



Paediatric Gastroenterology Clinical Network

Management of Inflammatory Bowel Disease in Children and Adolescents in New Zealand

A Clinical Guideline

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DISCLAIMER

The content of this guideline do not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

PUBLIC DOMAIN NOTICE

This document aims to provide an evidence-based guide to the diagnosis and management of Inflammatory Bowel Disease (IBD) in children and adolescents across New Zealand. The recommendations within this document provide a guide and may not encompass every clinical situation. The guideline is formulated within the framework of the Paediatric Gastroenterology Clinical Network.

GLOSSARY

ASA	Aminosalicylates
ASC	Acute severe colitis
CD	Crohn's disease
EEN	Exclusive enteral nutrition
GI	gastrointestinal
IBD	Inflammatory Bowel Disease
IBDU	IBD-Unclassified
MRE	MRI Enterography
PCDAI	Paediatric Crohn Disease Activity Index
PGA	Physicians Global Assessment
PUCAI	Paediatric Ulcerative Colitis Activity Index

INTRODUCTION

What is IBD?

The Inflammatory Bowel Diseases (IBD) are chronic inflammatory conditions of the gastrointestinal (GI) tract and comprise two main types - Crohn's Disease and Ulcerative Colitis.

Crohn's disease (CD) can involve any part of the GI tract, from the mouth to the anus. CD typically features transmural inflammation and skip lesions. The presence of granulomata on histology is a feature seen in CD but not UC.

Ulcerative Colitis (UC) typically involves the colon extending proximally from the rectum for a variable distance. Inflammatory changes in UC are superficial and continuous. UC is less common than CD in children.

In addition, the term IBD-Unclassified (IBDU) is used when children clearly have IBD, but the biopsy or other results are not able to clearly distinguish between CD and UC at that time. It usually becomes clear over time whether this is actually CD or UC.

What is the cause of IBD?

The precise cause of IBD is not clear. Three important factors are involved in causing IBD: **genes, bacteria and innate immune system responses in the digestive tract**. IBD likely occurs in people with a certain combination of genes: these genes alter the defences in the surface of the bowel, leading to a different response to bacteria in the intestines, resulting in uncontrolled inflammation. Certain genes are important in early-onset (paediatric) IBD.

What is important about paediatric IBD?

Over the past decades IBD has become increasingly common around the world. CD, in particular, is now seen much more frequently. Initially these changes in the pattern of IBD were seen in "western" countries, but more recently increased rates of IBD have also been seen in other countries. As well as becoming more frequent overall, IBD is now also occurring more commonly in younger children.

IBD in children differs greatly from adult onset disease in many ways, including disease location and patterns. For example, the majority of children with UC have pan-colonic involvement at diagnosis and few children have isolated proctitis. In children with CD, upper gut involvement in CD is also seen much more commonly than in adults (60% vs 5%).

IBD in children and adolescents commonly impacts upon growth, nutrition and pubertal development. Almost all children with CD and at least half of children with UC have poor weight gain or weight loss prior to diagnosis. Children with UC and CD may also have impaired linear growth at diagnosis or subsequently. This may lead to a reduction in final adult height. Furthermore, because IBD commonly causes problems during adolescence, it may impact upon the onset and progression of puberty. These factors influence the key aspects of the management of IBD in children and adolescents.

1. APPROACH TO DIAGNOSIS

1.1 Typical Presentation patterns

The classical symptoms in children with CD are abdominal pain, diarrhoea and weight loss, whilst typical symptoms in paediatric UC are abdominal pain and bloody diarrhoea. Children with IBD may present with a variety of these more common symptoms, but may also present with a wide array of other atypical symptoms. Atypical symptoms may include poor linear growth, delayed puberty, isolated perianal disease, or oral/perioral changes. In addition, some children may present with extra-intestinal symptoms prior to bowel symptoms (e.g. joint symptoms or rashes).

Children most commonly present with IBD in early adolescence. However, IBD can present at any age from early infancy onwards. Consequently, IBD needs to be considered across a range of presentations and ages, meaning that paediatricians need to be alert and consider IBD as a possible diagnosis. Depending on the presentation, differential diagnoses to be considered include Irritable Bowel Syndrome, Coeliac disease, infectious enteritis or colitis, carbohydrate intolerance, primary immune deficiencies or allergic processes.

1.2 Initial Investigations

In the majority of children presenting with persisting diarrhoea (with or without blood), the initial approach should be to exclude infection and to measure inflammatory markers (**Table 1**).

