Henoch-Schönlein purpura: problems in surgical diagnosis and management

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SUMMARY
The clinico-pathological features of 133 consecutive cases of Henoch-Schönlein purpura are presented, with emphasis on the gastrointestinal manifestations. The potential pitfalls of contrast radiography are underlined with respect to management of intussusception and a plea is made to re-establish clinical assessment of the abdomen as the prime indicator in deciding to undertake laparotomy.

INTRODUCTION
In 1837 Schönlein described what he called 'peliosis rheumatica', a purpuric rash associated with arthritis, and 37 years later Henoch added his classic description of a rash associated with colicky abdominal pain, gastrointestinal haemorrhage and arthritis. Now, almost 150 years later, Henoch-Schönlein purpura is the term given to a syndrome characterised by the typical purpuric rash affecting the lower extremities associated with gastrointestinal, joint and renal involvement. The aetiology is unknown, and, although allergies to drugs (particularly antibiotics), some foods, or a hypersensitivity reaction to the beta-haemolytic streptococcus have been suggested, nothing has been proven. Pathologically the characteristic lesion is a vasculitis affecting the small vessels and this gives rise to the various manifestations of the syndrome.

Surgical intervention is required only in a minority of cases even when gastrointestinal symptoms are severe. However, the decision to operate or to continue with conservative management is often difficult. This review of cases at the Royal Belfast Hospital for Sick Children was undertaken to clarify and correlate the spectrum of gastrointestinal symptoms and pathology.

MATERIALS AND METHODS
Only those cases where the consultant in charge had classified the final diagnosis as Henoch-Schönlein purpura were included in this study. The patients' age, sex, date of admission, point of onset of illness and discharge were noted. The presence or absence of the typical skin rash involving the buttocks and the extensor surfaces of the lower limbs, joint pain and swelling, haematuria or proteinuria were recorded, and also the past medical history, family history and

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Henoch-Schönlein purpura details of recent antibiotic administration. The specific gastrointestinal symptoms of abdominal pain, vomiting, diarrhoea and gastrointestinal bleeding were detailed.

RESULTS
Data were collected for the period 1972 – 1985 inclusive. A total of 133 patients were studied. There were 80 males and 53 females, a male : female ratio of 3 : 2. Ages ranged from one to 13 years with a mean of six years. The age incidence is presented in the Figure. Length of hospital stay was less than one week in 60 (45%) cases, 1 – 2 weeks in 35 (26%) cases and more than two weeks in 37 (28%) cases.

![Figure - Age Incidence](image)

In the two weeks prior to admission 53 (40%) of cases gave a history of upper respiratory tract infection and 33 (25%) cases had been prescribed antibiotics, 82% of which were of the penicillin type. The typical rash was present in 129 (97%) cases, an atypical rash in three, and in one there was no rash. In 47 (36%) cases, the rash appeared after the onset of abdominal pain. Joint pain and/or swelling was recorded in 107 patients (80%). In 73 (55%) only the lower limbs were affected. Isolated upper limb involvement was present in seven (5%), and in 53 (40%) both upper and lower limbs were involved.

Haematuria (overt or microscopic) or proteinuria were taken as evidence of renal involvement, and 52 patients (40%) showed these features. Haematuria was present in 23 (17%), proteinuria in 33 (25%) and 77 (58%) had both. In 84 (63%) of these patients, renal manifestations had resolved by the time of discharge, but, in the remainder, haematuria and/or proteinuria persisted for a variable period. Three patients developed renal failure. One, a six-year-old boy three months after the acute illness, did not require dialysis but he still shows biochemical evidence of renal impairment six years after the acute illness. Another, a 10-year-old boy, required dialysis and had a successful renal transplant. A third died with a nephrotic syndrome three months after his acute illness.

