Congenital Pyloric Stenosis, Webs and Strictures

117

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Congenital gastric outlet anomalies (CGOA) are extremely rare and represent only about 1 % of all gastrointestinal atresias [1]. Embryologically, these malformations are most likely caused by a developmental disturbance of the distal foregut, which forms the antrum and the first, supraampullary portion of the duodenum [2]. In pathological nomenclature, the term represents several different entities like congenital pyloric atresia (CPA), a congenital pyloric stenosis (not to be interchanged with a hypertrophic pyloric stenosis) and intraluminal mucosal webs with or without a central hole (windsock anomaly) [2]. Even a non-folded stenotic ring separating two gastric chambers has been described [3]. Associated anomalies are common [4]. Especially hereditary multiple intestinal atresias (HMIA) are of major clinical importance [5], because these patients often suffer from severe forms of immunodeficiency [5, 6]. Today it remains unclear, whether such immunological defects occur primarily or secondary to the intestinal dysfunction [4]. Finally, an association of the CPA with an epidermolysis hereditaria bullosa (EHB) [2, 7] is well known.

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The first suspicion of a gastric outlet obstruction is raised during prenatal ultrasound investigations. Detection of polyhydramnion, presence of an extremely distended stomach or absence of intestinal filling and motility offer clues towards an impaired gastrointestinal (GI) passage in the foetus. Postnatally the clinical symptoms vary considerably in terms of onset, severity (acute versus chronic), tolerance of feedings and vomiting. A neonate with a complete obstruction (e.g. due to a CPA) usually presents with low birth weight, a small abdominal cavity and non-bilious vomiting. Older children (e.g. those with gastric webs) may complain about chronic and unspecific abdominal pain for some time, non-bilious vomiting and failure to thrive.

Most CGOA can be diagnosed by an upper GI study and an endoscopy. Plain abdominal films possibly demonstrate a “single bubble” and the absence of gas in the distal gut. Even webs, membranes and windsock deformities should be detected on fluoroscopic investigations. Endoscopy has an added advantage that it not only confirms the diagnosis but also enables the possibility for immediate treatment by laser coagulation or sharp dissection of webs. Recently Lin et al. [3] reported about their experience with congenital outlet obstructions of the GI tract. In 37 cases they found 12 gastric, 22 duodenal and 3 jejunal obstructions. Ten of the 12 gastric anomalies underwent an

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endoscopy and a detection of the lesion was possible in every case. Three of these anomalies were webs and could be resected concomitantly. The remaining nine gastric obstructions comprised of non-folded rings or atresias, which were resected. Al-Sahem [1] published a similar experience with 11 cases of gastric outlet obstructions. Intraoperatively, five had a pyloric diaphragm, three had a CPA with a gap between the two segments and two had a CPA without a gap. The pyloric diaphragms were excised during a Heineke-Mikulicz pyloroplasty. The CPAs received a gastroduodenostomy. Although most of his patients did well after the procedures, there was a late 45 % mortality due to sepsis that resulted from an associated immune deficiency.

In summary, children with isolated gastric outlet obstructions have a good prognosis. Prompt endoscopic confirmation and surgical treatment are recommended. However, the associations of EHB or HMIA imply a high mortality due to sepsis and severe immunodeficiencies. These patients require a multimodal therapy beyond surgery.

References

1. Al-Salem AH. Congenital pyloric atresia and associated anomalies. Pediatr Surg Int. 2007;23(6):559–63.
2. Gahukamble DB, Adnan AR, Al Gadi M. Distal foregut atresias in consecutive siblings and twins in the same family. Pediatr Surg Int. 2003;19(4):288–92.
3. Lin HH, Lee HC, Yeung CY, Chan WT, Jiang CB, Sheu JC, Wang NL. Congenital webs of the gastrointestinal tract: 20 years of experience from a pediatric care teaching hospital in Taiwan. Pediatr Neonatol. 2012;53(1):12–7.
4. Cole C, Freitas A, Clifton MS, Durham MM. Hereditary multiple intestinal atresias: 2 new cases and review of the literature. J Pediatr Surg. 2010;45(4):E21–4.
5. Bass J. Pyloric atresia associated with multiple intestinal atresias and immune deficiency. J Pediatr Surg. 2002;37(6):941–2.
6. Moreno LA, Gottrand F, Turck D, Manouvrier-Hanu S, Mazingue F, Morisot C, Le Deist F, Ricour C, Nihoul-Feketé C, Debeugny P. Severe combined immunodeficiency syndrome associated with autosomal recessive familial multiple gastrointestinal atresias: study of a family. Am J Med Genet. 1990;37(1):143–6.
7. Dank JP, Kim S, Parisi MA, Brown T, Smith LT, Waldhausen J, Sybert VP. Outcome after surgical repair of junctional epidermolysis bullosa-pyloric atresia syndrome: a report of 3 cases and review of the literature. Arch Dermatol. 1999;135(10):1243–7.