Pyloric Stenosis

Congenital hypertrophy of the pyloric sphincter leading to reduced size of gastric outlet, impeding emptying which can cause abnormal nutrition, repeated vomiting & dehydration; this is a medical emergency, NOT a surgical emergency, so there is time to optimize these patients prior to providing anesthesia for surgery.

ANESTHETIC CONSIDERATIONS:

- Medical, not surgical, emergency due to severe fluid deficits
- Full stomach → aspiration risk
  - Consider emptying stomach with red rubber catheter pre induction
  - Awake intubation vs rapid sequence induction
- Metabolic derangement
  - Hypochloremic metabolic alkalosis
  - Hyponatremia and hypokalemia/hyperkalemia if severe dehydration
- Considerations of neonate / infant
- Risk of postoperative apnea (persistent metabolic or CSF alkalosis)

ANESTHETIC GOALS:

- Preoperative optimization: Correct fluid and acid-base deficits ($Na^+ > 130$; $K^+ > 3.0$; $Cl^- > 100$; $HCO_3 < 27$; U/O 1 cc/kg/hr; normal HR / BP / RR)
- Protect against / prevent aspiration
- Ensure adequate postoperative monitoring (overnight apnea monitoring)

HISTORY

- Prenatal history and family history
- Birth history, especially any difficulties or resuscitation
- Onset of illness; frequency, character and amount of vomiting; last feeding; diarrhea; urine output; activity of the newborn (active or lethargic), birth weight
- The most important aspect on the history and physical exam is to **determine the degree of dehydration** as this is a medical emergency
- There are many different approaches to assess volume status; in the neonate and infant, percentage of body weight is widely used (ideal if most recent weight is known to compare to baseline)
  - If baseline weight not known, degree of deficit can be estimated with table below
  - Note this is only applicable to **non-hemorrhagic volume depletion**

Finding	Mild	Moderate	Severe
Percent body weight	**< 5%**	**5-10%**	**>10%**
Mucous membranes	Slightly Dry	Dry	Dry
Capillary Refill	N	↓	↓↓
Tears	Normal	↓	Absent
CNS	Normal	Irritable	Lethargy
Pulse	N	↑	↑↑ and Weak
SBP	N	N or Slightly ↓	↓↓
Respiration	N	Slightly ↑	↑
Anterior Fontanel	N	↓	↓↓
Skin	N	Cool	Acrocyanosis
U/O	Mildly ↓	↓↓	Anuria
Volume Required	**25-50 cc/kg**	**50-100 cc/kg**	**100-150 cc/kg**

- Example, a 6 week old, 5 kg child presents with non-bilious vomiting, is irritable with moderately delayed capillary refill → moderate volume depletion
  - 5-10% of body weight → 250-500 mL
  - Thus, will require volume resuscitation of 50-100 mL/kg
- Many different opinions on how to resuscitate patient, but generally:
  - Use NS in 20 cc/kg boluses to replace deficit and titrate to above criteria and U/O of 1 cc/kg/hr
  - Maintenance of D5½NS + KCl 20-40 mEq/L at 1 to 1.5x maintenance requirement of 4 cc/kg/hr
- Recent retrospective review of patients presenting with pyloric stenosis (Miozzari 2001):
  - 62% euvolemic
  - 27% mild dehydration
  - 7% moderate dehydration
  - 4% severe dehydration
- Patients are presenting earlier with less severe volume deficits and electrolyte abnormalities
- Associated conditions
  - Unconjugated hyperbilirubinemia (2%) which tends to resolve after surgery
  - Midgut malrotation (up to 5%)
  - Congenital heart disease, congenital diaphragmatic hernia → rare

PHYSICAL

- **VITALS** – HR, BP, RR, SpO_2, temperature, current body weight (to assess weight loss)
- **HEENT** – mucous membranes, tears, anterior fontanel, eyes (normal or sunken)
- **CVS** – standard including capillary refill and skin turgor, overall volume status as above
- **RESP** – standard exam, cyanosis
- **GI** – pyloric "olive" palpable in upper abdomen
- **GU** – urine output

- **CNS** – activity (irritability, lethargy etc.)

