Don't Forget The Bubbles

LOWER GI BLEEDING & IBD

Facilitators Guide

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Topic: **Lower GI Bleeding & IBD**
Author: **Thomas Conway**
Duration: **up to 2 hours**
Facilitator level **ST4+/ANP equivalent+**
Learners level **Most useful for those experience seeing paediatric patients regularly: FY1+, band 5+ nurses.**
Equipment required: **None**

OUTLINE

- **Basics (10 Minutes)**
- **Main session (2x 15 Minutes) case discussions covering the key points and evidence**
- **Advanced session(2 x 20 minutes) case discussions covering grey areas, diagnostic dilemmas; advanced management and escalation**
- **Sim scenario (30-60 mins)**
- **Quiz questions (10 Minutes)**
- **Infographic sharing (5 Minutes): 5 take home learning points**

We also recommend printing/sharing a copy of your local guideline for sharing admission criteria.

PRE-READING FOR LEARNERS

Expectation is for the learners to have watched or read one of the basic pathophysiology links before the session.

PEM Playbook "GI Bleeding in Children"

https://dontforgetthebubbles.com/how-to-work-up-a-patient-with-bloody-diarrhoea/

IBD in Children - Podcast (39mins 54secs)

In My Shoes App - experience Crohn's and Colits (**Apple store** | **Google play**)

BASICS (DOES NOT NEED TO BE INCLUDED BUT FOR INFORMATION IF IT COMES UP IN DISCUSSION)

Common IBD and GI bleeding presentations:

Inflammatory Bowel Disease (IBD) is a term that describes Crohn's Disease and Ulcerative Colitis. These are both chronic relapsing conditions.

Ulcerative Colitis (UC) is diffuse mucosal inflammation involving colon and rectum. It usually presents with abdominal pains, bloody diarrhoea and weight loss. Nocturnal stools are common. Systemic effects can include fatigue, and anaemia. These appear alongside a raised CRP and ESR. There can be extraintestinal symptoms such as growth failure, erythema nodosum, uveitis and iritis although these are less common at presentation. Complications of UC can include toxic megacolon (a surgical emergency) and colon cancer. Where steroids are used as part of treatment, growth failure and osteoporosis can develop. Incidence in children range from 1:25,000 to 1:50,000 and it is more common in females than males. There are genetic associations with DR2, HLA-B27 and HLA-Bw35. A useful objective symptoms monitoring tool is the PUCAI (Paediatric Ulcerative Colitis Activity Index) score as shown below:

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 Remission	
	10-34 Mild Ulcerative Colitis	
	35-64 Moderate Ulcerative Colitis	
	65-85 Severe Ulcerative Colitis	

Crohn's Disease (CD) affects any part of the gastrointestinal tract from mouth to anus. It is twice as common as ulcerative colitis. Its presentation is similar to UC with gradual onset abdominal pain, diarrhoea (that can contain both blood and mucus), weight loss and poor growth. Unlike UC, presentation can involve perianal inflammation and aphthous mouth ulceration.

More than half the cases present with ileocolitis and the incidence is 1:25,000 with equal distribution between male and female. The mainstay of remission induction treatment is with exclusive enteral nutrition (EEN) using a protein based liquid feed - usually Modulen. This feed contains vitamins and minerals as well as fats, medium chain triglyceride oils and is casein based. As well as providing an easily absorbable method of nutrition, it has been shown to promote gut mucosal healing. It is used exclusively for 6 weeks before normal foods are slowly reintroduced. A useful objective symptoms monitoring tool is the PCDAI (Paediatric Crohn's Disease Activity Index) score - similar to the one used in ulcerative colitis. It is used to objectively measure disease activity in relation to therapeutic interventions. It can be useful to establish a baseline score so that when treatment is started the progress can be monitored and it may inform therapeutic decisions.

