



# **PEDIATRIC SURGERY HANDBOOK**

**Humberto L Lugo-Vicente, MD, FACS, FAAP\***  
**Professor/Associate Director of Pediatric Surgery**  
**University of Puerto Rico School of Medicine**  
**University Pediatric Hospital**  
**Chief - Section Pediatric Surgery**  
**San Pablo Medical Center**  
**<titolugo@coqui.net>**

CONTENT

<p><b>I. INTRODUCTION</b></p> <p>A. Neonatal Physiologic Characteristics</p> <p>    1. Water Metabolism</p> <p>    2. Fluid and Electrolytes Concepts</p> <p>B. Variations in Individual Newborns</p> <p>    1. Types of Newborns Infants</p> <p>    2. Metabolic and Host Defenses</p> <p>    3. Surgical Response of Newborns</p> <p><b>II. HEAD AND NECK LESIONS</b></p> <p>A. Cervical Lymphadenopathy</p> <p>B. Congenital Torticollis</p> <p>C. Thyroglossal Duct Cysts</p> <p>D. Branchial Cleft Fistulas</p> <p>E. Cystic Hygroma</p> <p>F. Cat's Scratch Disease</p> <p><b>III. OBSTRUCTIVE and GI PROBLEMS</b></p> <p>Logical Approach to Neonatal Intestinal Obstruction (by Jordan Weitzman, MD)</p> <p>A. Esophageal Atresia and Stenosis</p> <p>B. Achalasia</p> <p>C. Gastro-Duodenal Anomalies</p> <p>    1. Gastric Anomalies</p> <p>    2. Pyloric Stenosis</p> <p>    3. Duodenal Malformations</p> <p>D. Malrotation and Volvulus</p> <p>E. Intestinal Atresias</p> <p>F. Meconium Ileus</p> <p>G. Hirschsprung's disease</p> <p>H. Imperforate Anus</p> <p>I. Duplications</p> <p>J. Intussusception</p> <p>K. Appendicitis</p> <p>L. Chronic Intestinal Pseudo-obstruction</p> <p>M. Bezoars</p> <p>N. Carcinoid syndrome</p> <p>O. Meconium-related disorders</p> <p>    1. Meconium Ileus</p> <p>    2. Meconium Peritonitis</p> <p>    3. Meconium Plug syndrome and Left hypoplastic colon syndrome</p> <p>P. Foreign Body Ingestion</p> <p><b>IV. HERNIAS, THORACIC CONDITIONS AND ABDOMINAL WALL DEFECT</b></p> <p>A. Diaphragmatic hernias</p> <p>    1. Congenital Diaphragmatic Hernia (Bochdalek)</p> <p>    2. Morgagni Hernias</p> <p>    3. Hiatal Hernias</p> <p>B. Lung Bud Anomalies</p> <p>    1. Congenital Lobar Emphysema</p> <p>    2. Pulmonary Sequestration</p> <p>    3. Cystic Adenomatoid Malformation</p> <p>    4. Bronchogenic Cyst</p> <p>C. Chylothorax</p> <p>D. Empyema</p> <p>E. Spontaneous Pneumothorax</p> <p>F. Pneumatocele</p> <p>G. Inguinal hernias, Hydroceles, Undescended Testis and epigastric hernias</p> <p>H. Mediastinal Cysts</p> <p>I. Umbilical hernias</p> <p>J. Omphalocele and Gastroschisis</p> <p>K. Femoral Hernias</p>	<p><b>V. GASTROINTESTINAL BLEEDING</b></p> <p>A. Upper GI bleeding (Newborn)</p> <p>B. Lower GI bleeding (Newborn)</p> <p>    1. Necrotizing Enterocolitis (NEC)</p> <p>C. Upper GI bleeding (Older child)</p> <p>D. Lower GI bleeding (Older child)</p> <p>    1- Anal Fissure</p> <p>    2- Meckel's Diverticulum</p> <p>    3- Polyps</p> <p>    4- Familial Adenomatous Polyposis</p> <p><b>VI. PANCREATIC HEPATIC, BILIARY and SPLENIC DISORDERS</b></p> <p>A. Annular Pancreas/Pancreas Divisum</p> <p>B. Pancreatitis</p> <p>C. Pancreatic Cysts</p> <p>D. Hepatic Cysts</p> <p>E. Biliary Atresia</p> <p>F. Biliary hypoplasia</p> <p>G. Choledochal Cyst</p> <p>H. Cholelithiasis</p> <p>I. Idiopathic Perforation Bile Duct</p> <p>J. Splenic Cysts, splenoptosis, Spherocytosis and Asplenia</p> <p>K. Splenic Trauma</p> <p>L. Adrenal Hemorrhage</p> <p><b>VII. TUMORS</b></p> <p>A. Wilms tumor</p> <p>B. Neuroblastoma</p> <p>C. Rhabdomyosarcoma</p> <p>D. Liver Tumors</p> <p>E. Teratomas</p> <p>F. Ovarian Tumors</p> <p>G. Thyroid Nodules</p> <p>H. Burkitt's Lymphoma</p> <p><b>VIII. GYNECOLOGIC and INTERSEXUAL CONSIDERATIONS</b></p> <p>A. Labial Adhesions in Infants</p> <p>B. Ovarian Cysts</p> <p>C. Breast Disorders</p> <p>D. Congenital Adrenal Hyperplasia</p> <p>E. Testicular Feminization Syndrome</p> <p>F. Mixed gonadal Dysgenesis</p> <p>G. Mullerian Duct Syndrome</p> <p>H. Hydrometrocolpos</p> <p><b>IX. PRENATAL CONGENITAL MALFORMATIONS</b></p> <p>A. Fetal Surgery</p> <p>B. Fetal Intestinal Obstruction</p> <p>C. Fetal Abdominal Wall Defects</p> <p><b>X. SUGGESTED READING</b></p> <p>    Specific Reading</p> <p>    General Reading</p>
---	---

## I. INTRODUCTION

### A. Neonatal Physiologic Characteristics

#### 1. Water metabolism

Water represents 70 to 80% of the body weight of the normal neonate and premature baby respectively. Total body water (TBW) varies inversely with fat content, and prematures have less fat deposits. TBW is distributed into extracellular fluid (ECF) and intracellular fluid (ICF) compartment. The ECF compartment is one-third the TBW with sodium as principal cation, and chloride and bicarbonate as anions. The ICF compartment is two-third the TBW with potassium the principal cation. The Newborn's metabolic rate is high and extra energy is needed for maintenance of body temperature and growth. A change in body water occurs upon entrance of the fetus to his new extrauterine existence. There is a gradual decrease in body water and the extracellular fluid compartment with a concomitant increase in the intracellular fluid compartment. This shift is interrupted with a premature birth. The newborn's body surface area is relatively much greater than the adults and heat loss is a major factor. Insensible water loss are from the lung (1/3) and skin (2/3). Transepithelial (skin) water is the major component and decreases with increase in post-natal age. Insensible water loss is affected by gestational age, body temperature (radiant warmers), and phototherapy. Neonatal renal function is generally adequate to meet the needs of the normal full-term infant but may be limited during periods of stress. Renal characteristics of newborns are a low glomerular filtration rate and concentration ability (limited urea in medullary interticium) which makes them less tolerant to dehydration. The neonate is metabolically active and production of solute to excrete in the urine is high. The kidney in the newborn can only concentrate to about 400 mOsm/L initially (500-600 mOsm/L the full-term compared to 1200 mOsm/L for an adult), and therefore requires 2-4 cc/kg/hr urine production to clear the renal solute load. The older child needs about 1-2 cc/kg/hr and the adult 0.5-1 cc/kg/hr.

#### 2. Fluid and Electrolytes Concepts

Cellular energy mediated active transport of electrolytes along membranes is the most important mechanism of achieving and maintaining normal volume and composition of fluid compartments. Infants can retain sodium but cannot excrete excessive sodium. Electrolytes requirements of the full-term neonate are: Sodium 2-3 meq/kg/day, potassium 1-2 meq/kg/day, chloride 3-5 meq/kg/day at a rate of fluid of 100 cc/kg/24 hrs for the first 10 kg of weight. As a rule of thumb, the daily fluid requirements can be approximated too:

prematures	120-150 cc/kg/24 hrs
neonates (term)	100 cc/kg/24 hrs
Infants >10 kg	1000 cc+ 50 cc/kg/24 hrs.

Special need of preterm babies fluid therapy are: conservative approach, consider body weight changes, sodium balance and ECF tonicity. They are susceptible to both sodium loss and sodium and volume overloading. High intravenous therapy can lead to patent PDA, bronchopulmonary dysplasia, enterocolitis and intraventricular hemorrhage. Impaired ability to excrete a sodium load that can be amplify with surgical stress (progressive renal retention of sodium). Estimations of daily fluid requirements should take into consideration: (1) urinary water losses, (2) gastrointestinal losses, (3) insensible water losses, and (4) surgical losses (drains).

Blood Volumes estimates of help during surgical blood loss are: premature 85-100 cc/kg, term 85 cc/kg, and infant 70-80 cc/kg. The degree of dehydration can be measured by clinical parameters such as: body weight, tissue turgor, state of peripheral circulation, depression of fontanelle, dryness of the mouth and urine output. Intravenous nutrition is one of the major advances in neonatal surgery and will be required when it is obvious that the period of starvation will go beyond five days.

Oral feeding is the best method and breast is best source. Newborn infants requires 100-200 calories/kg/day for normal growth. This is increased during stress, cold, infection, surgery and trauma. Minimum daily requirement are 2-3 gm/kg of protein, 10-15 gm/kg of carbohydrate and small amount of essential fatty acids.

### B. Variations in Individual Newborns

#### 1. Types of Newborns Infants

a) The full-term, full-size infant with a gestational age of 38 weeks and a body weight greater than 2500 grams (TAGA)- they received adequate intrauterine nutrition, passed all fetal tasks and their physiologic functions are predictable. b) The preterm infant with a gestational age below 38 weeks and a birth weight appropriate for that age (PreTAGA); c) The small-for-gestational-age infant (SGA) with a gestational age over 38 weeks and a body weight below 2500 grams- has suffered growth retardation in utero. d) A combination of (b) and (c), i.e., the preterm infant who is also small for gestational age.

The characteristic that most significantly affects the survival of the preterm infant is the immature state of the respiratory system. Between 27 and 28 weeks of gestation (900-1000 grams), anatomic lung development has progressed to the extent that extrauterine survival is possible. It is only after 30 to 32 weeks of gestation that true alveoli are present. Once there is adequate lung tissue, the critical factor that decides extrauterine adaptation and survival of the preterm infant is his capabilities to produce the phospholipid-rich material, surfactant that lines the respiratory epithelium.

## **2. Metabolic and Host Defenses**

Handling of the breakdown products of hemoglobin is also a difficult task for the premature infant. The ability of the immature liver to conjugate bilirubin is reduced, the life span of the red blood cell is short, and the bilirubin load presented to the circulation via the enterohepatic route is increased. "Physiologic" jaundice is, therefore, higher in the preterm infant and persists for a longer period. Unfortunately, the immature brain has an increased susceptibility to the neurotoxic effects of high levels of unconjugated bilirubin, and kernicterus can develop in the preterm baby at a relatively low level of bilirubin.

Other problems affecting the baby include the rapid development of hypoglycemia (35 mg%), hypocalcemia and hypothermia. Newborns have a poorly developed gluconeogenesis system, and depends on glycolysis from liver glycogen stores (depleted 2-3 hrs after birth) and enteral nutrition. Immature infants can develop hyperglycemia from reduced insulin response to glucose causing intraventricular hemorrhage and glycosuria. The preterm and surgical neonate is more prone to hypocalcemia due to reduced stores, renal immaturity, and relative hypoparathyroidism (high fetal calcium levels). Symptoms are jitteriness and seizures with increase muscle tone. Calcium maintenance is 50 mg/kg/day.

Human beings are homeothermic organisms because of thermoregulation. This equilibrium is maintained by a delicate balance between heat produced and heat lost. Heat production mechanisms are: voluntary muscle activity increasing metabolic demands, involuntary muscle activity (shivering) and non-shivering (metabolizing brown fat). Heat loss occurs from heat flow from center of the body to the surface and from the surface to the environment by evaporation, conduction, convection and radiation. There is an association between hypothermia and mortality in the NICU's. The surgical neonate is prone to hypothermia. Infants produce heat by increasing metabolic activity and using brown fat. Below the 35°C the newborn experiences lassitude, depressed respiration, bradycardia, metabolic acidosis, hypoglycemia, hyperkalemia, elevated BUN and oliguria (neonatal cold injury syndrome). Factors that precipitate further these problems are: prematurity, prolonged surgery, and eviscerated bowel (gastroschisis). Practical considerations to maintain temperature control are the use of humidified and heated inhalant gases during anesthesia, and during all NICU procedures use radiant heater with skin thermistor-activated servo-control mechanism.

The newborn's host defenses against infection are generally sufficient to meet the challenge of most moderate bacterial insults, but may not be able to meet a major insult. Total complement activity is 50% of adults levels. C3, C4, C5 complex, factor B, and properdin concentration are also low in comparison to the adult. IgM, since it does not pass the placenta, is absent.

## **3- Surgical Response of Newborns**

The endocrine and metabolic response to surgical stress in newborns (NB) is characterized by catabolic metabolism. An initial elevation in catecholamines, cortisol and endorphins upon stimulation by noxious stimuli occurs; a defense mechanism of the organism to mobilize stored energy reserves, form new ones and start cellular catabolism. Cortisol circadian responsiveness during the first week of life is diminished, due to immaturity of the adrenal gland. Cortisol is responsible for protein breakdown, release of gluconeogenic amino acids from muscle, and fat lipolysis with release of fatty acids. Glucagon secretion is increased. Plasma insulin increase is a reflex to the hyperglycemic effect, although a resistance to its anabolic function is present. During surgical stress NB release glucose, fatty acids, ketone bodies, and amino acids; necessary to meet body energy needs in time of increase metabolic demands. Early postoperative parenteral nutrition can result in significant rate of weight gain due to solid tissue and water accumulation. Factors correlating with a prolonged catabolic response during surgery are: the degree of neuroendocrinological maturation, duration of operation, amount of blood loss, type of surgical procedure, extent of surgical trauma, and associated conditions (hypothermia, prematurity, etc.). They could be detrimental due to the NB limited reserves of nutrients, the high metabolic demands imposed by growth, organ maturation and adaptation after birth. Anesthetics such as halothane and fentanyl can suppress such response in NB.

## **II. HEAD AND NECK LESIONS**

### **A. Cervical Lymphadenopathy**

An enlarged lymph node is the most common neck mass in children. Most are anterior to the sternocleidomastoid muscle. Infection is the usual cause of enlargement; viral etiology and persist for months. Acute suppurative submandibular adenitis occur in early childhood (6 mo-3 yrs), is preceded by pharyngitis or URI, the child develops erythema, swelling and cellulitis, and management is antibiotics and drainage. Chronic adenitis: persistent node (> 3 wk., tonsillar), solitary, non-tender, mobile and soft. Generally no tx if < 1 cm, for nodes above 2 cm sizes with rapid growth, clustered, hard or matted do biopsy.

Other causes are: (1) Mycobacterial adenitis- atypical (MAIS complex), swollen, non-tender, non-inflamed, positive skin test, excision is curative, chemotx is of no value. (2) Cat-Scratch adenitis- caused by *A. Felis*, transmitted by kittens, positive complement fixation test, minimally tender, fluctuant regional nodes, spontaneous resolution. (3) Hodgkin's disease mostly teenager and young adults, continuing growth, non-tender node, associated to weight loss, biopsy is diagnostic.

### **B. Congenital Torticollis**

Congenital muscular torticollis is a disorder characterize by shortening of the cervical muscles, most commonly the sternocleidomastoid (SCM) muscle, and tilting of the head to the opposite side. This is the result of endomysial fibrosis of the SCM muscle. There is a relationship between birth position and the side affected by the contracture. Congenital torticollis causes: plagiocephaly (a craniofacial deformity), fascial asymmetry (hemihypoplasia), scoliosis and atrophy of the ipsilateral trapezius muscle if not corrected. Torticollis can develop at any age, although is more common during the first six months of life. The SCM muscle can be a fibrous mass, or a palpable tumor 1-3 cm in diameter within the substance of the muscle is identified by two to three weeks of age. Management is conservative in most cases using early physiotherapy exercises' a mean duration of three months to achieve full passive neck range of motion. The severity of restriction of motion is the strongest predictor of treatment duration. Those children with failed medical therapy or the development of fascial hemihypoplasia should undergo surgical transection of the SCM muscle.

### **C. Thyroglossal Duct Cysts**

Thyroglossal duct cyst (TDC) is the most common congenital anterior midline neck mass usually (2/3 of cases) presenting before the second decade of life. Symptoms appear at an average age of four with the sudden appearance of a cystic mass at the angle of neck level moving with tongue protrusion and swallowing. Males are more commonly affected than females. TDC is an embryologic anomaly arising from epithelial remnant left after descent of the developing thyroid from the foramen cecum. The lining is cuboidal, columnar or pseudostratified epithelium. TDC is associated to discomfort, infection and a slight probability of malignancy. A legally protective requirement is to document that the mass is not ectopic thyroid gland. Diagnosis is physical. Sonograms will show a cyst between 0.4 and 4 cm in diameter, with variable sonographic appearance and no correlation with pathological findings of infection or inflammation. Once infected surgical excision is more difficult and recurrence will increase. Management is Sistrunk's operation: Excision of cyst with resection of duct along with the central portion of hyoid bone (a minimum of 10-15 mm of hyoid bone should be removed) and some muscle surrounding the proximal ductules (the length of single duct above the hyoid bone spreads into many ductuli as it approach the foramen cecum). Extensive dissection can cause pharyngodynia. The greatest opportunity for cure is surgery at initial non-inflamed presentation. Inadequate excision is a risk factor for further recurrence.

### **D. Branchial Cleft Fistulas**

Branchial cleft fistulas (BCF) originate from the 1st to 3rd branchial apparatus during embryogenesis of the head and neck. Anomalies of the 2nd branchial cleft are by far the most commonly found. They can be a cyst, a sinus tract or fistulas. Fistulas (or sinus tract if they end blindly) display themselves as small cutaneous opening along the anterior lower third border of the sternocleidomastoid muscle, communicates proximally with the tonsillar fossae, and can drain saliva or a mucoid secretion. Management consists of excision since inefficient drainage may lead to infection. I have found that dissection along the tract (up to the tonsillar fossa!) can be safely and easily accomplished after probing the tract with a small guide wire in-place. This will prevent injury to nerves, vessels and accomplish a pleasantly smaller scar. Occasionally a second step-ladder incision in the neck will be required. 1st BCF are uncommon, located at the angle of the mandible, and communicating with the external auditory canal. They have a close association with the fascial nerve. 3rd BCF are very rare, run into the piriform sinus and may be a cause of acute thyroiditis or recurrent neck infections.

### **E. Cystic Hygroma**

Cystic hygroma (CH) is an uncommon congenital lesion of the lymphatic system appearing as a

multilocular fluid filled cavity most commonly in the back neck region, occasionally associated with extensive involvement of airway or vital structures. The etiology is intrauterine failure of lymphatics to communicate with the venous system. Prenatal diagnosis can be done during the first trimester of pregnancy as a huge neck tumor. Differential diagnosis includes teratomas, encephalocele, hemangiomas, etc. There is a strong correlation between prenatal dx and Turner's syndrome (> 50%), structural defects (Noonan's syndrome) and chromosomal anomalies (13, 18, 21). Early diagnosis (< 30 wk gestation) is commonly associated to those anomalies, non-immune hydrops and dismal outcome (fetal death). Spontaneous regression is less likely but can explain webbed neck of Turner and Noonan's children. Prenatal dx should be followed by cytogenetic analysis: chorionic villous sampling, amniocentesis, or nuchal fluid cell obtained from the CH itself to determine fetal karyotype and provide counseling of pregnancy. Late diagnosis (>30 wks) should be delivered in tertiary center prepare to deal with dystocia and postnatal dyspnea of newborn. The airway should be secured before cord clamping in huge lesions. Intracystic injection of OK432 (lyophilized product of *Streptococcus pyogenes*) caused cystic (hygromas) lymphangiomas to become inflamed and led to subsequent cure of the lesion without side effects.

#### **F. Cat Scratch Disease**

Cat Scratch Disease (CSD) is a self-limited condition transmitted by a *Bartonella* species (*Rochalimaea henselae*) present in unaffected kitten paws. Following inoculation by a scratch and one to two weeks of incubation period, malaise, fever, headache, anorexia and swelling of the regional lymph nodes follow. The adenopathy generally develops in the upper extremity (epitrochlear, axilla) or head/neck areas, is minimally tender and can develop fluctuation. Median age is 14 years with highest attack rate in children less than ten years of age. The diagnosis relies on the presence of symptoms, signs, physical exam (characteristic papule at the site of the scratch), history of exposure to a cat, and a positive immunofluorescent assay for *Bartonella* antibodies. Most patients with clinically diagnosed CSD developed an immunologic response to *Bartonella* species. Conservative symptomatic management is recommended for most children since the node will eventually disappear spontaneously. In other cases' aspiration of fluctuant nodes is alleviating. Antibiotics are recommended during severe cases. Overall prognosis is good.

