Hiatus hernia in children

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THE term congenital diaphragmatic hernia tends to be applied to the dramatic herniation of small intestine and other abdominal organs through the foramen of Bochdalek into the left pleural cavity with consequent acute respiratory disturbances. Such children commonly present as acute emergencies during the first few days of life. However, hiatus hernia is also frequently of congenital origin and this is not only a much more common condition but also one that can lead to much more morbidity and even mortality.

Hiatus hernia can be defined as a displacement of the cardio-oesophageal junction above the level of the diaphragm, or a protrusion of part of the stomach through the oesophageal hiatus. Both these types of the abnormality have been demonstrated in the newly born and are being demonstrated with increasing frequency in young children. It is difficult to regard similar conditions presenting in older patients as an entirely different disease but at the same time an explanation must be given to the fact that so many of these cases do not present until the patients reach the age of 50-60 years. The diagnosis can only be made positively by demonstrating cardio-oesophageal displacement or herniation of the stomach through the oesophageal hiatus; such demonstration may be achieved at necropsy, at surgical exploration or at radiological examination. The reported incidence of the condition thus tends to vary with the enthusiasm of the radiologist and the availability of special equipment but there is no doubt that continuing improvement in radiological techniques have brought to light many more cases of hiatus hernia in children than had previously been demonstrated as well as many more cases occurring in adults.

The increased frequency with which supra-diaphragmatic pouches of stomach have been demonstrated on radiological examination has led to an acceptance of the idea that gastro-oesophageal reflux in the newly born and young child is probably an early stage of hiatus hernia; indeed it might be that a hiatus hernia was present but had not been demonstrated. Both in adults and children it is increasingly accepted that the actual hiatus hernia is

of far less importance than the consequent functional disturbance at the cardio-oesophageal junction leading to free gastro-oesophageal reflux, oesophagitis and possibly oesophageal stricture.

Incidence

It is thus difficult to estimate the true incidence of hiatus hernia since almost certainly a number of cases are not demonstrated radiologically and equally a number of clinical diagnoses are made without radiological demonstration. Skinner & Belsey, however, in 1967 reported 119 children in their series of some 1500 cases operated on and Carré in 1970 suggests that in the United Kingdom the incidence of the disease in children is 1 in 1000.

There is no doubt that the condition is a great deal more common than is generally suspected and warrants rather more attention than it is given in the average textbook of paediatrics. While probably more than two-thirds of the children with the condition even untreated become symptom-free by the age of 2 years, usually improving when they get on to a weaning diet (Carré, 1959), there is nevertheless a three-fold increase in pulmonary infections in children who vomit during the night, and the other common complication of oesophagitis can lead to severe stenosis of the oesophagus necessitating major replacement surgery.

Aetiology

The common surgical finding in these children is that there is an abnormally large oesophageal hiatus with lax margins. It is possible that this may be associated with delay in the developmental descent of the stomach relative to the closing of the various elements of the diaphragm which occurs about the seventh week of foetal development; if this were so the descent of the stomach must continue after the inadequate closing of the diaphragm because the occurrence of a congenitally short oesophagus is almost universally accepted as occurring excessively rarely, if at all. Free gastro-oesophageal reflux is an almost universal feature of the neonatal period and it is perhaps rather more attractive to suggest an endocrine factor relating this to the loosening up of

mesenchymal tissues in pregnancy and in the menopause, which may play a part in the development of hiatus hernia in adults (Allison, 1970). The newly born child shows evidence at birth of the effect of its mother's hormones in its enlarged genitalia and the free gastro-oesophageal reflux in the newborn period which lasts only a few days may be similarly produced.

Pathological anatomy

Three types of hiatus hernia have been described resulting from this lax oesophageal hiatus (Fig. 1). First of all the sliding hernia in which a loculus of stomach slides up and down through the oesophageal hiatus. Secondly the rolling type in which a portion of the greater curvature of the stomach passes up alongside the oesophagus through the hiatus, sometimes called a para-oesophageal hernia, and thirdly a combination of these two types where the cardio-oesophageal junction has moved above the level of the diaphragm but there is in addition a rolling element. The sliding type accounts for at least 90% of all hiatus hernias, both in adults and in children.

