Original Article

Congenital pyloric atresia: our experience

Sirajuddin Soomro, Sikandar Ali Mughal

Department of Pediatric Surgery Chandka Medical College Larkana, Pakistan

Objectives: To assess the clinical presentation, associated anomalies and outcome of patients born with congenital pyloric atresia.

Methodology: A case series study conducted at the Department of Pediatric Surgery, Chandka Medical College, Larkana, Pakistan from 2006 to 2014. All patients with Congenital Pyloric Atresia (CPA) were analyzed for above mentioned objectives.

Results: During the study period, 7 patients (4 Male and 3 Female) had CPA. Six (85.7%) babies were products of consanguous marriages. Most had low birth weight. All presented with non-bilious vomiting and epigastric distention. Epidermolysis bullosa, anorectal malformation and cardiac anomaly were noted. All patients underwent surgery. Type I CPA was the common anatomical lesion (71.42%). Heineke-Mickulicz pyloroplsty was performed in five cases while in two patients, gastroduodenostomy was used to bypass the obstruction. Five (71.4%) patients survived while 2 (28.6%) patients expired.

Conclusion: Congenital Pyloric Atresia is a rare cause of non-bilious vomiting in the newborn babies. If the associated anomalies are less severe and patient presented early and managed timely, the outcome can be better. (Rawal Med J 2014; 39:456-458).

Key words: Gastric outlet obstruction, congenital pyloric atresia, non bilious vomiting.

INTRODUCTION

Congenital Pyloric Atresia (CPA) is a rare congenital malformation representing less than 1% of all atresias of gastrointestinal tract.[1] In 18th century, Calder described the 1st case of complete pyloric obstruction.[2] The reported incidence of CPA is 1:100,000.[2] Familial occurrence of pyloric atresia has also been reported. Congenital pyloric atresia may have a membrane occluding the lumen, a fibrous cord intervening between the stomach and duodenum or a gap in continuity between the stomach and duodenum. Commonly, CPA presents as isolated lesion but associated anomalies such as epidermolysis bullosa, atresia of other parts of intestine and anorectal malformation have been reported in 30% of cases.[3] Early diagnosis and timely surgical intervention have improved the outcome. The surgical options for correction of CPA are pyloroplasty and gastroduodenostomy. Mortality has been associated with delayed diagnosis and presence of associated anomalies.[1] Here we are presenting our experience with CPA at our institution.

METHODOLOGY

A case series study was conducted at the Department of Pediatric Surgery, Chandka Medical College, Larkana, Pakistan from June 2006 to June 2014. All the patients who were diagnosed and managed as CPA were included in the study and analyzed for clinical presentation, associated anomalies and outcome.

RESULTS

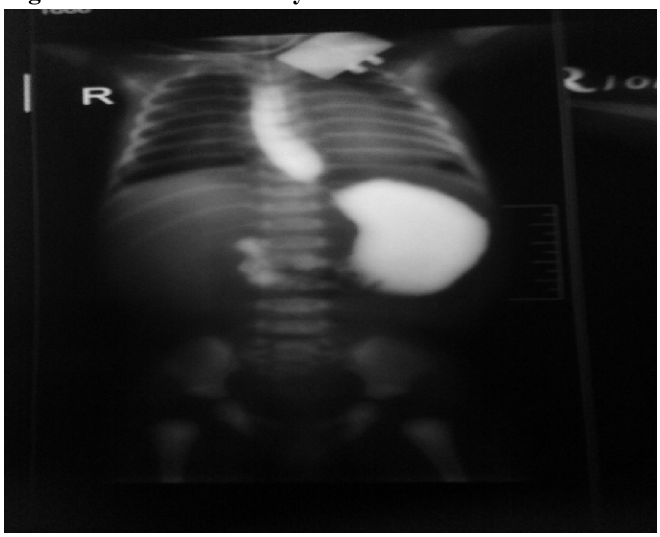
During the study period, seven cases of CPA were managed. There were 4 male and 3 female. Six were products of consanguineous marriages. Antenatal ultrasound of the mothers confirmed polyhyderamnios in 5 mothers. Four patients had low birth weight and 4 belonged to a single family.