Item	Points
1. Abdominal Pain:	
No pain	0
Mild - brief, does not interfere with activities	10
Moderate/Severe - daily, longer lasting, affects activities, nocturnal	20
2. Stools per day:	
0-1 liquid stools, no blood	0
≤ 2 semi formed with small blood or 2-5 liquid	7.5
≥6 liquid, gross blood or nocturnal diarrhoea	15
3. General function:	
No limitation of activities	0
Occasional difficulty with activities	10
Frequent limitation of activities	20
4. Weight:	
Weight gain (or voluntary stable/reduction)	0
Weight loss <10% (or involuntary stable)	5
Weight loss ≥10%	10

5.	**Height velocity:**	
	≤ -1 standard deviation from normal	
	-1 - < -2 standard deviation from normal	
	≥ -2 standard deviations from normal	
6.	**Abdominal examination:**	
	No tenderness or mass	0
	Tenderness, or mass without tenderness	5
	Tenderness, involuntary guarding, definite mass	10
7.	**Peri-anal disease:**	
	None, asymptomatic tags	0
	1-2 indolent fistula, scant drainage, non-tender	5
	Active fistula, drainage,tenderness or abscess	10
8.	**Extra-intestinal:**	
	None	
	1 manifestation	
	≥2 manifestation	
9.	**Haematocrit %:**	
	>33 if <10yrs / ≥34 if female 11-19yrs / ≥35 if male 11-14yrs / ≥37 if male 15-19yrs	0
	28-32 if <10yrs / 29-33 if female 11-19yrs / 30-34 if male 11-14yrs / 32-36 if male 15-19yrs	2.5
	<28 if <10yrs / <29 if female 11-19yrs / <30 if male 11-14yrs / <32 if male 15-19yrs	10
10.	**Erythrocyte sedimentation rate:**	
	<20mm/hr	0
	20-50mm/hr	7.5
	>50mm/hr	10
11.	**Albumin:**	
	≥35g/dl	0
	31 - 34g/dl	10
	≤30	20
10.	**Sum of PCDAI (0-150)**	
	<10 Remission	
	10 - 27.5 Mild Crohn's Disease	
	30 - 37.5 Moderate Crohn's Disease	
	≥40 Severe Crohn's Disease	

Juvenile Polyposis Syndrome is another possible cause of lower GI bleeding and can occur in 1% of preschool and school age children. Its presence is characterised by multiple benign growths usually in the colon. It can present with painless rectal bleeding and can be associated with or without stool. Abdominal pain and pain around defecation can be a feature. It is autosomal dominant and can be diagnosed on endoscopy. The presence of more than 5 polyps in the colon or at least one polyp and a positive family history can confirm the diagnosis. In generalised juvenile polyposis the incidence of gastric, colon, duodenal and pancreatic cancer is increased. Diagnosis and treatment can happen simultaneously with endoscopy and polypectomy.

CASE 1 (15 MINS): A NEW DIAGNOSIS OF CROHN'S DISEASE

An 8 year old boy presents to A&E with severe generalised abdominal pains. This has been going on for about 4 months. He also says he has "very runny poos and they are a bit sticky and sometimes have some blood!"

What other questions would be relevant?

Although PCDAI (Paediatric Crohn's Disease Activity Index) is specific to Crohn's, if there is no histological diagnosis then it is advisable to use the PUCAI (Paediatric Ulcerative Colitis Activity Index) score even if the history suggests Crohn's. This still gives a reasonable objective idea about disease severity.

- Questions can be based on PUCAI, including:
- Abdominal pain
- Rectal bleeding
- Stool consistency
- Number of stools per 24 hours
- Nocturnal stools
- Impact on daily activities

What relevant investigations could you do now?

- Bloods to include FBC, specifically to investigate haematocrit, ESR, U&E to assess electrolytes and LFT to assess albumin

Do you think he needs admitting?

This decision should be made on the basis of the severity of the symptoms and the length of the history in addition to the inflammatory markers and electrolytes on the bloods. If the inflammatory markers are high and consistent with severe symptoms then admission for Gastroenterology review and urgent endoscopy is advisable. Additionally if there are electrolyte disturbances with changes such as hypoalbuminaemia then admission is also advisable. These patients are more likely to be nutritionally deplete and at risk of more serious complications requiring more serious interventions. If the history is short, the symptoms do not seem to be affecting everyday life and the inflammatory markers are not particularly elevated then the patient may be able to be discharged and then reviewed at a later date in a paediatric gastroenterology outpatient clinic with a view to elective admission for endoscopy.

