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## **PODCAST TITLE: Infantile Hypertrophic Pyloric Stenosis (IHPS)**

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### **Introduction:**

Hello! Our names are Christine Miller and Sarah Mavin and we are fourth-year medical students at the Northern Ontario School of Medicine in Thunder Bay and Sudbury, Ontario, Canada. This podcast will discuss infantile hypertrophic pyloric stenosis (also known as IHPS), including a case presentation and general overview of risks, symptoms, consequences, and management. It has been created under the guidance of pediatrician Dr. Chantal Corbeil, Assistant Professor of Pediatrics within the Section of Child and Adolescent Health at the Northern Ontario School of Medicine.

### **Learning Objectives:**

The objectives of this podcast are to:

- Define infantile hypertrophic pyloric stenosis (IHPS)
- Review the pathophysiology associated with IHPS.
- Discuss etiology and risk factors of IHPS.
- Outline the management of IHPS.

### **Case Presentation:**

Let's begin with a clinical case scenario. During your Pediatric rotation, you are called for a consult in the Emergency Department. The patient is a 5-week-old male infant with a chief complaint of non-bilious vomiting for the past week. His mother reports the vomiting occurs after most feeds and can "shoot across the room". Sometimes, he will vomit the entire feed. After vomiting, he appears to still be hungry. The infant is exclusively formula fed since birth. In assessing the infant's hydration status, you discover he has been producing fewer wet diapers and stools than his usual. There is no blood in the stools. Until the recent episodes of vomiting, he has been gaining weight well and developing normally. He was born at term via spontaneous vaginal delivery and there were no complications at birth.

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### Differential Diagnosis for infantile non-bilious emesis:

- Hypertrophic pyloric stenosis
- Gastroesophageal reflux with or without a hiatal hernia
- Duodenal stenosis proximal to the ampulla of Vater
- Gastroenteritis
- Cow milk protein intolerance (blood-tinged stools)
- Adrenogenital syndrome causing adrenal insufficiency
- Inborn errors of metabolism causing recurrent emesis
- Liver disease (vomiting, poor weight gain, jaundice)
- Other congenital anomalies of the stomach, including:
  - Gastric, antral, or pyloric atresia
  - Pyloric or antral membrane/ web
  - Anomalies with rare incidence:
    - Microgastria
    - Gastric diverticulum
    - Gastric duplication
    - Gastric teratoma
    - Gastric volvulus
    - Congenital absence of the pylorus
- Overfeeding (however infants who are overfed do not show signs of hunger after feeding, are not dehydrated, and do not lose weight).

### Clinical Case:

Now that we have a differential, let's get back to the case.

Remembering that bilious emesis in an infant is a surgical emergency as it suggests Intestinal obstruction (malrotation, volvulus, Hirschsprung disease, intussusception) distal to the papilla of Vater, we confirm with his mother that the emesis is NOT coloured green or dark yellow.

You then begin your physical exam. Length is 59 cm (50th percentile), weight is 4 kg (25th percentile; previously 50th) and head circumference is 37 cm (50th). He appears to be well-nourished and is not in any acute distress. He is afebrile and his other vital signs are normal. No jaundice is noted. HEENT exam is normal. Auscultation of the heart demonstrates tachycardia in a regular rhythm. Lungs are clear. His abdomen is slightly distended with active bowel sounds. There is no hepatosplenomegaly or signs of peritonitis. You are able to palpate an olive mass within his right upper quadrant. Femoral pulses are strong bilaterally. Normal external male genitalia. Extremities are normal. Neurologic examination is grossly intact.

We suspect that our patient has IHPS but we are not ready to complete this case without working through our differential diagnosis and considering what we know about

IHPS. As we delve into the topic of IHPS, keep in mind the possible causes and risk factors for the development of this condition in the infant discussed above.

### **Literature Review:**

We will begin this review with a discussion of the definition and pathophysiology of pyloric stenosis, before moving onto risk factors, etiology, diagnosis and treatment strategies.

Hypertrophic pyloric stenosis is the most common cause of gastric outlet obstruction in infancy from corrected gestational ages 2 weeks to 12 weeks and is rare after 12 weeks of age. Infants are generally well prior to the gradual onset of non-bloody, nonbilious post-feed vomiting. The time course of symptomatology represents pathophysiology. It is rare for occurrences earlier than 2 weeks of age in a term infant. The pyloric musculature continuously hypertrophies over days to weeks until it causes gastric outlet obstruction, thus leading to the gradual maturation of symptoms. Projectile vomiting in the middle of a feed represents maximum obstruction.

Research suggests that impairment of neuronal nitric oxide synthase synthesis may be a cause of IHPS. This synthase causes relaxation of smooth muscle of the myenteric plexus, and in its absence, it is suggested that hypertrophy occurs. Gastric hyperacidity has also been implicated as a causal mechanism. Although the exact etiology of IHPS is unknown, there are some known risk factors and associations. Male gender and age between 2 to 12 weeks are the most significant associations. Molecular and twin studies have demonstrated an association between maternal history of pyloric stenosis and 7-20% incidence in her children. It is also more prevalent in certain genetic syndromes including:

- Cornelia de Lange syndrome
- Smith-Lemli-Opitz syndrome
- Apert syndrome
- Down syndrome
- Trisomy 18 syndrome

Exposure to macrolides, specifically erythromycin and azithromycin, in utero and in infancy are additional risk factors for IHPS. There is an association of increased incidence in infants born via cesarean section, in formula-fed infants compared to breast-fed infants, as well as a decreasing incidence with increasing birth order.

