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NHS
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THE PEN

Paediatric Education Newsletter

A VOMITY BABY...

An 8-week-old, full-term male infant presents with a dropping of weight centile from 91st to less than 0.04th centile. (BW 4178g -> 3460g). He has a history of vomiting which is present after each feed which is non-bilious but forceful. He was initially diagnosed as having reflux and was started on an anti-reflux formula which mum felt had helped somewhat. He was later tried on Neocate for cows milk protein intolerance. He is currently having 145ml/kg/day.

What is the differential diagnosis? What investigations will you do? What is the treatment of your top differential? (Answers on page 3)

Tummy Troubles Issue

A CASE OF CHRONIC ABDOMINAL PAINS

A 14 year old girl presented with:

- 18 month history of intermittent lower abdominal pain
- Passing type 5-6 stools x 2/day.
- No blood/mucous PR
- No fever/vomiting

Background:

- Iron deficiency anaemia - lowest Hb 69g/l - ?? secondary to menorrhagia.
- Previous faecal calprotectin: 920ug//g
- Previous normal coeliac screen and TFTs.

Current admission:

- CRP 161mg/l, ESR of 67mm, WCC 12.6g/l, Albumin 29g/l.
- Normal AXR + abdominal USS.
- Discussed with gastro, commenced on IV cefuroxime and Metronidazole.
- Blood and stool cultures were taken, Calprotectin repeated -> 2670ug/g.
- Transferred to Sheffield Children's Hospital for diagnostic endoscopy.
- Macroscopy: Inflammation with extensive strictures.
- Microscopy: Transmural inflammation, lymphoid infiltrates, and granuloma.

What is the likely diagnosis? Answers on page (3)

UPPER GI BLEEDING

A 5 year old girl presents with a history of 5 large coffee ground vomits which started this morning. There is no history of temperature or intercurrent illness. She has had her bowels opened today and there was no diarrhoea or melena. She has a background of hydrocephalus with VP shunt and bilateral cerebral palsy involving all 4 limbs with GMFCS level 5.

When you assess her she has a patent airway, sats are 96% in air with normal RR. She is peripherally cool and has a capillary refill time of 3-4 seconds. Her bp is stable but she is tachycardic at 160bpm. She is alert and responding in her normal way to mum. Her abdomen feels soft. Her bloods show Hb 123g/l (was 144g/l yesterday), with normal white cells and platelets. Her U+E/LFT and clotting are normal, as is her gas.

Initial management:

ABCDE approach: 15L oxygen, 2 x large bore cannulas, baseline bloods inc. FBC, U+E, LFT, clotting + cross match

Fluid resuscitation: treat shock with fluid bolus - aim systolic age x 2 + 70mmHg (not too high so you don't interrupt clot formation).

Blood products if shock + large Hb drop/ongoing bleeding/derranged clotting/low plts (give RBC, cryoprecipitate or plts)

NBM, NGT, IV fluids, calculate severity, consider medications below, transfer for therapeutic endoscopy if indicated

Sheffiled Scoring System for Upper GI Bleed

History:

Significant pre-existing condition: 1

Presence of melaena: 1

Large amount of hematemesis: 1

Clinical assessment:

Heart rate > 20 above mean for age: 1

Prolonged capillary refill time: 4

Laboratory findings:

Hb drop of >20g/l: 3

Management/resuscitation:

Need for fluid bolus: 3

Need for blood transfusion (Hb <80g/l): 6

Need for other blood products: 4

Total score: 24

Cut of for endoscopic intervention: 8

Octreotide: 5mcg/kg over 30 mins then 3-5mcg/kg/hr

Vitamin K: 300mcg/kg IV (max 10mg)

IV omeprazole 2mg/kg OD
OR ranitidine 200mcg/kg/hr or 1mg/kg 6 hourly IV (if >1month)

Sucralfate oral/NG
- **1 month – 2 years: 250mg QDS**
- **2 yrs – 12 yrs: 500mg QDS**
- **>12 yrs: 1 gram QDS**

INFANTILE HYPERTROPHIC PYLORIC STENOSIS

Hypertrophy of the pyloric sphincter results in narrowing of the pyloric canal.
It is the most common gastric outlet obstruction in the 2-12 weeks old age group.

Risk factors of pyloric stenosis

1) First-born male infant
2) Family history of pyloric stenosis
3) Prematurity
4) Early exposure to erythromycin
5) Bottle feeding

Investigations

1) Test feed
2) Capillary blood gas (CBG): hyponatraemic, hypokalaemic , hypochloraemic metabolic alkalosis (May be normal)
3) Ultrasound Abdomen: diagnostic if muscle thickness >4mm and muscle length >14mm in term baby)