IV antibiotics

The aetiology of Crohn's Disease is unknown but immunological response to gut microbiota is thought to play a role. Antibiotics have therefore been used to help aid induction of remission and to prevent relapse by reducing concentration of certain bacteria and altering the composition of the microbiota. Ciprofloxacin and metronidazole are the most commonly used antibiotics for this and can be started on the advice of the Paediatric Gastroenterologist.

ECCO Guidelines for New Crohn's Disease

Below is the link to the European Crohn's & Colitis Organisation (ECCO) guidelines for Crohn's disease. This website has a comprehensive collection of informative guidelines for IBD and is a reliable evidence based source of information.
http://www.e-guide.ecco-ibd.eu/algorithm/new-presentation-1

CASE 2 (15 MINS): JUVENILE POLYPOSIS SYNDROME

A 6 year old girl presents with painless fresh blood PR. She is otherwise completely well. **What could this be and what management options would you consider?**

Discussion can be based around the paper from the European Society of Gastroenterology, Hepatology and Nutrition's **"Management of Juvenile Polyposis Syndrome in Children and Adolescents: A Position Paper From the ESPGHAN Polyposis Working Group".**

Summary of paper

Juvenile polyposis syndrome (JPS) is a pre-cancerous condition and carries a 38%-68% risk of GI malignancy. It is diagnosed based on the following three criteria;

1. Five or more JPs of the colon or rectum, or
2. JPs in other parts of the GI tract, or
3. Any number of JPs and a positive family history

In infants it can be diagnosed clinically with symptoms comprising diarrhoea, failure to thrive and hypoalbuminaemia. The most common type of polyp in children are solitary juvenile polyps comprising 90% of cases and high grade dysplasia is rarely encountered.

Although genetics are inconsistent, the SMAD4 gene is associated with more aggressive disease and can be associated with Hereditary Haemorrhagic Telangiectasia (HHT).

JPS is autosomal dominant and although no formal guidelines exist it is thought that children who are at risk of JPS should be screened from the ages of 12 to 15 years although children who are symptomatic (explained anaemia, PR bleeds, abdominal pains, rectal prolapse or diarrhoea) should undergo colonoscopy earlier.

