Gastroschisis, Pyloric stenosis and Congenital Hernia

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Introduction

- **Gastroschisis** – usually presents as a *neonatal emergency* in *premature infants* (around 60% incidence of prematurity) soon after birth.

- **Pyloric stenosis**- a common *medical* but not a *true surgical emergency*. Usually present around 2-6 weeks of life depending on the parents’ recognition of symptoms.

- **Congenital hernia**- the child may present in infancy or may be in toddler age group.
Gastroschisis

- Greek word for `Belly cleft`.
- Evisceration of gut through 2-3 cm defect in anterior abdominal wall lateral to umbilicus.
- Usually on the right.
- Absence of covering or sac.
- Incidence: 1:15,000-30,000 live births.
Clinical presentation

- Usually contains small bowel with no surrounding membrane.
- Infarction/atresia of bowel is common.
- Liver is rarely in herniated contents.
- Herniated bowel- non rotated and devoid of secondary fixation to posterior abdominal wall.
- Bowel exposed to amniotic fluid.

Complications- chemical peritonitis, edema, heat loss, infection and thickening of intestine.
Gastroschisis
<table>
<thead>
<tr>
<th></th>
<th>Gastrochisis</th>
<th>Omphalocele</th>
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</thead>
<tbody>
<tr>
<td>Incidence</td>
<td>1:15,000-30,000</td>
<td>1:6,000</td>
</tr>
<tr>
<td>Peritoneal covering/sac</td>
<td>Absent</td>
<td>Present</td>
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<tr>
<td>Location of defect</td>
<td>Periumbilical</td>
<td>Within the umbilical cord</td>
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<tr>
<td>Herniated bowel</td>
<td>Matted, edematous</td>
<td>Normal</td>
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<tr>
<td>Associated anomalies</td>
<td>Low (10-15%)</td>
<td>High (40-60%)</td>
</tr>
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<td></td>
<td>Intestinal atresia (15%)</td>
<td>Congenital heart ds. Beckwith-Weidemann syndrome</td>
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Etiology

• Exact cause unknown.

• Theories:
  • Intrauterine occlusion of omphalomesenteric artery → ischemia and atrophy of abdominal muscle.
  • Early fetal rupture of an omphalocoele.
  • Rupture of umbilical cord at site of resorbed Rt. Umbilical vein.
Maternal predisposing factors

- Age <20 yrs
- Primiparas
- Smoking
- Alcohol
- Recreational drugs- Mephamphetamine, LSD, Cocaine, Morphine etc.
- Medications (NSAIDs, Pseudoephedrine)
- Genetic- Trisomy 13, 18, 21 and Monosomy 22
Associated anomalies

- Hydronephrosis
- Arthrogryposis
- Hypoplastic gall bladder
- Meckel’s diverticulum
- Oligohydramnios (IUGR)
- **CVS anomalies**: Persistent Pulmonary HT, Peripheral Pulmonary Stenosis, SVT
Diagnosis

• **Antenatal**

• ↑ maternal serum Alpha Fetoprotein more than 200-300 times.

• **USG** before 20 weeks gestation
  
  - **Early pregnancy**- bowel loops seen floating in amniotic fluid
  - **Later**- bowel obstruction, peritonitis, bowel perforation, fetal growth restriction

• **Postnatal**- obvious appearance
Treatment modalities

• *No intrauterine treatment* modality available for Gastroschisis at present.
• It is a *neonatal emergency* requiring urgent closure.
• The sooner the bowel is reduced, the more likely the primary closure can be achieved and less severe the degree of bowel edema and fibrinous coating.
Primary closure

• **Advantages**: reduced sepsis, sac dehiscence, prolonged ileus

• **Disadvantages**: increased hypotension, bowel ischemia, anuria, respiratory failure

• **Contraindications**: intragastric or intravesical pressure >20cmH₂O, change in CVP ≥ 4mmHg, EtCO₂ ≥ 50mmHg, PIP ≥ 35cmH₂O
Staged closure

