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RADIOLOGICAL CASE OF THE QUARTER*

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A five-day-old boy was admitted to Hacettepe Children's Hospital because there had been persistent vomiting since birth. Physical examination revealed severe dehydration. There was no abdominal distention and bowel sounds could not be heard. There was no history of polyhydramnios. The family history was unremarkable. Measurement of electrolytes showed no abnormality. Direct abdominal roentgenograms were as shown below.

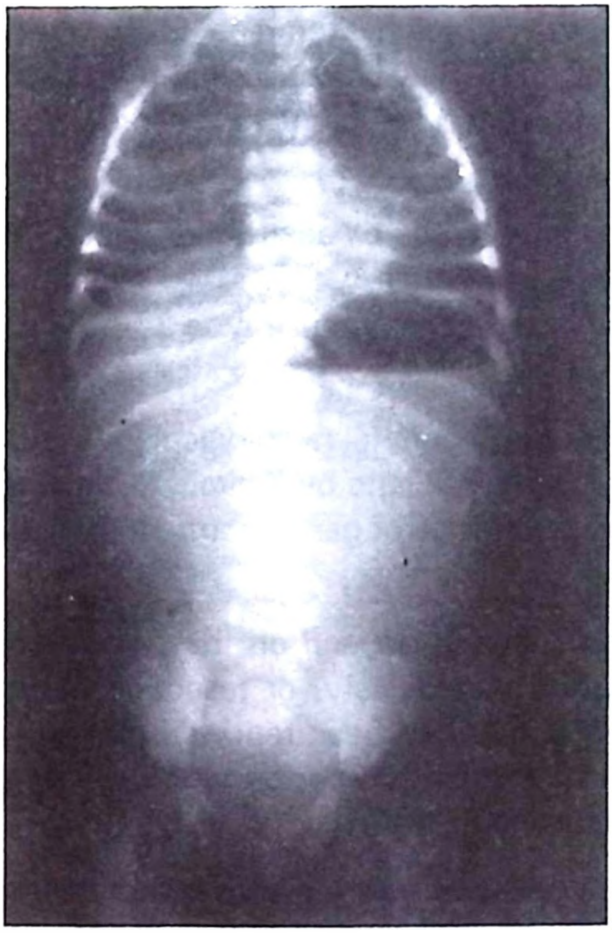

Figure 1

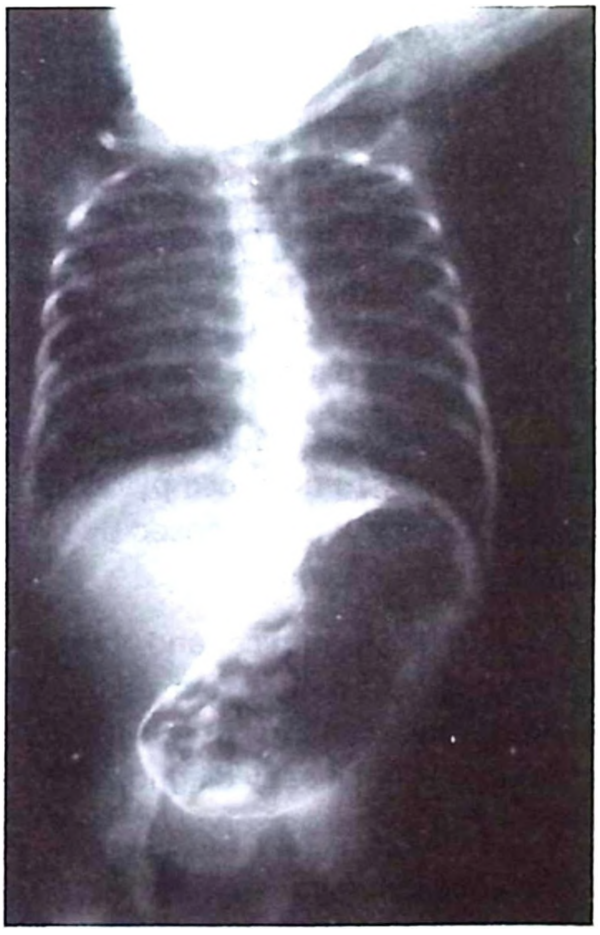

Figure 2

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Denouement and Discussion

PYLORIC ATRESIA

Figure 1. The abdominal X-ray showing an absence of air beyond the stomach.

Figure 2. The abdominal X-ray showing gastric dilation and absence of air beyond the stomach after air has been instilled.

Atresia of the pylor and of the stomach is rare, constituting only about 1% of all atresias of the gastrointestinal tract[1]. A congenital gastric outlet obstruction due to atresia was first reported by Bennett[2] in 1937. His report concerned the case of a four-day-old infant who was treated, unsuccessfully, for what was thought to be a hypertrophic pyloric stenosis. The autopsy of this infant showed a complete prepyloric septum.

The inheritance of pyloric atresia occurs by way of an autosomal recessive trait[3]. In approximately half of the cases, there is a history of polyhydramnios[4]. It has been suggested that the pathogenesis of this clinical entity may be a failure of recanalization of the intestinal tract or a mechanical or a vascular injury to the previously normal fetal intestine[1].

Pyloric atresia can be classified according to three types of obstruction[5]. A complete absence of lumen and two blind ends is known as type 1. Two separated segments connected by a fibrous cord or membrane characterize type 2. In type 3 there is a web or narrowed area.

Since air is the most useful contrast medium during neonatal radiological investigations, it is easy to diagnose pyloric obstruction in a neonate by a direct abdominal roentgenogram. It is diagnosed by the complete absence of gas or contrast medium beyond the stomach.

The treatment is surgical and the procedures required depend on the type of the obstructing lesion: excision of the septum and pyloroplasty, or resection of the atretic segment and gastrointestinal anastomosis[6]. It has been reported that prognosis is excellent if early diagnosis and surgical treatment are performed[7]. The prognosis is poor in the presence of other congenital anomalies.

In some cases congenital pyloric atresia is associated with epidermolysis bullosa letalis[5,6]. In a recent report[5] it has been emphasized that out of 70 cases of congenital pyloric atresia reported in the literature there were also 13 cases of epidermolysis bullosa letalis. The reasons for the association of these two rare disorders are unknown, but it has been suggested[8] that the concomitant occurrence of these two conditions represents the pleiotropic effect of a single gene, or that there is a close gene linkage between pyloric atresia and epidermolysis bullosa letalis.

Although pyloroduodenal atresia is a rare and usually isolated anomaly, oesophageal atresia has also been reported in association with pyloric atresia[9]. Since the presence of a second obstructing lesion, especially if it is unsuspected, can easily be overlooked at operation, this kind of combination is worth bearing in mind in cases of obstruction in neonates.

Key word: pyloric atresia

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