REVIEW

Inflammatory bowel disease

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Twenty five per cent of inflammatory bowel disease presents in childhood. Growth and nutrition are key issues in the management with the aim of treatment being to induce and then maintain disease remission with minimal side effects. Only 25% of Crohn's disease presents with the classic triad of abdominal pain, weight loss, and diarrhoea. Most children with ulcerative colitis have blood in the stool at presentation. Inflammatory markers are usually although not invariably raised at presentation (particularly in Crohn's disease). Full investigation includes upper gastrointestinal endoscopy and ileocolonoscopy. Treatment requires multidisciplinary input as part of a clinical network led by a paediatrician with special expertise in the management of the condition.

Twenty five per cent of inflammatory bowel disease (IBD) presents in childhood, often in adolescence, and usually as Crohn's disease (CD) or ulcerative colitis (UC). Family history is common. The diseases run a chronic relapsing course, with a significant disease burden particularly during the adolescent growth spurt. Growth and nutrition are key issues in the management with the aim of treatment being to induce and then maintain disease remission with minimal side effects. The treatment priority thus is not only symptom control and quality of life, but also to ensure that disease control is sufficient to facilitate normal growth and pubertal development.

This article reviews some of the current issues in children with IBD and provides practical guidance on the investigation and management.

EPIDEMIOLOGY AND PATHOGENESIS

UK incidence

Between June 1998 and July 1999, 5.2/100 000 children less than 16 years of age in the UK and Republic of Ireland were newly diagnosed as being affected by IBD.¹ Sixty per cent had Crohn's disease, the remainder being ulcerative colitis (28%) and indeterminate colitis (12%). The incidence has increased over the last 30–40 years.²

Aetiology

The aetiology of IBD is not known. However, it is likely that genetic predisposition, environmental factors, and immune dysfunction are all important. Most believe intestinal bacterial flora is vital for persistence of the inflammatory process.³

The genetic basis of the disease has long been implied by the high prevalence of disease within

families (see below). Numerous environmental factors have been implicated; these include cigarette smoking (or passive smoking) (protective for UC, risk factor for CD), appendectomy (protective for UC, risk factor for CD), breast feeding (protective), diet (high fat/sugar), perinatal or early childhood infections, wild type measles infections, MMR vaccine, *Mycobacterium paratuberculosis*, and oral contraceptives.²

Ethnicity

While IBD appears to be more common in northern latitudes, recent studies have suggested this difference has been overestimated. In the UK it has been reported that the proportion of children with UC is higher in South Asian than Caucasian patients.^{1,4}

Family history and genetics

Familial occurrence in IBD is well established and is mostly explained by genetic factors. Both diseases can occur in the same family. In CD, monozygotic twins have 44–58% disease concordance, while only 0–3.8% concordance is seen in dizygotic twins;^{5,6} this is less strong in UC (monozygotic twins 14–19%, dizygotic twins 0–5%).^{6,7}

The lifetime risk of Crohn's disease for first degree relatives is 7% in white, non-Jewish populations, but 16.8% in white, Jewish families.⁸

As most environmental triggers are not directly relevant in studies of paediatric onset IBD, it remains likely that genetic factors outweigh environmental ones. Polito *et al* reported that 30% of those presenting before 20 years of age had a family history of IBD, compared with only 14% of those presenting over age 40.⁹

IBD loci

Genome wide linkage studies have identified seven IBD susceptibility loci, including chromosomes 16q (IBD1), 21q (IBD2), 6p (IBD3), 14q (IBD4), 5q (IBD5), 19p (IBD6), and 1p (IBD7).¹⁰ Some loci are more specific for UC (for example, IBD2), and some for CD (for example, IBD1), although there is a clear need to replicate studies. These susceptibility loci may encode hundreds of genes, hence complicating the identification of specific genes and their function.

