modulate inflammation in healthy and dysregulated IBD tissue by distinct mechanisms. This may explain the varied response to T1IFN as a therapy in IBD.
Aims and Methods
A retrospective, population-based cohort study of children who became dependent on Infliximab (IFX) and/or Adalimumab (ADA) for at least 12 months, was reviewed. Risk factors for dependency and the impact on clinical outcomes were assessed in a Scottish paediatric IBD population. 

Results
Data was collected from 1.1.00 until 01.09.12 with follow up until 31.12.12. 188 children were given Infliximab (IFX) and/or Adalimumab (ADA) in a paediatric centre. Dependency was defined as ceasing biologicals prior to 12 months due to lack of response or remission, or due to adverse event. 31/62 (50%) stopped treatment after 12 months (61% IFX and 32% ADA). Of these, 16 (26%) patients had a reassessment colonoscopy 6 months after starting on Infliximab which showed complete mucosal healing (Rutgeerts score i0), 3 were on treatment with Infliximab and 10 were on Adalimumab. Seven children had reassessment colonoscopy after their right hemicolectomy and we have used CDEIS to assess their mucosal healing. 4 children had higher Rutgeerts endoscopic grading scale to assess their mucosal healing status. 7 children had reassessment colonoscopy from October 2012 to September 2013. In 51 children, we have used CDEIS to assess mucosal healing. 20 (65%) of these children had achieved complete mucosal healing and their CDEIS score varied from 3 to 25. Significantly higher proportion of children in the complete mucosal healing group had received treatment with anti-Tumour Necrosis Factor (anti-TNF) agents compared to the children in the other group (65% v 32%, p value=0.04). The anti-TNF agents used in the children in both groups were Infliximab. There were no significant difference in disease distribution, peri-anal involvement, Haemoglobin, CRP, ESR, platelets and Azathioprine use when comparing children in both the groups.

Conclusion
In our experience 42% (24/57) children with Crohn’s disease on long term maintenance treatment achieved complete mucosal healing. Children who had achieved complete mucosal healing did not need bowel resection surgery and Rutgeerts endoscopic grading scale for children who had right hemicolectomy.

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The epidemiology and management of acute pancreatitis in children

Dr Farah Mushtraq (Princess of Wales Hospital, Bridgend), Dr Mike Coagrove (Morriston Hospital Swansea), Mr Kim Hutton, Dr Huw Jenkins & "Dr Ieuan Davies (all University Hospital of Wales).

Introduction:
The aetiology of acute pancreatitis in children is diverse and the clinical features vary. There is a paucity of accurate prospective epidemiological data regarding this condition although studies suggest the incidence appears to be rising.

Aims:
To identify and investigate all cases of acute pancreatitis over a 12 month period in Welsh children.

Method:
This was a prospective Welsh Paediatric Surveillance Unit study. To be included children needed at least two of the following three criteria: abdominal pain, an elevated amylase >3 times the upper limit of normal and / or confirmatory findings on abdominal imaging. Affected children were notified by consultant paediatricians in Wales using the “green card” system. In addition, neighbouring secondary and tertiary colleagues from England were involved as well as the Welsh Chemical Pathology departments to ensure complete capture.

Results:
There were only 15 confirmed cases identified in 1 year (12 female / median age 11 years). 3/15 cases had hereditary pancreatitis and all these children had had previous episodes of acute pancreatitis (2 children had the PRSS mutation and 1 child had the SPINK mutation).

The other causes included: Gallstone related disease (5), idiopathic (2) and iatrogenic (2). Of the two iatrogenic related episodes the drugs were Sodium Valproate in 1 case and a combination of treatments in a child with acute leukaemia in the other case. Diabetic ketoacidosis, severe sepsis and haemolytic uraemic syndrome were seen in 1 child each. Older children presented with more classical adult like features of the condition.

Amylase levels ranged from 369 to 3481. The duration of hospital admission varied from 3 days to 63 days (mean 13 days). Parenteral nutrition was only given in 2 very sick children each with a prolonged illness course.

Intravenous fluid and analgesia was provided in all cases. Fluid resuscitation was required in 3 cases. Eight children were kept nil enterally for 2-3 days with 5 children nil enterally for 4-6 days. 7/15 children were given antibiotics with 6 of those children showing signs of infection. There were no obvious pancreatitis related complications identified during our study.

Summary and conclusions:
The incidence of acute pancreatitis in children was estimated as 2.7 per 100,000 / year. The most common risk factors were gallstone disease or hereditary pancreatitis. The management always included hospital admission, intravenous hydration and analgesia. No early complications were identified over the study period but 3/15 children are known to suffer from recurrent pancreatitis.

The effect of commonly used IBD drugs on autophagy induction using an in vitro cell culture system

Henderson, Paul; Satsangi, Jack; Wilson, David C; 1University of Edinburgh, Edinburgh Stevens, Craig; 2Edinburgh Napier University, Edinburgh

Background:
Genome wide association studies and functional experiments in inflammatory bowel disease (IBD) have delineated the importance of autophagy in IBD pathogenesis. We aimed to determine the effect of commonly utilised IBD drugs on autophagy induction and the pathways involved in vitro.

Methods:
Cells naturally expressing (HCT116) and not expressing (HEK293) NOD2, both stably expressing green fluorescent protein-labelled light chain 3 (LC3), were treated with varying concentrations of 6-thioguanine, azathioprine, methotrexate or infliximab at different time points; rapamycin, serum-starvation and bafilomycin A1 served as positive controls. Cells were also treated with ERK (U0126) and autophagy (3-methyladenine) inhibitors where appropriate. For immunofluorescent microscopy images were captured using an Axioskop 2 fluorescence microscope and ImageJ software used to identify cells with >5 punctate foci indicating autophagy induction. For western blot analysis cell lysates were immunoblotted with antibodies to LC3, p62, phospho-rpS6 or total rpS6. All statistical analyses were performed using GraphPad Prism.

Results:
All four drugs induced significant autophagy in HCT116 cells, with only azathioprine inducing autophagy robustly in both cell lines. Azathioprine induced autophagy in a dose-dependent manner in HEK293 cells with significant autophagy induction at all concentrations (30-90μM) in HCT116 cells. HCT116 cells treated with 6-thioguanine, azathioprine and methotrexate showed strong LC3-I to LC3-II conversion and a reduction in p62, with 6-thioguanine and azathioprine showing loss of phospho-rpS6 suggesting autophagy induction through the mTORC1 pathway. Use of U0126 and 3-methyladenine in HCT116 cells treated with azathioprine demonstrated that azathioprine may exert its autophagic effect via mTORC1 through the class I PI3K/Akt pathway.

Conclusion:
Common IBD drugs effect autophagy induction in vitro suggesting that manipulation of the autophagy pathway may be partly involved in the mechanism of action of many of these drugs, most convincingly azathioprine. Further work is now required to replicate these findings and further delineate the pathways in vivo.
Autoimmune pancreatitis in children; evolving diagnosis and management
Grammatikopoulos Tassos1; Zen Yoh2; Karani John3; Bogdanos Dimitrios4; Pavlidou Maria1; Harrison Phil5; Devlin John6; Miel-Vergani Giorgia1; Dhawan Anil7; 1Paediatric Liver Centre, King’s College Hospital NHS Foundation Trust, London; 2Institute of Liver Studies, King’s College Hospital NHS Foundation Trust, London; 3Department of Radiology, King’s College Hospital NHS Foundation Trust

Background:
Autoimmune pancreatitis (AIP) in children is an increasingly recognized diagnosis over the last few years. Diagnostic radiological, histological and serological criteria are adult derived with some limited application in children.

Aim:
To identify the features of children with AIP in our centre and describe the disease progression.

Methods:
Clinical, biochemical, histological and radiological data were reviewed in children diagnosed with AIP from 2006 to 2013.

Results:
Six (5 F) patients with AIP were identified. A combination of abdominal pain, raised serum amylase with autoimmune markers, bile duct and pancreatic radiological involvement with histological features guided us to the diagnosis. Hereditary causes were excluded. Median age at presentation was 11.5 years (range, 9.5–15.5). All patients presented with abdominal pain. Jaundice and pruritus was also present in 3. Ulcerative colitis was diagnosed in 2. No other organ involvement was reported. Median serum amylase, lipase, triglycerides and total bilirubin were 400 IU/L, 245 IU/L, 1,4mmol/L, and 45mmol/L, respectively. Anti-nuclear antibodies were positive in 2 and raised immunoglobulin G subclass-4 (IgG4) (2.8 g/L [nv, 0.23-1.1]) in 2 with normal total IgG (11g/L [nv, 5.4-16.1]) levels. Ultrasound Scan (USS) and Magnetic Resonance Cholangiopancreatography (MRCP) were performed in all with evidence of chronic pancreatitis. MRCP showed pancreatic atrophy (2), pancreatic head enlargement (5), pancreatic duct irregularity (3) and common bile duct dilatation (4). Endoscopic Retrograde Cholangio-Pancreatography was performed in 4 and 3 required stent insertion for biliary obstruction. Three patients underwent uneventful Endoscopic Ultrasound ampullary (2) and Computed Tomography pancreatic (1) biopsies with evident inflammation but limited number of IgG4+ve cells. Five were treated with corticosteroids; azathioprine was added in 1 as a steroid-sparing agent due to steroid induced diabetes. One patient who presented in 2006 underwent hepaticojejunostomy for biliary obstruction due to pancreatic head enlargement and AIP was diagnosed retrospectively. Clinical and serological remission (median amylase 48 IU/L, bilirubin 1.4mmol/L and 45mmol/L, respectively. Antinuclear antibodies were positive in 2 and raised serum amylase (2), lipase (2), triglycerides (2) and total bilirubin (1). Ultrasound Scan (USS) and Magnetic Resonance Cholangiopancreatography (MRCP) were performed in all with evidence of chronic pancreatitis.

Conclusions:
AIP is an evolving condition in children but diagnostic criteria still need to be established. They should include implementation of IgG4 serology, radiological imaging and lately histology, as prompt diagnosis is essential for effective immunosuppressive treatment.

Cost effectiveness of single visit parenteral iron (SVPI) infusions in children with Inflammatory bowel disease (IBD)
Rhona Hubbard, Dr P Narula, Dr M Thomson, Dr P Rao, Dept. of Paediatric Gastroenterology, Sheffield Children’s Hospital NHS Foundation Trust, UK

Background:
Iron deficiency is a common problem in children with inflammatory bowel disease. Intolerance to oral iron, abnormal absorption due to surgery or gastrointestinal disease, significant bleeding, and non-compliance may make oral iron treatment in some patients inadequate. We have previously published an abstract demonstrating the safety and efficacy of single visit parenteral iron infusions (SVPI) at our center with Monofer®.

Objectives:
To compare the costs and savings (if any) from the use of single visit parenteral iron preparations vs (previously used) multiple iron sucrose infusions vs blood transfusions for indicated iron deficient patients with IBD.

Methods:
Data on all patients with IBD and iron deficiency (Hb<10 gm/dl) that were given SVPI infusions (Monofer®) was retrieved from a database for the period April 2010- Sep 2013. Product costs were sourced from pharmacy. Human resource costs and Healthcare resource group codes were sourced from dept. of finance

Results:
10 paediatric patients with IBD (7 males, age range 7-16 years) had an uneventful 1 hour single parenteral infusion (SPI). The table below compares current treatment costs with previously used options to bring Hb up to a desired level.

Cost comparative and net income of 3 modalities on a per patient per treatment basis

<table>
<thead>
<tr>
<th>Blood Transfusion</th>
<th>Iron sucrose</th>
<th>SVPI (MONOPER®) 100mg via Infusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Product cost</td>
<td>£800 (2x2bags each of packed RBC)</td>
<td>£160 (10x100mg vials @ £10.20)</td>
</tr>
<tr>
<td>Daycare room usage</td>
<td>8 hours (4 hours 2)</td>
<td>40 hours (10 slots of 4 hours)</td>
</tr>
<tr>
<td>Daycare nursing cost Band 5 (£14 per hour)</td>
<td>£102</td>
<td>£560 (40 hrs)</td>
</tr>
<tr>
<td>Consultant (Mid-point – £60 per hour)</td>
<td>£480</td>
<td>£240</td>
</tr>
<tr>
<td>TOTAL COST PER PATIENT</td>
<td>£1382</td>
<td>£902</td>
</tr>
<tr>
<td>Payment from PCT (HRG)</td>
<td>£536 (10 x £63.60)</td>
<td>£936</td>
</tr>
<tr>
<td>Efficiency savings (Daycare hours saved)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Income generation by use of saved daycare hours for Infliximab infusions</td>
<td>700 (per IFX infusion x 10 = £7000)</td>
<td></td>
</tr>
<tr>
<td>NET INCOME</td>
<td>£-581</td>
<td>£5458</td>
</tr>
</tbody>
</table>

SVPI generated a cost saving and net income of 7584.60£ per patient. Blood transfusions resulted in a net loss of 581£ per case due to high product cost and use of daycare space.

Conclusion: Daycase blood transfusions and Multiple iron sucrose infusions consume significantly more resources and hence SVPI is a far more cost effective option in providing a higher net income. In our trust, the use of SVPI for 10 patients with IBD allowed for ‘freeing-up’ of 390 hours of daycare time which in turn was used for Infliximab infusions ( generating a total net income of 7568.60£ over 3 years).
MRI assessment of body composition in paediatric Crohn’s disease; intra-abdominal adipose tissue association with disease severity

Thangarajah D1; Chappell KE2; Gale C1; Parkinson JRC1; Epstein J3; Hyer W3; Soondrum K3; Frost G4; Fall JME3
1Section of Academic Neonatal Medicine, Imperial College, London; 2Department of Radiology, Chelsea and Westminster NHS foundation trust; 3Paediatric Gastroenterology department, Chelsea and Westminster NHS foundation trust; 4Nutrition and Dietetic Research Group, Faculty of Medicine, Imperial College Hammersmith Hospital

Background:
Paediatric Crohn’s disease (CD) is associated with malnutrition, poor growth and alterations in body composition. Intra-abdominal adipose tissue (IAAT) is the adipose compartment most strongly associated with chronic inflammation; intestinal adipose tissue expansion observed in surgical specimens is a recognised hallmark of CD. Adipocytes can function as macrophage-like cells in the inflammatory cascade, releasing adipokines such as IL-6, TNF-β.

Study aims.
For the first time we use magnetic resonance imaging (MRI) as a method of measuring body composition in paediatric CD, specifically for quantifying intra-abdominal adipose tissue (IAAT).

Methods:
Children (7-18 years) with CD were recruited from a tertiary Paediatric Gastroenterology department; healthy children were recruited to act as controls from general paediatric outpatients, Chelsea and Westminster hospital. Ethical approval was obtained. Volumes of the following abdominal compartments; Total abdominal adipose tissue (TAAT), IAAT, subcutaneous adipose tissue (SCA) and abdominal muscle (MU) were quantified from MR images for all subjects; volumes were expressed in litres.

Analysis:
Compartment volumes were adjusted for body size by derivation of a height (Ht) index for each compartment (Compartment/Ht²). They were also expressed as a ratio of TAAT:MU, and IAAT:SCA. Measures were analysed according to disease activity; remission/mild (PCDAI 0≤29), moderate/severe (PCDAI≥30). We have not yet fulfilled our recruitment target thus we present the descriptive statistics.

Results:
29 children were recruited; Mean age (±SD) (Controls (C): 14.4 ± 1.9yrs, n=6 (∧2); remission/mild (R/M); 14.2 ± 2.2yrs, n=12 (∧6); moderate/severe (M/S): 13.5 ± 1.4yrs, n=11 (∧5); Mean BMI (C): 19.3 ± 2.7kg/m²; R/M: 19.6 ± 4.3kg/m²; M/S: 16.6 ± 2.9 kg/m². The median [IQR] of TAAT:MU for each group; C: 0.61 [0.52-0.83]; R/M: 0.90 [0.60-1.50]; M/S: 1.26 [0.87-1.76]. For IAAT:SCA; C: 0.42 [0.27-0.54]; R/M: 0.37 [0.23-0.71]; M/S: 0.68 [0.36-1.20]. At the time of MRI scan no-child was on systemic steroids. The plots in Figure 1 represent the median Compartment/Ht² (bar), IQR (box) and range (error bars).

Conclusions:
Using MRI methodology we show that IAAT and related compartments can be quantified in children with CD. Our preliminary results indicate that severe disease is associated with lower muscle mass and higher IAAT. In severe disease despite lower BMI, there is evidence of higher IAAT; this implies that IAAT is mediated by local gastrointestinal inflammation.
Burden of care at night when living with a child on parenteral nutrition at home.

Hughes A, Koegelmeier J, Hill S.
Intestinal Failure Rehabilitation Unit, Dept Gastroenterology, Great Ormond Street Hospital, Great Ormond Street, London

Background

Children with severe Intestinal failure (IF) that has failed to respond to treatment are discharged home after extensive investigation in our specialist IF rehabilitation centre. The parents undergo a formal 2-week training programme to manage the PN and have sole responsibility for completing the PN connection and disconnection and over-night care. We endeavour to offer professional support with overnight care, but parents perform all PN connections and disconnections. These patients have complex medical needs and as well as the PN the parents will need to give medications, enteral feeds and deal with children who are often in pain and uncomfortable, both day and night. On discharge the team encourages parents and carers to return to ‘everyday’ life attending work and the children going to school and other usual childhood activities. We have been aware of a high incidence of night-time disturbances that has led us to try to understand if we are asking too much of the parents when undertaking all this care, in most cases without extra help at home.

Aim

The aim was to discover how much work parents and carers complete overnight for children who are dependent on PN.

Subjects and Method

Parents of all 32 patients who attend a specialist intestinal rehabilitation clinic and have been on treatment for PN at home for > 6 months were asked to complete a questionnaire regarding how often they are woken overnight and the tasks that they complete at night. All children were receiving PN as a single bag system using a portable pump. All school age children were attending full time school. 29 patients were receiving PN over 12 hours and 3 children for 24 hours. The children were analysed according to their underlying illness to see if this had an effect on volume of night work.

Results

18 of the 32 families responded to the questionnaire. The 18 children were aged 7 months -12.7 years (mean age 6 years). There were 9 female and 9 males. With the patients having been at home on PN for between 2 months and 12 years (mean 2.5 years). Parents reported that they were woken up from 2- 8 times every night (average 4 times a night). The reason for waking was toileting in 28% of all incidents, for pump alarming 24% and medication in 22%. Abdominal pain affected 16% and emptying a gastrostomy bag on free drainage was needed in 4%. When analysing according to underlying disease the children with multi-system disease with associated IF were disturbed the most overnight, with an average of 5.7 times a night, those with motility conditions 4.2 times a night, children with short bowel syndrome woke 3 times a night and those with an enteropathy an average of 2.3 times a night.

Conclusion

In conclusion, none of the children and parents questioned managed to get a complete night of sleep, although they are expected to complete normal activities of daily living every day. We also need to follow up the 14 families who have not yet responded. Giving medication at nights happens more frequently than expected and this should be discussed with parents and patients before discharge to coordinate their medication to within daytime hours. Perhaps the only exception should be analgesia treatment, but pain only accounted for 16% of the patients before discharge to coordinate their medication to within daytime hours. Perhaps we need to offer professional support with overnight care, but parents perform all PN connections and disconnections. These patients have complex medical needs and as well as the PN the parents will need to give medications, enteral feeds and deal with children who are often in pain and uncomfortable, both day and night. On discharge the team encourages parents and carers to return to ‘everyday’ life attending work and the children going to school and other usual childhood activities. We have been aware of a high incidence of night-time disturbances that has led us to try to understand if we are asking too much of the parents when undertaking all this care, in most cases without extra help at home.

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Long term outcome of children with intestinal failure in the UK

H. Gowen1, Lloyd, C1, JW Puntis1
1Birmingham Children’s Hospital, Birmingham, UK, and 1The General Infirmary at Leeds UK

Introduction

The British Intestinal Failure Survey (BIFS) was set up in 2005 to establish the incidence, causes and outcomes of IF in children in order to inform service planning. Incomplete case ascertainment has led recently to a change in the methodology of reporting (‘Paed elBans’). This study therefore summarises insights into IF gained during the eight years that BIFS has collected data.

Subjects and Methods

Children with IF (defined as parenteral nutrition dependant ≥28 days) were registered by 25 UK participating centres. Data collected included diagnosis/cause of IF, age at commencement of PN, length of time on PN, and status (alive, PN dependant, full enteral feeding, small bowel transplantation) at regular 6 monthly follow up. Out of a total of 707 registrants, this study reviewed outcomes in 422 who had at least 2 yr follow up data. The following patients were excluded from the analysis for the reasons stated:

- 158: <2 years duration of PN – (95.5% alive)
- 41: incomplete follow up
- 86: IF not directly related to primary gastrointestinal disease (e.g. cardiac, oncology, preterm NICU patients, etc.)

Results

Table 1 BIFS registrant diagnosis and status 2yr post initiation of PN, n = 422

<table>
<thead>
<tr>
<th>Diagnostic group</th>
<th>Sub-classification</th>
<th>Total</th>
<th>Full Enteral food</th>
<th>On PN†</th>
<th>RIP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Short Bowel Syndrome (n=328)</td>
<td>Gastrochisis</td>
<td>109</td>
<td>90</td>
<td>16</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Necrotising Enterocolitis</td>
<td>99</td>
<td>86</td>
<td>9</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>Other cause SBS</td>
<td>120</td>
<td>86</td>
<td>28</td>
<td>6</td>
</tr>
<tr>
<td>Disorder of Motility (n=54)</td>
<td>Chronic Intestinal Pseudo-Obstruction</td>
<td>16</td>
<td>11</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Long Segment Hirschsprung’s</td>
<td>16</td>
<td>10</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Other cause dysmotility</td>
<td>22</td>
<td>15</td>
<td>6</td>
<td>1</td>
</tr>
<tr>
<td>Enteropathy (n=40)</td>
<td>Auto-immune Enteropathy</td>
<td>8</td>
<td>6</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Microvillus Inclusion Disease</td>
<td>8</td>
<td>3</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Other cause enteropathy</td>
<td>24</td>
<td>16</td>
<td>8</td>
<td>0</td>
</tr>
<tr>
<td>Grand Total</td>
<td>422</td>
<td>323</td>
<td>84</td>
<td>15</td>
<td></td>
</tr>
</tbody>
</table>

*10 patients had undergone intestinal transplantation 138/84 were on full enteral feeds at 2yr but re-started PN at a later date.

Of the 407 patients alive at two years post enrolment in BIFS, 77% had achieved enteral autonomy and 20% remained PN dependant or reverted to need for PN after a period of attempted weaning. 35% of patients with enteropathy, 31% of those with dysmotility and 16% of SBS patients remained PN dependant after two years.

Conclusion

These data indicates that outcome of IF is good, with 4/5 of patients establishing enteral autonomy overall, including the majority of patients with dysmotility disorders and enteropathy. Some patients need long term PN, both in hospital and following discharge home; a small number develop life threatening complications and undergo small bowel transplantation. The BIFS demonstrates a successful model of cooperation between different UK centres in establishing outcomes for a group requiring both significant resources and multidisciplinary care. BIFS has provided a firm foundation for the development of ‘Paed elBans’ which has the potential to lead to further improvement in data collection at national level.
Introduction

HPN is often used in children to correct malnutrition or to sustain appropriate growth. Currently there is very limited data available regarding the outcome of children on HPN, as stated in the NCEPOD report. The current perception is that HPN is associated with high mortality and high rates of small bowel/liver transplantation.

Aim

To answer Q30 from the NCEPOD audit tool “What was the eventual outcome for this patient?” including their growth outcome, for a cohort of children on HPN.

