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Diagnosis and treatment of primary hypertrophic pyloric stenosis (HPS) in older children

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ABSTRACT

Hypertrophic pyloric stenosis (HPS) is the most important cause of non-bilious vomiting during infancy and it is usually idiopathic. It is very rare in older children and adolescents, in which gastric outlet obstruction (GOO) is more typically secondary to other conditions. Gastrointestinal malformation are frequently associated with Down syndrome and some of them (especially duodenal abnormalities) can be detected quite late. In infants the standard treatment of HPS is represented by a Ramstedt pyloromyotomy, while in adults a distal gastric resection is more indicated and it is usually associated to a Billroth I or II reconstruction.

Case report: We present an unusual case of a 12-year-old girl with Down syndrome, diagnosed with late-onset HPS. She came to our attention with periodic non-bilious vomiting and growth delay. She underwent multiple imaging examinations, which highlighted the presence of an HPS. Based on patient's age and the thickness of the pyloric muscle, we decided to perform a partial gastrectomy with a Roux-en-y reconstruction. Patient was discharged on the X day. She is now on follow up, gains weight and presents good conditions.

Discussion: HPS is rare but possible during childhood and adolescence. Diagnosis is usually performed through ultrasounds (US) and upper gastrointestinal radiological series (UGI), but sometimes further examinations are required (abdominal MRI, esophagogastroduodenoscopy). In older children since less invasive techniques are not effective, we have to consider partial gastrectomy. Based on our experience, Roux-en-y reconstruction is a valid option, associated with poor complication and good quality of life.

1. Introduction

Gastric outlet obstruction (GOO) refers to a broad spectrum of conditions that prevent the passage of gastric contents into the duodenum [1]. In pediatric population is a rare condition, with an incidence approximately of 2–5 per 1000, excluding infancy [2,3]. The usual clinical presentation consists of non-bilious vomiting, dehydration, and weight loss [4]. During infancy the most frequent etiology is idiopathic hypertrophic pyloric stenosis (IHPS), with over 90% of cases presenting between 3 and 10 weeks after birth [5]. After neonatal life GOO can be acquired as: peptic ulcer disease, gastric tumors, infections (such as tuberculosis), eosinophilic gastroenteritis and infiltrative diseases (such as amyloidosis) [2]. Late-onset primary hypertrophic pyloric stenosis (PHPS) is also reported in literature, although its etiology is unclear [4]. In this paper, we describe our experience in a case of HPS in a 12-years old girl with trisomy 21.

2. Case-report

A twelve-year-old female with trisomy 21 presented to our department with complaints of repeated vomiting and growth delay. She had a history of periodic vomiting and poor growth during infancy, partially solved after a period of gluten-free diet. Investigations for celiac disease and growth hormone deficiency came back negative. Therefore, she reintroduced gluten in the diet, with a good control of the symptoms until two years ago, when there was a deterioration.

Before coming to our department, she had already underwent an abdominal ultrasound (US), which showed a massive gastrectasia

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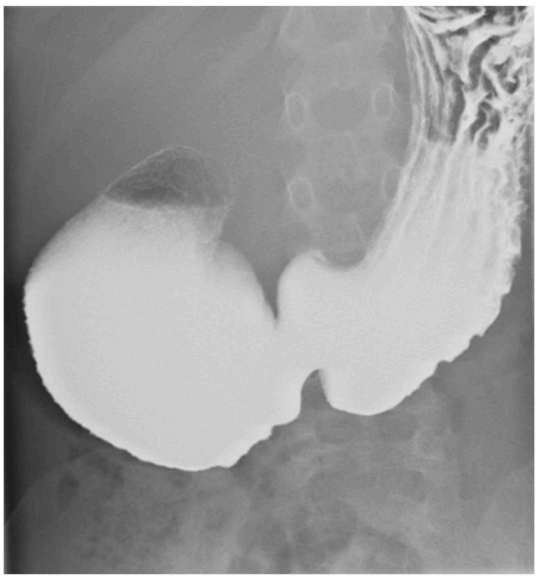

Fig. 1. Pre-operative UGI with evidence of tight duodenal stenosis and dilatation of the stomach.