NOD2/CARD15

After the publication of the human genome, Crohn's disease was one of the first diseases to identify disease susceptibility genes. Seminal studies in 2001 identified the gene encoding NOD2 as being important in white but not Asian

Abbreviations: CD, Crohn's disease; IBD, inflammatory bowel disease; UC, ulcerative colitis

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patients.^{11–12} NOD2 has now been renamed CARD15 (Caspase and Recruitment Domain) and is on chromosome 16q (IBD1 locus). Compared with 8–15% of controls, 10–30% of Caucasian patients with CD are heterozygous for one of the three common variants of CARD15; 3–15% are homozygotes or compound heterozygotes compared to 0–1% of controls. Homozygous or heterozygous carriage of the common variants of CARD15 has been associated with early onset, ileal and stricturing CD.^{13–15} It remains unclear how CARD15 increases the susceptibility to CD, although it has known functions in bacterial recognition, apoptosis, and inflammatory signalling (via NFκB).

Disease modifying genes

While some genes influence disease susceptibility, others affect only the course of the disease. Mutations in the IL-6 genotype are associated with an increased incidence of growth failure in Crohn's disease, but do not increase susceptibility to development of the disease.¹⁶

Advances in the genetics of IBD have yet to lead to significant changes in management or treatment. However, research continues on genotype–phenotype interactions, hoping to identify predictors of disease severity and treatment efficacy.

CLINICAL FEATURES

The signs and symptoms of an affected child will vary depending on the severity and chronicity of the condition, the segments of the gastrointestinal tract involved, whether the inflammatory process is mucosal or transmural, the presence of complications such as strictures or fistulae, and whether there are any extra-intestinal sequelae.

Clinical features: Crohn's disease

The variable and frequently non-specific nature of clinical presentation in children with Crohn's disease contributes to the delay between the onset of symptoms and the diagnosis. The majority complain of abdominal pain (72%), with only 25% presenting with the “classical triad” of abdominal pain, weight loss, and diarrhoea. Many children with Crohn's disease present in a “non-classical” manner with vague complaints such as lethargy or anorexia which may be associated with just mild abdominal discomfort. Other symptoms are fever, nausea, vomiting, growth retardation, malnutrition, delayed puberty, psychiatric symptoms, arthropathy, and erythema nodosum. These extra-intestinal manifestations may dominate the clinical presentation in children, causing diagnostic difficulty and delay in diagnosis.

It is essential that physical signs which could point to a diagnosis of inflammatory bowel disease are sought in all cases with these symptoms. There may be signs of pallor (from anaemia), clubbing, cutaneous involvement with erythema nodosum or pyoderma gangrenosum, or arthritis. Oral disease may manifest as lip swelling, aphthous ulceration, or gingival hyperplasia. There may be localising signs on abdominal palpation such as tenderness or a mass in the right iliac fossa. Examination of the perineum may reveal skin tags, fistulae, or abscesses. Recent weight loss may be evident from the child's growth chart, but growth failure may well be more difficult to ascertain at diagnosis without good quality historical growth data.¹⁷

Clinical features: ulcerative colitis

In children with ulcerative colitis, blood loss (84%), diarrhoea (74%), and abdominal pain (62%) are all common.¹⁷ Weight loss was less common in UC (35%) than CD (58%), as were other features suggesting systemic involvement such as anorexia or lethargy. The most commonly reported extra-intestinal symptom was arthropathy (10%). This survey also

reported that children with a diagnosis of indeterminate colitis had predominantly colitic symptoms.

The physical examination of a child with ulcerative colitis is frequently unremarkable. Mild lower abdominal discomfort (left iliac fossa) with signs of anaemia or arthritis (classically sacroileitis) are sometimes present. Where pain is more prominent, particularly if associated with systemic signs of inflammation, toxic dilatation of the colon must be suspected. This is a life threatening emergency and requires urgent medical intervention, and close liaison with experienced surgeons as colectomy may be required.

