Pediatrik Cerrahi Dergisi, 8:107-110, 1994

Pyloric atresia: Report of four cases and a review of the current literature

Haluk SARIHAN, Rahmi AKYAZICI

Karadeniz Technical University Faculty of Medicine, Department of Pediatric Surgery, Trabzon, Turkey

Summary

Four cases of pyloric atresia have been surgically treated at our institution. These were male neonates who presented either with non-bilious vomitting (2 patients) or hematemesis (2 patients). Three of the patients with this condition could be diagnosed by plain abdominal x-ray alone. Their presenting conditions consisted of severe dehydration, electrolyte imbalances and metabolic alkalosis. Malnutrition was an additional finding in one of the babies.

The patients received medical treatment for a minimum of 3 days before undergoing surgical correction. The common finding at exploration was a complete pyloric web. This web was excised and the operation was completed with a pyloroplasty in all four patients. These babies have been found to cathc up in growth in their follow up and have not needed further treatment.

Key words: *Intestinal atresia, pylorus, pyloroplasty, child*

Introduction

Pyloric atresia (PA) is a very rare congenital anomaly manifested by acute symptoms of gastric outlet obstruction. The prevalance has been reported to be less than 1 % of all gastrointestinal atresias [3,17]. At least 142 cases have been reported in the literature to date [1,7-12,14-20].

We present four additional cases with congenital pyloric atresia to highlight the surgical features of this unusual condition.

Case Reports

Case 1: *A-five-day-old boy was admitted to our clinic with nonbilious vomiting, haematemesis and convulsion. Mother was noted to have polyhydramnios. The baby was found to be dehydrated and peristaltic movements could be observed in the upper abdomen. The plain radiograph of abdomen showed a large, airfilled stomach without any gas distally (Fig. 1). He demonstrated metabolic alkalosis with hypopotasemia, hyponatremia and hypochloremia. His serum bilirubin was 8 mg/dl.*

Following three days of fluid and electrolyte resuscitation laparatomy was performed through a transverse supraumbilical incision. A greatly distended stomach filled the abdomen and the intestines were unused. The nasogastric tube could not be guided through the pylorus. Gastrotomy revealed a membrane at the entrance to the pylorus. The pylorus was opened longitudinally and the atretic segment excised. Gastric and pyloric mucosa were rejoined and an Heineke-Mikulicz pyloroplasty was carried out. Four days after the operation, oral feeding was tried with semiliquid food. Full infant formula could be introduced by the eight day. At reexamination with barium meal after three month, pyloric passage was observed to be normal (Fig. 2).

Case 2: *A-ten-day-old boy, one of twins, had nonbilious vomiting from first feed. At admission, he had severe dehydration and malnutrition. X-ray of abdomen showed a large gastric bubble without any gas distaly. His serum bilirubin was normal, but there were serum electrolyte imbalances, metabolic alkalosis, and hypoproteinemia.*

Parenteral nutrition was started. On the seventh day of admission, laparatomy was carried out with supraumbilical transverse incision. The pylorus obstructed by a thick web. This web was excised and an Heineke-Mikulicz pyloroplasty was performed. Three days after the surgery, haematemesis and me-

Address: Dr. Haluk Sarıhan, Karadeniz Teknik Üniversitesi Tıp Fakültesi Çocuk Cerrahisi Anabilim Dalı, 61080, Trabzon-Turkey

