CLEFT LIP & PALATE
- Definition: facial malformations that occur during embryonic development (6-12 weeks gestation) causing a failure of the maxillary and median nasal structure to fuse.

- Nonunion of the tissue and bone of the upper lip and hard/soft palate during embryologic development
- Familial disorder, often associated with other congenital abnormalities.
  - Cleft lip/palate 1 in 1000 births
  - Cleft lip with or without cleft palate affects more boys; cleft palate affects more girls.
CLEFT LIP & PALATE

- Etiology: unknown; may occur as part of a genetic disorder or environmental factors such as:
  a) Exposure to teratogens
  b) Familial tendency
  c) Increased incidence in Asians, Native Americans, lowest in African Americans.

CLEFT LIP

- Signs and symptoms
  a) May be unilateral or bilateral
  b) Varies from simple notching of vermillion border of the lip to a deep cleft, extending through the lip or into the nose.

CLEFT PALATE

- Signs and symptoms
  a) Midline fissure or opening in the hard and/or soft palate areas
  b) Difficulty forming a seal for sucking
  c) Coughing and choking
  d) Nasal distortion
  e) Congestion
  f) Failure to thrive (un-repaired defect with persistent feeding difficulties)
Figure 46-1A: Variations in clefts of lip and palate.

A. Bilateral cleft lip and palate. B. Unilateral cleft lip and palate. C. Unilateral cleft lip and palate. D. Cleft palate.

Figure 46-1B: Bilateral cleft lip.

Figure 46-1C: Cleft lip.
CLEFT LIP & PALATE

- Diagnostic tests and labs:
  - a) History and physical
  - b) Prenatal ultrasound
  - c) Pre-op laboratory data (CBC, electrolytes)
  - d) Wound and sputum culture if infection is suspected

Medical Management

- A team approach for therapy
  - Speech Therapist
  - Dentist and Orthodontist
  - Audiologist
  - Otolaryngologist
  - These children are prone to otitis media and possible hearing loss
  - Pediatrician

Surgical Management

- Timing varies with severity of the defect; early correction helps to avoid speech defects
  - Cleft Lip Repair
    - Usually performed at age 2 months
  - Cleft Palate Repair
    - Usually performed at 18 months in anticipation of speech development
Figure 46-2A  Repaired cleft lip.

Figure 46-2B  Repaired cleft lip.

Nursing Care Pre-op

- Feed in upright position to decrease possible aspiration and swallowing of air
- Burp frequently
- Special feeding devices as needed:
  - Cleft palate nipple
  - Syringe with soft rubber tubing
  - Soft nipple with enlarged hole
- Tube feeding as necessary
- Emotional Support to parents:
  - Demonstrate benefits of surgery
  - Reinforce that disorder is not their fault and will not affect child’s life span or mental ability
  - Prepare for surgery
**Nursing Care Pre-op**

- Position on side with head of the bed elevated
- Provide oral care and suction as needed
- Teach family to use bulb suction to clear airway

**Post-op Nursing Care**

- **Cleft Lip Repair**
  - Maintain patent airway
  - Position on back or side
  - **AVOID ANY STRAIN ON SUTURE LINE**

- Use elbow restraints to keep child’s hands away from the suture line
- Keep suture line clean
- Teach parents to keep suture line clean at home
Post-op Nursing Care

- Cleft Palate Repair
  - Position on side or abdomen for drainage of blood/mucus
  - Have suction at the bedside for emergency use
- Prevent injury or trauma to suture line
- Give water after feedings to clean suture line
- Rinse mouth with water after feeding
- Hold and cuddle these babies

Post-op Nursing Care

- Address pain management needs
- Avoid placing hard items in mouth (suction catheters, spoons, ice chips)
- Use a cup for drinking to avoid placing anything in mouth
- Monitor for bleeding, infection or breakdown at the surgical site

Complications

- Otitis media and hearing loss
- Speech difficulties
- Pulmonary complications from aspiration
- Malocclusion due to abnormal teeth eruption
Altered Connections between Trachea, Esophagus and Stomach