Results

All of the children requiring HPN in the northern region of England from 1st April 2008 until January 2013 were reviewed. In total there were 27 patients reviewed (17 on HPN < 1 year and 10 on HPN >1 year). Data was obtained by looking through clinic letters, prescriptions, and spread sheets collated by the nutrition team. Heights, weights and BMIs were all converted into z scores using the Children’s Growth Foundation software 2 months - 18 years old.

The outcome for children on HPN was good. 44% of the cohort are now fully enterally fed and using the Children’s Growth Foundation software 2 months - 18 years old.

The outcome of HPN is good. None of our children have chronic liver disease. Growth outcomes for children requiring HPN for <1 year are also good. Growth outcomes for children requiring HPN for longer than 1 year are less good. This could be due to compromises of nutritional intake compatible with family life, or we speculate may be due to alterations in gut hormone and other endocrine growth factors caused by overnight HPN.

Conclusion

Overall the outcome of HPN is good. None of our children have chronic liver disease. Growth outcomes of children requiring HPN for <1 year are also good. Growth outcomes for children requiring HPN for longer than 1 year are less good. This could be due to compromises of nutritional intake compatible with family life, or we speculate may be due to alterations in gut hormone and other endocrine growth factors caused by overnight HPN.

Fish-oil based intravenous lipid emulsion as a rescue in septic infants with intestinal failure and with or at risk of developing liver disease

Huoy Min Lai1, Ann Hickley2, Helen Callaby1, Maria O’Meara2, Lucy Thompson1, Jonathan Hind1; 1Paediatric Hepatology, 2Paediatrics, 3Pharmacy, King’s College Hospital, London, United Kingdom

Introduction/Background:

In infants with intestinal failure, it is known that episodes of sepsis can be accompanied by a significant deterioration in liver function. We hypothesised that an intravenous lipid emulsion (ILE) comprised solely of fish oil, such as Omegaven®, may protect the liver in these infants during episodes of sepsis.

Aim:

To describe our single centre experience with Omegaven® as a rescue therapy in septic infants with intestinal failure and with or at risk of developing liver disease.

Subjects and Methods:

A mixed source ILE containing both omega-3 and omega-6 fatty acids (SMOFlipid®) was used as first-line in infants at high risk of IFALD or severe liver disease. When these infants developed sepsis, Omegaven® was used as the sole ILE for up to 14 days. A retrospective review of their case notes was conducted.

Results:

Omegaven® was well tolerated in all infants. 10 infants had Omegaven® treatment during a 2-year period (August 2011-August 2013). Median birth weight was 965g (range 525–1960). Median gestation at birth was 28 weeks+5 days (range 24-34). Of the 10 patients, 2 had gastrochisis, 5 had necrotising enterocolitis (NEC), 2 patients had congenital infection with conjugated hyperbilirubininaemia, and 1 had conjugated jaundice associated with maternal liver failure. 1 patient with gastrochisis developed NEC. 2 patients were late transfers at 4-5 months of age from other hospitals with severe and progressive IFALD. Both subsequently died. Median age at start of Omegaven® was 38 days (range 2-189). 5 patients did not complete the full 2-week course of Omegaven®. 1 patient had 3 courses of Omegaven treatment due to recurrent episodes of sepsis. 9 patients had conjugated hyperbilirubininaemia with total bilirubin levels above 80µmol/l at commencement of Omegaven®. Out of these 9 patients, 5 had conjugated bilirubin levels above 100µmol/l when Omegaven was started. In the other patients, clinical assessment of the severity of sepsis led to the decision to commence Omegaven prophylactically to ameliorate potential liver damage. During their episodes of sepsis, bilirubin and CRP rose as expected in all patients. Transaminases were deranged in all. During the episodes of sepsis, those with established liver disease did not demonstrate the marked rise in bilirubin that would normally be anticipated. In those that were treated prophylactically, the liver function did not show marked deterioration despite the severity of sepsis. 7 patients showed improvement in bilirubin levels during treatment and this was maintained in the long term in 6. 1 patient’s total bilirubin levels improved initially but crept up again 10 weeks post Omegaven treatment. He died 6 months later due to E.coli sepsis. 1 patient was transferred to another centre for further medical treatment early in her Omegaven® course: her bilirubin was static.

Summary and Conclusion:

Use of Omegaven® as a short term rescue ILE in septic infants with intestinal failure and with or at risk of developing liver disease appears safe. The expected deterioration in liver function associated with sepsis was not seen in this series.
Five Year Experience of a joint multidisciplinary intestinal Failure and small Bowel Transplantation Assessment Centre – the Great Ormond Street and King’s College Hospital Referral Model

Eirini Serena1; Jain Vandana2; Hind Jonathan1; Heaton Nigel1; Vilaca-Melendez Hector1; Hill Susan1; Lindley Keith1; Köglmeier Jutta1; 1King’s College Hospital London; 2Great Ormond Street Hospital London

Background
In the last two decades the incidence of intestinal failure (IF) has constantly increased due to improved survival of extreme prematurity and raising incidence of gastrochisis. Improved management of structural, inflammatory and motility disorders has led to the need for long term parenteral nutrition (PN) in a larger number of children. At present around 150 British children are managed on home PN.

A new multidisciplinary intestinal failure assessment (IFA) and small bowel transplantation (SBT) centre was hence nationally commissioned in the United Kingdom. Clinical services started in April 2008. Patients are referred to Great Ormond Street Hospital for assessment of their intestinal function and sent to King’s College Hospital if criteria for SBT are fulfilled.

Aim
To report the experience of a new intestinal rehabilitation centre since the beginning of the program in 2008.

Methods
Retrospective case note review of all patients referred for IFA and SBT.

Results
A total of 48 children were referred for IFA of which 26/48 underwent assessment for potential SBT. 8/48 successfully weaned off PN with improved clinical management. 12/46 did not fulfill the criteria for SBT and remain on PN. 2/48 families declined the SBT offer. 10/48 underwent SBT. 1 child died post SBT due to severe graft rejection. 9/10 transplanted children are alive and well. 1/48 children was recommended for isolated liver transplant only due to good intestinal adaptation but irreversible liver disease. 4/48 are currently on the active transplant waiting list. No patient died on the waiting list.

Conclusion
Improved medical management by a highly specialized centre can prevent the need for small bowel transplantation in a significant number of patients.

Short term survival post SBT in this new centre is promising with 90 % of patients being alive and well following surgery.

Evaluation of Sedation versus General Anaesthesia for endoscopy in children and adolescents

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Backgrounds/Aim:
Debate continues about whether mild-moderate sedation or general anaesthesia is the ideal method to perform GI endoscopic procedures in children and adolescents. The study aim was to compare and evaluate intravenous sedation to general anaesthesia.

Materials and methods:
Retrospective and prospective studies were conducted to compare paediatric patients who underwent GI endoscopy under IV sedation or GA in a single centre. A patient satisfaction questionnaire was designed and submitted. The study was conducted in the gastrointestinal endoscopy unit at The Royal London Hospital in the UK.

Results:
329 patients (55 patients IV sedation and 274 patients in the GA group) were reviewed retrospectively. There were no significant differences in the two groups in the completion rate, complications or in the duration of gastroscopy. The duration of colonoscopy in the IV sedation group was significantly longer than in the GA group (p=0.001), this was possibly explained by the fact that a higher proportion of trainees started the procedures using IV sedation than GA. Forty five (8 IV and 37 GA) patients underwent detailed time analysis prospectively when no significant differences in the duration of gastroscopy or colonoscopy or whole procedure were seen. The duration of recovery in the IV sedation group was significantly shorter than that in the GA group (p= 0.005). When comparing a single endoscopist the time to complete procedures did not vary between GA and sedation.

16 patients and parents completed the patients satisfaction questionnaire with high levels of satisfaction about the endoscopic services. The major cost difference between the two procedures was the need for additional staff (1 consultant anaesthetist and one ODA) in the GA group.

Conclusion:
Intravenous sedation (midazolam+/-pethidine) can be used safely and effectively for paediatric GI endoscopy in appropriately selected patients. The retrospective study suggested that the duration of the colonoscopy was longer in the IV sedation group compared to the GA group, however this was not the case in the prospective study. GA costs more and in this study did not show any potential benefit in terms of increased numbers or shorter time in the endoscopy unit.
Paediatric Endoscopy Global Rating Score: Pilot data from 2 tertiary gastrointestinal centres

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Background:
The Joint Advisory Group (JAG) on gastrointestinal endoscopy aims to set standards for UK wide endoscopy units to provide safe, high-quality, timely, patient-centred care. JAG certification is underpinned by the Global Rating Score (GRS). The GRS was created in 2004 as a quality improvement and assessment tool for gastrointestinal endoscopy service. The endoscopy working group of BSPGAN recognises that paediatric endoscopy is lagging behind adult practice with regards to standardisation of endoscopy service. For this reason, the GRS has now been adapted to form a paediatric GRS that can be used to obtain a greater understanding of the variation of paediatric endoscopy practice nationally.

Aim:
To pilot the newly adapted paediatric GRS to assess the endoscopy services for children and young people in two tertiary paediatric gastroenterology centres.

Methods:
Two tertiary paediatric gastrointestinal units (Birmingham Children’s Hospital and The Royal London Hospital) piloted the newly adapted JAG-approved paediatric GRS. The scale comprises of four domains: 1) Clinical Quality; 2) Quality of Patient Experience; 3) Workforce and 4) Training. Levels of achievement (D-A) were used, where level D was basic and level A an excellent service. Level B or better is the current standard for an acceptable service.

Results:

<table>
<thead>
<tr>
<th>Area</th>
<th>Clinical Quality</th>
<th>Quality of Patient Experience</th>
<th>Workforce</th>
<th>Training</th>
</tr>
</thead>
<tbody>
<tr>
<td>Area</td>
<td>Consent</td>
<td>Safety</td>
<td>Comfort</td>
<td>Quality</td>
</tr>
<tr>
<td>Score</td>
<td>B&lt; D</td>
<td>RLH &lt; D</td>
<td>B&lt; D</td>
<td>D</td>
</tr>
<tr>
<td>Score</td>
<td>B&lt; C</td>
<td>RLH &lt; D</td>
<td>B&lt; D</td>
<td>A</td>
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<tr>
<td>Score</td>
<td>B&lt; D</td>
<td>RLH &lt; D</td>
<td>B&lt; D</td>
<td>D</td>
</tr>
<tr>
<td>Score</td>
<td>B&lt; D</td>
<td>RLH &lt; D</td>
<td>B&lt; D</td>
<td>B</td>
</tr>
</tbody>
</table>

Summary:
The results are almost identical across the four domains aside from workforce where Birmingham Children’s Hospital scored higher than The Royal London. As it stands, both centres would not attain JAG certification. These results are subject to bias but demonstrate the inadequacies of the current provision of paediatric endoscopy as compared to the standards set by JAG.

Conclusion:
These findings are similar to adult endoscopy centres where the majority of units do not pass on the first assessment where changes in practice can often be easily made to meet GRS approval. It was evident from our pilot study that many of our scores of D’s and C’s could achieve B status by implementing minor changes. Overall, a paediatric GRS is useful to identify areas of weakness that can be improved to raise the standards of patient care in endoscopy.

Title of Abstract: Evaluation of neonatal cholestasis in infants born pre-term

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Background:
Investigation of neonatal cholestasis in pre-term infants can be challenging. Although most often contributed to by prematurity, parentral nutrition (PN) and sepsis, other causes requiring urgent and specific management must be identified, including biliary atresia, transient or persistent cholestasis or transfer may be precluded by clinical condition and size. In 2010 we devised a structured proforma to record a summary of infants discussed by telephone with our Supra-Regional Referral Unit, in order to formalise documentation of history and advice given and to ensure need for transfer was carefully considered.

Aim:
(1) To evaluate a structured proforma by determining frequency of admission and final diagnosis. (2) To describe diagnostic evaluation of pre-term infants admitted and the characteristics of those with BA.

Subjects and methods:
All infants with neonatal cholestasis referred between 01/01/2010 and 31/03/2013 whose course was documented by proforma and/or were admitted.

Results:
Of 287 infants (182M:105F), 129 were preterm (85M:44F) with median gestation at birth 29 weeks (23-36) and birth weight 1.00 kg (0.52-3.7). Chronological age at referral was median 48 days (7-156). Excluding 15 referred from our own Neonatal Unit, decision to admit was made in 41/114 (36%) pre-term infants compared to 114/158 (72%) term infants (p<0.0001). Pre-term infants who were admitted were more likely to have no history of PN (66% v 8% p<0.001) and pale stools reported (61% v 14% p<0.001) than those not admitted. BA was diagnosed in 8/129 (6%) preterm infants referred compared to 41/158 (26%) term infants (p<0.0001). Those with BA were born at 27-36 (median 32) weeks gestation, referred at median age 32 (9-84) days and admitted after a median of 1 (0-13) days. One, who was ventilator dependent, died aged 9 months with chronic lung disease. Ultrasound scan (US) showed features of BA, but surgery was not attempted. In all of the remaining 7 US showed abnormal irregular gall bladder, triangular cord sign, and no identifiable CBD. Three also had US features of BASM syndrome. In all 7 BA was confirmed in the operating room. Donohoe syndrome (1) PFIC (1) congenital portosystemic shunt (2) and portal cavernoma (2).

Of 73 admitted who did not have BA, duration of admission was median 2 days (0-8). All underwent US, which in 32 was not suggestive of BA. One with US features of BA and non-excreting HIDA underwent liver biopsy which excluded BA. One with poorly distended GB on US but no other features of BA, and non-excreting HIDA, underwent cholangiogram to exclude BA. Sensitivity and specificity of US for BA in preterm infants were 100% and 97%. In addition to imaging by US, 12/20 with no previous HIDA underwent HIDA. Two infants in whom BA was not suspected underwent biopsy because of hepatosplenomegaly. US was the only investigation in 17 infants who were admitted for median 1 day (0-8). Overall, 16 had identifiable CBD: Cystic fibrosis (3) endocrine abnormality (5) CMV infection (2) alpha-1-antitrypsin deficiency (A-1-AT) (1) Donohoe syndrome (1) PFIC (1) congenital portosystemic shunt (2) and portal cavernoma (2).

Of 73 not admitted, none were subsequently diagnosed with BA. We are aware of additional diagnoses in 9: endocrine abnormalities (6) CMV infection (2) and A-1-AT (1). 44/45 with follow up information available had resolution or improvement of cholestasis. In one transfer was planned but precluded by clinical deterioration. Two infants were later seen as outpatients, one had resolved cholestasis, the other A-1-AT.

Conclusion:
Identification and timely transfer of pre-term babies with BA was facilitated by a structured proforma, which also minimised transfer of those without BA. Pre-term infants with final diagnosis of BA were referred earlier than those without BA (median 32 days v 50 days), were admitted promptly and had a good outcome following Kasai portoenterostomy. In all 7 BA was confirmed in US in pre-term infants for assessment of BA has good sensitivity and specificity, and facilitates short admission and the avoidance of more invasive procedures.
Growing up with biliary atresia without liver transplantation; a single centre experience.

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Background:
Biliary atresia is a unique condition to infancy and is the most frequent indication for liver transplantation (LT) in the paediatric age group.

Aims:
To evaluate outcomes in adulthood of biliary atresia not requiring LT in the paediatric age group. A single centre retrospective study was performed, comprising 268 patients who underwent Kasai Portoenterostomy (KP) between 1980-96.

Results:
After KP, 107/268 (40%) survived with their native liver for at least 16 years. Ninety-seven (43M:54F) of these patients are followed up at our centre; providing our study group; ten are being followed up at other adult liver centres. Of the remaining (n=161); 76% underwent LT<16yrs, 11% died and 13% were lost to follow up.

At a median age of 20.6yrs (range;16-32) at the last follow-up, 81 (84%) remain with their native liver (Group 1) whilst 16 underwent LT or are currently listed (Group 2).

In Group 1; 55 patients have normal SBR (<20umol/l) levels. Within this subgroup, patients with normal parameters are as follows; AST (<50 IU/l), 84%, platelet count (>100), 84%, synthetic function (albumin >35mg/dl and INR <1.2), 96% and no splenomegaly on USS, 44%. All parameters are normal in 21 patients. Within the subgroup of patients with abnormal SBR (>20umol/l) levels, only 1 had all normal parameters. Overall in Group 1, 7% of patients had at least 1 episode of cholangitis and 9% had documented varices. One patient died during pregnancy.

In Group 2, 14 patients underwent LT and 2 are currently listed for LT. Documentation for 15 patients was available. Median age at LT was 18.8yrs (range;16.5-27.1) after median waiting time for LT of 11.5mths (range;2-63). Median SBR and albumin at time of listing were 137umol/l and 32mg/dl respectively. Indications for LT were recurrent cholangitis with synthetic failure (n=7), synthetic failure (n=4), cholangitis (n=2), hepatocellular carcinoma (n=1) and portal hypertension (n=1). Two patients required re-transplantation for chronic rejection, of which one died following CMV infection.

Overall, four patients had 6 successful pregnancies of whom 1 is currently listed for LT and 1 is pregnant. One patient (SBR 29umol/l, portal hypertension) died during pregnancy from variceal bleeding.

Conclusions:
In our patient cohort, 84% are alive with their native liver, with 69% of patients demonstrating normal SBR levels and 26% showing no evidence of progressive liver disease. The most common indications for LT in adulthood, are recurrent cholangitis and/or synthetic dysfunction.
G2

A study of the impact of implementing the new coeliac guidelines in a single centre
Dr Anne Willmott, Consultant Paediatric Gastroenterologist, University Hospitals Leicester NHS Trust (UHL), Dr Suchandra Pande Consultant Paediatrician (UHL), Kristian Bravin, Paediatric Dietitian (UHL)

Introduction/Background
The diagnosis of coeliac disease has changed over the years from one requiring several biopsies to one needing only blood testing and a single biopsy. The vastly improved accuracy of the blood tests for this condition has led in 2012 to ESPGHAN and BSPGHAN agreeing new guidelines for the diagnosis of coeliac disease. In many cases this may avoid a biopsy altogether.

Aim
After discussing the new guidelines we decided to adopt them in Leicester for the calendar year 2013. The aim of this study was to look how practical the new guidelines were, to look at the impact of this decision on endoscopy rates, and to audit our practice against the new standards.

Subjects and methods
All new diagnoses of coeliac disease in children under 16 in our centre in the calendar year 2013 were analysed. These were recorded prospectively as they were referred. As well as this each month all raised TTGs in children, processed by our immunology department were reviewed, to check that there was a record of all possible new patients, and this was also cross checked with the dietetic coeliac database.

We recorded the blood tests done, the results and the speed of access to a definitive diagnosis (HLA result and repeat TTG, or biopsy result). We noted whether following the new pathway avoided endoscopy, and the cost implications involved in this.

Results
In 2013 34 patients were diagnosed with coeliac disease (to end Oct – updated figure will be included when available). Of these only 8 (24%) required endoscopy to confirm the diagnosis. After the initial screening test, diagnosis per patient with biopsy costs commissioners around £1400, (derived from tariff + top up) whereas diagnosis per patient without biopsy costs £130, thus there was a saving of approximately £36,000 in this year. It was hard to get accurate data regarding cost to the trust of an endoscopy as our PLICS data were clearly inaccurate.

The HLA result did not change the diagnosis in any of our patients, as it was positive in all patients with significantly raised TTG.

In reviewing our practice we followed the new pathway correctly in 31/34 (91%) of pts.

Three patients were wrongly started on gluten free diet after only one screening test. Two of these by a general paediatric colleague (identical twins), one in the private sector. All 3 had a repeat TTG test and HLA done within a few weeks of starting gluten free diet, after which coeliac disease was confirmed.

We are using this data to put a business case together to try to fund a new specialist nurse post, funded by the savings to commissioners of the change in practice.

Summary and Conclusions
Following the new pathway for diagnosis of coeliac was straightforward, providing a timely diagnosis, and was correctly followed in 91% of cases. We avoided 26 endoscopies, saving £36,000 to commissioners, although the inaccurate PLICS data did not allow us to confirm cost implications to the trust of the reduction in endoscopy. The HLA result did not change the diagnosis in any patients avoiding a biopsy, as it was positive in 100% of these patients, and we would suggest it’s use in the context of TTG high enough to avoid biopsy needs to continue to be reviewed. Only three patients with raised TTG were not referred immediately to paediatric gastroenterology. All were discussed with the physicians concerned. We would recommend the new pathway, with continuing education of colleagues and cross checking with immunology extra safety measures to ensure all patients are correctly referred to gastroenterology.
Coeliac Disease in Children – Screening, Assessment and Management
Dr Sarah Longwell, Dr Tasneem Khan, Dewsbury and District Hospital, Halifax Road Dewsbury, WF13 4HS. Sponsor: Dr Sherman Soman

Background
- The BSPGHAN 2012 guideline for the diagnosis and management of coeliac disease in children outlines the use of screening using quantitative TTG analysis and HLA subtyping, and also makes recommendations for post-diagnosis management.
- For the purpose of this audit we used the NICE clinical guideline 86 and the BSPGHAN 2012 guideline, to assess the diagnosis and management of children with coeliac disease presenting to the three DGHs at the Mid-Yorkshire Hospitals NHS trust.
- We have also evaluated the possible introduction of quantitative TTG analysis and HLA subtyping at the trust in the diagnosis of coeliac disease.

Methods
1. Retrospective case note analysis of a sample of 50 patients taken from the cohort of paediatric coeliac disease patients in the Mid-Yorkshire Trust.
2. Questionnaire given to 16 parents during attendance at a follow-up coeliac clinic in October 2013.

Results
In our audit 72% of our patients were female and 28% male. 52% made their initial presentation to paediatric department and 38% presented to their GP. 86% of our patients were asymptomatic on presentation, of which the most common symptoms were diarrhoea, faecal urgency, abdominal pain and lethargy. Out of our 4 asymptomatic patients, 3 were found on screening for affected first-degree relative and 1 had type 1 diabetes mellitus. 80% were officially diagnosed by biopsy and 18% on serology.

Post-diagnosis monitoring
- The Mid-Yorkshire trust had best results with clinician and dietician reviews, symptom monitoring and addressing any compliance issues.
- The areas where the trust still needs to improve are sibling screening, pneumococcal vaccination, school support and oats discussion for selected patients.

Questionnaire
- 15 out of the 16 patients had biopsies to confirm their diagnosis of coeliac disease.
- 15 out of 16 parents would have preferred HLA testing if it meant possibly avoiding biopsy.

Cost analysis – to move to numerical TTG reporting will incur no additional cost to immunology laboratories.
- Cost of biopsy = £858:
- Cost of HLA testing = £30

Conclusion
The Mid-Yorkshire Trust has scope for improvement to meet the updated standards introduced by the BSPGHAN guidelines in terms of screening siblings, discussing oats are pneumococcal vaccination. We have shown that HLA-typing can provide significant cost-reduction, and is a preferable option for parents.

Our recommendations are for the Mid-Yorkshire Immunology labs to report TTG titres in order for screening via HLA-typing to be commenced.