The profile of abdominal symptoms is presented in the Table. Abdominal pain and vomiting were by far the most common gastrointestinal problems. Melaena or haematemesis were uncommon, occurring in 26 (20%) cases, and 47 (35%) had other abdominal signs and symptoms without any evidence of gastrointestinal

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Gastrointestinal tract symptoms

| Symptoms     | Number of patients |
|--------------|--------------------|
| Pain         | 58 (44%)           |
| Vomiting     | 46 (35%)           |
| Melaena      | 14 (10%)           |
| Haematemesis | 12 (9%)            |
| Diarrhoea    | 8 (6%)             |

Total number of patients with gastrointestinal symptoms 73 (56%).

bleeding. Three children developed signs of intestinal obstruction and required laparotomy. Two were aged six years and one aged three years. They all had moderate to severe abdominal pain with vomiting; two had melaena, none had haematemesis. At operation two were found to have ileo-ileal intussusception and the third had an ileal submucosal haematoma. No other cases of intussusception were identified and no other children required surgery. Only three children received steroids as part of their therapy.

There were two deaths, but only one was directly related to Henoch-Schönlein purpura. The other occurred many years later from an unrelated cause.

DISCUSSION

Henoch-Schönlein purpura as an entity is essentially a clinical syndrome, there being no diagnostic laboratory test. The characteristic rash usually precedes abdominal pain but in our series the rash appeared afterwards in 36% of cases giving rise to obvious diagnostic difficulties. Despite this, no patient had an unnecessary laparotomy.

The cause of abdominal pain in this condition has always been assumed to be from involvement of the gastrointestinal tract. However, whilst 45% of those with abdominal pain had no evidence of kidney involvement, 73% of those with renal manifestations had abdominal pain without obvious gastrointestinal disease. Involvement of the kidneys may be responsible, at least in part, for the 'abdominal pain' associated with Henoch-Schönlein purpura in a significant percentage of cases.

The basic pathological process is a vasculitis resulting in perivascular oedema and haemorrhage. Involvement of the bowel, particularly the small bowel, may be of sufficient degree to cause a haematoma in the wall which may then give rise to perforation, subacute obstruction, intussusception or haemorrhage into the lumen.1-7 The incidence of obstruction and intussusception is difficult to assess from the literature as so few reported series are on unselected patients. In this study, there were two cases (approx. 2%), as compared with reported incidences of up to 10%.1,5,6

Severe abdominal pain was present in 15% of cases in this study. Only 2% came to laparotomy, so pain per se is not necessarily an indication for surgery. More important are the signs and symptoms of intestinal obstruction or of peritonitis, which are relatively uncommon in this disease.5
Henoch-Schönlein purpura

A straight radiograph of the abdomen is mandatory in all children with evidence of intra-abdominal pathology. However, diagnostically and therapeutically, barium studies in Henoch-Schönlein purpura are of limited value, as 50% of intussusceptions in this condition are ileo-ileal. This contrasts with 90% of all childhood intussusceptions which are ileo-colic, and although it is possible to reduce an ileo-ileal intussusception hydrostatically, we feel that it would probably be inappropriate to attempt this manoeuvre in the presence of obstruction and co-existing vasculitis of the gastrointestinal tract. Computerised axial tomography has also been used in the evaluation of the abdomen in Henoch-Schönlein purpura, and perhaps its most valuable future role would be accurately to distinguish a submucosal haematoma from an intussusception. As yet, computerised tomography has not superseded the existing methods of assessment.

Earlier series reported an unacceptably high mortality and morbidity associated with surgical intervention. This was probably caused by delay in laparotomy, which almost certainly stemmed from a reluctance to operate: firstly, because surgical complications were relatively common; and, secondly, because steroid therapy theoretically places patients at an increased risk of infection and poor wound healing. The role of steroids in contemporary management is now limited and their use should not cause any delay in the decision to operate. Although the incidence of significant surgical lesions in Henoch-Schönlein purpura is less than 5%, the morbidity in this group is high; we feel that with careful attention to clinical detail, it should not be necessary to perform laparotomy in the 10 – 15 per cent of those with more severe abdominal pain which has been suggested by other authors.

Contrast abdominal radiography may be misleading or inconclusive in Henoch-Schönlein purpura, and we recommend that laparotomy should be undertaken without delay when a child with this condition develops clinical signs which would normally indicate surgery. Attributing such signs to an 'ileus' or submucosal haematoma could have disastrous consequences in allowing intestinal gangrene, perforation or septicaemia to develop.

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