- Congenital rare malformation that represents a failure of the esophagus to develop as a continuous passage.
- Often found in:
  - Low birth weight infants
  - HX of maternal polyhydramnios

TYPES

- **Esophageal atresia**
  - Esophagus ends in a blind pouch; no entry route to the stomach
- **Tracheoesophageal fistula (TEF)**
  - Open connection between trachea and esophagus
- **Esophageal atresia with TEF**
  - Esophagus ends in blind pouch, stomach end of esophagus connects with trachea
Management

- Drug therapy
  - antibiotics for respiratory infections
- Surgery
  - Palliative
    - Gastrostomy for placement of a feeding tube
  - Corrective
    - End-to-end anastomosis to correct the defect and restore normal anatomy

Assessment Findings

- Esophageal atresia
  - Inability to pass an NG tube
  - Increased drooling and salivation
  - Immediate regurgitation of undigested formula/milk when fed
  - Intermittent cyanosis from choking on aspirated secretions
  - C-C-C (choking, coughing, cyanosis)
Assessment Findings

- TEF
  - Normal swallowing but some food/mucus crosses fistula, causing choking and intermittent cyanosis
  - Distended abdomen from inhaled air crossing fistula into stomach
  - Aspiration pneumonia from reflux of gastric secretions into the trachea

Nursing Interventions

Pre-operative
- Head of bed slightly lowered-minimize aspiration of secretions into trachea
- Keep NPO
- Provide suctioning
- IV fluids
- Maintain patent airway and lung expansion
- Recognize defect early (excess salivation)
- Prevent aspiration pneumonia

Nursing Interventions

Post-operative
- Provide nutrition
- Support parents
- Provide client teaching and discharge teaching
  - Alternative feeding methods (GT and then advance to oral feeding as child progresses)
  - Signs of respiratory distress
  - Suctioning
  - CPR
Nursing Interventions

- Promote respiratory function
  - a) Suction secretions as needed
  - b) Maintain care of chest tube and drainage apparatus
  - c) Administer oxygen as indicated
  - d) Prevent aspiration of feedings
- Prevent infections at the operative sites
- Maintain fluid and electrolyte balance
- Comfort measures (pacifier, tactile stimulation)

GASTROESOPHAGEAL REFLUX

GER

- Relaxation or incompetence of the lower esophagus sphincter which results in frequent return of stomach contents into the esophagus
GER

- Reversal of flow of stomach contents into lower portion of the esophagus
- Common in premature infants due to hypotonia
- Caused by relaxed cardiac sphincter or overdistension of stomach by gas or overfeeding
- Results in local irritation of lining of esophagus from gastric secretions

Manifestations of GER

- Infants
  - “Spitting up”/vomiting
  - Irritability
  - Weight loss
  - Frequent URIs
  - Respiratory problems
  - Life-threatening apnea
  - Bloody stools/emasisis
  - Anemia
- Older children
  - Heartburn
  - Abdominal pain
  - Chest pain (noncardiac)
  - Nocturnal asthma
  - Recurrent pneumonia
  - Chronic cough

Diagnostics

- History
- Fluoroscopic observation of reflux following a barium swallow
- Upper gastrointestinal endoscopy
- Direct measurement of pH (probe monitoring) of the distal esophagus
- Scintigraphy which detects radioactive substances in the esophagus after a feeding of the compound
Nursing Interventions
- Position with head elevated 30-45°
- Small, frequent feedings with adequate burping
- Provide client teaching and discharge planning
  - Teach parents how to position and feed infant
  - Administration of medications

Treatment
- Mild
  - Modification of feeding habits
    - Thicken feedings
    - Avoid fatty foods and citrus
  - Medication
    - Antacids, histamine blockers, metoclopramide
  - Positioning
    - Prone after feedings

Treatment
- Severe
  - Fundoplication
    - Wrapping of the fundus of the stomach around the distal esophagus
    - GT usually inserted during surgery and left in place for six weeks
PYLORIC STENOSIS

- Hypertrophy (thickening) of the pyloric sphincter causing stenosis and obstruction
- 5 in 1000 births, more common in Caucasian, firstborn, full-term boys
- Cause unknown; possibly familial