### **III. OBSTRUCTIVE PROBLEMS**

#### **LOGICAL APPROACH TO NEONATAL INTESTINAL OBSTRUCTION**

Prepared by: **Jordan J. Weitzman, MD ©**  
Los Angeles, California, USA

##### Signs and Symptoms

1. Bilious vomiting is always abnormal.
2. Abdominal distention (scaphoid abdomen possible).
3. Delayed, scanty or no passage of meconium.
4. Polyhydramnios in mother.
5. Down's syndrome
6. Family history
  - a. Hirschsprung's disease
  - b. Diabetic mother
  - c. Jejunal atresia

##### Work-up (Logical approach)

1. While the infant is being studied, it must be kept in mind that the problem may be "non-surgical".
  - a. Sepsis of the newborn with associated ileus is the most important cause of non-surgical bilious vomiting and abdominal distention.
  - b. Intracranial lesions
    - i. Hydrocephalus
    - ii. Subdural hemorrhage
  - c. Renal disease associated with uremia.
    - i. Renal agenesis

- ii. Polycystic disease
- iii. Other urinary tract anomalies which may be associated with severe hydronephrosis.

2. Plain roentgenograms of the abdomen.

- a. Diagnostic in complete high intestinal obstruction-no gas in distal small bowel.
  - i. Double bubble in duodenal obstruction.
  - ii. Few gas filled loops beyond duodenum indicates jejunal atresia.
- b. Many gas filled loops (requires 24 hours) indicates some form of low intestinal obstruction.
  - i. Ileal atresia
  - ii. Meconium ileus(an unfortunate misnomer)-obstruction of the distal small intestine by thick undigested meconium.
  - iii. Meconium plug syndrome-obstruction of colon by a plug of meconium.
  - iv. Small left colon syndrome.
  - v. Hirschsprung's disease-congenital aganglionosis of colon starting with the rectum.
  - vi. Colonic atresia.
- c. May be nonspecific in instances of malrotation of the intestines. This diagnosis must always be considered in neonates with unexplained bilious vomiting.
- d. Calcifications-at some time during fetal life meconium was (is) present in the abdomen.

3. Contrast enema will differentiate the various types of low intestinal obstruction.

- a. Microcolon-complete obstruction of the small bowel.
- b. Meconium plug syndrome-colon dilated proximal to an intraluminal mass.
- c. Hirschsprung's disease-although it may appear to be diagnostic, not reliable in the newborn.
- d. Small left colon syndrome-colon dilated to the splenic flexure, then becomes narrow.

4. Upper G.I.- the procedure of choice in diagnosing malrotation of the intestines. In the past a contrast enema was thought to be the diagnostic test of choice in instances of malrotation but the cecum and ascending colon can be in normal position in an infant or child with malrotation of the intestines.

5. Rectal biopsy- a pathologist competent in reading the slides is essential and should not be taken for granted.

- a. Suction biopsy of the rectal mucosa and submucosa- best screening procedure to rule out Hirschsprung's disease (ganglion cells are present in the submucosa), and is diagnostic in experienced hands.
- b. Full thickness biopsy of the rectal wall may be necessary if the suction biopsy is non-diagnostic or if the pathologist is unwilling or unable to make the diagnosis of aganglionosis on a suction biopsy specimen. This procedure is difficult in the small infant and has been replaced by the suction biopsy in most centers.
- c. All newborns who have delayed passage of meconium associated with a suspicious contrast enema should have a suction biopsy of the rectal mucosa and submucosa. With this technique, Hirschsprung's disease will be diagnosed early before it is complicated with enterocolitis. If delayed passage of meconium is "cured" by rectal stimulation(suppository, thermometer, or finger), it must be kept in mind that the diagnosis of Hirschsprung's disease is still a possibility. Whether or not a suction biopsy of the rectum is done before the infant goes home depends on the clinical setting but the safe course of action is to do the rectal biopsy before discharge. Parents may not call before the infant gets into trouble with enterocolitis.
- d. Suction biopsy of the rectum is probably indicated in all cases of so called meconium plug syndrome or small left colon syndrome. If the suction biopsy is not done, the infant must be observed for recurrent gastrointestinal symptoms. A breast-fed infant who has Hirschsprung's disease can "get by" for a prolonged period of time.

6. Concluding comments:

The newborn suspected of having intestinal obstruction should be studied in a logical step by step manner. It is important that it be definitely established that the infant has a surgical problem before surgery is performed. This is usually not difficult in instances of complete high small bowel obstruction or when plain films of the abdomen show calcification and/or a distal small bowel obstruction with the contrast enema showing a microcolon or a definite malrotation of the colon (cecum in upper mid-abdomen or left upper quadrant).

When plain films are suggestive of a high small bowel obstruction but there is gas in the distal small

bowel, an upper GI rather than a contrast enema should be performed. It is critically important that the diagnosis of malrotation of the intestines be always considered and ruled out in a neonate with bilious vomiting. Prompt recognition and treatment of malrotation of the intestines which is often associated with a midgut volvulus avoids the dire consequences of the problems associated with a massive small bowel resection.

Mistakes are frequently made when the contrast enema is interpreted as normal, meconium plug syndrome, small left colon syndrome or Hirschsprung's disease. In all of these clinical situations, a suction biopsy of the rectum is an excellent screening procedure. If ganglion cells are present, Hirschsprung's disease is ruled out and the infant probably has a non-surgical diagnosis. If ganglion cells are absent, the next step depends on the clinical picture and setting. If the pathologist is experienced and confident of the interpretation, the diagnosis of Hirschsprung's disease can be made with confidence. If there is any doubt about the absence of ganglion cells in the suction biopsy, a full thickness biopsy of the rectum (a difficult technical procedure requiring a general anesthetic) can be done to settle the issue. If Hirschsprung's disease is believed to be the problem, it must be diagnosed histologically before the infant is operated upon because at the time of surgery the site of obstruction may not be apparent and the abdomen may be closed because no obvious site of obstruction is found.

Hypothyroidism in the first two to three months of life can mimic Hirschsprung's disease in all aspects except for a normal rectal biopsy.

Another important point to remember is that duodenal atresia is a different disease from jejunal or ileal atresia in terms of their cause. Jejunal and ileal atresia occur as a result of a vascular accident in the small bowel mesentery during fetal life. Consequently, there is a relatively low incidence of other congenital anomalies except for cystic fibrosis. Duodenal atresia is a different disease in that there is a very high incidence of associated anomalies-- (Down's syndrome, imperforate anus, renal anomalies, congenital heart disease, etc.).

*Malrotation of the intestines and Hirschsprung's disease must be ruled out before a newborn with unexplained bilious vomiting and/or abdominal distention is sent home. It can be unsafe to rely on parents to observe their infant for problems resulting from the above conditions. If diagnosed late, malrotation of the intestines or Hirschsprung's disease can become life threatening or result in life long problems.*

## **A. Esophageal Atresia a/o Tracheo-esophageal fistula**

### **1. Embryology**

The trachea and esophagus initially begin as a ventral diverticulum of the foregut during the third intrauterine week of life. A proliferation of endodermal cells appears on the lateral aspect of this growing diverticulum. These cell masses will divide the foregut into trachea and esophageal tubes. Whether interruption of this normal event leads to tracheo-esophageal anomalies, or during tracheal growth atresia of the esophagus results because of fistulous fixation of the esophagus to the trachea remnant to be proven.

### **2. Classification**

EA with distal TEF (87%) - the most common anomaly, the NG tube coiled at T4-5 level and gas will be seen in the KUB. EA without TEF (8%) - pure esophageal atresia, NG coiled at T4-5 level with airless abdomen. TEF without atresia (4%) - pure tracheo-esophageal fistula. EA with proximal TEF (<1%). EA with proximal and distal TEF (<1%)

Congenital isolated tracheo-esophageal fistula (TEF) occurs as 4-6% of the disorders of the esophagus bringing problems during early diagnosis and management. More than H-type is N-type, due to the obliquity of the fistula from trachea (carina or main bronchi) to esophageal side (see the figure) anatomically at the level of the neck root (C7-T1). Pressure changes between both structure can cause entrance of air into the esophagus, or esophageal content into the trachea. Thus, the clinical manifestation that we must be aware for early diagnosis are: cyanosis, coughing and choking with feedings, recurrent chest infections, persistent gastrointestinal distension with air, and hypersalivation. Diagnosis is confirmed with a well-done esophagogram, or video-esophagogram (high success rates, establish level of the TEF). Barium in the trachea could be caused by aspiration during the procedure. Upon radiologic doubt bronchoscopy should be the next diagnostic step. Any delay in surgery is generally due to delay in diagnosis rather than delay in presentation. Management consists of surgical closure of the TEF through a right cervical approach. Hint: a small guide-wire threaded through the fistula during bronchoscopy may be of some help. Working in the tracheo-esophageal groove can cause injury to the recurrent laryngeal nerve with vocal cord paralysis. Recurrence after closure is rare.

### **3. Diagnostic characteristics:**



The incidence is one in every 2500 live births. We see between 8-10 per year at the University Pediatric Hospital. The mother might show polyhydramnios since the fetus is unable to swallow amniotic fluid. (May be responsible for early delivery). Polyhydramnios is most commonly seen in pure esophageal atresia type. Choking, coughing and regurgitation with first feed. Excessive salivation, cyanosis with feedings. Inability to pass feeding tube into the stomach. Contrast studies (UGIS, esophagogram) are rarely needed, and of potential disaster (aspiration of contrast material). Abdominal films should be obtained to rule out the occurrence of associated gastrointestinal anomalies. Isolated TEF is more difficult to diagnose and may require repeated lateral esophagograms, bronchoscopy and esophagoscopy.

#### **4. Management**

Correct dehydration, acid-base disturbances, respiratory distress and decompress proximal esophageal pouch (Replogle tube). Evaluate for associated conditions such as VACTERL association (3 or more):

- Vertebral anomalies i.e. hemivertebrae, spina bifida
- Anal malformations i.e. imperforate anus
- Cardiac malformations i.e. VSD, ASD, Tetralogy Fallot
- Tracheo-Esophageal fistula (must be one of the associated conditions)
- Renal deformities i.e. absent kidney, hypospadias, etc.
- Limb dysplasia

Early surgical repair (transpleural or extrapleural) for those babies with no evidence of pneumonia, adequate weight (>1200 gm) and no significant associated anomalies. Babies with Chest-X-Ray positive findings, but adequate ABG's can also be primarily repaired. Delayed repair (gastrostomy first) for all other patients. Surgical repair consist of a 4th intercostal space right muscle-sparing thoracotomy (side of thoracotomy is contralateral to side of aortic arch of patient), closure of tracheo-esophageal fistula and primary esophago-esophagostomy. Esophagogram is done 7-10 days after repair.

Complications after surgery are: Anastomotic leak, anastomotic stricture, gastroesophageal reflux, tracheomalacia, and recurrent TEF. The three most common anastomotic complications are in order of frequency: stricture, leakage and recurrent TEF. Recurrent TEF after surgical repair for esophageal atresia occurs in approximately 3-15% of cases. Tension on the anastomoses followed by leakage may lead to local inflammation with breakage of both suture lines enhancing the chance of recurrent TEF. Once established, the fistula allows saliva and food into the trachea, hence clinical suspicion of this diagnosis arises with recurrent respiratory symptoms associated with feedings after repair of esophageal atresia. Diagnosis is confirmed with cineradiography of the esophagus or bronchoscopy. A second thoracotomy is very hazardous, but has proved to be the most effective method to close the recurrent TEF. Either a pleural or pericardial flap will effectively isolate the suture line. Pericardial flap is easier to mobilize, provides sufficient tissue to use and serves as template for ingrowth of new mucosa should leakage occur. Other alternatives are endoscopic diathermy obliteration, laser coagulation, or fibrin glue deposition.

Esophageal stenosis in children can be of congenital (5%) or most commonly acquired nature (95%). Acquired stenosis is the result of repaired esophageal atresia, caustic injury, penetrating injury or reflux esophagitis. Congenital esophageal stenosis (CES) can be the result of a membranous diaphragm, segmental hypertrophy of the muscularis and submucosal layer (submucosal fibrosis), or presence of ectopic tracheobronchial rest. CES most commonly affect the middle and distal third of the esophagus and rarely cause symptoms in the neonatal period. Symptoms can be vomiting of undigested food, regurgitation, food impaction, difficulty swallowing solid and failure to thrive. CES affecting the upper third of the esophagus is very rare and usually produce respiratory symptoms such as stridor and repeated respiratory infections. Esophageal atresia is associated with one-third of cases of CES. To establish a diagnosis investigation has to include esophagogram (relatively long, smooth circumferential narrowing), esophagoscopy with biopsy, pH monitoring and in selected cases manometry. Recognition of the correct etiologic factor that caused the stricture will pave the way for adequate management. CES is managed with forceful dilatation or hydrostatic balloon dilatation, while resection with anastomosis will be needed for intractable (fibromuscular hypertrophy) cases and those harboring tracheobronchial rests. Most intractable cases are due to the presence of tracheobronchial rest.

#### **B. Achalasia**

Achalasia in children is an uncommon esophageal motor disorder distinguished by clinical, radiological and manometrics features. Incidence is estimated in 0.1 cases/year per 100,000 population under 14 years of age. Clinical presentation is characterized by progressive dysphagia, regurgitation, weight loss, chest pain and nocturnal cough. Infants exhibit failure to thrive. Diagnosis is established by barium swallow and confirmed by

manometry and motility studies. Ba swallow shows' esophageal dilatation, motility alteration and a small caliber (bird-beak) cardio-esophageal junction. Manometry reveals elevated E-G sphincter pressure, non- peristaltic esophageal contraction and failed relaxation of lower esophageal sphincter upon swallowing. Videofluoroscopy can be of help in the screening of esophageal motors disorders. Esophageal pneumatic balloon dilatation is not an effective method of treatment in children due to the high rate of recurrence of symptoms. Primary therapy is surgical (Heller's modified esophagomyotomy), and results are similar after a transabdominal or thoracic approach. Many authors favor a concomitant antireflux procedures in these patients. Nifedipine can be of help as a short management in preparation for surgery. Long-term result presents' a connection between achalasia and malignant disease of the esophagus.

## **C. Gastro-duodenal Anomalies**

### **1. Gastric Anomalies**

Congenital gastric outlet obstruction is extremely rare. It occurs either in the pyloric or antral region. Antral membranes (web or diaphragm) are thin, soft and pliable, composed of mucosa/submucosa, and located eccentric 1-3 cm proximal to pyloro-duodenal junction. They probably represent the developmental product of excess local endodermal proliferation and redundancy. The diagnosis should rely on history, contrast roentgenology studies and endoscopic findings. Symptoms are those of recurrent non-bilious vomiting and vary according to the diameter of aperture of the membrane. There is a slight male predominance with fair distribution between age groups in children. Associated conditions: pyloric stenosis, peptic ulcer and cardiac. History of polyhydramnios in the mother. Demonstration of a radiolucent line perpendicular to the long axis of the antrum is diagnostic of a web. Endoscopy corroborates the diagnosis. Management can be either surgical or non-surgical. Surgical Tx is successful in symptomatic pt. and consist of pyloroplasty with incision or excision of the membrane. Other alternative is endoscopic balloon dilatation or transection of the web. Non-obstructive webs found incidentally can be managed medically with small curd formula and antispasmodics. The presence of an abnormally dilated gastric bubble in prenatal sonography should alert the physician toward the diagnosis of congenital antro-pyloric obstruction.

### **2. Pyloric Stenosis**

Is an abnormality of the pyloric musculature (hypertrophy) causing gastric outlet obstruction in early infancy. The incidence is 3 per 1000 live births. The etiology is unknown, but pylorospasm to formula protein cause a work hypertrophy of the muscle. Diagnostic characteristics are: non-bilious projectile vomiting classically 3-6 weeks of age, palpable pyloric muscle "olive", contrast studies are not necessary when the pyloric muscle is palpated, enlarged width and length in ultrasonography.

The treatment consist in correction of hypochloremic alkalosis and state of dehydration and performing a Fredet-Ramstedt modified pyloromyotomy. Post-operative management consist of: 50% will have one to several episodes of vomiting, usually can feed and go home in 24-36 hours, initial feeds start 8-12 hours after surgery.

### **3. Duodenal Malformations**

Can be intrinsic (Atresia, Stenosis, Webs) or extrinsic (Annular pancreas, Ladd's bands). Occur distal or proximal to the ampulla of Vater. Most commonly distal to ampulla and therefore bilious vomiting is present. (Note: Bilious vomiting is surgical until proven otherwise in a baby).

"Windsock" webs have clinical importance because of their tendency to be confused with distal duodenal obstruction and because of the frequent occurrence of an anomalous biliary duct entering along their medial margin.

Embryology: The first major event in the differentiation of the duodenum, hepatobiliary tree, and pancreas occurs at about the third week in gestation, when the biliary and pancreatic buds form at the junction of the foregut and the midgut. The duodenum at this time is a solid cord of epithelium, which undergoes vacuolization followed by recanalization and restitution of the intestinal lumen over 3-4 weeks of normal development. Failure of recanalization of the second part of the duodenum results in congenital obstruction of the lumen, often in conjunction with developmental malformation of the pancreatic anlagen and the terminal part of the biliary tree. In support of this concept is the high incidence of annular pancreas observed, believed to represent a persistence of the ventral pancreatic anlage in association with intrinsic duodenal obstruction.

The diagnostic characteristics are: bilious vomiting, history of polyhydramnios in mother, KUB with classic "Double-bubble" appearance, a microcolon in barium enema study or malrotation.

Treatment consist in: (1) duodeno-duodenostomy bypass for atresias, annular pancreas, and some stenosis. (2) duodenoplasty for webs, and stenosis, and (3) lysis of Ladd's bands and Ladd's procedure for malrotation.

Associated anomalies are: Down's syndrome (20-30%), VACTERL syndrome, CNS anomalies and cardiac anomalies.

#### **D. Malrotation and Volvulus**

**Embryology:** The rotation and normal fixation of the intestinal tract takes place within the first three months of fetal life. In the earliest stages when the intestinal tract is recognizable as a continuous tube, the stomach, small intestine, and colon constitute a single tube with its blood supply arising posteriorly. The midgut portion of this tube, from the second portion of the duodenum to the mid-transverse colon, lengthens and migrates out into an extension of the abdomen, which lies at the base of the umbilical cord. Here this loop of bowel undergoes a 270-degree counterclockwise twist at its neck. In the center of the twisted loop lie the blood vessels that will become the superior mesenteric artery and vein. After rotation, the small intestine quite rapidly withdraws into the abdominal cavity, with the duodenum and the proximal jejunum going first. During this process the duodenojejunal junction goes beneath and to the left of the base of the superior mesenteric vessels. This leaves the upper intestine, including the stomach and the duodenum, encircling the superior mesenteric vessels like a horseshoe with its opening on the left side of the embryo. The small intestine then follows into the abdomen, and withdrawal of the right half of the colon takes place so that it lies to the left. At the next step, the cecum and the right colon begin to travel across the top of the superior mesenteric vessels and then down to the right lower quadrant. The colon now lies draped across the top of the superior mesenteric vessels, again like a horseshoe, with its opening placed inferiorly. The duodenojejunal loop is said to attach to the posterior abdominal wall soon after its turn, whereas the mesenteric attachments of the entire colon and of the remaining small bowel gradually adhere after they arrive in their normal positions. In malrotation the right colon can create peritoneal attachments that include and obstruct the third portion of the duodenum (Ladd's bands).

The diagnostic hallmarks are: bilious vomiting (the deadly vomit), abdominal distension and metabolic acidosis. A UGIS is more reliable than barium enema, most patients present in first month of life (neonatal), but may present at any time.

The treatment is immediate operation; volvulus often means strangulation. Needs fluid and electrolyte replacement. Ladd's procedure consist of: reduce volvulus with a counterclockwise rotation, place small bowel in right abdomen, lysed Ladd's bands, place large bowel in left abdomen, do an appendectomy. In cases of questionable non-viable bowel a second look procedure is required.

Failure to make early diagnosis and operate may lead to dead midgut with resultant short bowel syndrome.

