LRI Children's Hospital

Management of acute severe ulcerative colitis in children

Staff relevant to:	Paediatric trainees and consultants working within UHL Children's Hospital
Team approval date: AWP approval date:	23/02/2024 12/03/2024
Version:	5
Revision due:	March 2027
Written by: Reviewed by:	Dr H Bhavsar H Bhavsar & D Saxena
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1. Introduction and Who Guideline applies to

Paediatric-onset ulcerative colitis (UC) is often more extensive than adult-onset UC with 60-80% presenting as pancolitis [1]. Its progression is also more dynamic. Within five years of diagnosis a significantly higher percentage of children with UC are admitted to emergency units for acute severe colitis (ASC), compared to adult disease; and children are also more likely to fail intravenous steroids during an acute severe episode. This translates into higher colectomy rates in children compared to adult UC populations [2]. It is therefore vital to promptly diagnose, initiate treatment and monitor progress during an episode of ASC.

2. Guideline Standards and Procedures

To deliver standardised/evidenced based practice for all patients admitted with ASC in keeping with current guidelines and IBD standards [3].

These guidelines are intended to assist in the management of ASC in the first 24-72 hours of presentation.

Subsequent management needs to be individualised depending on the clinical improvement and is not covered in this document.

2.1 Criteria for Inclusion

This guideline applies to those patients who meet the following criteria

- Patient that has a confirmed diagnosis of UC.
- Patient suspected to have a severe flare of colitis as defined by the Truelove & Witts Classification Score [4]. Six bloody stools associated with one or more activity within the severe category.

Activity	Mild	Moderate	Severe
Number of bloody stools per day	<4	4-6	>6
Temp	Afebrile	Intermediate	>37.8
Heart Rate	Normal	Intermediate	>90
Haemoglobin (g/dl)	>11	10.5-11	<10.5
ESR (mm/h)	<20	20-30	>30

2.2 Clinical assessment

In addition to the usual clinical assessment of an acutely unwell child, it is important to consider the following points:

History

Frequency of stooling, stool consistency, blood in stools, nocturnal stools, weight loss, abdominal pain, and limitation of activity, which are used to calculate the Paediatric Ulcerative Colitis Activity Index (PUCAI) (see page 5), which is specific for UC (not Crohn's disease).
Infectious exposures e.g., sick contacts, food poisoning, foreign travel
Check doses of current medications and patient compliance.

Examination

- Look for vital signs – tachycardia, anaemia, jaundice, dehydration, abdominal tenderness (Toxic megacolon)
- If a patient presents with physiologic disturbance (ie fever, increased heart rate, low BP) and/or a tender abdomen then they should be considered to have toxic megacolon.– consider surgical review

Investigations

1. Send two stool samples, one sample of sufficient amount (>2.5ml) of stool for culture/microscopy for ova, cyst and parasites, Clostridiodes difficile and Entamoeba histolytica PCR to microbiology department and one for Faecal calprotectin to biochemistry department. Please do not test for Clostridiodes Difficile in patients less than 2 years of age.

1. Routine blood tests include FBC, Biochemistry including LFTs, CRP, amylase, Alpha 1 acid glycoprotein/ESR and blood culture. Consider sending Azathioprine metabolites (6TGN/6MMPN) if on AZT or 6-mercaptopurine therapy and poor compliance is suspected. Consider sending amoebic serology for first presentation with severe colitis, or if patient has travelled to an endemic area since the last test.

2. Abdominal x-ray (AXR) should be performed upon admission with a low threshold especially in children with abdominal tenderness or distension, significant pain and those with systemic toxicity. Repeat AXR should be dictated by the initial AXR and subsequent clinical condition.

   Toxic megacolon – this is defined as colon diameter > 5.6cms (>4cm in less than 10 year olds) on AXR. If present on admission, an urgent surgical referral must be made and the patient should be made nil by mouth and commenced on IV cefuroxime and metronidazole, with doses as per the BNFc. A period of 24-48 hours of intensive medical therapy may be considered provided patient is sufficiently stable, but failure to respond by 48 hours, or development of further dilatation during medical therapy mandates consideration of colectomy.