reaching the sub-umbilical region and also upper gastrointestinal radiological series (UGI) with the evidence of tight duodenal stenosis and dilatation of stomach and duodenal bulb [Fig. 1]. Upon admission, she was apyretic and in good general condition. On physical examination, there were no pathological signs. Laboratory studies, including electrolytes, were normal, except for C-reactive protein (CRP) that was 7.15 mg/dl (N: 0-0.00-0.50 mg/dl). An abdominal MRI was performed: it excluded ab extrinsic compression and showed a dilatation of the stomach and of the first duodenal tract, probably due to a duodenal web. An esophagogastroduodenoscopy (EGDS) confirmed the huge dilatation and the presence of an obstacle to the passage of the instrument between the first and second duodenal portion, but not completely consistent with duodenal webs. During the same procedure electrosurgical incisions and endoscopic pyloric balloon dilatation (EPBD) were attempted. After one week, an UGI check highlighted a persistent obstruction, with a total inability of the contrast agent to pass into the duodenum. For this reason, we performed an additional EGDS with biopsy in the region of the pylorus and duodenum. On endoscopic examination, the esophagus was normal, there was a distended stomach with a large amount of gastric contents and a hypertrophic pyloric stenosis (HPS) traversable with the neonatal instrument and measured to be 8 mm thick at the US intraoperative examination. She was discharged with Esomeprazole therapy, waiting for the surgery.

At this point, we elected to operatively explore child, six months later, to gain more insight into the etiology of persistent GOO. Upon laparotomy exploration we decided to perform a distal gastrectomy with pylorus resection and a Roux-en-y reconstruction (Fig. 2).

The histological examination confirmed a pyloric muscle hypertrophy, without malignancy or evidence of Helicobacter Pylori (see Fig. 3).

She was given bowel rest, total parenteral nutrition (TPN) and triple antibiotic therapy (Metronidazole, Cefazolin and Gentamicin). On post-operative day 10, she was discharged home on a free diet and Esomeprazole 20 mg once a day. A post-operative UGI was performed, with regular bowel transit and no contrast spreading [Fig. 4].

One year after surgery, she was completely free from the symptoms and gained 7 kg.

3. Discussion

Excluding infancy, GOO is a rare condition, despite its exact rate in children/adolescent is unknown [2]. Moreover, HPS is typical of infants younger than four months, while it is a rare cause of GOO in children and adolescents [6]. Most of the time, HPS during infancy is to be considered idiopathic, and its exact etiology is unknown and likely multifactorial [1], including genetic, hormonal and environmental factors. After neonatal life, GOO is more typically secondary to other conditions. In 2008, Sharma et al. [7] distinguished GOO in childhood in two groups: the congenital group (including aplasia, atresia, diaphragms or pyloric' webs) and the acquired group (mainly secondary to acid peptic disease and neoplasia, to which are added a few other rarer causes such as chemical injury or eosinophilic gastroenteritis or pyloric achalasia for neuromuscular incoordination). A separate rare condition, is the also known the Jodhpur disease, presents during infancy or also later, with projectile non-bilious vomiting. The main difference from HPS is that pylorus is normal, without muscular hypertrophy [6–9]. In the adult group of patients, causes of pyloric obstruction are most commonly secondary, including peptic ulcer disease, hiatal hernia, carcinoma, or inflammatory bowel disease [4].

In the literature, there are no many cases of primary HSP in older children/adolescent [Table 1].

Considering the rarity of primary HPS in older children, other causes of GOO, both congenital and acquired, should be investigated. The most frequent gastrointestinal malformation in Down syndrome (DS) are duodenal malformations (i.e. atresia and stenosis) found in about 3.9% of patients with DS. This type of malformation can stay silent for a long time, also until adolescence and adulthood, especially in case of duodenal membrane [10]. Although the rate of HPS has still increased in children with DS (0.3% against 5 in 1000 in the general population), it remains a rather infrequent pathology [11]. Certainly more likely in a 12-year-old patient was a pyloric stricture (PS) secondary to other conditions (e.g. peptic ulcer disease, neoplasia, eosinophilic gastroenteritis, amyloidosis, etc …). The first step in our diagnostic approach should be an US examination, to determine if there is hypertrophy of the pylorus. Nevertheless, the pyloric thickening is often not appreciable at the US examination, with only an indirect sign of GOO that is the gastric dilatation. An UGI can also help to show if there is a normal passage of the contrast from the stomach to the duodenum and reveal gastric emptying time. Additional we can use a second level imaging studies (e.g. TC or MRI) to be of further diagnostic utility, especially to highlight the presence of a compression of the duodenum ab-extrinsic such as pathological masses, annular pancreas, mesenteric aorta compass, preduodenal portal vein, choledochal cyst or anomaly of the biliary tree as possible cause of GOO. Instead, the endoscopic examination allows us to analyze the volume of the stomach, the gastric and duodenal mucosa, the gastric content, the anatomical aspect of the pyloric canal, the duodenum and also allows the acquisition of biopsies of the punch for the evaluation of hypertrophy muscle, *Helicobacter pylori*, eosinophilic cell infiltrate, chronic inflammatory cells [3,12,13].