We also recommend a proforma for use by clinicians as a prompt for post-diagnosis management in order for universal implementation of the guidelines. We recommend a re-audit once HLA-typing has been established and the proforma becomes available for use in practice.
G6

Immunological and histopathological changes associated with symptoms of an “inflammatory bowel disease – like” disease in a cohort of children with phenotypic diarrhoea

Whyte LA1; Brundler MA2; Hackett SJ2; Heart of England NHS Trust, Birmingham; Hartley J3; Protheroe SJ1; Birmingham Children’s Hospital, Birmingham; Heart of England NHS Trust, Birmingham

Background

Phenotypic diarrhoea (PD) or tricho-hepato-enteric syndrome (THES) is a rare congenital enterocyte defect with immune deficiency that usually presents with IUGR, facial and hair changes and protracted diarrhoea of infancy, that requires a period of parenteral nutrition (PN). In infancy, secretory diarrhoea appears within the first few months of life and is non-responsive to medical therapy (including immunosuppression). There is no characteristic histological pathology (e.g. enteropathy) to explain the diarrhoea. Later, the children develop skin changes, including café-au-lait macules. As our cohort grows older it is becoming apparent that they develop upper GI ulceration and inflammation, manifesting as problems of vomiting and haematemesis which limits their oral intake. Inflammatory changes have been noted in gastro-oesophageal, gastro-duodenal and ileo-caecal junctions, areas that are typically affected by Crohn’s disease and chronic granulomatous disease, but the histology has subtle differences. The gene defects identified include TTC37, a gene that was identified in our own centre and SKIV1, from a cohort in France.

Aims

1) To characterise the evolving clinical course, immunological deficiencies and associated congenital heart diseases of our cohort of patients with PD
2) To characterise the clinical and histological changes associated with the development of an inflammatory gastrointestinal pathology in older children with PD
3) To describe the treatment strategies and outcomes
4) To relate specific genetic defects to clinical phenotype
5) Discuss the potential causes of diarrhoea

Methods

We identified all patients cared for in our tertiary gastrointestinal centre who have a confirm diagnosis of PD and retrospectively reviewed histopathological, immunological, genotype and response to immunological therapy.

Results

We identified 9 patients with PD who were diagnosed between 2004 and 2013 in our unit. All the patients were still alive and all have short stature. Current age range is 18 months to 20 years. All received PN from infancy (range 1m-7 years of age). 3 have weaned off PN and are fully enterally fed. 5 have new onset symptoms that are PD-like in early childhood (vomiting, haematemesis, worsening diarrhoea), age of onset of symptoms range from 1 year to 7 years. Histological examination reveals similar inflammatory features which compromise a correlation in features, but inflammation that is distinct from inflammation seen in PD. 3 have received steroids with good effect. 3 are taking azathioprine and 2 have required IV immunoglobulins due to low IgG and bronchiectasis. All patients had delayed response to primary immunisations, but have normal overall T cell numbers and function; we therefore hypothesise a problem with T cell memory function. 4 have suffered with recurrent URTI's and have required tonsillectomy before the age of 7 years. 3 patients had congenital heart defects and 3 presented with chronic liver disease.

Conclusion

This is the first description of the phenotype-genotype in a large cohort of patients with phenotypic diarrhoea and PD-like symptoms in childhood. It would appear in our patients that the genotype does not correlate with the immunological function/clini phenotype. The enteropathy in infancy is not seen universally, and is not associated with a histopathological anomaly. We therefore hypothesise that the high stool sodium and chloride diarrhoea may be due to an unrecognised transport problem. The inflammatory bowel –like changes are likely to be due to immune dysregulation which has yet to be fully described.
Impact of socio-economic position on incidence of Inflammatory Bowel Disease

Mr Matthew Cole; Dr Jessica Whitburn; Dr Christine Spray; Dr Dharamveer Basude; Ms Sarah Sandmann; Dr Siba Prosad Paul; Dr Pramila Ramani; Prof Bhupinder Sandhu; Bristol Royal Hospital for Children, Bristol

Background:
Inflammatory bowel disease (IBD) is understood to result from the interaction of genetic, immunological and environmental factors. There has been a marked increase in the incidence of IBD over the last 25 years, suggesting environmental factors are important. A previous study found a higher incidence of Coeliac disease in the least deprived socioeconomic groups. The objective of this study was to investigate the relationship between IBD and socioeconomic position.

Methodology:
Bristol Children’s Hospital is the single regional centre where all children with suspected IBD from the South-west of England are referred. Data was collected prospectively on all children diagnosed between May 2004-March 2013. Socioeconomic status was determined by quintile rank of Index of multiple deprivation score (IMD-10 score) based on postcode at diagnosis. This has been shown to provide a nationally consistent measure of how deprived an area is. Population data was obtained from the 2011 Census. Data was analysed using Pearson Chi Squared test. Children with a postcode outside of the City of Bristol were excluded from the analysis.

Results:
Aged 0-17 years were diagnosed with IBD over the study period of which 50 had a postcode of residence within the City of Bristol. The incidence of IBS was higher in the three lower socio-economic classes compared to the two highest socio-economic classes. However, the difference in incidence between the socio-economic classes was not statistically significant.

<table>
<thead>
<tr>
<th>Socioeconomic Class</th>
<th>Number of IBD Patients</th>
<th>Exposed population</th>
<th>Cumulative incidence per 100,000</th>
<th>Incidence per 100,000 per year</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Most deprived</td>
<td>18</td>
<td>23374</td>
<td>77.01</td>
<td>8.72</td>
</tr>
<tr>
<td>2. Below average</td>
<td>15</td>
<td>22458</td>
<td>66.79</td>
<td>7.56</td>
</tr>
<tr>
<td>3. Average</td>
<td>9</td>
<td>10372</td>
<td>86.77</td>
<td>9.82</td>
</tr>
<tr>
<td>4. Above Average</td>
<td>5</td>
<td>13642</td>
<td>36.65</td>
<td>4.15</td>
</tr>
<tr>
<td>5. Least deprived</td>
<td>3</td>
<td>7202</td>
<td>41.66</td>
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<tr>
<td>Total</td>
<td>50</td>
<td>77048</td>
<td>64.89</td>
<td>7.35</td>
</tr>
</tbody>
</table>

p =0.204

Conclusion:
Our data suggests a higher incidence of diagnosed IBD in children from lower socioeconomic classes which may favour an environmental aetiology. However this did not reach statistical significance, possibly due to small numbers. A larger study is warranted.

References:

Improving time to diagnosis in children referred locally with suspected inflammatory bowel disease within a managed clinical network - a single centre, secondary care experience.

Victoria Rodulson (Medical Student, PCMD), Christopher Harbinson (Medical Student, PCMD), Dr Matthew Thorpe (Consultant Paediatrician, RCHT), Dr Christine Spray (Consultant in Paediatric Gastroenterology, BRHC), Mrs Sarah Sandman (Nurse Specialist, BRHC)

Introduction
All children with suspected inflammatory bowel disease (IBD) should undergo endoscopy to confirm diagnosis, requiring referral from secondary/satellite unit (DGH) to a tertiary paediatric unit, within a Managed Clinical Network (MCN). Delay in diagnosis can occur at three stages: 1) Referral from primary care to DGH, 2) from the time of secondary review to referring onwards 3) from referral to tertiary care to securing tissue diagnosis. Proposed national service specifications for paediatric gastroenterology suggest maintaining a Special Interest (SPIN) paediatrician within the DGH, acting as a link between the two services to streamline patient care. Children presenting to the Royal Cornwall Hospital (RCH) in Truro are referred to the Bristol Royal Hospital for Children (BRHC). Good working relationships between RCH and BRHC mean that the majority of referrals for suspected IBD go “straight to endoscopy”. Attention to effective triage of GP referrals via our Referral Management Service has also reduced time to initial review. A review of the literature did not find previously set standards against which to compare this data. Previous research relating to delay in diagnosis focused on delay between onset of symptoms to GP review as well as the delay between GP review to final diagnosis. Reducing this initial delay involves longer-term initiatives such as parent and GP education.

Aim
To establish time (in days) taken for diagnosis from initial referral, most commonly GP, in children with confirmed IBD following endoscopy within the CMN by analysing local data and to identify areas for improvement between 2010 and 2012.

Subjects and Methods
Sixteen patients diagnosed with IBD between 2010 and 2012 were identified from the DGH clinic managed by SPIN Consultant. Case notes were reviewed retrospectively to establish time in days from initial referral to date first seen by local paediatrician, date first seen by paediatrician to tertiary referral, and time in days from tertiary referral to endoscopy. Total time from initial referral to endoscopy was derived. Patients were then grouped by year for further analysis. GP referral times were compared with average referral times for first appointment.

Results
The graph below (figure 1) shows the total time to diagnosis over the 2010-12 time period with the time to diagnosis subdivided to demonstrate where the longest waits occur.

Figure 1 – Time to referral (days) shown on y axis and subdivided to show relevant waiting times 2010-12
Summary

Over the 3 year period, through excellent working relationships between RCH and BRCH the total time to endoscopy and therefore diagnosis was reduced by more than 50%, from 99 days to 41 days. In addition, systematic review of GP referrals meant average time to first review was fifteen days compared with fifty-six days for the remainder of general paediatric outpatients. Local review by a “SPIN” doctor reduced the time to tertiary referral. Individual cases with longer referral times were reviewed to look for avoidable factors. Referral pathways between surgeons and general paediatricians to “SPIN” paediatrician were refined.

Conclusion

This study has highlighted the need to review such data, in doing so demonstrating areas of good practice as well as those requiring improvements. With this in mind, and in the absence of relevant benchmarks, we propose other Managed Clinical Networks collect this data, so that analysis and service improvement may occur on a national scale.
Prevalence of Paediatric-onset Inflammatory Bowel Disease: A Systematic Review

1F Cameron, 2P Henderson, 1DC Wilson. 1Child Life and Health, University of Edinburgh, Scotland, UK and 2Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh, Scotland, UK

Introduction:
Previous systematic reviews have shown that the incidence of both adult-onset inflammatory bowel disease (IBD) and paediatric-onset IBD (PIBD) are increasing worldwide, particularly in developed countries, but little is known about PIBD prevalence. Although trends in incidence are important, the determination of prevalence is crucial to inform effective service provision and delivery and may provide clues to the aetopathogenesis of IBD.

Aims:
To assess the available evidence on worldwide prevalence of PIBD, assess geographic distribution and analyse trends using systematic review.

Methods:
A literature search was performed using MEDLINE (1950-2012), Medline in progress, Cochrane database and EMBASE (1980-2012) to identify relevant population-based studies. Studies were included that reported distinct paediatric prevalence data on total Inflammatory Bowel Disease (IBD), Crohn's disease (CD), Ulcerative colitis (UC), Inflammatory Bowel Disease Unclassified (IBDU) or any combination of these. Abstracts in all languages were considered but papers were only included in which full text was available for review. Data was extracted on source of prevalence data, age ranges, country of origin, diagnostic criteria and methodology for each eligible study.

Results:
4190 references were found and reviewed; 27 studies which presented data on the prevalence of PIBD and/or CD and/or UC from 11 countries were included. No prevalence data were presented on IBDU. The prevalence of PIBD ranged from 6.0-30.0 per 100,000 while CD ranged from 0.5 -85.3 per 100,000 and UC from 3.0-90.1 per 100,000. Only two studies provided trend analysis over time, showing an increasing PIBD prevalence during a 5 and 10-year period respectively. There was a preponderance of reports from developed countries: 8 from North America, 10 from Europe, 5 from the Middle East, 2 from Asia, 1 from Africa, and 1 from Australasia. Prevalence rates were highest in North America and lowest in Asia and Africa. Most studies (74%) were retrospective, used a variety of sources including national registries, insurance databases, retrospective case note review and physician survey. No true population-based studies were reported. Age ranges and diagnostic criteria used varied widely with reported age ranges from 0-16 combined to 0-19 years depending on the study, and with diagnostic testing varying from colonoscopy +/- radiological imaging +/- upper GI endoscopy.

Discussion:
Reported prevalence of PIBD was highest in North America and Europe with little data available from developing nations. There was insufficient data to analyse trends in prevalence of PIBD over time although it seems highly likely that, given the rising global incidence, a parallel rise in prevalence would be evident. Data reported from these studies were heterogeneous in terms of diagnosis of disease, method of case accrual and age ranges used making interpretation of prevalence challenging. Further work is needed in the form of well designed studies with clear diagnostic criteria and age ranges to confirm if the increase seen in the worldwide prevalence of adult IBD is mirrored in PIBD. The results of these studies can be used to enhance clinical service provision and drive future funding for research.
Background:

Coeliac disease (CD) is an immune mediated systemic disorder elicited by the ingestion of gluten and related prolamines in genetically susceptible individuals. Diagnosis has been based on small bowel histology as per ESPGHAN guidelines. In 2012 ESPGHAN guidelines were modified and recommended that in symptomatic patients a diagnosis of CD can be made without small-bowel biopsy if anti-tissue transglutaminase antibody (TTG) titre is greater than 10 times upper limit of normal (>10ULN), together with presence of HLA-DQ2 and/or DQ8. This study aimed to examine the relationship between TTG levels and the corresponding histological features.

Methods:

Data was collected prospectively at diagnosis of CD from 126 consecutive children between June’ 2011 – May’ 2012. TTG was measured using ELISA technique. Histological samples were obtained from endoscopic small-bowel biopsies and interpreted by paediatric histopathologists. The relationship between the modified Marsh criteria histological findings and contemporaneous TTG levels was analysed.

Results:

Out of 126 children, 13(10.5%) had positive TTG but no documented titres. In 121(98.3%) histological report did not specify Marsh classification. Complete data of histological report and TTG level were therefore available from 104 children (82%). The data (table) shows an association between TTG level and histological staging of CD. 58 (48%) children had TTG>10ULN (>100U/ml). 57/58 of these patients had biopsy proven CD. The sensitivity of the TTG level >100U/ml alone in correctly diagnosing CD in this cohort was 98.3%.

Modified Marsh Criteria identified on histology Mean TTG level (U/ml) (SD) (95% confidence level)

<table>
<thead>
<tr>
<th>Modified Marsh Criteria</th>
<th>3a</th>
<th>3b</th>
<th>3c</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean TTG level (U/ml)</td>
<td>95.9</td>
<td>110.0</td>
<td>167.4</td>
</tr>
<tr>
<td>(95% confidence level)</td>
<td>66.8-124.9</td>
<td>87.5-132.3</td>
<td>143.0-191.7</td>
</tr>
</tbody>
</table>

Conclusion:

98.3% of children with TTG>100U/ml had histologically confirmed CD although total villous atrophy was associated more often with TTG level of >200U/ml. It is essential to report TTG titres by all laboratories. This study supports the new ESPGHAN guidelines for the selective use of high TTG levels in diagnosing CD in symptomatic children without a biopsy.

References:


G13
The role of faecal calprotectin in paediatric non-IBD patients with gut pathology

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Aims:

The aims of this pilot study were to determine whether FC levels in non-IBD children with bowel symptoms correlate with significant histological and/or radiological abnormalities and to evaluate above what level FC can be used as a surrogate marker of gastro-intestinal disease severity.

Methods:

FC levels were assessed in children (0 to 16 years) with bowel symptoms (abdominal pain, diarrhoea) and/or faltering growth who presented to our institution over a 10-month period. Children with a diagnosis of IBD were excluded. A commercially available ELISA test, measuring calprotectin in mg/Kg of stool sample, was used. The included children had endoscopic and/or radiological assessment (MRI, ultrasound (US)) of the gastro-intestinal tract.

Results:

Eighteen children (9 male, median age of 5y 3m), had FC levels measured. Average FC was 756.4 mg/Kg. No children had FC equal to or less than 100 mg/Kg. FC was between 100 and 200 mg/Kg in 9 children with an average value of 141 mg/Kg (group 1). Nine children had FC levels equal to or above 200 mg/Kg (group 2), mean value of 1371.9 mg/Kg. In group 1, 7 children had histological assessment, 6 (85.7%) of which had no or mild pathology; those with mild mucosal changes had positive stool cultures for viruses. Six patients in group 1 had radiological investigations which did not reveal any significant gastro-intestinal abnormality. In group 2, 88.8 % had biopsies, all of which had mild or moderate mucosal abnormalities and negative stool cultures. Seven in group 2 had MRI and/or US; only 1 patient showed severe gastro-intestinal abnormalities on both tests.

Conclusions:

FC below 200 mg/Kg in non-IBD patients revealed no significant pathology; infectious causes must be excluded. A “wait and watch” approach to patient management would be advisable with a repeat FC. FC equal to or above 200 mg/Kg warrants further investigations with endoscopic evaluation and histology. US and MRI remain useful adjuvants, but do not increase the diagnostic yield. Further studies with larger number of patients will establish a higher upper limit of normal (ULN) value for calprotectin in the paediatric population.
Utility of Faecal Calprotectin in diagnosis of Inflammatory Bowel Disease (IBD): which patients require endoscopic evaluation?  

Nikaki K.¹, Rosie G.², Wilkes B. ², Butt A. ³ ¹ Paediatric Registrar, ² Foundation Year 1 Trainee, ³Consultant Paediatric Gastroenterologist, Royal Alexandra Children’s Hospital, Brighton, UK

Introduction:
Faecal Calprotectin (FC) is considered a non-invasive marker of GI inflammation, released from neutrophils during apoptosis/necrosis; it has a role in regulating inflammatory response but levels are not elevated in extra-digestive processes. In adults, role in differentiating IBD from non-IBD conditions is better established. In children, normal levels for age and an optimal cut-off point to guide decision for diagnostic endoscopy are not well defined.

Aim:
The aim of our study was to investigate the use of FC in our department, since it has become more routinely available, specifically to assess its value in accurately selecting patients for diagnostic endoscopy in IBD.

Subjects and methods
A retrospective review of laboratory requests and medical notes was undertaken for all FC test requests over a 12-month period (1st Jan 2012 - 31st Dec 2012). We identified 94 patients who had one, or more, requests for a FC test. Patient numbers excluded are; 27 (with known GI diagnosis); 5 (FC result unavailable) and 6 (notes unavailable). In total, data was analysed on 56 patients for monoclonal FC Ab EUSFA test with assessment of; biochemical inflammatory markers (Hb, WC, PLT, CRP, ESR, Alb), decision for endoscopy and final diagnosis, including, follow up period of 6-18 months until July 2013.

Results:
56 patients (pts) identified, 26 male; median age 12 y (range, 3 m – 17 y). FC values were categorised in 3 groups; (a) <50mcg/g, ‘normal’ in 18 pts, (b) 50-600 mcg/g, ‘intermediate’ in 25 pts and (c) >600 mcg/g, ‘highest’ in 13 pts. None of the 43 pts with ‘normal’ or ‘intermediate’ FC levels were diagnosed with IBD. 12/13 pts with ‘highest’ FC levels were diagnosed with IBD and 1/13 with post BMT gut CMV infection; all these pts were in the group of 21/56 pts who underwent diagnostic endoscopy based on standard clinical parameters. Additionally, 5/21 pts with ‘intermediate’ FC levels were diagnosed with, coeliac disease (1), rheumatological condition (1), gastroesophageal reflux disease (2) and irritable bowel syndrome (1) and 3/21 pts with ‘normal’ FC levels had other upper GI or normal pathology. ‘Intermediate’ levels of FC are seen in a range of non-IBD conditions e.g. IBS (25%), functional abdominal pain (7/25), GORD (1/25) and other (12/25). FC levels (>600 mcg/g) performed better as a marker to identify pts with IBD; all 12 pts and (c) >600 mcg/g, ‘highest’ in 13 pts. None of the 43 pts with ‘normal’ or ‘intermediate’ FC levels were diagnosed with IBD. 12/13 pts with ‘highest’ FC levels were diagnosed with IBD.

Summary and Conclusion:
The ‘highest’ FC levels (>600mcg/g) accurately identify all patients with final diagnosis of IBD. Furthermore, this level of FC identified all patients with IBD despite normal values of standard biochemical markers in 16% of cases. In our opinion, all patients with a FC >600mcg/g should be strongly considered for endoscopic evaluation. None of the patients with ‘intermediate’ FC levels (50-600mcg/g) were diagnosed with IBD; thus, we suggest that these patients remain under follow up and an alternative diagnosis is sought. Only when there are other clinical indicators and/or if FC subsequently rises >600mcg/g, should endoscopy be considered.

References:
2. Faecal calprotectin for screening of patients with suspected inflammatory bowel disease: diagnostic meta-analysis. P.F van Rheeinen et al. BMJ 2010; 341:c3369
Diagnostic efficacy of point of care rapid urease test (CLO®) in the diagnosis of H. Pylori in children

Rao, Prithviraj; Sharma, Shishu: Sheffield Children's Hospital, Sheffield.

Aims:
Primary: To evaluate the diagnostic efficacy of point of care rapid urease test (CLO®) in the diagnosis of H. Pylori in children.
Secondary: To identify if preceding treatment with proton pump inhibitors (PPIs) influences the diagnostic yield of CLO. To identify if there was an ethnic predilection in the South Yorkshire population.

Methods:
Single tertiary centre retrospective Audit over a defined period of 3 years (Sep 10 –August 13). Inclusion criteria: All CLO positive OR histology positive children (age range 12 months to 16 years) were identified through our endoscopy and histopathology database respectively.

Exclusion:
Absence of record of CLO in the endoscopy report. Case notes were retrieved and reviewed by a single researcher. The study was approved as a service evaluation project by the Sheffield Children's hospital clinical governance board.

Results:
A total of 57 episodes of CLO or histology +ve episodes were identified in 55 patients (males 60%, n=33), age range (12 m- 16 yrs). The highest ethnic predisposition, other than white british children (36.3%, n = 20) was seen in children of Somalian origin (25.4%, n = 14). All 14 Somalian children that were indicated for endoscopic evaluation were CLO and histology positive.

Summary
7 children were on hydrolysed milk in view of suspected Cow's milk protein intolerance with no benefit and 2 were breast fed. Apart from 3 preterm and 2 IUGR babies, no failure to thrive was noted.

On first consultation:
7 children were on hydrolysed milk in view of suspected Cow's milk protein intolerance with no benefit and 2 were breast fed. Apart from 3 preterm and 2 IUGR babies, no failure to thrive was noted.

Only 2 (6%) showed delayed passage of meconium (>48 hours) and 4 (12%) no data was available. Only 13 (37%) were subjected for rectal biopsy at Birmingham Children Hospital as 22 (63%) other children showed good response to medical treatment but required combination of laxatives along with general management. 3 neonates were diagnosed to have HD (all preterm babies).

14 children were on hydrolysed milk formulae with variable success. All children required more than 1 laxative medication more than recommended doses for the management of constipation. 6 children also required frequent suppository/microlax enema.

Over the period of 4-12 months, 16 (46%) children were fully recovered. 15 (43%) children were controlled with medication even after 18 months. 2 children had surgery (one for Hirchprung disease (HD) and another for abnormal anal position) 2 are still awaiting surgery.

Conclusion
This study suggests that CLO is a highly sensitive and specific point of care test for H. pylori during endoscopy in symptomatic children. Preceding PPI treatment does not appear to influence the diagnostic yield of CLO. There may be a relative higher prevalence of H. pylori infection in Somalian children resident in the South Yorkshire area and needs further prospective studies to explore the same.

<table>
<thead>
<tr>
<th>Histology positive</th>
<th>Histology negative</th>
</tr>
</thead>
<tbody>
<tr>
<td>CLO positive</td>
<td>42 8</td>
</tr>
<tr>
<td>CLO negative</td>
<td>7 374</td>
</tr>
</tbody>
</table>

Sensitivity = 85.71% Specificity = 97.9%
Type I error (False positive rate) = 2.1% Type II error (False negative rate) = 14.29%
Likelihood positive ratio = 40.81 Likelihood negative ratio = 0.14

Early onset constipation: what are the lessons learnt?

Dr Sukaina Asad, Foundation Year 2; Dr Felicity Boat; Foundation year 1 and Dr Subramanian Mahadevan-Bava, consultant paediatrician and paediatric gastroenterologist. Russell's Hall Hospital, Dudley, West Midlands D 7Y 2 HQ

Early onset constipation (EOC), developing constipation before 6 months of age, is not an uncommon condition but causes great distress to the child and carers. EOC is also a big challenge for Primary care team and paediatricians to manage. All clinicians in such referral would like to rule out congenital anomaly and Hirschsprung's disease (HD).