PYLORIC STENOSIS

- Pathophysiology
  - a) Hypertrophy of the pylorus muscle (stenosis of stomach lumen)
  - b) Lumen (inflamed and edematous/complete obstruction)
  - c) Projectile vomiting (dehydration and electrolyte depletion)

Assessment Findings

- Olive-size mass in RUQ
- Peristaltic waves during & after feedings (move left to right towards pylorus)
- Hyperactive BS
- Vomiting
  - As obstruction increases, vomiting becomes more forceful and projectile
  - Hungry after vomiting
- Dehydration
**Diagnostics**

- Upper GI
- Pyloric ultrasound
- Electrolyte imbalance
  a) Severe depletion of water and electrolytes from extensive vomiting
  b) Decreased serum levels of sodium and potassium
  c) Increased pH and bicarbonate levels (metabolic alkalosis)

**Interventions**

- Replacement of fluid and electrolytes
- NPO
  a) Promote gastric decompression
  b) Carry out lavage
  c) Maintain patency of NGT
  d) Measure and record amount and type of drainage
**Interventions**

- Surgery: Pylorotomy (the circular muscle fibers of the pylorus are released to allow the passage of food and fluids - incision through muscle fiber)
  - Clear liquids post-operatively; then advance
  - HOB elevated

**Interventions**

- Assess adequacy of intake
  - Weigh daily
  - Measure carefully: intake (PO and parental), output (vomitus, NGT drainage, stools, urine)
  - Urine specific gravity

**HIRSCHSPRUNG’S DISEASE (Aganglionic Megacolon)**

- Absence of autonomic parasympathetic ganglion cells in a portion of the large colon resulting in decreased motility in that portion of the colon and signs of functional obstruction
- Usually diagnosed in infancy
Hirschsprung’s Disease

- When stool enters the affected part of the colon, lack of peristalsis causes it to remain there until additional stool pushes it through; colon dilates as stool is impacted
- Familial disease; more common in boys; associated with Down’s syndrome

Assessment

- Failure or delay in passing meconium
- Abdominal distension
- Chronic constipation
- Foul, ribbon-like stools
- Bile-stained emesis
- Nausea, anorexia, lethargy
- Weight loss
### Assessment: Newborn
- Failure to pass meconium within 24-48 hours after birth
- Reluctance to ingest fluids
- Bile stained vomitus
- Abdominal distension

### Assessment: Infancy
- Failure to thrive (FTT)
- Constipation
- Abdominal distension
- Episodes of diarrhea and vomiting

### Assessment: Childhood
- Constipation
- Passage of ribbon-like-stool
- Foul smelling stool
- Abdominal distention
- Visible peristalsis
- Fecal masses easily palpable
- Poorly nourished
- Anemic
- Hypoproteinemic from malabsorption of nutrients
Diagnostics
- Barium enema
- In neonate, Dx is usually made based on clinical signs of intestinal obstruction and failure to pass meconium
- Rectal exam
  - Rectum empty of stool
  - Rectal biopsy confirms the diagnosis (absence of ganglia)

Treatment
- Drug Therapy: stool softeners
- Isotonic or Mineral Oil enemas
- Diet Therapy: low residue, diet modification (milder defect)
- Surgery:
  - Palliative: loop or double-barrel colostomy
  - Corrective: abdominal-perineal pull through; bowel containing ganglia is pulled down and anastomosed to the rectum

Nursing Interventions
- Administer enemas as ordered
- Administer TPN as ordered
- Provide low residue diet
- Provide client teaching and discharge planning concerning colostomy care and low residue diet
IMPERFORATE ANUS

- Congenital malformation caused by abnormal fetal development
- Many variations
- Often associated with fistula formation to rectum or vagina and other congenital anomalies

Imperforate Anus

- Surgical correction performed in stages with completion at about age 1 year
- May need temporary colostomy

Assessment

- No stool passage within 24 hours of birth
- Meconium stool from inappropriate orifice (appears in urine)
- Inability to insert rectal thermometer
- Absence of an anal opening
- Checking for patency of the anus and rectum is a routine part of the newborn assessment including observation or inquiries regarding the passage of meconium
Management

