JOHNS HOPKINS ALL CHILDREN'S HOSPITAL

Pyloric Stenosis Clinical Pathway

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Updated: March 24, 2026
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Rationale:

This clinical pathway was developed by a consensus group of Johns Hopkins All Children's (JHACH) physicians, advanced practice providers, nurses, and pharmacists to standardize the management of children hospitalized for pyloric stenosis. It addresses the following clinical questions or problems:

1. When to evaluate for pyloric stenosis
2. When to consider admission for further evaluation
3. When to consult Surgery
4. When to consider resuscitation and other disease processes

Background:

Pyloric stenosis is one of the more common causes of persistent vomiting in the first weeks of life and one of the most common surgical conditions of the newborn. The usual age at presentation ranges from 2–8 weeks, with occasional late presentations up to 12 weeks of age. The cause remains unknown, though it is thought to have a genetic predisposition with some environmental influence. It occurs more commonly in males than in females. Classically, the presentation is one of projectile, non-bilious vomiting. If bile is present in the emesis, it is NOT due to pyloric stenosis (Dalton, 2016; Jobson, 2016).

Definition:
Hypertrophic pyloric stenosis (HPS) occurs from hypertrophy of the circular and longitudinal muscularis of the pylorus and the distal antrum of the stomach with progressive narrowing of the pyloric canal. It is the most common surgical cause of vomiting in infants (Jobson, 2016).

Age range:
HPS typically presents in the first 2–8 weeks of life, with occasional late presentation up to 12 weeks of age (Dalton, 2016; Jobson, 2016).

Symptoms:
The classic symptom is progressively worsening projectile vomiting after feeding. The vomiting is usually non-bilious. A mass in the upper abdomen/gastric wave may or may not be palpable. Ongoing emesis left untreated leads to dehydration, including hypochloremia and hypokalemic metabolic alkalosis (Dalton, 2016; Jobson, 2016).

Epidemiology:
HPS is more common in males than in females, and it is more common in firstborn children. There is also a lower risk in African and Asian populations (Dalton, 2016; Jobson, 2016; Markel, 2017).

Differential diagnosis:
Includes overfeeding, gastroesophageal reflux, malrotation, duodenal stenosis, and intracranial lesions.

Diagnosis:

If pyloric stenosis is suspected:

1. Place nothing by mouth (NPO) order
2. Obtain basic metabolic panel (BMP)
3. Place a peripheral intravenous line (PIV)
4. Administer a normal saline (NS) bolus of 20 milliliters per kilogram (mL/kg)
5. Obtain pyloric ultrasound (US)
    a. Positive measurements include a pyloric muscle thickness greater than 3 millimeters (mm) and a pyloric canal length of 15 mm or greater, and no fluid moving through the pyloric canal
    b. If pyloric US is equivocal, recommend observation and repeat the pyloric US in 48 hours (hr)
    c. Consider an upper gastrointestinal (UGI) contrast study if pyloric US remains equivocal

If pyloric stenosis is confirmed:

1. Consult Pediatric Surgery if inpatient or transfer to JHACH for Pediatric Surgery consultation
2. Assess serum electrolyte results
3. Consider nasogastric (NG) tube placement; to be placed at the Surgical Team's discretion
4. Place the infant on an apnea monitor and admit to inpatient

Clinical Management:
Pre-operative resuscitation management:

Table 1: Laboratory Parameters and Management

Laboratory Parameters	Management
Cl < 85 mmol/L or HCO_3 ≥ 40 mmol/L	• Admit to PICU (Surgery to admit, co-manage with PICU) • Administer 20 mL/kg NS bolus x3, each bolus over 30 minutes, each bolus to be given 1 hour apart • Recheck BMP 1 hour after last bolus
Cl 85 – 97 mmol/L or HCO_3 33 – 39 mmol/L	• Admit to Pediatric Floor (Surgery to manage), unless infant has tachycardia, hypotension, or deemed necessary by the Surgical Team to admit to PICU. Surgery will co-manage with PICU. • Administer 20 mL/kg NS bolus x2, each bolus over 30 minutes, each bolus to be given 1 hour apart • Recheck BMP 1 hour after last bolus
Cl 98 – 99 mmol/L or HCO_3 31 – 32 mmol/L	• Admit to Pediatric Floor (Surgery to manage) • Administer 20 mL/kg NS bolus x1 • Recheck BMP 1 hour after bolus
Cl ≥ 100 mmol/L, HCO_3 ≤ 30 mmol/L, and K+ < 5 mmol/L	• Infant clear for OR

