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RESEARCH ARTICLE

APPROACH TO INFANTILE HYPERTROPHIC PYLORIC STENOSIS

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ABSTRACT

Infantile hypertrophic pyloric stenosis (IHPS) is a common surgical cause of vomiting in pediatric population. It usually occurs as an isolated condition or together with other congenital anomalies. There has been shown an association with genetic and environmental factors, younger maternal age, maternal smoking, bottle feeding, and erythromycin administration in the first two weeks of life. The patients typically present with projectile vomiting associated with symptoms of dehydration and acid-base abnormalities. On physical examination, an olive-like mass palpable in the right upper abdominal quadrant has decreased significantly over time, because of earlier diagnosis by imaging in ultrasound. Infantile hypertrophic pyloric stenosis is usually corrected through laparoscopic or open pyloromyotomy. However, preoperative preparation is essential to optimal outcome. Fortunately, the overall mortality after pyloromyotomy is less than 0.4% in most major centers and long-term sequels are rare.

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INTRODUCTION

Infantile hypertrophic pyloric stenosis (IHPS) is a common condition in infants characterized by an acquired narrowing of the pylorus and the most commonly encountered surgical disease in children (Al-Ansari, 2016). The key function of the pylorus is to be a gate-keeper (from the Greek pulo ̄rus). Its role is to avoid large undigested food particles from entering the small intestines. Besides, the pyloric muscle inhibits food from re-entering the stomach when there's contraction in the small intestine. When the pyloric muscle is in a state of relaxation, it allows small food particles to pass to the duodenum (Peters, 2014). Progressive hypertrophy of the pyloric muscle results in obstruction of gastric emptying. This causes the classical symptom of progressively worsening projectile vomiting after feeding (Jobson, 2016).The incidence of IHPS varies with geographic and ethnic populations for unclear reasons (Pandya, 2012). There have been reports in the literature citing a varied etiologic factors, some of which are discussed below. In the last decades, many advances have been made in the diagnostic and, most importantly, surgical techniques concerning IHPS. Therefore, to date, morbidity and mortality rates associated with IHPS are low (Peters, 2014).

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History: Infantile hypertrophic pyloric stenosis (IHPS) has been described throughout the medical literature as far back as the early 1700s. It was first documented by Harald Hirschsprung, a Danish Pediatrician, in 1888 in two post-mortem cases, his description of the clinical history and findings ignited a flux of scientific enquiry into every aspect of the disease (Taghavi *et al*., 2017; El-Gohary, 2018).In 1908,Fredetwas the first one to propose a full-thickness incision of the pylorus followed by a transverse closure. Even though this was successful, Ramstedt modified the technique and later described the sutureless, extramucosal longitudinal splitting of the pyloric muscle, which left the mucosa intact, this one is continuing to be the guiding surgical technique (Pandya, 2012)

Epidemiology

IHPS usually presents in the first 2 to 12 weeks of life, with a peak incidence occurring during the fifth week of age. The incidence of IHPS varies with geographic and ethnic populations. For example, pyloric stenosis occurs in approximately 2 to 4 per 1000 live births in the Western population, whereas the incidence has been reported to be approximately 4 times lower in the Southeast Asian and Chinese populations. This difference in incidence around the world is still largely unclear (Peters, 2014; Pandya, 2012). It affects two to four times more boys than girls, especially the first-born.

As a mather of fact 5.5% of the sons and 2.5% of the daughters of an affected father develop IHPS in comparison to 20% of the sons and 7% of the daughters of an affected mother. Its incidence is higher in infants with blood groups B and Oand in infants born preterm as compared with those born at term (Kliegman *et al*., 2020; Krogh *et al*., 2011).