Osteoporosis

Osteoporosis predisposes to fractures. Children with Crohn's disease have reduced bone density compared to normal controls¹⁸ and children with ulcerative colitis.¹⁹ The reported prevalence is variable (11–50%).^{19–21} This variability may partly be explained by the inherent difficulty in interpreting DEXA scans with limited normal paediatric data and variable height, weight, and pubertal status.

Total bone mass accumulates rapidly throughout puberty and adolescence, finally peaking at around 35 years of age. Disruption of this crucial process inevitably leads to a reduction in peak bone mass, with subsequent increased risk of fractures in later life. The aetiology of reduced bone mineral density is multifactorial, but includes corticosteroid use, release of cytokines (for example, IL-6), and a reduced intake of calcium and vitamin D. Significant corticosteroid exposure is a major factor increasing the fracture risk in adults²² and children.¹⁹

INVESTIGATION

Initial screening blood tests should include a full blood count, erythrocyte sedimentation rate (ESR), C reactive protein, serum urea and creatinine, serum albumin, liver function tests, and in certain instances a coeliac antibody screen. Characteristically inflammatory bowel disease is associated with a fall in haemoglobin and albumin, and a rise in platelet count, ESR, and C reactive protein levels.²³ These findings are however not always present, with UC having been reported with a normal haemoglobin, platelet count, and ESR.²⁴ Anti-saccharomyces cerevisiae antibodies (ASCA) and anti-neutrophil cytoplasmic antibodies (ANCA) may be elevated in Crohn's disease and ulcerative colitis respectively, with a diagnostic sensitivity between 60% and 80%.^{25–26}

The differential diagnosis is wide (box 1). Infectious causes of enteritis or colitis need to be excluded. Given the rise in tuberculosis in the UK this should be considered.

A full colonoscopy with intubation of the terminal ileum, and multiple biopsies of all segments of the colon (plus terminal ileum) is now considered essential by most paediatric gastroenterologists.²⁷ This approach increases the diagnostic yield and improves the diagnostic certainty when compared with a more limited recto-sigmoidoscopy.²⁸ For the same reasons an upper endoscopy has also now been advocated (IBD working group of ESPGHAN 2005). The macroscopic and histological appearances of the upper gastrointestinal tract (for example, the presence of aphthoid ulcers or giant cell granulomas) may confirm the diagnosis of Crohn's disease in up to 25% of cases.²⁹ Crohn's disease can affect any part of the gastrointestinal tract, and a patchy inflammatory process is common, whereas in ulcerative colitis inflammation is usually localised to the colon with inflammation typically extending proximally from the rectum, although rectal sparing has been reported.³⁰

Assessment of the small bowel is traditionally considered to be best performed radiologically with a barium meal and follow through, or by enteroclysis with intubation of the duodenum. This can identify small bowel pathology beyond

Box 1: Important differential diagnoses to consider during diagnostic work up of inflammatory bowel disease

- Infection
 - Campylobacter
 - Salmonella
 - Shigella
 - Yersinia
 - Amoebiasis
 - Tuberculosis
 - Clostridium
 - Giardia
- Eosinophilic gastroenteritis
- Vasculitis and autoimmune conditions
- Cows' milk/allergic colitis
- Immunodeficiency states (e.g. chronic granulomatous disease)
- Solitary rectal ulcer syndrome
- Benign lymphoid hyperplasia
- Intestinal lymphoma

the reach of the endoscope and better define the disease extent and presence of complications such as strictures or fistulae.³¹ If endoscopic and histological evidence allow a definitive diagnosis to be made, contrast studies may not be necessary, but are always indicated in children with diagnostic uncertainty (for example, failure to intubate the terminal ileum, indeterminate colitis) or suspicion of proximal small bowel disease, long segment ileal disease, or stricture.

