GUIDELINE

Pyloric Stenosis: Infantile Hypertrophic

Scope (Staff):	Nursing and Medical Staff
Scope (Area):	NICU KEMH, NICU PCH, NETS WA

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Aim

To provide standardized management protocol for neonates with Infantile Hypertrophic Pyloric Stenosis (IHPS).

Risk

Neonates with IHPS may have severe acid-base abnormalities. Failure to prevent and/or manage these abnormalities may increase morbidities in neonates.

Background

IHPS is a disorder presenting in infants due to hypertrophy of the pylorus leading to partial or complete gastric outlet obstruction. The incidence is 2 to 4 per 1000 live births.

Etiology

- Unclear, considered multifactorial, family history (48% siblings have h/o IHPS)
- First born children (RR:1.23; 95% CI: 1.07-1.4)
- Male gender (M:F 4:1; RR:2.71; 95% CI:1.93-3.78), higher in preterm females
- Neonatal hypergastrinemia and increased gastric acidity
- Exposure to Erythromycin and Azithromycin in the first 6 weeks of life
- Formula fed> breast fed
- Maternal smoking (RR:1.75; 95% CI: 1.54-2), young maternal age (<20 years of age),
- Co-morbid with congenital heart disease and use of prostaglandin E1.

Presentation

Classically infants present between 3 to 6 weeks of age. Occasionally they may present in the first 2 weeks or later than 6 weeks of age, especially in premature babies.

Recurrent vomiting soon after feeds. Typically, the vomiting is described as projectile, non-bilious and happens soon after feeds. The baby is generally very hungry and wants to feed despite recurrent vomiting. Some infants, rarely, can present with bilious vomiting. There may be weight loss and signs of dehydration on clinical examination. It is important to elicit history of wet nappies. Abdominal palpation may reveal the presence of an “olive mass” in the right upper quadrant but is less commonly seen now a days because of early diagnosis. A recent systematic review reported that palpation has limited sensitivity in diagnosing IHPS.

Blood gas findings

- Hypochloraemic metabolic alkalosis with hypokalaemia, not typical in preterm
- Measure urea and creatinine levels on admission. It will help when deciding commencement of potassium infusion.

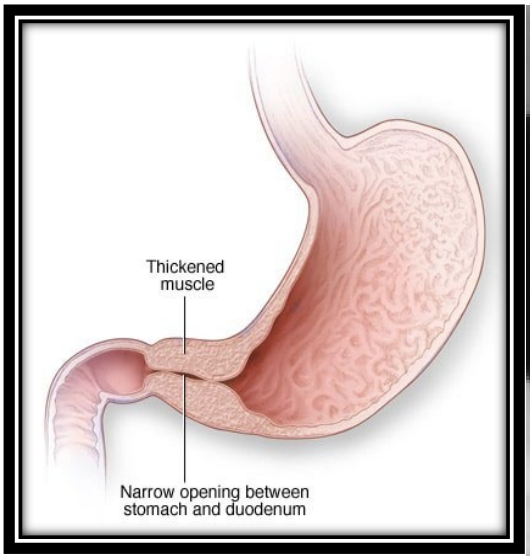
Special consideration for prematurity

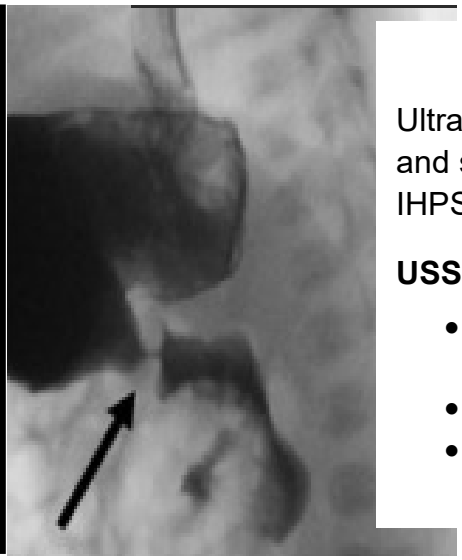
Premature babies with IHPS can present well before the infant even becomes a “term baby”. The characteristic projectile vomiting, VGP and metabolic alkalosis, and palpable pyloric olive (despite thin abdominal wall) are usually absent in preterm infants with IHPS. Dehydration is more common in preterm infants. Because standard ultrasound criteria for the measurement of pyloric muscle size in children with HPS may not be met in preterm infants, and also these criteria may not be valid for children with congenital HPS, an upper gastrointestinal contrast study may be needed to confirm or refute the presence of IHPS in a preterm infants.