Relationship of gastro-oesophageal reflux to hiatus hernia

The anti-reflux mechanism of the cardio-oesophageal junction is a very complicated one and at least five factors have been described as playing a part in preserving the competence of this mechanism. The pinchcock mechanism of the sling of the right crus of the diaphragm (Allison, 1951) can only function if the cardia lies below the hiatus. Similarly the closing of the distal oesophagus by positive intra-abdominal pressure can only function if that distal

oesophagus lies within the abdominal cavity, and the entry angle of the oesophagus into the stomach is changed from an acute one to an obtuse one when the cardio-oesophageal junction slides above the hiatus. The rosette formed by folds of mucosa at the point of entry of the oesophagus into the stomach is believed to play some part in preventing reflux and is probably dependent to a certain extent on an intrinsic sphincter mechanism in the lower oesophagus. These last two factors are probably much weaker than the first three and it has certainly been shown in dogs that the intrinsic oesophageal sphincter is interfered with by reflux of gastric juice into the lower oesophagus (Shatz & Bane, 1970). Neuhauser & Behrenberg (1947) demonstrated cardio-oesophageal relaxation as a cause of vomiting in infants and showed that a column of barium moved up the oesophagus on inspiration and on expiration was split, some of it passing on upwards and some going back into the stomach. Children with this oesophageal chaliasia presented with vomiting starting between the third and the tenth day of life, effortless in nature and closely associated with posture after feeding. The symptoms were exactly similar to those occurring in children where a hiatus hernia had actually been demonstrated.

In the rolling type of hernia the angle between the lower oesophagus and the stomach remains unchanged and in fact the bulk of the herniated stomach tends to put pressure on the lower oesophagus making reflux much more difficult, so that the predominant feature in this type of child will be dysphagia rather than reflux vomiting.

Pathological complications

In the rolling type of hernia the lesion resembles most other hernias from the abdominal cavity. There is usually a peritoneal sac and the protruding stomach creates a space-occupying lesion in the pleural cavity, usually rotating to the right side with an ever-present danger of further rotation of this protruding loculus producing a volvulus of the stomach with acute gastric distention. Thus, in addition to the tendency to bleeding from the congested gastric mucosa as it is compressed in the chest cavity, the child's life may be endangered by the very severe collapse following acute distention of the stomach associated with such a volvulus.

In the sliding hernia the pathological complications are the result of gastro-oesophageal reflux and vomiting. These may be simple failure to thrive as a result of inadequate nourishment because of frequent vomiting, repeated respiratory infections as a result of inhalation of vomitus, particularly when there is vomiting during the night, and lastly oesophagitis and its complications. Whilst it is clear that acid is a highly important factor in the production of oeso-

FIG. 1. Diagrammatic representation of types of hernia. (a) Normal. (b) Sliding hernia. (c) Rolling hernia. (d) Combined rolling and sliding.

phagitis the degree of acidity of the gastric contents and the duration of exposure of the lower oesophagus to that gastric content has not been shown in adults, at least, to have much relationship to the severity of the oesophagitis (Collis, 1970). This unexplained variation in response to persistent gastro-oesophageal reflux is a most important feature of the disease in children because whilst it is true that in a large number of cases the symptoms settle without leaving any permanent damage to the lower oesophagus and even of those cases operated upon a simple procedure is sufficient to bring the majority under control, there remains a small number, probably less than 10% of all cases, in whom peptic oesophagitis produces severe results with scarring and eventually demands resection of the lower oesophagus.