Table. Case summaries

No	Age in days	Gender	Birth weight	Consanguinity	Antenatal Polyhyderamnios	Associated Anomalies	Type of Atresia	Opeation	Outcome
1	3	Male	2kg	+ve	+ve	None	Type I	Pyloroplasty	Expired
2	2	Female	2.2 kg	+ve	+ve	None	Type I	Pyloroplasty	Survived
3	1	Male	2.6 kg	+ve	-ve	None	Type II	Gastroduodenostomy	Survived
4	2	Male	2.3 kg	+ve	-ve	None	Type I	Pyloroplasty	Surgvived
5	2	Female	2.5 kg	+ve	+ve	Imperforate anus and Epidermolysis bullosa	Type II	Gastroduodenostomy	Survived
6	3	Female	2.5 kg	+ve	+ve	None	Type I	Pyloroplasty	Survived
7	5	Male	2 kg	-ve	+ve	VSD	Type I	Pyloroplasty	Expired

Case number 1, 2 and three were siblings and case number 4 was maternal cousin to them. All presented with non bilious vomiting and epigastric fullness. Epidermolysis bullosa, imperforate anus and Ventricular septal defect were the associated anomalies noted. Plain X-ray abdomen and contrast meal X-rays (Fig 1) were the main investigations used for diagnosis.

Fig. 1. Contrast meal X-rays.



All the patients underwent surgery. Type I pyloric atresia was found in 5 patients while in 2 patients the anomaly was type II. Heinecke-Mickulicz pyloroplsty was done in 5 patients and gastroduedonostomy was done in two. 5 patients survived while 2 expired due to septicemia.

DISCUSSION

Pyloric Atresia is a rare congenital malformation representing less than 1% of all atresias of gastrointestinal tract.[1] In 1987, Lorenzet and Morzer compiled 140 cases of pyloric atresia published in literature.[3] Almost all cases of gastric atresia occur in the pyloric region.[5]

There are three types of pyloric atresia (i) isolated pyloric atresia, (ii) pyloric atresia associated with other gastrointestinal atresias and (iii) pyloric atresia associated with genetic disorders such as epidermolysis bullosa and aplasia cutis.[6] Most of our cases had isolated CPA, however, one patient suffered from association of imperforate anus and epidermolysis bullosa and in one patient congenital septal defect of heart.

Anatomically there are three types of pyloric atresia: Type I = Pyloric membrane (57%), Type II = Pyloric tissue replaced by solid tissue (34%) and Type III = Pyloric atresia with a gap between stomach and duodenum (9%).[7,8] Five (71.42%) of our patients had type I anatomical lesion while in 2 (28.57%) it was type II.

It is reported that the association of epidermolysis bullosa is the commonest.[9,10] The associations we found were epidermolysis bullosa (EB), anorectal malformation and VSD. It is believed that pyloric atresia in association with EB results from intrauterine sloughing of the mucous membrane with subsequent healing by fibrous tissue leading to obliteration of the pyloric region.[8]

A familial occurrence and consanguineous marriages has been described as a contributing factors.[1,3,8] [3] Six of our patients were products of consanguineous marriages and three of them were consecutive siblings and one a maternal cousin to them, indicating the role of consanguineous marriages and that the condition runs in families.

Pyloric atresia can be diagnosed antenatally by ultrasonography in the 2nd trimester of pregnancy.[10] It confirms excessive amount of liquor and may demonstrate absence of normal echo pattern of pyloric muscle and the pyloric canal besides pyloric membrane.[1] In all of our patients, antenatal ultrasound was done which confirmed polyhyderamnios in 5 (71.4%) cases. Postnatally, the patients present with non-bilious vomiting and epigastric distention. Respiratory problems may or may not be present. Radiological diagnosis of CPA is based on presence of a single gastric air bubble with complete absence of gas in rest of the abdomen on plain X-ray of abdomen and absence of beak sign and presence of pyloric dimple on contrast study.[4,11] All of our patients presented with typical X-ray findings of pyloric atresia.

