Congenital pyloric atresia

A report on a successfully treated case and existing research

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Summary

Congenital pyloric atresia is a rare malformation that occurs with a frequency of about one per million births. One hundred fifty-one cases of the anomaly have been reported including 28 cases in Japan. This is a report on an additional case of membranous atresia that was treated successfully by incision of the diaphragm with pyloroplasty. The familial occurrence in siblings and the association of the abnormality with hereditary epidermolysis bullosa suggests a genetic etiology. Treatment should be surgical and the choice of an operative procedure should depend on the anatomical type of atresia. Excision or incision of the diaphragm with pyloroplasty and gastroduodenostomy offer the most promising opportunity for successful treatment. The mortality rate is 49.3% and the prognosis is dependent upon such factors as early diagnosis, the appropriate operative procedure, the extent of prematurity of the infant and association of other congenital anomalies or complications.

Key words: pyloric atresia, pyloroplasty, gastroduodenostomy

Introduction

Congenital pyloric atresia is a rare condition. According to Gdanietz', it was first referred to by Calder in 1749. Since then 151 cases have been reported including 28 cases in Japan. We report here a case of membranous atresia which required an incision of the diaphragm with pyloroplasty and obtained a successful result.

Case Report

A 3-day old female infant was transferred to the Department of Surgery, Kennan General Hospital, Tochigi, Japan...
General Hospital, Tochigi, Japan, with a history of bile-free, non-projectile vomiting and upper abdominal distension since birth. The patient's mother had no polyhydramnios and the prenatal course was uneventful. The baby was born at 39 weeks gestation by spontaneous vaginal delivery without complications. At birth, the infant weighed 2,900 g had an Apgar score of 10. Slight meconium was passed by enema on the second day. The family history showed neither consanguinity nor pertinent abnormalities.

A physical examination disclosed a 2,600 g well-developed infant with mild dehydration and jaundice. Respiratory distress and cyanosis were not noted. The upper abdomen was moderately distended without evidence of peristaltic waves. A plain X-ray of the abdomen showed a single gastric air bubble with no air beyond the stomach (Fig. 1). An upper gastrointestinal series confirmed the diagnosis of complete pyloric obstruction.

In surgery, performed on the fourth day after birth, the stomach was markedly dilated and showed thickened walls. The pylorus appeared normal, however, the entire duodenum and the intestines had collapsed. The contents of the stomach could not be pushed into the duodenum. A longitudinal incision was made in the distal end of the stomach and an unsuccessful effort was made to pass a soft probe from the incision antegrade into the duodenum. A second longitudinal incision was then made in the proximal portion of the duodenum but the probe could not be maneuvered into the stomach. There was a membranous diaphragm completely occluding the lumen in the region of the pylorus. No atresia was detected below the duodenum. The two incisions were joined in one duodenopylorotomy about 3 cm in length. The diaphragm was incised and the posterior mucosa was sutured with interruption using No. 3-0 silk. A Heineke-Mikulicz pyloroplasty was then performed and a tube was inserted into the stomach. The postoperative course was uncomplicated. The baby could be fed through the tube on the third postoperative day and bowel movements were satisfactory. An abdominal roentgenogram revealed intestinal gas (Fig. 2). The infant was discharged two weeks postoperatively.

Discussion

Congenital pyloric atresia is an extremely rare abnormality. In the entire gastrointestinal tract, atresia is allegedly
stated to occur once in every 10,000 births. Of all the atresias, pyloric atresia accounts for less than 1 percent, therefore, it occurs with a frequency of about one per million births.

The first case of surgery for pyloric atresia was published by Little and Helmholz in 1905, and the first successfully treated patient was described by Touroff and Sussman in 1940.

There are various theories as to the pathogenesis of intestinal atresia; a failure of recanalization of the intestinal tract during embryogenesis, a mechanical or vascular injury to a previously normal fetal intestine, or a redundant endodermal lining and formation of an epithelial fold. These theories also can be applied to pyloric atresia but proof of etiology is lacking.

Classifications of the anatomical types of pyloric atresia by Kornfield or Gerber are generally accepted. Kornfield classified atresias into the following three different variations: Type 1 - web or diaphragm; Type 2 - fibrous cord bridging two blind ends; and Type 3 - segmental defect with blind ends. We have distinguished from Type 1 the double membranous type of atresia as Type 4, as Kume and co-workers did because the operative procedure for this type differs from that used in the treatment of Type 1 atresias. Of the 131 clearly described cases found in reports including our case, the modified Kornfield classification are: Type 1; 61.1% (80 cases), Type 2; 28.2% (37 cases), Type 3; 6.9% (9 cases), and Type 4; 3.8% (5 cases).

Gerber's classification was as follows: Pyloric membrane (IA), Pyloric atresia (IB), Antral membrane (II A), and Antral atresia (II B). The most common form consists of IA which comprises 53.8% (64 cases) of the 119 clearly described cases collected by us including our case. The percentage of the other three atresias are IB; 26.1% (31 cases), II A; 15.1% (18 cases), and II B; 5.0% (6 cases). Kadowaki stated that Gerber's classification seemed most practical in regard to operative treatment. In the presence of a diaphragm (type IA and II A), a pyloroplasty combined with either incision or excision of the membrane appears to be a satisfactory operation. When atresia is present (type IB and II B), an end-to-end gastroduodenostomy is the choice of procedure.

