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Clinical Topics

Chronic inflammatory bowel disease in childhood

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Abstract

The diagnosis of Crohn's disease in childhood has been facilitated by the use of fiberoptic endoscopy with biopsies, complemented by double-contrast radiology. Clinical suspicion leads initially to several relevant blood tests. These are followed by endoscopy and multiple colonic biopsies or barium follow-through studies depending on whether large-bowel or small-bowel disease is suspected. The present approach to diagnosis is based on corroborative investigative techniques—endoscopy, radiology, and histology. The availability of paediatric colonoscopes of small diameter should make it possible for paediatricians to perform limited examinations, but when more extensive endoscopy is indicated the child should be referred to special centres.

Introduction

Until recently Crohn's disease was thought to be uncommon in children. This may be because systemic manifestations dominate the early clinical picture¹ and many paediatricians in the past have been reluctant to embark on all the procedures necessary for diagnosis. Children, especially those aged from 2 to 8, tolerate proctosigmoidoscopy and rectal biopsy badly except under general anaesthesia. The air contrast barium

enema necessary for accurate assessment of inflammatory changes is uncomfortable, and most paediatric radiologists rely on the inferior single-contrast method. Either technique gives an appreciable dose of irradiation. Inadequate results from these existing methods of examination are in our view now superseded by a single fiberoptic endoscopy with biopsies. This is complemented when necessary by high-quality double-contrast radiology adapted to paediatric use to show inflammatory changes. These techniques permit accurate and early diagnosis of chronic inflammatory bowel disease—that is, Crohn's disease or ulcerative colitis—in childhood. We present the current approach to the management of a child with chronic inflammatory bowel disease at St Bartholomew's Hospital.

Clinical assessment

The early symptoms suggestive of Crohn's disease in childhood² (based on a previous study from our unit) are non-specific—lethargy, abdominal pain, and chronic diarrhoea. Nevertheless, at diagnosis, often many months or even years later, more symptoms are present, including, in decreasing order of frequency, loss of weight, abdominal pain and diarrhoea, lethargy, slow growth and delayed puberty, variable bowel habits with constipation, and psychological problems.

In the previous study physical examination at the time of diagnosis showed growth retardation with weight below the third percentile, perianal abnormality (anal fissures, perianal skin tags, and anal fistulae), clinically obvious anaemia, abdominal tenderness, and erythema nodosum. In some children with Crohn's disease no abnormality may be found.

In contrast, the early symptoms of children with ulcerative colitis are more specific—diarrhoea, bleeding per rectum, abdominal pain, tenesmus, anorexia, and loss of weight. Most children do not have any physical signs, but pallor is fairly common and abdominal tenderness is present. Psychological symptoms and failure to grow are uncommon.

Investigative approach

The flow diagram illustrates the current approach to investigations. In a child with any clinical features suggestive of

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chronic inflammatory bowel disease the first step is to exclude specific infections—for example, amoebiasis, yersinia enterocolitis, shigellosis, salmonellosis, and tuberculosis—by conventional stool microscopy and culture, serological investigations for amoeba, salmonella, and yersinia, and a Mantoux test for tuberculosis.

Haematological investigations include measurement of full blood count; erythrocyte sedimentation rate; acute-phase protein, such as C-reactive protein; serum iron and total iron-binding capacity; concentrations of serum and red-cell folate, serum vitamin B₁₂, total proteins, immunoglobulins; and a liver function test.

Classically a child with Crohn's disease has an anaemia characteristic of a chronic disorder with a haemoglobin concentration below 11 g/dl, a raised erythrocyte sedimentation rate and C-reactive protein concentration, a low serum iron concentration and low to normal total iron-binding capacity, and low red-cell folate concentration. All these investigations, however, may show normal results.

Further investigations at this point will depend on whether the clinician considers that the clinical features suggest small-intestinal or large-intestinal disease, or both (see flow diagram). Small-bowel disease is suggested by recurrent abdominal pain and shortness of stature, while chronic bloody diarrhoea with anal abnormality is more diagnostic of large-bowel disease.

Large-bowel disease

All patients with symptoms suggestive of a colitis or when inflammatory bowel disease is strongly suspected have colonic endoscopy and biopsy performed as the procedure of first choice, as this will immediately provide the clinical diagnosis on anatomical grounds of ulcerative colitis or Crohn's disease without having to expose the child to unnecessary irradiation. Colonoscopy not only permits assessment of colonic and rectal disease with biopsies but also allows definition of the terminal ileum with ileal biopsy.