• Silo chimney
• Silastic silo prosthesis
• Defect closure after 7-10 days as the bowel edema gradually decreases and abdominal wall expands.
• **Advantages** - avoids abdominal visceral compression, allows early extubation.
Silo prosthesis
Anaesthesia management

- Anaesthesia concerns
  - Patient related: neonatal age group usually premature
  - Severe dehydration due to massive fluid loss
  - Hypothermia due to heat loss
  - Sepsis
  - Metabolic abnormalities specially hypoglycemia
  - Associated congenital anomalies
• **Surgery related:** difficulty in surgical closure
• Risk of hypotension due to traction on liver (if herniated)
• Direct trauma to herniated bowel
• **Anaesthesia related:** premature neonate
• Anticipated difficult airway
• Altered drug metabolism
• Risk of aspiration
• Coagulation abnormalities due to prematurity or associated sepsis
• Almost always need post op mechanical ventilation for 24-48 hrs.
Anaesthetic goals

- **Preoperative:** prevention of infection
  Minimization of fluid and heat loss

- **Intraoperative:** maintenance of body temperature
  Fluid replacement
Preoperative evaluation

• Gestational age, birth history and weight
• Inspection of protruding viscera
• Airway assessment
• Assessment of associated congenital anomalies
• Assess for hemodynamic stability and hydration status (peripheral pulses, skin turgor, capillary refill time, urine output etc.)
• CVS and Respiratory system evaluation
Preoperative stabilization

- Neonatal **emergency**, needs optimization before surgery
- **Cover the exposed viscera** with plastic wrap or damp gauze to ↓ infection and loss of fluid and heat.
- *Bowel not to be twisted* - it impairs vascular integrity
- Cover the neonate’s lower half of body to reduce heat loss.
Preoperative stabilization continued...

- **Airway support**- only if the child requires resuscitation.
- **Gastric decompression**- prevents aspiration. Air going past pylorus is irretrievable and causes difficulty in repair.
- Initial fluid requirement is increased up to 2-4 times daily maintenance requirement (≥ 8-16ml/kg/hr).
- Losses replaced by isotonic saline.
Preoperative stabilization continued...

- **Hypoglycemia** is very common, give 10% Dextrose at 5-7mg/kg/min.
- 5% albumin should constitute 25% of replacement fluids to maintain oncotic pressure.
- Correct **acid base** and **electrolyte** imbalance.
- **Vitamin K**
- **Antibiotic** prophylaxis
Preoperative Investigations

• CBC and cross match 1 unit of blood
• Bld. glucose, Serum electrolytes
• ABG
• CXR
• ECG, ECHO
• Renal USG if associated renal anomaly
• Coagulation profile
• Head USG to rule out intracranial hemorrhage in premature infants
Anaesthesia technique

- Primary closure
- OT preparation
- **Monitoring:**
  - *Routine:* ECG, SpO₂, NIBP, EtCO₂
  - Precordial stethoscope
  - Temperature
  - CVP in very sick neonates, arterial cannulation for IBP, ABG
  - Intragastric and airway pressures
  - Urine output
• **Preinduction:** Nasogastric tube aspiration  
  i.v line preferably in upper limb  
  Inj Atropine 0.02 mg/kg i.v  

• **Induction:** Inj Fentanyl 1-2 mcg/kg iv  

• Inhalational induction with Sevo/ Halo with O₂ in air followed by Inj Atracurium 0.5mg/kg iv  

• IV induction with Inj Thiopentone (2-4mg/kg i.v) + Inj Atracurium  

• Awake intubation under sedation followed by muscle relaxant (anticipated difficult airway).
Anaesthesia technique continued...

- **Maintenance**: O₂ + Air + Sevo/Iso + Inj Atracurium
- N₂O contraindicated
- **Intraop analgesia**: Inj Fentanyl 3-5mcg/kg i.v (if post op ventilation anticipated).
- Maintain SpO₂ 95-97% in term neonate 90-93% in preterm
- Adjust FiO₂ to maintain PaO₂ 60-80mmHg
Anaesthesia technique continued...

