

## Congenital pyloric atresia: a report of two cases

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Congenital pyloric atresia (CPA) is a very rare malformation that constitutes less than 1% of all upper gastrointestinal atresias.<sup>1</sup> It is a unique malformation that is commonly seen as an isolated lesion, but can also occur in association with other genetically determined conditions such as epidermolysis bullosa and/or aplasia cutis congenita or form part of the hereditary multiple intestinal atresias (HMIA).<sup>2,3,4</sup> This is a report of two cases of isolated CPA, outlining aspects of diagnosis and management. The literature on the subject is also reviewed.

### Cases

A 2-day-old female, a product of 35-weeks gestation via a low cesarean delivery due to a transverse lie to a 25-year-old mother who had gestational insulin-dependent diabetes and polyhydramnios was referred because of non-bile stained vomiting. Her birth weight was 2.1 kilograms. Clinically, there was no evidence of other abnormalities and she had mild upper abdominal distension. Her abdominal x-ray showed dilated stomach with no gas distally (Figure 1). Gastrograffin meal (Figure 2) confirmed the diagnosis of gastric outlet obstruction. She underwent surgery, and intraoperatively was found to have pyloric atresia due to an intact diaphragm. This was excised via a longitudinal incision in the pylorus, which was then closed transversely. Subsequently, she did well and was discharged home 3 weeks postoperatively.

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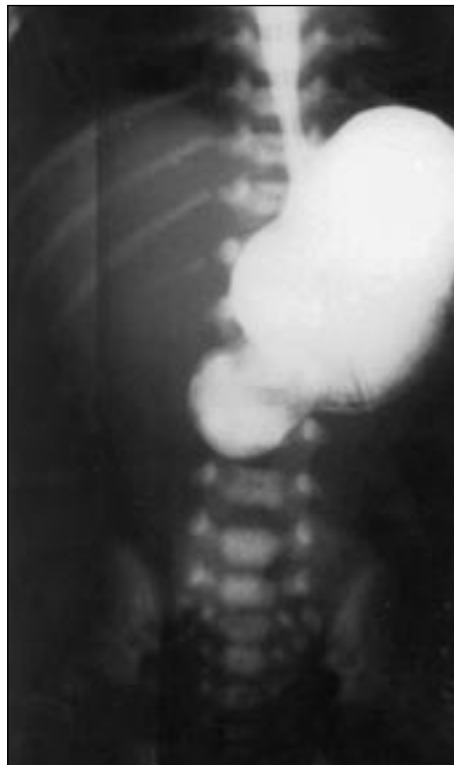
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*Accepted for publication:  
January 2004*

Ann Saudi Med 2005;25(2):165-167



**Figure 1.** Plain abdominal radiograph showing a dilated stomach with no gas distally.



**Figure 2.** Upper gastrointestinal study showing dilated stomach with gastric outlet obstruction.

A 1-day-old female, a product of full term delivery via a low cesarean delivery due to breach presentation to a 29-year-old mother with gestational insulin-dependent diabetes and polyhydramnios was evaluated because of persistent non-bile stained vomiting. Clinical examination revealed no abnormalities apart from mild upper abdominal distension. Abdominal x-ray showed dilated stomach with no gas distally and a barium meal confirmed the diagnosis of gastric outlet obstruction. She was operated on and during surgery was found to have congenital pyloric atresia secondary to a complete diaphragm. This was excised via a longitudinal incision in the pylorus, which was then closed transversely. Subsequently, she did well and was discharged home 3 weeks postoperatively.

### Discussion

In 1749, Calder first described CPA, which is a very rare malformation with an estimated incidence of 1 per 1 000 000 newborns.<sup>1,5,6</sup> Our hospital is the main maternity and children hospital in the Eastern Province of Saudi Arabia with an estimated 10 000 annual deliveries. During the last 20 years there were a total of about 200 000 deliveries and during the same period, we saw only 2 cases of CPA. This gives a local hospital based incidence of CPA of 1:100 000 newborns.

CPA can occur in one of three distinct forms:<sup>7</sup> as an isolated lesion,<sup>2,8</sup> in association with other gastrointestinal atresias as part of the HMIA, and in association with epidermolysis bullosa and/or aplasia cutis congenita. As an isolated lesion, it usually has a good prognosis. In association with other gastrointestinal atresias as part of the HMIA is hereditary, transmitted as an autosomal recessive inheritance. The atresias are often multiple and involve different regions of the gastrointestinal tract. Lambrecht and Kluth in 1998, in an extensive review of the literature, found only 35 well documented cases of HMIA.<sup>4</sup> Unfortunately, none of the reported cases of HMIA have survived. Sepsis was the main cause of death in the majority of them, and because of this, the possibility of associated combined cellular and humeral immunodeficiency should be excluded.<sup>9,10</sup> An association with epidermolysis bullosa and/or aplasia cutis congenita is genetically determined and inherited as an autosomal recessive.<sup>11</sup> Patients

with epidermolysis bullosa generally have a fatal outcome due to sepsis and fluid loss, and because of this, many surgeons do not advocate surgery when there is an associated pyloric atresia.<sup>12</sup> Recently however, there have been encouraging reports of infants surviving with epidermolysis bullosa.<sup>3,14</sup> This is especially the case with the use of phenytoin and steroids.<sup>3,15</sup> Both our patients had an isolated CPA due to a diaphragm and both have survived. In both our patients there was a history of gestational insulin-dependent diabetes. The significance of this is not known.

The diagnosis of CPA is based on the presence of non-bile stained vomiting and a single gastric air bubble with no gas distally on plain abdominal radiographs (Figure 1). This can be confirmed by an upper gastrointestinal contrast study (Figure 2). The association of CPA with epidermolysis bullosa and/or aplasia cutis congenita is evident clinically, but the association with other gastrointestinal atresias must always be kept in mind. This is especially true in the presence of scattered calcification on plain abdominal radiographs, which is seen in the majority of patients with HMIA.<sup>4</sup> These patients should have a contrast enema as well to rule out or locate the site of a possible associated colonic atresia.

Anatomically, CPA is divided into 3 types: 1) pyloric membrane or pyloric diaphragm, 2) pyloric atresia without a gap, and 3) pyloric atresia with a gap. Both our patients had pyloric diaphragms. Congenital pyloric diaphragm is usually single, but there are reports of double pyloric diaphragms.<sup>3</sup> This is of great importance intraoperatively as failure to recognize this will result in persistence of symptoms. The treatment of CPA is surgical and depends on the type of atresia. For those with pyloric diaphragm or pyloric atresia without a gap, the treatment is excision of the diaphragm and Heineke-Mikulicz pyloroplasty. Intraoperatively, it is important to locate the site of obstruction especially in those with a diaphragm, and to obviate missing a windsock diaphragm, a catheter should be passed distally via a small gastrotomy. This is also of importance in case there is more than one diaphragm. For those with pyloric atresia with a gap, if the gap is short, they should be treated with a Finny or Heineke-Mikulicz pyloroplasty, but if the gap is long then a gastroduodenostomy becomes the treatment of choice.

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