Several other methods of assessing, in particular the small bowel, which involve less or no radiation exposure have been reported, although as yet none has achieved universal acceptance. For example capsule endoscopy may become more widely used in cases where a stricture has been excluded.³² This approach cannot however replace endoscopy since histological assessment is required. Gadolinium enhanced magnetic resonance imaging or magnetic resonance imaging enteroclysis can also be used to assess the small intestine.³³ Ultrasound is another attractive option since it is often more readily available, avoids irradiation, and may reveal colonic or ileal (particularly terminal ileal) wall thickening or inflammation.³⁴ It may however miss mild inflammatory changes. Technetium labelled leucocyte scintigraphy may also identify intestinal inflammation, but there are concerns as to its sensitivity and specificity.³⁵

MANAGEMENT

This has been extensively reviewed by previous authors both from the paediatric and adult perspective, including evidence based reviews, Cochrane reviews, and management guidelines.^{36–40} There is widespread acceptance that the published evidence base is poor, with no available evidence based or consensus based guidelines in children. Much of paediatric practice is adapted from adult evidence, small paediatric studies, and the experience of paediatric gastroenterologists. The management strategy as outlined below reflects practice in the authors' units.

It is now recognised that children with IBD should be managed in centres led by a paediatrician with special expertise in the condition, with IBD being listed as a specialist condition in the National Specialised Services definition set.⁴¹ The care should be provided as part of a

clinical network,⁴² with regular audit, case discussion, and shared-care clinics. A key function of such networks is protocol development.

A successful network should allow rapid access for assessment and management of children in a setting with appropriate support services. Multidisciplinary team support is essential in managing these children and their families successfully, in particular to optimise their nutritional status, bone health, and growth, but also to address factors such as school attendance, psychological status, and preparation for transition of care to adult services.

Appropriate arrangements need to be in place for transition, involving both adult and paediatric teams, over a period of time allowing the young adult to get to know the adult team prior to transfer.

The general principles of the management of paediatric IBD are shown in box 2.

GROWTH FAILURE

Growth failure and nutritional impairment is more common in CD than UC at diagnosis, although in the longer term growth impairment is seen in both groups.

Growth failure is characterised by delayed skeletal maturation and delayed onset of puberty. Achieving disease remission results in improved growth and nutritional status, allowing a reasonable expectation of achieving final adult height. Particular care must be taken to provide appropriate therapy during the mid-pubertal growth spurt.^{43–44} Impressive increases in height velocity, with catch-up growth, are seen particularly following surgical resection.⁴⁵

The aetiology of growth failure is multifactorial and poorly understood, but poor nutritional state, systemic consequences of gut inflammation, disturbances of the growth hormone/insulin-like growth factor axis, and corticosteroid use are all implicated.

Accurate growth should be monitored every 3–4 months, with careful pubertal staging occurring at least every 6 months. Growth expectations must be seen in context of the mid-parental height. There needs to be a low threshold to reinvestigate if there is impaired linear growth and to optimise therapy using steroid sparing strategies if possible.

In the child with impaired linear growth there is a need for close collaboration with paediatric endocrinology which is

Box 2: General principles of the management of paediatric IBD

- (1) Provide a rapid, accurate, and expert diagnostic service
- (2) Provide timely, sensitive, and patient relevant disease information
- (3) Establish care within an MDT including surgical, dietetic, and nursing expertise
- (4) Institute effective therapy to achieve prompt disease remission
- (5) Minimise drug adverse effects and optimise nutritional status at all times
- (6) Maintain effective disease remission, aiming to achieve mucosal healing
- (7) Monitor growth and pubertal development
- (8) Ensure clearly defined pathway to access relevant expertise
- (9) Facilitate psychosocial/educational support
- (10) Prepare for appropriate transition to adult medical services

facilitated by joint assessment in combined clinics set up to deal with difficult cases.

Children often continue to grow into late adolescence and early adult life, outside the traditional age range for paediatricians. These key issues must remain priorities during transition of care to adult gastroenterology services and remain an issue in the adult clinic in some instances, as the potential for catch-up growth may be very delayed.