In our case, US and UGI highlighted a huge gastrectasia, until the sub-umbilical region, confirming the suspect of GOO, despite not giving information about the nature of this obstruction. MRI excluded the existence of masses compressing the gastric outlet, and suggested the execution of an endoscopic examination to evaluate the likely presence of duodenal webs. However, the EGDS performed ruled out the possibility of a duodenal obstruction and directed the diagnosis to a primary HPS, later confirmed by the histological examination.

The etiology of late-onset primary hypertrophic pyloric stenosis is not clear, but it's supposed to be infantile form, in which symptoms do not present until an intercurrent event occurs, such as oedema or inflammation [4,14,15]. In this regard, it is notable that our patient had suffered from suggestive symptoms already during infancy, but after a first medical management with a good control of the symptoms no further investigations where considered necessary.

Since it was described in 1912, the standard treatment for infantile HPS is Ramstedt pyloromyotomy [6]. Alternative procedures, after infancy, are balloon dilatation associated or not with endoscopic pyloromyotomy [16], laparoscopic or endoscopico or laparotomic

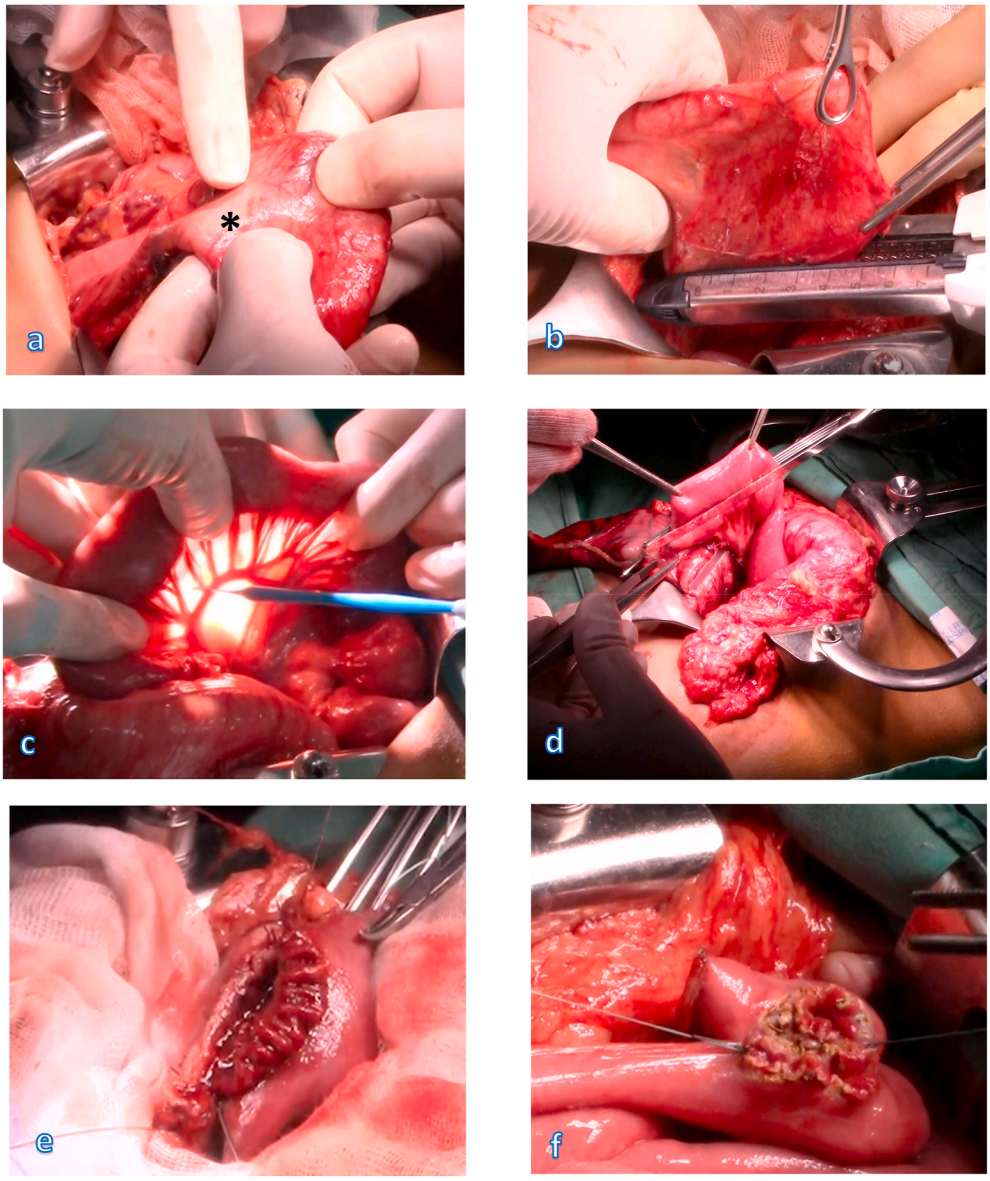

Fig. 2. (a) identification of the hypertrophic pylorus (*); (b) duodenal resection; (c,d) jejunal loop selection; (e) gastro-jejunal anastomosis; (f) jejuno-jejunal anastomosis.

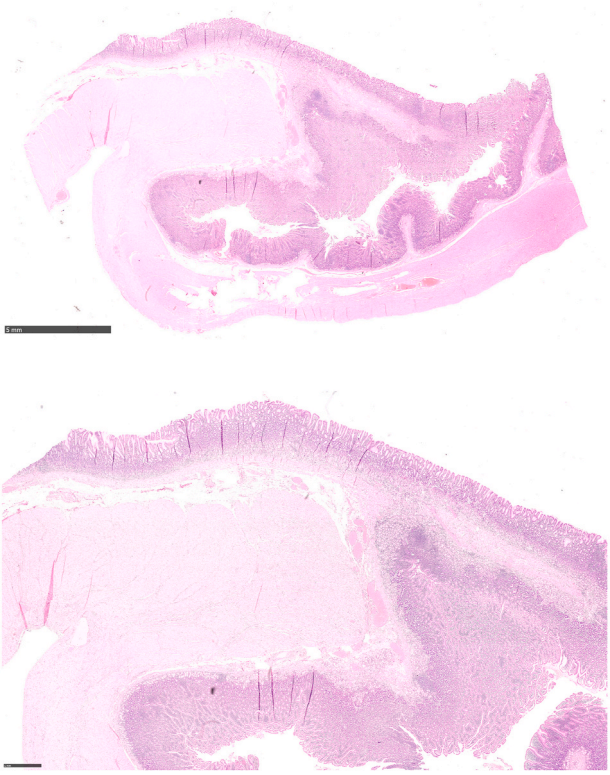

Fig. 3. Intraoperative specimens' cross sections showing muscle hypertrophy at the transition between the gastric and the duodenal mucosa.

pyloromyotomy, open (i.e. Heinke-Miculicz [7,17], Finney [8], Jaboulay [2], double [18]) or laparoscopic pyloroplasty [4,19], gastroduodenostomy (Billroth I) [2,6,8] or gastrojejunostomy (Billroth II) [6]. Different is primary HPS in adults, where most authors favor a limited distal gastric resection with Billroth I or II anastomosis, especially if the pylorus wall is very thick, making a pyloroplasty technically difficult [19].