Several studies have illustrated that the sensitivity and specificity of serum inflammatory markers varies widely in children, especially in those with UC. Whilst platelets, ESR and CRP may be elevated with gut inflammation, albumin levels may fall (as a negative acute phase reactant). Measurement of multiple markers can increase the utility of these tests. The presence of white cells (ascertained on a fresh stool sample) may indicate gut inflammation. Faecal calprotectin is a sensitive and specific indicator of gut inflammation. However, false positive levels may be seen in certain situations, such as young children, following NSAID usage and during/after an acute enteric infection.

Table 1: Initial investigations in children suspected of IBD

Stool culture - on two or three separate occasions
(consider specific request for *Clostridium difficile*, *Yersinia* and *Aeromonas*)
Stool microscopy (for white cells)
Full blood count (especially Hb, MCV, Platelets)
ESR
CRP
Albumin
Faecal calprotectin

Other relevant investigations may include *Yersinia* serology, coeliac screening tests (to exclude this), or others as clinically indicated.

1.3 Definitive diagnostic investigations

The diagnosis of IBD is based upon specific standard endoscopic, histologic and radiological findings (**Table 2**). In the vast majority of children a tissue diagnosis is required to confirm inflammatory bowel disease.

The timing for undertaking these definitive investigations will depend on the clinical situation. In general, it is appropriate to facilitate these tests within 2-4 weeks of presentation.

However, if the patient is unwell and requiring inpatient admission, endoscopic assessment within a week would be considered reasonable.

Inpatient admission and assessment should be considered in children with significant pain, very frequent stooling or blood loss (sufficient to lead to anaemia, tachycardia or dehydration). In addition, children with features of Acute Severe Colitis (ASC: see **Section 2.3**) will always require admission.

Based upon the well-described phenotypes of CD and UC in children and adolescents, international recommendations include assessment of the whole gut as initial investigations. Endoscopic assessment of both the upper gut and the lower gut is required. Diagnosis and/or management may be altered significantly according to the findings in the upper gut.

Small bowel imaging is also required to assess involvement of this segment of the gut. MRI Enterography (MRE) (small bowel series MRI) is preferable, given the three-dimensional and functional information provided. Such studies should be undertaken with oral contrast (to achieve small bowel distension) following standard protocols. The performance and interpretation of MRE are highly specialised skills, so the study should be carried out by a Radiology team with experience in the procedure. For younger children in whom MRE may not be feasible, barium meal and follow-through is an alternative.

Where available and appropriate, capsule endoscopy may provide mucosal assessment, but this is limited by the age of the child, funding and availability. Although ultrasound also provides a radiation-free assessment, this method is operator-dependent and is not universally recommended. CT (and consequent radiation exposure) is rarely indicated and should be avoided wherever possible.

Table 2: Investigations required in establishing a diagnosis of IBD in children

Upper Gastrointestinal Endoscopy and biopsies Ileo-Colonoscopy and biopsies Small bowel imaging (e.g. MRI Enterography)

1.4 Endoscopic Assessment

As above, upper and lower endoscopy is required in the assessment of possible IBD. Endoscopic assessment may be contraindicated in some individuals, especially those with features of acute severe colitis (see Section 2.3).

Endoscopic procedures in younger children (e.g. those aged less than 14 years of age) should be carried out under general anaesthesia or deep sedation in a child and family-friendly setting by staff that have experience and expertise in paediatric endoscopy, with support from other paediatric trained staff (including anaesthetist, nursing staff, theatre staff, recovery staff and other relevant personnel).

Endoscopy should be accompanied by multiple biopsies from all segments of the upper and lower gut, regardless of whether the endoscopic appearance is normal or abnormal. These biopsies should be reviewed by a pathologist with expertise in paediatric IBD (or consider review of histology slides in tertiary centre) along with regular clinical-pathology meetings.

1.5 Differentiation between Crohn's disease and ulcerative colitis

The findings of endoscopic, histological and radiological investigations, along with the clinical context, can be used to classify children as CD or UC. Various features can help to differentiate between UC and CD. Disease location, skip lesions, the presence of granulomata and other features (such as the presence of perianal disease) are key features.

The finding of colitis with some features that are unable to differentiate between CD and UC may be termed IBD Unclassified (IBDU). Other investigations (such as serological tests) may be helpful in IBDU.

1.6 Baseline assessment in children diagnosed with IBD

A range of baseline tests and assessments are indicated in all children at the time of or immediately following the diagnosis of IBD (**Table 3** and **Appendix 1 – Diagnosis Checklist**). These include establishing the patterns of other organ involvement (e.g. liver disease) and assessment of micronutrient levels (e.g. malabsorption).

1.7 Liaison with tertiary services during assessment of possible IBD

Physicians are encouraged to communicate with the relevant tertiary Paediatric Gastroenterology service following the initial assessment of a child with potential IBD and/or after review of initial investigations (as listed in **Table 1**) in order to jointly plan the next steps, with consideration of age/clinical status/geography.