2.3 Management

1. Intravenous methylprednisolone 1 mg/kg/day (up to 40 mg/day) once daily in the morning or in 2 divided doses, is recommended as the initial treatment at admission. A higher dose of 1.5 mg/kg/day (up to 60 mg/day) in 2 divided daily doses should be reserved for patients at the more severe end of the spectrum or for children who have failed oral steroids before admission

2. Empirical E.histolytica treatment with oral metronidazole (if tolerated) should be started in all patients who need immunosuppression (methyl prednisolone) before the stool PCR result could be available. This is due to high prevalence of E. histolytica in Leicester than elsewhere in the country. Intravenous Metronidazole should be used if oral Metronidazole is not tolerated. If stool PCR result is subsequently noted to be negative for E histolytica, empirical Metronidazole can be stopped.

   Please note that if stool PCR for E histolytica is positive, metronidazole should be continued (in higher doses as per BNFc for amoebiosis) for total duration of 10 days.

   Please discuss with microbiology if E histolytica is confirmed (either histologically or stool PCR/serology positive) as a second intra-luminal agent may be indicated.

   Intravenous Cefuroxime (in ‘severe infection’ dose) and Metronidazole are an agreed initial broad spectrum antimicrobial cover to consider when toxic megacolon or severe bacterial infection is suspected. In patients with a history suggestive of anaphylactic reaction to penicillin (which should only be a small proportion of paediatric patients), Meropenam, as a single agent antimicrobial can be started instead.

   Clostridiodes difficile infection confirmed on stool culture should be treated as per trust protocol - Clostridioides Difficile (formerly Clostridium) UHL Childrens Hospital Guideline

All antibiotic prescriptions should be reviewed at 48 hours with a maximum of 5 days as a standard duration. Antibiotic plans for individual cases, based on their culture results and clinical progress can be discussed with Microbiology as necessary.

3. All Mesalazine preparations (oral and rectal) should be discontinued upon admission to exclude Mesalazine intolerance, especially when Mesalazine has been commenced during the preceding few weeks; Reintroduction should be considered after significant improvement in the clinical condition

4. Nutritional support

    - Regular diet should be continued in most cases. Enteral (or parenteral in those not tolerating enteral) nutrition may be used if oral feeding is not tolerated or in the context of malnutrition.
    - Body weight, caloric intake, and hydration status should be monitored over the course of an admission.
    - Electrolytes should be monitored in line with baseline values and clinical status.
    - Oral or enteral feeding is contraindicated in cases of megacolon, or when surgery is imminent

5. Pain management

    - Non-steroidal anti-inflammatory drugs (NSAIDs) should be avoided in ASC.
    - Opiates should be used exceptionally with caution and close monitoring, in doses equivalent to 0.1 mg/kg morphine, given the remote risk of facilitating megacolon.

6. Monitoring - Calculate PUCAI score (see table 1) and reassess daily to monitor progress. It is the best validated predictive and decision-making tool in children with ASC [5].

7. Consider IV fluids if clinically dehydrated.

8. Subcutaneous low molecular weight heparin (LMWH) should be considered in adolescents with ASC when 1 or more risk factors are present: smoking, oral contraceptives, complete immobilization, central venous catheters (including PICC line), obesity, concurrent significant infection, known prothrombotic disorder, previous VTE, and family history of VTE.

    For further details, please refer to UHL Guidelines for Pharmacological and Mechanical Thromboprophylaxis for venous thromboembolism[9]

9. Request surgical review if AXR suggestive of toxic megacolon

10. Anaemia is a recognised complication in ASC and blood transfusion should be considered in discussion with the on-call Paediatric Gastroenterologist when haemoglobin level is below 8 mg/dL. Iron replacement without the need for transfusion should be considered in children whose rectal bleeding has ceased.

11. Discuss with the Paediatric Gastroenterology Consultant on service at Leicester Royal Infirmary (LRI) at the earliest opportunity via switchboard. Consider transfer to LRI if clinically indicate

COVID 19 in Patients with Acute severe UC[8]

1. Methylprednisolone is appropriate as a first-line management in all patients with ASC including those with symptomatic COVID-19.
2. Sigmoidoscopy is recommended prior to escalation to second-line therapy or colectomy. Delaying colectomy is not appropriate in a COVID 19 patient
3. Once patient is clinically well, oral prednisolone with tapering over 8-10 weeks is appropriate for COVID 19 patients.
4. After successful corticosteroid rescue, thiopurine maintenance is appropriate in patients with negative SARS-CoV-2 confirmed by lateral flow test and asymptomatic patients with positive PCR test but uncertain in symptomatic COVID-19 positive patients.

