Simplified Method for Diagnosis of Hirschsprung's Disease*

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The purpose of this report is to describe a practical, inexpensive method which is capable of reproducing the results of a technique, previously described for the diagnosis of Hirschsprung's disease (Tobon *et al.*, 1968). The procedure is a simplification of the earlier method for recording rectosphincteric reflexes and can be performed in the physician's office or the patient's home.

Methods and Materials

Pressures are recorded from the internal anal sphincter by a double balloon device tied around a hollow steel cylinder forming two separate compartments (Fig. 1). With both balloons deflated, this device is gently inserted into the anus. The internal balloon is inflated with 10 ml. air through a three-way stopcock and kept inflated by closing the stopcock. The recording device is then gently pulled caudad until resistance is met. At this point, the external balloon is inflated with 10 ml. air and its stopcock is closed. The recording device will then be in the correct position and remain there. A third balloon is inserted 5 cm. beyond the recording device through its hollow core. Rectosphincteric reflexes are initiated by transiently distending the rectal balloon by rapidly injecting and withdrawing 10–50 ml. air via a hand syringe. The time required to depress and withdraw the plunger of the syringe is approximately two seconds. Pressure changes are recorded by an aneroid manometer, which is attached to the stopcock of the inner balloon (Fig. 2). When the stopcock is opened to provide communication with the manometer, pressure recordings of 50–90 mm. Hg indicate that the balloon is in position.

Studies were made on 6 subjects (age 19 months to 80 years) with functional constipation and 3 subjects (14 months to 7 years) with Hirschsprung's disease proven by biopsy, and the results of this simplified method were compared to those obtained by the pressure transducers† and Sanborn recorder‡.

Results

In normal subjects and patients with functional constipation transient distension with 50 ml. air produces an immediate relaxation of the internal sphincter which can be measured by the manometer (Fig. 3) as a pressure decrease. The pressure decrease averaged 8 mm. Hg by direct manometry

FIG. 1.—Schematic diagram of pressure sensing device. The internal sphincter surrounds the internal sphincter balloon. The external sphincter balloon is surrounded by the subcutaneous bundle of the external sphincter and helps to anchor the device in place. Rectosphincteric reflexes are initiated by transient distension of the rectal balloon.

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† Sanborn Model 267B.

‡ Sanborn Model 964.

FIG. 2.—Device for recording internal sphincter pressure by aneroid manometer. The manometer is in communication with the internal balloon. The stopcock to the external balloon is closed after this balloon is inflated. Rectosphincteric reflexes are initiated by transient distension of the rectal balloon via the syringe.

FIG. 3.—Normal internal sphincter response. Internal sphincter relaxation induced by transient rectal distension (at the arrow) is recorded on the electronic recorder as pressure decrease in the internal balloon. Resting pressure is arbitrarily assigned a zero value. Pressure increases (contraction) are designated as positive and decreases (relaxation) as negative. Pressure recordings from the aneroid manometer (upper readings) correlate well with pressures recorded by the transducer and electronic recorder (first tracing).

and 10 mm. Hg by the pen-recording technique. The pressure decrease recorded by either technique varied in a given patient by ± 2 mm. Hg with repeated distensions. Internal sphincter relaxation does not occur in patients with Hirschsprung's disease (Tobon *et al.*, 1968), but, instead, a contraction is sometimes seen (Fig. 4). The pressure changes associated with this contraction are recorded faithfully by the aneroid technique (Fig. 4 and Table).

Discussion

Constipation associated with megacolon is frequently encountered, especially in paediatric practice; and functional constipation (psychogenic megacolon) is often difficult to differentiate from Hirschsprung's disease. Until recently the diagnosis has required full thickness biopsy of the rectum and search for ganglion cells, but now idiopathic or psychogenic megacolon can be differentiated from Hirschsprung's disease by manometric techniques (Tobon *et al.*, 1968). By these techniques both normal subjects and