Abbreviations: Cl, chloride; HCO_3, bicarbonate; K+, potassium; mmol/L: millimoles per liter; OR, Operating Room; PICU, Pediatric Intensive Care Unit

- Multiple boluses should be given 1 hour apart (all boluses should be normal saline)
- All patients should have a BMP drawn 1 hour after the last ordered bolus
- All patients should receive 1.5 times maintenance IV fluids (mIVF) utilizing D5W 0.45 NS with 20 milliequivalents per L (mEq/L) KCl
    - K+ should be included in the mIVF if the patient has adequate urine output (UOP) ≥ 2 mL/kg/hr, or wet diapers in the preceding 6 hours
        - May use caregiver history (if no wet diapers in the preceding 6 hours, do not add in K+ to IVFs)
- Place all patients on an apnea monitor while inpatient
- Patients should proceed to pyloromyotomy once they meet institutional anesthesia requirements (HCO_3 ≤ 30 mmol/L, K < 5 mmol/L, AND Cl ≥ 100 mmol/L)

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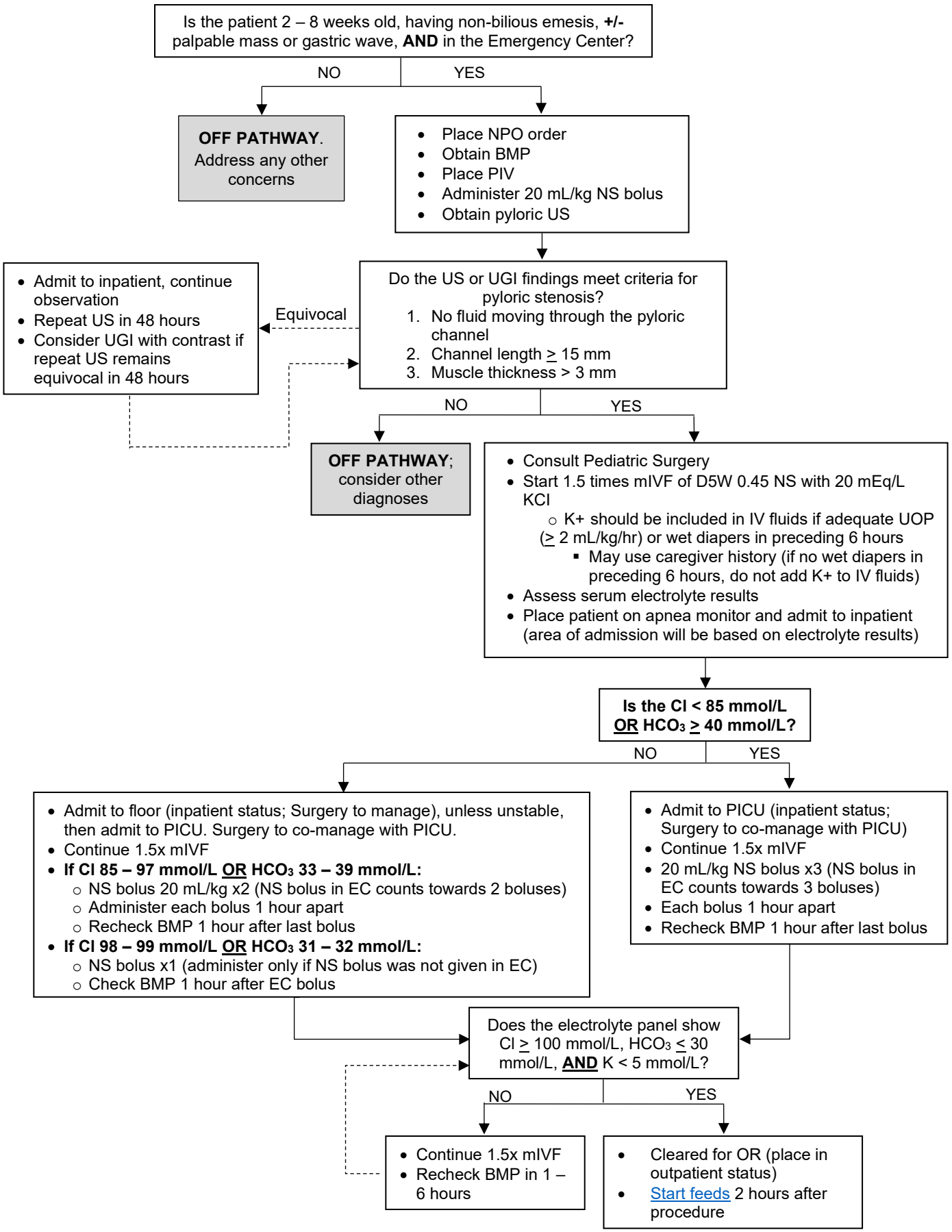