Etiology: The etiology of IHPS is still poorly understood despite its relatively high incidence. The cause is suggested to be multifactorial, with an important role for both genetic and environmental factors (Peters *et al*., 2014). IHPS may be present as an isolated entity, but is also associated with multiple genetic syndromes, suggestingthat genetic factors play a role in its etiology. Genetic syndromes such as Smith-Lemli-Opitz and Cornelia de Lange as well as chromosomal abnormalities including partial trisomy of chromosome 9, partial trisomy of chromosome 13 and partial monosomy of chromosome 18, and a translocation of chromosome 8 and 17. However, variations in the incidence of IHPS and the numerous reported environmental associations with IHPS suggest that it must, at least in part, be an acquired, as opposed to a congenital, condition. This is can be explained as various genetic and environmental factors contribute to an individual's "responsibility" towards the development of a disorder. Each factor has a small effect, but the effects add up and when a critical threshold liability threshold is crossed, illness occurs (Jobson *et al*., 2016; Panteli, 2009).

Pharmaceutical agents, environmental factors, hormones, and growth factors have all been related to IHPS through small case reports. For example, exposure of the lactating mother to erythromycin does show increased rates of IHPS in whom the drug is clearly contraindicated. Prostaglandins have also been implicated as causative agents. Other hormones, like prostaglandins at high levels have been found to be present in infants with IHPS, which suggests a positive link. Maternal risk factors that have been reported include: hyperthyroidism; nalidixic aciduse, young age, smoking, raised pre-pregnancy BMI (Pandya *et al*., 2012). Recently, the prone sleeping position has been suggested as a possible risk factor given the fact that it has been associated with increased risk of SIDS and the launch of the ‘‘back to sleep’’ campaign to prevent SIDS has coincided with the decline in the incidence of both IHPS and SIDS in Denmark and Sweden (Panteli, 2009). Bottle-fed infants experienced a 4.6-fold higher risk of IHPS compared with infants who were not bottle-fed. The result adds to the evidence supporting the benefit of exclusive breastfeeding in the first months of life (Krogh, 2012).

Clinical Presentation: Patients are commonly term infants who are otherwise healthy. The classical presentation is described as an infant with projectile, non-bilious vomiting just after being fed with formula or breast milk. They associate hypokalaemic, hypochloraemic metabolic alkalosis and a palpable, hypertrophied pyloric muscle, and an ‘olive’ in the abdomen. The palpating olive at physical examination has a 99% positive predictive value, whereas positive test feeds has inadmissibly high false- positive and false-negative rates and is therefore not used with frequency any more (2, 3).During examination, the physician must take advantage of the moment when the infant is sleeping or restingto palpate the mobile pyloric mass or olive. The abdominal wall must be completely relaxed or else palpation is nearly impossible. Emptying the stomach with a nasogastric or orogastric tube can sometimes make the olive more palpable (Pandya, 2012).

Several studies have shown that the classic electrolyte abnormalities of hypochloremic, hypokalemic, metabolic alkalosis is present in less than 50% of the patients with pyloric stenosis These interpretations were recently confirmed in a retrospective chart review; where theinfants with IHPS showing that normal laboratory values were the most common finding in these population and that metabolic alkalosis was found more commonly in the latter part of the decade and in older infants.This shows that, the increased use of ultrasound has led to an earlier confirmation of diagnosis with less opportunity for dehydration and electrolyte disorders (Peters , 2014; Jobson, 2016).Greater knowledge of pyloric stenosis has allowed earlier identification of patients, which has decreased the number of cases of chronic malnutrition and severe dehydration (Kliegman, 2012). It’s very important to remember that the absence of a palpable olive or negative radiographic evaluation for IHPS should prompt the surgeon to initiate a more extensive workup to evaluate the suspected intestinal obstruction (Pandya, 2012).

