SVOA Paediatrics

ISSN:2755-1660

ScienceVolks Open Access

Case Report

Prepyloric Diaphragm in Children: A Diagnosis Not to be Missed.

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DOI: https://doi.org/10.58624/SVOAPD.2023.02.035

***Received:** May 29, 2023* ***Published:** June 13, 2023*

Abstract

Background: Prepyloric diaphragm is a rare cause of gastric outlet obstruction (GOO). Accurate diagnosis is often difficult and is usually confirmed after surgery. To avoid delayed management, this entity, with its specific clinical and radiological features, should be kept in mind.

Case presentation: We report a case of prepyloric diaphragm in a 7-month year old boy. He presented with non-bilious vomiting since birth. A first upper gastrointestinal series was initially misinterpreted as gastroesophageal reflux. After re-examination of the gastrointestinal series, a pyloric obstacle was suspected, and a second Contrast study evoked a pyloric obstacle which gave rise to the indication for surgical exploration after resuscitation. Endoscopy was suggestive of the diagnosis and the accurate diagnosis was done during surgery.

Conclusion: This report illustrates the complexity of this diagnosis. Knowledge of this entity and a good, detailed analysis of contrast study in combination with ultrasound and endoscopy could lead to a rapid diagnosis and an adequate care with a good prognosis.

***Keywords:** Gastric outlet obstruction; Prepyloric diaphragm; Non-bilious vomiting; Antral web.*

Background

Prepyloric diaphragm is a rare cause of gastric outlet obstruction (GOO) (2,3,4).

Symptoms of recurrent non-bilious vomiting can appear in the first days of life or later in children and even in adults (2,6).

Symptoms usually correlate with apertures of 1 cm or less (2,6).

Accurate diagnosis is often difficult and is usually confirmed after surgery. To avoid delayed management, this entity, with its specific clinical and radiological features, should be kept in mind.

We present a case report of a prepyloric diaphragm with a delayed diagnosis.

Case Presentation

A 7-month-old boy was admitted in our unit for nonbilious vomiting and failure to thrive.

The baby and his parents came from another neighboring country.

He was the product of full-term gestation. No antenatal examinations were done during the pregnancy.

He presented these vomiting since birth with increasing projectile vomiting for the last two months. He was initially treated conservatively by antiemetics by physicians without any clinical improvement. The child lost significant weight during this period.

On physical examination the child weighed 4.5 kg. He was noted to be dehydrated, lethargic and cachectic. He was afebrile with normal vital signs. The abdomen was soft, non-tender and not distended.

Laboratory investigations showed hypokalemic hyponatremic alkalosis. Upper gastrointestinal series performed in another center was initially interpreted as a massif gastroesophageal reflux (figure 1).

After re-examination of the gastrointestinal series a pyloric obstacle was suspected (figure 1). Another barium meal confirmed an obstruction at the level of the pylorus with a prepyloric transitional zone and a delayed passage of contrast from the stomach to the duodenum and an important gastro esophageal reflux (figure 2.a and b).

After resuscitation and adjustment of the malnourished state, the child underwent exploration under general anesthesia.

Initially, a gastroscopy was performed and showed a gaping cardia with a grade 2 peptic esophagitis and was suggestive of a prepyloric diaphragm but without certainty (figure 3).

A right transversal supraumbilical laparotomy was performed. We noted a hugely dilated stomach without any hypertrophy of pylorus. And a small mobile mass of 1 cm was palpated inside the stomach next to a narrowing of the pylorus (figure 4.a).

The pylorus was opened longitudinally over the stricture. The diagnosis of prepyloric diaphragm was confirmed (figure 4.b). This expanded and incomplete diaphragm prolapsed until pylorus was resected (figure 4.c) and the pylorus was transversally stitched (figure 4.d).

The baby was fed gradually with a thickened milk from the second post operative day and was discharged on the 5th post operative day with a gain of weight of 500 g.

Histopathology of the excised specimen showed the presence of mucosal and submucosal gastric mural layers.

The patient improved well postoperatively. Vomiting ceased and he started thriving well. He was under follow-up for the past 5 years.