Delayed growth and onset of puberty (looking younger than your peers) will have a significant negative impact on the child's psychosocial development and self-image. The transition from early (dependent on parents) to late adolescence (dependent on self) can be particularly difficult for a child with chronically active IBD.

The optimal management of growth equates to the optimal management of disease activity in childhood.

SPECIFIC MANAGEMENT: CROHN'S DISEASE

The management will depend on disease severity and extent.

Enteral nutrition

Exclusive enteral nutrition (EEN) is an appropriate initial therapy in most cases. Whole protein, polymeric nutrition given as sole therapy for 6–8 weeks will induce disease remission in 60–80% dependent on case selection.⁴⁶ Crohn's colitis without ileal involvement may respond less well. This nutritional treatment is non-toxic and steroid sparing. The administration requires the support of a multidisciplinary team in order to ensure the correct amount is given and compliance assured. Nasogastric tube feeding can be avoided in most cases. Between 110% and 120% of the recommended nutritional intake is usually required.⁴⁷ The feed is best drunk chilled and can be flavoured if necessary. Food is usually reintroduced slowly (allowing identification of possible food triggers) while weaning the EEN over 2–4 weeks at the end of the treatment course.

Continued nutritional support has a role in the ongoing management, particularly if there is a nutritional deficit and/or growth delay. Further courses of EEN can be given for disease flare-ups.

Corticosteroids

Although they have significant adverse effects, corticosteroids are as effective as EEN at inducing remission in active Crohn's disease.⁴⁸ They are an appropriate alternative treatment, and the treatment of choice in severe pancolitis. Oral prednisolone is given as 1–2 mg/kg, maximum 40 mg, for 2–4 weeks and then weaned by 5 mg/day each week as the disease enters remission. Intravenous steroids are required if there is a severe pancolitis with systemic disturbance. Supplements of calcium and vitamin D should be considered during corticosteroid therapy,⁴⁹ particularly if intake is suboptimal or insufficient. There is no role for corticosteroids as maintenance therapy.⁵⁰

Budesonide may have a role as a less toxic alternative to prednisolone, given its potential reduction in toxicity due to first pass metabolism. This has not been confirmed in large paediatric studies.

If children develop steroid dependent disease, additional immunosuppression should be used.

5-Aminosalicylates

Adult guidance suggests that systemic 5-aminosalicylate (5-ASA) derivatives are appropriate as initial monotherapy in mild to moderate disease.³⁶ Disease severity in children tends to be more severe, and either enteral nutrition or steroids are frequently used in combination with 5-ASA therapy. There are numerous preparations and dosing regimens available.^{36–51} Sulphasalazine is associated with more side effects

than newer 5-ASA preparations (for example, mesalazine), which are therefore preferable in most cases.

5-ASA derivatives are still used as long term maintenance treatment, although the evidence base in CD remains poor and dosing regimens are unclear.

Azathioprine

Azathioprine (AZA) or 6-mercaptopurine (6MP), are highly effective maintenance agents for moderate to severe Crohn's disease.^{52–53} They are indicated in frequently relapsing disease and steroid dependency, particularly if there is significant steroid toxicity. They should be considered at diagnosis in children with severe and/or panenteric disease as relapse rates are almost halved at 18 months compared to conventional therapy with corticosteroids.⁵⁴ Maximum efficacy may not be reached until 12–16 weeks after starting treatment.

Families need to be aware of potential toxicity, including bone marrow suppression and pancreatitis. Regular monitoring of full blood count and amylase is required.⁵¹ Children who become unwell on AZA need to be aware of the need to seek urgent medical assistance. Thiopurine methyltransferase (TPMT) metabolises AZA; the risk of myelosuppression is increased in those with low activity of the enzyme. TPMT status should ideally be checked before starting therapy. 6MP is sometimes helpful in children intolerant to AZA.

The need for long term maintenance therapy should be reviewed annually and may be discontinued if pubertal, psychosocial, and educational development allow.