Aim
The aim of this study was to evaluate the outcome of EOC and relation to first bowel action, medical treatment, role of rectal biopsy, as well as significant congenital anomalies associated with it.

Methods
A retrospective study of 35 patients diagnosed as EOC between 2008 and 2013 referred to the Paediatric gastroenterology clinic at Russell’s Hall Hospital. The patient demographics, age of onset of symptoms, investigations and management were evaluated.

Results
Of 35 children, there were 20 (57%) males and 15 (43%) females, all Caucasian except one mixed race. Three (8%) were preterm babies stayed in neonatal unit for few weeks. 1/35 (3%) was found to have abnormally placed anus that was missed at the time of newborn baby check. The age of onset of symptoms was reported as since birth in 24 (69%) babies, 11 (31%) between 1-5 months. Median age of referral (many seen by other professionals before referral to GI clinic) was 5 months. 5 (15%) children had associated symptoms (GORD/poor feeding) and 4 (11%) had developmental delay.

On first consultation:
Over the period of 4-12 months, 16 (46%) children were fully recovered. 15 (43%) children were controlled with medication even after 18 months. 2 children had surgery (one for Hirschsprung disease (HD) and another for abnormal anal position) 2 are still awaiting surgery.

Conclusion
EOC is a common condition and has significant impact on quality of life for both infant and carer. In our study, there was no correlation between the timing of meconium at birth and EOC. We would like to stress the importance of being vigilant for the possible diagnosis of HD especially in preterm babies as this can be easily missed unless suspected. Also requires education of trainees related to abnormally placed anus that can be missed at birth as it is uncommon. Change in hydrolysed formulae is not an option. All required close support and medical management is the key to success.
**G18**

"Polyuria" with glucose-galactose malabsorption: a case report
Kalamchi, Sarmad, Marzouk, Hanaa, Lambert, Heather, Urs, Arun, GNCH, Newcastle Upon Tyne

**Background:**

Glucose-galactose malabsorption (GGM) is a rare autosomal recessive disorder due to selective defect of intestinal transport of glucose and galactose, leading to severe, watery, acidic diarrhoea, dehydration, failure to thrive, or early death. The disease was first reported in 1962 by Lindquist and Meeuwisse from Sweden and Laplane et al. from France. Since then over 300 cases are reported worldwide. Nephrocalcinosis, nephrolithiasis and proximal renal tubular dysfunction in association with GGM have been reported but only in handful of cases.

**Case:**

A one week old boy presented to local hospital with weight loss (20%), lethargy and signs of dehydration despite adequate bottle feeding with no apparent diarrhoea. He was the first-born child of first-cousins of Caucasian origin. He was delivered at term, with birth weight of 3650 g. The pregnancy, delivery and immediate neonatal period were uneventful. His initial investigations revealed metabolic acidosis, profound hypernatremia (184 mmol/dl), hypercalcaemia (total / ionized calcium 3.2 /1.6 mmol/l) and renal failure (urea 13mmol/l creatinine 83umol/l). Abdominal ultrasound showed bilateral medullary nephrocalcinosis. He was transferred to our centre at 13 days old as intermittent improvements were not sustained, despite careful rehydration. He was initially thought to be polyuric and his profuse watery diarrhoea, was not recognised until he underwent bladder catheterisation to aid fluid balance management. The resolution of diarrhoea upon termination of enteral feeds and stool chromatography positive for glucose raised the clinical suspicion of GGM. He was started on fructose-based formula (Galactomin 19, Nutricia®) with full resolution of biochemical abnormalities and weight gain of 3 kg within 6 weeks (50th centile). An analysis for SGLT1 gene mutation has been requested.

**Conclusion:**

This case illustrates a typical picture of rare disease where early recognition and treatment is life saving. Careful clinical observations together with laboratory investigations led to the diagnosis. The pathogenesis of hypercalcaemia and nephrocalcinosis remains unclear, with resolution of hypercalcaemia along with other biochemical abnormalities after institution of glucose-galactose free feeds.

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**G19**

A case series of post endoscopic duodenal haematoma and its natural history
Dr. Nandhini Kumaraguru, Dr. Sakthivel Gnanasambandam, Dr. Fevronia Kiparissi, Dr. Neil Shah, Dr. Mamoun Elawad, Great Ormond street Hospital NHS Trust, London.

**Introduction**

Upper gastrointestinal (GI) endoscopy or gastroscopy is a commonly performed procedure in most of the gastrointestinal units but intramural duodenal haematoma (IDH) is not frequently reported following this procedure. But it is being increasingly recognised as a complication following upper GI biopsies (1).

**Aim**

To determine the symptoms at presentation, clinical course, investigations, interventions and outcome on children who developed IDH following endoscopy.

**Methods**

Electronic hospital records were searched with search word “duodenal haematoma”. We retrospectively reviewed six children who developed IDH and presented with various abdominal symptoms following an upper GI endoscopy from a single tertiary gastroenterology centre from May 2003 to August 2013. Nearly 10,000 children have had gastroscopies during this period.

**Results**

All six patients were conservatively managed successfully with predominantly prolonged recovery period and didn’t require any surgical intervention.

**Summary**

All cases presented within 48 hours of endoscopy with abdominal pain, bilious or coffee ground vomiting or fluid intolerance. Hypernatremic dehydration and haemorrhagic shock were unusual presentations seen in our study group. Diagnosis should be made promptly with low index of clinical suspicion and the support of different available imaging modalities like ultrasonogram, upper GI series, CT and MRI scan depending on availability and prognosis. All children were conservatively managed and required parenteral nutrition due to feed intolerance. Laparotomy is not indicated in uncomplicated IDH.

**Conclusion**

Presently there are no predictors identifiable which can predispose to this condition and we do not recommend investigations like coagulation screen and full blood count routinely before the endoscopic procedure. Varying presentations of IDH should be borne in mind by physicians if a child presents to hospital following this common day case procedure as it is crucial to recognise this condition which has a good outcome following conservative management.

**References:**

**G20**

**Life threatening GI bleeding in Klippel-Trenaunay-Weber: initial response to thalidomide but not to sirolimus**

Tamhne S, Malik M, Ragunath K, Charlton CPJ; Departments of Paediatric and Adult Gastroenterology and Paediatric Nephrology Nottingham Children’s Hospital and Queens Medical Centre, Nottingham, NG7 2UH.

**Background**

Klippel-Trenaunay syndrome is a congenital vascular anomaly of unknown aetiology characterized by a triad of varicose veins, cutaneous capillary malformation, and hypertrophy of bone and/or soft tissue. Gastrointestinal vascular malformations in this syndrome may present with gastrointestinal bleeding. Sirolimus is a mTOR inhibitor immunosuppressant used in transplantation with some case reports of use in various vascular abnormalities and Thalidomide is a VEGF inhibitor of new vessel growth with case reports of use in a number of various rare conditions. 

**Methods**

Rhia 4y thought to have KTW and renal dysplasia with chronic anemia, was given erythropoetin and iron infusions. Though with normal stools and no GI symptoms, endoscopy showed multiple oozing gastric angio-dysplasias (normal duodenum, ileum and colon). Argon Photo Coagulation ablated the lesions but overt GI bleeding then required repeated transfusions. Repeat endoscopy showed multiple new bleeding lesions which were ablated without reducing transfusion requirement. Capsule endoscopy showed lesions only in upper GI tract. Because her renal dysplasia would ultimately require renal transplant we tried Sirolimus which had been reported to control abnormal vessel regrowth. Six weeks treatment showed no benefit with increasing bleeds and repeat endoscopy showing new lesions requiring APC and transfusions. Thalidomide was tried with decrease in bleeds and transfusion requirement within 2 weeks, and none during next 5 weeks treatment. The visible vascular lesions in the cheeks disappeared in first 2 weeks of treatment. Also on treatment with prednisolone (weaned down and stopped) tranexamic acid and sc octreotide. Rhia has had cramps in her hands which have been reported in Thalidomide treatment along with serious longterm peripheral neuropathy which we are monitoring. Unfortunately Rhia has just been readmitted with drop of Hb and awaits endoscopy assessment.

**Results**

Rhia’s GI bleeding and angiodysplasias deteriorated further while on 6 week sirolimus course and had initial response to course of thalidomide.

**Conclusion**

This is a single case report in a very rare complication of the rare condition KTW syndrome. Her care has necessitated discussion and advice in our own team, colleagues in BSPGHAN including Neil Shah and frequent help from our adult team. We share this experience with the family’s consent to encourage sharing of information to help Rhia and other children. We need to identify the mechanism of this condition and treatments to help these patients in the future.

**References**


**G21**

**Accuracy of the Strobel and Modified Strobel formulae in pH catheter placement in children: A prospective study**

Saha, Amit; Bhanduri, Bim (Prof); Maidstone Hospital, Maidstone, Kent, UK

**Background:**

Gastro-oesophageal reflux is a common, physiological and self limiting condition in infants. However early diagnosis and management is essential for prevention of complications. Oesophageal pH monitoring is the gold standard technique for the detection of acid gastro-oesophageal reflux episodes, and correct placement of the catheter is crucial for accuracy of the readings. The Strobel formula (0.252 × height in cm + 5) is frequently used as a guide to determine distance from the nostrils to the lower oesophageal sphincter(LES). However, a modified Strobel formula was introduced for more accuracy as follows: (1) infants <12 months (height in cm x 0.252 + 2); (2) older children >12 months (height in cm x 0.226 + 4.6) x 0.87.

**Aim:**

To compare the accuracy of Strobel formula and modified Strobel formula in achieving correct placement of the catheter tip, in children more than 1 year of age.

**Subjects and Methods:**

A previous study on 15 infants less than 1 year of age showed that the modified Strobel formula was more accurate. We did a further prospective study with 14 children to determine the accuracy of these formulae in older children. Initial catheter length was calculated using the modified Strobel formula, and the actual catheter placement was adjusted and confirmed either by direct measurement of length at endoscopy and/or confirming its tip position between T8 and T10 level on chest X-ray.

**Results:**

The 14 patients included in this study ranged in age from 1 year 2 months to 15 years (male n=8, female n=6), with a mean age of 7 years 7 months. The actual catheter position (as determined by endoscopy or chest radiograph or both) was compared with the predicted position as determined by the above formulae, and their deviation from actual LES position was calculated. The mean deviation for the Strobel formula was +6.9 (+2.8 to 11.0) cm, whereas the mean deviation for the modified Strobel formula was -0.99 to +3.61 cm. Therefore, our data shows that the Strobel formula overestimated the LES distance by an average of 6.9cm, whereas the modified Strobel underestimated it 0.99 cm on average.

**Summary and Conclusion:**

This study showed that the modified Strobel formula is more accurate than the Strobel formula in pH probe placement in children more than 1 year of age. Data from a similarly designed previous study had also shown it to be more accurate in infants less than 1 year of age. We recommend that the modified Strobel formula be used in all age groups of children for initial pH probe placement. However, in order to do a reliable pH study, radiographic confirmation of the catheter tip position must still be considered when using any formula.
An Interesting Cause of Secondary Intestinal Lymphangiectasia.

R Taylor1, A Godse2, S Bunn1, S Hodges1. 1Department of Paediatric Gastroenterology, Great North Children's Hospital Newcastle upon Tyne; 2Department of Paediatric Surgery, Great North Children's Hospital Newcastle upon Tyne.

Introduction

Primary intestinal lymphangiectasia is caused by a congenital malformation, it can also occur secondary to an acquired obstruction of intestinal lymphatic drainage. We present a case of a 22 month old child who developed intestinal lymphangiectasia as a result of an undiagnosed congenital diaphragmatic hernia.

Case

A previously well 22-month-old female presented with a 6-month history of diarrhoea, weight loss and lethargy. Initially she was investigated for coeliac disease with duodenal biopsies. Histological results did not support this diagnosis. At presentation she was hypoalbuminaemic, lymphopenic and further investigation had revealed hypogammaglobulinaemia and very low T-cells. A diagnosis of intestinal lymphangiectasia was postulated and she was commenced on an MCT feed and low fat diet. This resulted in a dramatic improvement in her symptoms of diarrhoea. On outpatient follow up she was noted to be tachyoeic with absent breath sounds on the left side. A chest x-ray showed opacification of the left hemithorax with gas filled lucencies. A diagnosis of congenital diaphragmatic hernia was confirmed with chest ultrasound.

At laparotomy a 3x3cm diaphragmatic defect was identified and repaired. Small and large bowel loops were present in the chest. 150mls of lymphatic fluid was drained from the abdomen. The left lung was small. She was discharged following an uncomplicated postoperative course. Initially she was kept on a low fat diet and MCT feed. Subsequently she was re challenged with a normal diet and had no recurrence in symptoms. Two months post operatively she had normal plasma albumin level, normal lymphocyte level and a normal chest xray. The herniation of her bowel had resulted in secondary lymphangiectasia which following repair of the hernia has resolved.

Conclusion

This case highlights the importance of considering intestinal lymphangiectasia in children with a protein losing enteropathy. It also demonstrates the difficulty in diagnosing young children with small bowel pathology such as lymphangiectasia where modalities such as capsule endoscopy and push enteroscopy would be beneficial.

Dysautonomia and Foregut Dysmotility in the Joint Hypermobility Syndrome

Protima Amon1, Rebecca Irvine2, Nigel Meadows1, Patricia Taraborrelli3, Phang Boon Lim3, Nelly Ninis and David Rawat1
1Department of Paediatric Gastroenterology, The Royal London Hospital; 2Department of Paediatrics, St Mary’s Hospital, Paddington; 3Imperial College Syncope Diagnostic Unit, Hammersmith Hospital

Background:

The joint hypermobility syndrome (JHS) is a multi-system inherited connective tissue disorder. In addition to the skin and musculoskeletal manifestations, gastrointestinal (GI) symptoms and autonomic nervous system-related symptoms are commonly seen which can cause significant debilitating disease. Postural orthostatic tachycardia syndrome (POTS) is a form of dysautonomia which is well recognised to be associated with JHS in adults but there is very little data in children. POTS is often difficult to diagnose. A tilt table test is vital in the diagnosis of POTS, although all symptoms must be considered before a final diagnosis is made. The presentation may be so varied and subtle that clinicians often fail to recognize autonomic disturbance as an individual clinical entity within the disorder leading to inadequate management.

Aims:

The purpose of this study was to investigate the presence of cardiovascular autonomic dysfunction in paediatric patients with JHS and describe the association with foregut symptoms.

Methods:

We conducted a retrospective review of the medical notes and electronic patient records for all children diagnosed with JHS (Beighton score >4/9) referred to the syncope service based at Hammersmith Hospital. The study period was June 2011 to October 2013. Cardiovascular autonomic function was evaluated using the tilt table test.

Results:

A total of 53 children were recruited with a median age range of 15.2 yrs (range 8.1yrs-18.7 yrs). The gender distribution was 28 females (53%) and 25 males (47%). All patients had autonomic nervous system-related symptoms including postural dizziness or syncope (53%), palpitations (40%), migraine (25%) and chest pain (19%).

<table>
<thead>
<tr>
<th>GI Symptoms</th>
<th>Tilt test result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nausea/bloating</td>
<td>POTS</td>
</tr>
<tr>
<td>Reflux</td>
<td>Post and vasovagal syncope</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>Vasovagal Syncope</td>
</tr>
<tr>
<td>Constipation</td>
<td>Test not completed/inconclusive</td>
</tr>
<tr>
<td>Diarrhoea</td>
<td>Normal</td>
</tr>
<tr>
<td>Feeding difficulties</td>
<td>Normal</td>
</tr>
<tr>
<td>No GI Symptoms</td>
<td>Normal</td>
</tr>
</tbody>
</table>

Of the 53 JHS patients with autonomic symptoms, 48 (91%) patients had GI-related symptoms. 45 (85%) of these patients had confirmed cardiovascular autonomic dysfunction on tilt testing. All these patients also complained of foregut symptoms, predominantly nausea and bloating.

Summary:

Foregut dysfunction was a comorbid presentation in all JHS patients with cardiovascular dysautonomia.

Conclusion:

Children with JHS presenting with foregut symptoms are likely to have associated cardiovascular autonomic dysfunction. Recognition of these symptoms by paediatricians is important as formal evaluation and treatment of the dysautonomia may improve the quality of life for these patients. Further studies are required to evaluate the objective improvement of GI symptoms following treatment for dysautonomia.
Phlebectasias are a rare cause of gastrointestinal bleeding in children.

We present the case of a 12 year old boy presenting with melaena. Upper Gastrointestinal (GI) endoscopy was normal, but colonoscopy demonstrated signs of ulcerative colitis and biopsy confirmed this. His symptoms responded well to medical treatment. Repeat colonoscopies and biopsy, when well, reaffirmed the diagnosis. On discontinuing medication he represented with melaena. Repeat colonoscopy showed ulcerative colitis and a small juvenile polyp. After resolution of symptoms he was discharged. However, he presented a year later with melaena, haemodynamic instability and Haemoglobin 5.7. Both lower and upper GI endoscopy and biopsies on this occasion were entirely normal. It was not until capsule endoscopy, in conjunction with CT angiography, was performed that the diagnosis of phlebectasia was made. Surgical resection of the affected jejunum was curative. A diagnosis of phlebectasia type cavernous haemangioma was made on histology. Retrospective re-evaluation of the initial histology suggests reactive rather than ulcerative colitis as the histology on the initial colon biopsies. He remains well with no readmissions, and has been discharged from follow up.

The first case of phlebectasia of the jejunum in a child was reported in 2007. This is the second paediatric case reported in the English literature. Our case highlights the importance of a multimodal approach to investigating ongoing obscure gastrointestinal bleeding.

Phlebectasia of the jejunum caused by cavernous haemangioma

Multifocal lymphangioendotheliosis with thrombocytopenia (MLT) or Cutaneovisceral angiomatosis with thrombocytopenia (CAT): a rare cause of gastrointestinal bleeding in neonatal period

Severe gastrointestinal bleeding in newborn is rare but serious. In 2004 North et al described MLT on histological sections of skin biopsy in children presenting with congenital multifocal cutaneous and gastrointestinal vascular anomalies, thrombocytopenia and GI bleeding. Around twenty cases have been described in the English literature, although other cases are reported under different names. The aetiopathogenesis is still unknown with reported mortality rate estimated at 65%. The refractory nature for treatment with multiple therapies poses significant challenges to clinicians involved in the care of the child. We describe a case that exemplifies the rare clinicopathological entity.

Case presentation:

A full term baby girl was born by C-section for maternal antepartum haemorrhage following an uneventful pregnancy. She weighed 2.37 kg and had multiple well-defined macular telangiectatic lesions on trunk and extremities. Some of the lesions were bright red and blanched on pressure whereas darker, red brown lesions did not. There is no family history of similar lesions or bleeding diathesis. Initial laboratory investigations revealed thrombocytopenia (nadir 23,000/L) with normal Hb (15.6g/dl), clotting profile including fibrinogen. At 4 weeks of life, she presented with upper GI bleeding (hematemesis and melena) with significant drop in haemoglobin (7.2g/dl) and platelets. Initial resuscitation included multiple blood and platelet transfusions, tranexamic acid, octreotide, omeprazole and steroids. She was placed NBM with total parenteral nutrition (TPN) before gradual reintroduction of enteral feeds. Endoscopic examination revealed multiple, red haemangiomatous macules of varying sizes with active oozing of blood in the stomach. Magnetic resonance imaging of head showed foci of haemorrhage above the body of the right lateral ventricle without evidence of an arterio-venous or venous malformation. MR angiogram of small intestine showed no further lesions and a chest CT showed bilateral nodules in lower lobes. A skin punch biopsy revealed prominent blood vessels in the papillary dermis, with a slightly ‘hobnail’ appearance to the lining endothelial cells, with fibroplasia and haemosiderin deposition seen around blood vessels in the deeper dermis. CD31 immunostain highlighted the lesional endothelial cells with D2-40 immunostain negative for lymphatics. At 3 months of age, she continues to have small tarry stools requiring regular weekly blood transfusion to maintain Hb > 10gm/dl. She has been treated with propranolol, methylprednisolone and lately vincristine which seemed to have reduced the frequency of blood transfusion. A repeat endoscopy and skin lesions remain unchanged.

Conclusion:

MLT and CAT are the same condition and approximately 40% have negative D2-40 immunostaining. GI bleeding is a major source of morbidity and mortality in patients in first year of life. In infants who survive, the severity of bleeding appears to decrease in first 2 or 3 years of life. A number of other multifocal vascular skin disorders are described in childhood but not associated with thrombocytopenia. Besides the skin and GIT, multiple other organs can be involved. Various therapies have been used with variable outcomes but information coming from a large registry suggests it is unclear if it is treatment response, spontaneous resolution or both. The clinicopathological features, natural history and treatment of this rare disease have yet to be established.
Quality of Life in Children assessed for Chronic Intestinal Pseudo Obstruction.
Barkley Lisa; Lewis Hannah; Great Ormond Street Hospital, London

Background

Chronic Intestinal Pseudo-Obstruction (CIPO) is a rare condition within paediatric populations (Bursch & Hyman, 2006). Living with associated symptoms (including pain, incontinence, diarrhoea and difficulties with feeding), anecdotaly leads to high levels of family distress and impedance in the child's daily functioning and emotional well-being. A paucity of research exists in this area; one US study indicated children with a diagnosis of CIPO reported high levels of emotional distress and poor perceived health related quality of life (QoL) (Schwankovsky et al., 2002). Similarly, adults diagnosed with CIPO have reported poorer HRQoL than patients with other gastroenterological conditions (Cogliandro et al., 2011). This audit presents data collated as part of a UK wide paediatric assessment pathway for CIPO. The aim was to identify the impact of symptoms on family's perceived quality of life and daily living to better understand the needs of families coming into the service.

Method

Proxy-report Likert scale questionnaires of the child’s QoL were completed by parents (Paediatric Quality of Life Inventory, PEDSQoL, Infant version and Young Child version,Varni,1998). Scores from the PEDSQoL related to social, emotional, physical and educational domains. Families also participated in a semi-structured assessment interview conducted by the Clinical Psychologist working in the service. Thematic analysis of reports generated was carried out by an independent clinician.

22 patients completed a psychology assessment as part of their admission over a 16 month period. Data from patients with significant concurrent conditions or disabilities were excluded (n=2). As older patients referred were likely to have had greater variation of experiences leading to assessment and disease/symptom progression, for the purpose of this audit, data collated children over 11 years old were excluded (n=4). The data presented here represents 16 patients (8 boys and 8 girls, Age range: 0.4-7.6 years, mean=3.6 years).

Results

Data from the PEDSQoL were scored against normative data from healthy children and compared to published data relating to children with inflammatory bowel disease (IBD)(Upton et al., 2005). Compared to healthy children the children undergoing CIPO assessment reported lower levels of QoL across all domains. Scores were also found to be consistently lower than children with an IBD across all domains.

Themes impacting upon child and family quality of life were: the negative impact of symptoms on child emotional wellbeing and functioning, hospital stays and their implications, difficulties within relationships and the impact upon family functioning, and the process of seeking a diagnosis (including being "heard").