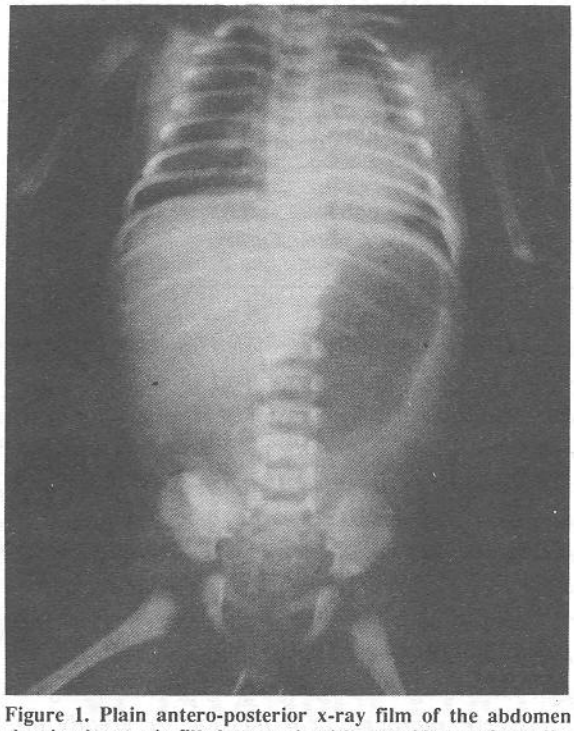

Figure 1. Plain antero-posterior x-ray film of the abdomen showing large, air-filled stomach without evidence of gas distally.

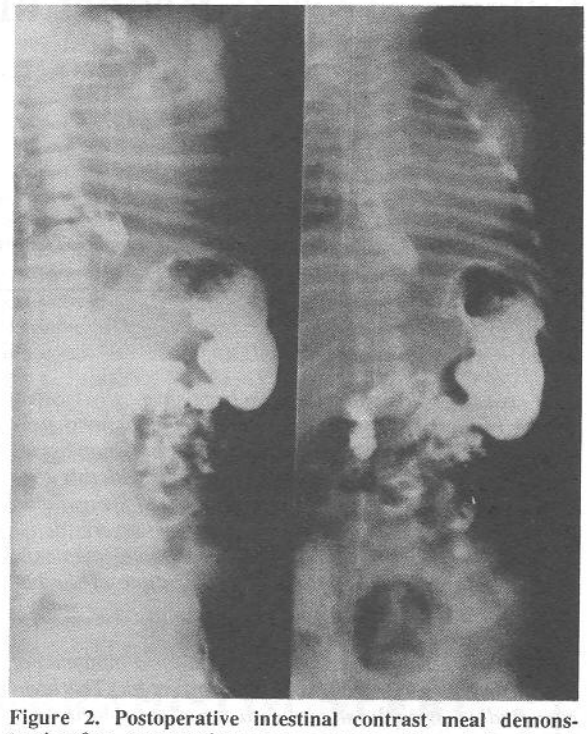

Figure 2. Postoperative intestinal contrast meal demonstrating free passage through a normaly wide pyloric chanel.

lena followed by stress ulcer developed. Conservative treatment including H2-receptor blocker led to prompt recovery. He was discharged on the twelfth day of his admission.

Case 3: *A fullterm boy was admitted to our clinic with hematemesis in the first day of life. Physical examination was unremarkable. After resuscitation with nasogastric decompression and saline irrigation, vitamin K injection, and H2-receptor blocker, hematemesis disappeared. Plain x-ray of abdomen demonstrated a "double-bubble" but contents of the nasogastric suction was nonblious and suggested complete pyloric obstruction. A contrast study showed a large stomach with air-fluid level and complete pyloric obstruction.*

Laparatomy revealed a distended stomach. The rest of the gastrointestinal tract remained collapsed. There was complete pyloric mucosal web extending into the duodenum. After web excision and mucosal repair a Heineke-Mikulicz pyloroplasty was completed. Postoperative period was uneventful and he was discharged in the seventh day.

Case 4: *A-four-day-old boy weighting 2.2 kg, was admitted to our clinic again with nonbilious vomiting. The mother was reported to have polyhdramnios. On examination he had dehydration and the stomach was palpable. He had serum electrolyte disturbance, metabolic alkalosis and hypoglycemia. Plain film of abdomen showed a large gastric buble without any gas distally.*

After resuscitation for two days he was operated; the pylorus was found to be obstructed by a thick web. Excision of the web was followed by a Heineke-Mikulicz pyloroplasty. In sixth day he was discharged with full feeding.

Discussion

Congenital gastric outlet obstruction due to PA is an rare malformation that reserves to be reminded. The first publication of this anomaly was made in 1749 by Calder and the first successful operation was described by Tourof and Susman in 1970 [12,19]. Etiology of PA is still obscure and the pathoembryogenesis is usually expained by the theory of re-

canalization failure of the gastrointestinal tract. Another suggestion is the fetal ischemic necrosis of the gut [4,12].