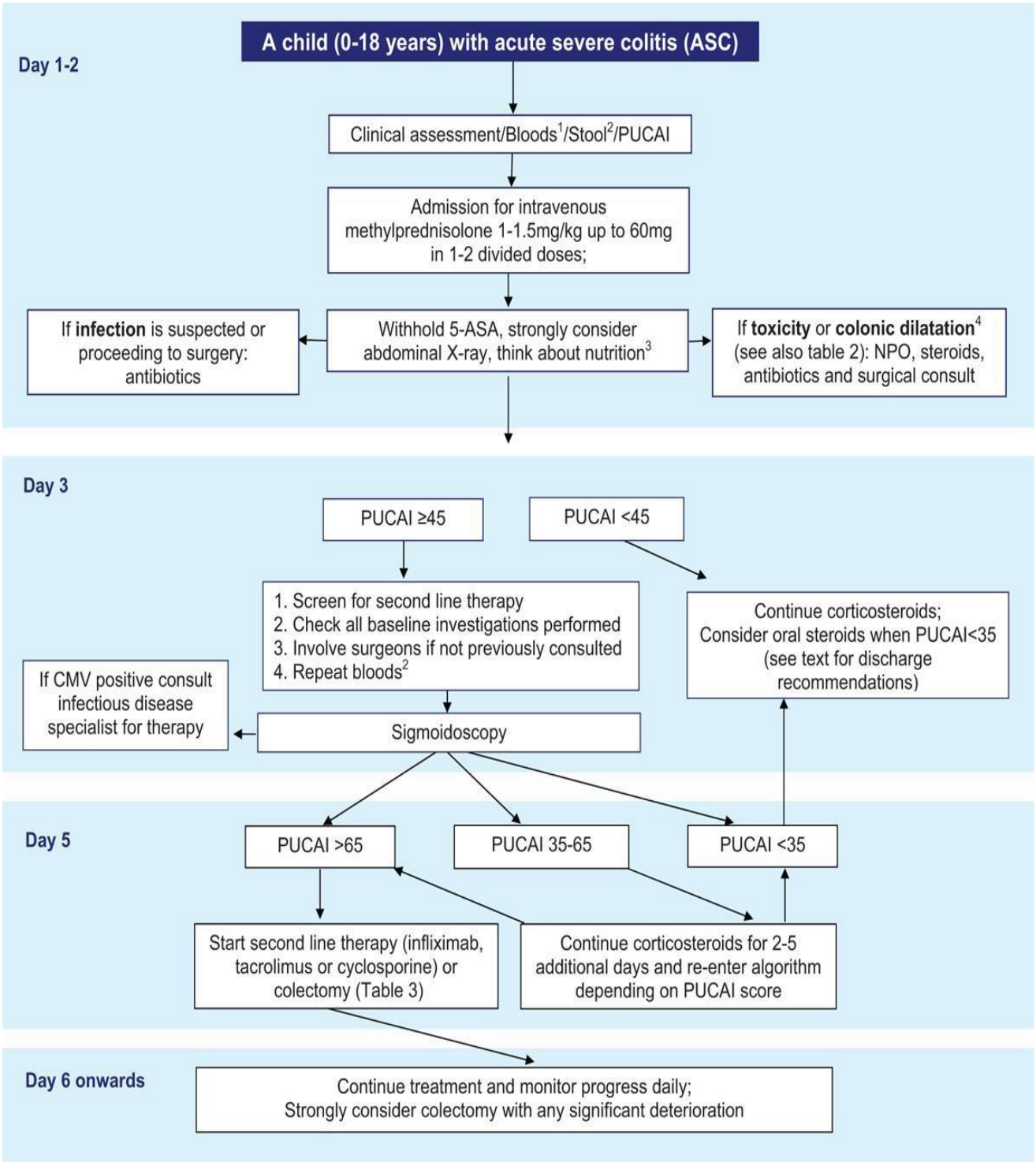
Please note this guideline is only intended for patients with ASC and not for those who have mild to moderate UC. Please discuss with senior colleagues for advice.

Table 1: **PAEDIATRIC ULCERATIVE COLITIS INDEX (PUCAI)** [5]

Name/DOB: **Date**

	Points							
(1) Abdominal pain No pain Pain can be ignored Pain cannot be ignored	 0 5 10							
(2) Rectal bleeding None Small amount in <50% stools Small amount with most stools Large amount (>50% stool)	 0 10 20 30							
(3) Stool consistency of most stools Formed Partially formed Completely unformed	 0 5 10							
(4) Number of stools per 24h 0-2 3-5 6-8 >8	 0 5 10 15							
(5) Nocturnal stools (any episode causing wakening) No Yes	 0 10							
(6) Activity level No limitation of activity Occasional limitation of activity Severe restricted activity	 0 5 10							
SUM OF PUCAI (0-85)								