South Island centres should liaise with Christchurch, whilst North Island centres should communicate with Auckland. (**Contact details see Appendix 2**)

Table 3: Baseline assessment at the time of diagnosis of IBD in children

Investigations

Iron and Ferritin, B12 & Folate
Vitamin D
Calcium, Magnesium, Phosphate & Zinc
Liver chemistry
Urea, electrolytes & renal function
Varicella serology
TPMT assay (activity)

Consider

Neutrophil function testing (especially in younger children presenting with granulomatous inflammation)
Measles serology (if history of incomplete vaccination)
Bone age (left wrist), particularly if pubertal delay
Bone densitometry (consider if history of recurrent fractures or significant malnutrition) (*see Section 3.5*)
Serological tests (e.g. pANCA and ASCA) may be helpful in IBDU, but is not considered as first line testing)

Document

Disease phenotype, including presence of perianal disease and extraintestinal manifestations
Document disease activity score (PCDAI or PUCAI) (*see Appendices 10 and 11*)
Past medical history
Vaccination status (standard schedule)
Previous varicella infection or vaccination
Risk factors for TB
Family history of IBD
Historical growth data
Parental heights (and calculate mid-parental height)
Tanner status

Measure

Weight and Height
Derive BMI

2. MANAGEMENT OF CHILDREN WITH IBD

2.1 Model of care

For any child with a chronic disease it is vital to deliver as much care as possible as close to their home. However, the management of IBD is complex and these children greatly benefit from the input of a multidisciplinary team with specialist experience and expertise in IBD.

All children with IBD should be managed either by, or on a shared care basis with, a tertiary paediatric gastroenterology centre. For children living outside Auckland and Christchurch, this will necessitate some form of shared care arrangement for medical care. Although the precise nature of the shared care arrangement may vary from centre to centre, the following is recommended as a minimum standard:

- A named local paediatrician
- A named tertiary gastroenterologist
- At least 3-4 monthly follow-up by local paediatrician
- At least annual review by tertiary gastroenterologist
- Clear lines of responsibility and pathways of care if concerns about acute deterioration

It would be anticipated within each DHB that there will be a number of children with IBD. Their local care may be most effectively coordinated through one paediatrician at each hospital, managing them in conjunction with a specialist centre. This will have a number of benefits, including:

- “upskilling” of the general paediatrician in the management of IBD
- facilitating good working relationships and clear lines of responsibility
- ensuring rapid access to the full range of relevant investigations
- ensuring rapid and appropriate management of disease exacerbations
- coordination of multidisciplinary care

Reviews by tertiary gastroenterologist may be in the form of Outreach clinics in the regional centre, ideally carried out jointly with a nominated local paediatrician. The frequency of these clinics will vary depending on local need. Various methods to ensure effective communication between outreach clinics should be utilised: these might include email correspondence, telephone calls or regular video conferencing.

Paediatric dietitians and nursing staff have critical roles in the care and management of all children with IBD. All children in regional centres should be seen regularly by a paediatric dietitian locally along with support from a dietitian in the relevant tertiary centre. Similarly, children should have access to specialist nursing care locally with support from their tertiary centre. Many children also require access to other disciplines (e.g. psychology). Staff in regional centres should also link with relevant personnel in the two tertiary centres for support, resources and information.

It is also essential that each patient and family is provided with a list of key people, along with relevant contact details. (**Appendix 3**). This should include contact details in the event of a relapse or flare of symptoms, or if they have queries or concerns.

2.2 Liaison with primary health care services

The GP should maintain a key role in the overall health-care of the child with IBD. Following diagnosis the GP should be updated with regards key details and provided with an overview of standard expectations and overall management goals.

2.3 Initial Management steps following diagnosis of IBD

Education

Children and their parents should begin initial education steps following diagnosis. Age-appropriate tools and resources should be utilised. Two or more education sessions may be required, with further review of understanding and opportunities for questions during subsequent review appointments. Written resources and information about electronic resources should be provided (**Appendix 13**)

Dietetic assessment

All children should have a formal dietetic assessment following diagnosis : this should include assessment of growth and review of diet. This is especially relevant in those with substantial nutritional impairment at diagnosis. However, in children managed initially with EEN, it may be best to review normal eating patterns and dietary choices after the period of EEN not before.

Allied health referral

Referral to social work, psychology or other allied health services should be considered as required.

Support Group

Peer support and/or an introduction to local support groups should be offered following diagnosis. However, some children and their parents may not wish to undertake this until a later time.

Other

Referral to Health School should be considered for children with significant disruption to their usual schooling, especially when this disruption is expected to be prolonged in the coming months.

2.4 Induction of remission

The first step in management of newly-diagnosed IBD is the induction of remission. Various interventions can be employed to induce remission (**Table 4**). The choice of therapy will reflect individual patient factors, disease type and disease severity.