Conclusions

This sample is a small and heterogeneous one, however; this preliminary data suggests symptoms led to high perceived impact on the child's quality of life, and both the child and family members' daily functioning. Data suggests this population experience poorer QoL than other families where children have a similar condition. Service changes were implemented to better take these factors into account. As additional data becomes available, research to compare experiences of those who ultimately received a diagnosis of CIPO to those without clear pathology would be beneficial.
Growth Hormone Deficiency Presenting De Novo as Non-Alcoholic Fatty Liver Disease

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Background:
The high prevalence of childhood obesity has resulted in increased referral rates of patients with the triad of transaminitis, overweight and fatty liver infiltration on ultrasound with presumed non-alcoholic fatty liver disease (NAFLD). Fatty liver due to growth hormone deficiency in children is an uncommon but well recognised cause of NAFLD. We report 2 cases of growth hormone deficiency previously unrecognised in two children who were referred to hepatology due to obesity and presumed NAFLD. The importance of accurate anthropometry in reaching this diagnosis is discussed.

Case 1:
A 15 year old male was referred to the liver clinic with transaminitis, having been under follow up locally at a DGH for some years with non-specific problems including migraine. His height was on the 9th centile and weight on the 98th centile. He had not attained puberty. His liver ultrasound revealed fatty liver. In view of the discrepancy between height and weight centiles, and delayed pubertal onset, a full metabolic screen along with pituitary function tests were performed. He was found to be hypoglycaemic on prolonged fasting and had marked glyceroluria. The cause for this remains unexplained with normal glyceral kinase (GK) levels and no abnormality identified on genetic testing of the GK deficiency locus. He was subsequently found to be growth hormone deficient on stimulation (peak GH = 0.20 µg/l, IGF=66 µg/l) and has now commenced growth hormone injections.

Case 2:
An 11 year old male was referred by his GP with transaminitis (AST of 198, ALT of 270) (normal 0 – 40), elevated alkaline phosphatase (295 U/L) and normal liver ultrasound. Initial investigations revealed a normal thyroid profile and normal fasting glucose and insulin levels. He initially presented at another hospital with skin rash and was discharged worsened. He was referred to the endocrinology team in view of his discordant weight and height. Anterior pituitary stimulation testing revealed isolated growth hormone deficiency (peak GH =4.00 µg/l, IGF=69 µg/l). He was started on growth hormone therapy and his liver function tests normalised. He is reviewed on a 6 monthly basis.

Discussion:
These cases reiterate the importance of plotting height and weight relative to a target centile range and mid-parental height in patients referred with obesity and suspected NAFLD. Obese children are usually tall for age. Reduced height for target centile range and high body mass index with central obesity may indicate that NAFLD has resulted from growth hormone deficiency. This should be confirmed by anterior pituitary stimulation testing.

The first case of fatty liver caused by growth hormone deficiency was reported by Takano et al1 in 1997. Following a trial of daily growth hormone therapy the transaminitis and the fatty liver normalised after four months due to suppression of lipogenesis in favour of lipolysis. The association between GH deficiency and fatty liver is now well recognised. However, NAFLD is equally common in patients with GHD and age and BMI-matched controls2. Therefore accurate anthropometry is of fundamental importance in reaching a diagnosis of GH deficiency in patients presenting with transaminitis and obesity to hepatology clinics.


Hepatic Involvement in Langerhans Cell Histiocytosis

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Background
Langerhans cell histiocytosis (LCH) is a rare, non-malignant disease of unknown aetiology, characterised by extreme heterogeneity and an unpredictable course. Hepatic involvement in LCH is associated with worse prognosis.

Methods
A retrospective review of children with LCH referred to a tertiary paediatric hepatology centre from 1999 to 2012.

Results:
Ten children (7 girls with LCH were referred at a median age of 30 months (range 10 – 72). Median age at diagnosis was 17 months (range 8 – 27). Multisystem disease was documented in all but one, where initial information was not available. At presentation, skin rash was found in all and hepatic involvement was noted in 8, including 4 with hepatomegaly and 1 with hepatosplenomegaly. Diagnosis was confirmed by CD1a/S100 immunostaining on skin biopsies. All patients except one underwent a liver biopsy, suggestive of sclerosing cholangitis (SC) in 7 patients, with CD1a-positive cell infiltrates in 3. Paucity of intrahepatic bile ducts and biliary changes compatible with graft-versus-host disease (GVHD) were the findings in the other 2 biopsies.

Systemic chemotherapy was the initial treatment for 8 patients. Two children had a very aggressive LCH in liver disease and died at 23 and 31 months without liver transplant (LT). Six patients with severe SC and progressive portal hypertension (PHT) received LT at median age of 84.5 months (range 13 – 139). All except one were jaundiced (median bilirubin 193 µmol/L, range 13 – 732). Splenomegaly, thrombocytopenia and eosinophagial varices were present in 5, 4 and 3 patients, respectively. Liver explants showed biliary cirrhosis in all. One patient died shortly after LT from fungal sepsis. He also had lung LCH involvement at the time of LT. Two girls had biopsy-proven LCH recurrence in the graft at 6 and 60 months after LT, respectively. One of them had been successfully treated for post-transplant lymphoproliferative disease. They received further chemotherapy which controlled the disease in one, but the other required re-LT for progressive disease 3 years later. Five of the transplanted patients are alive after a median follow up of 14 years (range 0.75 - 18) post LT.

One of the patients had no preceding liver involvement and was diagnosed with post-haematopoietic stem cell transplant GVHD. Another patient normalised his liver function tests after chemotherapy. They are both asymptomatic 41 and 56 months after diagnosis, respectively.

Conclusion
Hepatic involvement is almost universal in multisystem LCH. Progression of PHT often heralds a requirement for LT. This is a viable management option, if the LCH is in prolonged remission.
Paediatric intestinal transplantation in the modern era: King's College Hospital Experience

**Introduction:**

We established an intestinal transplant service in 2009 in collaboration with a tertiary intestinal rehabilitation centre (Great Ormond Street Hospital) to complement surgical and medical strengths of both units. We report our 4 year experience.

**Methods:**

A retrospective review of the children who received an intestinal transplant at King's College Hospital from 2009 to date.

**Results:**

From August 2009 to October 2013 10 children (5 male) underwent intestinal transplantation at median age 5 years (range 0.5-16). The diagnoses were: short gut syndrome (gastroscisis (3), strangled diaphragmatic hernia (1)), chronic intestinal pseudo-obstruction (3), intestinal lymphangiectasia (1), microvillous inclusion disease (1) and antenatal midgut volvulus with biliary atresia splenic malformation syndrome (1). Eight received an isolated small bowel transplant and two a combined liver and small bowel transplant.

Complications:

Two patients developed severe acute cellular rejection treated with ATG. One responded and one died from sepsis after graft removal. Four patients developed post-transplant lymphoproliferative disease. All were treated successfully; three with rituximab, one with chemotherapy. One patient died from sepsis after graft removal. Four patients developed post-transplant lymphoproliferative disease. All were treated successfully; three with rituximab, one with chemotherapy. One patient died from sepsis after graft removal.

Immunosuppression:

ATG was given to the donor for the first 4 transplants. All had basiliximab induction with tacrolimus and prednisolone maintenance. Sirolimus was added to all isolated bowel transplant recipients after 1 month, but was stopped in 7 of 8 due to neutropenia. One is also on azathioprine. Three patients were given infliximab due to ongoing graft inflammation without apoptosis. One improved after two doses. Two improved and remain on 8 weekly doses.

**Conclusion:**

An intestinal transplantation programme set up in units with liver transplant and intestinal rehabilitation experience can achieve excellent results comparable to large centres.

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**H4**

The use of bone marrow aspiration as an investigative tool in paediatric liver disease.

**Introduction:**

Bone marrow aspiration (BMA) has traditionally been an important tool in the investigation of paediatric liver disease, particularly in the diagnosis of storage conditions or Haemophagocytic lymphohistiocytosis (HLH). It is an invasive measure however, and diagnostic yield is uncertain. We reviewed experience of BMA in a tertiary paediatric centre to determine its best application in practice.

**Methods:**

Paediatric liver patients who had a BMA for suspected storage conditions over a 10 year period were identified through a haematology database. Clinical, metabolic, haematological and histological data were reviewed. SPSS v 17.0 was used for analysis.

**Results:**

During the study period, 154 children (105 boys) underwent 182 BMAs for suspected storage conditions. Median age of presentation was 0.39 years (IQR 0.19, 1.63). Presenting features were conjugated hyperbilirubinaemia in 75 (38 also splenomegaly (SM), 3 hepatomegaly (HM) and 14 hepatosplenomegaly (HSM), acute liver failure in 29, isolated HM in 7, SM in 10 and HSM in 23. Final diagnosis was Niemann Pick C (NPC) in 13, NPA or B in 3 and Wolman disease/cholesteryl ester storage disease in 2, Glycogen storage disease in 4, mitochondrial cytopathy in 6, HSM in 4. Aetiology was not identified in 70 (46%), 7 of whom have died and the remainder discharged or followed with stable disease. BMA was haemodilute/inadequate in 20(13%). The sensitivity of initial BMA for detection of storage conditions/H/HLH in the setting of ALF was 57% (86% on repeat) with a positive predictive value (PPV) of 100% and a negative predictive value (NPV) of 83%. In infants < 1 year with conjugated hyperbilirubinaemia (HSM), the sensitivity for detection of storage disorders was 71% with a PPV of 91% and a NPV of 76%. In children > 1 year, the sensitivity was 83% with PPV of 100% and NPV of 95%. At the time of writing, 49 had a previous diagnostic BMA or WCE. Time from biopsy to results was a median 3 months.

**Conclusions:**

The overall sensitivity PPV and NPV for BMA as an investigative tool for investigation of paediatric storage disorders were high, particularly in the setting of acute liver failure when a rapid diagnosis was necessary. Inadequate samples requiring repeat procedure are frequent however, and for non-urgent indications, WCE and skin biopsy may be preferred.
Overview of Combined Liver and Kidney Transplantation in Children in the UK.
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Aim
To describe a single UK centre's experience of Combined Liver and Kidney Transplantation in Children (CLKT).

Introduction
The 1st Combined Liver and Kidney Transplant (CLKT) worldwide took place in 1984 (Margreiter et al 1984). Since then CLKT has been carried out as a recognised treatment for End Stage Renal Disease (ESRD) with associated liver disease. There is little published data on CLKT in children.

Method
A retrospective study was carried out on all patients who underwent CLKT at our centre.

Results
Children were usually referred from local renal units and were assessed for transplant jointly by the paediatric hepatology and renal teams.

The first CLKT was undertaken in March 1994. Since then a total of 38 children have received a CLKT. The most common indications for CLKT were ESRD due to Fibropolycystic Disease (n=24) and Primary Hyperoxaluria type 1 (n=11).

Median age (range) and weight at transplant was 5.5 years (1.5-15 years) and 20.5 kg (9.1-57.25 kg). Median survival since transplant is 5 years (1 month - 14 years). Actuarial 14 year patient and renal graft survival is 80%. Of the 5 deaths, 2 were in the peri-operative period and the other 3 late deaths due respectively to vascular thrombosis, sepsis and seizures.

10% had an episode of histologically proven acute liver rejection and 5% acute kidney rejection. All rejection episodes were successfully treated with pulsed corticosteroids.

Currently all patients are well although one is receiving dialysis whilst waiting for a renal retransplant. 7 patients have been successfully transferred to adult services.

Conclusion
In our experience CLKT is a highly successful long-term treatment for ESRD with associated liver disease. Subsequent quality of life is excellent and there is a low incidence of renal rejection.

Late referral of Intestinal Failure Associated Liver disease; an ongoing issue?
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Background:
Cases of intestinal failure associated liver disease (IFALD) requiring combined liver bowel transplantation have decreased with earlier referral to intestinal rehabilitation (IR) units.

Methods:
We describe our recent experience of children with complex intestinal failure referred to a paediatric hepatology unit with end-stage liver disease having missed opportunity for IR management.

Case Studies/Results:
2 patients (A and B) were ex-premature (30+5 week gestation female and 24 week male) infants who developed necrotizing enterocolitis in the first week of life; patient A managed conservatively and B surgically. Both required parenteral-nutrition (PN), being unable to tolerate nutritive volumes of enteral feeding. Both had recurrent sepsis with deterioration of liver function. Both were managed in tertiary neonatal units, neither was discussed with or referred to an IR unit.

Hepatology referral was made after finding an abnormal gallbladder on ultrasound of patient A and stomal variceal bleeding in B, at 4 and 5 months of age respectively. At referral, both had severe cholestasis, portal hypertension (splenomegaly/ascites), and coagulopathy. Liver biopsy was not performed due to coagulopathy.

Results
(A): total bilirubin (TB) 429 µmol/l, direct bilirubin (DB) 359 µmol/l, ALT 108 IU/l, albumin 40, platelets 66, INR 1.9.
(B): TB 582 µmol/l, DB 461 µmol/l, ALT 279 IU/l, albumin 33, platelets 64, INR 2.1

The IR team was subsequently involved and IF management instituted with further GI investigation, re-commencement of enteral feeding and optimization of PN. Neither patient established enteral feeding successfully. Transplant was not possible due to lack of venous access and severe multi-organ dysfunction. Both patients died at 3 months after referral.

Conclusions:
Late recognition of complex IF and IFALD with consequent delayed appropriate referral and management remains a problem outside of IR and transplant centres. We have reported 2 such cases to one unit within the last year - is this the tip of an iceberg?
High risk behaviour in young people – an opportunity for targeted intervention with hepatitis B vaccination in the UK

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Background

The prevalence of blood borne viruses (BBV), hepatitis B (HBV), hepatitis C (HCV) and Human Immunodeficiency Virus (HIV) in adults involved in intravenous (IV) drug use or unsafe sexual behaviour is 50-80% in the UK and accordingly, HBV vaccination is advised for these adults. The incidence of BBV infection in at risk young people <18 years old in the UK is unknown.

Aim:

To identify the prevalence of BBV infection and establish the need for health promotion including HBV vaccination in young people involved in at risk behaviour.

Study Population:

All young people (age <18 years) attending the Young Person’s Substance Misuse service in Birmingham.

Design:

Non-randomised, quantitative study and descriptive questionnaire to ascertain risk behaviour and screen for BBV infection.

Method:

Study information was provided to the young people by their key workers. Those who agreed to take part were seen by the study team and informed consent was obtained. Data collected included: demographics details of risk behaviour, serology for HBV, HCV and HIV. Information about BBV prevention was provided. Non-immune young people were offered HBV vaccination and those found to be infected were referred for clinical care.

Results:

170/500 young people were approached by their case worker and 65 (40%) (28F, 56M; Median Age 17.3; Rang 13 to 18 years) agreed to take part; Caucasian 66%, Afro-Caribbean 13% and Asian 9%. Refusal was either due to fear of needles or they had been tested. Risk behaviour included: 6 IV drug users; 58 cannabis users; 59 had multiple sexual partners (40 had 1 - 5 sexual partners and 19 had > 6 sexual partners), 51 had engaged in unprotected sex. 56 were negative for HBV, HCV, and HIV. 8 were HBV immune following vaccination and 1 was naturally immune.

HBV vaccination was recommended to 56 non-immune young people, but was declined by most of them; The main reasons were that as vaccination was not available at the centre, the young people were reluctant for their confidential information to be disclosed to their family doctors.

Summary:

The study found that no young person screened had yet been infected by BBV which may be related to the small number of IV drug users. The main risk factor for acquiring BBVs in this population was related to unprotected sex with multiple sexual partners. There was no routine provision of hepatitis B vaccination to this at risk group. Refusal by young people to accept screening for BBVs may be due to fear and inadequate counselling.

Conclusion:

The prevalence of BBV infection was low in this group of young people involved in high risk behaviour, but is a major problem in older adults. There is need for better awareness and education about prevention of BBV infection in key workers and at risk young people. This is an opportunity for targeted intervention of hepatitis B vaccination in this group of young people.
Portoduodenostomy for biliary atresia splenic malformation and midgut deletion

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Introduction

Portoduodenostomy can be performed in patients with biliary atresia but insufficient jejunum to create a Roux-en-Y portoenterostomy. We report a case of a 2 year old girl who underwent this procedure for biliary atresia splenic malformation and ultrashort gut, followed by later combined liver small bowel transplantation.

Case report

The patient was noted to have echogenic bowel on antenatal ultrasound scans. Exploratory laparotomy on day 2 of life revealed biliary atresia splenic malformation with midgut deletion from presumed intrauterine volvulus. Only 10cm of jejunum was left intact. At the time, no therapeutic option was given but parents demanded a second opinion so parenteral nutrition was commenced.

On day 11 of life the patient was transferred to our centre. She underwent a portoduodenostomy for biliary drainage. A jejunal stoma and a colonic mucous fistula were created. She was commenced on parenteral nutrition and slowly increasing enteral feeds. The jejunostomy effluent was re-fed into the distal colon. A system of intermittent clamping of the jejunal tube was introduced in an attempt to dilate the proximal small bowel for subsequent lengthening but this failed.

At 9 months of age a third surgical procedure was performed with reestablishment of intestinal continuity and creation of a gastrostomy. The duodenocolic anastomosis did not function well, and bilious fluid refluxed out of the gastrostomy. To maintain colonic health this fluid was refed into the colon via a colonic tube. A fourth operation at 10 months of age found a cirrhotic liver and it was not possible to improve the bowel anastomosis. Her liver function began to deteriorate and she was listed for transplantation.

She underwent liver small bowel transplant at 19 months of age. Induction immunosuppression was basiliximab followed by tacrolimus and prednisolone maintenance. Transplant course was uncomplicated. Enteral feeds via nasogastric tube were started one week post transplant and increased with reduction of parenteral nutrition. At discharge 28 days after transplant she was fully enterally fed with finger foods, top-up nasogastric tube feeds and a supplemental overnight feed. Her body weight was 12 kg. The serum albumin was 42 g/L, the white cell count was 6.51 x 10^9/L, and lymphocyte count was 2.79 x 10^9/L. Surveillance ileoscopy at 1 month after operation showed well preserved villi and crypt architecture. There has been one subsequent complication of abdominal wall abscess which was treated with antimicrobial therapy.

At nine months of post transplant follow-up, she remains clinically well. She has normal growth without nutritional support and has normal developmental milestones.

Conclusion

Portoduodenostomy in biliary atresia, splenic malformation with ultra-short gut can be performed for biliary drainage and does not preclude later successful liver small bowel transplantation.

H10

Cholestatic liver disease in children with Kabuki syndrome

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Background:

Kabuki syndrome (KS) is a rare disorder with multisystem anomalies and developmental delay. Mutations in the Myeloid Leukemia Lineage 2 gene (MLL2) underlie KS. The protein encoded by MLL2 is a histone methyltransferase that methylates the Lys-4 position of histone H3 and belongs to the ASCOM complex, a transcriptional regulator of the nuclear farnesoid X receptor (FXR), which regulates several genes involved in bile acid homeostasis. Individual reports describe a variety of cholestatic liver diseases (CLD) in KS with no genetic confirmation.

Aim and methods:

To describe hepatobiliary disease in KS and to corroborate its genetic association with MLL2 mutations, we reviewed our centre’s cohort of paediatric patients with CLD. We identified four individuals with KS. All had high serum B-glutamyltransferase (GGT) activity. Clinical, laboratory and demographic data were collected and MLL2 sequencing was undertaken.

Results:

Patients 1-3 were of European, Patient 4 of Indian origin; the parents of Patient 4 were consanguine. There was no family history of CLD or of KS in any. Each patient was heterozygous for a predictedly pathogenic MLL2 mutation. In Patient 1, disease manifested in late childhood as severe portal hypertension and idiopathic cholestasis. In Patients 2-4, cholangiopathy was present neonatally (see Table). Patient 4 had neonatal sclerosing cholangitis. Patients 2 and 3 had biliary atresia and underwent hepatic portoenterostomy, followed by liver transplantation at age 11 years in Patient 3. Patients 1, 2, and 4 have on-going CLD.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at presentation (years)</th>
<th>Portal tract expansion</th>
<th>Inflammation</th>
<th>Bile duct proliferation</th>
<th>Cholestasis</th>
<th>Bilirubin (mmol/L)</th>
<th>ALT (IU/L)</th>
<th>GGT (IU/L)</th>
<th>MLL2 Mutation</th>
<th>Alpha acid glycoprotein</th>
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</thead>
<tbody>
<tr>
<td>1 (F)</td>
<td>10 yrs</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>17</td>
<td>53</td>
<td>148</td>
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<td>2 (F)</td>
<td>3 wks</td>
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<td>+</td>
<td>-</td>
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<td>134</td>
<td>614</td>
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<td>+</td>
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<td>159</td>
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</tbody>
</table>

Conclusion:

This is the first report of CLD in genetically defined KS. One novel (truncating) and three known (1 splice site, 2 truncating) MLL2 mutations were identified in four patients with KS. Clinico-pathologic features did not suggest FXR dysregulation. In previous reports, early-onset cholangiopathy also has predominated among CLD of KS. Factors conducing to CLD in KS remain obscure.
Eosinophilic Oesophagitis Post Liver Transplantation.

R Taylor, S Rajasek, S Davison, P McClean, SV Karthik. Children’s Liver Unit Leads General Inflammatory.

Introduction

Eosinophilic oesophagitis (EOE) is an increasingly recognized condition that maybe associated with upper gastrointestinal tract symptoms and is characterized by dense eosinophilic infiltration of the squamous oesophageal epithelium or deeper oesophageal tissue. (1) There is a significant association between EoE and atopy. Both environmental and food-specific IgE is present in a significant proportion of patients with eosinophilic oesophagitis. EoE has also been described in patients following solid organ transplantation. (2)

Case Report.

A 6 yr old boy with Tyrosinaemia type 1 underwent Orthotopic liver transplant aged 14 months for severe liver impairment. One year post transplant, he presented with chronic intermittent bloody diarrhoea. He was investigated with upper GI endoscopy and colonoscopy that revealed a diffuse increase in chronic inflammatory cells with numerous eosinophils in both the upper and lower GI tract. It was postulated that this may be due to allergy and he was commenced on a milk and egg free diet with some improvement in his symptoms. He has had a persistently elevated plasma IgE level. Coeliac screen has persistently been positive but duodenal biopsies at this time showed normal crypt to villous ratios. An empiric trial of growth hormone had been tried earlier in view of short stature but was unfortunately unsuccessful.

He was re admitted 4 yrs later with hypoalbuminaemia and ascites. Upper GI endoscopy revealed oedematous white exudates from mid oesophagus to cardia. Histology suggested severe reflux oesophagitis and a possible evolving eosinophilic oesophagitis. Repeat endoscopy 6 months later again demonstrated white plaques. Histology was similar in the lower oesophagus to features seen before however the biopsies of the proximal oesophagus showed severe, haemorrhagic, fibrinous and eosinophilic necrosis, together with sheets of eosinophils. There was colonisation of Candida but no active mucosal invasion. Biopsies from the second part of the duodenum showed normal preserved villous architecture whereas duodenal bulb biopsies showed villous atrophy and significant lymphocytosis. No eosinophils were seen in the duodenal mucosa.

The current recommended pharmacological treatment for EoE is swallowed fluticasone administered via a meter dose inhaler or viscous budesonide. (1) In this case extreme caution needs to be taken prior to use of any steroid containing medication in view of his existing poor growth plus Candida colonisation of the oesophageal mucosa. The absence of food impaction, dysphagia or any other upper gastrointestinal symptom tilted the balance in favour of watchful expectancy as the risks of further exposure to steroids could potentially outweigh the benefits.