Retrospective adult evidence suggests benefit of continuing therapy for at least four years, and the longer term use of AZA is well described in adult practice. Children on AZA should not receive live vaccines.

Antibiotics

Antibiotics (broad spectrum, for example, ciprofloxacin and metronidazole) are indicated in fulminant disease, particularly if the child is pyrexial, and mandatory if toxic megacolon is suspected. Oral metronidazole alone or in combination with ciprofloxacin is also helpful in peri-anal disease, often required for weeks or months. Long term metronidazole can cause peripheral neuropathy.

Second line immunosuppression

In children unresponsive to the above therapies, alternative immunosuppressive agents can be considered. These include methotrexate, cyclosporin, tacrolimus, and anti-TNF monoclonal antibodies (infliximab). Infliximab has been a significant advance in the management of Crohn's disease and has proven efficacy at inducing and maintaining remission. Repeated doses are frequently needed, with children requiring infusions every eight weeks to remain asymptomatic. Indications for its use in adults have been recommended by NICE, and should be extended to include children. Active disease unresponsive to immunosuppressant therapy and not amenable to surgery (that is, localised resection), or treatment resistant fistulising disease should all be a consideration for infliximab. An additional paediatric indication is severe growth failure with pubertal delay in the context of disease resistant to medical therapy.

There are still significant concerns about the long term safety and potential toxicity of infliximab. At present the drug should only be administered in a centre familiar with its use and according to a strict protocol. Antibody formation may lead to diminished response and anaphylaxis. This can be minimised by concomitant immunosuppressant use, pretreatment with corticosteroids, and regular dosing schedules.

Surgery

Surgery is indicated for either localised (for example, stricturing) disease, abscess, or disease resistant to medical

therapy. Surgery should only be considered if conventional medical therapy has failed, although impressive catch-up growth is seen following surgical resection in early to mid-puberty.

Preoperative nutritional status should be optimised and steroid dose minimised. Surgery is usually a small bowel resection, right hemicolectomy, or sub-total colectomy with ileostomy formation and preservation of the rectal stump. Postoperative recurrence risk is high, but may be reduced by the continued use of anti-inflammatories or immunosuppressives.

SPECIFIC MANAGEMENT: ULCERATIVE COLITIS

The treatment strategy for ulcerative colitis is similar, although enteral nutrition is not effective. The notes on management below emphasise the aspects of drug use specific to UC, but do not repeat the information listed under CD where similarity with UC exists.

Corticosteroids

Corticosteroids are widely used and effective in the management of UC either at diagnosis or during disease flare-ups. Dosing regimens and reducing courses are similar to CD, and aim to minimise toxicity by discontinuation after 8–10 weeks. Intravenous steroids are indicated for severe disease and fulminant colitis. Local therapy can be given for distal disease (see distal colitis). There is no role for corticosteroids as maintenance therapy.

5-ASA derivatives

Systemic 5-ASA derivatives are appropriate initial therapy in mild disease, and may be effective as single agents. Efficacy is greater with higher doses. 5-ASA derivatives are used as long term maintenance treatment with good evidence for efficacy.³⁶ Long term 5-ASA use probably protects against the development of colonic cancer.³⁶

Azathioprine

Azathioprine (or 6-mercaptopurine) is a highly effective steroid sparing maintenance agent in the management of UC.

Second line immunosuppression

Cyclosporin and tacrolimus can induce a rapid remission in a subgroup of children with acute colitis and can be used to transition patients to AZA. Infliximab, which is highly effective in active Crohn's disease, may have a role in achieving remission in acute UC.

Surgery

Subtotal colectomy and ileostomy, usually with a view to subsequent ileal pouch formation, is performed for cases where medical management is failing, as it offers a potential cure. Persistently active disease may lead to increased steroid requirements, drug toxicity, growth failure, and pubertal delay. Deteriorating quality of life frequently prompts the decision for surgery and is mostly seen as a welcome relief from severe symptoms and failing medical therapies.