Exclusive enteral nutrition (EEN) is the preferred initial therapy for newly diagnosed CD, regardless of disease location. Newly diagnosed UC may require corticosteroids or aminosalicylates (ASA), or infrequently EEN.

Table 4: Common Therapies for IBD in children and adolescents

Induction of Remission

Exclusive Enteral nutrition
Steroids
Antibiotics
ASA
Infliximab/Adalimumab

Maintenance of Remission

Supplementary enteral nutrition
ASA
Azathioprine / 6-MP
Methotrexate
Infliximab/Adalimumab

2.4.1 Exclusive Enteral Nutrition

This therapy involves the exclusive administration of a liquid diet (typically a polymeric formula) for up to 8 weeks using a standard protocol (**Appendix 4**). This approach is as effective as steroids in inducing clinical remission of active CD, but leads to much more effective mucosal healing and avoids any side-effects related to steroids. As well as anti-inflammatory effects, enteral nutrition has great benefits upon nutrition and growth. EEN tends to be more effective when used as the first treatment for CD: but it can still be useful

for management of disease flares in children who have had CD for some time. EEN does not have a routine role for the induction of remission of active UC.

Key aspects important in the success of EEN are a multidisciplinary approach, with clear education and ongoing review and support. Standard information sheets for children are available (**Appendix 5 A and B**). An additional handout covers the reintroduction of standard diet at the end of the course of EEN (**Appendix 6**).

2.4.2 Corticosteroids

Steroids may be administered orally (prednisone, prednisolone, budesonide), rectally (prednisone, methylprednisone) or intravenously (hydrocortisone, methylprednisolone) (See **Appendix 7: Pharmacopoeia**). Oral steroids may be required to induce remission in severe CD (e.g. if EEN proves unsuccessful or is not tolerated) or moderate-severe UC. Intravenous steroids may be required in a child who does not respond to oral therapy, or who has severe disease. IV steroids are particularly indicated in acute severe colitis (ASC), as detailed in Section 2.3.3. Topical steroids may have a role in distal colonic disease, but tend to be poorly tolerated in children.

When required for active disease, oral prednisone should be commenced at a maximal dose of 40 mgs daily (in the morning). After establishing control, the dose of prednisone should then be progressively weaned, so that the course of steroids extends over 8-10 weeks.

Oral Budesonide may be used as an alternative to oral prednisone for terminal ileal CD (subject to Special Authority criteria). Although budesonide has less systemic absorption, it may not be as effective as prednisone and therefore has a limited role.

Although traditionally steroids have been used as the principal way to manage active disease, they should not be part of routine maintenance therapy and must be used sparingly.

2.4.3 Aminosalicylates (ASA)

This family of medicines can be utilised to induce remission (in mild disease), especially in UC. The family of drugs includes Sulphasalazine (also known as Salazopyrin), Mesalazine (Pentasa, Mesasal) and Olsalazine (Asacol). ASA drugs are most often administered orally, but are also available as rectal therapies. (**Appendix 8**)

2.4.4 Antibiotics

Antibiotics have a role in the induction of remission of mildly active CD, but probably do not have any role in UC. Metronidazole (Flagyl) and ciprofloxacin (Ciprofloxin) are the two most commonly utilised antibiotics (**Appendix 7**). Both antibiotics tend to be used for short courses for luminal disease, but may be utilised for more extended periods in perianal disease.

2.4.5 Biological or focused therapies

Two biological drugs (infliximab and adalimumab) are currently available in NZ on special authority according to set criteria. In general, the decision to start biologics should be taken only after discussion with the tertiary gastroenterologist on the multidisciplinary team.

Both infliximab and adalimumab are supported with paediatric data for use in CD in individuals with severe disease that is unresponsive to standard therapy, or for fistulising disease. Infliximab is supported with paediatric data for use in intractable UC. Induction regimens and patient information resources are available for both drugs.

2.4.6 Other drugs to induce remission

The calcineurin inhibitors (tacrolimus and cyclosporin) may be indicated in severe colitis unresponsive to other therapies, including corticosteroids. Mycophenolate and Thalidomide are used rarely in those unresponsive to other therapies.

2.5 Management of acute severe colitis (ASC)

2.5.1 ASC is a medical emergency

Acute severe (or fulminant) colitis is a medical emergency and can lead to significant morbidity and potential mortality. ASC can present as the first presentation of IBD or can present at any subsequent stage.

2.5.2 Definition of ASC

ASC is defined in adults as more than 6 bloody stools a day, with fever (over 37.8°), tachycardia (above 90), anaemia (Hb less than 105), or elevated ESR (over 30).