Management of these children is designed to avoid the pathological complication in the rolling hernia or the combined type. Spontaneous regression will not occur and surgical correction is indicated in all cases. In the sliding hernia, however, when as many as two-thirds of the cases may be expected to resolve spontaneously, surgical treatment in all cases would be unjustifiable. At the same time it is very important that those children likely to develop severe oesophagitis should be dealt with before irreversible changes have occurred in the oesophageal mucosa; and less obviously, recurrent inhalational respiratory infections and failure to thrive must be recognized before they interfere seriously with the child's ultimate development.

These then are the indications for surgical intervention. The institution of conservative management and its various aspects depends on the diagnosis of the condition and a knowledge of its natural history.

Signs and symptoms

Vomiting

The classical presentation of hiatus hernia in children is with a history of vomiting from birth. Over 90% of the children will certainly vomit during the first month of life and the majority will in fact start during the first day or two. The vomiting is usually forceful in character and may even be projectile, resembling the vomiting due to congenital hypertrophic pyloric stenosis, but the very early onset tends to differentiate the condition from hypertrophic pyloric stenosis, in addition to the fact that the child with hiatus hernia frequently vomits early during a feed when taking the first few mouthfuls, and that posture influences the vomiting which often occurs the moment the child is laid down after a feed. It must be remembered that congenital pyloric stenosis may occur as an associated condition with congenital hiatus hernia.

The nature of the vomitus is also of some importance: it consists of mucus or gastric content un-

stained by bile but frequently containing some altered blood.

Failure to thrive

The vomiting in the neonatal period before treatment may be severe enough to cause considerable dehydration in addition to preventing weight gain. Most children in fact are already underweight when they present and show some degree of malnutrition.

Anaemia

Anaemia is a common finding especially in those patients with oesophagitis. Although the daily loss of blood may be very slight and even though occult blood is not always demonstrable in the stool it is likely that this very small but persistent loss, as in the child losing a little blood from colostomy dressings daily, is sufficient to account for the presence of anaemia, and that the malnutrition does not play such an important part. The poor correlation between the severity of oesophagitis and the degree of anaemia has led some workers to suggest an iron absorption deficiency (Michaelides & Philis, 1959).

Frank haematemesis is rare and practically confined to those cases with para-oesophageal rolling hernias.

Dysphagia

Dysphagia again is an uncommon symptom unless the child with a sliding hiatus hernia has developed oesophagitis. Occasionally a baby is seen to cry on feeding suggesting the presence of dysphagia. In the rolling hernia, dysphagia would be more commonly expected because of the pressure of the herniated fundus of the stomach on the lower oesophagus and indeed this does occur in some older children with the rolling type of hernia; in the neonate, however, the child with the rolling type of hernia still tends to present with vomiting.

Progress of the disease

There is a strong tendency for the condition to subside by the time the child reaches a year of age and in fact the mother commonly will note that, first when the child sits up for more of the time during the day, and later when he starts to walk there is a noticeable decrease in the frequency of the vomiting. The time when he sits up is also related to the time when he takes more solid foods. Although vomitus may often be found on the pillow at the end of a night's sleep, the child gradually vomits much less during the day. Waterston (1969) rightly warns of the danger of too much optimism when daily vomitus changes to episodic attacks which may indicate that the child has developed a stricture in his lower oesophagus, not that the hernia is cured.

Oesophagitis

Oesophagitis with its risk of stricture formation and the tendency to progress to severe fibrosis is the most undesirable complication, and it is important that those cases liable to develop severe oesophagitis should be dealt with surgically before irreversible fibrotic changes have been allowed to occur: persistent blood-staining of the vomit and dysphagia are warning signs as are the episodic severe attacks mentioned above.

Respiratory infections

Recurrent aspiration pneumonia is a well recognized complication and is especially important in the children with mental retardation who form an appreciable number of most series of hiatus hernias (Moncrieff & Wilkinson, 1954). In addition to this, both in those children who have been operated on and those who have been treated conservatively, there is a notable tendency for the child to have an episode of vomiting for several days whenever he has upper respiratory infection.

