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Pyloric atresia: report of four cases and review of the literature

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Abstract. Pyloric atresia is a very rare malformation. Up to 1987, 140 cases of complete pyloric obstruction had been described in the available medical literature [9]. Four patients with congenital pyloric atresia are presented. On the basis of our own cases and those described in the literature, clinical features, diagnostic procedures, and therapy are summarized and discussed. The typical radiological and clinical signs should in most cases lead to the correct diagnosis.

Key words: Pyloric atresia – Rare congenital pyloric malformation

Introduction

In the 18th century, Calder described for the first time the rare condition of complete pyloric obstruction. Over 100 years later, in 1940, Touroff and Sussmann [14] were able to operate successfully in a case of pyloric atresia; they excised a membrane and subsequently performed a Heinecke-Mikulicz pyloroplasty.

The incidence of total intestinal obstruction in the upper gastrointestinal tract is given in the literature [13, 15] as approximately 1:10,000 neonates. Pyloric atresia accounts for about 1% of these atresia cases, an incidence of 1:100,000.

In 1987, the 140 cases of pyloric atresia published up to that time in the medical literature were compiled by Lorenzet and Morger [9] and evaluated with regard to clinical features, diagnostic procedures, therapy, and associated malformations [5, 7, 8, 16].

Of the 114 children with pyloric atresia operated on since 1940, only 53% survived. In 12 cases the pyloric obstruction was at first overlooked. The diagnosis was only established at reoperation or autopsy.

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The following four case histories demonstrate the typical clinical features, diagnostic procedures, and therapy. An early diagnosis is decisive for the success of the operation and the chance of survival of these children.

Case reports

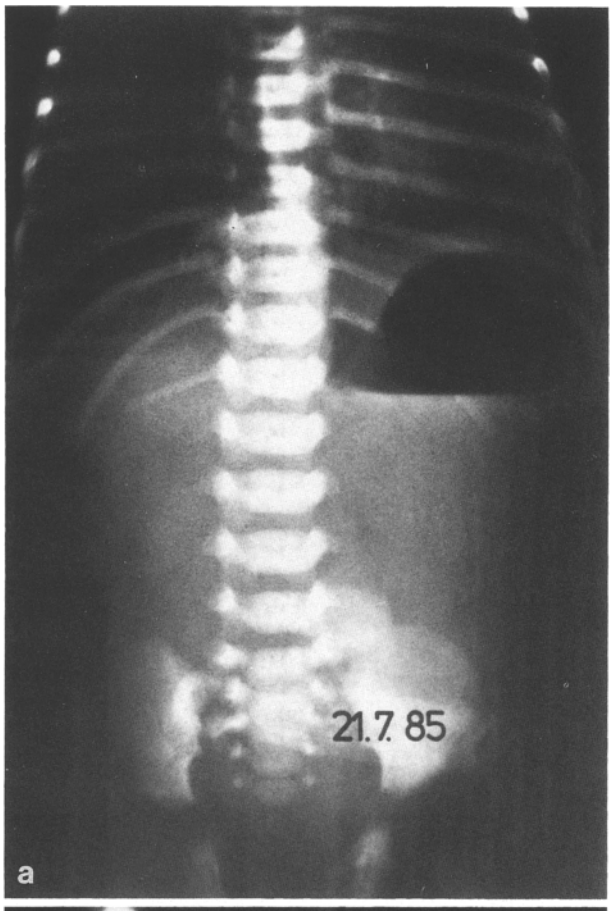
Case 1. (Children's Hospital, St. Gallen) M. I. is the second child of a 27-year-old gravida 2, para 2 mother. The ultrasound examinations during pregnancy were normal. Ceserean section has necessitated by a pathological cardiotocogram and yellow amniotic fluid following artificial rupture of the membranes. An exceptionally large amount of amniotic fluid (4.51) was observed. The Apgar score was 7/10/10. The first feeding, which took place 12 h after birth, was followed by vomiting of bile-free stomach contents; vomiting recurred at subsequent feedings. Micturition and passage of meconium were normal. In the left epigastrium and mid-abdomen peristaltic movements could be observed at times; there was also slight distention of the epigastrium. On admission to the Children's Hospital at the age of 36 h the patient showed signs of dehydration. The abdomen was soft, percussion hyperressonant, bowel sounds quantitatively diminished but qualitatively normal. The stomach was palpable.