Case scenario - Approach to a vomity baby

Pyloric Stenosis

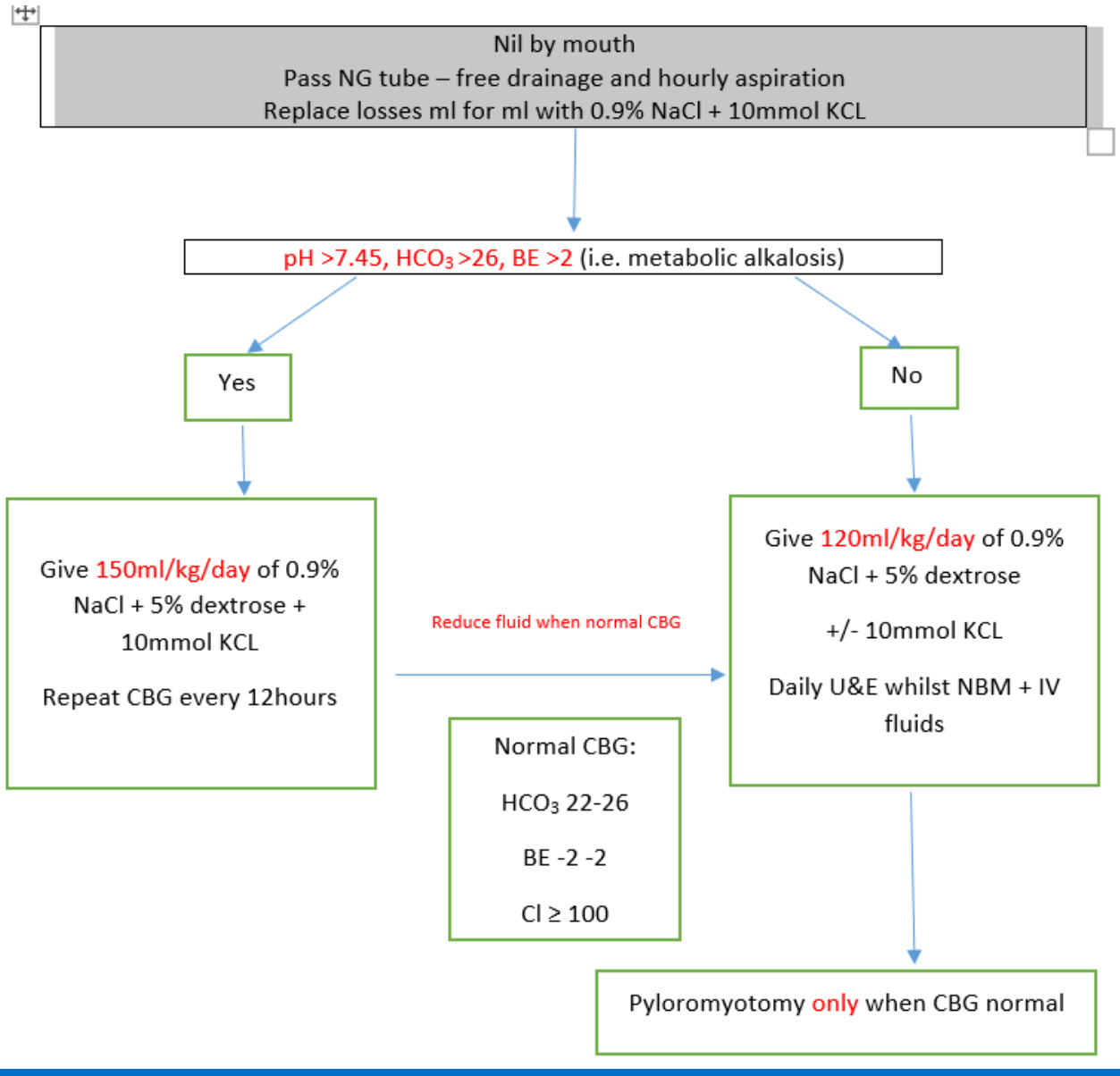
In this case, the baby had normal baseline bloods with a blood gas that was essentially normal with pH 7.45. However, his USS revealed an overly distended stomach. There was no movement through the pylorus throughout the scan. The pylorus measured length 20mm, width 13mm, muscle thickness 5mm : Suggestive of pyloric stenosis.

Differentials : CMPA, GERD

Investigations

Faltering growth' baseline bloods: FBC, U+E, LFT, TFT, bone profile
Blood gas
USS abdomen

MANAGEMENT



CROHN'S DISEASE

In the case scenario, she has Chron's disease. Here are the ways to distinguish between the two IBD presentations

	Crohn's disease	Ulcerative colitis
Most common site	Terminal Ileum	Rectum
Distribution	Mouth to anus	Rectum->colon, 'backwash' ileitis
Spread	Discontinuous, 'skip lesions'	Continuous
Gross features	- Focal aphthous ulcers with intervening normal mucosa - Linear fissures - Cobblestone appearance - Thickened bowel wall	- Extensive ulceration - Pseudopolyps
Microscopy	Noncaseating granuloma	Crypt abscesses
Inflammation	Transmural	Limited to mucosa and submucosa
Complications	- Strictures - Obstruction - Fistulas - Sinus tracts - Abscesses	Toxic megacolon
Genetic association		HLA B27
Cancer risk	1-3%	5-25%
Extraintestinal manifestations	Common e.g. arthritis, primary sclerosing cholangitis, erythema nodosum, pyoderma gangrenosum, uveitis	