Diagnosis: The diagnosis has usually been established by the palpation of the pyloric mass. The mass is firm, hard, movable, olive shaped and approximately 2cm in length. Best palpated from the left side, and located above and to the right of the umbilicus in the mid epigastrium beneath the liver's edge (Kliegman *et al*., 2012) Positioning the infant supine and the legs bent to relax the abdominal muscles, the examining hand must be placed on the epigastrium. After the edge of the liver has been recognized with the fingertips, gentle pressure deep to the liver and progressing caudally in the midline a third of the distance between the umbilicus and xiphoid should expose a palpable pylorus if IHPS is present (Coran, 2012) The olive is easy to palpate after an episode of vomiting (Kliegman *et al*., 2020). This palpation of the olive can be of great value in diagnosis of this etiology, but this is not always possible, even in skilled physicians the rate of a successful palpation varies between 40 and 100% (Peters, 2014). The next step in diagnosing IHPS in most of the cases is imaging by means of ultrasound (Peters, 2014). The use of this imaging technique has increased since 1977 when Teele and Smith first published their paper on the use of diagnostic ultrasonography (Jobson, 2016). US has become not only the most common initial imaging technique for the diagnosis but also the standard for diagnosing IHPS (Coran, 2012). Besides the fact that is aninexpensive diagnostic modality, ultrasound is the most accepted and widely used method in diagnosing IHPS, especially when combined with the history of projectile emesis and the hypochloremic metabolic alkalosis (Peters, 2014)

The specificity and sensitivity of US in diagnosing IHPS, performed by an experienced pediatric radiologists, are very high with 98% and 100%, respectively. (Peters, 2014) Criteria for diagnosis by ultrasound are: pyloric thickness 3-4 mm, an overall pyloric length of 15-19 mm, and pyloric diameter of 10-14 mm (Kliegman *et al*., 2020). Also, several studies have advocated that the pyloric muscle thickness and length directly correlate with age and weight of the infant (Peters, 2014). This is why, patients younger than 3 weeks of age should be observed and re-assessed in 1–2 days when the lesion could be more clinically or radiologically apparent (Peters, 2014). Dehydration resulting from the protracted vomiting can also cause a low measurement of muscle thickness, which may increase after fluid resuscitation (Ranells, 2011). However, if US is not available or it’s not diagnostic, an upper gastrointestinal contrast examination is highly effective in

making the diagnosis of IHPS. Barium is commonly preferred compared with water-soluble contrast to avoid the chemical pneumonitis. This procedure should demonstrate an elongated pyloric channel (also called the "string sign"), a bulge of the pyloric muscle into the antrum ("shoulder sign"), and parallel bands of barium seen in the narrowed channel, producing a "double tract sign" (7, 10). The increasing reliance on imaging has resulted in diagnoses being made before alkalosis has developed, and in a shorter clinical course, less morbidity and a shorter postoperative hospital stay. (Ranells, 2011)

Differential diagnosis: Pylorospasm and gastroesophageal reflux could be difficult to differentiate from IHPS without any imaging evaluation. Other medical causes of nonbilious vomiting include gastroenteritis, increased intracranial pressure, and metabolic disorders. (Coran, 2012) An endocrine disorder, adrenal insufficiency from the adrenogenital syndrome can simulate pyloric stenosis, but the lack of a metabolic acidosis and elevated serum potassium and urinary sodium concentrations of adrenal insufficiency aid in differentiation (Kliegman *et al*., 2020). On the other hand, surgical causes of non-bilious emesis include antral webs, pyloric atresia, duplication cyst of the antropyloric region, and ectopic pancreatic tissue within the pyloric muscle, all far less common than IHPS. (Coran, 2012). For the differential diagnosis, it is important to differentiate if the vomit is bilious or greenish. In that case, hypertrophic pyloric stenosis is unlikely. And, if it is bilious, the physicians must rule out malrotation, performing imaging with contrast studies (Peters *et al*., 2014; Hernanz-Schulman, 2003).