Prolonged medical treatment associated with delayed diagnosis and malnutrition increase the postoperative complications and recovery time which is due to physiological immaturity of the organs and LBW of a preterm rather than IHPS. Early surgical intervention can decrease the morbidity and mortality. IHPS should always be considered in the differential diagnoses in a preterm with feed intolerance, GORD, or recurrent vomiting.

Diagnosis

Ultrasonography is the diagnostic modality of choice, and it shows hypertrophied and elongated pylorus. (Figure 1a). Upper GI contrast (Fig 1b: string-sign) may be performed if ultrasonography is inconclusive or if associated gut anomalies are clinically suspected.





Ultrasound has 95% sensitivity and specificity in diagnosis of IHPS

USS criteria for diagnosis

- Pyloric muscle Thickness >3 mm
- Pylorus Length > 15 mm
- Pylorus Diameter >10 to 14 mm

Fig 1a. Showing Thickening of the pylorus muscle and hypertrophy causing Gastric outlet obstruction.
Fig 1b. Narrowed pyloric canal on upper GI contrast (String-sign)

Immediate management

- Admit to hospital, NICU/Paediatrics ward based on age, gestation and weight criteria for admission.
- Keep nil by mouth.
- Insert size 8F nasogastric tube and allow it to drain freely.
- Arrange an ultrasound of the abdomen: diagnostic modality of choice.
- If USG is inconclusive and clinical suspicion persists, upper GI contrast could be considered in consultation with surgeons (especially in preterm infants).
- Insert intravenous cannula. Send blood investigations - venous blood gas (VBG), UEC, FBC, Cross match.
- Measure urea and creatinine levels and monitor urine output. It will help when deciding commencement of potassium infusion.
- Start intravenous antibiotics if considering differential diagnosis such as sepsis, UTI, volvulus, NEC especially in preterm infant, gastroesophageal reflux disease etc.
- Meta analyses demonstrate no benefit of prophylactic antibiotics before pyloromyotomy in preventing surgical site infections in children with IHPS.
- Assess the degree of dehydration and fluid management:
    - Management of dehydration and metabolic alkalosis with fluid resuscitation (Refer to Figure 2)
    - The fluid management is generally guided by the severity of dehydration and the blood gases and electrolyte abnormality (Figure 2)

- Chloride is the most sensitive and specific indicator of the need for additional saline boluses preoperatively.
- Nasogastric fluid losses should be replaced mL for mL every 4 hours with 0.9% Sodium Chloride.
- Correct hypokalaemia if present (Refer to Figure 2). Ensure that urine output is adequate (more than 1 ml/kg/hour) and creatinine levels are normal before commencing potassium infusion.
- Monitor blood glucose levels, if <3 mmol/L, increase the glucose infusion rate.

Surgical management

- The definitive management of IHPS is Laparoscopic or open Ramstedt's pyloromyotomy.
- It is preferable to correct alkalosis prior to surgery to decrease the risk of post-operative apnoea's. Preferable pre-operative blood gases targets:
  - Ph <7.45, Base excess <3, Bicarbonate < 26 mmol/L, Sodium >132 mmol/L, Chloride > 100 mmol/L and Potassium > 3.5 mmol/L, Blood glucose>3.5 mmol/L.

Note: This may not be achievable in all cases and need to be considered in discussion with surgeons.

Post-operative care

- Monitor the baby for apnoea for at least 24 hours post-surgery.
- Cease antibiotics post-operatively unless otherwise specified.
- Some infants may have post-op Gastroesophageal reflux or vomiting which should be managed on case-to-case basis.

Post-operative feeding guide

Feeds can generally be started after 4 hours unless specified in the operation notes. Adjusting the IV fluid rate as oral feeds are tolerated.

Feeds are usually tolerated by 24 hours post operatively.

This is a guide only and will need to be adjusted if vomiting persists.

- Feeding:
  - Feeds can generally be started after 4hours unless specified in the operation notes and post-operative instructions.
  - Adjusting the IV fluid rate as oral feeds are tolerated.
  - Start 2 or 3 hourly feed as per the gestation and weight of the baby.
  - Generally, infants will tolerate starting feed at 5 ml 3 hourly feed

- Grade up feeds 5ml every 3 hours.
- Breast feeding can be commenced 3 hourly for 5 to 10 minutes in addition as per parental preferences.
- Transition to full breast feed gradually with the grading up of feeds.
- Full feeds can be graded as tolerated over 24 hours unless other concerns exist, requires clinician discretion. Majority will tolerate full feeds.