In children, ASC is defined as a PUCAI of greater than 65 (see **Section 4.2 & Appendix 11** for explanation of PUCAI).

2.5.3 Early referral to tertiary centre

Children with ASC should be managed in a tertiary paediatric centre, with access to specialist paediatric gastroenterology and paediatric surgical expertise.

Following initial assessment and evaluation, children fitting criteria of ASC should be discussed directly and promptly with the appropriate tertiary service. Transfer should be considered as soon as feasible and safe: however, prior to transfer, children should have initial fluid stabilisation (as required), key investigations and commence medical therapy.

2.5.4 Management of ASC

Initial assessment in ASC includes detailed abdominal examination, measurement of vital signs and assessment of disease activity score (PUCAI). Stool samples should be sent, with standard culture and specific screening for *C difficile*. CMV infection should be excluded endoscopically in children with steroid-resistant disease.

Colonoscopic assessment should be considered **only** when feasible and safe to undertake, particularly in children without an established diagnosis. In some cases with severe colitis, it may be safe to only undertake a limited colonoscopic assessment (e.g. sigmoidoscopy). A subsequent full endoscopic assessment would be required in such cases.

Disease activity should be monitored regularly during admission, with frequent assessment of vital signs, daily completion of PUCAI scores and monitoring of key blood tests (ESR, full blood count, albumin and electrolytes) at admission and regularly over the following days. Early paediatric surgery referral is required. Regular clinical review (at least daily) is also essential.

Initial treatment of ASC should be with intravenous corticosteroids, with methylprednisolone (1-1.5 mg/kg/day, as 1 or 2 daily doses to max of 60 mgs daily) favoured due to less mineralocorticoid effect. Antibiotics are not indicated routinely but should be considered when sepsis suspected or when toxic megacolon present. There is no evidence for the routine use of prophylactic heparin to prevent thromboembolic events in children, but this may be considered in particular circumstances. ASA therapies should be discontinued at admission in patients with known IBD and introduction should be delayed in newly diagnosed patients.

There is no therapeutic benefit in bowel rest for the majority of children with acute severe colitis. Regular diet should be continued, and nutritional support (enteral or parenteral) considered if oral intake is inadequate. Oral intake should be ceased when surgery is imminent and is contraindicated in toxic megacolon.

Complications (such as perforation or toxic megacolon) should be considered in children with increasing or severe pain. Narcotics or non-steroidal anti-inflammatory drugs are not recommended in the setting of acute severe colitis.

PUCAI scores can be used to monitor response and the need for secondary therapy. A score of >45 at day 3 indicates likely poor response to corticosteroids and a need to prepare for rescue therapy. This includes introduction to, and review by, a paediatric surgeon, if this has not already occurred. A score of >65 on day 5 indicates a need to commence rescue therapy on that day. In children with PUCAI scores of between 35 and 60 at day 5, steroids can be continued for a further 2-5 days before secondary therapy should be considered. Children with scores of less than 35 points on day 5 are not likely to require rescue therapy

Plain radiographs of the abdomen should be obtained in any child with clinical signs of toxicity and then repeated subsequently as indicated. The diagnostic criteria for toxic megacolon in adults comprise radiological evidence of colonic dilatation (≥ 56 mm), along with signs of toxicity. Children may develop toxic megacolon with lesser degrees of dilatation. Urgent surgical review is required in all children with suspected or established toxic megacolon. Conservative management is appropriate if the child has stable vital signs and there are no signs of sepsis. If signs of toxicity worsen, then immediate colectomy should be undertaken. Rescue medical therapies are not indicated in the setting of toxic megacolon.

Rescue therapies for ASC unresponsive to initial therapies include medical (infliximab or calcineurin inhibitors) and surgical (colectomy) options. Sequential medical rescue therapies are not recommended in children. If colectomy is required in acute severe colitis in children, subtotal colectomy and ileostomy is recommended. Pouch formation can subsequently be considered. Surgical complications can be reduced by avoiding delays in colectomy to enhance nutrition or to wean corticosteroids, and the use of perioperative broad spectrum antibiotic coverage.

2.6 Maintenance of remission of IBD

Following the induction of remission, the next phase of therapy of IBD is the maintenance of remission and prevention of relapse. The choice of maintenance therapy is also dependant on disease type, disease severity and patient factors.

2.6.1 Aminosalicylates (ASA)

These drugs (as above) may have roles in the maintenance of remission, especially in mild or moderate disease, with particular role in UC (**Appendix 8**).

2.6.2 Thiopurines

These drugs (Azathioprine and 6-MP) have roles in moderate to severe CD and UC (**Appendix 9: Thiopurine protocol and Checklist**). They are considered when ASA drugs are not sufficient to maintain control or in children with moderate-severe disease at diagnosis. Both drugs have a slow onset of action, and are associated with various potential side effects. Standard information sheets for children and families should be provided prior to commencing treatment.