Pathologically and anatomically, there are three distinct types of pyloric obstruction [6,17,19];
(i) membranous pyloric obstruction, **(ii)** longitudinal segmental atresia, and **(iii)** pyloric aplasia. Patients with double membranes have also been reported [13]. Recently, Moore [12] has described a more comprehensive classification. The membraneous type of pyloric obstruction is more common and all of our patients were in this category.

Familial occurrence of PA has been reported. The inheritance occurs through an autosomal recessive trait [2]. The most constant feature of patients with PA is low birth weight [12,17]. The prominent feature is the finding of nonbilious vomiting. Maternal history of polyhydramnios is also possible. Haematemesis due to hemorrhagic gastritis has been observed occasionally [12,13,17]. These neonates frequently experience apnea, aspiration or respiratory distress from aspirations thick copious gastric secretion.

The diagnosis of PA is generally confirmed with plain radiographs of the abdomen. A stomach distended with air in an otherwise gasless abdomen is diagnostic [3,6,17,19]. When there is prolapse of a membrane into the duodenum (wind sock) a double-bubble sign can imitate duodenal atresia [12]. This condition was seen in our third patient. With the routine use of prenatal US, diagnosis can be made by demonstrating maternal hydramnios and fetal gastric dilatation [1,16].

Epidermolysis bullosa is a condition that has been associated with PA and carries a high mortality rate [9,10,12]. One explanation given for this association is that PA is acquired as an intrauterine complication. Mechanical and chemical injury of the pylorus could lead to denudation of the mucosa and consequent development of synechiae and fibrous cicatrization [5]. Thirty of the reported 142 cases with PA had epidermolysis bullosa [1,8-18]. PA associated esophageal atresia [12] and down syndrome [17] have also been reported.

The management of a neonate with PA is complex. Early recognition and intervention for PA with nasogastric decompression are imperative. Surgical correction is not urgent requirement and should be delayed until complete stabilization. Correction of dehydration and metabolic alkalosis will be needed even in the prenatally diagnosed case.

Standard techniques of reconstruction have yielded highly satisfactory results. These are Heineke-Mickulicz pyloroplasty for the pyloric web or short segment atresias and resection with gastroduodenostomy for long-segment luminal gaps [12]. In our experience that patients with PA should not receive gastrostomies jejunal tubes for feeding as a part of their treatment. After appropriate surgical correction, an excellent prognosis can be expected for isolated lesion.

References

1. Achiron R, Hamiel-Pinchas O, Engelberg S, et al: Aplasia cutis congenita associated with epidermolysis bullosa and pyloric atresia: The diagnostic role of prenatal ultrasonograph. Prenat Diagn 12:765, 1992
2. Bar-Maor JA, Nissan S, Nero S: Pyloric atresia: A hereditary congenital anomaly with autosomal recessive transmission. J Med Genet 9:70, 1972
3. Bronsther B, Nadeque MR, Abrams MW: Congenital pyloric atresia-A report of three cases and a review of the literature. Surgery 69:130, 1971
4. Campbell JR: Other condition of the stomach. In Welch KJ, Rowe MI, Ravitch MM, et al (Eds): Pediatric Surgery. Chicago, Year Book Med Publ 1986, p.821
5. Chang CH, Perrin EV, Bove KE: Pyloric atresia associated with epidermolysis bullosa: Special reference to pathogenesis. Pediatr Pathol 1:449, 1983
6. Çağlar MK, Ceyhan M, Dilmen U, Jenses A: Radiological case of the quarter. Turk J Pediatr 27:49, 1985
7. Hasegawa T, Kubota A, Imura K, et al: Prenatal diagnosis of congenital pyloric atresia. J Clin Ultrasound 21:278, 1993
8. Hayashi AH, Galliani CA, Gillis DA: Congenital pyloric atresia and junctional epidermolysis bullosa: A report of long term survival and a review of the literature. J Ped Surg 26:1341, 1991
9. Ishigami T, Akaishi T, Nishimura S, Yokoo T: A case of pyloric atresia associated with junctional epidermolysis bullosa. Eur J Pediatr 149:306, 1990
10. Lacour JP, Hoffman P, Bastiani-Griffet F, et al: Lethal junctional epidermolysis bullosa with normal expression of BM 6000 and antro-pyloric atresia: A new variant of junctional epidermolysis bullosa? Eur J Pediatr 151:252, 1992
11. Lestringant GG, Akel SR, Qayed KI: The pyloric atresia-junctional epidermolysis bullosa syndrome. Report of a case and review of the literature. Arch Dermatol