INVESTIGATIONS

- **Labs**
  - CBC, electrolytes, BUN, Cr, glucose, ABG, bilirubin unconjugated, U/A
  - Classic laboratory derangement:
    - **Hypochloremic metabolic alkalosis**
    - Hyponatremia
    - Hypokalemia
      - Due to secondary hyperaldosteronism (to maintain ECF and Cl^-)
      - Although one review demonstrated 36% of patients had hyperkalemia on presentation (Swart 2003)
    - Initially, urine is alkalotic due to HCO_3^- and Na^+ loss to maintain electrical neutrality with gastric Cl^- loss
    - However, if severe volume depletion, can have metabolic acidosis
  - However, metabolic and electrolyte abnormalities are less common than in past due to earlier diagnosis
- **Imaging**
  - EKG – for marked hypokalemia
  - Ultrasound often used to confirm diagnosis although may still have upper GI barium study
    - Barium swallow shows elongated and narrowing of pyloric canal

OPTIMIZATION

- It may take 12 to 72 hours to stabilize fluid and electrolytes
- **Restoration of volume deficit** → most important!
- A large-bore IV should be placed and an infusion started immediately to correct the deficits and provide maintenance fluids
- Many different opinions on how to resuscitate patient, but generally:
  - Use NS in 20 cc/kg boluses to replace deficit and titrate to clinical criteria outlined above and U/O of 1 cc/kg/hr
  - Maintenance of D5½NS + KCl 20-40 mEq/L at 1 to 1.5x maintenance requirement of 4 cc/kg/hr
- **Restoration of metabolic and electrolyte abnormalities**
  - References difficult to find to delineate what levels should be targeted
  - Barash:
    - Na > 130 mEq/L
    - K > 3.0 mEq/L
    - Cl > 85 mEq/L
  - Other papers in literature are more conservative in their numbers and seem more reasonable:
    - Na > 130
    - K > 3.0
    - Cl > 100
    - HCO_3 < 27
- **Prevention of aspiration**
  - A nasogastric tube (14F catheter or red rubber catheter) should be inserted and suctioned with child in various positions to thoroughly empty the stomach → NOTE: this does not ensure empty stomach
- Pre-operative acetaminophen 20-40 mg/kg rectally

ANESTHETIC OPTIONS

- Generally, GA performed, however, multiple series with neuraxial anesthesia
  - SAB (Somri 2003) - 25 children
    - 0.8 mg/kg of isobaric bupivicaine 0.5%
    - Conversion to GA in 2/25
  - Caudal anesthesia (Moyao-Garcia 2002)
    - Commonly done in Mexico and case series published
    - Large doses of 1.6 mL/kg (4 mg/kg) of 0.25% bupivicaine

ANESTHETIC SETUP

- **Drugs**
  - Routine emergency drugs (atropine & SCh)
- **Equipment**
  - Routine CAS monitors + temperature probe
  - Neonatal setup: neonatal circuit, warm room, warming blanket and lights

MANAGEMENT OF ANESTHESIA

- **Induction**
  - Suctioning stomach preoperatively with 14F catheter (can use red rubber catheter) until no further gastric contents obtained
  - Intubation
    - Pre-treat with **atropine 20 mcg/kg**
    - Awake intubation vs. RSI
      - Benefits of awake intubation: preservation of protective reflexes, able to provide oxygen during intubation and ability to ventilate in unanticipated difficult airway
        - Cook-Sather (1998) compared awake intubation vs. RSI vs. modified RSI (allowed gentle PPV with cricoid)
        - Awake intubation required more attempts and had longer time to intubation vs. RSI / modified RSI
        - No advantage to modified RSI vs. RSI with episodes of hypoxemia
      - RSI

        - Preoxygenate and apply cricoid
        - Propofol 4-6 mg/kg
        - SCh 2 mg/kg (can consider rocuronium 1-1.5 mg/kg and / or remifentanil 2-4 mcg/kg)
    - **Succinylcholine**
        - Concern is unrecognized myopathy
        - However, for short procedure rocuronium in adequate doses may require post-operative ventilation
    - **Conflict**: patient with pyloric stenosis and difficult airway
- **Maintenance**
    - Could consider adjuvant caudal anesthesia but requires large dose to achieve level
    - Maintenance IV: D5½NS at 4-5 cc/kg/hr
    - Maintenance anesthesia: users preference (volatile or TIVA), avoid N_2O
    - Ventilation: avoid hyperventilation to avoid worsening respiratory alkalosis
    - Surgeon should infiltrate incision with bupivicaine at end of case
- **Emergence**
    - Extubation should be performed when patient is completely awake and protecting airway
    - Post-operative Apnea
        - Several cases of post-operative apnea in this population
            - Majority of cases were with halothane maintenance
        - However, recent study of infants undergoing pyloromyotomy had pre- and postoperative apnea studies
            - None of the patients had abnormal studies
            - Concluded that risk of post-operative apnea in term infants does not exist (however, they only tested 30 children)
        - May be prudent to watch these children post-operatively
        - May be worse if preoperative metabolic alkalosis (may need concomitant respiratory acidosis as stimulus to breath)