Admission and Rehydration:

Pyloric stenosis is a medical, not a surgical, emergency. The classic metabolic derangement is a hypokalemic, hypochloremic metabolic alkalosis (from gastric Cl losses). This will be evident by the results of the electrolyte panel (Jobson, 2016). The patient's clinical status determines rehydration strategies. Dehydrated patients should receive an NS bolus of 20 mL/kg over one hour, in addition to 1.5 mIVF with D5W 0.45 NS with 20 mEq/L KCl at 1.5 times the mIVF rate. Assess for adequate UOP ($\geq$ 2 mL/kg/hour) (Dalton, 2016). Surgery will not be performed until the electrolytes are corrected (K < 5 mmol/L, Cl $\geq$ 100 mmol/L, AND $HCO_3 \leq$ 30 mmol/L) (Jobson, 2016).

Inpatient Management:

- Once the patient is adequately hydrated and the electrolytes are corrected, surgical intervention is indicated
    - The surgical procedure is a pyloromyotomy, which can be performed open or laparoscopically
- Acetaminophen only for pain management

Post-operative feeding management:

- Feeding regimen per Attending discretion.
    - Caregivers may provide the formula of choice pre-operatively (typically breast milk or Enfamil®).
    - Emesis may occur with the first few feedings. Continue the planned feeding regimen and provide caregiver reassurance unless otherwise directed by the Pediatric Surgery Fellow or Attending.

1. NPO for 2 hours following the procedure.
2. Initiate breast milk or formula at 60 mL (2 oz) every 3 hours.
3. Place all patients on an apnea monitor.
4. Discharge criteria:
    - Tolerating 2 oz for two consecutive feedings without emesis, and
    - Meeting institutional post-operative monitoring requirements (Dalton, 2016).

Discharge:

- Anticipate discharge on post-operative day (POD) 1 or 2

Documentation Recommendations:

- Providers should be sure to capture any electrolyte or acid-base abnormalities in their assessment and plan (e.g., hypokalemia, metabolic alkalosis)
- Document if dehydration was present on admission, and if so, the degree/severity

- Document if shock was present on admission

Patient Class Recommendations:

- Patients with stable volume status and electrolytes after evaluation and fluid resuscitation in the Emergency Center (EC) who will proceed to the OR without delay for clinical indications should be placed in observation status
- Patients admitted to the PICU and/or patients requiring ongoing fluid resuscitation and electrolyte monitoring beyond what is performed in the EC should be admitted to inpatient status

References:

Dalton, B. G., Gonzalez, K. W., Boda, S. R., Thomas, P. G., Sherman, A. K., & St Peter, S. D. (2016). Optimizing fluid resuscitation in hypertrophic pyloric stenosis. *Journal of Pediatric Surgery, 51*(8), 1279–1282.

Jobson, M., Hall, N., & Bchir, M. B. (2016). Contemporary management of pyloric stenosis. *Seminars in Pediatric Surgery, 25*(4), 219–224.

Markel, T. A., Scott, M. R., Stokes, S. M., & Ladd, A. P. (2017). A randomized trial to assess advancement of enteral feedings following surgery for hypertrophic pyloric stenosis. *Journal of Pediatric Surgery, 52*(4), 534–539.

Outcome Measures:

1. Time from first BMP to time electrolytes are corrected for OR
2. Post-operative length of stay

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Pyloric Stenosis Clinical Pathway
Johns Hopkins All Children's Hospital

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Date Approved by Clinical Pathway Development Committee (CPDC): February 3, 2026

Date Approved by JHACH Clinical Practice Council (CPC): February 18, 2026

Date Content Last Revised: March 24, 2026

Date Available on Webpage: March 24, 2026

Historical Submission:
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Date Approved by JHACH Clinical Practice Council: March 16, 2021

Content Last Revised: June 1, 2021

Date Available on Webpage: June 1, 2021

Disclaimer:

Clinical Pathways are intended to assist physicians, physician assistants, nurse practitioners, and other healthcare providers in clinical decision-making by describing a range of generally accepted approaches for the diagnosis, management, or prevention of specific diseases or conditions. The physician must make the ultimate judgment regarding the care of a particular patient, taking into account the patient's individual circumstances.

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