BOWEL PREPARATION

Preparation of the bowel for colonoscopy is simple but important.³ No food is allowed for 24 hours before colonoscopy. Only clear fluids should be taken. A senna syrup (X-Prep) at a dose of 1 ml/kg body weight is given 24 hours before the examination. On the morning of the colonoscopy one or two large-volume tap-water enemas are given depending on results. Oxyphenisatin (Veripaque) 0.75–1.5 g/l water may be added to the second enema.

An alternative method is possible, on the morning of the colonoscopy, using 5% mannitol solution taken four to five hours before the procedure. This may be preferable in the presence of diarrhoea. Metoclopramide (not exceeding 0.5 mg/kg body weight) is given an hour before 500–1000 ml of 5% mannitol is taken by mouth. This is followed by liberal amounts of fluids in the form of water or flavoured drinks. Though bowel preparation with mannitol is less traumatic to the child, it may be difficult to get smaller children to drink all of the solution.

SEDATION

Droperidol (5–10 mg) or pethidine compound elixir (0.5 ml/kg/dose) is given by mouth as premedication one hour before the colonoscopy. Before the procedure in the colonoscopy room, a butterfly needle is inserted to give intravenous diazepam emulsion (Diazemuls) 5–10 mg, which is less painful than the ordinary preparation, and intravenous pethidine 1–2 mg/kg/body weight. General anaesthesia is not used, but the sedation is titrated until the child is extremely drowsy.

ENDOSCOPY TECHNIQUE

Colonoscopy is easier in children than in adults provided that a gentle technique is used to avoid loop formation. An adult colonoscope can be used for all except small babies, though paediatric instruments are available. Examination to the caecum and terminal ileum is possible in over 90% of children, and transillumination of the thin abdominal wall makes localisation easy.

The endoscopic characteristics of Crohn's disease are small aphthoid ulcers or sometimes deeper ulcers in an otherwise normal mucosa. In childhood ulcerative colitis, generalised abnormality with reddened and friable mucosa and loss of normal vasculature is usual. Biopsy specimens are, however, always taken because differentiation is not possible in every case.

If Crohn's disease is diagnosed on endoscopy or histologically a barium follow-through study is necessary to confirm or refute accompanying small-bowel disease.

Small-bowel disease

If Crohn's disease affecting the small intestine is considered likely a barium meal and follow-through should be performed.

Early minor abnormalities can be shown on barium studies only if the examination is technically adequate for confident radiological appraisal. Small-bowel meals are often of poor quality, as not enough barium is given, with the result that the small bowel is underfilled, transit is slow, especially if no agent is given to increase the rate of transit, and flocculation of the barium suspension may occur; compression studies are seldom performed, so that overlapping loops may obscure early changes. Compression studies allow better definition of the distal small bowel and may help distinguish between Crohn's disease and lymphoid nodular hyperplasia, which is common in childhood.

RADIOLOGICAL TECHNIQUE

The following technique for small-bowel studies is recommended. After an overnight fast the child is given 5–10 mg of metoclopramide by mouth about 10 minutes before the barium. A non-flocculent barium (Baritop G) is taken by the child, volume according to weight (10 ml/kg/body weight). The patient is placed on the right side to aid gastric drainage and prone films are taken 10, 30, and 50 minutes later. Spot films with manual compression of the abdomen with a lead glove are obtained of the distal ileal loop or any suspicious segments.

Abnormalities typical of Crohn's disease include aphthoid ulceration or longitudinal ulcers, cobblestoning, stricturing, and pseudodiverticular formation. Skip lesions are common, but these may be diffuse and often there is a mixture of changes.

If the small bowel is shown to be abnormal radiologically colonoscopy is the next examination of choice to provide histological confirmation by biopsy of the terminal ileum, and to show any colonic disease.

Histology

Ideally, the histological picture should be the final arbiter of the nature of an inflammatory bowel disorder, but this can be true only when a surgically resected specimen is available for examination. When the pathologist is restricted to examining small mucosal biopsy specimens differential diagnosis between inflammatory bowel disorders cannot rest on histological findings alone because of the limited nature of the sample. Differentiation between inflammatory disease and normal is, however, always possible. There is no single histological feature that is diagnostic for Crohn's disease or ulcerative colitis, though the presence of non-caseating granulomas is important in diagnosing Crohn's disease.