Treatment

Preoperative preparation: The preoperative treatment is focused toward correcting the fluid, acid–base and electrolyte disturbances. Therefore, correction of the alkalosis is vital to prevent postoperative apnea, which may be related with anesthesia (Kliegman *et al*., 2020). Benson and Alpern defined three levels of severity; mainly on the basis of the serum carbon dioxide (slight: <25 mEq/L; moderate: 26 to 35 mEq/L; and severe: >35 mEq/L). In addition to the elevated bicarbonate, hypochloremia, hypokalemia, dehydration and possibly malnutrition may be present. (Coran, 2012). In addition, oral feedings should be discontinued. A nasogastric tube should not be placed routinely because it removes extra fluid and hydrochloric acid from the stomach, which perpetuates the electrolyte and acid-base imbalance. (Coran, 2012) Vomiting usually stops when the stomach is empty, and only an occasional infant requires nasogastric suction (Kliegman *et al*., 2020). The resuscitation with fluids should be based on the level of dehydration and the level of electrolyte imbalance. Also, most infants with IHPS should be able to be resuscitated within a 24-h period. But, in the presence of severe metabolic and fluid irregularities, an aggressive resuscitation should be avoided because it may produce rapid fluid and electrolyte shifts, possibly leading to complications like seizures. (Peters, 2014). The ideal resuscitation regimen for fluid and electrolyte replacement is the administration of 5% dextrose in 0.45 normal saline containing 20 mEq/l of potassium chloride, given intravenous (IV) . In the scenario of severe hypokalemia, the concentration of potassium chloride can be increased to 30 mEq/l. In this case, the serum potassium level should be carefully monitored, mainly because the IV fluid rate possible will be above maintenance rates.

Other electrolyte disturbances like hyponatremia is rarely a problem. (Peters, 2014). Although, it is usual that normal saline is given as an initial bolus, there is little rationale for the use of normal saline because it enhances the hypokalemia by dilution and provides an excess amount of sodium. Therefore, resuscitation should be correlated with the level of dehydration. At the beginning, the rate for fluid resuscitation is 1.25- to 2-times the normal maintenance rate, until adequate fluid resuscitation and urine output are achieved. Also, the concentration of potassium chloride in the IV fluid should be based on the amount of hypokalemia and the rate of infusion, while keeping in mind that a potassium chloride concentration exceeding 30 mEq/l is rarely an indication. It is essential to monitor urine output and serum electrolytes because potassium should be administered only in the presence of diuresis. Normalizing the level of bicarbonate in serum (goal of decreasing level below 30 mEq/dl) frequently lags behind normalization of fluid volume and serum potassium and chloride (over 100 mEq/dl). Finally, when serum chloride and serum bicarbonate are corrected, it's safe to performed anesthesia and surgery. (Peters, 2014)

Operative procedure: The operative procedure of choice remains the Ramstedt pyloromyotomy. This procedure has stood the test of time because it is straightforward, curative, and associated with remarkably low morbidity. Regardless of abdominal access techniques, the myotomy created is identical. (Coran, 2012). The traditional Ramstedt procedure is performed through a short transverse skin incision. The underlying pyloric mass is cut longitudinally to the layer of the submucosa; the constriction is relieved and allows normal passage of stomach contents into the duodenum (7, 2). The fascial layers of the abdominal wall are closed with running absorbable suture. The skin is closed with subcuticular suture and Steri-Strips and covered with a dressing (Coran, 2012). This operation is effective at providing excellent exposure of the pylorus but results in an abdominal scar that grows with the patient and becomes quite significant with time. (Peters, 2014) Some other methods have been introduced, such as described by Tan and Bianchi, in which the pyloromyotomy is executed through a supraumbilical skin-fold incision. This technique reaches an excellent cosmetic outcome with an apparently minimal scare in the abdomen. In 1991, Alain *et al*. introduced the laparoscopic approach. Both surgical modalities have gained wide acceptance in the Western world. (Peters, 2014) The laparoscopic technique is equally successful. And in one study resulted in a shorter time to full feedings and discharge from the hospital as well as better parental satisfaction (Kliegman *et al*., 2020). Several comparative studies on laparoscopic pyloromyotomy (LP) and open pyloromyotomy (OP) reported a higher rate of complications with LP, leading to the conclusion that the operation is equal to OP only after the surgeon has sufficient experience (Jia *et al*., 2011).