Figure 2: Treatment algorithm for acute severe UC [6]



3. Education and Training

None

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Audit to review inpatient management of acute severe ulcerative colitis patients	Discussing results of audit and learning from it	Dr Bhavsar	yearly	Departmental audit meetings

5. Supporting References

1. J Van Limbergen, R K. Russell, et al. Definition of Phenotypic Characteristics of Childhood-Onset Inflammatory Bowel Disease, Gastroenterology 2008,Vol135,Issue 4

2. Fumery M, Singh S, Dulai PS , *et al*. Natural history of adult ulcerative colitis in population-based cohorts: a systematic review. Clin Gastroenterol Hepatol 2018; **16**:343–56.

3. Turner D , Ruemmele FM , Orlanski-Meyer E , *et al* Management of paediatric ulcerative colitis, part 2: acute severe Colitis-An evidence-based consensus guideline from the European Crohn's and colitis organization and the European Society of paediatric gastroenterology, hepatology and nutrition. J Pediatr Gastroenterol Nutr 2018;**67**:292–310

4. Truelove S C, Witts L. Cortisone in ulcerative colitis: final report on a therapeutic trial. BMJ. 1955;2:1041–1048

5. Turner D, Walsh CM, Benchimol EI, et al. Severe paediatric ulcerative colitis: incidence, outcomes and optimal timing for second-line therapy. Gut 2008;57:331–8.

6. Turner D, Travis SP, Griffiths AM, et al., European Crohn's and Colitis Organization; Porto IBD Working Group, European Society of Pediatric Gastroenterology, Hepatology, and Nutrition. Consensus for managing acute severe ulcerative colitis in children: a systematic review and joint statement from ECCO, ESPGHAN, and the Porto IBD Working Group of ESPGHAN. Am J Gastroenterol 2011;106:574–88.

7. Fell JM, Muhammed R, Spray C, et al. Management of ulcerative colitis. Arch Dis Child 2016;101:469–74.

8. Hansen R.et.al. Adaptations to the current ECCO/ESPGHAN guidelines on the management of paediatric acute severe colitis in the context of the COVID-19 pandemic: a RAND appropriateness panel. Gut. 2021 Jun;70(6):1044-1052. doi: 10.1136/gutjnl-2020-322449. Epub 2020 Sep 1. PMID: 32873696.

9. Paediatric VTE risk assessment tool, University Hospitals of Leicester

6. Key Words

Ulcerative colitis, Paediatric Inflammatory bowel disease, Acute severe colitis, PUCAI

The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title) DR BHAVSAR – Consultant Paediatrician	**Executive Lead** Chief medical officer

Version 5 (March 2024)
Details of Changes made during review:

Page 2 - Examination

- Look for vital signs – tachycardia, anaemia, jaundice, dehydration, abdominal tenderness (Toxic megacolon)
- If a patient presents with physiologic disturbance (ie fever, increased heart rate, low BP) and/or a tender abdomen then they should be considered to have toxic megacolon.– consider surgical review

Additional points on Covid 19 patients presenting with ASC with new reference
COVID 19 in Patients with Acute severe UC[5]

- Methylprednisolone is appropriate as a first-line management in all patients with ASC including those with symptomatic COVID-19.
- Sigmoidoscopy is recommended prior to escalation to second-line therapy or colectomy. Delaying colectomy is not appropriate in a COVID 19 patient
- Once patient is clinically well, oral prednisolone with tapering over 8-10 weeks is appropriate for COVID 19 patients.
- After successful corticosteroid rescue, thiopurine maintenance is appropriate in patients with negative SARS-CoV-2 swab and asymptomatic patients with positive swab but uncertain in symptomatic COVID-19.

2.3 Management

1. Empirical E.histolytica treatment with oral metronidazole (if tolerated) should be started in all patients who need immunosuppression (methyl prednisolone) before the stool PCR result could be available. This is due to high prevalence of E. histolytica in Leicester than elsewhere in the country. Intravenous Metronidazole should be used if oral Metronidazole is not tolerated. If stool PCR result is subsequently noted to be negative for E histolytica, empirical Metronidazole can be stopped.

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All antibiotic prescriptions should be reviewed at 48 hours with a maximum of 5 days as a standard duration. Antibiotic plans for individual cases, based on their culture results and clinical progress can be discussed with Microbiology as necessary.

References revised and updated.

Minor edits to wording throughout.