- Manual dilation for anal stenosis
- Surgery
  - Anoplasty (reconstruction of the anus)
  - Colostomy for higher anomalies in infants
- Prophylactic Antibiotics

Nursing Interventions

- If suspected, do NOT take rectal temperature because of risk of penetrating wall and causing peritonitis
- Perform manual dilation as ordered, instruct parents in proper technique
- After surgery keep incision clean as possible
- Use side-lying/prone position post-surgery
- For colostomy teach parents colostomy care (dressing change, skin care, correct application of collection devices)

CONSTIPATION

- Decrease in number of bowel movements with large, hard stools
- May be caused by high fat and protein and low fluid diet
- May cause bowel obstruction if severe
Assessment
- Less frequent stools
- Difficulty eliminating stool
- Hard consistency compared to normal pattern
- Bleeding with stoolsing
- Abdominal pain

Management
- Drug therapy
  - Stool softeners
  - Suppositories
  - Enemas
- Diet
  - Increased fiber and fluids

Nursing Interventions
- Assess for other pathologic causes
- Dietary modification
- Apply lubricant around anus
- Remove stool digitally if possible
- Provide prune juice (1 oz), add fruits to diet
DIARRHEA

- Inflammatory process of GI lining
  - Acute caused by:
    - Infections (bacterial, viruses)
    - Antibiotic therapy
    - Diet conditions
  - Chronic caused by:
    - Malabsorption disorders
    - Structural defects
    - Allergic disorders

Assessment

- Frequent watery stools
- Anorexia
- Dehydration
- Weight loss

Nursing Interventions

- Mild diarrhea – home treatment
  - ORT (oral rehydration therapy)
  - Dietary advancement (clear liquids, electrolyte solutions)
  - Bland/BRATT diet
  - Infants may need soy formula
  - Teach parents signs & symptoms of dehydration
**Nursing Interventions**
- Severe diarrhea – hospitalize esp. infants
  - Isolation (enteric) until organism isolated
  - NPO to rest bowel
  - Fluid & electrolyte replacement
  - Strict I & O
  - Daily weight
  - Skin care

**Gastroenteritis**

- **Definition:** an inflammation of the mucous membranes of the GI tract characterized by vomiting and diarrhea resulting in F&E losses that lead to dehydration and electrolyte imbalances.

- **Predisposing factors:**
  - Age (NBs and infants)
  - Impaired health
  - Climate
  - Environment
Gastroenteritis

Pathophysiology:
- Pathogens infect the cells
- Produce enterotoxins that damage the cells
- Or, enterotoxins adhere to the walls of the intestines

Enteropathogenic organisms:
- Shigella
- Salmonella
- Escherichia Coli

Transmission:
- Fecal-oral route, person to person
- Contaminated water and food supplies
- Exposure to day care facilities increased risk
- Travel to other countries
Gastroenteritis

- Other causative factors:
  - Viruses
  - Stress
  - Fever
  - Parasites
  - Food intolerance
  - Overfeeding
  - Medications
  - Colon disease

Gastroenteritis

- Major physiological disturbances:
  - Dehydration
  - Electrolyte imbalance
  - Metabolic acidosis

Gastroenteritis

- Diagnostic evaluation:
  - Stool specimen
  - Serum electrolytes
  - Hematocrit
  - BUN
Gastroenteritis

- Signs of dehydration:
  - Sunken fontanel
  - Sunken eyes
  - Poor skin turgor
  - Dryness of mouth
  - Loss of weight
  - Increased heart rate
  - Concentrated urine
  - Irritability

Gastroenteritis

- Treatment:
  - Home care
  - Oral hydration therapy (Pedialyte>1/2 st formula>FS formula)
  - BRATT diet contraindicated

Gastroenteritis

- Contraindicated foods:
  - Carbonated beverages and those containing sugar
  - Caffeinated soda
  - Chicken or beef broth
Gastroenteritis

- Nursing interventions:
  - Assessment and history
  - Weight and vital signs
  - Monitor IV fluids
  - I & O
  - Description of stools, vomitus, and urine
  - Skin care
  - Good hand washing