DISTAL COLITIS

A similar management strategy can be adopted for distal colitis whether due to UC or CD. Distal colitis can be more resistant to therapy; systemic therapy is often required in the first instance. Topical therapy with enemas or suppositories, using either 5-ASA or corticosteroids, can be highly effective. It is important however that children and families are adequately supported in the administration technique and positioning required. Enemas are best given with the patient lying on their left side with the foot of the bed elevated and ample time given for the enema to act locally—that is, 30–45 minutes before getting up and going to the toilet.

Suppositories and small volume enemas are only effective for rectal disease.

ACUTE TOXIC COLITIS

Either CD or UC can present as toxic colitis. The management is outlined in box 3.

PROGNOSIS

There are no prospective studies of outcome in paediatric IBD. While UC can be “cured” by removal of the colon, CD cannot. Both run a chronic relapsing course with significant disease morbidity as a consequence. There is no known cure for Crohn's disease and there is a high relapse rate following surgical intervention. The majority of children with Crohn's disease have a good quality of life and final height data is encouraging (provided steroid therapy is minimised during the pubertal growth spurt).

In 1984 Puntis *et al* reported that 83% of children with Crohn's disease required surgical intervention within four years of diagnosis.⁵⁵ Freeman (2004) reported that 56.3% required surgery within a mean time of 4.2 years. The likelihood of surgery therefore remains high despite advances in treatment. The more widespread use of immunosuppressive regimens like azathioprine and infliximab over the last 5–10 years may impact on this. Emerging genetic factors may help predict those likely to have a more severe disease course and thereby allow patient specific introduction of more potent immunosuppressive regimens.⁵⁶

Polito *et al* retrospectively studied records of 552 adult patients and found greater small bowel involvement (88.7% *v* 57.5%), more stricturing disease (45.8% *v* 28.8%), and a higher frequency of surgery (70.6% *v* 55.3%) associated with a younger age of diagnosis.⁹ Freeman more recently reviewed 224 patients and has attributed increased complications in the younger group to the fact that disease duration is simply longer. His analyses showed that the incidence of strictures (28.6%) and penetrating complications (46.4%) in the younger group were similar to those in adults when both groups were followed up for a similar period of time.⁵⁶

In a retrospective case note review of 171 children with UC, Hymans *et al* (1996) found a colectomy rate of 25% at 5 years in those with moderate to severe disease at diagnosis. Disease severity at diagnosis was predictive of long term outcome.⁵⁷

Box 3: Practical management of acute toxic colitis

- *Fluid resuscitation* with saline bolus if required
- *Blood transfusion* if Hb <8 g/dl
- *Intravenous fluids*—initially 0.45% saline/dextrose with 20 mmol KCl per 500 ml modified depending on fluid balance and electrolyte results
- *Intravenous hydrocortisone*—10 mg/kg in four divided doses; maximum 100 mg qds
- *Broad spectrum intravenous antibiotics* if pyrexial
- *Plain AXR*—if toxic megacolon (colonic dilation), repeat every 12–24 hours or if clinical deterioration
- *Ultrasound scanning of the abdomen* to document bowel wall thickening
- *Surgical review* (at presentation) as up to 50% with toxic megacolon will require colectomy. Most with an acute toxic colitis (not megacolon) will not require surgery

Once the child is stable fluids and light diet can be allowed

CONCLUSION

Twenty five per cent of IBD presents in childhood, often during adolescence. Clinical features are not always classical. Growth failure is common. The morbidity in terms of disease course and treatment toxicity is high. The challenge for paediatricians is to support children with IBD through childhood and adolescence for transfer to adult services. Diagnosis and treatment require a multidisciplinary approach, best led by centres with expertise in the management of these children. This is best provided in the form of clinical networks, enabling the patients to access most of their healthcare needs near their home. Like many paediatric conditions the evidence base for management is limited and multicentre research initiatives with consensus based guidelines for treatment would benefit children and adolescents with this condition.

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