References


Chicken Pox in a Post Liver Transplant patient on standard immunosuppression with demonstrable varicella immunity

E. Griffiths1; G. Gupta2; P. Narula1; M. Patel1; P. Rao1; F. Shakley1 (‘Sheffield Children’s Hospital (SCH)’; 2 ‘Birmingham Children’s Hospital (BCH)’)

Introduction:

Varicella Zoster (VZV) infection can be a life-threatening condition in immunosuppressed children. Therefore VZV immunity status is commonly checked before starting immunosuppression and vaccination is offered if absent. If vaccination is not possible, the recommendations are for post-exposure immunoglobulin and intravenous (IV) acyclovir if the patient develops clinical signs of chicken pox. Development of chicken pox in the VZV immune child is rare. We present a case of chicken pox in an immunosuppressed child who had demonstrated VZV immunity.

Case presentation (methods):

A 5 year old, immunosuppressed girl presented with a macular rash 16 days after exposure to chicken pox. She had a background of gastroesphisis and short gut syndrome and had received 2 liver transplants aged 2 and 3 years respectively. She was on a standard regime of tacrolimus. Her mother had developed chicken pox and following this exposure the child was started on prophylactic aciclovir pending serology results. There was no clear history of previous VZV infection. Her serology came back positive for IgG antibodies for VZV but the rash appeared two weeks later. (Her mother still had active lesions at this point.) The rash was reviewed by our infectious diseases team who felt it was suspicious (although not typical) of early chicken pox infection. We increased the dose of her oral aciclovir to five times per day and sent skin and throat swabs. Her skin swabs were negative but VZV was isolated (by polymerase chain reaction (PCR)) on her throat swab. She completed 3 weeks of aciclovir and remained well throughout this period without further progression of her symptoms.

17 months later she presented again with a vesicular rash on her thigh and labia in a dermatomal distribution. This was felt to be herpes zoster (shingles) and she was commenced on IV aciclovir for 4 days, completing a course of high dose oral aciclovir when access was lost. This was confirmed as VZV on PCR.

Discussion:

There are two aspects to VZV immunity: humoral and cell-mediated immunity. It is possible that those who develop chicken pox despite demonstrable antibodies may have deficient cell-mediated immunity. It is also recognised that in the immunocompromised patient, the infection may represent reactivation rather than a newly acquired VZV. Given the clear association between exposure and symptoms in our case, reactivation is unlikely.

Conclusion:

There is little in the literature discussing recurrent chicken pox in the immunocompromised or suppressed child with humoral immunity. This case highlights the need for further research into varicella immunity in the immunosuppressed community and prevention management of this potentially fatal disease.
Inhibition of ileal Bile Acid Reabsorption with SC-435 Improves Liver Function in a Rat Model of Partial Bile Duct Ligation.

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Background
Elevated serum and hepatic bile acids play an important role in the development and progression of cholestatic liver disease. Total bile duct ligation (tBDL) has been extensively used to study cholestasis but this model completely obstructs bile flow into the intestine and therefore does not represent many cholestatic liver diseases including Alagille syndrome and progressive familial intrahepatic cholestasis (PFIC). It also prevents evaluation of blocking bile acid recycling from the intestine as a therapeutic approach to lower serum and hepatic bile acids and provide benefit in cholestatic diseases.

Aim
The aim of the present study was to develop a partial bile duct ligation (pBDL) model of cholestasis to better represent the human disease and to test the efficacy of a minimally-absorbed apical sodium-dependent bile acid transporter inhibitor (ASBTi), SC-435, to reduce serum bile acids (SBA) and reduce liver injury.

Methods
Harlan Sprague Dawley (HSD) rats were anesthetized with isoflurane, the common bile duct exposed by midline laparotomy and a short length of PE-10 tubing placed parallel to the bile duct. A ligature of 4-0 silk suture was tied tightly around the duct and tubing after which the tubing was removed resulting in constriction of the duct lumen without complete obstruction. At 3, 7 and 14 days after surgery, blood samples were taken for analysis of SBA and liver function parameters. Liver tissue was removed and flash-frozen at 14 days for histopathology evaluation.

Results:
Three days after pBDL surgery, SBA were increased by 29-fold. Serum liver alkaline phosphatase (ALP), aspartate aminotransferase (AST), alanine aminotransferase (ALT) and gamma-glutamyl transferase (GGT) were increased by >5-fold and serum total bilirubin levels by >100-fold. Seven and fourteen days after surgery, liver enzymes had begun to decrease but SBA, GGT and total bilirubin levels were still markedly elevated compared to the sham surgery group. Once daily oral gavage of SC-435 (10 mg/kg) caused dramatic reductions in SBA at both seven (52%) and fourteen (71%) days. ALP, GGT and total bilirubin were also significantly reduced at both time points with the decreases at day 14 being 75%, 65% and 67%, respectively, compared to the vehicle group (p < 0.05 for all parameters). The effect on reducing SBA was similar if dosing with SC-435 was started at days 0, 1, 2 or 3 after the pBDL surgery. Consistent with the improvement in liver function parameters, histopathology analysis of liver tissue showed SC-435 reduced necrosis, inflammatory cell infiltration and the number of proliferating cholangiocytes.

Conclusions:
The pBDL model results in significant increases in SBA and liver function parameters from 3 to 14 days after surgery that are characteristic of cholestasis and liver injury. This model is therefore more representative of the cholestatic liver injury observed in Alagille syndrome and PFIC. Blocking bile acid recycling with an ASBTi prevents dramatic increases in total SBA and liver biomarkers suggesting an ASBTi may provide a new therapeutic option for the treatment of cholestatic liver disease by decreasing the accumulation of toxic bile acids and reducing the severity of cholestatic liver injury.

Conflict of Interest
Vice President Research, Lumena Pharmaceuticals, Entrepreneur in Residence, Rivervest Venture Partners
Cohort Study of Childhood Obesity and Inflammatory Bowel Disease in UK
A McCorquodale, H Dogra, D Rawat, S Naik, Paediatric Gastroenterology, Barts and the London Children’s Hospital, Royal London Hospital Barts Health NHS Trust, UK.

Background
Large cohort epidemiological studies of childhood onset Inflammatory Bowel Disease (IBD) are limited. Those that exist generally relate to specific population cohorts most recently in China1. Prevalence of obesity and the proposed relationship to IBD has been described in American children2.

We cumulated data from a 10 year cohort of newly diagnosed paediatric IBD attending the paediatric gastroenterology unit at the Royal London Hospital. Our primary objective was to describe the prevalence of obesity at diagnosis and analyse any contributory characteristics specific to the culturally diverse referral population from North East/North Central London, Kent and Essex.

Methods
Paediatric IBD diagnoses over a 10 year period from January 1st 2001 to December 31st 2010 inclusive were obtained. All diagnoses made on or before a patient’s 18th birthday were included. Data was obtained via the hospital electronic records system, infloflex database or manually from written healthcare notes. BMI measurements were calculated using the standard formula and RCPCH UK BMI centile charts were used to stratify children into standardised weight categories.

Results
Over the 10 year sample period there were 400 IBD diagnoses; 60% male, 40% female. 244 (61%) were Crohn’s disease (CD), 127 (32%) were ulcerative colitis (UC) and 29 (7%) were indeterminate colitis. Age range at diagnosis was 1.6-18 years, median 13.2 years, mean 12.4 years, mode 15 years. Number of diagnoses per year increased over the study duration and this was statistically significant using the Mann-Kendall test (p<0.01).

Ethnicity - White Caucasian 57%, Bangladeshi 13% (reflecting immediate local population) Asian/Pakistani 15%, African/Caribbean 8%, European 5% and 3% mixed or unstated.

From n=400 accurate height and weight data within 1 month of diagnosis was available on 249 patients. 73% of patients had normal BMI, 6% obese and 4% as overweight (table 1). At diagnosis 3% of CD patients were obese compared to 11% with UC (p<0.05).

From the cohort of 249, 65% had follow up BMI data at 1 year (77% normal, 9% overweight and 8% obese) (table 2). Only 24% had 5 year data mostly due to discontinued height documentation after transition to adolescent clinic (79% normal, 11% overweight and 8% obese).

Table 1. BMI Classification at diagnosis

<table>
<thead>
<tr>
<th></th>
<th>Crohn’s Disease Number</th>
<th>Ulcerative Colitis Number</th>
<th>Indeterminate Colitis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Very thin</td>
<td>17</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Low BMI</td>
<td>2</td>
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<tr>
<td>Normal</td>
<td>104</td>
<td>61</td>
<td>16</td>
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<tr>
<td>Overweight</td>
<td>6</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Obese</td>
<td>3</td>
<td>6</td>
<td>1</td>
</tr>
<tr>
<td>Severely Obese</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Morbidly Obese</td>
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<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>151</td>
<td>80</td>
<td>18</td>
</tr>
</tbody>
</table>

Table 2. BMI Classification at 1 year

<table>
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<th>Crohn’s Disease Number</th>
<th>Ulcerative Colitis Number</th>
<th>Indeterminate Colitis</th>
</tr>
</thead>
<tbody>
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<td>Very thin</td>
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<td>Low BMI</td>
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</tr>
<tr>
<td>Normal</td>
<td>76</td>
<td>76</td>
<td>80</td>
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<tr>
<td>Overweight</td>
<td>11</td>
<td>11</td>
<td>4</td>
</tr>
<tr>
<td>Obese</td>
<td>6</td>
<td>9</td>
<td>10</td>
</tr>
<tr>
<td>Severely Obese</td>
<td>3</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Morbidly Obese</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>100</td>
<td>49</td>
<td>12</td>
</tr>
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</table>

Conclusion
Over 10 years of IBD diagnoses from a single centre in London the diagnostic yield increases with time. This supports a rise in IBD prevalence suggested by other published data. BMI results suggest a correlation between obesity and newly diagnosed UC prior to any treatment bias. This differs to the EPIC study in adults and warrants further investigation.

References:
Introduction:
Anaemia is common in children with inflammatory bowel disease (IBD) with a reported prevalence between 41% and 75%. Several mechanisms have been proposed for this, including decreased oral iron intake, impaired mucosal absorption, increased gastrointestinal tract loss and altered iron homeostasis due to the inflammatory response seen in IBD. Adult studies have shown quality of life can be independently improved by higher haemoglobin concentration, irrespective of disease activity.

Oral iron has traditionally been used in the treatment of iron-deficiency anaemia in children with inflammatory bowel disease. It has been proposed that oral iron may generate radical oxygen species that might provoke inflammation and exacerbate symptoms in these children. There are few studies demonstrating the safety and efficacy of intravenous iron in children as an alternative therapy.

Aim
The authors reviewed iron-deficiency anaemia in all children with inflammatory bowel disease, requiring intravenous iron in a tertiary children’s hospital over a 14-month period. The aim was to characterise its safety and efficacy in this group.

Methods
All children with IBD and iron-deficiency anaemia who were given intravenous iron between January 2012 and March 2013 were retrospectively identified and included. Iron-deficiency anaemia was confirmed through haemoglobin, ferritin, transferrin and serum iron concentrations. The response to treatment was assessed through repeat bloods at 4-6 weeks following infusion. Disease type, location and severity were identified through clinical records.

Children were given intravenous iron infusion (ferric carboxymaltose) as an infusion over 15-30 minutes. Any adverse reactions were recorded in clinical records. Symptom improvement was ascertained from clinical records and a retrospective patient survey.

Results
Between January 2011 and March 2013, 48 children with IBD were identified who received intravenous iron for the management of their iron-deficiency anaemia. 24 had a diagnosis of Crohn’s disease, 8 of ulcerative colitis and 16 had IBD-unclassified. 6 had disease involvement of the ileum, 27 had colonic involvement and 15 had both the ileal and colonic disease involvement.

There was a significant improvement in blood indices in children following intravenous iron infusion - mean haemoglobin rise of 2.62g/L (range: 0.3-5.7), mean ferritin rise of 72.4mg/L (range: 2.4-151.8) and mean serum iron rise of 9.7μg/L (range: 2.4-29).

No early or late adverse reactions were recorded.

Conclusions
Iron-deficiency anaemia is prevalent in children with a diagnosis of IBD. The use of intravenous iron in its management has not been previously characterised in children with inflammatory bowel disease. This study demonstrates that the use of intravenous iron is both safe and efficacious in the treatment of iron-deficiency anaemia in children with inflammatory bowel disease.

N3
Specialist Dietitian Care Pathway for Liquid Nutrition Therapy in Paediatric Crohn’s Disease
Norton, Haidee1; O’Shea, Caitriona1; Stanley, Ruth1; Bunn, Su2; 1Dietetic Department, GNCH, Newcastle upon Tyne. 2Paediatric Gastroenterology, GNCH, Newcastle upon Tyne

Background:
There are no national guidelines for the dietetic management of Liquid Nutrition Therapy (LNT) in paediatric Crohn’s disease (CD). Following the dedicated dietitian appointments in gastroenterology patient support for LNT has been delivered consistently and a care pathway was developed (2010). Care pathway includes admission for initiation of graduated feed introduction and intensive dietetic support whilst inpatient and outpatient. Feed volumes were minimised by realistic kcal aims and concentrating feed.

Aim:
To establish if the provision of dedicated dietetic support and the implementation of a care pathway for LNT in children improves compliance, clinical outcome and has a positive impact on patient experience.

Methods:
Data collection retrospective from dietetic notes 2008 - 2010, 2010-2012 prospective data collection in database.

Results:

<table>
<thead>
<tr>
<th></th>
<th>2008</th>
<th>2009</th>
<th>2010</th>
<th>2011</th>
<th>2012</th>
</tr>
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<tbody>
<tr>
<td>No. started on LNT</td>
<td>27</td>
<td>20</td>
<td>9</td>
<td>5</td>
<td>25</td>
</tr>
<tr>
<td>No. oral feed</td>
<td>11</td>
<td>10</td>
<td>8</td>
<td>3</td>
<td>18</td>
</tr>
<tr>
<td>No. oral + tube feed</td>
<td>4</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>No. tube feed</td>
<td>12</td>
<td>6</td>
<td>1</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>No. completed course</td>
<td>16</td>
<td>11</td>
<td>6</td>
<td>2</td>
<td>17</td>
</tr>
<tr>
<td>No. failed to respond</td>
<td>Unknown</td>
<td>6</td>
<td>1</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>No. failed to complete due to poor compliance</td>
<td>Unknown</td>
<td>3</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Disease specific feeds</td>
<td>2</td>
<td>5</td>
<td>6</td>
<td>5</td>
<td>24</td>
</tr>
<tr>
<td>Average weight gain (kg)</td>
<td>Unknown</td>
<td>3.4</td>
<td>5.4</td>
<td>6.7</td>
<td>2.7</td>
</tr>
</tbody>
</table>

Summary:
Prior to 2010 dietetic records were incomplete due to patients, having been initiated on LNT, not being followed up. Compliance improved to oral acceptance of LNT and in completing course. Weight gain was not sacrificed by reducing volumes to aid compliance. Patient experience and cost improved by reducing numbers reliant on full or partial tube feeding.

Conclusion:
We conclude that the support and intense input of dedicated paediatric dietitians and the use of a standardised care pathway for children commenced on LNT provided the IBD multidisciplinary team with a standardised approach to LNT and improved the delivery of this treatment.
The impact of a dedicated nutrition team on the outcome of paediatric home parental nutrition
Paediatric Nutrition Team, Great North Children's Hospital, Newcastle upon Tyne, NE1 4LP
Presenting author – Sarah Cunningham

Introduction
A multi disciplinary team is recommended to facilitate home parenteral nutrition (PN). Prior to April 2008 children in Newcastle were monitored on an adhoc basis. From April 2008 an appointment of a nutrition nurse to work with x 2 consultant gastroenterologists, paediatric surgeon, pharmacist and dietitian provided a specialist service for children requiring home PN

Aim
Our aim was provide home PN when necessary with progression to full enteral feeding whenever possible and to decrease the rate of central line infection aiming for a catheter related bloodstream infection (CRBSI) rate of <5 per 1000 catheter days.

Results
5 children needed home PN prior to April 2008. By April 2008, 2/5 remained on home PN. Of the other 3 children, 2 children had their PN discontinued and 1 child transferred to adult services. A total of 31 patients (20 short gut, 3 complex IBD, 8 other – including dysmotility and enteropathy) have needed home PN from April 2008-June 2013. 20 patients were discharged from hospital between 0-1 years of age, 3 patients 1-5 years of age and 8 patients 10-18 years of age. We currently have 12 children on home PN (9 short gut, 19 are no longer under our service; 15 went on to full enteral feeding, 2 were transitioned to adult services and remain on PN, 1 underwent small bowel and liver transplant and 1 died (not related to PN). None of the remaining home PN patients have developed chronic liver disease and all of these patients were discharged on Tauroloch and SMOM lipid.

The use of central venous catheters for home PN increased throughout July 2008 to July 2013, which included 16837 catheter days and 67 CRBSI. This is an overall CRBSI rate of 3.97 per 1000 catheter days.

• 5 patients have had no CRBSI
• 5 patients <5 CRBSI per 1000 catheter days
• 5 patients >5 CRBSI per 1000 catheter days (3 patients still on home PN and are on 70% alcohol locks)

Conclusion
The use of PN has increased, as shown by the number of home PN catheter days and we have been able to reduce the infection rate despite an increase in catheter usage. Chronic liver disease is an avoidable complication of patients requiring home PN. It is possible by using a multi-disciplinary approach, targeted strategies and protocols to manage a home PN service in children that has very low catheter infection rates, no patients with chronic liver disease and a good outcome with regards to progressing to full enteral feeding.

What Do Doctors Know? - Cow’s Milk Allergy and Lactose Intolerance
Walsh, Joanne: Castle Partnership, Norwich; Queckett, James: Director of Education at Doctors.net.uk

Introduction / Background
It is perceived that there is confusion about the symptomatology and management of cow’s milk allergy and lactose intolerance. With up to 7% of infants presenting with symptoms suggestive of cow’s milk allergy and the impact of managing them having a potential huge impact on NHS resources, it is imperative that these infants are managed efficiently and effectively, often entirely within primary care. (Ludman BMJ 2013). For this to happen, primary care clinicians require knowledge of the symptoms and their management.

Aim
By developing a series of online questions, we aimed to get insight into doctors’ interest in this area and their baseline knowledge. These insights were then used to develop online learning materials to improve any areas where knowledge was particularly poor.

Method
JW, with input from Doctors.net.uk, developed a quiz with an initial set of ten multiple choice questions (MCQs) to determine baseline knowledge and identify learning needs. After each question, the correct answers and an explanation were given. The results of the quiz were used to determine particular knowledge gaps.

A further module was then produced to test knowledge more specifically related to symptoms of lactose intolerance and those of cow’s milk allergy, and also to assess knowledge about the use of replacement formulas for use in cow’s milk allergy. Any doctor choosing to undertake the module performed a pre-module assessment which included a set of five MCQs before working through a series of case histories with related questions, answers and explanations. A post-module assessment with the same five MCQs was then completed. Baseline knowledge and knowledge improvement were then determined. The quiz and module were signposted specifically to general practitioners using the website but were available to any member of Doctors.net.uk.

Results
1317 doctors completed the initial quiz between July 2012 and October 2013.

One of the noticeable areas of poor knowledge was question 2, “Which of the following are true of lactose intolerance?”. Across all regions, less than 30% of doctors answered correctly that gastroesophageal reflux is not a symptom of lactose intolerance.

By question 9 of the quiz however, 70% correctly identified as “false” a similarly themed claim, “Gastroesophageal reflux is a symptom of lactose intolerance”.

The subsequent learning module was completed by 1,005 doctors (763 GPs, 99 paediatricians, 144 doctors in other specialties) between April and October 2013. The percentage of doctors who passed the pre-module assessment was 64%; this increased to 87% in the post-module assessment (pass mark 70%). Of interest in this module, only 33% correctly answered that constipation is listed by NICE (2011) as a symptom of food allergy. After completing the module, 71% of doctors were aware of this fact.

Summary and Conclusion
The initial quiz showed poor knowledge of the symptoms of lactose intolerance across all regions and supports the belief that there is confusion among doctors about the presentations of lactose intolerance and cow’s milk allergy. It identified that there were learning gaps and the subsequent learning module with pre-and post-assessment showed how that knowledge can be improved as there was an increased proportion of doctors answering the post-module assessment questions correctly.

The large number of doctors completing the module shows that it is an area of interest and possibly recognised as one where learning is needed. As well as many GPs completing the module, other specialties are obviously interested in education in this area. The answers to some questions highlighted poor knowledge and that although this approach to e-learning saw improvements in the level of knowledge, more education is still needed with regard to lactose intolerance and milk allergy.

Conflicts of Interest:
Dr Walsh has received honoraria for consultancy work from MJN and Nutricia. She has received funding from Doctors.net for her role in the production of educational material.

Doctors.net.uk as a commercial organisation has received an educational grant from Mead Johnson Nutrition, it has also received funds for promotion activities from both Mead Johnson Nutrition and Nutricia.
Congenital Sucrase-Isomaltase Deficiency A five patient case series.

John Puntis1, James Sawyer2; 1Department of Paediatric Gastroenterology, The Leeds General Infirmary, Great George Street, Leeds, West Yorkshire, LS1 3EX, UK; 2BS Orphan Ltd, Beam Heath Way, Nantwich, Cheshire, CW5 6PQ.

Congenital sucrase-isomaltase deficiency (CSID) is a rare genetic condition characterised by an absence or deficiency in the brush-border SI enzyme. Sufferers from the condition are therefore unable to metabolise certain di- and polysaccharides.

The diagnosis of CSID has hitherto required laboratory confirmation of reduced enzymatic activity, either from within small bowel biopsies or alternatively using faecal chromatography to demonstrate the presence of undigested sugars. A less invasive and practically simpler technique is required since current approaches are often only considered following unsuccessful programs of dietary restriction focused upon more common disorders of absorption. Dietary control is a valuable tool for the management of patients with CSID.

However, it is highly restrictive for patients and carers and difficult to exclude all sugars that may precipitate symptoms this may also increase the diagnostic challenge. Enzyme replacement therapy with sacrosidase (Sucrads®) to facilitate the breakdown of sucrose into simpler components for intestinal absorption provides an effective therapeutic approach to management.

We present five cases of children with CSID who received sacrosidase treatment at the Leeds General Infirmary. All of the patients experienced prolonged periods of illness and investigation prior to the diagnosis of CSID. Treatment with Sucrads® resulted in clinically significant improvements in gastrointestinal symptoms in all five patients. Furthermore parents reported additional benefits such as improvements in sleep, improved child happiness and increased self confidence, whilst some noting that missed doses were associated with symptoms recurrence. No significant adverse events were reported during long term therapy ranging from 5 to 15 years.

There are few case series of patients with CSID reported from within the UK, belying the reported prevalence of the condition from epidemiologic studies elsewhere. The availability of a non-invasive test for CSID and an effective treatment as experienced by the patients we report would be a significant advance in the management and diagnosis of this debilitating condition.

Genetic Testing of Congenital Sucrase-Isomaltase Deficiency

Stefan Maaser1, James Sawyer2; 1Cantogene AG, Schillingallee 68, 18057 Rostock, Germany; 2BS Orphan Ltd, Beam Heath Way, Nantwich, Cheshire, CW5 6PQ.

Congenital sucrose-isomaltase deficiency (CSID) is an autosomal recessive inherited disorder caused by mutations in the sucrase-isomaltase enzyme complex which is exclusively expressed in the small intestine microvillus membrane. Sucrase-isomaltase is the primary enzyme for degradation of dietary starch and sucrose, together representing over 90% of carbohydrates in the Western diet. Sucrase hydrolyzes alpha-1,2 and alpha-1,4 glucosidic bonds and isomaltase cleaves alpha-1,6 linkages.

It is estimated that CSID affects about 0.2% of individuals of European decent and about 5% of indigenous Greenlanders. The differential diagnosis of CSID includes a variety of disparate conditions including cystic fibrosis, coeliac disease, irritable bowel syndrome (IBS) and food allergy. Some studies have suggested that up to 5% of patients diagnosed with irritable bowel syndrome may actually have CSID.