The plain X-ray film of the abdomen showed a large, air-filled stomach without any gas distally (Fig. 1 a and b).

An upper median laparotomy was carried out for suspected pyloric atresia. On opening the abdomen, a greatly distended stomach was found; distal to the pylorus the intestine was gas-free and collapsed. On attempting to push a nasogastric tube through the pylorus a resistance was met that could not be overcome. After incision of the stomach wall, a membrane could be seen at the entrance to the pylorus. The pylorus was opened and the atresia resected by means of a longitudinal incision. Gastric and pyloric mucosa were joined and a pyloroplasty was performed. A transanastomotic silicon-7 catheter was inserted followed by closure of the gastrostomy.

Peri- and postoperatively antibiotic coverage was administered for 5 days. After 48 h enteral nutrition was begun through the transanastomotic tube. Passage of stool occurred spontaneously and weight gain was normal. Postoperatively a contrast meal showed unimpeded intestinal passage through a normally wide pylorus. A follow-up study after 6 months also showed completely normal passage (Fig. 2).

Cases 2 and 3. (Marien Hospital, Bochum) H. St. is the second child of a secundigravida who in 1986 gave birth to a girl weighing 2000 g in the 35th week of pregnancy. Postpartal respiratory distress occurred in connection with an amnion-infection syndrome.



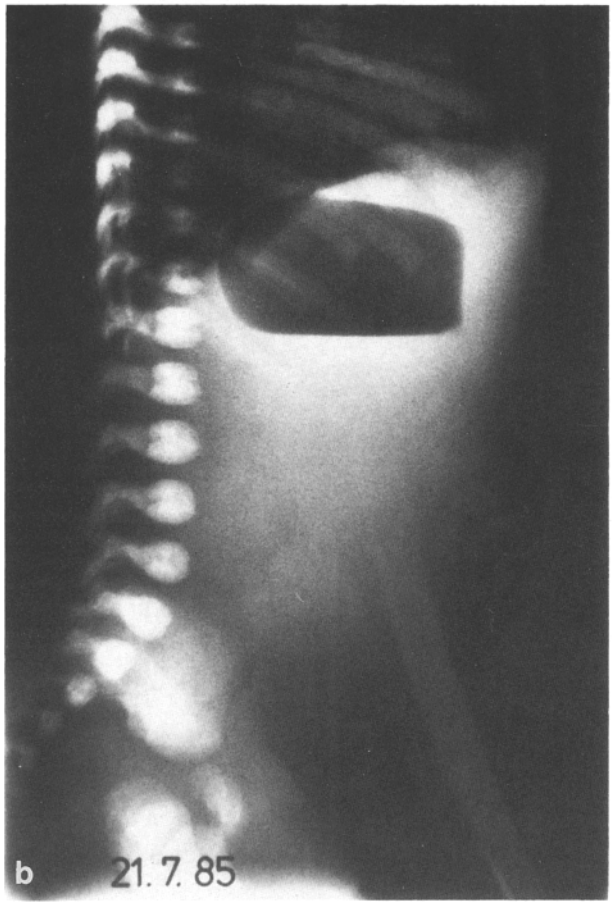


Fig. 1. a Plain AP X-ray film of the abdomen. **b** Plain lateral X-ray film of the abdomen. Note large, air-filled stomach without evidence of gas distally

