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Infantile Hypertrophic Pyloric Stenosis in Infancy: A Review Article

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Abstract

Infantile hypertrophic pyloric stenosis (IHPS) is the most common cause of nonbilious projectile vomiting in infants. Majority of cases present between 3 - 5 weeks of age and pyloromyotomy introduced by Ramstedt is the definitive treatment. In this review article in addition to historical aspects it is aimed to review the etiology, epidemiology, clinical manifestations and complications of IHPS in children under the light of relevant literature.

Keywords: *Infantile Hypertrophic Pyloric Stenosis; Children; Ramstedt's Pyloromyotomy; Complications*

Introduction

Infantile hypertrophic pyloric stenosis (IHPS) is a disorder of young infants which is caused by thickening of the pyloric portion of the stomach producing obstruction of gastric outlet. It is the most common surgical condition causing nonbilious projectile vomiting in infants. Pyloromyotomy is the preferred method of management since its description more than a century ago and this surgical intervention itself has remined relatively unchanged. In this review article, in addition to historical aspects of IHPS, a brief overview is given with regard to the etiology, epidemiology and current treatment options of IHPS under the light of relevant literature.

History

Although IHPS was first documented by Hirschsprung in 1888, both Hildanus in 1627 and Blair in 1717 had described the entity long before [1-3]. First division of pyloric muscle was performed by Stiles, a British Surgeon, in 1910 but the procedure was introduced by Conrad Ramstedt, a German Surgeon, in 1911 [4,5]. In this case he had used an omental "Graham patch" to cover the defect of mytomized pyloric muscle. One year later he performed the procedure without using omental patch leaving the pyloric muscle incised alone and since then the procedure itself has remined unchanged although the approach to the abdomen has continued to evolve.

Epidemiology

The incidence of IHPS is 2 - 5 per 1000 live births with geographic variation [6]. It is less common in Black and Asian population than White patients in the United States and because of this, the entity is considered as a "western disease" [7,8]. With a male predilection of 4:1 - 6:1 IHPS occurs usually (30 - 40% of cases) in first-born children and less common in infants of older mothers [6,9-17]. Typically symptoms begin between 3 and 5 weeks of life and bottle-fed infants are more commonly affected than babies fed by breast milk.

Etiology

Although the etiology of IHPS is unclear, there are several risk factors implicated in IHPS including genetic predisposition, environmental factors. Familial aggregation and associaton of IHPS with multiple genetic syndromes has been reported [9]. There are several genetic loci that are related to IHPS including APOA1 gene together with MBNL1 and NKX2-5 loci [18,19]. Cornelia de Lange syndrome is one of the geetic syndromes that has an association with IHPS [20]. Environmental factors include maternal factors including smoking and alcohol consumption during pregnancy, hyperthyroidism, nalidixic acid usage, young age, intranasal decongestant use [15,21-24]. Bottle feding of infants appears to be associated with increased risk with regard to IHPS [25]. Infants younger than 2 weeks of age medically treated with macrolide group antibiotics including erythromicin and azithromycin have increased risk for IHPS [14,26-30]. On the other hand prenatal exposure to erythromicin during pregnancy and premature birth are not associated with development of IHPS [31]. A disorder in nitric oxide pathway producing hypertrophy of the muscle and infectious etiology have also been suggested as other implicated factors producing IHPS [32-34].

Clinical Presentation

Characteristic symptoms include projectile nonbilious vomiting, palpable mass in the abdomen called "olive" and visible peristalsis especially after feding. Only 14% of children with IHPS present with all 3 signs of classic triad [35]. These children are usually hungry after vomiting. Palpation of "olive" has a 99% positive predictive value, and the frequency of this hypertrophied pyloric mass was high (up to 92 percent) in previous reports but, with the advances of imaging technology especially ultrasonography, these children are more commonly diagnosed earlier and palpation of "olive" in the diagnosis of IHPS is less common in subsequent reports and the clinicians have less practice at palpating the pyloric mass [10,36-38]. Jaundice may be seen in 2% of infants and is thought to be secondary to defective hepatic glucuronyl transferase activity [39,40].