128:1083, 1992
12. Moore CCM: Congenital gastric outlet obstruction. J Ped Surg 24:1241, 1989
13. Müller M, Morger R, Engert J: Pyloric atresia: Report of four cases and review of the literature. Pediatr Surg Int 5:276, 1990
14. Nazzaro V, Nicolini U, De Luca L, et al: Prenatal diagnosis of junctional epidermolysis bullosa associated with pyloric atresia. J Med Genet 27:244, 1990
15. Nezir Z, Attar ZB, Moazam F: Congenital pyloric atresia and epidermolysis bullosa. J Pak Med Assoc 41:254, 1991
16. Paled Y, Hod M, Friedman S, et al: Prenatal diagnosis of familial congenital pyloric atresia. Prenat Diagn 12:151, 1992
17. Patel SB, Milstein JM, Schwartz ML: Radiological case of month. Congenital pyloric atresia in Down syndrome. Am J Dis Child 147:307, 1993
18. Roman MEO, Gutierrez JDS, Rodriguez JC, et al: Pyloric atresia-congenital epidermolysis bullosa syndrome: Report of a new case. An Esp Pediatr 35:419, 1991
19. Şenyüz OF, Söylet Y, İskit S, et al: Congenital pyloric atresia: Report of a successful case. Presental at XV. Annual International Meeting of Greek Association of Paediatric Surgeons, Greece. Abstracts 1990. p.53
20. Zorludemir Ü, Yücesan S, Olcay I: Pyloric atresia. A case report. Turk J Pediatr 30:119, 1988

WORKSHOP

PEDİATRİK ÜRODİNAMİ

2 Ekim 1995, İzmir

Düzenleyen

Ege Üniversitesi Tıp Fakültesi Çocuk Cerrahisi Anabilim Dalı • Pediatrik Üroloji Bilim Dalı

PROGRAM

2 Ekim 1995, Pazartesi

09.00-09.30	**Kayıt**	
09.30-09.40	**Açılış**	
09.40-10.45	Pediatrik ürodinami: Tanım ve yorum	İ. Ulman
	VUR ve enürezisde ürodinami	A. Avanoğlu
	Myelodisplazilerde nöroürolojik yaklaşım	S. Mutluer
	Temiz aralıklı kateterizasyon ve İYE	C. Kabasakal
10.45-11.00	**Tartışma**	
11.00-11.30	**Ara**	
11.30-12.30	Pitfalls in urodynamic investigations in children	R.M. Nijman
	Urodynamic investigations in the management of neupothic bladder	R.M. Nijman
12.30-13.00	**Tartışma**	
13.00-14.00	**Öğle yemeği**	
14.00-18.00	Klinik uygulamalar	R.M. Nijman
19.00	**Kapanış kokteyli**	

Konuk Konuşmacı

Rien J.M. Nijman (Erasmus Üniversitesi, Rotterdam)

Kurs yeri: M. Erel amfisi ve Çocuk Cerrahisi A.D. Ürodinami Odası, E.Ü. Tıp Fakültesi
Katılım ücretleri: Uzman 450.000.-TL, Asistan 250.000.-TL, Öğrenci ve Hemşire 150.000.-TL.
Başvuru adresi: Y. Doç. Dr. İbrahim Ulman, Ege Üniversitesi Tıp Fakültesi Çocuk Cerrahisi Anabilim Dalı, 35100 Bornova-İzmir Tel: 232 388 14 12 Fax: 232 342 21 42