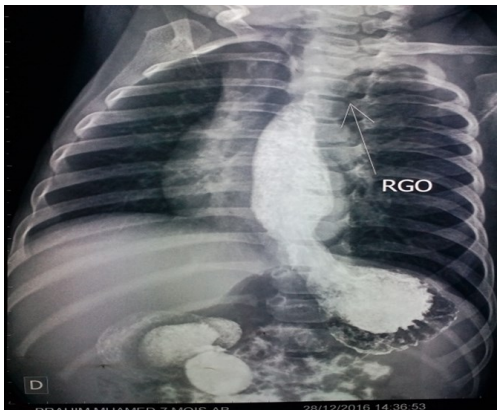

Figure 1: *The first contrast gastrointestinal radiographic studies interpreted as a massive gastroesophageal reflux and after re-examination a dilated antrum was noted and a non-specific obstacle at the level of the pylorus was suspected.*

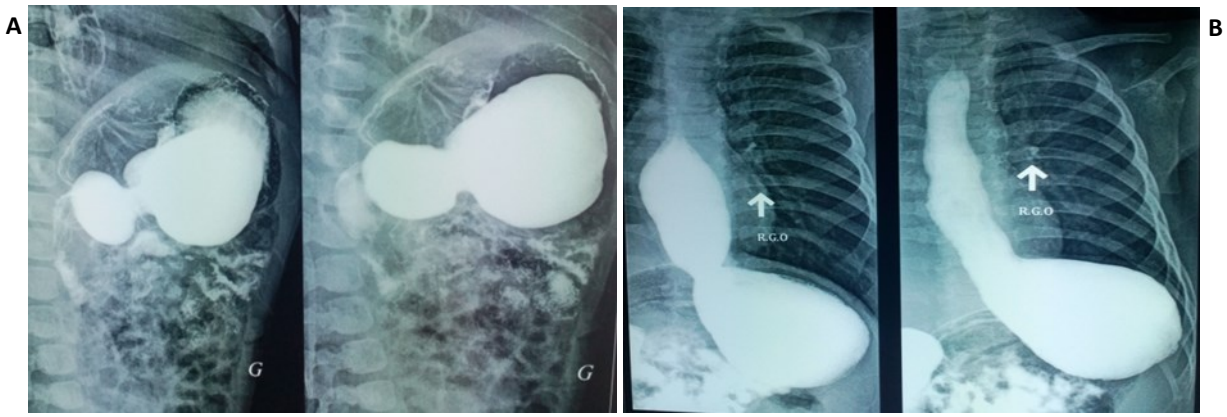

Figure 2: ***A.*** *upper gastrointestinal series done in our center showing a prepyloric transitional zone, a delayed passage of contrast from the stomach to the duodenum and a distended antropyloric region.*

B. *Important Reflux with a Dilated stomach and esophagus.*

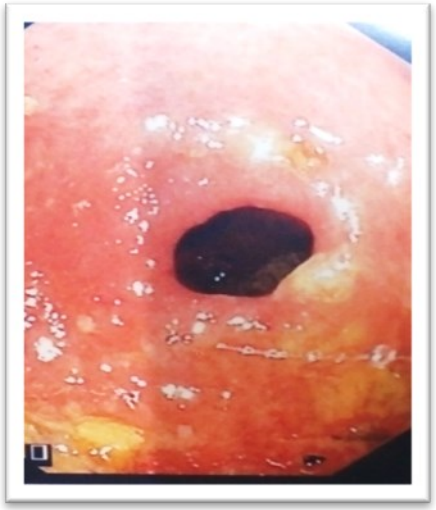

Figure 3: *Endoscopy suggestive of a prepyloric diaphragm.*

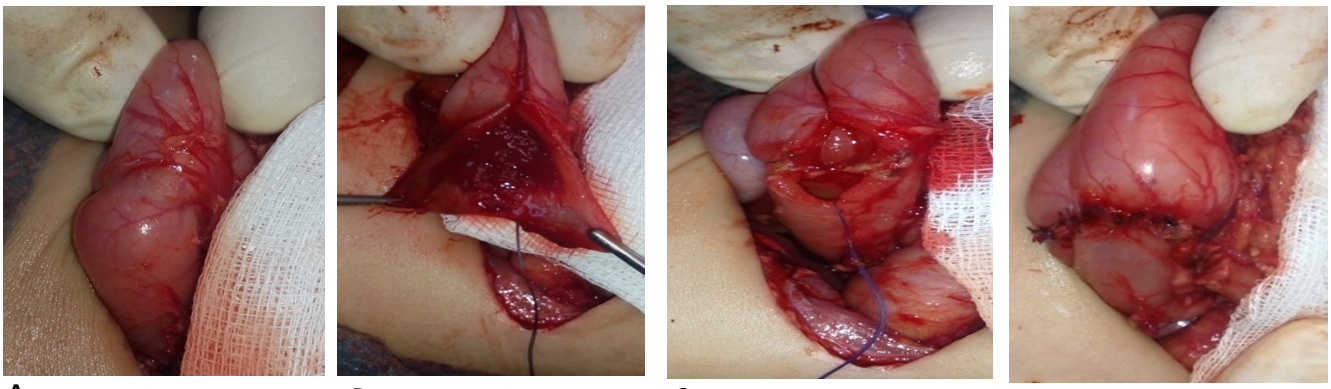

Figure 4: A. *Prepyloric stricture of stomach;* ***B****. The pylorus was opened longitudinally. An expanded antral incomplete diaphragm 1 cm proximal to the pylorus was found:* ***C****. The diaphragm was resected;* ***D****. Pyloroplasty.*