• Intraop fluids- 5% dextrose with 0.18% saline for maintenance.
• RL 8-15ml/kg/hr for third space losses.
• Intraoperative requirement upto 25% of estimated blood volume expected during repair of large defects.
• Target Hb- 10-12 g/dl. Give warmed blood when required.
• Platelets and FFPs- 10ml/kg if low platelet count or coagulation profile abnormal
Criteria for post-operative mechanical ventilation

- Size of patient
- Prematurity
- Intraop events
- Associated pathology
- Hemodynamic status
- Magnitude of abdominal defect
- **Extubation**: small defect, fully awake, regular spontaneous breathing, vigorous movement of all limbs and maintaining SpO₂ with stable hemodynamics.
Postoperative care

- **Fluid management**: 60% of maintenance requirement immediately postop.
- Check fluid balance and electrolytes for subsequent fluid requirement.
- Initially 10% dextrose or 5% dextrose with 0.18% saline
- GI loss with NS/RL
- Colloids for third space loss.
Postoperative care continued...

- **Glucose control**: regular Bld. Glucose monitoring and treatment with 1-2 ml/kg of 10% dextrose if required.
- **Prolonged ileus expected**: TPN till full feeds established.
- **Pain relief**: wound infiltration
  - PCM rectal suppository
  - iv Fentanyl (if on ventilatory support).
- **Antibiotics**
Postoperative complications

- Necrotizing enterocolitis
- Adhesive intestinal obstruction
- Gastroesophageal reflux
- Abdominal compartment syndrome
- Abdominal wall breakdown
- Wound infection, sepsis
- Renal insufficiency
- Pneumonia
- TPN related: metabolic derangement, cholestasis
Abdominal compartment syndrome

- Occurs as a result of closure of abdominal wall defect under pressure.
- Tight abdominal closure impairs diaphragmatic excursion leading to ventilatory compromise.
- IVC compression $\rightarrow$ impaired venous return $\rightarrow$ hypotension
- Aortocaval compression $\rightarrow$ bowel ischemia and necrosis, $\downarrow$ cardiac output, hepatic and renal insufficiency
Abdominal compartment syndrome continued...

- **Diagnosis**: signs of decreased peripheral perfusion, lower extremity congestion and cyanosis
- Increased intra-abdominal and intravesical pressures > 20cmH₂O
- Increased airway pressure and decreased compliance
- **Treatment**: removal of fascial sutures and closure of only skin or addition of prosthesis.
Hypertrophic Pyloric Stenosis

- One of most common GI disorders during early infancy.
- Described by Hirschsprung in 1888.
- Hypertrophy of circular muscles of pylorus results in constriction and obstruction of gastric outlet.
Epidemiology and Etiology

- **Incidence**: 1-2/1000 live births
- **Epidemiology**: more in first born males
  - M:F - 4-5:1
- **Etiology**: Unknown
- Genetic- 11q14-22 and Xq23
- Familial
- Gender
- Ethnic origin- more in whites
Associated anomalies

- Esophageal atresia
- Tracheoesophageal fistula
- Hirschsprung disease
- Exomphalos
- Inguinal hernia
- Hypospadias
- Undescended testis
Clinical presentation

• **History**: 2\textsuperscript{nd} - 8\textsuperscript{th} week of life
• Projectile, frequent episodes of non-bilious vomiting 30-60 minutes after feeding
• Weight loss
• Persistent hunger
• Jaundice (2%)- due to decreased hepatic glucoronosyl transferase associated with starvation
• **Examination:**
  • Palpable *olive shaped* mass (1.5-2cm) to the right of epigastric area.
  • Visible gastric peristalsis from Lt. upper quadrant to epigastrium
  • **s/o dehydration**
Pathophysiology

- Vomiting → loss of H⁺ and Cl⁻ → Hypochloremic hypokalemic metabolic alkalosis
- Protracted vomiting → ECF volume deficit → urinary excretion of K⁺ and H⁺ to preserve Na⁺ and water
- Initial alkalotic urine becomes acidotic - Paradoxical aciduria
- *Hypochloremic hypokalemic metabolic alkalosis with paradoxical aciduria with secondary respiratory acidosis*
- Hyponatremia may not be evident because of hypovolemia
Diagnosis

