Case Report

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Pyloric atresia: A report of ten patients

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ABSTRACT

Pyloric atresia (PA) is uncommon. It occurs in 1:100000 live births. Neonates usually present soon after birth with copious non-bilious vomiting. The treatment is surgical and its prognosis is poor, especially, when it is associated with epidermolysis bullosa (EB). The aim of this study was to evaluate the clinical presentation, diagnosis, operative management, post-operative courses, and outcome in infant with PA, based in our cases and literature review. Charts of 10 patients who underwent surgery for PA in the department of paediatric surgery in a Teaching Hospital in Tunisia (Monastir) between 1990 and 2012 were reviewed. Data were analysed for demographic, clinical, therapeutic, and prognostic characteristics. The average of age at presentation was 2 days and there were six males and four females. The main presenting symptoms were non-bilious vomiting in 90% of cases. Abdominal X-ray showed gastric dilatation with an absence of gas in the rest of the intestinal tract in 90%, and a pneumoperitoneum in one. The surgical approach was laparotomy in all cases. Gastric perforation was observed in one patient and was completely repaired. The distribution of the anatomic variations was type A in nine cases and type B in one. Five patients underwent excision of the diaphragm and Heineke-Mikulicz pyloroplasty and gastroduodenostomy in the other five cases. Identified associated anomalies were Down's syndrome in one and EB in 2 (20%), one family has three affected sibling. Post-operative mortality rate was 70%. No standard surgical approach can be adopted a better management of PA or the associated anomalies depends on an early diagnosis and the availability of neonatal intensive care unit.

Key words: Epidermolysis bullosa, pyloric atresia, surgery

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INTRODUCTION

Pyloric atresia (PA) is uncommon and it occurs in 1:100000 live births.[1] Neonates usually present soon after birth with copious non-bilious vomiting. Its prognosis is poor, especially, when it is associated with other abnormalities, the most common being epidermolysis bullosa (EB). The aim of this report was to document our experience in management of PA over the within a 22 year period.

MATERIALS AND METHODS

We reviewed 10 patients who underwent surgery for PA in the department of paediatric surgery in a Teaching Hospital in Tunisia (Monastir), between 1990 and 2012. Data were analysed for demographic, clinical, therapeutic, and prognostic characteristics.

RESULTS

The average age at presentation was 2 days (extreme between 1 and 7 days) and there were six males and four females and were all full-term born. Among the five patients who underwent prenatal ultrasonography, four had signs of gastric outlet obstruction (polyhydramnios and gastric dilatation). The main presenting symptoms were non-bilious vomiting in 90% of cases. Four patients had upper abdominal distension at the time of presentation and one presented with gastric perforation and signs of hypovolaemia. Associated anomalies included Down's syndrome in one and EB in 2 (20%); there was a positive family history in one, affecting three siblings.

Routine laboratory test results showed leukocytosis in 20% of cases, severe anaemia in two cases. Electrolyte disturbances were noted in half of these cases. Radiological explorations included plain abdominal–X-ray, which was performed in all cases; an ultrasound in 4 patients (40% of cases), and an upper gastro-intestinal contrast in 6 cases (60% of cases).

Abdominal X-rays showed gastric dilatation with an absence of gas in the rest of the intestinal tract in9/10 [Figure 1] and a pneumoperitoneum in one. Upper gastro-intestinal contrast study revealed a dilated double loop of Stomach with significant constriction at prepyloric region in one case [Figure 2].

Children were resuscitated with nasogastric aspiration and intravenous fluid, transfusion, and antibiotics until the serum electrolyte correction, after which they all underwent laparotomy. Gastric perforation was observed in one patient and was completely repaired. The distribution of the anatomic variations was type A in nine cases and type B in one [Figure 3]. Five patients had excision of the diaphragm and Heineke-Mikulicz pyloroplasty, while the remaining five had gastroduodenostomy.

The mean hospital stay was 10 days (range: 1-days). The overall mortality rate was 70% (including the two patients with EB and the patient with Down syndrome) and the main cause of death was septicaemia. The average follow-up was 8 years. At 6-15 months after the operation, three patients are well, and no gastrointestinal disorders are present.

