HIATUS HERNIA IN CHILDHOOD*

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The first description of a hiatus hernia was probably that of Bright (1836), who recorded the autopsy of a girl aged 19 years in whom a large portion of the stomach was in the chest and the cardia was at the level of the fourth dorsal vertebra. In the early part of the present century the condition was thought to be rare; Eppinger's (1911) series of 650 diaphragmatic herniae contained only 11 cases of hiatus hernia and in Quénu's (1920) series of 112 cases of diaphragmatic hernia treated surgically there were only two cases of hiatus hernia. With the introduction and development of radiology it was recognized (Åkerlund, 1926) that sliding hiatus hernia, in which the cardia and part of the stomach lie a varying distance above the diaphragm, is the most common form of diaphragmatic hernia and occurs frequently. The condition is found chiefly in the infant and in the middle-aged adult and it has generally been assumed that, in both age groups, similar factors are concerned in the aetiology and the mechanism of production of symptoms. There are, however, significant differences in the clinical features, the natural course of the disease and the response to treatment which suggest that, although the complications may be identical, hiatus hernia in the child is pathogenetically quite distinct from the majority of adult cases.

There is little doubt that infantile hiatus hernia is either present at, or develops shortly after birth; symptoms usually date from the first few days of life and the condition has been demonstrated radiologically during the first week. It was considered by Findlay and Kelly (1931) that hiatus hernia in the child is a congenital, developmental anomaly in which the oesophagus fails to lengthen synchronously with the caudal migration of the septum transversum so that the diaphragmatic hiatus comes to surround the stomach instead of the oesophagus. Botha (1958) thought that a large proportion of hiatus herniae is due to such a developmental short oesophagus. The rapid elongation of the oesophagus in the normal embryo occurs during the

sixth and seventh weeks (Keith, 1948) and a gross anomaly occurring at such an early period of development would be expected to be accompanied frequently by other major abnormalities as is oesophageal atresia which arises about this time. The evidence concerning associated malformations with hiatus hernia is somewhat conflicting, but the reports of multiple anomalies, which include hiatus hernia, have been mostly of isolated cases or of small series (Williams, 1945; Radloff and King, 1947; Barrett, 1950). In our own clinical series of 76 cases no child has a known major congenital abnormality and a low incidence of associated lesions was found by Thomsen (1955) and by Bonham Carter (1956), although mental deficiency was not uncommon in their series.

The anatomical features of hiatus hernia in the child suggest that a developmental shortness of the oesophagus is an unlikely aetiological explanation. The almost invariable presence of a sliding peritoneal sac covering the anterior and left aspects of the intra-thoracic portion of the stomach, the fact that this part of the stomach derives its vascular supply from the coeliac axis and not from the thoracic aorta, and the operative finding that the vagi are not taut and, unless involved in peri-oesophageal fibrosis, do not interfere with reduction of the hernia appear to indicate that the cardia was originally in the abdomen and only later migrated into the chest. In the acquired hiatus hernia of the adult, in which the pathological anatomy is identical with that of the child, an important aetiological factor in most cases, and the primary one in many, is relaxation of the diaphragmatic hiatus due to degenerative changes in the musculature. Barrett (1954) and Forshall (1955) have therefore considered that the essential lesion in the infantile case is a congenital weakness or laxity of the hiatus which allows herniation of the cardia shortly after birth when a negative intra-thoracic pressure develops with the onset of respiration. An objection to this hypothesis is that hiatus hernia in the child can undergo gradual spontaneous reduction (Figs. 1 and 2) and eventual cure; spontaneous recovery of

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FIG. 1.—Long, tubular hiatus hernia in child aged 3 months. Cardia at level of sixth dorsal vertebra.

FIG. 2.—Same patient as in Fig. 1, aged 18 months. Symptomless. Marked spontaneous reduction in size of hernia.

FIG. 3.—Autopsy specimen of hiatus hernia in still-born anencephalic infant. Forceps in sliding peritoneal sac

FIG. 4.—Same specimen as in Fig. 3. Stomach and cardia withdrawn from above diaphragm, demonstrating sliding sac.

a diaphragmatic weakness, if possible at all, would be unlikely to lead to the reduction of a large hernia. The autopsy finding of a hiatus hernia in a still-born anencephalic infant indicates that a pressure difference between the abdomen and the thorax is not an essential pathogenetic factor. The gross associated anomaly in this case at first suggested that the condition was a 'congenital short oesophagus' but dissection revealed the typical anatomical features of a sliding hernia (Figs. 3 and 4). As the child had never breathed, the cardia must have become displaced during pre-natal life.

An important agent in the production and maintenance of some cases of adult hiatus hernia is pressure from below, displacing the cardia upwards through the hiatus; symptomatic and even radiological cure may follow reduction of intra-abdominal pressure by the elimination of obesity, the abandonment of corsets or the evacuation of a pregnant uterus (Dutton and Bland, 1953). There is no evidence of an unduly high intra-abdominal pressure in the infant with hiatus hernia but the not infrequent association with hypertrophic stenosis of the pylorus, the phreno-pyloric syndrome of Roviralta (1952), suggests that raised intra-gastric pressure or increased gastric peristalsis may similarly force the cardia into the chest. The coincidence of pyloric stenosis in our hiatus hernia series is 10.5%. In Bonham Carter's (1956) series of 136 cases of hiatus hernia five children had pyloric stenosis and he suggested that the vomiting caused by the latter leads to the development of the hernia when there is a congenital laxity of the oesophageal hiatus. In our eight patients with the double lesion, however, vomiting began shortly after birth and not, as occurs with pyloric obstruction alone, after the second week of life.