Discussion

Gastric outlet obstruction (GOO) due to prepyloric diaphragm was first described in 1937 by Bennett (1).

It is also called antral web (4). Its thickness is usually 2-4 mm and is located 1 to 3 cm proximal to pyloroduodenal junction (2,4,5).

It is a fenestrated diaphragm made up of two layers of mucosa and usually lacking muscular component (5).

It is an uncommon finding with an incidence of 1 in 100,000 births (4, 5).

If occlusion is complete, symptoms appear at birth, however, Incomplete prepyloric diaphragm are usually diagnosed later as in our case (3).

The etiology of the webs remains unknown. An incomplete recanalization of foregut during 5-6 gestational weeks has been evoked. Another hypothesis states that they may result from an excessive local endodermal proliferation early in gastric development (1,4,5).

Prepyloric diaphragm can be associated with other associated congenital anomalies such as duodenal stenosis, hypertrophic pyloric stenosis, malrotation, Meckel's diverticulum, cardiovascular anomalies like coarctation of the aorta and ventricular septal defect (1,4).

Symptoms usually correlate with aperture of 1 cm or less (6). Lumen sizes larger than 1 cm do not result in obstructive symptoms (5). Most children are symptomatic in the first year of live but other cases in later children or adulthood are reported (2,6). Persistent non bilious vomiting are the main frequent symptoms (1,4).

The diagnosis can be established using barium meal study by demonstrating the classic feature of the double-bulb appearance (4,5,6). It will show a lucent band across a contrast filled antrum, up to 7 cm from the pylorus. (2,6) as well as spraying of barium through a central aperture with a "jet effect" (4). Distension of the antrum beyond the aperture may be seen also (4). In our case, contrast gastrointestinal radiographic studies had led initially to a misdiagnosis of gastroesophageal reflux. It was only by reanalyzing the first exam and repeating it that we manage to evoke the diagnosis.

Endoscopy could be helpful for the diagnosis (2). In our report, it has not established with absolute certainty diagnosis.

Accurate diagnosis is difficult and often delayed even if patient undergoes upper gastrointestinal series and endoscopy (5).

Ultrasound might also be useful for the diagnosis. It could show the diaphragm as an echogenic structure (6, 4, 5), gastric dilatation, delay in gastric emptying and normal pylorus are additional us signs that may be seen (2,4). We haven't performed this exam for our patient.

Association of polyhydramnios dilated blind fluid filled sac of upper abdomen and absence of double bubble sign in antenatal ultrasound could raise suspicion of diagnosis (1). In our case, as the patient came from another country, prenatal follow-up was lacking.

Surgery remains the primary treatment for symptomatic prepyloric diaphragm with GOO (3). It includes simple diaphragm excision with or without pyloroplasty (1,4,5). In our case we performed excision of the diaphragm and a pyloroplasty.

The endoscopic intervention is another option for treatment. However, diaphragm transection is possible if the mucosal structure is uniform without major vessels or muscular or serosal layers (1,4,5).

Prenatal diagnosis, although difficult for this condition (1), should be improved and widespread, especially in developing countries. We think that this could be an additional diagnostic criterion that could help evoke diagnosis in addition to clinical, radiological and endoscopic features.

The prognosis after surgical treatment of this condition is good (4,5).

Conclusion

Prepyloric diaphragm should be considered as a possible cause of persistent non bilious vomiting in children. Its diagnosis remains challenging.

This entity must be known by practitioners especially pediatric surgeons, radiologists, endoscopists and pediatricians.

We highlight through this report that a good « in-depth » analysis and even repeating of radiologic investigations could lead to this rare Diagnosis to avoid delayed management.

Conflict of Interest

The authors declare no conflict of interest.

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Citation: Masmoudi M, Ben Jmaa K, Hsairi M, Guermazi M, Mhiri R. Prepyloric Diaphragm in Children: A Diagnosis Not to be Missed. *SVOA Paediatrics* 2023, 2:3, 68-72.