DISPOSITION & MONITORING

- Monitored bed for apneas for 12-24 hrs if metabolic alkalosis is only partially corrected or if infant is ex-premature (though, case reports of healthy infants requiring monitored bed also)
- Analgesia
    - Caudal
    - Tylenol
    - Local anesthetic infiltration by surgeon
    - Avoid narcotics in postoperative period

COMPLICATIONS

- Fluid, electrolyte & metabolic imbalance uncorrected
- Aspiration d/t full stomach
- Apnea postoperatively

PATHOPHYSIOLOGY

- Elongation and thickening of the pylorus leading to near-complete gastric outlet obstruction
- Occurrence 1:500 live births and more common in males (4-6:1) with ~30% occurring in first born males
- Presents in 2^{nd} to 6^{th} week of life with non-bilious vomiting resulting in loss of hydrogen ions, chloride, sodium, and potassium ions
- The effects of the metabolic alkalosis include:
    - ↑ pH results in shift of the oxygen dissociation curve to the left, thereby binding more O_2 to Hb and unloading less O_2 at the tissue level
        - This assumes even more importance in newborns because at 3 weeks they still have up to 70% fetal Hb with an already low P_{50} (i.e. 20 to 22 mmHg)
    - Respiratory compensation is accomplished by hypoventilation with resultant increased potential for atelectasis, as well as periods of apnea
    - Decrease in ionized calcium
    - Increased potential for seizures
- Differential diagnosis: achalasia of the esophagus, hiatus hernia, duodenal atresia, jejunal atresia, ileal atresia, pancreatic annulus, malrotation of the gut, intra-abdominal hernias, extra-abdominal hernias, and Meckel's diverticulum

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Tracheoesophageal Fistula/Esophageal Atresia & VACTERL

Tracheoesophageal fistula and esophageal atresia is a congenital malformation which may present in several ways including a variety of connections between the esophagus and trachea as well as varying degrees of esophageal atresia leading to feeding difficulties (volume depletion / electrolyte abnormalities) and recurrent aspirations (with or without the development of pneumonitis; challenges include rapidly securing the airway to prevent further aspiration by attempting to place an ETT distal to the fistula while avoiding PPV; this malformation is part of the VACTERL association which should be further investigated in these patients (especially cardiac and renal abnormalities).

ANESTHETIC CONSIDERATIONS:

- Considerations of the neonate / possibly prematurity
  - Transitional circulation, postoperative apnea, hypothermia, hypoglycemia, altered pharmacokinetics
  - Considerations of prematurity (IVH, BPD, ROP, NEC)
- Aspiration risk: acute and chronic +/- pneumonitis (sepsis, hypoxemia)
- Volume depletion with electrolyte abnormalities
- Difficult ETT placement and difficulty with ventilation (subglottic stenosis, stomach insufflation)
- Shared airway for bronchoscopy / esophagoscopy, and intraoperative compression of lung and great vessels
- Coexisting conditions:
  - VACTERL
  - Cardiac (in 25%): VSD, ASD, PDA, TOF – Antibiotic prophylaxis
  - Right aortic arch in 5%
- In patient with prior TEF repair now going for surgery - they may have **tracheomalacia**, **tracheal diverticulum** at site of fistula, **reflux**, and **esophageal stricture** at site of anastomosis

ANESTHETIC GOALS:

1. Preoperative optimization
   a. Evaluation for congenital heart disease
   b. Optimize pulmonary status
   c. Volume resuscitate and correct electrolyte abnormalities
2. Specific airway and ventilation concerns
   a. ETT placement / positioning – **rapidly establish airway** (prevent further aspiration) / tip just above carina
   b. **Ventilate without inflating stomach** (avoid PPV – minimize gastric insufflation & secure A/W distal to fistula)
   c. Risk of subglottic stenosis
3. Avoid perioperative increases in pulmonary vascular resistance
4. Avoid hypoglycemia, hypothermia
5. Early extubation if possible

HISTORY

- Usual perinatal / anesthetic history
- Establish post conceptual age
- Prenatal history, gestational age, post-partum age
- Polyhydramnios
- Concerns during pregnancy / labor / delivery
- Events leading up to diagnosis & details of fistula anatomy (imaging)
- Identify associated anomalies i.e. VACTERL
  - CVS: CV decompensation, cyanosis, CHF, murmur
  - Resp: pneumonia, subglottic stenosis, aspiration history (choking, cough)
  - GI: gastric distension, anal atresia, bowel obstruction
- Discuss investigations regarding type of fistula with surgeon
- Investigations performed (CXR, ECHO, renal U/S)