#### **E. Intestinal Atresias**

Intestinal atresias are the product of a late intrauterine mesenteric vascular accident (blood supply was not received by a portion of bowel) as attested by Louw and Barnard in 1955. They are equally distributed from the ligament of Treitz to the ileocecal junction. Colonic atresias are very rare. There is proximal bowel dilatation, with distal (unused) micro-bowel. The diagnosis is suspected with maternal history of polyhydramnios (the higher the atresia), bilious vomiting, abdominal distension and obstipation. KUB shows "thumb-size" dilated bowel loops, and barium enema a microcolon of disuse. Louw classified them into: Type I: an intraluminal diaphragm with seromuscular continuity. Type II: cord-like segment between the bowel blinds ends. Type IIIA: atresia with complete separation of blind ends and V-shaped mesenteric defect (see figure), the most commonly found. Type IIIB: jejunal atresia with extensive mesenteric defect and distal ileum acquiring its blood supply entirely from a single ileocolic artery. The distal bowel coils itself around the vessel, giving the appearance of an "apple peel" deformity. Type IV: multiple atresias of the small intestine. After preoperative stabilization (GI decompression, electrolytes disturbances' correction, antibiotherapy, and normothermia), treatment consists of exploratory laparotomy, resection of proximal dilated intestine, and end to oblique anastomosis in distal jejuno-ileal atresias. Tapering jejunoplasty with anastomosis is preferred in proximal defects.

## **F. Meconium Ileus**

Meconium ileus is a neonatal intraluminal intestinal obstruction caused by inspissated meconium blocking the distal ileum. Occurs in 10-15% of all patients with cystic fibrosis, and 85-95% of patients with meconium ileus have cystic fibrosis. The meconium has a reduced water, abnormal high protein and mucoprotein content, the result of decreased pancreatic enzyme activity and prolonged small bowel intestinal transit time.

Meconium Ileus is classified into two types: (1) Simple meconium ileus: The distal small bowel (10-30 cm of distal ileum) is relatively small, measuring less than 2 cm in diameter and contains concretions of gray, inspissated meconium with the consistency of thick glue or putty. It is often beaklike in appearance, conforming to the shape of the contained pellets. Proximally, the mid-ileum is large, measuring up to 7 cm in diameter. It is greatly distended by a mass of extremely thick, tenacious, dark green or tarry meconium. The unused small colon (microcolon) contains a small amount of inspissated mucus or grayish meconium. (2) Complicated meconium ileus: usually occurs during the prenatal period associated to volvulus, atresias, gangrene, perforation or peritonitis. A cystic mass or atresia of the bowel may occur.

The degree of obstruction varies, may be cured in mild cases by rectal irrigations. Failure to pass meconium, abdominal distension and vomiting are seen in more severe cases. The diagnosis is suspected with findings of: multiple loops of dilated small bowel and coarse granular "soap-bubble" appearance on plain abdominal films. Some cases may show calcifications in the peritoneum (Meconium peritonitis). The Sweat Test is diagnostic of cystic fibrosis (value over 60 meq/L of sweat sodium or chloride are diagnostic). This test is not useful in infant during first weeks of life.

Therapy is either: (1) Nonoperative- should be tried first. It consist of a careful gastrografen enema after the baby is well-hydrated. Gastrografen is a hyperosmolar aqueous solution of meglumine diatrizoate containing 0.1% polysorbate-80 (tween-80, a wetting agent) and 37% iodine. Its success is due to the high osmolarity (1700 mOsm/liter) which draws fluid into the bowel and softens and loosens the meconium. (2) Surgical therapy that has included: ileostomy with irrigation, resection with anastomosis, and resection with ileostomy (Mikulicz, Bishop-Kopp, and Santulli). Post-operative management includes: 10% acetylcysteine p.o., oral feedings (Pregestimil), pancreatic enzyme replacement, and prophylactic pulmonary therapy. Long-term prognosis depends on the degree of severity and progression of cystic fibrosis pulmonary disease.

## **G. Hirschsprung's Disease**

Hirschsprung's is the congenital absence of parasympathetic innervation of the distal intestine. The colon proximal to the aganglionic segment, in an effort to overcome the partial obstruction, becomes distended and its wall markedly thickened because of muscle hypertrophy. Occurs 1 in 1000-1500 live births with a 4:1 male predominance. 96% are TAGA. 4% prematures.

The parasympathetic ganglion cell network located between the circular and longitudinal muscle layers is referred to as Auerbach's plexus, whereas Meissner's plexus is the submucosal layer of ganglion cells just beneath the muscularis mucosa. In Hirschsprung's disease, ganglion cells are absent from all layers. That aganglionic segment usually involves the terminal intestine, i.e. the rectum or rectosigmoid. The aganglionic segment may, however, include the entire large bowel and even small bowel.

Hirschsprung's disease (HD) is characterized by lack of enteric ganglion cells, hyperplasia of abnormal nerve fibers and a non-propulsive, non-relaxing segment of bowel. Classically the etiology is attributed to a failure of cranio-caudal migration of parasympathetic neural crest cells to the distal bowel. A plausible explanation for the failure of relaxation of the bowel involved is a deficiency of enteric inhibitory nerves that mediates the relaxation phase of peristalsis. These nerves are intrinsic to the gut and are classified as non-adrenergic and non-cholinergic. Nitric oxide (NO) has recently been implicated as the neurotransmitter which mediates the relaxation of smooth muscle of the GI tract in HD. Its absence in aganglionic bowel might account for the failure of relaxation during peristalsis. Besides, adhesions molecules (absent in aganglionic bowel) during early embryogenesis might restrict the neuro-ectodermal origin involved in the initial contact between nerves and muscle cell (synaptogenesis) suggesting that developmental anomaly of innervated muscle and absent NO causes the spasticity characteristic of HD.

Symptoms usually begin at birth, frequently with delayed passage of meconium. Any newborn who fails

to pass meconium in the first 24-48 hours of life should be evaluated for possible Hirschsprung's disease. In some infants, the presentation is that of complete intestinal obstruction. Others have relatively few symptoms until several weeks of age, when the classic symptom of constipation has its onset. Diarrhea is not uncommon but differs from the usual infantile diarrhea in that it is associated with abdominal distension. Occasionally the patient will go many years with mild constipation and diagnosis will be delayed.

The diagnosis is first suspected based on history and physical examinations (characteristically there is no stool in rectum and abdominal distension is painless). Initial evaluation includes an unprepared barium enema (the first enema should be a barium enema!). The aganglionic rectum appears of normal caliber or spastic, there is a transition zone and then dilated colon proximal to the aganglionic segment. 24-hrs delayed films shows poor emptying with barium throughout the colon, as opposed to the child with psychogenic stool holding in whom the barium generally collects in the distal rectosigmoid. Rectal suction biopsy is then performed. This can be done without anesthesia and the submucosal plexus is examined for ganglion cells. With experience, a good pathologist (should be an expert!), can identify the presence or absence of ganglion cells in this specimen without a full thickness biopsy. Difficulty in interpreting the specimen or not enough to include several slides of submucosa would require a full-thickness biopsy for definitive diagnosis generally done under general anesthesia. Some centers employ manometry, histochemical studies or special stains for diagnosis. These special studies are only as good as the person performing them and interpreting the results.

The initial treatment requires performing a "leveling" colostomy in the most distal colon with ganglion cells present. This requires exploration with multiple seromuscular biopsies of the colon wall to determine the exact extend of the aganglionosis. The colostomy is placed above the transition zone. Placement of the colostomy in an area of aganglionosis will lead to persistent obstruction. Once the child has reached an adequate size and age (6-12 months; 20 pounds or more), a formal pull-through procedure is done. Some of this are: Swenson, Duhamel and Soave procedures. Current preference is for Soave procedure (modified endorectal pull-through) and consist of resection of the majority of aganglionic bowel except for the most distal rectum, the mucosa and submucosa of this rectum is excised and the normally innervated proximal bowel is pulled through the seromuscular coat of retained rectum and suture immediately above the dentate line. Recently a laparoscopic pull-through procedure avoiding the colostomy is being used in early life with promising results.

Intestinal Neuronal Dysplasia (IND) is a colonic motility disorder first described in 1971 by Meier-Ruge associated to characteristic histochemical changes of the bowel wall (hyperplasia of submucous & myenteric plexus with giant ganglia formation, isolated ganglion cells in lamina propria and muscularis mucosa, elevation of acetylcholinesterase in parasympathetic fiber of lamina propria and circular muscle, and myenteric plexus sympathetic hypoplastic innervation), also known as hyperganglionosis associated to elevated acetylcholinesterase parasympathetic staining. The condition can occur in an isolated form (either localized to colon or disseminated throughout the bowel), or associated to other diseases such as Hirschsprung's (HD), neurofibromatosis, MEN type IIB, and anorectal malformations. It is estimated that 20-75% of HD cases have IND changes proximal to the aganglionic segment. Clinically two different types of isolated IND have been described: Type A shows symptoms of abdominal distension, enterocolitis, bloody stools, intestinal spasticity in imaging studies (Ba Enema) since birth, is less common and associated with hypoplasia of sympathetic nerves. Type B is more frequent, symptoms are indistinguishable from that of HD, with chronic constipation, megacolon, and repeated episodes of bowel obstruction. Management depends on clinical situation; conservative for minor symptoms until neuronal maturation occurs around the 4th year of life, colostomy and resectional therapy for life threatening situations.

#### **H. Imperforate Anus**

Embryology- Between 4-6 weeks, the cloaca becomes the common depository for the developing urinary, genital and rectal systems. The cloaca is quite promptly divided into an anterior urogenital sinus and a posterior intestinal canal by the urorectal septum. Two lateral folds of cloacal tissue join the urorectal septum to complete the separation of the urinary and rectal tracts.

Diagnostic evaluation include physical exam for clues such as: meconium "pearls", bucket handle anus, a fistula or meconium at meatus (urethra). Radiography could be of help initially by using the

Wangensteen-Rice "upside-down" film with opaque marker, sacral films, urogram (IVP and cystourethrogram). Through the distal stoma of the initial colostomy a contrast study (colostogram) can be done to further delineate the recto-urethral fistula associated.

Associated Anomalies: (1) Gastrointestinal- 10-20% of patients with imperforate anus have another GI lesion such as esophageal atresia, intestinal atresia or malrotation. (2) Cardiovascular- approximately 7% have associated CV lesions. (3) Skeletal- approximately 6% have skeletal lesions such as spina bifida or agenesis of the sacrum. (4) Genitourinary- 25-40% of patients will have associated genitourinary anomalies. The incidence is higher with supralevator lesions than with infralevator lesions.

The repair has been revolutionized by Peña approach (Posterior sagittal anorectoplasty procedure). The most important decision in the initial management of Imperforate Anus (IA) male patient during the neonatal period is whether the baby needs a colostomy and/or another kind of urinary diversion procedure to prevent sepsis or metabolic derangements. Male patients will benefit from perineal inspection to check for the presence of a fistula (wait 16-24 hours of life before deciding). During this time start antibiotherapy, decompress the GI tract, do a urinalysis to check for meconium cells, and an ultrasound of abdomen to identify urological associated anomalies. Perineal signs in low malformations that will NOT need a colostomy are: meconium in perineum, bucket-handle defect, anal membrane and anal stenosis. These infants can be managed with a perineal anoplasty during the neonatal period with an excellent prognosis. Meconium in urine shows the pt has a fistula between the rectum and the urinary tract. Flat "bottom" or perineum (lack of intergluteal fold), and absence of anal dimple indicates poor muscles and a rather high malformation needing a colostomy. Patients with no clinical signs at 24 hours of birth will need an invertogram or cross-table lateral film in prone position to decide rectal pouch position. Bowel > 1 cm from skin level will need a colostomy, and bowel < 1 cm from skin can be approached perineally. Those cases with high defect are initially managed with a totally diverting colostomy. Diverting the fecal stream reduces the chances of genito-urinary tract contamination and future damage.

The most frequent defect in female patient with imperforate anus (IA) is vestibular fistula, followed by vaginal fistulas. In more than 90% of female cases perineal inspection will confirm the diagnosis. These infants require a colostomy before final corrective surgery. The colostomy can be done electively before discharge from the nursery while the GI tract is decompressed by dilatation of the fistulous tract. A single orifice is diagnostic of a persistent cloacal defect usually accompanied with a small-looking genitalia. Cloacas are associated to distended vaginas (hydrocolpos) and urologic malformations. This makes a sonogram of abdomen very important in the initial management of these babies for screening of obstructive uropathy (hydronephrosis and hydroureter). Hydrocolpos can cause compressive obstruction of the bladder trigone and interfere with ureteral drainage. Failure to gain weight and frequent episodes of urinary tract infections show a poorly drained urologic system. A colostomy in cloacas is indicated. 10% of babies will not pass meconium and will develop progressive abdominal distension. Radiological evaluation will be of help along with a diverting colostomy in these cases. Perineal fistulas can be managed with cutback without colostomy during the neonatal period.

### **I. Duplications**

Duplications of the gastrointestinal tract are considered uncommon congenital anomalies usually diagnosed or unexpectedly encountered intraoperatively during the first two years of life. The duplicated bowel can occur anywhere in the GI tract, is attached to the mesenteric border of the native bowel, shares a common wall and blood supply, coated with smooth muscle, and the epithelial lining is GI mucosa. May contain ectopic gastric or pancreatic tissue. Most are saccular, other tubular. Theories on their origin (split notochord syndrome, twinning, faulty solid-stage recanalization) do not explain all cases of duplicated bowel. Three-fourth are found in the abdomen (most commonly the ileum and jejunum), 20% in the thorax, the rest thoraco-abdominal or cervical. Symptoms vary according to the size and location of the duplication. Clinical manifestations can range from intestinal obstruction, abdominal pain, GI bleeding, ulceration, or mediastinal compression. Ultrasound confirms the cystic nature of the lesion (muscular rim sign) and CT the relationship to surrounding structures. Management consists of surgical excision avoiding massive loss of normal bowel and removing all bowel suspected of harboring ectopic gastric mucosa.

### **J. Intussusception**

Although intussusception can occur at any age, the greatest incidence occurs in infants between 4-10 months of age. Over half of the cases are in the first year of life. Frequently occurs after a recent upper respiratory infection, by Adenovirus type 3 that causes a reactive lymphoid hyperplasia that act as lead point (of Peyer's patch).

A definite lead point is identified in about 5% of patients. These include: Meckel's diverticulum, polyps, Henoch's Schönlein purpura, hematoma, lymphoma, foreign bodies, and duplications. Most children have no lead point and it is felt that enlarged mesenteric nodes or swollen Peyer's patches may be the cause. The baby has intermittent periods of severe discomfort with screaming, stiffening and drawing up of the legs, followed by periods of rest. Vomiting may occur and bloody, mucoid (currant jelly) stool may be passed. The baby may become dehydrated and appear acutely ill. Frequently, lethargy may be an early sign. The diagnosis is made by barium enema, and hydrostatic reduction of the intussusception with barium is successful in approximately 50% of cases. To be successful, the barium must reflux into the terminal ileum. The surgeon should be notified before an attempt at barium reduction, and should be present at the time of study. Recently the use of gas enema reduction has been successful in patients with: (1) symptoms less than 12 hours, (2) no rectal bleeding, (3) absence of small bowel obstruction, and (4) normally hydrated. Ultrasonography can be used as a rapid sensitive screening procedure in the initial diagnosis of intussusception. Previous adverse clinical features that precluded barium reduction can be replaced during gas reduction. Predictors of failure of reduction are: (1) ileocolic intussusception, (2) long duration of symptoms, (3) rectal bleeding, and (4) failed reduction at another institution. Air reduction (pneumocolon) is a very effective alternative method since it brings less radiation (shorter fluoroscopy time), less costs and less morbidity in cases of perforations.

Failure of hydrostatic reduction requires urgent operation through a right lower quadrant horizontal incision. The intussusception is reduced by pushing on the distal bowel like a tube of toothpaste rather than pulling the proximal bowel. Most cases are ileo-colic intussusception, and a few are jejuno-jejunal or ileo-ileal intussusception.

### **K. Appendicitis**

Included in this group because is caused by obstruction of the appendiceal lumen, most commonly by fecaliths. When obstruction occurs, secretions from the appendix accumulates and acutely distends the lumen. The pressure eventually produces arterial obstruction leading to infarction. Bacteria initially invade the mucosa and produce intramural infection. Other causes include pinworm infestation, carcinoids and lymphoid hyperplasia.

Initially periumbilical pain secondary to distension of the lumen of the appendix occurs. Pain impulses from the wall of the distended appendix are carried by visceral afferent sympathetic fibers through the celiac ganglion to T10 and then referred to the umbilical area in the tenth dermatome. Later the pain shifts to the right lower quadrant of the abdomen, where it localizes. The shift in location is an important diagnostic sign and indicates the formation of irritating exudate around the inflamed appendix that stimulates the pain receptors of the peritoneum locally. Anorexia, nausea and vomiting follow the onset of abdominal pain.

Physical findings include an obviously ill-appearing child who usually will walk slowly and bent over. Motion, heel tap, or bouncing on the heels will elicit pain in the right lower quadrant. Point tenderness in the right lower quadrant (or the persistence of right lower quadrant pain) is the most reliable physical finding. There is usually rebound and referred pain to the right lower quadrant, indicating peritoneal inflammation. Fever is usually present. Laboratory findings are an elevated white blood cell count in most instances. Very high WBC's > 18,000 may indicate perforation. Urinalyses is generally clear, but occasionally RBC's or WBC's may be associated with the inflamed appendix adjacent to the bladder or ureter. Radiographic findings may include ileus, appendicolith (pathognomonic finding), splinting, abdominal wall edema, and only very rarely, free air.

Initial treatment is rehydration to establish adequate urinary output. Any evidence of possible perforation should mandate the use of appropriate antibiotics. Once adequate then surgical intervention proceeds quickly. Most patients are approached through a right lower quadrant horizontal muscle splitting incision. Removal of the appendix, irrigation and, when localized abscesses are identified, institution of drainage.

Appendicitis is usually diagnosed from signs, symptoms, results of simple laboratory tests a/o simple

abdominal films. After simple abdominal films an appendicolith (coprolith, fecalith, retained barium or foreign body) is sometimes found in the symptomatic child with right lower quadrant pain or less commonly in an asymptomatic situation. In the child WITH SYMPTOMS of low abdominal pain this finding should be followed by appendectomy. Appendiceal fecaliths and calculi play a role in the pathogenesis of appendicitis and are associated with perforation and gangrene. In the ASYMPTOMATIC situation a prophylactic appendectomy is NOT justified when an appendicolith, retained barium or another foreign body within the lumen of the appendix is identified. A normal appendix will expel the appendicolith or barium in a variable period. The parents should be informed that appendicitis may develop and that the child should seek a physician if abdominal symptoms develop. A note should appear in the record explaining this conversation.

A word on incidental appendectomy: Removing a normal appendix incidentally during a surgical procedure done for reasons other than abdominal pain is associated with a small but definite increase in adverse postoperative outcome. In this respect incidental appendectomy has been found to increase the incidence of postoperative septic complications (wound infection). It is neither cost-effective as an estimated 36 incidental procedures would be needed to prevent one case of appendicitis. As any procedure it increases adhesion formation from surgical manipulation in the right lower quadrant fossa. In potentially contaminated primary procedures the addition of incidental appendectomy does not increase operative morbidity or mortality. Incidental appendectomy is indicated in procedures where a potential diagnostic pitfall can occur such as Ladd procedure for malrotation, diagnostic laparoscopy for right quadrant pain and surgically reduced ileo-colic intussusception.

#### **L. Chronic Intestinal Pseudo-Obstruction**

Chronic Intestinal Pseudo-Obstruction is a rare disorder of intestinal motility in infants and children characterized by recurrent attacks of abdominal pain, distension, vomiting, constipation and weight loss in the absence of obvious mechanical lesions. The disease can be familial or sporadic. Suggested etiology is degeneration of enteric nervous or smooth muscle cells. The diagnosis is based on history, physical exam, radiographies and motility studies. X-Ray hallmarks are: absent strictures, absent, decreased or disorganized intestinal motility, and dilated small/large bowel loops. Associated conditions identified in 10-30% of patients are bladder dysfunction (megacystis) and neurological problems. Histologic pattern portrayed: myenteric plexus hyperplasia, glial cell hyperplasia, and small ganglion cells (hypoganglionosis). Management is primary supportive: intestinal decompression (NG), long-term TPN and antibiotic prophylaxis. Motility agents are unsuccessful. Venting gastrostomy with home parenteral nutrition has shortened the high hospitalization rate associated to this disease process. A similar condition can be seen in early fed prematures due to immaturity of intestinal motility.

#### **M. Bezoars**

Bezoars are rare foreign body concretions formed in the stomach and small bowel composed mainly of hair (tricho), vegetable matter (phyto) or milk curds (lacto). Most cases are females children, 6-10 years old, with bizarre appetite (trichophagia) and emotional disturbances. Originally the mass forms in the stomach and can move to the small bowel by fragmentation, extension or total translocation. Diagnosis can be confirmed by UGIS, CT-Scan or endoscopy. The child can develop an asymptomatic palpable abdominal mass, pain, obstruction or perforation. Other children will reduce intake and develop weight loss. Predisposing conditions to bezoar formations are: gastric dysmotility and decreased acidity. Management can consist of mechanical or pulsating jet of water fragmentation via the endoscope, operative extraction, shock-wave lithotripsy (ESWL) with subsequent evacuation, or dissolution by oral ingestion of proteolytic enzymes (papain, acetylcysteine, cellulase). With ESWL the shock wave pressure needed is less than half used for urolithiasis cases.