- History and physical examination
- **Abdominal USG**: Pyloric muscle thickness >3-4mm or pyloric length > 15-18mm in presence of functional gastric outlet obstruction - *Diagnostic*
- **Upper GI study** when atypical presentation or negative USG
- *Diagnostic*: narrowed, elongated pyloric channel with pyloric mass effect on stomach and duodenum – *String/ Double tract/ Beak/ Pyloric teat sign*
Barium swallow

- Air filled fundus
- Duodenal bulb
- Narrowed pyloric channel
- Barium filled antrum

String sign

Normal stomach
• **Treatment**: medical emergency but NOT surgical emergency
• **Definitive treatment**: Ramstedt Pyloromyotomy

• **Anaesthetic considerations**
  • *Patient related*: infant age group
  severe dehydration
  electrolyte imbalance
  • *Surgery related*: open/ laparoscopic
  Celiac reflex
  • *Anaesthesia related*: pulmonary aspiration
  PONV
Preoperative investigations

• Hemoglobin
• S. Electrolytes
• BUN
• ABG
• Bld. Sugar
Preoperative preparation

• **Correction of fluid deficits** - over 24-48 hrs
  - **Deficit**: isotonic fluid 0.9% saline (20ml/kg bolus)
  - **Maintenance**: 0.45% saline in 5% Dextrose at 1.5 times maintenance rate +10-40 meq/L KCL added once urine output established

• Correction of electrolyte imbalances

• **Prevention of aspiration**: aspiration through NGT

_Surgery should only take place when dehydration corrected, normal S. Na and K, Cl⁻ > 90mmol/L, HCO₃ <28mmol/L and BE <+2._
Anaesthetic management

- **Technique**: GA with controlled ventilation with endotracheal intubation
- **Goals**: normoxia
  - Normocapnia
  - Normothermia
  - Normovolemia
  - Electrolyte balance
- Avoid bradycardia, aspiration of gastric contents
Anaesthesia technique

• OT Preparation
• **Monitoring:** Precordial stethoscope
  ECG, NIBP, SpO₂
  Capnography, temperature
  Urine output
  ABG

• **Preinduction** – Nasogastric tube aspiration, Inj Atropine 0.02 mg/kg iv

• **Induction** – Inj Fentanyl 1-2 mcg/kg iv
• Inhalational induction with Sevo/ Halo with O₂ ± muscle relaxant.
• IV induction with Inj Thiopentone + Inj Atracurium
• Awake intubation under sedation (if anticipated difficult airway)
• NGT reinserted after orotracheal intubation.
• **Maintenance**: inhalational agents + muscle relaxants (if paralyzed)
• **Intraop fluids**: isotonic fluids
• **Awake extubation** after reversal with Neostigmine (50-70mcg/kg i.v) and Atropine (0.02mg/kg i.v)
Postoperative care

• **Post op pain relief:**
  - Acetaminophen 30-40mg/kg rectal suppository
  - LA infiltration of surgical incision

• **Post op concerns:** respiratory depression and apnea due to CSF alkalosis and intraop hyperventilation
  - Hypoglycemia
  - PONV - usually self limiting. Early feeding is recommended post op.
  - Avoid hypothermia
Congenital Inguinal Hernia

• Most common congenital disorders managed by paediatricians and paediatric surgeons.
• Can result in loss of testis/ ovary/ portion of bowel (if incarceration or strangulation occurs).
• Timely diagnosis and operative treatment is important.
Embryology

- **Processus vaginalis** is a peritoneal diverticulum extending through the internal inguinal ring.
- Present in fetus at 12 wks in utero.
- Gives rise to indirect inguinal hernia.