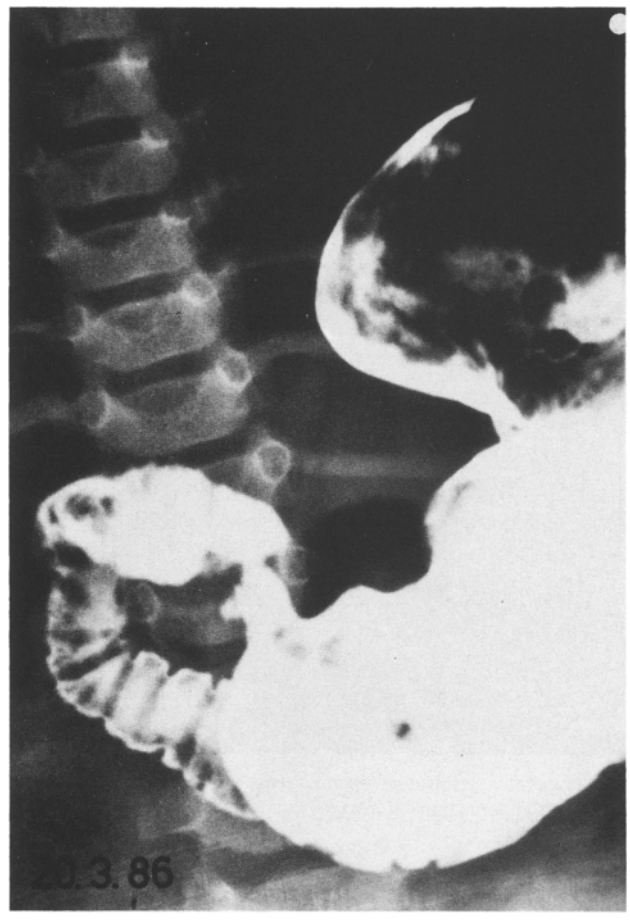


Fig. 2. Postoperative intestinal contrast meal showing free passage through a normally wide pyloric canal

On the 2nd day of life a laparotomy was carried out for suspected pyloric atresia. Intraoperatively, two membranes were found, which were penetrated and dilated. Postoperatively lethal cardiopulmonary complications arose. The autopsy showed fresh scarring of the myocardium suggestive of a possible viral myocarditis.

H. St. had a birth weight of 2640 g after an uneventful pregnancy with marked hydramnios. One day postpartum repeated vomiting began. Passage of meconium occurred twice. The diagnosis of suspected pyloric atresia was made sonographically and radiologically (Fig. 3). Laparotomy confirmed the diagnosis of a long-segment pyloric atresia. Additionally, dispersed pancreatic anlages were found in the region of the duodeno-jejunal flexure and 3 cm further distally on the cranial, anterior wall of the jejunum. An arcuate incision was made on the bulbus duodeni and antrum pylori. The lumen in the pyloric region was totally absent for a length of 13 mm. The incision was extended in order to create a pedicle flap of the anterior gastric wall. After mobilization, the flap was transferred up to the incision on the anterior duodenal wall. By leaving the dorsal portion of the pylorus, the pedicle flap could be used for reconstruction of the anterior circumference of the pyloric canal. In addition, the lesser curvature near the pylorus was mobilized through an incision and adapted corresponding to the defect (Fig. 4 a and b). A Charrière 8 jejunal tube was inserted transanastomotically for splinting. Histologi-

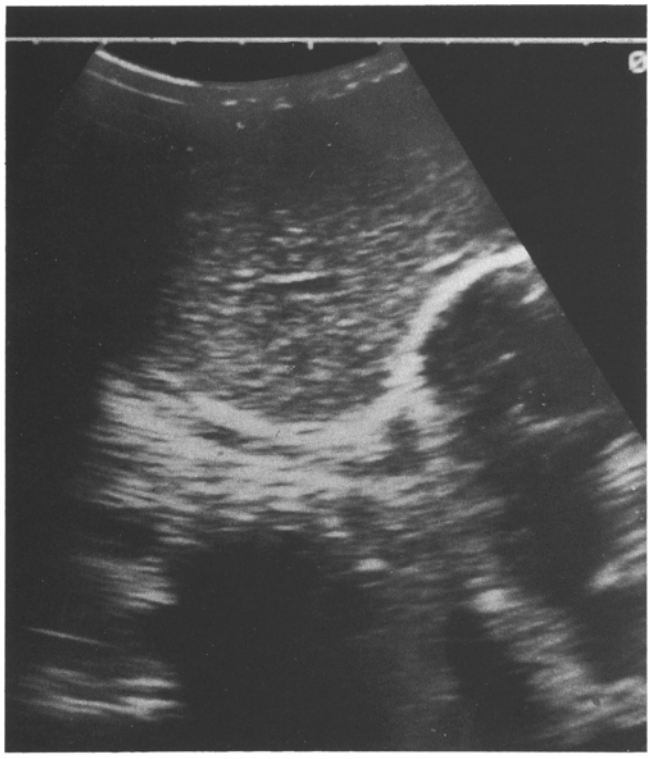


Fig. 3. Preoperative ultrasound examination showing long, stretched-out pyloric canal without connection to the stomach

