

and intermyenteric plexuses. Serial sectioning failed to show any ganglion cells. The suction biopsy diagnosis of Hirschsprung's disease was confirmed in every case at full thickness biopsy.

In one further case, normal cholinergic nerve fibres in the lamina propria and muscularis mucosae were seen at suction biopsy and no ganglion cells could be identified in the small amount of submucosa present in the biopsy specimen. The clinical course, however, demanded further investigation. Serial sections of full thickness specimens sampling the whole colon, including the ileocaecal junction, failed to show any ganglion cells, but the nerve trunks in the submucosal and intermyenteric plexuses were normal. A diagnosis of total aganglionosis of the colon (Zeulzer-Wilson syndrome) was made.<sup>6</sup>

### Discussion

Suction rectal biopsy has been used as a screening method for patients with symptoms and signs suggestive of Hirschsprung's disease in this department for six years. It has proved to be a more accurate screening method than radiology and/or anorectal manometry. There were no false negative or false positive diagnoses in 17 cases of Hirschsprung's disease, but full thickness biopsy was required to make the diagnosis of total aganglionosis in one case of non-Hirschsprung's constipation.

A review of the anorectal manometry on some of the patients shows the typical lack of reflex inhibition seen in Hirschsprung's disease but repeated suction and full thickness biopsies failed to show any

histological abnormality. These are a 'physiological' or 'pseudo-' Hirschsprung's disease and respond well clinically to sphincterotomy. It is therefore essential that any patient suspected of having Hirschsprung's disease on anorectal manometry be submitted to rectal biopsy to confirm the diagnosis.

In the screening of patients with chronic constipation for Hirschsprung's disease, suction biopsy is as accurate as full thickness biopsy. It should be accompanied by anorectal manometry for more detailed analysis. Providing facilities for histochemistry are available, full thickness biopsy need have no further place as a screening method.

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## Acquired toxoplasma encephalitis

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**SUMMARY** Toxoplasma was the cause of encephalitis in a 4 year old boy. He recovered completely after treatment with pyrimethamine and sulphadimidine. Toxoplasma encephalitis has a high mortality, and active treatment is recommended.

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In childhood encephalitis it is unusual to discover a treatable cause. I report a case of encephalitis in which there was serological evidence of a toxoplasmic aetiology.

### Case report

A 4 year old boy was well until October 1983 when he had a generalised convulsion. Three weeks later he began to have recurrent convulsions, which increased in frequency, and he became ataxic. On transfer to Sheffield in mid-December he seemed to be fully orientated, but had gross truncal ataxia, bilateral intention tremor, and dysarthria, although no nystagmus. There was no evidence of raised intracranial pressure, cranial nerve palsy, or limb weakness. He was receiving sodium valproate and phenytoin.

Table *Toxoplasma* serology results

| Date             | Reciprocal titres |      |      | Specific IgM<br>ELISA† (EIU‡) |
|------------------|-------------------|------|------|-------------------------------|
|                  | Latex             | HA*  | Dye  |                               |
| 15 December 1983 | 2048              | 1024 | 2048 | 0                             |
| 23 January 1984  |                   |      | 1024 | 38 (Positive)                 |
| 13 April 1984    | 256               |      |      |                               |

\*HA=Haemagglutination antibody.

†ELISA=Enzyme linked immunosorbent assay.

‡EIU=Enzyme international units.

There was no abnormality on computed tomography or examination of cerebrospinal fluid. An electroencephalogram in October had been normal, but a further electroencephalogram now showed disintegration of the background rhythm with generalised bursts of spike activity, suggestive of encephalitis. Investigation revealed no evidence of a neurodegenerative disorder or metabolic abnormality. Blood concentrations of anticonvulsants were below normal therapeutic ranges. The only positive finding on paired viral serology was a rise in adenovirus titre from 1/10 to 1/40. *Toxoplasma* serology was as shown in the Table.

Before the first toxoplasma result and drug concentrations were known, it was suspected that he was suffering from drug toxicity. In view of his severe condition and frequent convulsions, therefore, previous drugs were withdrawn and prednisolone 40 mg/day and phenobarbitone were started, producing a dramatic reduction in convulsions and ataxia within three days. As the dose of prednisolone was reduced, however, both convulsions and ataxia recurred, and the convulsions began to assume a myoclonic nature. For several days he was drowsy with almost continuous myoclonic jerking. On receipt of the initial toxoplasma serology result, treatment with pyrimethamine, sulphadimidine, and calcium folinate was begun. After two weeks of this treatment he began to improve and then rapidly returned to normal, although some myoclonic convulsions continued until clonazepam was added.

After six months, anti-toxoplasma treatment and anticonvulsants were withdrawn, and he has remained well. Electroencephalograms after three and a half months' and six months' treatment showed progressive improvement, although some right sided predominance remained.

## Discussion

Acquired toxoplasmic encephalitis in childhood was

first reported in 1941.<sup>1</sup> In recent years toxoplasmic encephalitis has become a recognised opportunistic complication of a variety of immune deficient states,<sup>2</sup> but of 45 published cases of acquired central nervous system (CNS) toxoplasmosis reviewed by Townsend *et al* in 1975, 22 had no known pre-existing immune deficiency.<sup>3</sup>

The manifestations of CNS toxoplasmosis vary considerably, from non-specific encephalopathy or diffuse meningoencephalitis to features of space occupying lesions.<sup>3</sup> Cerebrospinal fluid pleocytosis is not invariable.<sup>4</sup>

In this case the high initial toxoplasma latex titre with subsequent fall, the conversion of the specific IgM ELISA test during the illness, and the response to anti-toxoplasma treatment provide convincing evidence of an acute toxoplasma infection, which was probably the cause of the encephalitis. The rise in adenovirus titre was probably related to a minor intercurrent illness.

The usual course of acquired toxoplasmosis without CNS involvement is spontaneous recovery, but published reports suggest that when the CNS is involved there is a high mortality.<sup>3,4</sup> We recommend, therefore, that evidence of toxoplasma infection be sought in every case of unexplained encephalitis, and that, when found, the condition should be treated actively with pyrimethamine and a sulphonamide. Calcium folinate may be given simultaneously to prevent haematological side effects of pyrimethamine. Although steroids were given in this case before diagnosis, in toxoplasmosis they may be deleterious and should be avoided. There are no clear guidelines on duration of treatment, but it was decided to treat this boy for six months, as recommended by Luft *et al*,<sup>4</sup> and no adverse effects of treatment were observed.

I thank Dr B L Priestley and Dr J M Davies for permission to report this case, which was under their care.

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