FIG. 4.—*Abnormal internal sphincter responses in Hirschsprung's disease. Rectal distension (at the arrow) fails to induce internal sphincter relaxation. Instead a contraction is seen in the first 2 distensions. Internal sphincter contraction, when present, occurs in the absence of external sphincter contraction as seen after the second rectal distension. In the third distension the internal sphincter failed to relax but did not contract. Pressure recording from aneroid manometer (upper readings) correlate with pressures recorded electronically (first tracing).*

patients with psychogenic megacolon respond to rectal distension with reflex relaxation of the internal sphincter (Tobon *et al.*, 1968; Schuster *et al.*, 1965). This reflex is absent in Hirschsprung's

disease (Tobon *et al.*, 1968; Howard and Nixon, 1968). Therefore, patients showing normal responses need not be subjected to the hazards of full thickness biopsy, and only those patients with an abnormal response require further investigation.

We have used two sizes of balloons and hollow steel cylinders depending upon the patient's age. Both devices are 95 mm. long. The larger device has a steel core with an outer diameter of 12 mm. and an inner diameter of 11 mm. When inflated with 10 ml. air the internal balloon has a diameter of 31 mm. and the external balloon a diameter of 12 mm. at its apex and 31 mm. at its base. The smaller device with an outer diameter of 5 mm. is used for patients under 3 years of age. 30 ml. air has been found to be the optimal amount for rectal distension in infants and children under 3 years and 50 ml. for patients above 3–4 years of age.

Since one or two test distensions may be required for the recording device to assume automatically its proper position, the first two rectal distensions should be ignored. For valid results the patient must remain quiet for 15–20 seconds from the beginning of rectal distension. Sedation is useful in the young uncooperative patient, since a straining movement with contraction of the buttocks may be recorded as pressure increase in the internal balloon. Phenobarbitone sodium (4 mg./kg.) or chloral hydrate may be used for sedation without altering the normal reflex relaxation of the internal sphincter.

The external balloon primarily serves to anchor the recording apparatus in the proper position in the anal canal, and recording from this balloon is unnecessary since the external sphincter is not abnormal in Hirschsprung's disease.

All of the patients with biopsy-proven Hirschsprung's disease showed the absence of reflex internal sphincter relaxation, which is characteristic of Hirschsprung's disease (Tobon *et al.*, 1968; Howard and Nixon, 1968). One of these patients showed the internal sphincter contraction sometimes seen in this disorder. Both the failure to relax and the contraction were equally well recorded by the electronic recorder and the aneroid manometer, and our results indicate that the simplified method is as reliable as the electronic technique for the diagnosis of Hirschsprung's disease.

Summary

A simple, inexpensive technique is described for recording rectosphincteric reflexes by an aneroid manometer. This test provides a rapid,

TABLE

Comparison of Electronic and Aneroid Techniques

Case No.	Age (yr.)	Electronic Recording (mm. Hg change)	Aneroid Manometer (mm. Hg change)
A. Functional constipation			
1	22	–12	–12
2	25	–12	–12
3	45	–8	–10
4	80	–12	–10
5	1½	–7	–5
6	74	–6	–5
B. Hirschsprung's disease			
7	1½	0	0
8	½	+5	+5
9	1½	+3	+5

Note: Average pressure changes in internal sphincter balloon are given for each patient. Negative values represent pressure decrease associated with sphincter relaxation, and positive values represent pressure increase accompanying sphincter contraction. There is close correlation between results obtained by the two recording techniques.

easily interpretable method for the diagnosis of Hirschsprung's disease, and can be performed on an out-patient basis.

REFERENCES

- Howard, E. R., and Nixon, H. H. (1968). Internal anal sphincter: observations on development and mechanism of inhibitory responses in premature infants and children with Hirschsprung's disease. *Arch. Dis. Childh.*, **43**, 569.
- Schuster, M. M., Hookman, P., Hendrix, T. R., and Mendeloff, A. I. (1965). Simultaneous manometric recording of internal

- and external anal sphincteric reflexes. *Bull. Johns Hopk. Hosp.*, **116**, 79.
- Tobon, F., Reid, N. C. R. W., Talbert, J. L., and Schuster, M. M. (1968). Non-surgical test for the diagnosis of Hirschsprung's disease. *New Engl. J. Med.*, **278**, 188.

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