Currently diagnosis requires a biopsy specimen for the determination of the sucrase-isomaltase activity, which represents a substantial hurdle for the appropriate management of patients. Consequently they may remain undiagnosed until adulthood, many patients report a long history of severe and disabling gastrointestinal problems, often accompanied by a significant healthcare consumption.

The underlying cause of the disease is mutations in the sucrase and or isomaltase coding region. In the former case enterocytes of patients lack the sucrase activity whereas in the latter enzymatic activity can vary from absent to normal. Mutant phenotypes vary in their posttranslational processing, cellular localization and function. This condition is generally inherited in an autosomal recessive pattern. Therefore most patients are homozygous for single mutations but also heterozygous mutations have been reported although this is atypical.

Genetic screening for CSID is an alternate route to diagnosis that can help avoid the time and costs associated with trials of dietary restriction followed by the invasive procedure of small bowel biopsy. In one study, 32/56 (57%) mutant alleles found amongst 31 patients diagnosed clinically with CSID were accounted for by one of four mutations, V577G, G1073D, R1124X and F1745C, the most commonly represented being G1073D (31%). Although not fully inclusive, testing for a panel of eleven following mutations, P348L, V577G, E613X, T694P, R774G, F875S, G1073D, Q1098P, R1124X, C1229Y, F1745C is expected to cover more than 80% of symptomatic CSID patients. Larger epidemiologic studies are required to confirm this estimate.

In case patients with clinical features suggesting a diagnosis of CSID, but for whom one of these mutations is not present, sequencing of the entire gene is a further alternative to biopsy. The gene is not prone to large deletions or insertions therefore additional genetic testing by MLPA or qPCR is not needed.

The inclusion of genetic testing for CSID should be considered in the triage of patients with recurrent diarrhoea of unknown cause. The optimum scheduling of testing within the management of such patients warrants inclusion in care algorithms.
Long-term Outcome of Intestinal Epithelial Cell Dysplasia Tufting Enteropathy

Aim:
To review the very long-term outcome of children with tufting enteropathy and age at which enteral autonomy is gained.

Subjects and methods:
Twenty children who had presented in infancy with watery diarrhoea and severe intestinal failure (IF) that required long-term PN treatment were reviewed. All cases had the histological appearance of enterocyte tufting on small intestinal +/- colonic biopsies. Age, sex, country of origin, dependence on PN, survival and age of weaning off PN were recorded.

Results:
Of the 20 patients 11 were male and 9 female. Ten patients were Maltese, 4 White British, 5 Arab and one Afghan. One patient died aged 2 years. The patients all presented with diarrhoea and severe faltering growth. They were all commenced on treatment with long-term PN. In 15 cases parents were trained and PN was continued at home. Five children remained on long-term hospital PN.

When reviewed the surviving 19 patients were aged from 3 – 27 (mean 13) years. Eight children were aged under 10 years (3–9 years), 6 children were aged 10 – 20 years and 5 over 20 years. One of the 8 children under 10 years of age had weaned off PN. Two of the 6 children aged 10-20 years had weaned off PN and 3 of the 5 patients over 20 years of age had done so.

All 13 children still on PN had some enteral intake as well. Six/13 children had 7 infusions/week, 3/13 had 6 infusions/week, 3 others had 5/week and one, 2 infusions/week. The two patients aged over 20 years still on PN were infusing 5 nights/week.

The six children (30%) who had gained enteral autonomy had done so when aged from 3-22 years. They are now aged from 10-24 years (mean 16.5 years). No child had needed to restart PN after stopping it.

Summary and Conclusion:
Children with tufting enteropathy have an increasing chance of weaning off PN with increasing age ranging from 2/20 or 10% chance of doing so before 10 years of age to 40% aged 10-20 years and 3/5 or 60% of those aged over 20 years. It is possible that an even greater number of children could wean with best possible medical care and psychological support. Given the good long-term outcome, intestinal transplant should be avoided if at all possible.

Table 1. Reported complications.

<table>
<thead>
<tr>
<th></th>
<th>Hardly ever (%)</th>
<th>Sometimes (%)</th>
<th>More than expected (%)</th>
<th>Frequently (%)</th>
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<td>Psychological problems</td>
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<td>12.5</td>
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<td>Deterioration in academic performance</td>
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<tr>
<td>Confusion about care outcomes</td>
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<td>35</td>
<td>12</td>
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</table>

We estimate about 50% paediatric IF centres, and the largest adult IF centres responded to the survey. The number of teenagers approaching transition is currently around 52 with a similar number of young adults (44) aged 20-30yrs who are currently receiving PN. The practices and processes of transition reported were highly variable. One third of respondents identified confusion around care routines and psychological problems at the time of transition.

Conclusions:
Better documentation and service standards relating to timing of initial stages of transition and employment of key worker should be developed. Consideration should be given to check lists for practical aspects (e.g. pumps) as well as psychology input to enhance emotional resilience of the young people and support for carers.

Current practices across the UK regarding the monitoring and supplementation of sodium in surgical infants and children

Danielle Petersen1; Vanessa Shaw1; Anne Payne2; 1Great Ormond Street Hospital; 2Plymouth University

Background:

Infants and children with excessive sodium losses, for example, premature infants, those with ileostomies or cystic fibrosis are at risk of sodium depletion which may result in metabolic acidosis and poor weight gain. Experts therefore suggest that urinary sodium (UNa) should be monitored and if it is low, sodium supplements should be started. However, there are no guidelines suggesting how this is best carried out and limited evidence is available within the literature. Practices widely differ within the UK and with a lack of consensus in the literature, the development of guidelines or further research is limited. This study therefore aims to describe current practices regarding the monitoring and supplementation of sodium in paediatric surgical centres across the UK, from which guidelines or further research could be developed.

Methods:

The study design was a cross-sectional descriptive survey. A questionnaire was developed and electronically sent to the Paediatric Surgical Dietitian in each of the 26 paediatric surgical centres across the UK. Total population sampling was used with one Paediatric Dietitian from each of these centres invited to partake in the survey. Completed questionnaires were returned, data analysed and the results of each of the 18 questions summarized and discussed. Where possible, the results from the survey were compared to the recommendations within the literature.

Results:

100% response rate was achieved. Results showed that all paediatric surgical centres across the UK monitored UNa and started sodium supplements if the UNa was found to be low. There was also consensus regarding the importance of sodium in relation to growth. All of these are consistent with the recommendations found in the literature. Other practices, however, varied more between centres and often between Consultants within the same centre. The two areas of practice which were found to be the most inconsistent between centres and to that suggested within the literature were: the method used to monitor UNa; and the reference values used to define a low UNa. These are two essential aspects regarding the monitoring and supplementation of sodium.

Conclusions:

There is limited literature and evidence regarding the monitoring and supplementation of sodium in surgical infants and children. Furthermore, the results from this survey show that even within the areas of practice where evidence is available, current practices are not always based on these recommendations and vary significantly across the UK.

Outcome of neonatal short bowel syndrome (SBS) requiring long-term PN: necrotising enterocolitis (NEC), volvulus/ataresia & gastrochisis compared.

Vanessa La Vela, Clinical Fellow, Susan Hill, Gastroenterology Consultant, Gastroenterology Dept, Great Ormond Street Hospital for Children, London WC1N 3JH

Introduction/Background:

There is limited information about the length of time that parenteral nutrition (PN) support may be required for neonates who undergo small intestinal resection for either intestinal necrosis due to volvulus or necrotising enterocolitis (NEC), or for intestinal strictures, atresia or gastrochisis.

Aim:

To review length of time on treatment with parenteral nutrition (PN) in infants presenting with intestinal failure (IF) according to underlying diagnosis.

Subjects and methods:

All patients attending an intestinal rehabilitation clinic who underwent intestinal resection for necrotising enterocolitis (NEC) or volvulus and/or atresia or gastrochisis who required long-term PN for >28 days were reviewed. Gestational age was recorded. Length of time on PN was calculated and compared in the two groups.

Results:

Eighteen infants (9 male: 9 female) were identified. 9 patients (3 male: 6 female) had NEC. Eight were born prematurely and one at term (range 23-40 weeks, mean 27, median 25 weeks). Three patients had intestinal volvulus (2 male: 1 female) and 4 atresia (2 male: 2 female) born from 27-35 weeks gestation (mean 31, median 30 weeks). Three other male infants had gastrochisis.

Small intestinal length was from 30-85cm (mean 45cm, median 40cm) at time of resection in 8 NEC patients (1 unknown), from 30-120cm (mean 63, median 60cm) in volvulus/ataresia patients and 22 and 50cm in 2 gastrochisis cases (1 unknown).

PN treatment was given for 3-42 months (mean 16, median 10 months) in NEC patients, 1-33 months (mean 14, median 8 months) in volvulus/ataresia patients and 22 and 144 months in gastrochisis patients. There was no significant correlation between remaining length of small intestine and time on PN (Pearson’s R was 0.21, p=0.59 for NEC; -0.01, p=0.99 for volvulus and -0.02,p=0.8 for atresia).

Summary and Conclusion:

There was no clear correlation between length of remaining small intestine and time on PN.

Children with SBS secondary to NEC, volvulus or atresia with a remaining small bowel length from 30-120 cm who require long-term PN are likely to continue to need PN for an average 15 months and up to 42 months whereas gastrochisis cases may need treatment for over 10 years.
**N12**

A pilot study on weaning practices amongst semi-urban community in Southwest England.

Dr Siba Prosad Paul1, Dr Megan Eaton2, Dr Manjunath K Sanjeevaiah2, Yeovil; Dr Rachel Debono2, Mrs Christine Routley2, Ms Julia Johnson2, Bristol Royal Hospital for Children, Bristol (previously at Yeovil District Hospital, Yeovil); 2Yeovil District Hospital, Yeovil;

**Background:**

The Department of Health, UK recommends that all children from 6-months to 5-years of age are given supplemental vitamin (A,C,D) drops and are weaned on to solids from 6 months. This pilot study assessed the actual practice followed in a semi-urban community in the Southwest of England.

**Methods:**

Fifty-three parents (children aged 6-months to 2-years) responded to a questionnaire survey given to them while they visited the paediatric services. The survey questionnaire was designed with 11 questions: timing of introduction of solids, food first used for weaning, awareness about national guidelines on weaning and vitamin supplements, advice received from community health practitioners (CHP) and reasons for introduction of early solids.

**Results:**

Age ranges of children participating in the survey were almost equally distributed. Exclusive breast feeding was reported only in 16/53 (30%) infants. Most children (40/53 i.e. 75%) were weaned aged 4–6 months, 1/53 aged <4-months and 7/34 (23%) aged >6-months. Hungry baby (47%) and advice from others (13%) were most common reasons cited for early weaning. Baby rice (77%) and vegetables (41%) were the most common weaning foods used by parents. 75% parents responded positively about advice received from CHP. Majority (85%) of parents responded correctly being aware of national guidelines regarding time of weaning. However, only 32% parents responded having been told about vitamin supplementation and only 20% parents actually administer it to their child.

**Conclusion:**

This pilot survey demonstrated that although majority of parents were aware of current UK guidelines regarding timing of weaning but chose not to adhere to it. Only 20% of parents actually gave vitamin supplementation to their children and this has been addressed with the CHP. There is plan to extend the survey to 200 respondents to get a better overview of the adherence to national guidelines.

**N13**

Central venous catheter related infection in children receiving home parenteral nutrition. A single centre experience

Mutalib, Mohamed; Victoria Evans; Anna Hughes; Donna Forbes-Penfold; Alice Spano; Susan Hill; Great Ormond Street Hospital NHS Foundation Trust

Intestinal failure in children is defined as a reduction of functional gut mass below the minimum amount necessary for adequate absorption of nutrients, fluids and electrolytes to maintain health and growth. Children with intestinal failure depend on parenteral nutrition for their calories and nutritional needs. Home parenteral nutrition (HPN) allows the transfer of care of long term PN patient to home which improves school attendance, sport participation and overall improvement in quality of life. However, long term administration of PN requires central venous access which can be hampered by catheter related complications such as infection, thrombosis, blockage and breakage.

**Methods**

We retrospectively reviewed case notes and electronic records of our cohort of patients on home PN between Nov 2012 and Nov 2013

We identified 36 patients. 19 males and 17 females. Their age range between 1.1 years to 18.2 years with the average age of 7.3 years. They were on PN for a period of 52 weeks with a combined 13140 days of PN. There were 55 episodes of infections of which 29 were catheter related blood stream infection, the remaining 26 episodes were urine, skin and GI fluids infections without bacteremia or sepsis. Of the 29 catheter related blood stream infections, 15 episodes of infections occurred in two patients only with underlying severe immune dysregulation. Including those two patients, our central line infection per 1000 PN days is 2.2. However excluding those two patients will reduce the incidence to 1.1 central line infection per 1000 PN days. Organisms isolated were: staph epidermidis in 20.7%, Staph aureus and Kelbsella in 13.8%, pseudomonas in 10.3%, candida in 10.3%, streptococcus 6.9%, Ecoli and Kocunia Rhizophilia in 3.4% while Stenotrophomonas was isolated in 17.2% (all the stenotrophomonas infections were isolated from single patient).

**Conclusion**

Catheter related blood stream infection CRBSI is one of the commonest complications of long term PN patients. While removal of the infected catheter can ultimately clear the source of infection, frequent line changes can lead to venous thrombosis and occlusion compromising access sites and increasing overall patient morbidity. Lack of central venous access is considered one of the indications of small bowel transplant independent of underlying disease severity. Prompt recognition and early antibiotics therapy can save central venous catheter and reduce complications.
Maddison, Karen; Ferry, Sarah; Embleton, Nick; Thomas, Julian: Royal Victoria Infirmary, Newcastle

Background
The Special Care Baby Unit uses standard formulations and guidelines for parenteral nutrition (PN) in pre-term neonates. The guidelines are designed so that the baby should start PN on day one of life (including lipid) and build to full energy and fluid requirements by day three; 150ml/kg/day, 3g/kg/day protein and 100kcal/kg/day. Due to other complications, many of the pre-term infants do not reach full fluid requirements from PN. For example, they may be fluid restricted or have concurrent drug infusions. The practice on the unit is also to reduce glucose concentration of PN if hyperglycaemia develops.

Aim
To investigate the actual energy provision from PN by looking retrospectively at fluid balance charts and calculating energy provided.

Method
Data was collected for twenty consecutive babies on PN. The energy provision from glucose, protein and lipid was calculated over each 24 hour period. This was compared to the ideal energy provision that the standard formulations should provide and percentages were calculated. When enteral nutrition (EN) was initiated, the energy provision from EN was subtracted from 100kcal/kg/day to allow for the new PN target.

Results
On average, 9 days were spent on PN (range 4-28). Overall, 74% of the energy provision intended from standard PN was achieved.

Discussion
The standard PN regime provides a good level of energy provision overall, but in the days after birth the energy provision is less than achieved from day 4 onwards.

Day 1 energy provision appears to be very low, but this can be accounted for by the 24 hour period of data collection. Initiation times are obviously different throughout day 1 depending on when baby is born.

The energy provision varied greatly for some babies, as illustrated by the error bars on the graph. This was generally caused by the reduction in glucose in the PN when the baby was hyperglycaemic. The glucose is reduced to 5% or 7.5% concentration in the PN.

The possibility of having a central line only standard bag which aims to give 120kcal/kg/day and 4g protein/kg/day should be explored so the nutrition energy can be optimised for babies with adequate access.

Health-related quality of life in children on home parenteral nutrition: parent and child perspectives
Protima Amor, Katie Knight, Kate Blakely, Jami Khair, Rachal Bowman, Protima Amon, Katie Knight, Kate Blakely, Jami Khair, Rachel Bowman, Minal Patel, Nigel Meadows, Sandhia Naik; Department of Paediatric Gastroenterology, The Royal London Hospital

Background:
Health-related quality of life (HRQoL) is increasingly recognised as important to reflect the impact of an illness and its treatment on a patient from the patient's perspective. However, there may be times when it is difficult to obtain this information directly from paediatric patients, and parents are therefore used as substitutes. However, the perceptions of parents can vary from the self assessment by the patient with the latter often reporting a better quality of life. Survival of children with intestinal failure (IF) has increased in the past decade; however, data on their HRQoL are lacking. As in other chronic diseases children with intestinal failure are at risk of depression, anxiety, altered self image and social isolation which can all have a negative impact on their HRQoL.

Aim:
The aim of this study was to evaluate the HRQoL among children with intestinal failure who are on home parenteral nutrition (HPN) as assessed by parents and the children themselves and consider the impact of the condition on affected families.

Subjects and Methods:
Seventeen out of eighteen children on HPN were surveyed for parent and child responses using The Paediatric Quality of Life Inventory (PedsQL) version 4.0. One child was excluded from the study as the foster carer was very recently allocated.

In addition to the questionnaires, parents were asked 5 open-ended questions to address more specifically the effects of HPN on children and their families and assess the suitability of the PedsQL to evaluate their and their child’s HRQoL.

Results:
The 23-item (21 items for ages 2-4 years) PedsQL 4.0 Generic Core Scales (physical, emotional, social and school functioning) were completed by 13 out of the 16 families (N.B. one family has two children on HPN). Items are scored on a scale from 0 to 100. Higher scores indicate better HRQoL.

Seven children aged 5-16 years self-reported their HRQoL. The majority of parents (67%) said the PedsQL failed to address specific effects of HPN on children and their families.

Summary:
There were no statistically significant differences in HRQoL based on parent proxy report and child self report. Summary score findings suggest that parents have concerns about their child’s HRQoL, particularly with respect to physical and school functioning.

Conclusion:
Our findings demonstrate there can be similarities between child and parent perspectives as measured by the PedsQL questionnaire. A disease specific quality of life survey taking into account different age groups as well as both child and parent views would be useful in better determining the HRQoL of children with IF and their parents.

### Table: HRQoL Scores

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<th>Parameter</th>
<th>Parent Mean (SD)</th>
<th>Child Mean (SD)</th>
<th>Mean Difference</th>
<th>p value</th>
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<td>44.4 (34.2)</td>
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<td>Social Functioning</td>
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<td>Emotional Functioning</td>
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<td>50 (21.1)</td>
<td>44.2 (23.3)</td>
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</table>
Improved nutrient provision with naso jejunal feeds in extremely low birth weight infants
Gandhi Rashmi; Tavener Kate; Ient Georgina; Hickey Ann; King’s College Hospital NHS Foundation Trust

Introduction/Background
Achieving adequate growth with enteral feeds in extremely low birth weight (ELBW) infants remains challenging. Those with significant respiratory morbidity are often nutritionally compromised. Lower oesophageal sphincter immaturity and aspiration of milk feeds may result in reduced enteral and thus caloric intake. Parenteral nutrition (PN) administration with its associated complications may be prolonged in this group.

We describe our practice of aiming to achieve adequate enteral caloric intake and growth by introducing naso-jejunal (NJ) feeding in this high risk group of patients.

Patients and methods
9 ELBW infants received total or partial NJ feeding between January 2011 – March 2013. Median gestation was 24(24 - 28) weeks. Median birth weight was 705(550 – 960)g. Median Birth Z score was 0.49(-1.74 - 0.62). All infants had significant respiratory disease and remained ventilated at the time of commencement of NJ feeding.

Results
All infants were fed maternal expressed breast milk (MEBM). This was commenced at a median of 5(1-9) days. Full nasogastric feeds were established at a median of 20(13-33) days. Median nasogastric volume occurred was 150(135-180)ml/kg/day. Median nasogastric caloric intake was 104(93-140)kcal/kg/day. Median nasogastric protein intake was 2.3(2 - 4.1)g/kg. In one infant breast milk fortifier (BMF) was introduced.

NJ feeds were commenced at a median of 40(19-97) days. Median weight at this point was 1.19 (0.75-1.49)kg; Z-score; -1.89 (-3.57 - 0.54). Documented reasons for commencement of NJ feeds were gastro-oesophageal reflux in 7 and recurrent aspiration in 2 infants. All infants had radiological evidence of broncho-pulmonary dysplasia at the time of commencement of NJ feeds.

NJ feeds were commenced at a median of 40(19-97) days. Median weight at this point was 1.19 (0.75-1.49)kg; Z-score; -1.89 (-3.57 - 0.54). Documented reasons for commencement of NJ feeds were gastro-oesophageal reflux in 7 and recurrent aspiration in 2 infants. All infants had radiological evidence of broncho-pulmonary dysplasia at the time of commencement of NJ feeds.

Median NJ volume achieved was 180(160-180)ml/kg/day. Median NJ caloric intake was 135(110- 140)kcal/kg/day and protein intake was 3.6(2.4-4.2)g/kg. Three additional infants had BMF introduced and tolerated, one infant had peptiprotein (50%) introduced and the remainder were fed MEBM. No infant received supplementary PN during NJ feeding period.

The median number of days on NJ feeds was 33(10-58). Minimal NG feeds were continued in all infants during NJ feeding. No complications of NJ feeds were noted. All infants gradually re-established oral and NG feeding. Feeding assessments performed suggested safe swallow in 8 infants, but 2 remained NG feed dependent at the time of discharge due to disorganised and inefficient suck pattern.

Conclusion
The introduction of NJ feeds in this high risk population facilitated an improved caloric and protein intake and avoided the reintroduction on PN. ESPGHAN guidelines (2010) on nutritional requirements in premature infants suggest an intake of 110-135 kcal/kg/d and 3.5-4.5 g protein/kg/d which was achieved on NJ but not on NG feeds in our group.

Improvement in gastrointestinal symptoms on an exclusion diet.
Shergill-Bonner Rita, Katherine, Great Ormond Street, London

Introduction
Multiple food exclusion diets are used as treatment for non IgE mediated food allergies, to obtain relief from a vast range of gastrointestinal symptoms. The empirical exclusion involves eliminating milk, egg, wheat and soya from the diet for a period of 6-8 weeks, but the number of foods excluded and length of time for the dietary exclusion may vary according to the clinician.

The evidence to support which particular foods to exclude and the length of time to exclude them for is lacking.

Background
To carry out an audit to evaluate the efficacy of the exclusion diet, change in symptoms and time taken for symptom improvement to occur. The combination of foods excluded may also influence symptom improvement and this was recorded for all patients.

Methods
40 children were selected from gastrointestinal outpatient clinics, who had never trialled an exclusion diet before. Symptoms on initial assessment were documented and scored from 1-5 for severity (5 being the most severe). The patients were reviewed by telephone at 2, 4, 6 and 8 weeks after their initial appointment and were asked to recall symptoms and severity.

Results
2 patients were excluded from the audit. 1 had previously been on an exclusion diet and 1 returned to a normal diet during the audit period. In total 58% of children reported an improvement in symptoms, with decreased severity scoring. Of those that improved 36% were symptoms free at the end of eight weeks. The majority of patients showed improvement between weeks 6-8, 10/22 patients reported an improvement in this time (39%).

The most popular exclusion combination was milk, egg, wheat and soya with 13 of the children being placed on this exclusion and out of the 13, 7 improved. Those with more severe symptoms initially, were put on medications as well as the diet. There was little difference in improvement between those on medications and those not on medications. 12/22 of those children that improved were on the exclusion diet alone.

Conclusion
Dietary exclusions can lead to improvement in gastrointestinal non-IgE mediated symptoms. Accurate diagnostic assessments are essential to avoid burdening individuals and families with exclusion diets for long periods of time. To reduce the time taken to identify offending food groups all four food groups should be excluded initially followed by structured reintroduction. The most significant improvement was seen between 6-8 weeks which confirms the length of time the child should be kept on the diet before an improvement may be seen. Further improvement may have been seen between 8-10 weeks, further research and the time period of future audits should to be extended to determine this.
Nutritional adequacy of PWS children on restricted diets. A 10 year review from an MDT clinic.