Postoperative management: Vomiting in the postoperative stage occurs in half the patients and it seems to be secondary to edema and inflammation of the pylorus at the incision site (Kliegman *et al*., 2020). In the vast majority of the patients, feeding can be started within 4 hours after the surgery. Must be taken into consideration, the infants with hematemesis from gastritis; which may benefit by delaying feeding for an additional 6 to12 hours after the procedure. (Coran, 2012).

Nonoperative treatment: For patients who are not good surgical candidates, the conservative management with

nasoduodenal feedings or atropine is desirable (Kliegman *et al*., 2020). One pharmacological therapy that has been described when surgical expertise is not available is oral and intravenous atropine sulfate (pyloric muscle relaxant), which has 80% success rate described in some studies. Those studies also describe that oral atropine has less adverse effects than IV atropine for treating IHPS. In conservative protocols, atropine is given intravenously at a dose of 0.01 mg/kg 6 times a day 5 min before feeding. During atropine infusion, the heart rate has to be continuously monitored by electrocardiography. Also, oral feeding is started at a volume of 10 mL formula during 6 times a day. The volume is increased day by day until the infant tolerate 150mL/kg/day unless vomiting occurs more than twice a day (Kliegman *et al*., 2020). Later, when patients are able to tolerate the full volume of formula without vomiting more than twice a day, 0.02 mg/kg atropine is administered orally 6 times a day before feeding. As the conservative management takes longer and oral feedings may not be tolerated at the beginning, deteriorating of the nutrition status may occur and total parenteral nutrition could be required (Kliegman *et al*., 2020)

Complications: Complications after pyloromyotomy should be minimal if executed by experienced pediatric surgeons. Vomiting, which is frequent in the early postoperative period, is thought to be secondary to discoordination of gastric peristalsis, gastro-esophageal reflux or gastric atony and should not be considered a complication. However, frequent vomiting lasting beyond 3 to 4 days may suggest an incomplete myotomy or an unsuspected perforation. With that being the case, a postoperative contrast study may demonstrate a leak but is not helpful in evaluating the completeness of the myotomy. For that reason, it's important to remember that it takes several weeks for the radiographic appearance of the pylorus to improve. Therefore, persistent and frequent vomiting 1 week beyond the pyloromyotomy may require reexploration. (Coran, 2012). The complication rates vary between 4.6 and 12%. The incidence of major complications such as: perforation of the mucosa, incomplete pyloromyotomy, post- operative bleeding or wound-related problems (dehiscence of the fasciae or severe infection of the wound requiring operative drainage) is 1.2-3.4 %. And the incidence for minor complications is 7.3–23.8%. (Peters, 2014). The procedure of dilation with an endoscopic balloon has been effective in patients with persistent vomiting secondary to incomplete pyloromyotomy (Kliegman *et al*., 2020). The surgical treatment of pyloric stenosis is curative, withan operative mortality of 0–0.5% (Kliegman *et al*., 2020)

Conclusion

IHPS is a benign condition presenting with projectile, non-bilious emesis in the newborn infant, usually begin between three and five weeks of age and very rarely occur after 12 weeks of age and remains as one of the most frequently treated pediatric surgical conditions. Although several potentially causative factors have been explored, the pathogenesis of IHPS is not yet fully understood and remains unclear, leading to the conclusion that is a mix of environmental and genetic factors.In particular, IHPS has been associated with the administration of macrolide antibiotics to infants during the first few weeks of life and perhaps to their mothers during last gestation or lactation. This hypothesis is further supported by the success of atropine sulfate, which reduces muscular spasms of the pylorus, as a medical treatment for IHPS and a rescue treatment for incomplete pyloromyotomy. Increasing use of ultrasound has likely led to earlier diagnosis of infants with IHPS; classical biochemical abnormalities may be seen less frequently yet remain important to recognize, that's because the correction of acid-base disturbances are vital to prevent complications. Appropriate fluid resuscitation followed by pyloromyotomy has transformed IHPS from a lethal condition into a readily diagnosed and treated malady with nearly 100% survival rate. Conflict of interest: The authors declare that there is no conflict of interest.

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