and that a total dosage of 22·5 g might be sufficient for cure. It has the advantage that it can be given orally on an outpatient basis.

The adult flukes live in the biliary system of sheep and cattle, ova being excreted in the host's fæces. These ova hatch out into miracidia, which seek out and penetrate the host snail, in Britain usually the species *Lymnæa truncatula*. After asexual multiplication the parasite emerges from the snail as a minute tadpole-like creature, a cercaria which encysts on waterside vegetation. This is the form infecting mammals, in whose gut the young flukes develop to invade the liver and biliary system.

An increase of snail population dependent on warmth and moisture predisposes to outbreaks. Last year's high rainfall in the west midlands was followed by an unusually high incidence of fascioliasis in animals.

A working party of the Public Health Laboratory Service (1966) was set up to study the hygienic conditions under which watercress was grown. They commented that in general watercress appeared to be a safe and wholesome food, but that watercress growing wild should not be sold for human consumption.

Acknowledgment: We are grateful to Mr P H Everall FIMLT for his detailed technical assistance at all stages.

REFERENCES
Dawes B (1966) *Nature (Lond.)* **209**, 424
Lienert E (1962) *Z. Tropenmed. Parasit.* **13**, 338
Public Health Laboratory Service (1966) *Mth. Bull. Minist. Hlth Lab. Serv.* **25**, 146
Swiēzawska E, Kaliszewicz S & Grott J W (1967) *Wiad. Parazyt.* **13**, 707

Dr A W J Houghton (*Shrewsbury*) said that watercress beds should be supplied with running water from sources known to be free from the possibility of fluke infestation. The chief points in the diagnosis of the disease were fever, enlargement and tenderness of the liver coupled with eosinophilia. There was usually a history of having eaten wild watercress. The disease very often 'went a begging' for a diagnosis and was probably commoner, especially in country areas, than was generally supposed.

Ehlers-Danlos Syndrome

David Harland FRCS
(*Luton and Dunstable Hospital, Luton*)

Miss C W F, aged 17

Past history: Born three weeks prematurely with left talipes equinovarus. Has always tended to bruise very easily.

History: In June 1966 she developed sudden severe abdominal pain and vomiting and, on admission to hospital, had the signs of pelvic peritonitis. It was also noted that she had numerous bruises and scars, particularly over her legs, arachnodachtyly with hypermobility of the fingers and wrists, blue sclera and a high arched palate.

Operation (12.6.66): A localized perforation of the sigmoid colon was found and repaired and a transverse colostomy was performed. This was subsequently closed.

July 1968: She again developed sudden severe lower abdominal pain and was admitted with the signs of pelvic peritonitis. Laparotomy (19.7.68) revealed a further perforation at the apex of the sigmoid colon; this was closed and a left iliac colostomy was performed. On 29.11.68 a total colectomy and ileorectal anastomosis was performed. Apart from a severe secondary hæmorrhage, she made an uneventful recovery from this operation and is now very well.

Family history: The patient's father was also born prematurely and is said to have closely resembled his daughter in facial appearance and the length of his fingers; he also had a tendency to bruise easily, so that he was never able to play games. At the age of 33 he was admitted to hospital with acute abdominal pain and at laparotomy was found to have a perforation of the sigmoid colon. He died on the second postoperative day.

Discussion

The spontaneous recurrent colonic perforations at the ages of 13 and 15, the appearance of the patient and the history of the father's death from colonic perforation suggest a heritable connective tissue disorder, and from the clinical point of view Ehlers-Danlos syndrome and Marfan's syndrome seem to be the most likely. Beighton & Horan (1969) and Barabas (1967) have both mentioned spontaneous perforation as complications of the severe form of the Ehlers-Danlos syndrome, and Cook (1968) has recorded a family with Marfanoid stigmata in which two unaffected members died of spontaneous colonic perforations in their 20s, one of them having had a previous perforation in childhood.

The absence of specific bacteriological, biochemical and radiological features, the possibility of overlap of one of these syndromes with the other, and the fact that dislocation of the lens is not a *sine qua non* of the Marfan syndrome, make it difficult to give an exact label to this case. It is, however, believed that she falls into the category of the severe form of the Ehlers-Danlos syndrome in spite of the absence of hyperelasticity of the skin and the fact that at operation the tissues were not of the 'wet blotting paper'

variety previously described. Undoubtedly she also has associated Marfanoid features.