The treatment is surgical correction of the obstruction. The options available are Heinecke-Mikulicz pyloroplasty or gastro duodenostomy.[8,10,11] Gastrojejunostomy to bypass the obstruction is not advised and should be avoided, as it is associated with a high morbidity.[8] Most of our patients underwent Heinecke-Mikulicz (71.4%), while in 2

(28.6%) patients gastroduodenostomy was done to bypass the obstruction. Postoperatively, all patients did well initially but subsequently two of them developed signs of septicemia and expired making the mortality rate of 29%, which is on lower side as compared to literature. The reason of lower mortality could be due to the fact that the most of our cases were suffering isolated CPA and the associated anomalies were less in number and less severe.

Early diagnosis and surgical intervention with the neonatal supportive care have improved the survival of the patients born with CPA. The mortality has been associated with delayed diagnosis and mainly associated anomalies. The prognosis of isolated CPA is much better than the patients with associated anomalies. The overall mortality rate described in literature is around 50%.[7-9]

CONCLUSION

Congenital Pyloric Atresia is a rare cause of gastric outlet obstruction in newborn babies. The surgical options available are pyloroplasty and gastroduodenostomy. Isolated CPA carries a good prognosis. If presented early and managed timely the outcome can be improved.

Author Contributions:
Conception and design: Sirajuddin Soomro, Sikandar Ali Mughal
Collection and assembly of data: Sirajuddin Soomro
Analysis and interpretation of the data: Sirajuddin Soomro
Drafting of the article: Sirajuddin Soomro
Critical revision of the article for important intellectual content: Sikandar Ali Mughal
Statistical expertise: Sirajuddin Soomro
Final approval and guarantor of the article: Sirajuddin Soomro
Corresponding author email: soomrosiraj@yahoo.com
Conflict of Interest: None declared
Rec. Date: Jul 11, 2014 Accept Date: Sep 15, 2014

REFERENCES

1. Jasonni V. Pyloric atresia and prepyloric antral diaphragm, in Newborn Surgery by Puri P. Arnold. Edition 2nd, London, UK, 2003:383-387.
2. Samad L, Siddiqui EF, Arain MA, Atif M, Parkash J, Ahmed S, et al. Pyloric atresia associated with epidermolysis bullosa- three cases presenting in three months. J Pediatr Surg 2004;39:1260-69.
3. Puri P, Guiney EJ, Carroll R. Multiple gastro-intestinal atresias in three consecutive siblings: observations on pathogenesis. J Pediatr Surg 1985;20:22-4.
4. Nawaz A, Matta H, Jacobsz A, Al-Salem. Congenital pyloric atresia and junctional epidermolysis bullosa: a report of two cases. Pediatr Surg Int 2000;16:206-8.
5. Sencan A, Mir E, Karaca I, Günşar C, Sencan A, Topçu K. Pyloric atresia associated with multiple intestinal atresias and pylorocho-ledochal fistula. J Pediatr Surg 2002;37:147-51.
6. Al-Salem A, Nawaz A, Matta H. Congenital pyloric atresia. The spectrum. Int Surg 2002; 87:147-51.
7. Bhatnagar SJ. Pyloric atresia Type II. Neonat Surg 2013;2:36-8.
8. Al-Salem AH. Congenital pyloric atresia and associated anomalies. Pediatr Surg Int 2007;23:559-63.
9. Al-Salem AH. Congenital pyloric atresia, presentation, management and outcome: a report of 20 cases. J Pediatr Surg 2014;49:1078-82.
10. Agarwala S, Goswami JK, Mitra DK. Pyloric atresia associated with epidermolysis bullosa, malrotation, and high anorectal malformation with recto-urethral fistula: a report of successful management. Pediatr Surg Int 1999;15:264-5.
11. Bader I, Haq A, Akhter N, Abbas Z. Congenital pyloric atresia: Report of two cases, with review of literature. Pak J Med Sci 2004;20:256-8.