

# ON THE ÆTIOLGY OF HIRSCH- SPRUNG'S DISEASE.

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The signs and symptoms of congenital dilatation of the colon are well enough recognized to require no elaboration in this short communication, the purport of which is to describe a pathological basis for the disease and to report the results of the histological examination of two examples. I can find no reference to such histological changes as are recorded here in the relevant literature.

Both cases investigated were in boys, aged 10 and 7 years respectively, and were typical in every respect. The second case is described here.

#### *Macroscopic Examination.*

The colon is distended from the cæcum to the pelvi-rectal sphincter; its width being five times that of the sphincter. The wall above the sphincter is about four times the thickness of the wall at the sphincter, and the enlargement is chiefly confined to the circular muscular coat.

#### *Microscopic Examination.*

(a) *Through the distended area.* The mucosa, the muscularis mucosæ, and the submucous layers all show some fibrous thickening; the circular muscular fibres are greatly hypertrophied, the longitudinal to a lesser degree. The cells of Auerbach's plexus appear healthy but some fibrous change is apparent in their surroundings.

(b) *Through the sphincter.* No changes of importance are noted in the epithelial layer and glands, the muscularis mucosæ, or the submucous layer, although a few inflammatory cells are found throughout. The circular muscle coat is in sharp contrast to the area above and is of normal thickness, as also is the longitudinal layer, and shows little alteration apart from some fibrous and fatty degeneration. In the intermuscular plexus of nerve cells the changes are of a striking character. As the accompanying microphotographs (Figs. 1 and 2) show, the ganglia are replaced by inflammatory cells. In the ganglion which is shown under the higher magnification, only one nerve cell, shrunken and degenerated, is to be seen.

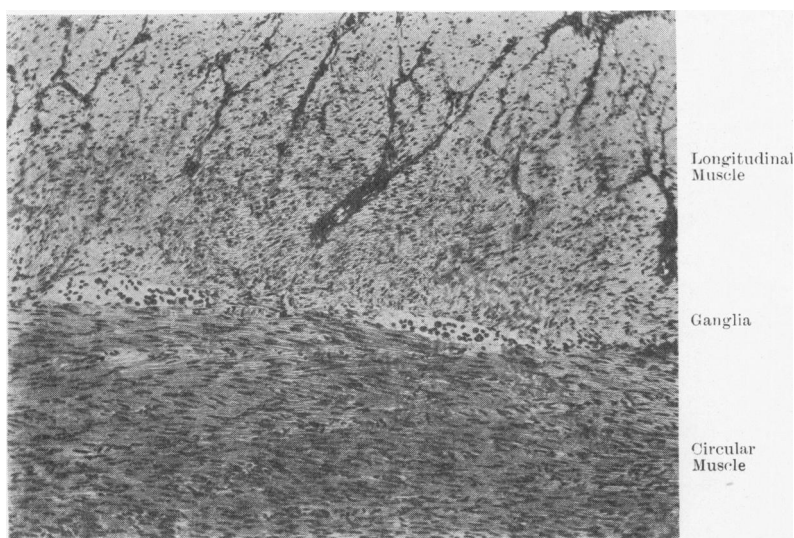


Fig. 1. Low power microphotograph through pelvi-rectal sphincter,

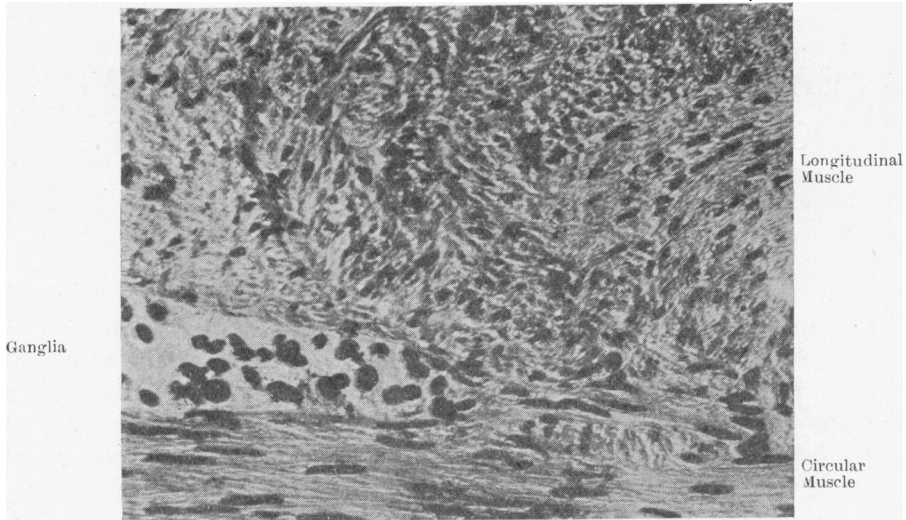


Fig. 2. High power microphotograph of ganglion in Auerbach's plexus.

A destructive lesion of the nerve ganglia at the pelvi-rectal sphincter will act as a direct obstacle to the descending peristaltic waves. Above it stasis of the contents will take place, and in an attempt to expel the accumulated and unaccustomed burden, the wall and especially the principal contracting part of it, the circular muscle layer, will hypertrophy.

The initial cause of the condition may be found in some inflammatory lesion of the rectum or anus, but as most of these cases are congenital, the probability is that meconium, retained in the relatively long and lax pelvic colon of the newly born, undergoes bacterial decomposition and sets up inflammatory changes in the mucosa. The inflammatory cells in their outward spread involve the ganglia, and an interruption of peristalsis results. Once a balance is struck between the distending bowel and the hypertrophy of its wall, a return to normal conditions is attempted, and at the sphincter the mucosa and muscular layers may recover from the inflammatory process, while the more highly organized layer of nervous tissue fails.

Hirschsprung's disease is therefore exactly comparable in its pathological basis to cardiospasm, as Hurst on clinical grounds has previously suggested. Although the lesion that I have recorded here is comparatively of old standing, it is so similar to the first cases of cardiospasm I described, that I have little doubt that if a sufficiently early case of megacolon is examined the results will be the same as in a case of acute cardiospasm published last year, in this journal<sup>1</sup>, and the complete line of invasion by inflammatory cells will be found stretching from the epithelium to Auerbach's plexus.

It is not my province to deal with the surgical treatment of this condition, but so generally successful has dilatation of the cardia proved in cases of cardiospasm, that one naturally feels that similar dilatation of the pelvi-rectal sphincter might be given a reasonable trial.

#### REFERENCE.

1. Cameron, J. A. M., *Archiv. Dis. Child.*, Lond., 1927, II, 358.