HIRSCHSPRUNG'S DISEASE ASSOCIATED WITH IMPERFORATE ANUS*

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The association of Hirschsprung's disease with various other congenital anomalies is well known. The combination reported hereunder is, however, comparatively uncommon.

Report of a Case

An 8 lb. 10 oz. male child with a high type of imperforate anus was operated by us within seven hours of birth, on 16th November, 1970. A one-stage abdomino-perineal pullthrough procedure was carried out. At operation the bowel was found to be grossly dilated and the meconium was grapelike in character. This dilatation was believed to be due to discontinuity of the bowel. The post-operative period was uneventful and when he was discharged from the hospital, the child possessed a good sphincter control.

After about 4 months of the operation, the parents noticed that the child was not passing stools regularly and required the administration of laxatives. Simultaneously, they also noted a gradually increasing abdominal distension. Initially these symptoms were believed to be due to the formation of a post-operative stricture but when the symptoms did not abate in spite of regular finger dilatations, another pathology was suspected and barium studies were carried out. The barium enema revealed a picture consistent with Hirschsprung's disease (Fig. 1 Plate II) and a rectal biopsy confirmed the diagnosis.

The patient was prepared and operated on at the age of 1 year and 4 months in March, 1972. At this time, the child had a grossly distended abdomen, and the anal opening was not stenosed. The child was in good general health. At operation, the aganglionic segment was well demarcated and Soave's (1964) procedure was carried out, whereby the rectal mucosa was denuded from the pelvic brim to the anorectal line and the proximal colon was drawn through the denuded rectal cuff and was united to the upper margins of this muscular cuff by interrupted sutures. The distal end was tied around the anal tube.

On the 13th post-operative day, the patient developed a burst abdomen. On the table, this time, it was found that the obstruction involved only the small bowel, loops of which were matted together. The colon, on the other hand, was not distended at all and apparently there was no obstruction at this level.

Subsequently the patient developed respiratory tract infection and succumbed to bronchopneumonia in the 4th week post-operatively.

Review of Literature and Discussion

A case of Hirschsprung's disease associated with an imperforate anus is a rarity. This is evident by the scarcity
of case reports on the subject. In 1957, Ragins and Van Prohasky reported a solitary case. Parkkulanien et al. (1959) reported 15 such cases.

Paucity of the literature stimulated Kiesewetter et al. (1965) to collect all the available statistics from American and Canadian paediatric surgeons. In their own experience there was only one such case out of 44 cases of imperforate anus. Out of the 32 paediatric surgeons who responded to their questionnaire, 19 stated emphatically, that they had never encountered this combination of conditions. Thirteen reported on specific histopathological examination and only 6 surgeons reported having met the coincidence of these conditions and they reported 10 cases out of a total of 296 cases of imperforate anus.

Kiesewetter et al. (1965) thus felt that in Parkkulanien et al's (1959) series the high incidence needed confirmation. They felt that it should be positively ascertained that no degree of anal stenosis or stricture existed and the diagnosis should be confirmed by a biopsy taken at least 2 cm. above the mucocutaneous junction.

Thus, Kiesewetter et al. (1965) estimated an incidence of 3.4% for the association of Hirschsprung's disease with imperforate anus.

In our experience at Indore, we encountered this solitary case among 82 cases of imperforate anus operated on by us in the last 5 years. This gives an incidence of about 1.2%. However, this incidence may be fallacious as histopathological studies were not carried out in all the cases of imperforate anus.

Ehrenpreiss (1966) stated that megacolon may be produced secondary to ischaemia and in support of this belief, he cited a case of Hirschsprung's disease in whom the symptoms recurred four years after an abdomino-perineal pull-through procedure. Barium studies revealed recurrence of a narrow segment, and histopathology on the second occasion demonstrated that the ganglion cells were sparse and degenerated, while the blood-vessels had inclusions of hyaline material. He explained this pathology on the basis of a temporary obstruction of vascular supply to the distal colon because of the previous pull-through procedure.

That the case presented by us was not of such a nature can be concluded from the fact that de Villiers (1956) carried out experiments on 18 dogs, in whom he produced temporary ischaemia, and though he observed a variety of lesions, intramural ganglion cells were consistently present in adequate numbers and were of a normal structure in both the plexuses.

We are thus inclined to agree with Louw (1969) who questions Ehrenpreiss's (1966) solitary case and states that the initial transection may have been carried out through the transitional zone.

From this it appears that Ehrenpreiss's (1966) hypothesis has a doubtful validity and our case appears to have been an authentic association of the two lesions. Even if Ehrenpreiss's (1966) suggestion be accepted, histopathology, in his case, revealed degenerative changes in the ganglia present, whereas in our case there was total aganglionosis.