In our patient, a first endoscopic approach (i.e. electrosurgical incisions and EPBD) was attempted without success. Later, we took into account several therapeutic options, but the extent of the obstruction and the poor impact of the previous treatment, together with the progressive worsening of the symptoms, made us choose a distal gastrectomy and resection of the pylorus. After a revision of the literature, we considered a Billroth I or a Billroth II reconstruction. However, the choice of reconstruction following a distal gastrectomy has to take into consideration the complications related to the specific post-gastrectomy physiology that will result. In particular, Roux-en-Y reconstruction appears to lead to reduced complaints of reflux gastritis and better quality of life compared with Billroth reconstruction (Billroth I or Billroth II) [20]. Another issue is represented by cancer risk after distal gastrectomy. Although absolute consensus is lacking, physiologic changes related to gastrectomy could produce a pro-carcinogenic state, maybe secondary to bile reflux, known to promote gastritis and metaplasia [21]. Moreover, many authors noted that this risk was not increased in the first post-operative years, but after more than 20 years [22]. This is why for a long time a Roux-en-Y reconstruction is preferred, especially in young patients. Therefore, we chose to perform a distal gastric resection with a Roux-en-Y anastomosis that to our knowledge is the first performed in a late-onset HPS.

Table 1
Review of the literature. In this table are shown studies that reported cases of late-onset primary hypertrophic pyloric stenosis in children/adolescents.

Paper	Number of cases	Mean age (years)	Treatment
Selzer et al. 2009	1	14	Heineke-Mikulicz pyloroplasty
Mahalik et al. 2010	1	4.5	Open pyloromyotomy
Boybeyi et al 2010	11	3.6	Balloon dilatation/Billroth I
Bajpai et al. 2013	1	8	Heineke-Mikulicz pyloroplasty
Parnall et al. 2016	1	15	Billroth I
Wolf et al. 2016	1	17	Laparoscopic pyloromyotomy
Bartlett et al 2018	1	12	Heineke-Mikulicz pyloroplasty
Oswari et al. 2020	1	11	Pyloric resection

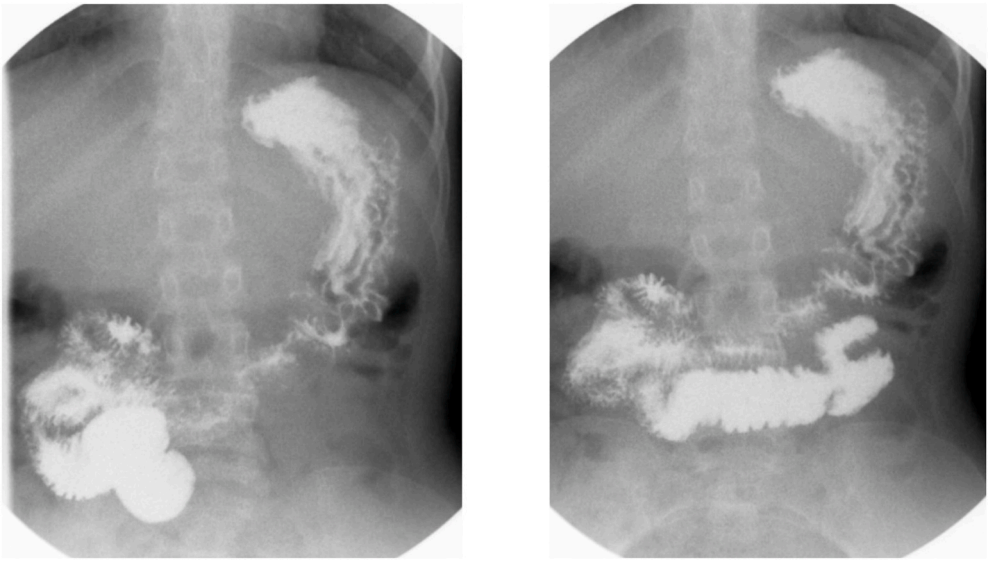

Fig. 4. Post-operative UGI with evidence of regular bowel transit and no contrast spreading.

4. Conclusions

GOO in older children is a condition as rare as disabling for the patient and the family. It is essential to perform a prompt diagnosis and treatment in order to ensure that children's physical and psychological development. Though late-onset primary HPS is a rare condition, we always have to consider it in the differential diagnosis of older children with symptoms of gastric outlet obstruction. In case of failure of less invasive procedures associated with a very hyperplasia of the pylorus, we suggest to perform a distal gastrectomy, choosing the type of anatomical reconstruction that best suits the surgeon's case and experience. We present our experience in a case of primary HPS in a 12-year-old female treated with distal gastrectomy and Roux-en-y reconstruction, which is known to be associated to fewer complications and better quality of life.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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