Thus, in summary, the highest risk non-syndromic infant would be a first-born male, aged 2 to 5 weeks at the onset of symptoms, exclusively formula fed from birth, with a parental family history of pyloric stenosis, and who was treated with macrolide in the neonatal period.

### **Case Presentation: Potential Causes and Diagnosis:**

Back to our case! The relevant risk factors that placed our male infant at a higher risk for IHPS, include his age, male gender and being formula fed. We are now convinced that our patient has IHPS, but how do we make sure our diagnosis is correct?

What do you think we should do to complete our diagnosis of pyloric stenosis?

- A. Complete an observed feeding trial to assess for vomiting episodes
- B. Ultrasound of the pylorus
- C. Upper gastrointestinal study
- D. Basic metabolic panel with electrolyte assessment and bilirubin
- E. No further investigations - clinical diagnosis is sufficient

If you selected any of the answers, you could be right! But this depends on the patient. For our case, the infant had a palpable pyloric olive, thus no further imaging is required for definitive diagnosis in the setting of his clinical presentation, however it may be helpful for the surgical team to have ultrasound imaging to evaluate the pylorus. Peristaltic waves seen across the abdomen prior to emesis is also suggestive of IHPS. In patients where there is no palpable mass, approximately half, ultrasound of the pylorus is indicated and a muscle thickness greater than 3-4 mm, or length greater than 15mm, in the presence of gastric outlet obstruction is diagnostic. If ultrasound is not available, an upper GI study may also be used. An observed feeding study would be indicated if you were unable to gather a clear feeding history from the infant's caregiver, or if imaging was inconclusive. And finally, any infant showing signs of dehydration should be tested with a basic metabolic panel with electrolyte assessment for hyponatremia, hypokalemic metabolic alkalosis. A serum bilirubin is indicated for an infant with jaundice.

Working through our differential diagnosis, he is less likely to have upper gastrointestinal tract atresia or webbing due to the rare incidence of these anomalies to the presence of a palpable mass in the region of the pylorus. Although radiographic studies could distinguish between GERD and IHPS, the feature of projectile vomiting and sustained hunger after vomiting makes IHPS more probable. His presentation is not consistent with metabolic acidosis or hyperkalemia, making adrenal insufficiency and inborn errors of metabolism unlikely. Milk protein allergy typically presents with blood-positive stool. Finally, gastroenteritis is unlikely without an infectious source, significant diarrhea, or other infectious features.

### **Treatment:**

All infants with hypertrophic pyloric stenosis are admitted to hospital. The three main goals of treatment are:

1. Screen for and correct any metabolic and electrolyte abnormalities
2. Assess hydration and correct fluid deficit
3. Pyloromyotomy

Assessment of hydration status is imperative in IHPS. Although hypovolemic shock is not common in IHPS, severe dehydration, altered mental status, impaired end-organ perfusion, and/or decreased blood pressure are all clinical signs that rapid fluid resuscitation is needed.

The name of the game, of course, is surgery (laparoscopic or open pyloromyotomy). Surgery is curative and may take place immediately if the infant is well hydrated. Fluid resuscitation and electrolyte correction are required before surgical intervention can be initiated. Most surgeons will request that a nasogastric tube be inserted at the time of diagnosis prior to surgical intervention. Infants can begin oral feeds within a few hours of surgery and the prognosis is excellent. Some regurgitation with feeds is common. IHPS alone is relatively benign, however delays in diagnosis can lead to failure to thrive, hypochloremic-hypokalemic metabolic alkalosis, and uncompensated hypovolemic shock. If vomiting persists >5 days post-surgery, further investigations should be completed such as an upper GI series.

### **Case Presentation: Completion**

Upon discovery of the infant's IHPS, you discuss with your attending that the next steps are to correct his fluid deficit and to consult the pediatric surgery team. Diagnosis of IHPS is confirmed via abdominal ultrasound which shows thickening of the pyloric muscle, along with elongation of the pyloric canal. He is admitted to the hospital and laparoscopic pyloromyotomy is planned for the next day. Your attending particularly mentions to you the importance of keeping an eye on the infant's hydration status overnight and the possibility of adjusting his IV fluids.

### **Learning Points:**

Let's review some main points of this podcast!

- IHPS is defined as the gradual hypertrophy of the pyloric muscle, which progressively obstructs the gastric outlet. This leads to non-bilious projectile vomiting, typically seen after feeds.
- History and physical exam are positive for age between 2 to 12 weeks, non-bilious vomiting with increased hunger post-emesis.
- Risk factors for IHPS include age between 2 to 5 weeks at the onset of symptoms, male gender, formula feeding, in utero and neonatal exposure to macrolides, first born, positive parental history, and C section delivery.
- Treatment: pyloromyotomy is the only definitive treatment for pyloric stenosis. Before surgical intervention, metabolic, electrolyte, and fluid abnormalities must be corrected.

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