Assessing Severity

Paediatric Ulcerative Colitis Activity Index

Item	Points
1. Abdominal pain:	
No pain	0
Pain can be ignored	5
Pain cannot be ignored	10
2. Rectal bleeding	
None	0
Small amount only, in less than 50% of stools	10
Small amount with most stools	20
Large amount (>50% of the stool content)	30
3. Stool consistency of most stools	
Formed	0
Partially formed	5
Completely unformed	10
4. Number of stools per 24 hours	
0–2	0
3–5	5
6–8	10
>8	15
5. Nocturnal stools (any episode causing wakening)	
No	0
Yes	10
6. Activity level	
No limitation of activity	0
Occasional limitation of activity	5
Severe restricted activity	10
Sum of PUCAI (0–85)	

<10 normal , 10-34 mild
35-64 moderate.
65-85 severe

MANAGEMENT

Mild to Moderate disease

First-line

- Exclusive enteral nutrition (EEN).
  This uses a complete liquid formula as the sole source of food for 6-8 weeks. Several studies have shown there is no statistically significant difference between induction of remission between EEN and steroids.

Second line

- Prednisolone for a minimum of 4 weeks, and then tapered. Care has to be taken regarding the risk of immunosuppression.

High risk for severe disease

First-line

- Induction is commenced with anti-TNF agents ie Infliximab.

Maintenance therapy

- Methotrexate, azathioprine, and 6-mercaptopurine.
- If infliximab is used as a maintenance treatment it should be given with methotrexate or azathioprine because this reduces the risk of developing antibodies.
- Patients with strictures and fistulae may need surgical management.

ESPGHAN review

Can Calprotectin differentiate between IBD and other gut conditions?

No. It has excellent sensitivity but low specificity. Calprotectin has a role in the regulation of inflammatory reactions, found in tissues & fluids - particularly within neutrophils. It is superior to any blood marker for the diagnosis of GI inflammation meaning it can distinguish inflammatory bowel disease (IBD) from other non-inflammatory gut conditions and inflammation outside the GI tract.

Clinical intervention should be based not only on calprotectin levels but on the clinical features, as younger children have naturally raised calprotectin levels in the absence of inflammation. The recommended cut off is >50mcg/g, but normal children can have faecal calprotectin levels > 100mcg/g. Therefore endoscopy is diagnostic when there is strong suspicion of IBD and should not be delayed for calprotectin results.

EXTRA RESOURCES AND JOURNAL ARTICLES

BMJ Best Practice: Pyloric stenosis

Best practice with the approach to pyloric stenosis, differential, investigations and management. Highlights the postulated causes of pyloric stenosis from hyperacidity as a result of antral distention with feeding and hypertrophy of the pylorus from repeated contraction, poor pyloric muscle neuronal innervation, nitric oxide synthase deficiency as well as lack of intestinal-pacemaker cells of Cajal.

Azithromycin in early infancy and pyloric stenosis

A retrospective cohort study that reviews the association between exposure to oral azithromycin and erythromycin and subsequent development of pyloric stenosis. The babies involved were exposed to these antibiotics in the first 3 months of life, between the years 2001 and 2012. Results showed an increased risk of pyloric stenosis to newborns exposed within the first 14days of life for both azithromycin and erythromycin, with an adjusted odds ratio [aOR], 8.26; 95% confidence interval [CI], 2.62-26.0 for azithromycin; while erythromycin had an aOR of 13.3 (95% CI, 6.80-25.9). The association was strongest during the first 2 weeks but persists to a lesser degree between 2 and 6 weeks of age.

The association of prenatal and postnatal macrolide exposure with subsequent development of infantile hypertrophic pyloric stenosis: a systematic review and meta-analysis

A systematic review with meta-analysis of the association between macrolides use, mainly erythromycin and subsequent occurrence of infantile hypertrophic pyloric stenosis (IHPS). 14 studies (12 retrospective cohort studies and two case-control studies) that investigated, the association between perinatal exposure to macrolides and pyloric stenosis were included. Seven cohort studies indicated that postnatal exposure had a statistically significant association, While for prenatal exposure there were 6 cohort studies and 2 case-control studies, the meta-analysis demonstrated a statistically significant association in the cohort studies but not in the case-control studies. Overall, the results demonstrated good evidence of an association between development of IHPS and direct postnatal exposure to macrolides but the evidence for prenatal and postnatal maternal exposure via breastfeeding was inconclusive.

Faecal calprotectin testing for differentiating amongst inflammatory and non-inflammatory bowel diseases: systematic review and economic evaluation.

In this systematic review which included 11 studies with a total of 1,284 adults and children with either inflammatory or non-inflammatory bowel disease, the sensitivity and specificity of calprotectin (with a cut off of 50mcg/g) for distinguishing between the two was 99% (95% CI 95%-100%) and 74% (95% CI 59%-86%) respectively.

Noninvasive Tests for Inflammatory Bowel Disease: A Meta-analysis.

This systematic review found that the pooled diagnostic performance of faecal calprotectin for detection of inflammatory bowel disease in an analysis of 10 studies with 867 patients showed a sensitivity of 99% (95% CI 92%-100%) and specificity of 65% (95% CI 54%-74%)

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