Method

PWS children attending our specialist MDT clinic were asked to complete weighed 3 day food diaries as part of their assessments. Data was analysed against UK reference nutrient intakes (RNIs) adjusted for age and sex. Analysis was completed using a specific computer package with data input completed by 1 of 2 trained dietitians. Results were expressed as % of intake against RNIs.

Results

35 food diaries were returned from 11 PWS children (age range 1.1-12 years) 5 males and 6 females between August 2002 - March 2013. Mean caloric intake was 69.5% RNI (Range 38-102%). A mean lower caloric % of RNI was seen in the younger children (<5 years, n=20) 66.8% compared to the older children (6-12 years n=15) 73.1%. There was no obvious difference in the intake between males and females (68.8% vs. 70.8%). The mean proportions of % energy from fat, carbohydrate and protein (compared to UK recommendations) were 25% (35%), 58.8% (50%) and 18.8% (15%) respectively. 4 patients contributing a total of 10 food diaries over the collection period showed <20% total energy from fat. A Spearman correlation co efficient showed a significant (r=-0.57) inverse trend for greater micronutrient deficiencies with greater caloric restrictions. Incidence of <100% RNI for calcium and iron were 20% and 31% respectively appearing to have no relation to age or sex.

Conclusion

Children in our specialist PWS clinic have on average 30% less caloric intake compared to RNIs. The contribution proportions of macronutrients identified some concern of over restriction of fat (<20%) associated with increased risk for inadequate intake of PUFA. A reasonably strong correlation between caloric restriction and micronutrients intakes below the RNI should be noted and all results highlight care is required to carefully balance the vulnerable restricted diets of PWS children.
A survey of UK clinical experiences of grid trainees in PGHAN
Dr L Whyte on behalf of CSAC

Introduction:
The PGHAN CSAC are required to monitor and improve standards of training and work with trainers and other stakeholders to do this. The CSAC were concerned that trainees in PGHAN across the UK have different training experiences.

Aim:
To document training opportunities during March - September 2013 of PGHAN Grid trainees in the UK and to highlight if there might be major discrepancies between training centres.

Methods:
Survey designed by CSAC and sent to all PGHAN Grid trainees September 2013. The centres and trainees remained anonymous. The endoscopy questions were based on recommendations from the Joint Advisory Group (JAG) regarding the electronic assessment system known as JETS.

Results:
There are 17 grid trainees in post currently, 4 are working out of programme and so were excluded from the survey. 9 out of the remaining 13 surveys were returned.

<table>
<thead>
<tr>
<th>Question</th>
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<tr>
<td>Do you use JETS?</td>
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<td>Are trainers logged on as assessors on JETS?</td>
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</tr>
<tr>
<td>Number of procedures/6m</td>
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<td></td>
</tr>
<tr>
<td>Number of lists/week</td>
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<td>Number of lists attended/wk</td>
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<td>Number of clinics scheduled/ wk for SpR to attend</td>
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<td>Number of clinics attended/ wk</td>
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</table>

The majority of trainees use the JETS system for assessment of endoscopy training. Formal teaching is provided once a week for all the trainees surveyed, but this is not bleep free. Trainees did not cover a general paediatric service in normal working hours achieving the standard that the majority of day time training is spent in the speciality. Trainees were required to provide out of hours cover with a range of 6-12 weekend nights and 8-16 weekday nights in 6 months.

Discussion:
PGHAN is a small speciality and so the survey was limited by the low numbers of trainees currently in post; individual training centres were not identified and not all training centres were included. Opportunities for clinical experience across the UK in the speciality appear to vary. Teaching and endoscopy opportunities may be limited due to time commitment to in-patient care. JETS is used to record endoscopy training but has not yet used by all trainees. The trainee that did not undertake colonoscopies was working in hepatology at the time. Recommendation: – It is the responsibility of each trainee, educational supervisor and grid coordinator to work together to target specific aspects of training on an individual basis in order to facilitate achievement of competencies. This may require targeted training, e.g. organisation of time to attend specialist clinics and provision of bleep free dedicated endoscopy training lists and teaching sessions.
Adenovirus gastroenteritis can kill... a severe case of a common problem.

Background

Viral gastroenteritis is a common killer worldwide. Rotavirus is the most common in children under 5yr. Other known viruses are Human astrovirus, calcivirus, enteric adenovirus, corona, toco, picobirnavirus, aichi etc.1

Adenovirus related diarrhea accounts for 20% of viral gastroenteritis worldwide and is known to cause protracted diarrhea.

We present a unique case of Adenovirus gastroenteritis in an infant with severe form of the illness. His journey from initial resuscitation at presentation to being discharged home highlights important clinical problems of intussusception, subacute intestinal obstruction, food aversion and failure to thrive which brings out the challenges arising from a perceived common illness.

Aim

To present a case of Adenoviral gastroenteritis with severe illness and highlight the complications arising from it.

Methods

Clinical presentation

A 9-month-old infant presents with 2 days history of profuse diarrhea with blood in stools and vomiting and needs fluid resuscitation due to significant hypovolemia. Though Ultrasound abdomen and contrast study do not suggest intussusception but exploratory laparotomy shows bruising at terminal ileum to suggest a spontaneously reduced intussusception.

On going diarrhea with passage of blood and intestinal mucosal slough establishes intestinal failure and need for TPN for 6 weeks.

Over the next 6 months of hospital stay he has had intermittent episodes of abdominal pain and vomiting and needs second laparotomy and excision of small bowel adhesions and removal of a foreign body (iv drip set clamp).

With improvement of symptoms, enteral feeds were introduced and progressed gradually via nasogastric feeding initially and then orally.

Since being discharged home, he has had further episodes of diarrhea and vomiting to suggest postadenoviral enteritis.

At present he is overcoming aversion to solids, secondary to his illness, but is achieving good catch up growth.

Investigation Results

1 Abdominal X-ray- suggestive of proximal high-grade small bowel obstruction
2 US abdomen- distended thick walled small bowel loops, no intussusception
3 Electron microscopy of biopsy- viral gastro-entero-colonopathy
4 Faecal virology- Positive for Adenovirus, difficult to serotype
5 NPAb- positive for Rhino and Adenovirus
6 Pediatric Immune deficiency panel- Low total IgG but subsets normal.

Conclusion and Discussion

Adenovirus type 40 and 41 of subgenus F are responsible for enteric gastroenteritis.2

Presentation varies from asymptomatic carriage to acute and protracted gastroenteritis. There is evidence to report complications in the form of mesenteric adenitis and intussusception and related mortality.3 Enteric adenovirus though difficult to grow in culture media, can be isolated better by highly sensitive and specific monoclonal antibody based Elisa methods.4

Management is mainly supportive. Antiviral treatment is used for adenovirus related enteritis in immunocompromised children but high quality evidence to support this is lacking.

References:


Acute watery diarrhoea continues to cause significant morbidity and mortality in infants and young children both in developing and developed countries despite advances in oral rehydration therapy, which is the mainstay of treatment. Racecadotril is an antisecretory agent that selectively inhibits intestinal enkephalinase and therefore can prevent fluid/electrolyte depletion from the bowel as a result of acute diarrhoea, without affecting intestinal motility. There have been previously published reviews of the literature that included some meta-analyses, but these were limited by a lack of consideration of adverse events and methodological issues. An up-to-date systematic review using the Cochrane Collaboration format is therefore indicated to summarise the current evidence on the use of Racecadotril for the treatment of acute diarrhoea in children.

Methods

Randomised controlled trials comparing racecadotril to placebo or other interventions in children with acute diarrhoea, as defined by the primary studies, were included. Data sources were MEDLINE, EMBASE, Cochrane Central Register of Controlled Trials, reference lists of retrieved articles and drug company contacts. When inadequate data was presented, authors were contacted. Data extraction and assessment of methodological quality were performed independently by two reviewers. Methodological quality was assessed using the Cochrane risk of bias tool. Data was analysed according to the intention to treat principle.

Results

7 Randomised controlled trials were included, 5 comparing racecadotril with placebo or no intervention, with 1 having inpatient and outpatient populations that were analysed separately. There was 1 study comparing racecadotril with pectin / caolin and 1 with loperamide. Moderate to high risk of bias was present in all studies. No significant difference in efficacy or adverse events was found in 1 study comparing racecadotril to loperamide. Meta-analysis of 4 studies with 806 participants showed significantly shorter duration of symptoms in the racecadotril group compared to placebo or no intervention (SMD -6.30 hours, 95% CI -6.52 to -6.08). Meta-analysis of 2 studies with 405 participants showed significantly less stool output in the first 48 hours of treatment in the racecadotril group compared to placebo or no intervention (SMD -63.30 hours, 95% CI -65.23 to -61.36). Meta-analysis of 2 studies with 405 participants showed significantly less stool output in the first 48 hours of treatment in the racecadotril group compared to placebo or no intervention (SMD -63.30 hours, 95% CI -65.23 to -61.36). Meta-analysis of 2 studies with 402 participants showed significantly shorter duration of symptoms in the racecadotril group compared to placebo or no intervention (SMD -6.30 hours, 95% CI -6.52 to -6.08). Meta-analysis of 5 studies with 949 participants showed no significant difference in adverse events between racecadotril and placebo or no intervention (OR 0.99, 95% CI 0.68 to 1.43).

Conclusions

There is some evidence from this review that racecadotril is more effective than placebo or no intervention in reducing the duration of illness and stool output in children with acute diarrhoea. However, the overall quality of the evidence and therefore strength of this conclusion is limited due to sparse data, heterogeneity and risk of bias in the studies. Racecadotril appears safe and well tolerated when compared to placebo and loperamide.
Shwachman-Diamond syndrome (SDS) is a rare genetic disorder characterized by exocrine pancreatic insufficiency, bone marrow dysfunction and skeletal abnormalities. This study is an overview of clinical experience of SDS in a National Clinic in the UK.

Methods

44 children identified from the Hospital’s National SDS Clinic database. 13 excluded because of alternative diagnoses or incomplete data. 31 patients (15 male) with a clinical and/or genetic diagnosis of SDS were included. Their clinical symptoms, psychological and genetic characteristics were analysed from initial presentation until early adulthood or death.

Results

All 31 patients had genetic screening for the disease; 11/31 were compound heterozygotes for the common SBDS mutation (c.183_184TA>CT and C258+2T>C). Less common genetic profiles were identified in 13 patients.

8 patients had mild or transient neutropenia with no recurrent or serious infections. 19 had moderate to severe neutropenia and of those, 11 had transient or persistent pancytopenia with 2 progressing to bone marrow failure and bone marrow transplantation.

The majority of patients required long-term pancreatic enzyme and vitamin/trace element supplementation due to severe pancreatic insufficiency. 5 patients became pancreatic sufficient at follow up, including one with mild pancreatic lipomatosis on imaging. Transient transaminitis with absent/mild radiological features but no clinical implications was seen in 63%. A non specific enteropathy was identified endoscopically in 22%, with no clear correlation with pancreatic or genetic status.

Growth issues were a common denominator either due to feeding difficulties or skeletal dysplasia, the latter seen in 48%. No true endocrinopathies were identified. 21 patients had significant dental involvement.

18 patients had identifiable learning and behavioural difficulties, ranging from mild speech delay to severe hyperactivity and depression.

Congenital abnormalities such as cleft palate, congenital hydrocephalus, and congenital heart disease were seen in a minority. Renal manifestations included neonatal nephrocalcinosis, hyperoxaluria and late onset proteinuria.

Conclusions

Phenotype and genotype descriptions and potential correlations in SDS from the largest cohort in Europe. Better understanding of spectrum of SDS manifestations allowing timely diagnosis and contributing to optimal management of this rare condition.
Gut and Mesenteric Lymph Node Involvement in HIV Infection pediatric patients
Cecilia Mantegazza MD 1, Giovanni Maconi MD 2, Vania Giacomet MD 1, Federica Furfaro MD 2, Chiara Mameli MD 1, Cristina Bezzio MD 2, Vania Giacomet MD 1, Gian Vincenzo Zuccotti MD 1; 1 Department of Paediatrics, L. Sacco University Hospital, Milan, Italy; 2 Gastroenterology Unit, L. Sacco University Hospital, Milan, Italy.

Introduction:
The gastrointestinal tract is a primary target for Human Immunodeficiency Virus (HIV). HIV infection causes a depletion of CD4+ T-lymphocytes in the gut-associated lymphoid tissue and affects gastrointestinal mucosal integrity and permeability. The gastrointestinal tract has also been suggested as the main reservoir of HIV despite highly active antiretroviral therapy (HAART).

Aim:
We performed a prospective case-control study to assess gut involvement in HIV-infected patients, either naïve or on HAART, using noninvasive methods such as bowel ultrasound and fecal calprotectin.

Subjects and methods:
Thirty HIV-infected children and youth underwent the following tests: CD4+ T-cell count and HIV viral load, fecal calprotectin and bowel ultrasound; the latter evaluated bowel wall thickness and mesenteric lymph nodes. Fecal calprotectin and bowel ultrasound were also assessed in thirty healthy controls matched for age and sex. In particular, faecal calprotectin was measured through a quantitative immunochromatographic point-of-care test (Buhlmann laboratories AG, Schonenbuch, Switzerland) and concentrations ranging from 0 to 200 µg/g were considered to be normal reference values in children.

Results:
Fecal calprotectin was normal in 29 HIV-infected patients and was not statistically different from controls (respectively mean value 63.8 ± 42.5 µg/g and 68.3 ± 40.5 µg/g; p: 0.419); calprotectin did not correlate with HIV viral load, CD4+ T-cell absolute count and percentage and HAART treatment. No significant changes on bowel ultrasound were found except for enlarged mesenteric lymph nodes, which were observed in 7 HIV-infected patients (23.3%) and 2 controls (6.6%). This last data significantly correlated with high HIV viral load (p: 0.001) and low CD4+ T-cell percentage (p: 0.004).

Summary and conclusion:
HIV-infected children did not have significant biochemical or ultrasonographic signs of mucosal inflammation. Few patients showed enlarged mesenteric lymph nodes, which correlated with uncontrolled HIV infection.

The prevalence of morbidity in older children who have undergone neonatal repair of congenital abdominal wall defects.
Jackson, RE (Principal Investigator); Parikh, DH (Consultant Paediatric Surgeon, Birmingham Children’s Hospital, Clinical Supervisor); Plunkett, A (Consultant Paediatric Intensivist, Clinical Supervisor); Turner, J (The university of Sheffield, Academic Supervisor).

Introduction:
Existing research into morbidity following repair of Congenital Abdominal Wall defect has focused on the first two years of life. Morbidity outcomes after this age are poorly described in the medical literature.

Aims:
To describe the prevalence of the following morbidities in a cohort of children aged 2-16 years, following neonatal repair of a Congenital Abdominal Wall defect: Gastro-oesophageal reflux disease; Chronic constipation; Vomiting; Failure to thrive; Faecal and urinary incontinence.

To describe the quality of life in this cohort, using a validated paediatric quality of life tool.

Methods:
Study design: observational, retrospective cohort study. Subjects: patients aged 2-16 years of age, with a history of surgical repair of Gastroschisis or Exomphalos.

Patients were identified from two sources:
1. The surgical database of a tertiary children’s hospital in the UK;
2. An online support forum Gastrobaby and Avery’s Angels that allowed me to put a post on their websites.

Presence of morbidities and quality of life score were assessed by parental questionnaire.

Results:
We sent out a total of 202 questionnaires; 183 were patients cared for at Birmingham Children’s Hospital; 19 were parents from Gastrobaby or Avery’s Angels who responded to our online post. We received 58 responses. A high prevalence of all measured morbidities was reported: Vomiting (3 times or more per week) – 22%; Chronic constipation – 26%; Urinary incontinence – 22%; Faecal incontinence - 29%; Under 10th percentile for weight - 26%; Under 10th percentile for height - 31%.

We also used the PEDsQL for gastrointestinal symptoms, to measure quality of life in this cohort. We found 35% of respondents scored under 600 score indicating that their quality of life was adversely affected by the presence of gastrointestinal symptoms.

Conclusion:
In this small cohort, the prevalence of morbidities amongst older children following repair of congenital abdominal wall defect is high, and quality of life scores are poor. The results demonstrate that further research is needed to describe this prevalence more precisely using larger cohorts.
O8

Use of ATM BridleTM to secure nasal feeding tubes in children.

Hanza Marzouk, Clinical Observer, Great North Children’s Hospital, Newcastle upon Tyne
Sarah Cunningham, Paediatric Gastroenterology Nurse Specialist, Great North Children’s Hospital, Newcastle upon Tyne; David Derry, Paediatric Gastroenterology Nurse Specialist, Great North Children’s Hospital, Newcastle upon Tyne; Su Bunn, Consultant Paediatric Gastroenterologist, Great North Children’s Hospital, Newcastle upon Tyne

Background:
The ATM BridleTM was developed to secure nasal feeding tubes in confused / agitated adults. Tape is passed via one nostril, around the vomer bone and out the other nostril. The two ends of tape are then clipped to the nasal feeding tube to prevent inadvertent removal. It has clear application in paediatrics where children, due to young age or neurodisability, frequently remove nasal feeding tubes.

Aim:
To review the early experience in one paediatric centre of the use of ATM BridleTM to secure nasal feeding tubes.

Methods:
Retrospective case note review of nasal bridles inserted between September 2012 and November 2013

Results:
10 children (8 boys) had 16 nasal bridles inserted for feeding tubes, 3 between Sept 2012 – March 2013 and 13 between April – Nov 2013. One boy also had a pH probe secured for 48 hours using a bridle. The median age of first ATM BridleTM insertion for nasal feeding tube was 9.5 months (range 2-169). The bridle was used to secure 3 nasogastric tubes (NG) and 13 nasojejunal tubes (NJ). NG / NJ placement and bridle insertion were performed under GA with endoscopy in 11 (NG=1, NJ=10), with xray screening in 2 (NJ=2) and on ward in 3 (NG=2, NJ=1). Sizes of nasal feeding tube and ATM BridleTM were 8 French in 9 children and 6 French in one. 6/10 children had history of frequent nasal tube removals prior to bridle insertion. Underlying conditions were as preterm 2, cardiac surgery 3, neurodisability 2, renal disease 2, food allergy 3, GI dismotility 1. Indicators for tube feeding included vomiting in 10 (100%), faltering growth in 9 (90%) and low intake due to feed aversion in 4 (40%). The median length of time a single bridle and nasal feeding tube was in place was 32 days (range 5 – 144). Three children required more than 1 bridle due to i) long term NJ feeding requirement - NJ and bridle electively changed 4 times each after approximately 90 days; ii) NJ tube pulled out despite bridle after 5 days, probably due to clip not being closed completely – NJ and bridle replaced; iii) NJ blocked after 45 days - NJ and bridle replaced.

Presently 4 children still have a NJ and bridle, one is awaiting fundoplication and gastrostomy; one is awaiting percutaneous endoscopic gastrostomy (PEG) with trans-gastrostomy jejunal extension (PEJ) insertion. Of the 6 remaining children who no longer need the bridle, 1/6 have been transitioned from NJ to NG feeding, 2/6 have a PEG, 2/6 (food allergy with feed aversion & severe transient gastroparesis) no longer require tube feeding and are eating orally and 1/6 is on total parenteral nutrition for severe gut dysmotility and attempts at jejunal feeding have been aborted.

There have been no significant complications. One child (as above) removed his NJ despite bridle after 5 days and required repassage. One family asked for bridle to be removed after 56 days as they felt that nasal tape was uncomfortable and child would no longer pull out a NG tube secured without bridle.

Summary:
The ATM BridleTM has proven safe and effective in securing short – medium term nasal feeding tubes in children aged from 2 – 169 months. It has proved especially useful in securing of NJ feeding tubes and has allowed long term naso-jejunal feeding. Experience of insertion and confidence in its effectiveness has seen an increase in applications, most markedly in children with cardiac and renal conditions where NJ feeding is required for growth but vomiting will often resolve with management of their underlying disease.

Conclusions:
The ATM BridleTM is a useful tool in paediatric enteral nutrition and we would recommend it to other centres. Our experience has shown that, unlike in adults, the bridle must be passed prior to the nasal feeding tube as young children’s nostrils are too small for feeding tube and ATM BridleTM introducer. Additionally, placing a bridle on a non-cooperative, moving child is challenging and experience should be gained initially in anaesthetised children. However, whilst initially we placed most bridles under GA, the last 4 bridles in 3 children have been placed quickly and successfully in non-anaesthetised children.

O9

Superior Mesenteric Artery Syndrome in a Patient with an Eating Disorder

Dr Neil Briscoe, Dr Anna Pigott, Dr Mary Jones University Hospital of North Staffordshire

Background:
Superior mesenteric artery syndrome (SMA syndrome) is a rare but life threatening complication associated with children and adults who have had recent weight loss, and as such, can occur in patients with eating disorders. Early diagnosis and treatment is essential for good clinical outcomes. We present a case of a 14 year old girl with an eating disorder who developed SMA syndrome, the causative factor being an intentional 10kg weight loss in the preceding three weeks prior to admission. We discuss the investigation and treatment of SMA syndrome, as well as the difficulty in making an early diagnosis. We further discuss if the use of the junior MARSIPAN guidelines in this case could have prevented the missed diagnosis which occurred at first presentation of this patient to the paediatric unit.

Method:
Ultrasound imaging of the aorto-mesenteric angle showed a narrowing to 20 degrees. An upper GI series was subsequently performed which showed a filling defect in the third part of the duodenum.

Result:
The clinical presentation and imaging findings in this case were consistent with SMA syndrome, and allowed for reasonably prompt treatment with total parenteral nutrition, and enteral feeding.

Conclusion:
Clinician knowledge and application of the junior MARSIPAN risk assessment framework tool in this case, would have resulted in earlier admission and treatment for this patient.

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Group meetings
(May be subject to last minute day and room changes)

Thursday morning:
Research – Mortimer Suite
Tower 1 – Education
Xibar – Endoscopy
Nutrition – Xibar

Friday morning
PoGHAN – Tower 1
IBD nurses – Neville
Gastroenterology – Mortimer
e-BANS – Xibar

Poster walk judges:

Wednesday 29th January 2014:
Team 1 – Posters G1 to G12:
Dr Mike Cosgrove
Dr Lucy Howarth
Dr Hemant Bhavsar
Team 2 – Posters G13, G14, N15, N20 to N23, O16 to O19
Dr Ieuan Davies
Dr Ed Giles
Ms Tracey Johnson

Thursday 30th January 2014
Team 1 – Posters G1 to G4, H5 to H12
Dr Jenny Epstein
Dr Fiona Cameron
Ms Sarah Tizzard
Team 2 – Posters H13 to H17, N17, N18, N19, N22, N23, O20 and O21
Dr Indra van Mourik
Dr Nadeem Afzal
Dr Anna Pigott

Friday 31st January 2014
Team 1 – Posters G10 to G10, N11 and N12
Dr Sue Protheroe
Dr Paul Henderson
Ms Kay Crook
Team 2 – Posters O13 to O15, N16 to N23
Dr Rajeev Gupta
Dr Anthony Wiskin
Mr Mick Cullen

NB. Plan not to scale and may be subject to changes
Exhibitors:
Norgine
BioHit
GBUK

Charities:
Children’s Liver Disease Foundation
Orphan Europe
CICRA
Crohn’s and Colitis UK

Brochure produced by
Louise Dilloway

and printed by
Prestige Print & Design, Birmingham