The pathogenesis of hypertrophic pyloric stenosis remains unknown. Thomson (1933) considered that the muscular hypertrophy of the pyloric canal occurs as a result of a preceding functional obstruction at the pyloric sphincter. Friesen, Boley and Miller (1956) claim to have demonstrated immaturity of ganglion cells in the pyloric tumour. As pyloric stenosis, unlike hiatus hernia, occurs predominantly in the male and has a high familial and hereditary incidence, there is obviously no all-embracing aetiological explanation for the two conditions but their frequent co-existence lends support to Thomson's theory of a prenatal functional pyloric obstruction; the increase in intragastric pressure so produced may then lead to prolapse of the cardia into the chest especially since, at this period of development, the gastro-oesophageal junction is probably not yet stabilized in its normal position.

Some of the clinical features of infantile hiatus hernia, even when there is no associated hypertrophic pyloric stenosis, also suggest the presence of a gastric emptying disorder. That the hernia and the incompetent cardia, although responsible for the development of oesophagitis, are perhaps not the primary cause of the main symptom of vomiting is indicated by the fact that, in those children treated medically and in many of those treated surgically, the hernia and gastro-oesophageal reflux may still be demonstrable radiologically many months after vomiting has ceased. Interference with gastric emptying is suggested by the frequent clinical finding of visible gastric peristalsis and by the fact that the vomiting itself, unlike the passive postural reflux of a small quantity of gastric contents which occurs in the adult, is usually forcible or even projectile, of large volume and not immediately controlled by postural treatment.

Medical management of hiatus hernia in infancy is often successful in relieving the symptoms and in allowing spontaneous cure of the hernia and the development of a competent gastro-oesophageal sphincter. The disadvantage of conservative management is that hospital treatment must often be prolonged, and it is difficult to decide when the patient may be considered free of the risk of complications developing. It is often considered that cure has been achieved when vomiting ceases and the child gains weight normally. The hernia and the incompetent cardia persist, however, for some considerable time after vomiting has stopped and the symptomless patient may still be exposed to the risks of oesophagitis and its sequelae. Severe complications, if they are going to occur, usually develop early but this is not invariably the case; in one of our patients an oesophageal stricture first appeared at the age of 10 years (Figs. 5 and 6). Chronic penetrating ulcer of the intra-thoracic portion of the stomach (Barrett's ulcer) may also develop in later childhood (Fig. 7).

The conventional surgical procedure for hiatus hernia in the child, as in the adult, is transthoracic, transdiaphragmatic reduction of the hernia and repair of the enlarged oesophageal hiatus. In our experience of 40 cases this operation has not given satisfactory results; vomiting has persisted after operation in over 50% of patients and radiological examination, carried out one year or more after operation, has shown recurrence of the hernia or persistence of gastro-oesophageal reflux in over 70%. Unsatisfactory results of hiatal herniorrhaphy in childhood have also been recorded by Husfeldt (1953) and by Thomsen (1955).

Although there is no radiological confirmation of

FIG. 5.—Hiatus hernia without stricture in boy aged 6 years.

FIG. 6.—Same patient as in Fig. 5, aged 10 years. Severe oesophageal stricture.

a gastric emptying disorder in cases of infantile hiatus hernia, there would appear to be sufficient clinical evidence of its temporary presence, responsible at least in part for the development of the lesion and the production of the symptom of vomiting, to justify the therapeutic application of this hypothesis. The operation of pyloroplasty has therefore been applied to the treatment of hiatus hernia in childhood. The rationale of this procedure is that facilitation of gastric emptying will discourage

FIG. 7.—Operation specimen. Chronic penetrating peptic ulcer (Barrett's ulcer) of intra-thoracic part of stomach in boy aged 9 years with recurrent haematemesis.

vomiting and encourage spontaneous cure of the hernia and the development of a competent gastro-oesophageal sphincter. Peptic oesophagitis is due to reflux of pepsin and acid from the stomach; the most severe effects are probably produced by reflux of unbuffered gastric juices when the stomach is empty of food. In order to inhibit this neurogenic peptic secretion and so render inert fluid regurgitating into the oesophagus, bilateral vagotomy has been performed in addition to pyloroplasty when endoscopy revealed the presence of oesophagitis. These procedures have so far been applied to only a small series of cases. The early response has been satisfactory but more prolonged study of the patients is required before full assessment of the results is possible.

Summary

The pathogenesis of infantile hiatus hernia is discussed. It is considered that an important factor in the causation of the hernia and in the production of vomiting is a disorder of gastric emptying. The operation of pyloroplasty and, in the presence of peptic oesophagitis, vagotomy has therefore been carried out in a small series of cases.

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