Management

There is no doubt that a considerable number of children with gastro-oesophageal reflux settle with conservative methods and are not investigated. If a child is possetting a good deal after feeds it may well be that he has gastro-oesophageal reflux but provided none of these possets are blood-stained and the child's weight-gain is normal then there is little indication for further investigation, and the mother is encouraged to persist in the simple management of the child. Investigation is indicated when there is evidence that the child is failing to gain weight or when there is some blood-staining of the vomitus.

The diagnosis should be suspected in all those children who present with vomiting in the first week of life, particularly those who vomit gastric contents alone and in whom no systemic cause of the vomiting has been demonstrated. Confirmation of the diagnosis is achieved by radiographic study after the swallowing of contrast medium, and the type of treatment is similar whether gastro-oesophageal reflux alone has been shown or whether a sliding type of hiatus hernia has been demonstrated. In the first place management will be on conservative lines; the child is nursed in an upright position by day and by night making use of one of the commercially available plastic baby chairs, and the feeds are thickened either by the addition of Benger's food or by the use of Nestargel. Frequent small feeds should be given and if the child should vomit, either during or at the end of the feed, he should be given another feed immediately.

The aims of treatment are two-fold—to prevent loss of nourishment by vomiting and to avoid oeso-

phagitis from the bathing of the lower oesophagus in refluxed acid gastric juice. Medical methods of avoiding reflux oesophagitis include the use of antacids and the more recent introduction of a low specific gravity colloidal gel (Gaviscon) designed to float on top of the feed in the upper part of the stomach and to form a protective coat adhering to the lower oesophageal mucosa. Oesophageal ulceration, however, can occur very rapidly with grave consequences and when there is evidence of oesophagitis the possibility of surgical intervention must be closely borne in mind.

Surgical intervention

In a review of 3 years in The Children's Hospital, Sheffield, forty-five cases were referred to the paediatric surgical unit. Thirty of these children were treated successfully by medical means, fifteen of them came to operation.

(a) *Rolling hernia.* In a rolling hernia the herniated stomach forms a space-occupying lesion in the thoracic cavity. Four of the fifteen cases operated upon fell into this class and all were symptomatically cured as well as being shown to be cured radiologically. Three presented under 2 years of age, the fourth was a child of 8 who developed acute distention of the herniated stomach after trauma and was operated upon as an emergency. This was the only child who had an abdominal approach to his lesion, the others having been operated upon through the left pleural cavity, and having had the hernial sac excised and the oesophageal hiatus narrowed by a few non-absorbable sutures in the right crus posterior to the oesophagus.

This group of cases presents little problem. In those children in whom the rolling hernia is combined with a sliding one a fundal plication can be added to the operation, the fundus of the stomach

FIG. 2. Diagram of fundal plication.

FIG. 3. Pre- and postoperative contrast study X-rays showing rolling hernia corrected by reduction, repair and fundal plication.

being sutured around the lower oesophagus to convert the gastro-oesophageal junction into an invertible inkwell type of entrance (Fig. 2). This procedure will make reflux very much less likely to occur and also provides a buttress below the oesophageal hiatus which discourages recurrence (Fig. 3).

(b) *Sliding hernias*. Sliding hernias produce a rather less satisfactory picture. They came to surgery on two broad indications.

(1) *Failure of conservative treatment*. In four of the eleven children aged from 3 months to 6 years, surgical correction was undertaken because of failure to gain weight in spite of nursing in the upright position in the first few weeks of life, or failure to control vomiting in the older child. All these children were cured by relatively minor procedures, two had a pyloroplasty alone and two had pyloroplasty, vagotomy and fundal plication. All are cured of their symptoms though not necessarily of their hernia.

(2) *Dysphagia and bleeding*. Dysphagia indicates either stenosis of the lower oesophagus or severe oesophagitis with some spasm. The severe oesophagitis may also produce persistent anaemia, repeated melaena and, rarely, haematemesis. The occasional blood-stained vomit, particularly vomit containing altered blood, is of far less significance than dysphagia: this type of blood-stained vomit was seen in two of the children who were operated on because of vomiting and in many of the children who were treated medically.

