

## CONGENITAL PYLORIC ATRESIA (CPA): Report of two cases, with review of literature

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### ABSTRACT

We are reporting two neonates with isolated pyloric atresia, with an objective to emphasize the importance of considering this rare condition in the differential diagnosis of upper intestinal atresias. Both had non-bilious vomiting and epigastric fullness. X-ray showed a dilated gastric shadow with no evidence of gas in the rest of the abdomen. On exploration they had pyloric atresia which was corrected with gastroduodenostomy.

Congenital pyloric atresia is a rare condition, which presents with features of gastric outlet obstruction. It can be diagnosed antenatally but the picture can mimic other conditions. Epidermolysis bullosa has a strong association with it and require skin biopsy for diagnosis.

**KEY WORDS.** Congenital pyloric atresia, Gastric outlet obstruction

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### INTRODUCTION

Congenital Pyloric Atresia (CPA) is a rare condition representing less than 1% of all atresias of the alimentary tract and incidence is usually 1 in 100,000 live births.<sup>1,2</sup> Almost all

cases of gastric atresia occur in the pyloric region.<sup>2</sup>

CPA may present as a membrane occluding the lumen, as a gap in continuity, or a fibrous cord intervening between patent portions at the gastroduodenal junction.

CPA commonly occurs in isolation, it has rarely been reported in association with other alimentary tract atresias.<sup>2</sup> When occurring in isolation, the clinical course usually is uncomplicated after surgical treatment. It has a strong association with Epidermolysis Bullosa which has to be excluded in all cases of CPA.

### CASE I

A full term male baby was referred with history of vomiting, upper abdominal distention and respiratory distress. On examination the baby was active and pink but had tachypnoea. He did not have any skin lesion. There was fullness in the epigastrium. The abdomen was otherwise soft. Plain radiograph of the abdomen showed only the gastric bubble. Rest of the abdomen was gasless. An abdominal

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ultrasonography also showed gastric dilatation. Esophageal atresia was ruled out by passing a nasogastric tube.

A diagnosis of proximal duodenal atresia was made.

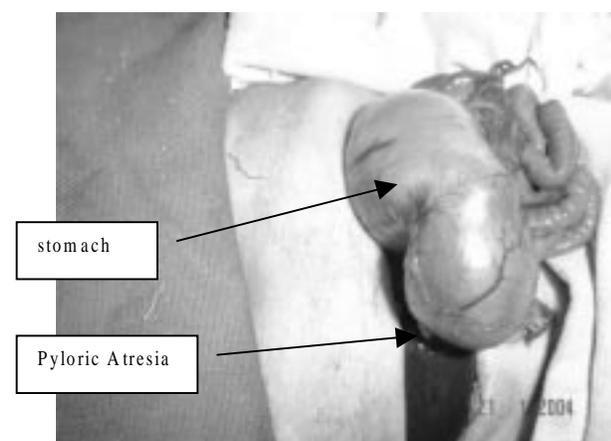
After initial resuscitation the baby was explored. Stomach was found enormously distended with atresia at the pyloric region due to a membrane. Distal patency was confirmed flushing saline into the distal gut. A gastroduodenostomy was performed bypassing the pyloric atresia. The baby had an uneventful recovery and was sent home.

Skin biopsy was not performed because the patient had no skin lesion and no family history of epidermolysis bullosa.

### CASE II

A 5 days old male baby presented with persistent non-bilious vomiting since birth. The baby had passed meconium on second day of life. On examination the baby was otherwise normal. Abdomen was flat, soft and no mass was palpable. X-ray abdomen showed a large distended stomach shadow. Rest of abdomen was gasless. Contrast study showed only the gastric shadow. Rest of the gut did not fill with the contrast material.

On exploration a complete pyloric atresia was found with a 3-cm. long cord connecting the dilated stomach to the atretic duodenum.



Picture showing enormously dilated stomach with pyloric atresia

A gastro duodenostomy was performed. The baby recovered uneventfully and was sent home after a week.

### DISCUSSION

Pyloric atresia is a congenital anomaly manifested by features of gastric outlet obstruction. There are three distinct groups of pyloric atresias: (i) isolated pyloric atresia, as in our case (ii) pyloric atresia in association with other alimentary tract atresias; and (iii) pyloric atresia in association with genetic disorders such as epidermolysis bullosa (EB) and aplasia cutis congenita.<sup>3</sup> The association between EB and CPA is rare, but is a known distinct clinical entity with autosomal recessive inheritance. The outcome of such an association is universally fatal.<sup>4</sup>

Clinically CPA presents with non-bilious vomiting since birth and upper abdominal distension. Maternal polyhydramnios occurs in approximately 50% of cases. Respiratory problems are common with CPA. Dyspnea, tachypnea, cyanosis and excessive salivation are often mistaken for esophageal atresia.<sup>5</sup>

A familial occurrence of CPA has been reported in 13 infants in 6 families.<sup>6</sup> Antenatally pyloric atresia can be diagnosed by presence of double bubble sign, which disappears on antral peristalsis. This can be misdiagnosed as congenital duodenal obstruction. To differen-



Contrast study showing the dilated stomach shadow. Rest of abdomen being gasless

tiate CPA from congenital duodenal obstruction, whole stomach configuration needs to be studied by continuous observation. This can be performed at different times when gastric peristalsis is active as well as quiet.<sup>7</sup> In our patients antenatal scans were not performed so both the cases were diagnosed after birth.

Plain x-ray abdomen is diagnostic and shows gas filled stomach, with complete absence of gas in the remainder of the gastrointestinal tract. Our patient had similar radiological findings. In the erect abdominal film this gives rise to a single air-fluid level in the stomach, which has been named as topped single bubble sign, the pyloric dimple sign and the absence of the beak sign. Neonatal gastric hypotonia may give a similar appearance.<sup>7</sup>

An antenatal ultrasound in the second trimester can pick up the pathology by noticing polyhydramnios and a cystic mass in the upper abdomen (dilated stomach) of the fetus.<sup>8</sup> Our patient had single gastric bubble with no flow distally.

CPA can recur in cases of consanguineous parents so there should be emphasis on prenatal diagnosis and counseling. A high index of suspicion in affected families helps in early diagnosis and treatment with better outcome.<sup>9</sup> Definitive surgery for CPA is gastroduodenotomy or Mikulicz procedure. All children with pyloric atresia should be screened for other anomalies. Association with EB should not preclude surgical treatment. A skin biopsy specimen should be taken if there is a family history of EB or bullous skin lesion.<sup>10</sup> The prognosis of isolated CPA has improved remarkably in the last two decades.<sup>11, 12</sup>

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