#### **N. Carcinoids**

This argentaffin cell tumor causes interest because of its diverse presentation, hormonal secretion, and malignant potential. The carcinoid is the most common neoplasm of the GI tract in childhood and may occur at any site along the alimentary tract. Above the diaphragm is commonly identified in the bronchus, and below the diaphragm in the appendix. Female predominates, the tumor is seldom life-threatening, and children rarely develop hormonal hypersecretion of 5-hydroxy indole acetic acid (Carcinoid syndrome). Carcinoids are usually discovered as an incidental finding during surgery done for other reasons. The appendix tumor arise



from subepithelial endocrine cells with exclusive growth in the lamina propria beneath the epithelial crypts. Most tumors are found in the tip of the appendix. Simple appendectomy is curative in most cases. Tumors larger than 2 cm invading neighboring structures may need right hemicolectomy. Long term follow-up is imperative.

The carcinoid syndrome (facial flushing, diarrhea, tricuspid regurgitation, pulmonic stenosis, valvular fibrosis and wheezing) is the result of serotonin overproduction by a carcinoid tumor. Carcinoid tumors arise from enterochromaffin cells (APUD cells from the neural crests), occur in virtually every organ, could be multiple, metastatic and associated with a second malignancy. Patients are diagnosed biochemically from increased urinary excretion of 5-hydroxyindoleacetic acid (5-HIAA). Platelet serotonin levels are more sensitive for detecting carcinoids that secrete small amounts of serotonin. Jejunum-ileum, bronchus and appendix are the most common sites of origin. Carcinoid of the appendix is the most common neoplasm of the GI tract in childhood. Metastasis to liver of midgut carcinoids produce the syndrome. Tumors greater than 2 cm are more prone to metastasis needing aggressive surgical management. Octreotide scan and I-131 MIBG are useful in determination of location and extent of some carcinoid tumors, particularly those of midgut origin. A positive scan may predict the ability of

Octreotide therapy to control symptoms of hormonal hypersecretion. Scans provide localization of the primary tumor that should be widely excised including lymph nodes. Higher survival rates are found for patients with midgut lesions who undergo intra abdominal debulking procedures excluding the liver. For single liver lesion resection is justified, otherwise with multiple diffuse disease hepatic artery ligation or embolization has been tried. Symptomatic metastasis should be managed with Octreotide. Prognosis is associated with the presence of liver metastasis, syndrome development and level of tumor markers (chromogranin A).

## **O. Meconium-related Disease**

### **1. Meconium Ileus**

Meconium Ileus (MI) is a neonatal intraluminal intestinal obstruction associated with Cystic Fibrosis (10-20%). The distal ileum is packed with an abnormally thick, viscous, inspissated meconium. The meconium has a reduced water content the result of decreased pancreatic enzyme activity and a prolonged small bowel intestinal transit time. MI can be classified as simple or complicated. Simple MI appears in the first 48 hrs of life with abdominal distension and bilious vomiting. Complicated MI is more severe (< 24 hrs) with progressive abdominal distension, respiratory distress, and peritonitis. X-Ray findings are: dilated bowel loops, absent air-fluid levels, "soap-bubble" granular appearance of distal ileum due to a mixture of air with the tenacious meconium. Therapy consists of Gastrografin enema for simple cases: hyperosmolar solution draws fluid to the bowel lumen causing an osmotic diarrhea. Operative therapy is reserved for failed gastrografin attempts and complicated cases (associated to volvulus, atresias, gangrene, perforation or peritonitis). Surgical procedures has included: ileostomy with irrigation, resection with anastomosis, and resection with ileostomy (Mikulicz and Bishop- Kopp). Post-operative management includes: 10% acetylcysteine p.o., oral feedings (pregestimil), pancreatic enzyme replacement, and prophylactic pulmonary therapy. Long-term prognosis depends on the degree of severity and progression of cystic fibrosis pulmonary disease.

### **2. Meconium Peritonitis**

Meconium peritonitis (MP) is a chemical peritonitis that occurs following bowel perforation during fetal life. It is generally looked upon as benign, resulting in no long-term sequelae. The peritonitis occurs when the meconium leaves the bowel, enters the peritoneal cavity and spreads throughout causing a sterile inflammatory reaction. Most common site of bowel perforation is the distal ileum, and 50% of babies with MP develop intestinal obstruction. Prenatal ultrasound findings include ascites, intraabdominal masses, bowel dilatation and the development of intraabdominal calcifications. Bowel disorders which lead to MP in utero are those resulting in bowel obstruction and perforation, such as small bowel atresias, volvulus and meconium ileus. MP can be divided into simple or complex. Cases with spontaneously healed perforation (simple MP) need observation as they rarely develop symptoms. Newborns with complex MP are born with bowel obstruction a/or pseudocyst formation (localized collection of meconium contained in a cyst made of fibrous granulation tissue). Complex MP needs surgical therapy.

### **3. Meconium plug syndrome/Left hypoplastic colon syndrome**

Colonic obstruction in the newborn child could be the result of necrotizing enterocolitis, atresia,

meconium plug syndrome, duplication cyst, Hirschsprung disease or the small left colon syndrome. In meconium plug syndrome the baby expels a grey-meconium and the obstruction subsides. Meconium plug syndrome (MPS) was first described by Clatworthy in 1956 as a transient form of distal colonic or rectal obstruction in newborns caused by an inspissated, immobile meconium. The plug is white and chalky and rarely involves the small bowel. Clinical manifestations include progressive abdominal distension, vomiting (sometimes is bilious) and failure to pass meconium during the initial two days of life. Though most cases are idiopathic, MPS has been associated with prematurity, hypotonia, hypermagnesemia (reduces acetylcholine release with subsequent myoneural depression), diabetic mother, Hirschsprung's disease and cystic fibrosis. Colonic contrast study suggests the diagnosis (filling defect) and can be therapeutic in most cases to relieve the obstruction. Gastrografin instillation is highly effective in moving the obstructing long, thick plug, even in tiny premature infants with MPS. Suction rectal biopsy to exclude the diagnosis of Hirschsprung's disease along with cystic fibrosis screening is warranted in all cases of MPS. The diagnosis of MPS is made after all the above causes are excluded. Need for surgery is extremely rare.

The left (small) hypoplastic colon syndrome (LHCS) is a very rare cause of colonic obstruction identified in newborns with characteristic roentgenographic features resembling those of Hirschsprung's disease. Manifesting in the first 24-48 hours of life, LHCS is a functional disturbance related to immaturity of the intrinsic innervation of the colon that is especially common in low birth weight neonates or of diabetic mothers. Intestinal perforation, sepsis, hypoglycemia and death may occur. The diagnosis is suggested in a barium enema when the caliber of the left colon is small with a transitional zone at the splenic flexure. Management consists of hypoglycemia correction, antibiotics, nasogastric decompression and observation. In most babies the obstruction clears in 48-72 hours. When the clinical diagnosis is not readily apparent a rectal biopsy and sweat chloride test should be done to differentiate LHCS from Hirschsprung disease and cystic fibrosis respectively. The narrowed left colon remains narrow in follow-up.

#### **P. Foreign Body Ingestion**

Accidental foreign body (FB) ingestion such as - coins, fish bones, toys plastic parts, jewels, batteries, safety pin, needles, etc. - is a common problem in children, specially infants and toddlers. Infants usually swallow button batteries while coins are the most frequently swallowed objects in children over the age of three years. No child ingests more than one FB. Management of esophageal FB differs from the rest of the gastrointestinal tract. Diagnosis is made by either chest-x-ray, barium swallow or esophagoscopy. They should be suspected when the child develops excessive salivation, vomiting, respiratory distress, recent-onset asthma, dysphagia and hematemesis. FB in the esophagus should be removed urgently to avoid erosion and perforation, specially those lodge in the upper-third of the esophagus. They can be removed using flexible fiberoptic endoscopy, balloon catheter or bougienage. Beyond the stomach, foreign body should be managed conservatively. This means follow-up visits until the FB spontaneously appears in the feces. In cases of coins ingestion serious complications are extremely rare. There is no need to x-ray monitor coins or any other metallic FB. More than 85% of all ingested FB passes spontaneously through the rectum despite nature or length. Surgery will be needed in less than 2% of all ingested FB. Patients with previous abdominal surgery are at increased risk. Development of abdominal pain, distension or profuse bleeding is an indication to remove the FB surgically.

### **IV. HERNIAS, THORACIC CONDITIONS AND ABDOMINAL WALL DEFECTS**

#### **A. Diaphragmatic Hernia**

##### **1- Congenital Diaphragmatic Hernia (Bochdalek)**

The most common congenital diaphragmatic hernia (CDH) is that which occurs through the postero-lateral defect of Bochdalek. It is caused by failure of the pleuroperitoneal membrane to develop adequately and close before the intestines returning to the abdomen at the tenth week of gestation. The intestines then enter the pleural cavity and cause poor lung development leading to pulmonary hypoplasia (a reduced number of alveoli per area of lung tissue). This defect is postero-lateral in the diaphragm and may vary in size. Stomach, liver or spleen may be partly in chest as well. Frequency is 1:2000 live births and the natural history in prenatally diagnosed CDH is that 60% will die. The clinical presentation is that the newborn becomes rapidly cyanotic,

acidotic, and has poor ventilation. Major findings relate to the degree of pulmonary maldevelopment. Chest films will show intestines in the chest. Left sided hernias are more common than right (90% on left). Placement of a radiopaque nasogastric tube may show the tube coiled in the lower left chest. Higher risk factors are: early appearance of symptoms in life, prematurity and associated anomalies. Treatment consist of rapid intubation and ventilation with use of muscle relaxants, placement of a nasogastric tube to prevent gaseous distension of the intestines and preoperative stabilization of arterial blood gases and acid-base status. Surgery can be undertaken when one of the following objectives are met: (1) blood gases normalize with no significant changes between preductal and postductal samples, (2) echocardiogram demonstrate reduce pulmonary pressure and pulmonary peripheral resistance.

Operative management consist of abdominal approach, closure of hernia by primary repair or use of mesh, and correction of malrotation. Postoperative management is very difficult. Due to hypoplastic lungs, there is frequently pulmonary hypertension leading to right-to-left shunting and progressive hypoxemia, hypercarbia, and acidosis that worsens the pulmonary hypertension. The use of chest tubes may cause overstretching of the already hypoplastic alveoli causing: increase pulmonary hypertension, reduce functional residual capacity and reduce lung compliance. Postoperatively, the infant should be kept paralyzed and ventilated and only very slowly weaned from the ventilator. The severity of pulmonary hypoplasia, both ipsilaterally and contralaterally, is the main determinant of outcome. ECMO (extracorporeal membrane oxygenator) has come to reduce somewhat the mortality of this condition.

The mortality of CDH is directly related to the degree of lung hypoplasia associated. Death is caused by persistent pulmonary hypertension and right ventricular failure. Prospective studies of prenatally diagnosed fetus prior to 25 wk. gestation has shown that 60% will die despite optimal postnatal care. This unsolved problem has prompted investigators to develop new treatment options such as preoperative stabilization, jet-frequency ventilation, and ECMO. Another area of development is intrauterine fetal surgical repair. To achieve success fetal surgery should: (1) pose no risk to the mother (innocent bystander) or her future reproductive capacity; (2) tocolytic therapy in the post-op weeks should proved effective to avoid prenatal stillbirths; and (3) the procedure should be superior to conventional therapy. Intrauterine repair has meet with limited success due to herniation of the fetal liver into the chest through the defect. Disturbance of the umbilical circulation during or after liver reduction causes fetal death. Positive-pressure ventilation after birth reduces the liver before the baby comes for surgical repair. Dr. Harrison (USFC Fetal Treatment Center) has devised separate fetal thoraco-abdominal incisions to deal with this problem ("two-step dance"), reducing or amputating the left lateral segment of the liver. Another less invasive approach is enlarging the hypoplastic lungs by reducing the normal egress of fetal lung fluid with controlled tracheal obstruction called PLUGS (Plug Lung Until it Grows).

Delayed presentation beyond the neonatal period is rare, estimated to occur in 4-6% of cases. Infants and children will present with either respiratory or gastrointestinal symptoms such as: chronic respiratory tract infection, vomiting, intermittent intestinal obstruction, and feeding difficulty. Occasionally the child is asymptomatic. The small size of the defect protected by either the spleen or the liver and the presence of a hernial sac may delay the intestinal herniation into the chest. A rise intraabdominal pressure by coughing or vomiting transmitted to any defect of the diaphragm makes visceral herniation more likely. Diagnosis is confirmed by chest or gastrointestinal contrast imaging. Management consists of immediate surgery after preop stabilization. Most defects can be closed primarily through an abdominal approach. Chest-tube placement in the non-hypoplastic lung is of help. Surgical results are generally excellent. A few deaths have resulted from cardiovascular and respiratory compromise due to visceral herniation causing mediastinal and pulmonary compression.

## **2. Morgagni Hernias**

Morgagni Hernias (MH) are rare congenital diaphragmatic defects close to the anterior midline between the costal and sternal origin of the diaphragm. They occur retrosternally in the midline or more commonly on either side (parasternally) of the junction of the embryologic septum transversum and thoracic wall (see the figure) representing less than 2% of all diaphragmatic defects. Almost always asymptomatic, typically present in older children or adults with minimal gastrointestinal symptoms or as incidental finding during routine chest radiography (mass or air-fluid levels). Infants may develop respiratory symptoms (tachypnea, dyspnea and

cyanosis) with distress. Cardiac tamponade due to protrusion into the pericardial cavity has been reported. The MH defect contains a sac with liver, small/ large bowel as content. Associated conditions are: heart defects, trisomy 21, omphalocele, and Cantrell' pentalogy. US and CT-Scan can demonstrate the defect. Management is operative. Trans-abdominal subcostal approach is preferred with reduction of the defect and suturing of the diaphragm to undersurface of sternum and posterior rectus sheath. Large defects with phrenic nerve displacement may need a thoracic approach. Results after surgery rely on associated conditions.

### **3. Hiatal Hernia**

Hiatal hernia is rarely a problem in infants unless associated with gastroesophageal reflux or severe anatomic dysfunction of the stomach (upside-down intrathoracic stomach).

Two types of esophageal hernia recognized are the hiatal and paraesophageal hernia. Diagnosis is made radiologically always and in a number of patients endoscopically. The hiatal hernia (HH) refers to herniation of the stomach to the chest through the esophageal hiatus. The lower esophageal sphincter also moves. It can consist of a small transitory epiphrenic loculation (minor) up to an upside-down intrathoracic stomach (major). HH generally develops due to a congenital, traumatic or iatrogenic factor. Most disappear by the age of two years, but all forms of HH can lead to peptic esophagitis from Gastroesophageal reflux. Repair of HH is determined by the pathology of its associated reflux (causing failure to thrive, esophagitis, stricture, respiratory symptoms) or the presence of the stomach in the thoracic cavity. In the paraesophageal hernia (PH) variety the stomach migrates to the chest and the lower esophageal sphincter stays in its normal anatomic position. PH is a frequent problem after antireflux operations in patients without posterior crural repair. Small PH can be observed. With an increase in size or appearance of symptoms (reflux, gastric obstruction, bleeding, infarction or perforation) the PH should be repaired. The incidence of PH has increased with the advent of the laparoscopic fundoplication.

## **B. Lung Bud Anomalies**

### **1. Congenital Lobar Emphysema**

Congenital lobar emphysema (CLE) is an unusual lung bud anomaly characterized by massive air trapping in the lung parenchyma that nearly always occurs in infancy and affects males more commonly (2:1). Lobar over distension causes compression of adjacent lung tissue, mediastinal shift and decrease in venous return. When this occurs persistent progressive respiratory distress (dyspnea, tachypnea, wheezing, cough and cyanosis) develops requiring lobectomy. Asymptomatic CLE exists, more commonly beyond infancy and associated with an acute viral respiratory infection. Lobar hyperinflation, flat diaphragms and retrosternal air, mediastinal shift in simple films suggests the diagnosis. CT scan depicts the abnormal anatomy (lung herniation) and the morphology of the remaining lung. V/Q scans confirm the non-functioning nature of the affected lobe. Upper and middle right lobes are more commonly affected. Etiology centers in a combination of bronchial (flap/valve) obstruction with congenital cartilage dysplasia. Most common associated defect is cardiovascular (VSD, PDA). Symptomatic patients nearly always require lobectomy. Asymptomatic children do not benefit from surgical treatment but need close follow-up. Prenatally diagnosed cases need referral to surgery centers.

### **2. Pulmonary Sequestration**

Pulmonary sequestrations refer to masses of abnormal lung parenchyma with anomalous systemic blood supply not communicating with the normal tracheobronchial tree. The abnormal lung parenchyma may be Intralobar (IS) or Extralobar Sequestration (ES). Intralobar is contained within the visceral pleural of a lower lobe receiving the blood supply from the abdominal aorta or other thoracic vessel. It is believed IS are acquired postinfectious process due to their association with chronic recurrent lung infection and reactive airway disease. ES is a congenital malformation with variable ectopic blood supply (aorta) having its own pleural investment separate from normal lung, containing typical features of CCAM-2 (40%) and associated malformations (40%). Both types can have patent communication with foregut. Prenatal diagnosis can be obtained with real-time US with Doppler imaging (can cause fetal lung compression, mediastinal shifts and hydrops). Postnatally, contrast-enhanced CT may establish the diagnosis eliminating the need for more invasive imaging (arteriography). Most presents in early infancy with a soft tissue opacity in the posterior basal segments of the lung on simple chest films. Management consists of resection to alleviate symptoms and avoid complications.

ES can be managed with resection alone, while IS needs lobectomy. Anecdotal cases of partial or total disappearance of these masses while asymptomatic has been reported.

### **3. Cystic Adenomatoid Malformation**

Congenital cystic adenomatoid malformation is a lung bud lesion characterized by dysplasia of respiratory epithelium caused by overgrowth of distal bronchiolar tissue. Prenatally diagnosed CCAM prognosis depends on the size of the lung lesion and can cause: mediastinal shift, hypoplasia of normal lung tissue, polyhydramnios, and fetal hydrops (cardiovascular shunt). Classified in two types based on ultrasound findings: macrocystic (lobar, > 5 mm cysts, anechoic, favorable prognosis) and microcystic (diffuse, more solid, echogenic, lethal). Occurs as an isolated (sporadic) event with a low rate of recurrence. Survival depends on histology. Hydrops is caused by vena caval obstruction, heart compression and mediastinal shift. The natural history is that some will decrease in size, while others disappear. Should be followed with serial sonograms. Prenatal management for impending fetal hydrops has consisted of thoraco-amniotic shunts (dislodge, migrate and occlude), and intra-uterine fetal resection (technically feasible, reverses hydrops, allows lung growth). Post-natal management consists of lobectomy.

### **4. Bronchogenic Cyst**

Bronchogenic cysts (BC), first described in 1911, are benign congenital lesions of the respiratory tract that have the potential to develop complications creating a dilemma in diagnosis and treatment. BC are commonly located in the mediastinum (2/3) or lung parenchyma (1/3) arising from anomalous budding along the primitive tracheobronchial tube (foregut duplication errors). Other atypical locations are cervical, subcutaneous, paravertebral, etc. Contain mucoid material lined with ciliated columnar epithelium (bronchial glands, smooth muscle, cartilage) not communicating with the respiratory tract. Clinical presentation may range from prenatal diagnosis, asymptomatic (1/3) lesions identified during routine work-up to symptomatic (2/3) cases. Infants may show respiratory distress: cough, dyspnea, cyanosis, hemoptysis or dysphagia. Older children present with chest pain, non-productive cough or pulmonary infection. Diagnosis relies on chest films and CT-Scan. Bronchoscopy and barium swallow are not very useful. Infection, hemorrhage, erosion, malignant potential and expansion mandate surgical management consisting of thoracotomy with excision of the lesion if mediastinal in location, and segmentectomy or lobectomy for intraparenchymal cysts. Marsupialization is associated with recurrence.

### **C. Empyema**

Thoracic Empyema (TE) is an infection (pus) of the pleural cavity. TE develops after complications of bacterial pneumonia (most commonly), thoracic trauma or surgery. Three distinct phases of TE development are recognized: exudative, fibrinopurulent and organizing. In the early exudative phase the fluid is thin with a low viscosity and cellular content. Intravenous antibiotics, aspiration or chest tube drainage accomplishes successful management as the lung expands rapidly. This phase can be followed by bacterial invasion, deposition of fibrin, increase turbidity and cellular content with fixed, less expandable, lung tissue known as fibrinopurulent stage. Loculations form, effective closed pleural drains become impossible and antibiotics are less effective in this phase. If left untreated (two to four weeks after primary infection) the TE goes through a final organizing stage with thickening of the fibrinous peel and complete lung entrapment. In this final stage open decortication may be required. Success in management of TE consists in identifying its early phases followed by thorough debridement and lysing of the pleural space of all fibrinous material, adhesions and loculations during the fibrinopurulent phase before fibrosis begins. Indication for video-assisted thoracoscopic debridement includes lack of medical response, pulmonary air leakage, localized effusion, persistent respiratory distress and pleural thickening without resolution on imaging (US or CT Scan). Thoracoscopic debridement and irrigation have accomplished this goal in several series of children reducing complications from open thoracotomy and hospital stay. Early thoracoscopy facilitates removal of restrictive purulent debris, decreases parenchymal injury, promotes rapid recovery and has a high rate of success. Benefits include good visualization of the entire thoracic cavity for more effective debridement and efficient drainage, and subjectively diminished postoperative pain and associated morbidity.