Seven children were treated in whom there was

radiological evidence of oesophageal narrowing; one appears to have been relieved by gastrostomy alone (a child aged 4 weeks), one by pyloroplasty, vagotomy and fundal plication, and two by gastrostomy and pyloroplasty followed at a later stage by vagotomy and fundal plication.

Three, however, remained in trouble. Two had gastrostomy and pyloroplasty followed by vagotomy and fundal plication and required dilatations for persistent stricture even after 2 years. The third had a pyloroplasty but developed a severe stricture for which a gastrostomy had to be made and eventually he required an oesophago-gastrectomy.

Discussion

In those children in whom operation was indicated basically to control vomiting the results were excellent; all had a pyloroplasty made which is possibly important with regard to the not infrequent associated pyloric hold-up (Waterston, 1969). The fundal plication procedure (Nissen & Rosetti, 1959) has proved a most satisfactory method of preventing reflux.

The rather surprising relief of symptoms in several cases by gastrostomy alone led us to believe that fixation of the stomach to the anterior abdominal wall might be an even simpler procedure and an adequate one. Results achieved by fixation of the lesser curvature of the stomach to the anterior abdominal wall were disappointing. Since the purpose of operation is to prevent gastro-oesophageal reflux

then gastropexy must be designed to have its effect by maintaining an abdominal oesophagus and thus preserving the exposure of lower oesophagus to the pinchcock action of the right crus and to the positive intra-abdominal pressure: Woodward, Rayl & Clarke (1970) point out that both these mechanisms are weak elements of the natural sphincteric mechanism and a high incidence of long-term recurrence would be expected.

In our series in Sheffield, 20% of those operated on remained in serious trouble from strictures. One had already required interposition of colon between the oesophagus and the stomach, and it seems likely that the other two will also require this procedure. The poor results are in those children in whom severe stricture of the lower oesophagus has occurred. A great deal of work has been done on the varying response of the oesophageal mucosa to gastric contents (Collis, 1970) but it is not clear why oesophagitis can be so severe in some cases and so mild in others. Hyperacidity is not essential but acid is highly important in the production of this oesophagitis and one might therefore suggest that an anti-reflux procedure should be carried out in all cases. This, however, would mean that in approximately two-thirds of children an unnecessary operation would have been carried out, since thirty out of our forty-five cases were successfully treated conservatively. Nevertheless, it would seem necessary that surgical intervention should be considered earlier in some cases than it has been in the past. It is difficult also to ignore the fact that a number of children who have developed severe strictures have in fact been shown to be high acid secretors (Lari, Lister & Duthie, 1968). It would seem justifiable in these cases not only to recommend fairly early operation, but also to include vagotomy in the surgical procedure. Ideally, then, an antireflux procedure of a simple nature as provided by the operation of fundal plication (Nissen) should be carried out in all those cases in whom there is evidence of persistent oesophagitis and selective vagotomy should be added if there is also evidence of high acid secretion.

In those cases in which some narrowing of the lower oesophagus has already occurred it is important to recognize whether that narrowing is reversible or not. Temporary gastrostomy plays a part in this differentiation. On more than one occasion when dealing with a child who had dysphagia, vomiting and malnutrition, we have made a gastrostomy essentially for feeding to improve the child's general condition before major operation; we have then

found that within a few days the child began to swallow normally demonstrating that the narrowing of the oesophagus was due more to spasm than to fibrotic stricture. This effect may have been due to neutralization of the gastric contents by frequent gastrostomy feeds, to resting the oesophagus, or to fixing the stomach to the anterior abdominal wall, but whatever the mechanism a rapid relaxation of the narrowed lower oesophagus would encourage a less radical surgical procedure to be undertaken, and failure of such a response is a good indication for proceeding to oesophageal resection. Colonic interposition between the oesophagus and the stomach, whilst being a major procedure, is probably preferable to the years of dilatation that may be required to overcome the repeated recurrence of severe strictures.

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