- *Patent processus vaginalis is only a potential hernia and becomes an actual hernia only when bowel or other intra-abdominal contents exit the peritoneal cavity into it.*
Incidence

- **Indirect hernia**- 1-5%, increased in premature infants, M: F - 8-10: 1
- **Associated diseases**- Cystic Fibrosis (15% incidence of inguinal hernia)
- Connective tissue ds.- Ehlers Danlos syndrome
- Mucopolysaccharidosis- Hunter- Hurler syndrome
- **Direct hernia**- rare in children (1/3 of direct hernias in children with previous indirect hernia repair)
Clinical presentation

- **Indirect hernia** - groin bulge extending toward the top of scrotum visible most frequently during periods of ↑ abdominal pressure.
- **Direct inguinal hernia** - groin mass extending towards the femoral vessels with exertion or straining.
- Mostly present for elective surgery but may present as emergency in case of incarceration or bowel obstruction.
- **Treatment** - Herniotomy/ Herniorrhaphy
Anaesthesia concerns

• **Patient related**- age group (infant or toddler)
• **Associated anomalies** - Cryptorchidism, Cleft lip and palate, Congenital heart ds. like VSD, valvular anomalies
• **Anaesthesia related**- usually a day care surgery
• Need for psychological preparation
• Assessment and exclusion of children with severe URI
Preoperative preparation

• Usually toddler age group
• **Psychological preparation** - to allay separation anxiety, fear of physical injury and fear of strange or unknown.
• Gentle examination preferably by anaesthesiologist involved in that particular surgery.
• Explain the procedure to the older child in familiar and positive terms.
• Allowing parents in induction room.
Anaesthesia management

• Check NPO status on morning of surgery.
• Premedication
• **Sedation** – Midazolam 0.25-1 mg/kg oral 15-20 min before surgery
• OT preparation
• **Monitoring**: Precordial stethoscope
• NIBP, ECG
• EtCO₂, SpO₂
• Temperature
Anaesthesia management continued...

• **Induction** – Inhalational induction with Sevo / Halo or iv with Propofol/ Thiopentone
• **Airway device** – LMA with spontaneous breathing/ ETT with muscle relaxant (emergency or < 1 year of age)
• **Maintenance** - Oxygen + N₂O + Sevo/Iso + muscle relaxant (if paralysed).
• Child should be well anaesthetized during spermatic cord manipulation as inadequate depth can result in laryngospasm.
• **Reversal**- Neostigmine + Glycopyrrolate/ Atropine iv (if muscle relaxant given)
• LMA should be preferably removed in deeper plane of anaesthesia.
• **Post op concerns**- pain relief, PONV
Modalities for post op pain relief

- Paracetamol rectal suppository
- Regional analgesia techniques:
  - Caudal analgesia
  - Ilioinguinal and Iliohypogastric nerve block
  - Transversus abdominis plane block

These are quite effective in providing postoperative pain relief and in decreasing intra-operative anaesthetic requirements.
Subarachnoid block for inguinal hernia repair

- Specially for premature infants in whom inhalation anaesthetic may be contraindicated.
- Spinal anaesthesia has been used successfully to avoid general anaesthesia and endotracheal intubation.
- It decreases the incidence of postoperative adverse events.
- Dose- 0.5% Bupivacaine 1mg/kg
Day care surgery

- **Indications**: elective peripheral surgery
- No associated medical/surgical comorbidity
- No previous history of anaesthesia problems or PONV
- Adequately controlled pain
- Social issues
Day care surgery continued...

- **Technique:** use of short acting inhalational agents like Sevo, opioids like Fentanyl, Remifentanil.
- Regional analgesic techniques where applicable
- **Discharge criteria:** conscious, comfortable
- No vomiting
- Clear written instructions to the parents.
- Accessibility to the hospital in case of any problem.
Summary

• *Gastroschisis* manifests as external herniation of abdominal viscera through a small (usually <5cm) defect in anterior abdominal wall.

• *This is a neonatal emergency requiring urgent surgical repair at specialised centres well equipped for management of these patients.*

• *Pyloric stenosis is a common medical but not a true surgical emergency presenting around 2-6 weeks of life.*
• This requires preoperative correction of fluid, electrolyte and acid base imbalances over 24-48 hours before definitive surgery.

• Congenital inguinal hernia repair is usually an elective surgical procedure in infancy or more commonly toddler age group.

• Psychological preparation, management of postop pain and PONV are the primary concerns in this age group.
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Thank you!