Diagnosis

Accurate diagnosis of IHPS can be established with a history and physical examination alone in these children. Before physical examination the child should be relaxed and distracted. Palpation of "olive" in the abdomen is highly diagnostic but this may not be possible in inexperienced hands. An ultrasonographic finding of a pyloric muscle thickness, pyloric muscle length and pyloric diameter greater than 3 - 4 mm, 15 - 19 mm and 10 - 14 mm respectively, is generally considered diagnostic for IHPS [41-44]. If physical examination and ultrasonography are nondiagnostic, upper gastrointestinal (UGI) series may be used for diagnosis. Elongated pyloric canal (the "string" sign), thin tracks of radio-opaque agent along the pyloric canal (the "double track sign"), narrowing of pyloric canal (the "beak" sign) are the usual findings usually seen in UGI series.

The differential diagnosis includes gastroesophageal reflux, cow's milk protein intolerance, adrenal crysis, liver disease, other causes of intestinal obstruction, viral gastroenteritis, pylorospasm, hiatal hernia and other causes.

Preoperative management

Definitive management of IHPS is surgical pyloromyotomy. The disease itself is not a surgical emergency but medical emergency and surgical treatment should be performed once the patient is adequately resusciated. Initial fluid resuscitation aims to correct dehydration and acid-base derangements. The clasical observation is that of hypochloremic, hypokalemic metabolic alcalosis. This derangement in electrolyte balance is due to vomiting of gastric contents containing sodium, potassium, chloride and hydrogen ions. If adequate rehydration is not commenced immediately, kidneys conserve potassium at the expense of hydrogen ions leading to paradoxical aciduria which enhances alcalosis further [45]. Metabolic alcalosis has been reported to be associated with apneas and extubation difficulties [46]. Infants with normal electrolyte values and no dehydration should receive maintanence IV fluids such as 5% dextrose with one-half normal saline

(%0.45 NaCl) and 10 - 20 mEq KCl per L. In children with moderate or severe dehydration more intensive management by administering fluids with higher NaCl concentrations may be necessary.

Treatment

The classical operation for IHPS is Ramstedt pyloromyotomy involving extramucosal longitudinal splitting of pyloric muscle. This procedure may be performed using an open surgery or laparoscopic intervention. In open surgery, right upper abdominal transverse muscle splitting incision or circumumbilical approach introduced by Tan and Bianchi are the options [47]. Laparoscopic pyloromyotomy is a minimal invasive technique. In a previous study with regard to operation time, time to full feeding or length of hospital stay no differences were found between the children with IHPS operated via either open surgery or laparoscopic intervention [48]. On the other hand, infants treated via laparoscopic approach had fewer episodes of emesis and lesser need for analgesia [49]. Moderate vomiting as regurgitation occurs in as many as 80% of infants after pyloromyotomy [10]. If vomiting after surgey persists beyond 5 days postoperatively, radiological evaluation should be performed for a possible complication of incomplete myotomy [50].

Most important complications of Ramstedt pyloromyotomy are mucosal perforation and incomplete myotomy. Other postoperative complications include general complications of surgical interventions like wound infection, postoperative vomiting, bleeding, dehiscence of wound or fascia and subcutaneous emphysema. Luckily these complications with an incidence of 0.5 - 1.9% are rarely seen. Postoperative management include monitoring, analgesia and reintroduction of enteral feeds [46]. Many variations in feeding regimens following surgical treatment of children with IHPS exist. After a glance at literature on this subject it seems that majority of infants tolerate feedings postoperatively well.

Conclusion

In conclusion, IHPS is one of the hallmark diseases of pediatric surgery and is successfully treated with surgical pyloromyotomy. The disease itself is a medical emergency but not a surgical emergency. If the child with IHPS has dehydration or electrolyte derangements the surgical intervention for IHPS should be delayed and appropriate fluid and electrolyte therapy should be commenced promptly. Physicians should keep this entity in their minds when they face infants with nonbilious projectile vomiting. Rapid pediatric surgical consultancy should be taken by the front liners of medical providers dealing with these patients and these children should be treated accordingly.

Conflicts of Interest

The author certifies that he has no affiliations with or involvement in any organization or entity with any financial interest, or non-financial interest in the subject matter or materials discussed in this manuscript.

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Author Contribution to the Manuscript

Idea/concept, design, control and processing, analysis and/or interpretation, literature review, writing the article, critical review, references and materials by Volkan Sarper Erikci.

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