### **D. Chylothorax**

Effusion of lymph (chyle) into the pleural cavity is known as chylothorax. Chyle is clear-milky fluid with an elevated total protein and albumin level, a specific gravity above 1.012, the presence of WBC with lymphocyte predominance (80%), and elevated triglyceride (chylomicrons). In children is a potentially life-threatening disorder that has profound respiratory, nutritional (hypoalbuminemia), electrolyte (hyponatremia) and immunologic (lymphopenia, hypogammaglobulinemia, T-cell depletion) effects. Chylothorax has a congenital (mediastinal lymphangiomatosis), acquired or idiopathic origin. Acquired chylothorax is most commonly found; the result of a direct lesion of the thoracic duct or lymphatic vessels by trauma (thoracotomy, central venous catheters or chest tubes insertions), during cardiac surgery, mediastinal malignancy (neuroblastoma) or infection, repair of a diaphragmatic hernia or associated with superior vena cava obstruction (thrombosis). Initial management consists of: 1- chest tube drainage after failed thoracentesis (pleural space tamponade), 2- medium-chain triglyceride enriched formula for a week (lymphatic decompression), 3- TPN if chylothorax increases or persists. More protracted course (4 week medical tx) will require surgery to locate and suture ruptured subpleural lymphatics, ligate the thoracic duct, do chemical pleurodesis or place a pleuroperitoneal shunt. Those associated with venous obstruction or increase right sided cardiac pressure produce more volume, persist longer and are more difficult to manage.

#### **E. Spontaneous Pneumothorax**

Most pneumothorax in children are the result of blunt/open chest trauma, mechanical ventilation (barotrauma), bronchial asthma or an infectious pulmonary process. Primary spontaneous pneumothorax (PSP) is rare in children with most cases seen in adolescent males with thin body habitus. Main presenting symptoms consist of chest pain, cough and shortness of breath. Recurrence is high in this older population of children. PSP is usually the result of: 1) a ruptured apical bleb or bullae in three-fourth cases, 2) destructive parenchymal disease (cystic fibrosis, AIDS), or 3) alveolar rupture due to proximal airway obstruction. Initial management consists of oxygen supplementation for small pneumothorax less than 15% with no tension physiology present. Chest tube drainage is needed for medium or large size pneumothorax. Recurrence or persistent pneumothorax is managed with video-assisted thoracoscopic surgery (VATS) by ablating with endoscopic stapling (Endo GIA), suturing or ligating using an endoloop technique the apical bullae followed by pleurodesis. Pleurodesis can be done chemically or surgically. Chemical pleurodesis is achieved with such agents as talc, tetracycline, bleomycin or quinacrine instillation. Mechanical pleurodesis carries a lower recurrent rate and can be achieved by abrasion or electrocoagulation. Most common complication is persistent air leak. VATS is a fast, cost-effective method of treatment for PSP with less morbidity.

#### **F. Pneumatocele**

A pneumatocele is a benign air-containing cyst in the lung most commonly the result of Staphylococcus Aureus pneumonia. Other less common bacteria associated are Hemophilus Influenza, Pseudomonas Aeruginosa and Strep Pneumonia. Pneumatoceles are most commonly seen in young children (almost 50% of cases are less than one-year of age) during the acute phase of the pneumonic process. In a low percentage of cases the pneumatocele can be the result of closed chest trauma. In the infectious pneumonic setting the inflammatory process causes necrosis and liquefaction of the lung parenchyma followed by air leak and subpleural dissection forming a thin-walled cyst. Fever and respiratory distress are the most common symptoms during initial presentation. Diagnosis is established with the help of simple chest-x-ray films. CT-Scan might be needed to differentiate between a congenital lung cyst or cystic adenomatoid malformation. Follow-up films will help determine if the pneumatocele is growing or not in size. Rapidly enlarging pneumatocele may need percutaneous catheter decompression. Surgery is indicated only if the child develops respiratory distress or the pneumatocele ruptures into the pleural space creating a tension pneumothorax, a bronchopulmonary fistula or an empyema. Fortunately most pneumatoceles gradually decrease in size and disappear after the acute pulmonary infection subsides in a period that may range between six weeks and six months.

#### **G. Inguinal hernias, Hydroceles, Undescended Testis and epigastric hernias**

A hernia is defined as a protrusion of a portion of an organ or tissue through an abnormal opening. For groin (inguinal or femoral) hernias, this protrusion is into a hernial sac. Whether or not the mere presence of a hernial sac (or processus vaginalis) constitutes a hernia is debated. Inguinal hernias in children are almost exclusively indirect type. Those rare instances of direct inguinal hernia are caused by previous surgery and floor

disruption. An indirect inguinal hernia protrudes through the internal inguinal ring, within the cremaster fascia, extending down the spermatic cord for varying distances. The direct hernia protrudes through the posterior wall of the inguinal canal, i.e., medial to deep inferior epigastric vessels, destroying or stretching the transversalis fascia. The embryology of indirect inguinal hernia is as follows: the duct descending to the testicle is a small offshoot of the great peritoneal sac in the lower abdomen. During the third month of gestation, the processus vaginalis extends down toward the scrotum and follows the chorda gubernaculum that extends from the testicle or the retroperitoneum to the scrotum. During the seventh month, the testicle descends into the scrotum, where the processus vaginalis forms a covering for the testicle and the serous sac in which it resides. At about the time of birth, the portion of the processus vaginalis between the testicle and the abdominal cavity obliterates, leaving a peritoneal cavity separate from the tunica vaginalis that surrounds the testicle.

Approximately 1-3% of children have a hernia. For infants born prematurely, the incidence varies from 3-5%. The typical patient with an inguinal hernia has an intermittent lump or bulge in the groin, scrotum, or labia noted at times of increased intra-abdominal pressure. A communicating hydrocele is always associated with a hernia. This hydrocele fluctuates in size and is usually larger in ambulatory patients at the end of the day. If a loop of bowel becomes entrapped (incarcerated) in a hernia, the patient develops pain followed by signs of intestinal obstruction. If not reduced, compromised blood supply (strangulation) leads to perforation and peritonitis. Most incarcerated hernias in children can be reduced.

The incidence of inguinal hernia (IH) in premature babies (9-11%) is higher than full-term (3-5%), with a dramatic risk of incarceration (30%). Associated to these episodes of incarceration are chances of: gonadal infarction (the undescended testes complicated by a hernia are more vulnerable to vascular compromise and atrophy), bowel obstruction and strangulation. Symptomatic hernia can complicate the clinical course of babies at NICU ill with hyaline membrane, sepsis, NEC and other conditions needing ventilatory support. Repair should be undertaken before hospital discharge to avoid complications. Prematures have: poorly developed respiratory control center, collapsible rib cage, deficient fatigue-resistant muscular fibers in the diaphragm that predispose them to potential life-threatening post-op respiratory complications such as: need of assisted ventilation (most common), apnea and bradycardia, emesis, cyanosis and re-intubation (due to laryngospasm). Independent risk factors associated to these complications are (1) history of RDS/bronchopulmonary dysplasia, (2) history of patent ductus arteriosus, (3) low absolute weight (< 1.5 Kg), and (4) anemia (Hgb < 10 gm- is associated to a higher incidence of post-op apnea). Postconceptual age (sum of intra- and extrauterine life) has been cited as the factor having greatest impact on post-op complications. These observations make imperative that preemies (with post conceptual age of less than 45 weeks) be carefully monitored in-hospital for at least 24 hours after surgical repair of their hernias. Outpatient repair is safer for those prematures above the 60 wk. of postconceptual age. The very low birth weight infant with symptomatic hernia can benefit from epidural anesthesia.

At times, the indirect inguinal hernia will extend into the scrotum and can be reduced by external, gentle pressure. Occasionally, the hernia will present as a bulge in the soft tissue overlying the internal ring. It is sometimes difficult to demonstrate and the physician must rely on the patient's history of an intermittent bulge in the groin seen with crying, coughing or straining.

Elective herniorrhaphy at a near convenient time is treatment of choice. Since risk of incarceration is high in children, repair should be undertaken shortly after diagnosis. Simple high ligation of the sac is all that is required. Pediatric patients are allowed to return to full activity immediately after hernia repair. Patients presenting with incarceration should have an attempt at reduction (possible in greater than 98% with experience), and then admission for repair during that hospitalization. Bilateral exploration is done routinely by most experienced pediatric surgeons. Recently the use of groin laparoscopy through the hernial sac permits visualization of the contralateral side.

Approximately 1% of females with inguinal hernias will have the testicular feminization syndrome. Testicular feminization syndrome (TFS) is a genetic form of male pseudohermaphroditism (patient who is genetically 46 XY but has deficient masculinization of external genitalia) caused by complete or partial resistance of end organs to the peripheral effects of androgens. This androgenic insensitivity is caused by a mutation of the gene for androgenic receptor inherited as an X-linked recessive trait. In the complete form the

external genitalia appear to be female with a rudimentary vagina, absent uterus and ovaries. The infant may present with inguinal hernias that at surgery may contain testes. Axillary/pubis hair is sparse and primary amenorrhea is present. The incomplete form may represent undervirilized infertile men. Evaluation should include: karyotype, hormonal assays, pelvic ultrasound, urethrovaginogram, gonadal biopsy and labial skin bx for androgen receptor assay. These patients will never menstruate or bear children. Malignant degeneration (germ cell tumors) of the gonads is increased (22-33%). Early gonadectomy is advised to: decrease the possible development of malignancy, avoid the latter psychological trauma to the older child, and eliminate risk of losing the pt during follow-up. Vaginal reconstruction is planned when the patient wishes to be sexually active. These children develop into very normal appearing females that are sterile since no female organs are present.

A hydrocele is a collection of fluid in the space surrounding the testicle between the layers of the tunica vaginalis. Hydroceles can be scrotal, of the cord, abdominal, or a combination of the above. A hydrocele of the cord is the fluid-filled remnant of the processus vaginalis separated from the tunica vaginalis. A communicating hydrocele is one that communicates with the peritoneal cavity by way of a narrow opening into a hernial sac. Hydroceles are common in infants. Some are associated with an inguinal hernia. They are often bilateral, and like hernias, are more common on the right than the left. Most hydroceles will resolve spontaneously by 1-2 years of age. After this time, elective repair can be performed at any time. Operation is done through the groin and search made for an associated hernia. Aspiration of a hydrocele should never be attempted. As a therapeutic measure it is ineffective, and as a diagnostic tool it is a catastrophe if a loop of bowel is entrapped. A possible exception to this is the postoperative recurrent hydrocele.

The undescended testis is a term we use to describe all instances in which the testis cannot be manually manipulated into the scrotum. The testes form from the medial portion of the urogenital ridge extending from the diaphragm into the pelvis. In arrested descent, they may be found from the kidneys to the internal inguinal ring. Rapid descent through the internal inguinal ring commences at approximately week 28, the left testis preceding the right. Adequate amounts of male hormones are necessary for descent. The highest levels of male hormones in the maternal circulation have been demonstrated at week 28. Thus, it appears that failure of descent may be related to inadequate male hormone levels or to failure of the end-organ to respond.

The undescended testes may be found from the hilum of the kidney to the external inguinal ring. A patent processus vaginalis or true hernial sac will be present 90% of the time. The incidence is about 0.28% of the population, approximately 50% occurs on the right, 25% on the left, and 25% occur bilaterally.

The diagnosis of undescended testes is usually made by the parents or first examining physician. The important point is the absolute necessity of distinguishing between retractile testes and the true undescended testes. Testes that can be drawn to the scrotum, even if they retract again, are retractile testes and not undescended, the squatting position may aid in helping descend the testes for exam. Retractable testis need no further surgical management.

Since Leydig cell degeneration can occur after age two, present recommendations are for orchiopexy before age 2. Although testicular malignancy is rare, undescended testes have a 40-50 times higher incidence of developing seminomas. This can occur in the contralateral normally descended testis as well as the undescended testis. Surgical repair does not reduce the incidence of malignancy, but does allow for examination and earlier detection. Another reason for surgical repair is the higher incidence of trauma and torsion in the undescended testis. Bilateral undescended testes may be initially treated with a four-week course of human chorionic gonadotrophin. Approximately 15-30% of patients will have descent with this therapy. Surgical repair is most commonly performed by a Dartos pouch technique. Laparoscopy helps in non-palpable undescended testis by identifying those testis that did not develop, suffered an ischemic intrauterine event, and in performing first stage Steven-Fowler technique.

Congenital epigastric defects occur anywhere in the linea alba from the navel to the xiphoid process. They represent almost 5% of all hernias defect that presents in children. Most epigastric hernias occur in the midline, are small (15-25 mm), asymptomatic and reducible. Multiple fascial defects can also be present in 20% of all cases. The defect might arise congenitally from an abnormally wide orifice of a blood vessel during development of the linea alba. The bump is the result of a piece of preperitoneal fat stuck through the fascial defect. Tenderness is an unusual symptom while growth of the defect occurs with time. Most surgeons



recommend repair of the defect at the time of presentation. Repair is an outpatient procedure done under general anesthesia with low morbidity and risk of recurrence. Voluminous epigastric hernia (5-10 cm) with a sac that contains epiploic appendages or viscera (ileum loops, stomach) has also been rarely reported in infants.

#### **H. Mediastinal Cysts**

Mediastinal cysts identified in children are classified according to the compartment where they arise as: anterior (extends to the sternum, thoracic inlet and anterior border of the heart), middle (between anterior mediastinum and anterior borders of the vertebrae) or posterior mediastinum. Although usually asymptomatic, they require excision for purpose of diagnosis and avoidance of symptoms such as chest pain, airway obstruction, hemoptysis or dysphagia. Diagnosis can be accomplished with the use of CT-Scan, US and esophagogram. Some of the most common encounter cysts in the mediastinum are: bronchogenic cysts, neurenteric cysts, pericardial cysts, cystic hygroma, thymic and dermoid cysts.

#### **I. Umbilical Hernias**

Between the sixth and tenth gestational week, the developing gastrointestinal tract is partially extruded into the umbilical cord with return into the abdomen by the tenth week. By the time of birth, the umbilical ring has become entirely closed by the developing abdominal wall except for the space occupied by the cord, which contains the umbilical vein, paired umbilical arteries, and the fibrous remnants of the urachus and omphalomesenteric duct (yolk sac). After ligation of the cord, the vessels thrombosed and the cord dries and sloughs off, leaving a granulating surface that heals by cicatrization and is covered by epithelium. This is followed by scar contraction and retraction of the umbilicus. It is believed that most umbilical hernias occur through the cephalad portion of the umbilical ring, where the contracted scar around the obliterated umbilical vein is less dense than in the caudal portion of the scar. Umbilical hernias are very common, especially in prematures, blacks, and certain syndromes, such as Down's. The incidence decrease with age since many will close spontaneously. The diagnosis is made by physical exam, there is a fascial defect at the umbilicus. Complications such as strangulation or incarceration are extremely rare in children.

Umbilical strapping should not be done since it does not promotes closure and may lead to skin erosion. Elective repair is usually delayed until after five years of age since closure may occur spontaneously or the defect may get smaller, allowing easier repair. Operative repair includes excision of the sac and horizontal one-layer closure through a small infraumbilical incision.

#### **J. Omphalocele and Gastroschisis**

The three most common abdominal wall defect in newborns are umbilical hernia, gastroschisis and omphalocele. Referral to tertiary centers with available neonatal intensive care is necessary in prenatally diagnosed cases. Changing the route of delivery does not affect outcome for either defect. Omphalocele has a high incidence of associated anomalies (cardiac, neurogenic, genitourinary, skeletal, chromosomal syndromes) that are the cornerstones of mortality. Detailed search for associated anomalies, fetal echocardiogram and karyotyping should be performed always. Cesarean section is justified in large omphaloceles (> 5 cm) to avoid liver damage, sac rupture and dystocia. Gastroschisis prenatal US appearance depends on gestational age and condition of extruded bowel. Fetal karyotyping testing is less important. Intestinal atresia complicates the defect, the result of an intrauterine vascular accident. Intestinal obstruction due to atresia or luminal constriction may cause polyhydramnios, fetal growth retardation and preterm labor, findings that can be monitored with serial US. No benefit has been found in recommending routine c-section for most cases of gastroschisis. Preterm deliveries by c-section have been found to prevent bowel damage in fetus with progressive bowel dilatation and thickening, a finding that has not been corroborated by others. Abnormal US appearance of fetal bowel is associated with more bowel edema, longer operative time and a higher incidence of postoperative complications. Omphalocele is a milder form of primary abdominoschisis since during the embryonic folding process the outgrowth at the umbilical ring is insufficient (shortage in apoptotic cell death). Bowel and/or viscera remains in the umbilical cord causing a large abdominal wall defect. Defect may have liver, spleen, stomach, and bowel in the sac while the abdominal cavity remains underdeveloped in size. The sac is composed of chorium, Wharton's jelly and peritoneum. The defect is

centrally localized and measures 4-10 cm in diameter. A small defect of less than 2 cm with bowel inside is referred as a hernia of the umbilical cord. There is a high incidence (30-60%) of associated anomalies in patients with omphalocele. Epigastric localized omphalocele are associated with sternal and intracardiac defects (i.e., Pentalogy of Cantrell), and hypogastric omphalocele have a high association with genito-urinary defects (i.e., Cloacal Exstrophy). All have malrotation. Cardiac, neurogenic, genitourinary, skeletal and chromosomal changes and syndromes are the cornerstones of mortality. Antenatal diagnosis may affect management by stimulating search for associated anomalies and changing the site, mode or timing of delivery. Cesarean section is warranted in large omphaloceles to avoid liver damage and dystocia. After initial stabilization management requires consideration of the size of defect, prematurity and associated anomalies. Primary closure with correction of the malrotation should be attempted whenever possible. If this is not possible, then a plastic mesh/silastic chimney is fashioned around the defect to cover the intestinal contents and the contents slowly reduced over 5-14 days. Antibiotics and nutritional support are mandatory. Manage control centers around sepsis, respiratory status, liver and bowel dysfunction from increased intraabdominal pressure.

The exact embryology of gastroschisis is unclear. The defect is always to the right of the midline with a normally attached umbilical cord. Theories include failure of the right lateral somatopleure to form properly, intrauterine rupture and intrauterine vascular accident leading to ischemia of the right developing rectus. Associated anomalies are rare, with an 11% incidence of atresia. Treatment is identical to omphalocele except more urgent to avoid problems with the exposed bowel (dehydration and hypothermia). More than 90% babies survived. Prenatal diagnosis has brought a controversy toward optimal mode of delivery (Cesarean vs vaginal). The appearance of the bowel is edematous, matted and foreshortened due to exposure to amniotic fluid, and the constrictive vascular effects of a small defect.

#### **K. Femoral Hernias**

Femoral hernias (FH) in the pediatric age are very rare accounting for less than 0.5% of all groin hernias in children. Children present with a recurrent groin lump that is usually reducible. The correct preoperative diagnosis is often overlooked. FH is defined as a protrusion of viscera, fat or omentum occurring through the femoral hiatus. Peak incidence occurs between five and ten years of age. Misdiagnosis includes inguinal hernia, lymphadenitis, and lymphangioma. Preoperative diagnosis is possible if the bulge appears in a location inferior and lateral to that of the commonly occurring indirect hernia. Early recurrence of a groin swelling after what seems to be an adequate inguinal herniorrhaphy should be suspected of having a missed femoral hernia. Mc Vay expressed that the etiology was a congenitally narrow posterior inguinal wall attachment into Cooper's ligament with a resultant enlarged femoral ring. Excision of the sac and repair of the femoral canal is curative. At surgery Cooper's ligament (Mc Vay) repair is the surgical treatment of choice, though some contend that simple repair of the femoral ring carries good long-term results.

### **V. GASTROINTESTINAL BLEEDING**

#### **A. Upper GI bleeding (Neonate)**

Initially do an Apt test to determine if blood comes from fetal origin or maternal origin (blood swallowed by the fetus). If its of fetal origin then consider a coagulation profile (PT, PTT). If this coagulation profile is normal the possibilities are either stress gastritis or ulcer disease. If the coagulation profile is abnormal then consider hematologic disease of the newborn and manage with vitamin K. The apt test is performed by mixing 1 part of vomitus with 5 part H<sub>2</sub>O, centrifuge the mixture and remove 5 ml (pink). Then add 1 ml 1% NaOH, wait 2 minutes and if it remains pinks is fetal blood, if it turns brown-yellow its maternal blood.