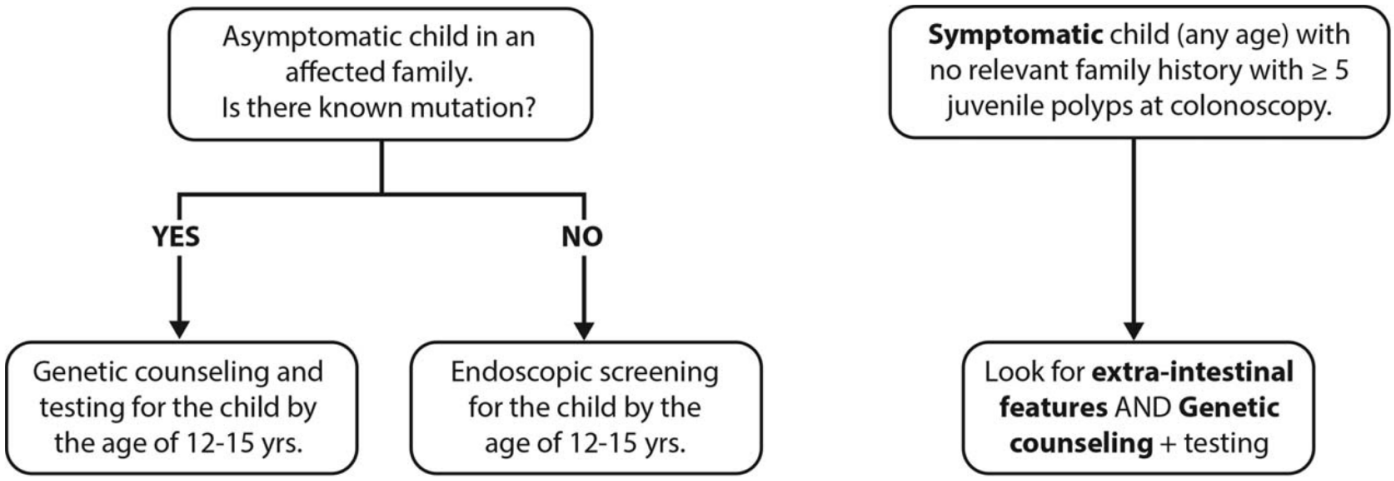

Figure 1

Management of Juvenile Polyposis Syndrome in Children and Adolescents: A Position Paper From the ESPGHAN Polyposis Working Group

Colonscopy should aim to remove all polyps >10mm and then be performed annually until no further polyps are found. If the patient has a single juvenile polyp then repeat colonoscopy is not usually required. If there is no prior family history but a new diagnosis is made in a symptomatic child then genetic testing should be offered to all first-degree relatives.

ADVANCED DISCUSSION (20 MINS PER CASE)

This is an opportunity to cover grey areas, diagnostic dilemmas and advanced management and escalation if there are more experienced trainees or senior registrars in your group.

ADVANCED CASE 1: TOXIC MEGACOLON

A 12 year old girl is transferred from another smaller hospital with a year's history of abdominal pains and PR bleeding with weight loss. She had not previously been referred to tertiary services. She underwent a colonoscopy yesterday afternoon confirming ulcerative colitis. She looks sick with a distended abdomen and peripheral oedema.

How would you manage this patient?

Questions to further discussion

1. What complication(s) do you think this girl may be suffering from?

2. What would you look for on an abdominal xray to confirm your diagnosis and how would this alter your management?

3. What could you do pre surgery to optimise this patient?

4. What do you understand by refeeding syndrome and what are the parameters you would want to correct urgently?

The major risks here in a patient with a long history of undiagnosed and therefore untreated ulcerative colitis, abdominal distention and peripheral oedema are toxic megacolon and refeeding syndrome. An abdominal x-ray should be performed: a bowel loop with diameter of more than 5.5cm is diagnostic of megacolon and should prompt a paediatric surgical review with a consideration for theatre and subtotal colectomy.

Refeeding syndrome is associated with electrolyte and metabolic derrangement in patients following a period of prolonged starvation which is then followed by reintroduction of nutrition. It would be important to correct albumin and electrolyte disturbances (the patient is likely hypoalbuminaemic, hypophosphataemic and hypokalaemic). The most commonly reported biochemical feature is hypophosphataemia although other electrolyte disturbances (including sodium imbalance, hypokalaemia, hypo magnesia and changes in glucose, protein, thiamine and fat metabolism) are usually present and are equally important. Refeeding syndrome is important to recognise as it can result in organ failure and ultimately death.

ADVANCED CASE 2: MECKELS DIVERTICULITIS

A one and a half year old boy presents to the paediatric ED. His parents are very frightened and show you his last nappy that contained a large amount of dark black stool. The infant is alert and active but appears to be a little tired. His parents report he is usually a happy baby and has never really been unwell.
He has had no vomiting and his parents do not report any concerns around possible abdominal pain. He has had previously normal stools. During examination you notice he is floppy and weak and on opening his nappy you find a large amount of melaena.

1. What are the possible complications of Meckel's diverticulum?
2. Consider how you might manage shock and stabilise this patient?

This presentation is most likely to represent a Meckel's Diverticulitis with painless rectal bleeding in an otherwise previously well child. The 'rule of 2' is a useful tool - it occurs in 2% of the population, within 2 feet of the ileocecal valve, is 2 inches in length, there are 2 types of ectopic tissue involved and presentation is generally before the age of two. Meckels can present in a variety of ways including gastro-intestinal bleeding, volvulus, intussusception, adhesions, hernia or perforation. Abdominal pain is sometimes, but not always, a feature. Ectopic gastric mucosa and pancreatic mucosa are the 2 structural possibilities in a Meckel's Diverticulum. The best imaging modality to use for detecting these is the technetium 99 scan.