Flow diagram of investigation in chronic inflammatory bowel disease in childhood.

The histological diagnosis of inflammatory bowel disease is therefore cumulative and based on groups of features; aggregate histological features being placed in a spectrum of certainty ranging from definite Crohn's disease to definite ulcerative colitis.⁴

HISTOLOGICAL CRITERIA

The criteria for diagnosis of Crohn's disease are focal inflammation, submucosal or transmucosal inflammation, retention of mucus, and lymphocyte aggregates without germinal centres, together with non-caseating granulomas or fissuring ulceration. In ulcerative colitis there is acute inflammation of diffuse distribution with mucus (goblet-cell) depletion in the acute phase and pronounced crypt distortion or mucosal atrophy in the inactive phase.

Follow-up

On follow-up of children with either Crohn's disease or ulcerative colitis, colonoscopy may be performed to define disease activity and complications and provide preoperative evaluation of the child in severe disease. The instant (Barium) enema may be useful, however, particularly in ulcerative colitis, to define the extent and severity of the colonic disorder and to show up fistulae and strictures, and may be necessary when it is not possible to perform total colonoscopy. Usually only one film is needed to record all the necessary diagnostic information. The examination causes minimal disturbance, discomfort, and irradiation to the child.

INSTANT ENEMA

Instant enema⁵ is a well-proved technique in adult radiology and provides a double-contrast examination of the affected colon without preparing the bowel. Barium at 100% weight/volume is introduced into the rectum via a soft rubber catheter until a residue is encountered or the splenic flexure is reached. The rectum is drained and then air is gently insufflated to obtain an adequate double-contrast view. A single prone film is usually sufficient to visualise the colonic abnormality.

Clinical results

During 1972-81, 81 children with chronic inflammatory bowel disease were seen at St Bartholomew's Hospital. According to the protocol (see flow diagram) 80% of the children had colonoscopy and biopsy as the procedure of first choice for large-bowel disease (chronic bloody diarrhoea with anal abnormality) while the remaining children had a barium meal and follow-through study performed initially for predominant small-bowel symptoms (recurrent abdominal pain and shortness of stature).

Of the 81 children, 54 were diagnosed as having Crohn's disease, 17 ulcerative colitis, and a further 10, with definite chronic inflammation on colonic biopsy, indeterminate colitis.

Conclusion

There are great rewards for thorough investigation of possible inflammatory bowel disease at an early stage, particularly in paediatric practice. Children show loss of weight, and often systemic features of inflammatory bowel disease occur more quickly than in adults, and earlier disease is easier to treat. The flow diagram summarises the approach that we have adopted. Clinical suspicion leads initially to several relevant blood tests. If their results are abnormal, endoscopy and multiple colonic biopsies or barium follow-through studies usually lead to a clinical diagnosis. In practice, the "final" clinical diagnosis of Crohn's disease rests on a combination of the three major diagnostic investigative techniques—endoscopic, radiological, and histological—as no one single means of investigation may be regarded as diagnostic.⁶ Until the fundamental aetiology of the two important inflammatory bowel diseases (Crohn's disease and ulcerative colitis) is known firm diagnostic criteria will be difficult.

Our present approach to the diagnosis of chronic inflammatory bowel disease in childhood is based on investigative techniques that are well tolerated by children and in combination give a definite diagnosis (or exclusion) in the minimum time. Nevertheless, the routine examination of the whole colon and the terminal ileum by colonoscopy needs practice and experience that will not be acquired by paediatricians outside a few special centres.

The recent availability of paediatric colonoscopes of small diameter should, however, make it possible for paediatricians interested in gastroenterology to perform limited examinations. Even fibre-proctoscopy and certainly fibre-sigmoidoscopy should be clinically valuable and often allow diagnostic colonic biopsy specimens to be taken. When more extensive examination is indicated the child should be referred to a paediatric gastroenterology centre or to a gastroenterologist who is skilled in colonoscopy in children. It is important that paediatricians should have easy access to such diagnostic techniques, which, in future, will need to be more widely available.

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