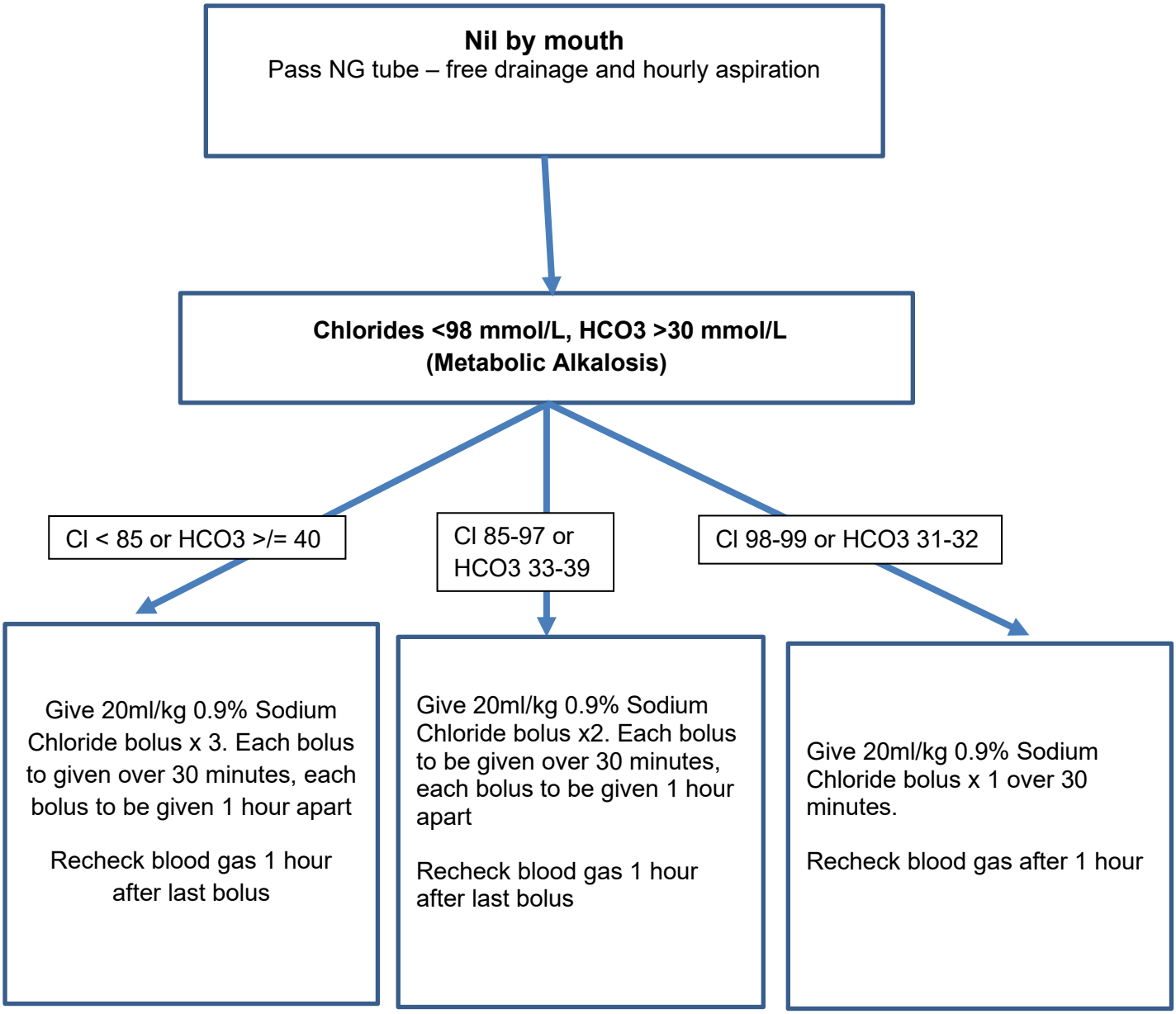


Fig 2: Management of Initial fluid resuscitation based on CBG results

- All patients should be put on 1.5 times maintenance fluid
- If hypokalemia (K^+ <3.0 mmol/L), run a sideline of Potassium Chloride infusion (as per Hypokalemia guideline) ensuring the patient has adequate urine output (>1 ml/kg/hour)
- All patients should be placed on cardiorespiratory monitoring
- TPN should be avoided in the initial rehydration and stabilization phase

Related CAHS internal policies, procedures and guidelines

Neonatology Clinical Guidelines

- Post-Operative Care
- Pre-Operative Care

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10. Johns Hopkins All Children's Hospital Pyloric Stenosis Clinical Pathway

Useful resources

Laparoscopic Pyloromyotomy (child)

This document can be made available in alternative formats on request.

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