Although hydramnios was not clinically detected in the mother of this infant, patients with pyloric atresia are usually born of a mother with polyhydramnios, the incidence of
which is greater than 50%\(^{16-13}\). Hydramnios was present in 77 cases (50.7%) among the series collected by us. This is comparable to the 45% incidence of mothers whose infants had duodenal or jejunal obstruction as reported by Clatworthy\(^{14}\). Lloyd\(^{15}\) stated that failure of the fetus to participate normally in the transfer of amniotic fluid, due to an obstructive lesion in the proximal portion of the gastrointestinal tract, leads to polyhydramnios.

The incidence of premature births is also greater than 50%\(^{16-13}\). A premature infant was present in 60 out of 108 clearly described cases (55.6%) in the series.

Sexual distribution is roughly equal\(^{17-19}\). In the series including our case, 74 out of 132 cases (56.1%) were male and 58 (43.9%) were female. Seventeen cases of consanguinity among parents have been reported\(^{19-23}\). Twenty-seven cases of familial occurrence in siblings in 13 families\(^{12,13,15,18,21,24-25}\) have been reported since the first report by Thompson\(^{20}\). These findings strongly suggest a genetic determination, probably an autosomal recessive mode of inheritance as previously suggested by Bar-Maor\(^{19}\).

It is of interest that 21 cases of pyloric atresia associated with epidermolysis bullosa (EB) and/or aplasia cutis congenita (ACC) have been reported\(^{17,19-22,27,37,39-40}\). Bull et al.\(^{20}\) support the existence of EB–pyloric atresia syndrome as a distinct clinical entity with autosomal recessive inheritance. The possibility suggested by De Groot et al.\(^{20}\) is that the concurrence of EB and the malformation represents the pleiotropic expression of a single gene. On the other hand, Peltier et al.\(^{21}\) describe the theory of a genetic linkage between these two disorders. Furthermore, Carmi et al.\(^{22}\) state a linkage between the gene for ACC and the gene for EB and pyloric atresia. The possibility of a genetic etiology in the abnormality is strongly suggested.

The diagnosis of pyloric obstruction is not generally difficult if the following clinical findings are noticed. A newborn infant, usually born of a mother with polyhydramnios, presents with upper abdominal distention and persistent bile-free emesis. Plain X-rays of the abdomen most often show a large distended stomach with a solitary gas bubble or an air fluid level, and no air beyond the pylorus. It is not necessary to use a contrast medium to make or confirm the diagnosis\(^{7}\). The differential diagnosis includes a number of conditions such as duodenal atresia, annular pancreas, brain damage, adrenocortical insufficiency, and maternal narcotic addiction. Talwalker\(^{40}\) described a case of congenital pyloric atresia showing a “double bubble” appearance. Failure to relieve the obstruction can lead to respiratory distress, aspiration pneumonia, metabolic alkalosis resulting from persistent vomiting, and even gastric perforation\(^{12,16,25,21,40}\).

Treatment should be surgical and the operative procedure should be individualized depending upon the anatomical type of malformation. For Type 1, a pyloroplasty combined with incision or excision of the diaphragm is the choice of procedure. When it is Type 2, an end-to-end or side-to-side gastroduodenostomy is preferred. A gastroduodenostomy or gastrojejunostomy is the method of choice for Type 3 or Type 4. However, gastrojejunostomy being done, the risk of a late anastomotic ulcer, occasionally a blind loop syndrome, and a high mortality rate must be considered\(^{40-12,50}\). Some authors\(^{11,13}\) favor adding gas-
trostomy which affords excellent decompression of the stomach.

Seventy out of 142 clearly described cases (49.3%) of pyloric atresia died in the series, which is higher than of previous reports\(^\text{10,13}\). The mortality rate of premature infants, 55.0% (33/60 cases), is very high in contrast to that of mature babies, which is 25.0% (12/48 cases) in the series. Surgical intervention was attempted in 123 infants with 50 resulting in death (mortality, 40.7%), that is about equal to that of Konvolinka's report\(^\text{43}\).

The death rate of each type of atresia according to the modified Kornfield classification is shown in Table 1. Seven cases of gastrojejunostomy are among the 14 cases of Type 3 and Type 4 atresias. All 7 patients died, which suggests that serious considerations should be exercised before this procedure is used.

The mortality rates after various surgical procedures are listed in Table 2. The 33.3% who died after pyloroplasty combined with incision or excision of a diaphragm include 4 who died from other anomalies or complications despite a successful operation. The mortality rates shown suggest that gastrojejunostomy should be reserved for cases where pyloroplasty or gastroduodenostomy can not resolve the atresia. Other operative procedures were used in the five cases that resulted in the deaths of all five. It is clear that pyloroplasty and gastroduodenostomy offer the greatest opportunity for success.

The mortality rate of pyloric atresia having other congenital anomalies or complications is very high (Table 3). In the 14 cases of pyloric obstruction in association with gastrointestinal anomalies\(^\text{1,15,23,25,42,43}\), only one survived, which was Haller's case\(^\text{49}\) with the duodenal atresia. Eight out of 13 deaths had multiple gastrointestinal atresia, of which Guttman\(^\text{43}\) described three consanguineous cases. He designated a new syndrome of hereditary multiple gastrointestinal atresia with an autosomal recessive trait. Only four patients\(^\text{4,12,43,44}\) showed features of Down's syndrome in contrast to the high incidence associated with neonatal duodenal atresia.

The prognosis of pyloric atresia will be dependent upon such factors as early diagnosis, operative procedure, prematurity of the infant and other anomalies or complications.
References

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