PHYSICAL

- **GENERAL** - Examine for stigmata of VACTERL
- **VITALS** - Including pre- / post-ductal SpO_2
- **HEENT** – Airway exam, presence of nasoesophageal tube or gastrostomy tube
- **CVS** - Murmur, precordial thrill,, cyanosis, enlarged liver, HTN, volume status (fontanels, mucous membranes, peripheral pulses, capillary refill)
- **RESP** - Pattern & evidence of distress / increased WOB & crackles suggesting aspiration

INVESTIGATIONS

- **Labs**
  - CBC, ABG
  - Group / screen
  - Electrolytes / BUN / Cr / Glucose
- **Imaging**
  - CXR
  - ECHO preoperatively for *all* TEF – also VSD / PDA / TOF

OPTIMIZATION

- Anticholinergic (antisialogogue / bradycardia defense)
- Aspiration of nasoesophageal tube
- Ensure adequate hydration and electrolyte balance (may be on TPN)
  - Continuation of dextrose IVF / TPN
- **Urgent** but **NOT a surgical emergency**
- Rigid bronchoscopy to define anatomy (>3 mm associated with increased risk of adverse events)
- If the neonate is in good condition, primary repair can be performed at 24–48hrs, otherwise delay surgery to optimize
- **Head up** and face down or lateral position to avoid aspiration

- Stop feeds, and **continuous suctioning** to upper esophageal pouch
- Ventilatory support, pulmonary toilet, antibiotics as indicated for respiratory distress
    - Note: positive pressure ventilation may lead to a distended stomach, affecting ventilation
    - Gastrostomy under local anesthetic may be required due to gastric distention
    - If **gastrostomy** in place, **open to air** before induction
    - A Fogarty catheter can be passed via G-tube to occlude esophagus / fistula to aid ventilation
- Consultations as appropriate for other congenital anomalies
- **Procedures**
    - Primary complete repair is preferred (ligation of fistula and anastomosis of esophagus)
        - Depending on anatomy and surgeon preference, procedure may either be done via right thoracotomy (?potentially left thoracotomy if right-sided Ao arch), cervical approach or endoscopically
    - A staged repair (gastrostomy and ligation of fistula with delayed esophageal anastomosis) may be done if the ends of the esophagus are too short to anastomose primarily

ANESTHETIC OPTIONS

- Local to insert gastrostomy tube
- GA for primary repair

ANESTHETIC SETUP

- **Drugs**
    - Emergency drugs (succ, atropine)
- **Equipment**
    - CAS monitors and temperature, precordial stethoscope on left chest wall (indicates R mainstem intubation)
    - Neonatal setup (warm OR, warming blanket and lights)
    - +/- art line

MANAGEMENT OF ANESTHESIA

- **Induction**
    - Preinduction intravenous +/- volume resuscitation
    - Consider gastrostomy tube pre or post induction; if in place, open to air
    - Airway
        - **Traditionally intubated awake with spontaneous ventilation** and ETT to right mainstem, withdraw until bilateral breath sounds heard
            - Miller suggests mild sedation with topicalization of airway
        - **Inhalational induction and maintenance of SV** – potential conflict with CHD
        - RSI with relaxant; position ETT fiberoptically or place ETT just above carina (hopefully below fistula) by **intentional endobronchial intubation and gradual withdrawal** while auscultating the left side
        - Note that it is possible to intubate the fistula
        - Confirm placement (fiberoptically) with all techniques
        - May use PPV if low inflation pressures (can use Fogarty to occlude fistula)
- **Maintenance**
    - Maintenance: air / oxygen / volatile / narcotic
        - If unstable, opioid anesthetic (e.g. fentanyl 10-15 mcg/kg)
    - Left lateral decubitus positioning
    - Consider **epidural** (by **caudal** or direct)
    - **Avoid N_2O**
    - One lung ventilation may be poorly tolerated, and may require the surgeon to interrupt the procedure intermittently
    - 100% Oxygen may be justified despite risks of retinopathy
    - Monitor ventilation and BP carefully as pressure from retractors can occlude trachea / bronchi / major vessels / and impair CO dramatically
    - Manipulation and ligation of fistula leads to bleeding and secretions (may require repeated suctioning)
- **Emergence**
    - Re-expand lung to minimize post-operative atelectasis
    - Avoid neck extension and passing deep suction catheters in the case of a new esophageal anastomosis
    - **Likely require post-op ventilation**
    - Vigorous infants may be extubated immediately postoperatively (preferred for maintenance of repair)