2.6.3 Methotrexate (MTX)

MTX may be indicated in children who are not able to tolerate a thiopurine or have failed thiopurine therapy. Evidence for the use of MTX in CD is much stronger than in UC (with only several case series in children with UC). MTX is administered once weekly as a subcutaneous injection (**Appendix 10: MTX Protocol and Checklist**). Once remission is well-established, it may be appropriate to consider a reduction in dosage. Standard

information sheets for children and families should be provided prior to commencing treatment. Standard handling precautions should be followed.

2.6.4 Biological drugs

When these drugs have been used to induce remission successfully, they can be continued as maintenance therapy. In general, the decision to continue biologics as a maintenance therapy should be taken after discussion, at an appropriate point, with the tertiary gastroenterologist on the multidisciplinary team.

The standard regimen for Infliximab is 5mg/kg every 8 weeks, whilst Adalimumab is given fortnightly (standard adult dose 40mg/dose - adjusted according to surface area for children). Standard information sheets for children and families should be provided prior to commencing treatment.

2.6.5 Supplementary enteral nutrition

When EEN has been successfully employed to induce remission in CD, the continuance of enteral support can help in the maintenance of remission. This can be achieved in several ways: the most straightforward way is the administration of daily oral supplements of up to 1000 mls of enteral formula in addition to normal diet.

2.7 Surgery

Surgical procedures may be required to induce remission in medically-intractable disease or to manage complications (such as stricturing disease in CD).

2.7.1 Overarching principles

The indications for surgery in inflammatory bowel disease are limited to specific clinical scenarios. Whilst surgery has a role to play in the management of IBD, inappropriate surgical management may be associated with significant complications and adverse clinical outcomes.

In general, the decision for surgery should be taken only after discussion with the tertiary paediatric gastroenterologist and the multidisciplinary team involved in the patient's care. Surgery should be performed by a surgeon with skills and experience in managing such procedures in children and adolescents. This will almost always entail involvement of a tertiary centre. Even within a tertiary centre, such intervention may also require consultation with or involvement of an adult colorectal surgeon.

2.7.2 Specific Surgical Indications in CD

Perianal Crohn's disease: Surgery has a role in delineating the extent of perianal disease and facilitating drainage of active perianal sepsis. This may involve examination under anaesthesia, incision and drainage of abscesses and placement of seton sutures. Surgical intervention in perianal CD should be seen as one component of overall coordinated care, in conjunction with medical therapy.

Medically intractable CD/steroid dependency: Resectional surgery is indicated if there is a failure of medical therapy to induce or maintain remission, particularly in the case of localised small bowel disease or isolated colonic disease. It may also be indicated in the context of steroid dependency (particularly if there is evidence of complications related to chronic use). Alternatively, a defunctioning procedure may be appropriate to consider.

Fistulising CD: Enteroenteric fistulae frequently require surgery as part of multidisciplinary management. In general, this will entail limited resection of the involved bowel with or without stoma formation.

Strictureing CD: Although inflammatory strictures may respond to medical therapy, fixed, fibrotic strictures require surgical intervention. Short strictures may be amenable to dilatation. However, this should only be carried out by experienced individuals and must be performed in a centre where there is recourse to surgery in the event of complications, such as perforation. Longer strictures are likely to require surgical resection, though occasionally stricturoplasty may be considered in cases where bowel length preservation is critical.

Intra-abdominal collections (phlegmons) in CD: Intra-abdominal abscess collections may complicate enteroenteric or enterocutaneous fistulae. Although they may respond to percutaneous drainage in combination with medical management, they may necessitate resection of the affected bowel. Management of collections requires a coordinated medical and surgical approach.

Masses in CD: A palpable mass may be found at diagnosis or follow-up in children with CD. Although inflammatory lesions may resolve with medical management, complex inflammatory masses require detailed evaluation and may require surgical resection.

Gastrostomy: Some children with ongoing additional nutritional requirements will require the formation of a gastrostomy and placement of an appropriate feeding device.

2.7.3 Specific Surgical Indications for UC

Ulcerative colitis: In contrast to operations for CD, colectomy for UC can eliminate the inflammatory disease, but may lead to other sequelae. Therefore, timing of operative intervention for paediatric UC should take into account symptom severity, quality of life issues, surgeon experience and patient/parent preferences. Operative management of UC is aimed at removing the colon and rectum with preservation of anal sphincter integrity and function. The most commonly performed operation for paediatric UC is total proctocolectomy with subsequent ileoanal pouch (J-pouch) anastomosis. This is generally carried out as a two or three stage procedure.