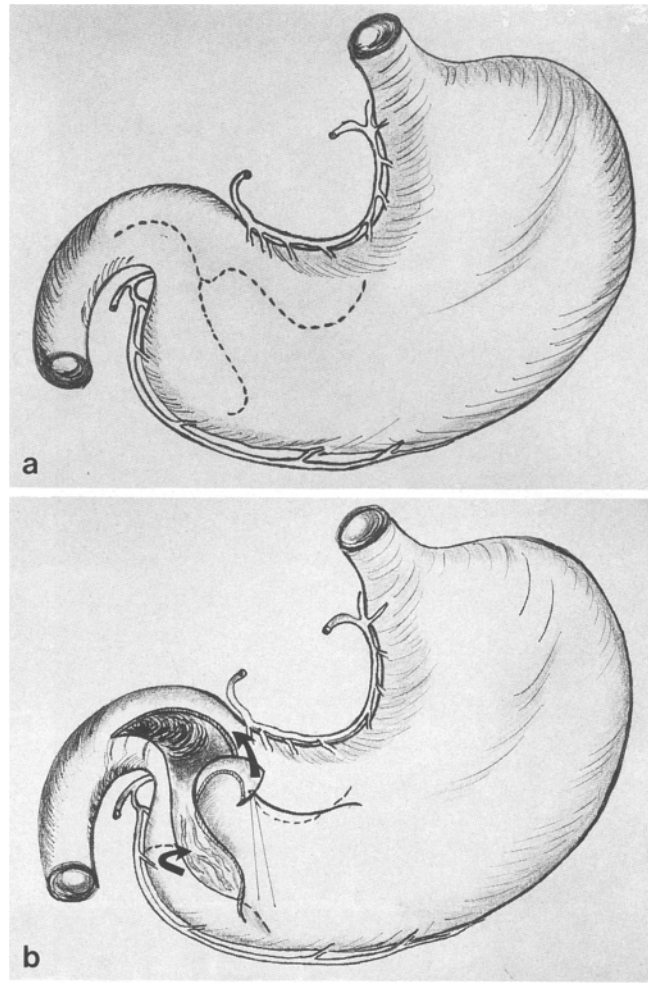


Fig. 4. a and b Schematic drawings of operative procedure

cal examination of the tunica muscularis showed three-dimensionally woven, single-fiber bunching with parts of the plexus myentericus.

Parenteral nutrition was carried out postoperatively. After 10 days enteral feeding was begun. A contrast meal after 4 weeks showed unrestricted passage with normal peristalsis of the stomach.

Case 4. (Children's Hospital, St. Gallen) A. P. is the second child of a 33-year-old secundigravida. The pregnancy was normal. One day before birth polyhydramnios was found and amniocentesis was carried out; 1.31 amniotic fluid was removed. Sonographically, a duodenal stenosis was suspected. Birth occurred spontaneously at 40 $^1/_7$ weeks. The amniotic fluid was clear. The Apgar score was 8/9/10. The stomach contained 70 ml amniotic fluid. On admission to the intensive care unit the abdomen was soft but distended. Percussion revealed hyperresonance and the bowel sounds were scanty. On rectal examination a small amount of meconium was passed.

The plain X-ray film of the abdomen showed a large, air-filled stomach without evidence of gas distally as in case 1 (Fig. 1).

An upper median laparotomy was carried out for suspected pyloric atresia; a very large stomach and a collapsed duodenum were found. Incision of the pylorus showed a membraneous obstruction at the border pylorus/duodenum. The membrane was opened and bile reflux occurred. A silicone catheter was inserted into the jejunum. The intestine was typically collapsed. The longitudinal incision on the pylorus was joined with the duodenum transversely and a gastrostomy was carried out. Postoperatively antibiotic coverage was administered and after 48 h enteral feeding was begun through the jejunal tube.

A contrast meal 10 days later showed a normally wide anastomosis and free passage into the normally positioned small intestine, which had a rather narrow lumen.