DISCUSSION

The aetiology of PA is unknown.[2] Anatomically, PA is divided into three different varieties: type A is a pyloric membrane, type B is a solid cord, and type C is a gap between stomach and duodenum.[3] Our finding agrees with reports in which type A is occurred most frequently.[2,4]

The typical presentation of PA is non-bilious vomiting soon after birth and minimal abdominal distension.[2,5,6] However, non-bilious vomiting may be confused with gastro-oesophageal reflux, leading to delay in diagnosis, which in turn may result in severe metabolic derangement and gastric perforation as was the case in this report.

An antenatal diagnosis of PA may be suggested by polyhydramnios, a dilated stomach, and narrowing of gastric outlet on ultrasonography.[5,6] These signs have been reported in 55% of PA.[2]

In post-natal, the diagnosis of PA may be suspected from plain abdominal films, which show a dilated stomach with the typical 'single bubble' appearance as seen in our cases.[6] Ultrasound may show a long stretched out pylorus, although, this was not encountered in this series.



Figure 1: Abdominal X-ray: gastric dilatation with an absence of gas in the rest of the intestinal tract

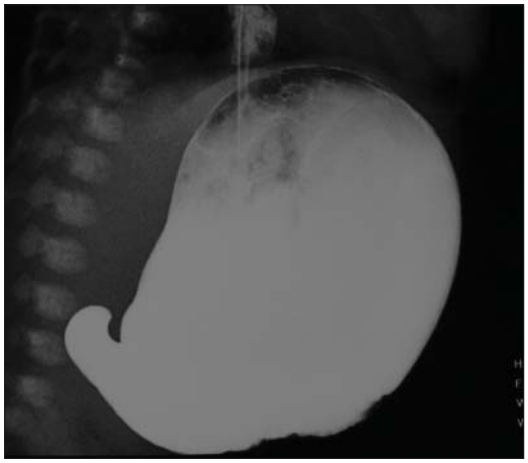


Figure 2: The upper gastro-intestinal contrast: a dilated double loop of stomach with significant constriction at pre-pyloric region

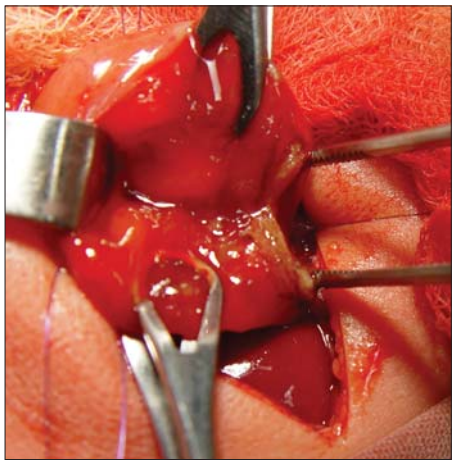


Figure 3: surgical exploration: pyloric atresia type A (pyloric membrane)

The treatment of PA is surgical and the approach varies. The treatment of choice for pyloric diaphragm is excision of the diaphragm and Heineke-Mikulicz pyloroplasty, while for type B or C, end-to-end or the diamond-shaped (Kimura method) gastroduodenostomy

is preferred.[2,6-8] Recently, an endoscopic technique has been developed to perforate the membrane and incise it with an electrocautery papillotome. Some authors advocate gastrostomy with local excision of the web to preserve the pyloric function, which is lost if a pyloroplasty is performed.[9,10]

PA may occur as an isolated condition or in association with other abnormalities. PA is associated anomaly in about 43.8-54.5% of cases.[2] The most common abnormality is junctional EB, an autosomal recessive genodermatosis.[11] Down's syndrome is also a common associate, whereas type A atresia predominates in non-EB cases, type B atresia predominates in association with EB. In our two cases of EB-PA syndrome, type A atresia was noted, this finding was also reported by Samad *et al*.[11]

The treatments of EB–PA syndrome are symptomatic including conservative managements such as appropriate dressing, infection control and nutritional supplements. Despite surgical treatment of concomitant PA, the prognosis is generally poor due to nutritional disturbance and sepsis in many cases. Therefore, if a prenatally diagnosis could be made and the denudation of fetal skin may be detected on ultrasound (a "snowflake sign"), a decision about terminating pregnancy could be considered.[2] Recently, genetic mutations as defect in genes for alpha 6 and beta 4 integrin subunit have been discovered.[12,13] Therefore, a genetic examination may be helpful for the diagnosis of EB.

The mortality rate is reported to be around 20-56.3%, 15.[2,14,15] This rate is lower than the 70% in our series. Higher mortality rate in PA is more when there is a presence of other congenital anomalies, especially, if there is neonatal intensive-care units and parental nutritional support.

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