DISPOSITION & MONITORING

- HDU or ICU

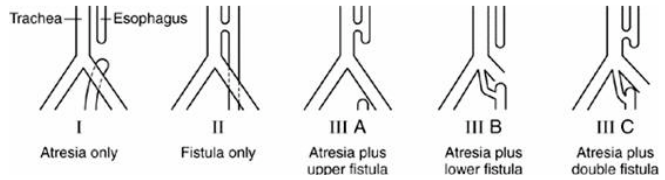
COMPLICATIONS

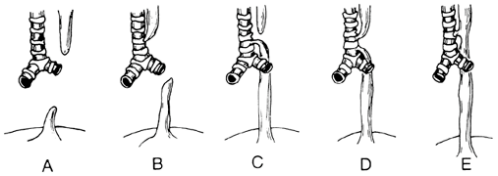
- **Difficulty ventilating**
    - This may be due to kinking of the trachea by the surgeon, which is relatively common especially when tracheomalacia is present
    - Severe gastric insufflation +/- pneumoperitoneum
        - Some may consider immediate needle decompressive gastrostomy in this situation (to left upper quadrant of abdomen)
    - Tube migration
- **Intra-operative hypoxemia**
    - Massive gastric distention
    - Surgical compression of lung
    - Endobronchial intubation
    - **Intubation of the fistula**
    - Pneumothorax
    - Underlying CHD (R to L shunt)
    - Aspiration
- Post-operative

- **Tracheomalacia** (expiratory wheeze, apnea – treat with CPAP)
- Recurrent aspiration (may need GERD surgery)
- Bronchospastic reactions
- Anastomotic leak
- RLN injury

PATHOPHYSIOLOGY

- Incidence of 1 in 3000 births
- Most (2/3rd) cases are now detected prenatally by U/S
- 50% of infants with EA have associated anomalies
- Most common forms (see figures)
  - Many different classification schemes (incidence below from Coexisting, 5th pg 595)
  - **86% of cases are esophageal atresia and fistula between the trachea and distal segment of the esophagus** (Type IIIB or **Type C**)
  - Second most common (14%) is proximal fistula (Type IIIA or Type B)
  - Third (8%) is pure esophageal atresia (Type I or ~~Type E)~~ Type A
- Less common forms
  - Proximal esophageal fistula to trachea with disconnected distal esophagus (Type IIIA or Type B)
  - Both proximal and distal esophageal components have fistula to trachea (Type IIIC or Type D)





- Associated conditions include:
  - Prematurity
  - Congenital heart disease (22%)
  - VACTERL syndrome
  - Other renal and GI anomalies
- Chronic aspiration and difficulty feeding mandate early surgical intervention
- Early surgical treatment starts with rigid bronchoscopy and is usually followed by primary repair (vs. staged repair)

VACTERL

- **VACTERL**: Newer term to include VATER syndrome
  - **V** – Vertebral anomalies (scoliosis, hemivertebrae, fused vertebrae & absent pedicles)
  - **A** – Anal atresia (imperforate anus & rectal fistulae)
  - **C** – Cardiac disease (VSD, ASD)
  - **T** – TEF
  - **E** – Esophageal atresia
  - **R** – Renal anomalies (unilateral renal agenesis)
  - **R** – Radial (dysplasia & absent digits)
  - **L** – Limb defects (typically lower limb)
- Non-Mendelian inheritance with multiple sites suggestive of early insult during embryogenesis
- Components of the VATER association may manifest in isolation but are more likely to be associated with other components
- Certain syndromes including Klippel-Feil syndrome, Sprengel deformity & Goldenhar syndrome have been associated with components of the VACTERL association
- A complete and intensive workup is necessary in any patient who manifests any component of the association with special attention to renal and cardiac assessment
- **Most common cause of death is cardiac arrest**
- Patients with VACTERL syndrome will require TEF surgery, and potentially other surgical procedures due to their associated defects
  - Diverting colostomy & a pull through procedure to repair the anal atresia may be necessary
  - Later surgeries involve repair of remaining radial, vertebral & urogenital defects
  - Patients remain at high risk for GERD following TEF repair

ANESTHETIC CONSIDERATIONS:

1. Considerations of the neonate and possible prematurity
2. A patient presenting with one of the above issues should prompt a search for other anomalies, particularly cardiac, TE fistula, and renal
3. Vertebral defects must be considered with regard to neuraxial anesthetics (lumbar U/S pre-op)

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