In this example the patient is floppy and weak and has a large amount of melaena in his nappy. This raises the likely possibility of shock secondary to major haemorrhage. Shock in an infant as a complication of Meckel's Diverticulum should be managed as you would any other resuscitation situation - starting with ABC. Airway and breathing should be able to be cleared reasonably quickly and oxygen should be applied. The main focus of this scenario is hypovolaemia which should be treated with fluid resuscitation with emergency blood as quickly as it is available. Below is a link to a very clear and informative guideline and algorithm for managing circulation from the Royal Children's Hospital in Melbourne.

https://www.rch.org.au/trauma-service/manual/circulation-management/

Once this patient is stabilised then it is important for the area of small bowel containing the Meckle's Diverticulum to be resected and any ulcer (the likely source of bleeding) to be oversewn. It is not unusual that in patients who present with abdominal pain an initial surgical exploration for appendicitis is undertaken only to find a normal appendix and the diagnosis of Meckel's Diverticulum is more elusive.

A useful summary for Meckels's diverticulum:
https://www.ncbi.nlm.nih.gov/books/NBK6918

QUIZ QUESTIONS: (10 MINUTES)

Question 1.

Juvenile polyposis syndrome is predominantly defined as:

A: Polyps in a child less than 10 years old

B: More than one polyp in the colon.

C: Five or more JPs of the colon or rectum.

D: Blood with stools.

Answer:

Juvenile refers to the histology of the polyps, not to the age of the patient. The following three criteria define juvenile polyposis syndrome: Five or more JPs of the colon or rectum, or JPs in other parts of the GI tract, or any number of JPs and a positive family history. Blood in stools can be a sign of JPS, UC or Crohns or just simple constipation.

Question 2.

A 10 year old boy presents to the emergency department with a history of loose stools with mucus, intermittent blood, abdominal pains and anorexia. What would you do to objectively document the patient's symptoms?

A: Use the PUCAI as there is no histological diagnosis yet.

B. Use PCDAI as this is likely to be Crohn's disease.

C. Use his descriptions from the history.

Answer:

The Paediatric Ulcerative Colitis Activity Index (PUCAI) score is the most useful objective assessment of symptoms in IBD and prior to histological diagnosis is the most useful way of quantifying a history.

Question 3.

What is a useful rule to remember the pathophysiology and epidemiology of Meckel's Diverticulitis in children?

A. Rule of 4.

B. Rule of 3.

C. Rule of 2.

Answer:

The rule of 2 reminds us that Meckel's occurs in 2% of the population, within 2 feet of the ileocecal valve, is 2 inches in length, 2 types of mucosa involved and presents before the age of two.

Question 4.

What is the most common biochemical marker that would raise the possibility of a diagnosis of refeeding syndrome?

A. Hypokalaemia.

B. Hypophosphataemia.

C. Hypernatreamia.

D. Hypocalcaemia.

Answer:

Hypophosphataemia is a well recognised biochemical derangement signaling refeeding syndrome. It can also occur with other electrolyte disturbances as well as abnormal fluid shift as the syndrome is complex. Treatment is usually supportive in terms of careful biochemical correction (see Mehanna et al).

INFOGRAPHICS (2 minutes)

1 If you suspect IBD use the PUCAI score to give a reasonable objective idea about disease severity (even if the history suggests Crohn's).

2 Remember the "rule of 2" for Meckel's diverticulum - 2% of the population, within 2 feet of the ileocecal valve, 2 inches in length, 2 types of ectopic tissue involved and presentation is generally before the age of 2.

3 JPS is an autosomal dominant condition. Children who are at risk of JPS should be screened from the ages of 12 to 15 years although children who are symptomatic should undergo colonoscopy earlier.

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