Acute severe colitis/toxic megacolon: Unlike adults, children with UC usually present with extensive disease and have a higher incidence of ASC, both at presentation and during follow-up. The management of ASC is addressed elsewhere (Section 2.3 above).

Chronically active disease/steroid dependency: Failure to maintain remission despite optimal medical treatment is an indication for colectomy. Optimal medical therapy may include ASA, immunomodulators and biologic therapies. The development of steroid dependency may be a further indication for colectomy, particularly if there is evidence of significant steroid toxicity.

Severe gastrointestinal bleeding: Haematochezia requiring transfusion is relatively common in severe paediatric UC. Failure to control bleeding may be an indication for emergency colectomy.

3. MONITORING OF CHILDREN WITH IBD

3.1 General review

Children with IBD should be reviewed regularly to assess progress. In most instances, this will involve review at least every 3 months. However, many children with unstable disease will need much more frequent review.

3.2 Blood testing

3.2.1 Regular blood tests

Many children will require regular blood tests. The frequency of blood tests will vary accordingly to clinical status and current medical therapy. Children on an immunosuppressive drug should have routine full blood count and liver chemistry every 3 months. All children on medical therapy will require at least annual bloods.

3.2.1 Annual screening bloods

Annual blood tests are recommended for all patients to screen for micronutrient deficiency, to document levels of inflammatory markers and detect side-effects of therapy (**Table 5** and **Appendix 13**). Completion of these on the anniversary of diagnosis or at the time of the child's birthday can help to ensure that these are completed each year.

Table 5 – Annual blood tests

FBC
Liver chemistry
Urea, creatinine and electrolytes
ESR
CRP
Vitamin D
B12, Folate, Iron
Ca, Mg, PO
Thiopurine metabolites (6-TGN and 6MMP) – if relevant

3.3 Annual review

All children and adolescents with IBD should have an annual review of key aspects of their progress (**Appendix 12**). In many instances, children will need much more regular attention to some of these aspects.

3.4 Growth

Weight and height should be measured at each review consultation, with plotting on appropriate growth charts. Calculation of weight for height scores and/or BMI should also be completed. In addition, the calculation of weight, height and BMI z scores (standard deviation scores) helps to place these parameters within the context of the wider population. Historical height trajectory and mid-parental height should be added to the child's growth chart at diagnosis.

Bone age determination is recommended at diagnosis especially in adolescents with apparent impaired linear growth. Subsequent annual repeat assessment of bone age is recommended, especially in those with delayed bone age initially.

3.5 Diet

Children should have regular review of their diet. Formal dietetic assessment should be conducted at the time of annual reviews, but may also be required more frequently.

General recommendations will be for a healthy balanced diet in children with IBD. A small number of children may require specific dietary changes (such as a low fibre diet in those with obstructive symptoms). Any dietary changes should be undertaken in conjunction with and under the supervision of a paediatric dietitian.

3.6 Bone Mineral Density

Particular indications for assessment of bone mineral density include children with significant steroid exposure, history of fractures, or persistent severe disease. In some children, a baseline assessment at diagnosis will be indicated.

Bone mineral density assessment should be undertaken within a unit experienced in the interpretation of bone densitometry in children. Results will need to be interpreted using age-appropriate normal ranges, and be adjusted for height and bone age.

3.7 Vaccination Status

The vaccination status of children should be ascertained at diagnosis. Annual review should provide an opportunity to review this, especially in younger children. Live vaccinations are contraindicated in children receiving immunosuppressive drugs (steroids, thiopurines and others).

3.8 Iron deficiency/Iron deficient anaemia

Iron deficiency is commonly seen in children and adolescents with IBD. It may reflect inadequate dietary intake, altered absorption (e.g. duodenal involvement) or ongoing enteric losses.

Parenteral iron replacement (given as an intravenous infusion) is preferred over oral iron supplementation in active disease. Oral iron supplementation should be considered only in patients in remission. If oral iron therapy does not lead to an adequate response, then parenteral iron should be administered. In those with iron deficiency, dietetic review is also required to optimise dietary intake of iron-containing foods.

3.9 Surveillance

Colitis-associated colonic cancer is rare in the paediatric population. The estimated risk of developing carcinoma with UC is approximately 1% per year after eight to 10 years of active disease: the risk for colonic CD is thought to be similar.

Endoscopic surveillance with multiple biopsies is required in patients with longstanding disease, following standard NZ Society of Gastroenterology and international guidelines. In general, children with either CD or UC should commence colonoscopic surveillance 8-10 years after diagnosis to monitor for the development of dysplasia and colitis-associated cancer. Surveillance colonoscopy should then be conducted every two years. In some specific circumstances (e.g. primary sclerosing cholangitis and colitis), surveillance should begin after 12 months of disease, reflecting the increased risks of colitis-associated cancer in this setting. The identification of high grade dysplasia is an indication for colectomy.