#### **B. Lower GI bleeding (Neonate)**

Again start with an Apt test, if its positive its maternal swallow blood, if its negative do a PT, PTT. If the coagulation profile is abnormal give Vit K for hematologic disorder of newborn. If it's normal do a rectal exam. A fissure could be the cause, if negative then consider either malrotation or Necrotizing enterocolitis.

##### **1- Necrotizing Enterocolitis (NEC)**

Although the exact pathogenesis of NEC is not known, the most widely held theory is that of perinatal stress leading to selective circulatory ischemia. The stress includes prematurity, sepsis, hypoxia, hypothermia, and jaundice. These babies frequently have umbilical artery, vein catheters, have received exchange

transfusions or early feeds with hyperosmolar formulas. The intestinal mucosal cells are highly sensitive to ischemia and mucosal damage leads to bacterial invasion of the intestinal wall. Gas-forming organisms produce pneumatosis intestinalis (air in the bowel wall readily seen on abdominal films). Full-thickness necrosis leads to perforation, free air and abscess formation. These usually premature infants develop increased gastric residuals, abdominal distension, bloody stools, acidosis and dropping platelet count. The abdominal wall becomes reddened and edematous. There may be persistent masses and signs of peritonitis. Perforation leads to further hypoxia, acidosis and temperature instability. The acid-base status is monitored for worsening acidosis and hypoxia. Serial platelet counts are obtained and, with increasing sepsis, the platelet count drops  $<50,000$ , indicating intravascular coagulation and decreased bone marrow production. The white blood cell count may be high, low or normal and is not generally of help. Serial abdominal films are obtained to look for evidence of free abdominal air, a worsening picture of pneumatosis intestinalis, or free portal air. Therapy consist initially of stopping feeds, instituting nasogastric suctioning and beginning broad-spectrum antibiotics (ampicillin and gentamycin). Persistent or worsening clinical condition and sepsis or free air on abdominal films require urgent surgical intervention. Attempts to preserve as much viable bowel as possible are mandatory to prevent resultant short gut syndrome.

Complicated NEC is the most common neonatal surgical emergency of modern times, has diverse etiologies, significant mortality and affects mostly premature babies. The use of primary peritoneal drain (PPD) in the management of NEC dates from 1977. The technique is used in the very low birth weight premature infant ( $<1500$  gm) with pneumoperitoneum, metabolic and hemodynamic instability. Consist of a right lower quadrant incision and placement of a drainage (Penrose or catheter) under local anesthesia with subsequent irrigation performed bedside at the NICU. Initially used as a temporizing measure before formal laparotomy, some patient went to improvement without the need for further surgery (almost one-third). They either had an immature (fetal type) healing process or a focal perforation (not associated to NEC?) which healed spontaneously. Those babies not improved by PPD either die (20%), go on to laparotomy and half die (20%) or develop complications (24%). Some suggestion made are: PPD should be an adjunct to preop stabilization, before placing drain be sure pt has NEC by X-rays, persistent metabolic acidosis means uncontrolled peritoneal sepsis, do not place drain in pts with inflammatory mass or rapid development of intraperitoneal fluid, the longer the drainage the higher the need for laparotomy.

### **C. Upper GI Bleeding (Older Children)**

In the initial evaluation a history should be obtained for bleeding disorders, skin lesions, and aspirin or steroid ingestion. The physical exam for presence of enlarged liver, spleen, masses, ascites, or evidence of trauma or portal hypertension. Labs such as bleeding studies and endoscopy, contrast studies if bleeding stops. Common causes of Upper GI bleeding are:

#### 1. Esophagus

(a) Varices- usually presents as severe upper gastrointestinal bleeding in a 2-3 year old who has previously been healthy except for problems in the neonatal period. This is a result of extrahepatic portal obstruction (portal vein thrombosis most commonly), with resulting varices. The bleeding may occur after a period of upper respiratory symptoms and coughing. Management is initially conservative with sedation and bedrest; surgery is rarely needed.

(b) Esophagitis- this is a result of persistent gastroesophageal reflux leading to inflammation and generally slow, chronic loss of blood from the weeping mucosa. Treatment consist of antacids, frequent small feeds, occasionally medications and if not rapidly improved, then surgical intervention with a fundoplication of the stomach.

© Mallory-Weiss- this is a tear of the distal esophagus and/or gastroesophageal junction secondary to severe regurgitation. This was thought to be uncommon in children because it was not looked for by endoscopy. It probably occurs more often than previously thought. Treatment initially is conservative and, if persistent, oversewing of the tear through an incision in the stomach will be successful.

(d) Duplication cysts- Rare cause, they are seen on the mesenteric side of the intestine anywhere from the esophagus to the anus. They bleed when there is ectopic gastric mucosa present. Total excision is curative.

#### 2. Stomach

- (a) Gastric Erosions- managed medically in most cases.
- (b) Ulcer- treated medically unless there is persistent hemorrhage, obstruction or perforation.
- © Hematoma- usually secondary to trauma or bleeding disorders.

### 3. Duodenum

- (a) Duodenitis- associated to acid peptic disease.
- (b) Hematobilia- secondary to blunt or penetrating abdominal injury. Occasionally requires surgical intervention with local repair or ligation of hepatic vessels.

## **D. Lower GI bleeding (Older Children)**

### **1- Anal Fissure**

Anal fissure is the most common cause of rectal bleeding in the first two years of life. Outstretching of the anal mucocutaneous junction caused by passage of large hard stools during defecation produces a superficial tear of the mucosa in the posterior midline. Pain with the next bowel movement leads to constipation, hardened stools that continue to produce cyclic problems. Large fissures with surrounding bruising should warn against child abuse. Crohn's disease and leukemic infiltration are other conditions to rule-out. The diagnosis is made after inspection of the anal canal. Chronic fissures are associated with hypertrophy of the anal papilla or a distal skin tag. Management is directed toward the associated constipation with stool softeners and anal dilatations, warm perineal baths to relax the internal muscle spasm, and topical analgesics for pain control. If medical therapy fails excision of the fissure with lateral sphincterotomy is performed.

### **2- Meckel's Diverticulum**

Meckel's diverticulum denotes those anatomic structures resulting from the umbilical and intra-abdominal persistence of the embryonic vitelline duct (yolk stalk). Meckel's diverticulum is clinically significant, either when discovered incidentally at celiotomy or recognize as the cause of symptoms, which are intestinal bleeding, abdominal pain, or intestinal obstruction. Children with symptomatic Meckel's present with hemorrhage (40%), intestinal obstruction (30%), diverticulitis (20%), and umbilical discharge or disease (6%). Diagnosis is related to presentation. Intermittent rectal bleeding may be diagnosed on occasion by <sup>99m</sup>Tc sodium pertechnetate. Barium studies are unreliable. Persistent bleeding requires laparotomy even if the Meckel's scan is negative. Diverticulitis or perforation presents with finding similar to appendicitis. Obstruction secondary to intussusception is diagnosed by clinical findings and barium enema. Incidental Meckel's diverticulum is generally handled by simple wedge excision in children. Bleeding Meckel's is managed by resection of the bowel and end-to-end anastomosis. Diverticulitis is handled by either method. Asymptomatic Meckel's diverticula identified incidentally should be removed if upon palpation there is questionable ectopic (gastric or pancreatic) mucosa.

### **3- Polyps**

Juvenile Polyps comprise 80% of childhood polyps. This is a mucous retention polyp and histologically features a cluster of mucoid lobes surrounded by flattened mucus-secreting glandular cells. There is no malignant potential. These polyps are most commonly seen in children age 3-10 with a peak at age 5-6. 85% of children have a solitary polyp. The most common complaint is rectal bleeding and occasionally the polyps may prolapse out the rectum. Diagnosis is by barium enema, rectal exam and/or endoscopy. Removal by endoscopy is the treatment of choice. rarely, exploratory lap with colotomy and excision is required. Lymphoid polyps are not polyps at all, but rather localized elevations of colonic mucosa that on barium enema produce filling defects resembling polyps. They are produced by hyperplasia of submucosal lymphoid tissue. These make up about 15% of childhood polyps. They begin to appear during the first year of life, peak at about the third year of life, and diminish in number by 5 years of age. These are multiple and may present with mild chronic blood loss. Diagnosis is made by barium enema. endoscopy and biopsy. No treatment is necessary since they will regress spontaneously.

Peutz-Jeghers syndrome are polyps associated with melanin hyperpigmentation of the lips and oral mucosa. The polyps are usually multiple and hamartomas. They will appear anywhere, but the majority are limited to the jejunum and ileum. Chronic blood loss anemia in a child with repeated bouts of colicky abdominal pain secondary to actual or incipient intussusception are typical. Diagnosis is by contrast studies. GI malignancy has been reported in 2-3% of patients. Females with Peutz-Jeghers syndrome seem predisposed to develop ovarian tumors, usually in adolescence. Treatment depends on the severity of symptoms and extend of

involvement.

Familial Polyposis Coli has an autosomal dominant inheritance. Hundreds of adenomatous colonic polyps with virtually all developing adenocarcinoma of colon by the third decade of life. The most common and best-recognized variant is patients with Gardner's syndrome, which combines the premalignant adenomatous colonic polyps with soft and hard tissue tumors. Diagnosis is by family history, and contrast studies followed by biopsy. Treatment is total colectomy with ileostomy or endorectal pull-through with ileo-anal anastomosis.

#### **4- Familial Adenomatous Polyposis**

Familial history of colon cancer is an important indicator of future risk for colorectal cancer. The more extensive and closer the affected relatives, the greater the risk. Highest risk is found in Familial Adenomatous Polyposis (FAP). Once the risk is appreciated screening for the disease must take place. This involves genetic analysis for members of syndrome families along with lower gastrointestinal endoscopy for the rest as polyps can occur throughout the gastrointestinal tract. FAP is a genetic (autosomal dominant) premalignant condition that will ultimately manifest with the development of colorectal carcinoma. FAP has been linked to germline mutations of the adenomatous polyposis coli (APC) gene. Initial presentation can be rectal bleeding. Besides multiple adenomatous polyps of the colon predisposing to malignancy at an early age, a variety of extra colonic manifestations are associated with this condition. Once the diagnosis of FAP is established endoscopic surveillance should be instituted. All polyps should be subjected to histopathological exam to determine presence of adenomatous epithelium. With the presence of dysplastic changes total colectomy with the creation of an ileorectal or ileoanal (pouch) anastomosis is recommended. Almost one-third of cases develops adenomas in the ileal pouch after proctocolectomy. Baseline small bowel enteroscopy should be done at the time of surgery and in the postop period in children with FAP and juvenile polyposis. With duodenal polyps enteroscopy should be done at the time of surgery. Biopsy and/or excision of larger polyps should be done as they may harbor a carcinoma.

## **VI- PANCREATIC, HEPATIC, BILIARY and SPLENIC DISORDERS**

### **A. Annular Pancreas/Pancreas Divisum**

The pancreas develops from an anterior and posterior anlage of the foregut early during gestation (28 days). The anterior bud leads to the liver and body and tail of the pancreas. The posterior diverticulum develops into the head of the pancreas. This bud rotates anteriorly and later fuses to achieve the relationship to the rest of the pancreas. Development of the pancreas in embryonic life requires a trophic stimulus from the associated mesenchyme. Under the influence of this mesenchyme the mature organ develops, being mainly composed of ductal, exocrine and endocrine cells. Exocrine and ductal pancreas are derived from the endoderm of the foregut. Recent evidence suggests that the endocrine cells derive also from the endoderm of the foregut as evidenced by the expression of the genes responsible for hormonal production. This challenges the theory that endocrine cells may originate from the neural crest cells (neuroectodermal) of the embryo reinforced by the enunciation of the amine precursor uptake decarboxylase (APUD) theory.

Annular pancreas is the most common congenital malformation of the pancreas in association with duodenal atresia. Embryologically the ring formation (annulus) originates from the ventral pancreas primordium (Lecco's theory). The pancreatic duct of the annular tissue passed from the anterior portion to the lateral and posterior portion finally joining with the main pancreatic duct. Two types of annular pancreas are recognized: 1) Extramural - causing high gastrointestinal obstruction; vomiting is the most common presenting symptom, and 2) Intramural - producing duodenal ulceration. Presentation at birth is affected by the degree of duodenal obstruction and coexistent anomalies. Polyhydramnios usually accompanies complete high intestinal obstruction in annular pancreas. Associated anomalies can range from malrotation, intrinsic duodenal obstruction, Down syndrome and duodenal bands. ERCP is the most important procedure to find the characteristic features and establish the therapeutic strategy in cases of annular pancreas. Experience militates against any direct attack on the offending annulus. Therefore, all children with this abnormality are generally treated with a bypass procedure, preferably a duodeno-duodenostomy. Long-term complications may include cholestatic jaundice, upper gastrointestinal motility disorder, failure to thrive, chronic diarrhea and chronic

relapsing pancreatitis due to an incomplete divisum anomaly.

Pancreas divisum (PD), believed the most common congenital anomaly of the pancreas, is an embryologic variation of pancreas development where the dorsal (Santorini) and ventral portions (Wirsung) ducts drain separately. Diagnosis is made with ERCP (short duct of Wirsung that does not communicate with main pancreatic duct of Santorini). Not everybody with this ductal anomaly develops pancreatitis. Likewise with the minor papilla draining the bulk of the pancreas in PD, a small orifice size (< 0.75 mm) plays a role in outflow obstruction and development of pancreatitis. Children with PD and recurrent episodes of pancreatitis will need endoscopic sphincterotomy of the minor and sometimes major papilla. If not feasible technically, surgical sphincteroplasty of both papillae along with cholecystectomy (bile stasis leads to gallstones) is indicated. Intraoperative pancreatogram will help determine if both papilla are stenotic. Once chronic pancreatitis is established, ductal drainage or resection may be necessary.

### **B- Pancreatitis**

Acute pancreatitis (AP) is unusual in the pediatric patient, can affect all age groups and should be considered in children presenting with acute abdominal complaints. Causes are diversely and clinical course less severe. The three most common etiological factors are: trauma, drug-induced, and biliary tract disorders. Other factors to consider are: infections (mumps, ascaris, adenovirus), metabolic (branched-chain organic acidemias), structural defects (anomalous union of pancreatico-biliary ductal system), and hereditary. Blunt abdominal trauma is the leading cause (20-30%) of AP by crushing the fixed organ between the spine. Drugs associated to the development of AP are: steroids, L-asparaginase, valproic acid, acetaminophen (drug withdrawal is treatment of choice). Biliary disorders related to AP are gallstone and choledochal cysts by causing transient ductal obstruction. Most common complaint of children with AP is abdominal pain. Diagnosis is confirmed with elevated amylase/lipase in serum and urine (lipase is more specific since pancreas is major source). Imaging studies of utility are US, CT-Scan and ERCP. The use of ERCP in previously idiopathic cases of AP have increased the yield of diagnosing anomalous pancreatico-biliary junctional defects. Management during early phase is supportive with IV therapy, NG decompression, NPO (to decrease acid stimulation and prevent secretin release), and nutritional (TPN). Surgery is rarely required except complications such as abscess and pseudocyst formation.

Uncommon disorder in childhood. Trauma (compressed injury against spinal column) and biliary tract disorders (choledochal cyst, cholelithiasis) are most common cause of pancreatitis. The most common congenital ductal anomaly leading to pancreatitis is pancreas divisum. Most common complaint is mid-epigastric abdominal trauma associated with nausea and vomiting. Diagnosis is confirmed with elevated levels of amylase and lipase. Ultrasound is useful to determine degree of edema and presence of pseudocyst formation. Treatment consists of: NPO, NG decompression, decrease acid stimulation (H-2 blockers), aprotinin, glucagon, and anticholinergics. Pain is relieved with meperidine. When pancreatic serum enzymes level return to near normal level patient is started in low-fat diet. Antibiotic prophylaxis use is controversial. Surgery is indicated for: abscess formation and pseudocyst. Pseudocysts are the result of major ductal disruptions or minor lacerations. Observation allows spontaneous resolution in 40-60% of cases. Percutaneous aspiration and catheter drainage is another alternative in management. Follow-up studies permit determine if cavity is decreasing in size. This can be done outpatient teaching parents to irrigate the catheter at home to assure patency. Persistency beyond 6 months may need resectional therapy. Additional option is internal drainage (cyst-gastrostomy, cyst-jejunostomy). Abscess should be drained and debride.

Pancreatic pseudocyst formation is an uncommon complication of pancreatic inflammatory disease (pancreatitis) or trauma in children. More than half cases are caused by blunt abdominal trauma. Ultrasound is the most effective and non-invasive way of diagnosing pancreatic pseudocysts. Acute pseudocysts are managed expectantly for 4-6 wk. until spontaneous resolution occurs. 25-50% will undergo spontaneous resolution. Medical therapy consists of decreasing pancreatic stimulation and giving nutritional support. Rupture is the major complication of conservative management. Chronic pseudocysts (> three mo.) will benefit from prompt operation and internal drainage since resolution is rare. Percutaneous catheter drainage under local anesthesia using Ultrasound or CT guided technique is an appropriate method of first-line therapy for non-resolving (chronic) or enlarging pancreatic pseudocysts. The approach is transgastric or transcutaneous.

Those cysts that fail to resolve with percutaneous drainage should go investigation of ductal anatomy to rule out disruption of the main pancreatic duct. The need for further surgery (drainage or resectional) will depend on the status of the duct of Wirsung.

### **C. Pancreatic Cysts**

Pancreatic cystic lesions are usually inflammatory pseudocyst (90%) or neoplastic process (10%). Distinguishing between them is essential for appropriate surgical therapy. Non-inflammatory neoplastic cysts in children are very rare ductal lesions with a spectrum of histologic characteristics and favorable outcomes. Histologically they include retention cysts, lymphoepithelial cysts, papillary cystic tumors, benign serous cystadenoma, mucinous tumors and mucinous cystadenocarcinoma. Most reported cases occur in females during adolescent years. Mode of presentation includes mild upper abdominal pain and palpable mass. Clinical, radiographic and intraoperative frozen section are non-reliable methods in distinguishing the different types of pancreatic cysts. Preoperative cyst fluid obtained by US or CT-guided percutaneous aspiration can be analyzed for viscosity (mucoid, viscous, serous), chemical (amylase, lipase), tumor markers (CEA, CA 19-9, CA125) and cytology characteristics. High CEA levels (> 25 ng/ml) indicate that the cyst is either malignant or mucinous (pre-malignant) type. Higher levels of CA 19-9 suggest pseudocysts and serous cystadenomas. Very high CA 125 levels appear predictive of malignancy. Viscosity above 1.63 suggests mucinous tumors. Amylase and lipase content should be low in true pancreatic cysts. Cytology analysis is insensitive unless positive for tumor cells. When the nature of the pancreatic cyst cannot be definitively establish by the above methods surgical resection is indicated.

### **D. Hepatic Cysts**

Hepatic cysts (HC) can be either parasitic (echinococcal) following infestation in endemic regions, acquired (after trauma or inflammatory processes), or nonparasitic (congenital) in nature. Congenital nonparasitic HC are uncommon, solitary, benign lesions that arise from aberrant development of intrahepatic biliary radicals after ischemic thrombo-embolic phenomena (vascular disruption theory). The cyst is lined with cuboidal or squamous epithelium, and there is a female and white children predominance. Although generally asymptomatic, children may manifest increased abdominal girth, vague abdominal discomfort, infection, or obstructive jaundice. Ultrasound and CT-Scan are diagnostic tools. Management may consist of: simple unroofing, complete removal by enucleation or hepatic lobectomy, internal Roux-en-Y drainage, or percutaneous aspiration and sclerosis (alcohol, minocycline). The surgical alternative to use will depend on size, location (central, peripheral or dumbbell), and presence of communication with biliary system of the cyst (see figure). Some cases diagnosed prenatally or during the neonatal period have undergone slow spontaneous regression.

### **E. Biliary Atresia**

Persistent conjugated hyperbilirubinemia (greater than 20% of total or 1.5 mg%) should be urgently evaluated. Initial evaluation should include a well-taken history and physical exam, partial and total bilirubin determination, type and blood group, Coomb's test, reticulocyte cell count and a peripheral smear.

Cholestasis means a reduction in bile flow in the liver, which depends on the biliary excretion of the conjugated portion. Reduce flow causes retention of biliary lipoproteins that stimulates hypercholesterolemia causing progressive damage to the hepatic cell, fibrosis, cirrhosis and altered liver function tests.