The decision to perform a total colectomy was not undertaken lightly or without expert proctological advice, but it is hoped that, with the resulting looser fæcal content, the small intestine will be less likely to perforate than was the large. This operation will give no immunity from a major arterial catastrophe, which would seem to be a distinct possibility.

REFERENCES
Barabas A (1967) MD thesis, Manchester
Beighton P & Horan F T (1969) *Brit. J. Surg.* 56, 255
Cook J M (1968) *Ohio St. med. J.* 64, 73

Mr A Barabas (*London*) said that total colectomy in this patient was fully justified, as recurrent perforation of the colon invariably took place.

The prematurity of both the patient and her father deserved attention. This was due to premature rupture of defective fœtal membranes. Because premature infants were now successfully reared, more patients with severe forms of inherited connective tissue diseases might be seen in the future.

Professor Harold Ellis (*London*) said it was interesting that the scar on the patient's knee following an old injury had stretched widely – this was a typical feature of Ehlers-Danlos syndrome.

Klippel-Feil Syndrome

G H Whitehouse[1] MRCP DMRD
(for R J Harrison FRCP)
(*St James' Hospital, London*)

Mr J S, aged 28. Scaffolder

History: August 1967, complained of six episodes of transient loss of consciousness over previous nine months, occurring without warning or apparent precipitating factors, and lasting from a few seconds to two minutes, with a subsequent rapid recovery. In addition, he had for the past nine months noticed a tendency to trip, especially on mounting the curb. His left arm had never swung as freely as his right when walking, and he had for some years noticed difficulty in standing on one leg.

A cleft palate had been repaired in infancy. He had always been noted to have a short neck with restricted head movements. Radiographs of the neck taken in adolescence for weakness of the shoulder muscles revealed an 'abnormality of the neck'.

On examination: The neck was shortened and the hairline was low with some webbing of the neck (Fig 1). All movements of the neck very restricted. Increased dorsal kyphosis. The only abnormality in the limbs was a slight weakness of the left shoulder and elbow movements, and weakness of all muscle groups in the left leg. He did not swing the left arm on walking, tended to fall to the left on closing his eyes, and was unable to stand on one foot.

Investigations: Radiographs (Fig 2) of the cervical spine showed fusion of the bodies and arches of all vertebræ, except at C5–6, where degenerative changes were present, and at C1, which remained separate and without evidence of occipitalization. Thoracic spine showed upper dorsal scoliosis with two hemivertebræ at this level and fusion of upper ribs on each side. The skull, which showed no basilar impression, and the lumbar spine were normal. Lumbar puncture, right carotid angiogram and air encephalogram normal. Myelography showed considerable, but not complete, obstruction to the passage of myodil at the level of uppermost disc below the fused vertebræ, i.e. C5–6, especially in extension, with extradural pressure mainly on the lateral and posterior aspects and only a slight impression anteriorly. The cord was of normal diameter.

Operation (December 1967, Professor V Logue): The fused vertebral mass was found to have three fused spinous processes and a lowermost bifid spine which led to two hemilaminæ, that on the right being fused to the mass and that on the left being fused with the upper border of the

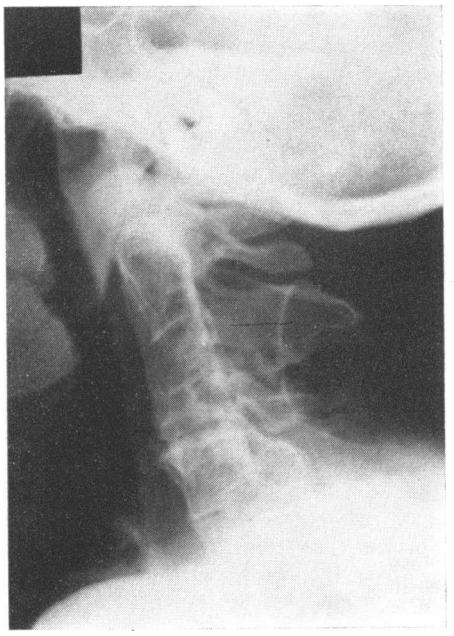

Fig 1 *Lateral tomogram to show fusion of 2nd–5th cervical vertebræ and degenerative changes in C5–6 disc space*

[1]Present address: Bristol Royal Infirmary, Bristol