4. ASSESSMENT OF DISEASE ACTIVITY

Clinical scores have been developed for CD and UC to document disease activity: these can be used to define disease severity.

4.1 Paediatric Ulcerative Colitis Activity Index (PUCAI)

This score for UC is based on recent symptoms (**Appendix 11**). Symptoms should reflect the average of the preceding two days, except in rapidly changing situations where the preceding 24 hours should be used. This score varies between 0 and 85.

A score of <10 indicates remission, whilst mild disease is 10-30, moderate is 30-60 and severe is ≥65. Acute severe colitis can be defined as a PUCAI score ≥65.

4.2 Paediatric Crohn Disease Activity Index (PCDAI)

This score for CD is based on recent symptoms over the preceding week, current growth findings (since last assessment), key examination findings and three key blood test results (**Appendix 12**). This index varies between 0 and 100. Remission is indicated by a score less than 12.5: moderate disease is indicated by a score of 30 to 45, whilst a score greater than 45 signifies severe disease.

4.3 Physicians Global Assessment (PGA)

The PGA is an index based upon the assessment of an experienced paediatric gastroenterologist. It is a four point score (0=remission, 1=mild, 2=moderate, and 3=severe activity).

5. TRANSITION FROM PAEDIATRIC CARE

Transition from the paediatric setting to adult care is a critical time in the management of adolescents with IBD. Transition should be a process and not just an event (i.e. not just transfer of care).

Generally, it is felt that the transition process should be a process of preparation, with a phased introduction of concepts, leading up to the final step of transfer.

In general the commencement of the transition process should begin before the age of 15 years. Subsequent steps include discussion regarding the differences between paediatric and adult care, progressive movement of responsibility from the parents to the adolescent, and outline of the process itself. During this time, the adolescent's understanding and disease-specific knowledge should be assessed, with opportunities provided for additional education steps as required, along with ample time for questions to be answered. As adolescents go through this process, they should begin to be seen by themselves, with subsequent inclusion of the parent(s) in a summary of the consultation. The adolescent should gradually take on more responsibility for adherence to therapy, arrangements for monitoring tests, completion of repeat scripts and arrangements for follow-up appointments. In addition, time should be made for discussions about healthy living (e.g. exercise, sleep, and management of stress) and lifestyle choices (e.g. smoking, alcohol, contraception etc).

The final stage of transition is often timed to coincide with the end of secondary school (but the actual age will depend on local practices). The final stages of transfer of care would generally be considered when all requirements have been fulfilled, and at a time of stable disease control and following the pubertal growth spurt.

As the transition process proceeds, there should be discussions with the young person about the options for who takes over their care. In some centres there may be only limited options: in others there may be numerous possibilities. In general, it is preferable to transfer care to an adult gastroenterologist who is aware of the issues faced by adolescents with IBD, who is understanding towards young adults, and with whom the young person "clicks". Although the practices may differ between centres, transition should always include an initial consultation with the selected adult gastroenterologist prior to the final transfer of care. Potential ways to manage this phase include designated transition clinics, one-off joint clinics with the relevant adult gastroenterologist, or an individual "getting to know you" appointment in the adult setting. In addition, a detailed summary of the young person's progress from the time of diagnosis should be prepared and made available to the adult gastroenterologist.

6. SUPPORT AND EDUCATION

6.1 Initial education and introduction to IBD and orientation to the treating team

In general, standard education processes after diagnosis of IBD would involve two or more sessions with the child and their parent(s) during which the key concepts of IBD are outlined, and management principles are outlined.

The child and parents should have ample time to ask questions and clarify concerns or worries. Appropriate written resources should be provided at this time. In addition, links to appropriate online resources should be mentioned.

Following these introductions to IBD, the child and their family should be made aware of the roles of key members of the management team. This will include outlining the expected roles of the paediatrician and paediatric gastroenterologist. In addition, the roles of allied health staff should be outlined, with introductions to key staff (e.g. dietitian) as required. The completion of applications for appropriate allowances should also be considered at this time.

6.2 Ongoing education

Ideally, the child and their family should have regular opportunities to attend further education meetings or presentations. Outpatient reviews, especially annual review appointments, should include the opportunity to confirm disease-specific knowledge and to provide time for the young person (or the parents) to ask questions.

6.3 Support

Generally young people benefit greatly from interaction with others with IBD, especially those of a similar age. Opportunities for such interaction may come from local support groups, or from informal and formal peer-support activities. Although a number of children will be hesitant to meet others soon after diagnosis, the offer of peer-support should be made at that time and subsequently. Joining Crohns and Colitis NZ will be another mechanism to provide support and information. (**Appendix 14**)