Biliary Atresia (BA) is the most common cause of persistently direct (conjugated) hyperbilirubinemia in the first three months of life. It is characterized by progressive inflammatory obliteration of the extrahepatic bile ducts, an estimated incidence of one in 15,000 live births, and predominance of female patients. The disease is the result of an acquired inflammatory process with gradual degeneration of the epithelium of the extrahepatic biliary ducts causing luminal obliteration, cholestasis, and biliary cirrhosis. The timing of the insult after birth suggests a viral etiology obtained transplacentally. Almost 20% of patients have associated anomalies such as: polysplenia, malrotation, situs inversus, pre-duodenal portal vein and absent inferior vena cava. Histopathology is distinguished by an inflammatory process in several dynamic stages with progressive destruction, scar formation, and chronic granulation tissue of bile ducts. Physiologic jaundice of the newborn is a common, benign, and self-limiting condition.

In BA the patient develops insidious jaundice by the second week of life. The baby looks active, not

acutely ill and progressively develops acholic stools, choluria and hepatomegaly. Non-surgical source of cholestasis shows a sick, low weight infant who is jaundiced since birth. The diagnostic evaluation of the cholestatic infant should include a series of lab tests that can exclude perinatal infectious (TORCH titers, hepatitis profile), metabolic (alpha-1-antitrypsin levels), systemic and hereditary causes. Total bilirubin in BA babies is around 6-10 mg%, with 50-80% conjugated. Liver function tests are nonspecific. Lipoprotein-X levels greater than 300 mg% and Gamma Glutamyl Transpeptidase (GGT) above 200 units% suggest the diagnosis. The presence of the yellow bilirubin pigment in the aspirate of duodenal content excludes the diagnosis of BA. Ultrasound study of the abdomen should be the first diagnostic imaging study done to cholestatic infants to evaluate the presence of a gallbladder, identify intra or extrahepatic bile ducts dilatation, and liver parenchyma echogenicity. The postprandial contraction of the gallbladder eliminates the possibility of BA even when nuclear studies are positive. Nuclear studies of bilio-enteric excretion (DISIDA) after pre-stimulation of the microsomal hepatic system with phenobarbital for 3-5 days is the diagnostic imaging test of choice. The presence of the radio-isotope in the GI tract excludes the diagnosis of BA. Percutaneous liver biopsy should be the next diagnostic step. The mini-laparotomy is the final diagnostic alternative. Those infant with radiographic evidence of patent extrahepatic biliary tract has no BA. Medical management of BA is uniformly fatal. Kasai portoenterostomy has decreased the mortality of BA during the last 30 years. Kasai procedure consists of removing the obliterated extrahepatic biliary system, and anastomosing the most proximal part to a bowel segment. Almost three-fourth of patients will develop portal hypertension in spite of adequate postoperative bile flow. They will manifest esophageal varices, hypersplenism, and ascites. Hepatic transplantation is reserved for those patients with failed portoenterostomy, progressive liver failure or late-referral to surgery.

#### **F. Biliary Hypoplasia**

Biliary hypoplasia is a rare cause of persistent neonatal conjugated hyperbilirubinemia. Pathologically, affected children have absent or reduced number of bile ductules with normal distribution of branches of the portal vein and hepatic artery within the liver parenchyma. Biliary hypoplasia is also identified as paucity of interlobular bile ducts (PILBD). Two types of PILBD are recognized: 1) syndromic (arteriohepatic dysplasia or Alagille's syndrome) with characteristic extrahepatic abnormalities (facial appearance, pulmonic artery stenosis, vertebral anomalies, embryotoxon and delayed weight-height development), and 2) non-syndromic biliary hypoplasia. Biliary hypoplasia is clinically indistinguishable from biliary atresia and can sometimes be confused. A definitive diagnosis is difficult to make in early infancy. Differentiation between biliary atresia, hypoplasia and neonatal hepatitis continues to require direct visualization of the biliary ducts. This mean laparoscopic or open intra-operative cholangiography and liver biopsy. The cholangiogram will show diminutive intra- and extra-hepatic biliary tree. Attempts to establish biliary flow by means of hepatic porto-enterostomy (Kasai procedures) in children with PILBD have been unsuccessful and contraindicated. Management is conservative and include predigested formulas, ursodeoxycholic acids (10 mg/kg/day), phenobarbital and A,D,K,E vitamin

replacement. Non-syndromic PILBD have better long-term prognosis. Children with syndromic PILBD identified in infancy because of cholestasis have a 50% probability of long-term survival without liver transplantation.

#### **G. Choledochal Cyst**

Choledochal cyst is a rare dilatation of the common bile duct, prevalent in oriental patients (Japan), where >60% of patients are less than 10 years old. The etiology is related to an abnormal pancreatic-biliary junction (common channel theory) causing reflux of pancreatic enzymes into the common bile duct (trypsin and amylase). Symptoms are: abdominal pain, obstructive jaundice, a palpable abdominal mass, cholangitis, and pancreatitis. Infants develop jaundice more frequently, causing diagnostic problems with Biliary Atresia. Older children may show abdominal pain and mass. Jaundice is less severe and intermittent. Diagnosis is confirmed with Ultrasound and corroborated with a HIDA (or DISIDA) Scan. Choledochal cysts are classified depending on morphology and localization. Management is surgical and consist of cyst excision and Roux-en-Y hepatico-jejunostomy reconstruction. Cyst retention penalties paid are: stricture, cholangitis, stone formation, pancreatitis, biliary cirrhosis, and malignancy. Long-term follow-up after surgery is advised.

#### **H. Cholelithiasis**

With the increase use of sonography in the work-up of abdominal pain, cholelithiasis is diagnosed more



frequent in children. Gallstones occur as consequence of loss of solubility of bile constituents. Two types are recognized: cholesterol and bilirubin. Those of cholesterol are caused by supersaturation of bile (lithogenic) by cholesterol overproduction or bile salt deficiency. Bilirubin stones occur due to hemolysis (Sickle Cell, thalassemia) or bacterial infection of bile. Other etiologies include: *Ascaris Lumbricoides* infestation, drug-induced (Ceftriaxone), ileal resection, TPN, etc. Gallbladder sludge is a clinical entity that when it persists can be a predisposing factor for cholelithiasis and cholecystitis.

Laparoscopic Cholecystectomy (LC) has become the procedure of choice for the removal of the diseased gallbladder of children. The benefit of this procedure in children is obvious: is safe, effective, well tolerated, it produces a short hospital stay, early return to activity and reduced hospital bill. Several technical differences between the pediatric and adult patient are: lower intraabdominal insufflation pressure, smaller trocar size and more lateral position of placement. Complications are related to the initial trocar entrance as vascular and bowel injury, and those related to the procedure itself; bile duct injury or leak. Three 5 mm ports and one 10 mm umbilical port is used. Pneumoperitoneum is obtained with Veress needle insufflation or using direct insertion of blunt trocar and cannula. Cholangiography before any dissection of the triangle of Calot is advised by some workers to avoid iatrogenic common bile duct injuries during dissection due to anomalous anatomy, it also remains the best method to detect common bile duct stones. Treatment may consist of: (1) endoscopic sphincterotomy, (2) opened or laparoscopic choledochotomy, or (3) transcystic choledochoscopy and stone extraction. Children with hemolytic disorders, i.e. Sickle cell disease, have a high incidence of cholelithiasis and benefit from LC with a shorter length of postop stay and reduced morbidity.

Acalculous cholecystitis (AC) is more commonly found in children than adults. Two-third of cases appear as a complication of other illness: trauma, shock, burns, sepsis, and operative procedures. Contributing causes mentioned are: obstruction, congenital tortuosity or narrowing of the cystic duct, decreased blood flow to the gallbladder, and long-term parenteral nutrition. Males are more commonly affected than females. Fever, nausea, vomiting, diarrhea, dehydration and marked subhepatic tenderness are the most common symptoms. Other less common sx are jaundice, and abdominal mass. Labs show leucocytosis and abnormal liver function tests. Recently (APSA 95), two distinct forms of this disease have been recognized: acute, with symptom duration less than one month and chronic, with sx greater than three months. US is diagnostic by demonstrating hydrops of gallbladder, increase wall thickness and sludge. HIDA scan with CCK stimulation may help diagnose chronic cases. In both situations management consist of early cholecystectomy which can be executed using laparoscopic techniques.

#### **I. Idiopathic Perforation of Bile Ducts**

Spontaneous perforation of the common bile duct is the second cause of surgical jaundice in infants. The perforation is generally identified at the junction of the cystic and common bile ducts. Most infants develop slowly progressive bilious ascites, jaundice, and clay-colored stools. Other patients develop an acute bile peritonitis.

Diagnosis is by ultrasound or HIDA scan showing extravasation. Paracentesis confirms the nature of the ascitic fluid. Management consist of intraoperative cholangiogram (to demonstrate area of leak), and adequate simple drainage of area. Periportal inflammation precludes vigorous surgical efforts that could be disastrous. Tube cholecystostomy placement help for post-op follow-up studies. The leak generally seals spontaneously during the ensuing 2-3 weeks. Prognosis is good with no long term biliary sequelae.

#### **J. Splenic Cyst, Splenoptosis, Spherocytosis and Asplenia**

Splenic cysts in children are either considered true epidermal (congenital), pseudocysts (post-traumatic), or infectious (*Echinococcus*) in etiology. They are rare, benign, solitary cysts often producing few symptoms. They may present as a palpable mass in the left side of the abdomen or during evaluation for another abdominal problem. Ultrasound (large unilocular sonolucent cyst) is the most important diagnostic method, and can be supplemented by CT-Scan. The lining of the cyst is a flattened endothelium surrounded by fibrous tissue. This mesothelium can produce carcinoembryonic antigen (CEA). Indications for surgery are: (1) risk of complications (rupture, bleeding), (2) size greater than 5 cm., (3) infectious etiology, and a (4) symptomatic child (pain, mass or splenomegaly). Their management formerly total splenectomy has changed to: interventional sonography with fluid aspiration (catheter placement), or partial splenic decapsulation

(cystectomy); the result of recognition of the physiologic importance (hematologic and immunologic) of the spleen, together with the development of radiological imaging and operative surgery. Long term follow-up with radionuclide scans is recommended.

Splenoptosis (Wandering spleen) is a rare congenital fusion anomaly of the dorsal mesogastrium of the spleen that results in failure and laxity of its normal attachment to the diaphragm, retroperitoneum and colon. Relatively more common in children than adults, and females outnumber males. The child presents with an asymptomatic mass (splenomegaly), mass and subacute gastrointestinal complaints or with acute abdominal symptoms. These are the result of torsion of the pedicle, ischemia and splenic sequestration. 50% of spleens are lost to acute ischemia from torsion. Other complications are: pancreatitis, hypersplenism and cyst formation. Lab tests are nonspecific, but may occasionally reveal evidence of hypersplenism or functional asplenia. Diagnosis needs a high index of suspicion, and is achieved with: Ultrasound, CT, and Scintigram. Management consists of splenectomy for frank splenic infarct, or splenopexy for viable organs. Splenopexy is achieved by creating an extraperitoneal pocket or wrapping the spleen in absorbable mesh and anchoring to the retroperitoneum (splenic nood).

Hereditary spherocytosis is a clinically heterogeneous autosomal dominant red blood cell membrane disorder that causes anemia. The genetic defect results in deficiency of spectrin, the largest and most abundant structural protein of the erythrocyte membrane skeleton. The affected RBC loses its biconcave shape, strength and flexibility to the stress of circulation, becomes round and is trapped and destructs early in the spleen. Initial symptoms are those of pallor, jaundice and chronic anemia, followed by splenomegaly. Hemolytic crises are triggered by intercurrent infections. Pigment gallstones are common after the first decade of life. Labs' findings are those of many spherocytes in the peripheral smear, 8-10 mg% hemoglobin, elevated reticulocyte count, increase erythropoiesis in the bone marrow, and negative Coomb's test. Erythrocytes' shows increased osmotic fragility with autohemolysis in hypotonic solutions. Definitive therapy consists of splenectomy. This condition is the most common indication for elective splenectomy in children. The risk of overwhelming postsplenectomy sepsis makes it advisable to delay splenectomy until after six years of age unless the child becomes transfusion dependent. At the time of splenectomy, it is important to identify and remove accessory spleens. If gallstones are present, cholecystectomy should be done. A low content of spectrin and high percentage of microcyte has been used as determinants of early splenectomy as judge by the clinical severity of the disease process. Howell-Jolly bodies in erythrocyte are identified after total splenectomy.

The absence of the spleen (asplenia) occurs after surgical removal, following chronic conditions or congenital. Trauma is the most common cause of removing the spleen in children and sickle cell disease is the most common cause of functional asplenia in children. Congenital absence of the spleen is usually associated with serious malformations, primarily cardiovascular and abdominal heterotaxia. The spleen contributes importantly to the normal and pathologic removal of blood cells from the circulation and to defense against infection with encapsulated bacteria. Asplenia increases the risk of fulminant bacteremia (post-splenectomy sepsis) and mortality with these organisms. This risk is also increased by the underlying condition that caused the removal of the spleen, i.e., trauma, malignancy or hematologic disease. Several recommendations have been given when dealing with an asplenic individual. These are to vaccinate the child against pneumococcus (Pneumovax vaccine), hemophilus influenza type b and meningococcus. Regarding Pneumovax use revaccination after 3-5 years is recommended for children with asplenia who are 10 years of age or younger and for older children and adults who were immunized at least five years before. Duration of vaccine-induced antibodies is unknown but may be shorter than that in normal persons. Long-term antimicrobial prophylaxis is also used. This carries the problem of compliance and for how long. Significant febrile episodes should be managed aggressively, and probably most important, the patient and family should be carefully educated about this complication (name tag). Most deaths from hyposplenia-related septicemia are preventable.

#### **K. Splenic Trauma**

Spleen is the most common injured organ in blunt abdominal trauma. Hematologic and immunologic importance of the spleen has changed the attitude of trauma surgeons toward preservation of this organ whenever hemodynamics physiology permits. Massive hemorrhage (> 50 cc/kg weight) and hemodynamic

instability are indications for surgery. CT-Scan continues to be the choice of imaging during blunt abdominal trauma to establish the diagnosis of solid organ rupture in blunt abdominal trauma and rule out other major abdominal injuries. Isolated splenic rupture can be managed conservative in almost 80-90% of cases reducing complications and post-splenectomy sepsis. Low velocity of injury, thicker capsule, ribs elasticity and transverse nature of the laceration explain propensity for spontaneous healing in children. Associated lesions are not a contraindication for conservative management. Should conservative management fails the next step is splenography or splenectomy. Child is admitted to intensive care for 48 hours, followed by in-hospital observation until stable to be discharge home. Vaccination (pneumococcus, hemophilus and meningococci) affords added protection. Sonography is helpful for sequential splenic imaging to show when the appearance returns to normal, though clinical exam suffices. Participation in body contact sports should be curtailed for at least three months after injury.

#### **L. Adrenal Hemorrhage**

Retroperitoneal adrenal Hemorrhage usually occurs at birth or during the first postnatal days the result of traumatic or breech delivery, large fetal size, disorders of hemostasis, perinatal asphyxia and fetal hypoxia producing anemia from blood loss accompanied by an enlarging flank mass. Other times adrenal Hemorrhage manifests as unexplained hyperbilirubinemia with a mass in the flank in a healthy infant. Most cases affect the right adrenal gland (75%). Differential diagnosis includes neuroblastoma, cortical renal cysts, adrenal abscess, obstructed upper cortical renal cyst, and obstructed upper excretory tract in duplicated kidney. Diagnosis of neonatal adrenal hemorrhage is based on ultrasound (echo-free mass superior to downward displaced normal kidneys with linear calcifications). Fine needle aspiration can be done to confirm the diagnosis. The hematoma resolves gradually under supportive management by three months of age as documented by serial sonography. Lesion evolution with progressive decrease in its size and development of calcifications on repeated sonographic follow-up studies may be the only reliable sign in preventing unnecessary surgery.

## **VII. TUMORS**

### **A. Wilms Tumor**

This is the most common solid intra-abdominal tumor (malignant) in children. It affects 450-500 children annually in the USA. Neuroblastoma is most common, but they are not all confined to the abdomen. It has a peak incidence at 3-5 years of age. Present as abdominal or flank mass with abdominal pain, asymptomatic hematuria, and occasionally fever. Other presentations: malaise, weight loss, anemia, left varicocele (obstructed left renal vein), hypertension. Abnormalities associated with Wilms tumor include hemihypertrophy, pseudohermaphroditism, aniridia, Beckwith-Wiedemann syndrome, trisomy 18 and other genitourinary anomalies.

The initial evaluation consist of: abdominal films, ultrasound, IVP, urinalysis, and chest-X-rays and tomography. The presence of a solid, intrarenal mass causing intrinsic distortion of the calyceal collecting system is virtually diagnostic of Wilms tumor. Sonography can be of help to evaluate the IVC and renal veins (venous extension of the tumor). Metastasis most common in the lung and occasionally the liver.

Operation is for both treatment and staging to determine further therapy. The abdomen is explored by a large transverse incision and both kidneys are visualized. Nodes are biopsied to determine extent of disease.

Staging by National Wilms Tumor Study Group:

Group I- tumor limited to kidney and completely resected.

Group II- tumor extends beyond the kidney but is completely excised.

Group III- residual non-hematogenous tumor confined to the abdomen.

Group IV- hematogenous metastasis.

Group V- bilateral tumors.

Further treatment with chemotherapy or radiotherapy depends on staging and histology (favorable vs non-favorable) of tumor. Non-favorable histologic characteristics are: anaplasia (enlarged nucleus 3X, hyperchromatism, mitosis), sarcomatous or rhabdoid degeneration.

Disease-free survival is 95% for Stage I and approximately 77% for all patients. Poor prognosis for

those with lymph nodes, lung and liver metastasis.

Congenital Mesoblastic Nephroma presents in infants under 30 days of age (< 6 months), is commonly benign and invasive locally. Operative removal is curative, ruptured of tumor increases recurrences. Chemo, radiotx not indicated.

### **B. Neuroblastoma**

Neuroblastoma is the most common solid tumor of infancy and childhood. Most appear during the first five years of life; over half occur in children under 2 years of age. Two-thirds of children over 2 years of age have disseminated disease at presentation.

Neuroblastomas can occur at any site where neural crest tissue is found in the embryo and are derived from primordial neural crest cells and neuroblasts migrating from the mantle layer of the developing spinal cord into the sympathetic ganglion chain and the adrenal medulla. The etiology is unknown. About three-fourths of neuroblastomas arise in the abdomen; half of these originate in the adrenal gland. About 20% occur in the posterior mediastinum. Other uncommon sites include the pelvis (4%), and the neck (4%). It's a solid, highly vascular tumor with a friable pseudocapsule.

Staging:

Stage I- tumor limited to organ of origin.

Stage II- regional spread that does not cross the midline.

Stage III- tumor extending across the midline.

Stage IV- distant metastasis.

Stage IV-S patients with a small primary and metastases limited to liver, skin, or bone marrow without radiographic evidence of bone metastases.

The clinical presentation is an abdominal mass (50-75%), hypertension (25%), weight loss, diarrhea, fever, bone pain. Rare: "opso-myoclonus" (dancing eye syndrome), Horner's syndrome, Panda's eyes, VIP syndromes.

Diagnostic work-up includes: IVP, ultrasound, chest films, KUB (fine stipple calcifications 50%), skull x-rays, urinalysis, CBC, Urine VMA, HVA, and bone marrow aspirate. Other markers: cystathione, homoserine, neuron-specific enolase and ferritin.

The surgical goal is complete removal of the tumor when possible. Unfortunately, metastases are present in 60-90% of patients at diagnosis. Even in these patients attempts to reduce the bulk of tumor is important. Further treatment with radiation and chemotherapy depends on stage and extent of metastases.

There is a 100% survival for stage I, although this stage is extremely rare. Survival for stage II is 75%, stage III is 35%, stage IV 10-20%, and stage IV-S is about 80%. Age is an important prognostic factor, with 75% survival in children less than one year; 50% in children 1-2 years of age; 25% in children 2-3 years of age, and 15% in children over 3 years. Other prognostic factors are related to stage, nutritional status, site of primary, maturity state of tumor, VIP tumor (+), positive lymph nodes (-), high ferritin, NSE, and Diploid DNA levels(-).

Routine use of prenatal sonography will increase the incidental diagnosis of fetal neuroblastoma. Most are detected during the third trimester of pregnancy as cystic/solid suprarenal mass. The tumor does not cross the placenta but can metastasize in utero to the fetal liver or placenta. After birth 50% of babies have elevated HMA/VMA levels. Most enjoy improved survival due to: lower stage of disease, cystic variety (in-situ), and higher stage IV-S (which has been associated with spontaneous immuno-regression. Adverse biologic features are: diploid tumor karyotype (cytometry) and amplify N-myc oncogene. They can be very difficult to differentiate from neonatal adrenal hemorrhage; T2 of MRI can be of help. Are they neuroblastoma in-situ, and will they regress spontaneously without treatment are question waiting answer in the near future.

