

Suction biopsy in Hirschsprung's disease

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SUMMARY Seventy two children with symptoms and signs consistent with Hirschsprung's disease had full thickness and suction rectal biopsies performed. Results were identical with both methods, except for one case of total aganglionosis of the colon. Full thickness biopsy no longer has a place as a screening method.

In the management of chronic constipation Hirschsprung's disease may be diagnosed on the basis of radiological investigation, anorectal manometry, and rectal biopsy. Though x ray films and manometric studies may be highly suggestive, the definitive diagnosis requires rectal biopsy.^{1 2}

The classical method of rectal biopsy involves taking a full thickness of rectal mucosa and underlying muscle, requiring general anaesthesia and suturing of the biopsy wound. The possible complications necessitate inpatient management, and its morbidity precludes its use as a screening procedure.

The introduction of suction rectal biopsy of mucosa and submucosa represents a considerable advance.^{3 4} Biopsy requires neither general anaesthesia nor suturing, may be carried out even on outpatients, is suitable for all ages, and rarely leads to complications.

Several teams have reported false negative results when compared with full thickness biopsy.⁵ We investigated over 2000 children with chronic constipation and suspected Hirschsprung's disease and performed suction biopsies on 400 of them. To assess the validity of the suction results we are reviewing all the cases (72) in which full thickness biopsies were also carried out, either at a formal resection or at upper partial sphincterotomy.

Patients and methods

Suction biopsy specimens were taken from 72 patients aged from a few weeks to 17 years. They were suspected of having Hirschsprung's disease. Some biopsy specimens were taken without anaesthesia and others under general anaesthesia just after manual evacuation, anorectal manometry, or anal dilatation. Biopsy specimens were taken from the posterior wall of the rectum at intervals of 2, 3,

4, and 5 cm from the anal verge with a Quinton multipurpose biopsy machine.

These patients either came to formal bowel resection or, on failing to improve after repeated anal dilatations, received a therapeutic extended upper partial internal sphincterotomy, during which a full thickness biopsy specimen was taken from the edge of the wound before suturing the mucosa.

All biopsies, full thickness or suction, were treated in the same manner. After orientation, they were frozen in Iso-pentane cooled in liquid nitrogen before 8 μ sections were cut on a cryostat. Sections were stained with a modified Karnovsky-Roots technique³ and also with routine haematoxylin and eosin stains. The tissues were then fixed in 10% formol saline, and serial sections of paraffin blocks were examined for the presence of ganglion cells. In some of the suction biopsy specimens in which enough submucosa was included ganglion cells could be seen in some of the sections.

A diagnosis of Hirschsprung's disease was made if larger than normal nerve fibres in increased numbers were seen in the lamina propria and muscularis mucosae at suction biopsy. At full thickness biopsy not only were the nerve fibres in the lamina propria and the muscularis mucosae assessed, but the presence or otherwise of ganglion cells and the size of the nerve trunks in the submucosal and intermyenteric plexuses were also assessed. All sections were examined by the same histopathologist without reference to previous biopsies.

Results were correlated independently by another person, and no patients who had had both sorts of biopsy were excluded.

Results

Seventy two patients were analysed. Of these, 54 showed normal nerve fibres on suction biopsy. In eight serial sectioning showed ganglion cells in the submucosa. Full thickness biopsy specimens showed normal nerve bundles and ganglion cells in the submucosal and myenteric plexuses. In 17 cases, cholinesterase staining showed an increased size and number of cholinergic nerve fibres both in the lamina propria and the muscularis mucosae at suction biopsy. In addition, full thickness biopsy specimens showed the typical hypertrophied nerve trunks of Hirschsprung's disease in the submucosal

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.⁶

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.

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.