Passage of stool occurred spontaneously and weight gain was normal. After 6 weeks a contrast meal showed free passage through a normally wide pyloric canal.

Discussion

Pyloric atresia is characterized by typical features, as documented in our own four cases and the cases of Lorenzet and Morger [9] (Table 1).

In making the diagnosis of pyloric atresia with corresponding clinical signs, polyhydramnios as well as the typical X-ray picture indicate, as in our cases, a suprapapillary obstruction. The plain X-ray film of the abdomen (hanging position) shows the one-bubble sign corresponding to a large, solitary gastric air bubble. This is not a specific radiological sign of pyloric atresia, but it indicates a stenosis of the gastric outlet. Vinz [15] described a patient whose ligamentum teres hepatis had blocked the outlet of the stomach, giving the deceptive impression of pyloric atresia. A long, stretched out peak at the pylorus is regarded as a pathognomonic sign of complete pyloric closure. The ultrasound examination, depending on the skill of the examiner is an important addition to the radiological diagnostic studies. When there is prolapse of a membrane into the duodenum, a double-bubble sign can imitate duodenal atresia.

Table 1. Clinical features of pyloric atresia

Symptoms	Occurrence (%)
Bile-free vomiting	100
Single stomach bubble (one-bubble) on X-ray	98
Passage of meconium	69
Distended epigastrium	68
Polyhydramnios	63
Birth weight less than 2500 g	53
Prematurity	45
Jaundice	21
Peristaltic movements in the epigastrium	18
Haemorrhagic vomiting	12

Table 2. Incidence of the different pyloric atresia types from review of the literature [9]; $n = 140$

Type A (membrane)	77 cases (57%)
Type B (atresia)	46 cases (34%)
Type C (aplasia)	12 cases (9%)
No details	5 cases (3.5%)

Pathologically-anatomically, there are three different types of pyloric obstruction: (1) membraneous pyloric obstruction; (2) longitudinal segmental atresia; and (3) pyloric aplasia. Apart from these, single case reports [2, 11] have described double membranes. In Table 2, the incidence of the different pyloric atresia types is shown [9]. Two of our patients (cases 1 and 4) had membraneous obstruction, one (case 3) had atresia, and one (case 2) a double membrane.

Corresponding to the classification of the pyloric atresias, different operative procedures were used in our cases. Our preoperative measures consisted of therapy for pulmonary complications, correction of dehydration, and insertion of a nasogastric tube. Splinting was performed in the operative area to avoid postoperative edema.

In the literature [9], the best results from operative treatment of a membraneous obstruction were obtained by excision of the membrane and pyloroplasty according to Heinecke-Mikulicz or Finney. With longitudinal segmental atresia, the operative method depends on the length of the atresia. When the atresia is short, a Finney pyloroplasty [9] should be carried out; as a rule, however, a Billroth I gastroduodenostomy [12] is required.

Table 3. Mortality with regard to type of pyloric obstruction [9]. Operated cases since 1940, $n = 114$

Type of atresia	n	successful	%	died	%
Membrane (type A)	67	41	61.2	26	38.8
Atresia (type B)	41	19	46.2	22	53.8
Aplasia (type C)	6	1	16.6	5	83.4

Total mortality: 47%

Pyloric aplasia, which is often associated with other malformations, carries a high mortality [9] (Table 3). The high incidence of pyloric aplasia associated with esophageal atresia is striking: 25% [9].

Of the malformations associated with pyloric obstructions, epidermolysis bullosa is the most prevalent [1, 3, 4, 10]. Of 140 children [9], 35.8% had an associated malformation; 10% had, in addition to complete pyloric closure, a second atresia proximal or distal to the pylorus. Our cases did not have such malformations.

Ultrasound is a good screening method for detecting associated malformations [6]. In view of these associated malformations, hereditary autosomal-recessive transmission of pyloric atresia has been suggested [2, 4]. Genetic counselling is indicated in view of the 25% risk of recurrence.

Prenatal diagnosis is possible by measurement of alpha-fetoprotein and ultrasound examination.

In summary, our cases support the general view that early diagnosis and subsequent operation are of decisive importance in cases of pyloric atresia.

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