Neuroblastoma (NB) in early stages of development (stage I & II) benefits from surgical excision. The role of surgery in the management of neuroblastoma stage III tumor (tumor infiltrating across the midline with or without lymph node involvement) is controversial. Many variables enter the formula of determining risk of disease, i.e., age, site, stage, N-myc status, DNA diploidy and Shimada classification to mention a few of the most important. Some reports have independently found that stage III managed initially with chemotherapy and radiotherapy and is responding benefits from eventual complete tumor excision despite site, age or histology. Complete surgical excision as determine by free margin of tissue has a significant survival advantage overall.

Preop chemotx converts a friable tumor into a firmer, more mature and easily resectable tumor. Surgical complications in advance stages are higher (bleeding, nephrectomy, adjacent organ removal, infection). Some have found that complete resection is not needed in biologically favorable children with NB less than one year of age. Biologically unfavorable patients one year of age or greater who undergo gross surgical resections has improved survival. Defining subgroups of patient with poor prognostic biologic markers and histology to decide whether surgery or bone marrow transplant is the next best option is pending trial randomization and study.

Stage IV Neuroblastoma (metastatic NB) refers to high risk group of children with the primary tumor in the adrenal gland, mediastinum or pelvis associated with disease progression in other sites (bone marrow, cortical bone, liver, lymph node). Role of surgery in stage IV NB is controversial. Cure will require control of the primary tumor and elimination of metastatic disease. For infants with metastatic NB a more than 95% resection has been found adequate surgical treatment either initially or after effective chemotherapy. Adding ipsilateral lymph node dissection does not appear to affect survival. Delayed surgery after several courses of chemotherapy may be as effective as initial resection and is associated with fewer complications statistically. Resection without induction chemotx results in significant blood loss. High risk NB usually invades blood vessels and surrounding structures precluding resection. Intensive preop chemotherapy reduces tumor size and invasiveness allowing surgical removal. A fibrotic capsule forms with less blood supply to the tumor. Stage IV NB is best managed with initial chemotx until distant metastasis are controlled followed by primary gross tumor removal (even in the face of significant tumor reduction) and completion chemotx. Gross complete resection is best accomplished when a good partial response is obtained. Radiotx is added to unresectable lesions. Even when chemotx changes the tumor histology (Shimada) from unfavorable to favorable this does not improve overall outcome. Resection is not confounded by biology of the tumor (n-myc status). Survival is improved with kidney preservation during surgery. Local control of disease is a prerequisite for successful bone marrow transplantation.

### **C. Rhabdomyosarcoma**

Most common soft tissue sarcoma in infants and children and represents about 5-15% of all solid malignant lesions. It has a peak incidence at age 2-5 years. Second surge between 10-15 years of age. Tumors of the pelvic organs and head and neck are more prevalent in infancy and early childhood, while the paratesticular rhabdomyosarcomas are largely a disease of adolescents and young adults.

Although classically described as occurring in striated muscle, rhabdomyosarcomas arise from a primitive cell type and occur in mesenchymal tissue at almost any body site (possibly excluding the brain), including many organs that normally do not have striated muscle. The predominant histologic type in infants and small children is embryonal. The botryoid rhabdomyosarcoma is a subtype of the embryonal variety, which ordinarily extends into body cavities such as the bladder, nasopharynx, vagina, or bile duct. The alveolar cell type, named for a superficial resemblance to the pulmonary alveoli, is the most common form found on the muscle masses of the trunk and extremities, and is seen more frequently as age advances.

The clinical findings, diagnostic evaluation and therapy employed are dependent upon the location of the primary tumor and is beyond the scope of this review. In brief, head and neck tumors are most common and occur in the orbit, nasopharynx, cheek, neck, middle ear, larynx and paranasal sinuses. Most are treated by simple biopsy followed by combined therapy or preoperative chemotherapy and radiation followed by conservative resection. Operations for extremity lesions include wide local excision to remove as much of the gross tumor as possible. Rhabdomyosarcomas can arise from the bladder, prostate, uterus, or vagina. The trend in treatment is more chemotherapy and conservative surgical management.

### **D. Liver Tumors**

Hepatoblastoma and hepatocellular carcinoma are the most common malignant tumor of liver. These represent about 2% of all malignancies in childhood and 15% of malignant abdominal masses.

Hepatoblastoma (HB) is the most common primary malignant neoplasm of the liver in children mostly seen in males less than four year of age. Diagnostic work-up (US, Scintigraphy, CT-Scan) objective is predicting resectability and tumor extension. Diagnostic laparotomy will decide resectability. Markers associated to this tumor are: alpha-fetoprotein and gamma-glutamyltransferase II. Only reliable chances of cure is surgical excision although half are unresectable at dx. Unresectable tumors can be managed with preop chemotx.

Disadvantages of preop chemotx are: progressive disease, increase morbidity, post-op complications, and toxicity. Advantages are: decrease in tumor size, convert three-fourth cases into resectable, although extent of surgery is not decreased. Tumor necrosis is more extensive in pt. receiving preop chemotx. Osteoid present in tumors after chemotx may represent an inherent ability of the tumor to mature and differentiate. Diploid tumors on DNA flow cytometry show a better prognosis.

Hepatocellular carcinoma in childhood is histologically identical to hepatoma seen in the adult and is associated 50% of the time to a prior liver disorder (i.e. tyrosinemia, hepatitis type B, etc.).

Associated anomalies and conditions are: hemihypertrophy, osteoporosis, lipid storage disease, glycogen storage disease, virilization in males. Clinical presentation is asymptomatic abdominal mass, with abdominal pain and weight loss in 25% of patients.

Diagnostic work-up includes: alpha-fetoprotein, chest x-ray, abdominal films, IVP, ultrasound, liver-spleen scan, CAT scan, and occasionally arteriogram.

Hepatic resection has provided the only cures. In patients with initially unresectable tumor or in post-resection patients, chemotherapy is employed. Among those patients in whom the entire tumor can be resected, survival is 80% at two years. Unresectable tumors have a dismal prognosis. ADR (adriamycin) is the principal chemotherapeutic agent.

#### **E. Teratomas**

Teratomas contains tissues derived from the three embryonic layers (endoderm, mesoderm, and ectoderm), found in a locus that does not normally harbor such tissues. It is not always possible to find tissue in each teratoma that is derived from all three embryonic layers.

Sacroccocygeal teratoma (SCT) is the most common extragonadal germ cell tumor in neonates with an incidence of one in 30-40,000 live births. Three-fourth are females. SCT present as a large, firm or more commonly cystic masses that arise from the anterior surface of the sacrum or coccyx, protruding and forming a large external mass. Histology consist of tissue from the three germ cell layers. SCT is classified as: mature, immature, or malignant (endodermal sinus) and produces alpha-feto protein (AFP). Prenatal sonographic diagnostic severity criteria are: tumor size greater than the biparietal diameter of the fetus, rapid tumor growth, development of placentomegaly, polyhydramnios and hydrops. Large tumors should benefit from cesarean section to avoid dystocia or tumor rupture. Management consist of total tumor resection with coccyx (recurrence is associated with leaving coccyx in place). Every recurrence of SCT should be regarded as potentially malignant. Malignant or immature SCT with elevated AFP after surgical resection will benefit from adjuvant chemotherapy. Survival is 95% for mature/immature tumors, but less than 80% for malignant cases. Follow-up should consist of (1) meticulous physical exam every 3-6 months for first three years, (2) serial AFP determination, (3) fecal/urodynamic functional studies. Long term F/U has found a 40% incidence of fecal and urinary impairment associated to either tumor compression of pelvic structures or surgical trauma.

#### **F. Ovarian Tumors**

Ovarian tumors are uncommon childhood malignancies (1%) characterized by recurrence and resistance to therapy. Aggressive surgery is limited to avoid compromising reproductive capacity and endocrine function. Low incidence and need of multinodal therapy encourages referral to centers dealing with effective cancer therapy. The most common histology is germ cell: dysgerminoma, teratoma, and endodermal sinus tumor. This is followed by the sex-cord stroma tumors with a low incidence of malignancy. They can cause feminization (granulosa-theca cell) and masculinization (androblastoma). Other types are: epithelial (older adolescent), lipid-cell, and gonadoblastoma. Ovarian tumors present with acute abdominal symptoms (pain) from impending rupture or torsion. They also cause painless abdominal enlargement, or hormonal changes. Preop work-up should include: human chorionic gonadotropin (HCG) and alpha-fetoprotein (AFP) levels. Imaging studies: Ultrasound and CT-Scan. The most important prognostic factor in malignant tumors is stage of disease at time of diagnosis. Objectives of surgery are: accurate staging (inspection of peritoneal surfaces and pelvic organs, lymph node evaluation), washing and cytology of peritoneal fluid, tumor removal, and contralateral ovarian biopsy if needed. Chemotherapy consists of: bleomycin, cis-platinum, and vinblastine. Radiotherapy is generally not effective, except in dysgerminoma. Elevation of tumor markers (AFP or HCG) after therapy signals recurrence..

### **G. Thyroid Cancer**

In spite of presenting with advanced, multicentric and larger tumors children have a better survival than adults. Populations at risk: past radiation to head and neck, nuclear waste radiation, MEN II kindred. Clinical presentation is a solitary cervical mass or metastatic lymph node. Diagnostic work-up should include: sonogram (cystic or solid), thyroid scan (cold or hot), Fine-needle aspiration cytology(FNA), and Chest-X-Ray (lung metastasis 20% at dx). Pathology of tumors: papillary (majority, psammomas bodies), follicular (vascular or capsular invasion), medullary (arise from C-cells, multicentric, locally invasive), anaplastic (rare, invasive and metastatic).

Management is surgical. Complications of surgery increase with decreasing age of patient: temporary hypoparathyroidism, recurrent nerve injury. Prognostic factors associated to higher mortality are: non-diploid DNA, psammomas bodies, over 2 cm diameter nodule, and anaplastic histology. Follow-up for recurrence with serum thyroglobulin level and radioisotope scans. Adjunctive therapy: thyroid suppression and radio-iodine for lymph nodes and pulmonary metastasis.

### **H. Burkitt's Lymphoma**

Burkitt's lymphoma (BL) is a highly malignant tumor first described during the late 50's in African children (jaw), endemic in nature, and composed of undifferentiated lympho-reticular cells with uniform appearance. The American BL variety is non-endemic, mostly attacks children between 8-12 years of age, predominantly (>75%) with abdominal disease such as unexplained mass, pain, or intussusception. The head and neck region follows. The tumor can appear as a localized, diffuse (multifocal, non-resectable) or metastatic abdominal mass (bone marrow and CNS). It's considered the fastest growing tumor in humans with a doubling time around 12-24 hrs. Chemotherapy is the primary treatment modality due to its effectiveness in rapidly proliferating cells. The role of surgery is to establish the diagnosis (using open biopsy), stage the tumor, remove localized disease, relieve intestinal obstruction and provide vascular access. Complete resection whenever possible offers the patient improved survival. Is more readily accomplished in patients with localized bowel involvement operated on an emergency basis due to acute abdominal symptoms. The only predictor of event free survival is extent of abdominal disease at diagnosis. Debulking (cytoreductive) procedures increases morbidity and delays initiation of chemotherapy worsening prognosis. Extensive tumors should be managed with minimal procedure and immediate chemotherapy (a/o radiotherapy). Bone marrow and CNS involvement are ominous prognostic signs.

## **VIII. GYNECOLOGIC and INTERSEXUAL CONSIDERATIONS**

### **A. Labial Adhesions in Infants**

Minor labial adhesions is a common pediatric gynecologic problem occasionally confused with imperforate hymen. Most cases are in children 2-6 y/o and involve labial adhesions secondary to diaper rash. The process causing fusion is a natural one: two normally covered surfaces with squamous epithelium in contact with each other is traumatized eventually forming a fibrous tissue union (agglutinate) between them when healing occurs. A small opening near the clitoris is always present through which urine escapes. This seldom causes symptoms except recurrent UTI if it covers the urethral meatus. Treatment consists of applying estrogenic creams (0.1%) for two weeks. Manual separation can be painful and adhesion recurs. Unless the urethral meatus is covered, there is no reason to be further aggressive in management. Prolonged use of estrogenic cream can cause precocious isosexual development.

### **B. Ovarian Cysts**

Ovarian cysts in fetus and infants are usually follicular in nature and less than 2 cm in size. They are commonly diagnosed between the 28th and 39th wk. of gestation by sonography. Hypotheses on etiology are: (1) Excessive fetal gonadotropic activity, (2) enzymatic abnormalities of the theca interna, and (3) abnormal stimulation by the mother HCG. Obstetric management consists on observation and vaginal delivery. After birth, diagnostic assessment and management will depend on the size and sonographic characteristics of the cyst. Simple anechoic cysts, and those less than 5 cm in size can be observed for spontaneous resolution. Cyst with fluid debris, clot, septated or solid (complex nature), and larger than 5 cm should undergo surgical excision due to the higher incidence of torsion, perforation and hemorrhage associated to them. Percutaneous

aspiration of large simple cysts with follow-up sonography is a well-accepted therapy, preserving surgery for recurrent or complicated cases. Surgical therapy is either cystectomy or oophorectomy that can result in loss of normal ovarian tissue.

### **C. Breast Disorders**

Most breast disorders in children of either sex are benign. Congenital lesions are: absent or multiple breast. Transplacental hormonal influence in neonates may cause hyperplasia of breast tissue with predisposition to infection (Mastitis neonatorum). Premature hyperplasia (thelarche) in females is the most common breast lesion in children. It occurs before the age of eight as a disk-shaped concentric asymptomatic subareolar mass. Remains static until changes occur in the opposite breast 6-12 mo later. It can regress spontaneously or stay until puberty arrives. Biopsy may mutilate future breast development. On the contrary, discrete breast masses in males cause concern and excision is warranted. Gynecomastia is breast enlargement cause by hormonal imbalance, usually in obese pre-adolescent boys. If spontaneous regression does not occur, it can be managed by simple mastectomy. Virginal hypertrophy is rapid breast enlargement after puberty due to estrogen sensitivity. If symptomatic, management is reduction mammoplasty.

Breast enlargement is commonly seen in newborns babies, a condition associated with clear or milky nipple discharge. Maternal hormones are considered the culprit. On rare occasions the mother of an infant will bring to you the attention that the child is having intermittent episodes of bleeding through the nipple. In infants it is a benign, self-limited condition that should be managed conservatively (it could take six months to go away). The main reason of bloody nipple discharge is mammary ductal ectasia, which extends down to the collecting tubules. Mammary duct ectasia was first reported in 1983, characterized by dilatation of the subareolar duct system, and by inflammatory reaction and fibrosis. The infant's own endocrine system is responsible for breast enlargement and mammary duct ectasia, though infection has also been postulated as etiologic factor. Surgical procedures should be avoided, because injury to the breast bud may cause permanent damage. Stimulation or massages to the breast should also be avoided. When associated with hypertrophied mammary glands, prepubertal gynecomastia should be sought.

### **D. Congenital Adrenal Hyperplasia**

Congenital adrenal hyperplasia (CAH) involves a functional defect in any of the five enzymatic steps required for cortisol synthesis, most commonly 21- (involved in 90-95% of cases) and 11-hydroxylase level. This primary genetic defect transmitted as autosomal recessive impairs the ability of the adrenal cortex to synthesize cortisol causing increase feedback secretion of ACTH and adreno-cortical hyperplasia of the gland. Increase output of steroids proximal to the block (androgenic precursors) causes virilization in affected males and females. Its more severe form is associated with aldosterone deficiency and life-threatening salt wasting. Female pseudohermaphrodite due to virilizing CAH is the most frequent form of intersexuality found. The phenotypic picture varies from mild clitoral enlargement alone to complete masculinization of the urethral meatus at the tip of the penis. Prenatal diagnosis (southern blotting of DNA) is based on finding the disease gene on the short arm of chromosome 6. Likewise management in the mother (dexamethasone) is started empirically until the affected status is known by chorionic villus sampling. After birth management consists of cut-back or flap vaginoplasty with clitoral recession at 3-6 months of age. Children with high vaginal entry proximal to the urethra external sphincter can undergo early one-stage reconstruction at 8-12 months of age. Long term surgical results of female children show adequate sexual identification, reproduction, intellectual functioning and acceptable genitalia.

### **E. Testicular Feminization Syndrome**

Testicular feminization syndrome (TFS) is a genetic form of male pseudohermaphroditism (patient who is genetically 46 XY but has deficient masculinization of external genitalia) caused by complete or partial resistance of end organs to the peripheral effects of androgens. This androgenic insensitivity is caused by a mutation of the gene for androgenic receptor inherited as an X-linked recessive trait. In the complete form the external genitalia appear to be female with a rudimentary vagina, absent uterus and ovaries. The infant may present with inguinal hernias that at surgery may contain testes. Axillary/pubis hair is sparse and primary amenorrhea is present. The incomplete form may represent undervirilized infertile men. Evaluation should include: karyotype, hormonal assays, pelvic ultrasound, urethrovaginogram, gonadal biopsy and labial skin bx



for androgen receptor assay. These patients will never menstruate or bear children. Malignant degeneration (germ cell tumors) of the gonads is increased (22-33%). Early gonadectomy is advised to: decrease the possible development of malignancy, avoid the latter psychological trauma to the older child, and eliminate risk of losing the pt during follow-up. Vaginal reconstruction is planned when the patient wishes to be sexually active.

#### **F. Mixed Gonadal Dysgenesis**

Mixed gonadal dysgenesis (MGD) is an intersexual genetic abnormality caused by a defect in the sex chromosomes (gonosomes) associated with dysgenetic gonads and retained Müllerian structures. The most common gonosomal aberration in MGD is 45 X0/46 XY mosaic karyotype. The external genitalia could be normal looking female and these children will present later in life with primary amenorrhea. Otherwise, it could be ambiguous: clitoromegaly and urogenital sinus to a sizable phallus with hypospadias. A uterus and one or both fallopian tubes may also be present. MGD is characterized by a streak gonad and a contralateral testis (that is typically cryptorchid) or bilateral streak testes. The testis might show prepubertal tubules lined by a few spermatogonia and immature Sertoli cells. Female gender assignment is usually preferred, but male assignment is an alternative in instances of extreme virilization. Dysgenetic gonads with the presence of a Y chromosome or a translocated fragment have a significant risk of developing malignant gonadoblastoma (though seminoma and dysgerminoma can occur). Routine early bilateral gonadectomy is advised in MGD. The child to be raised as a female will

need clitoral recession and vaginoplasty in early infancy. If it is to be raised as male, then various types of hypospadias repair can be done, gonads can be replaced with prostheses, the prepenile scrotum reconstructed and Müllerian structures removed.

#### **G. Müllerian Duct Syndrome**

Müllerian Duct Syndrome (MDS) refers to a genetic disorder of male pseudohermaphroditism (46 XY karyotype) characterized by normal masculinization of the external genitalia and the presence of uterus and fallopian tubes. Most cases are discovered during surgery for undescended testis, inguinal hernia or transverse testicular ectopia. MDS is caused by a deficient activity (most cases) or receptor insensitivity of anti-müllerian hormone (also known as Müllerian inhibitor factor). This hormone is produced by testicular Sertoli cells and is responsible of producing fetal regression of Müllerian structures (uterus & fallopian tubes) in genetic males. Most cases are transmitted as autosomal recessive restricted to males (sex-linked). Anatomic variants include fallopian tube or uterus within the inguinal canal, testis and tubes in a hernia sac or bilateral cryptorchidism with the testes embedded in the broad ligaments. The vas deferens is intimately adhered to the uterus lateral wall. Initial procedure consists of hernia repair, replacement of structure within pelvis and karyotype. After diagnosis follow-up management has been controversial. A few suggest partial removal of the uterus (leaving vas deferens intact on a thin pedicle of myometrium) and fallopian tubes with testicular fixation. Most content that surgical excision of persistent MDS structure may result in ischemic or traumatic damage to the vasa deferentia and testes and optimal management is orchiopexy leaving the uterus and fallopian tubes in situ. The testis in MDS are at risk of malignant degeneration.

#### **H. Hydrometrocolpos**

Hydrometrocolpos (HMC) is accumulation of secretions in the vagina and uterus caused by one of two mechanisms: 1- excessive intrauterine stimulation of the infant's cervical mucous glands by maternal estrogen (secretory HMC), or 2- accumulation of urine (urinary HMC) in the presence of a vaginal obstruction. HMC can arise from congenital or acquired pathology. Acquired causes include vaginitis from Diphtheria or measles, senile vaginitis, from radiation therapy and corrosive vaginitis. Congenital vaginal obstruction causing HMC is due to imperforate hymen, transverse vaginal septum and persistence of urogenital sinus with complete distal vaginal obstruction. The child may present with a lower abdominal mass from the dilated vagina and uterus, urogenital sinus, obstructive uropathy (hydronephrosis), dribbling, respiratory distress, bowel obstruction and lower extremity venous congestion. HMC usually occurs in the neonatal period and the majority of cases are caused by vaginal occlusion by a transverse septum combined with cervical secretion. HMC can be associated with congenital adrenal hyperplasia when there is a long urogenital sinus. Physical exam (obvious vagina septum of imperforate hymen or a urogenital sinus), US (large cystic anechoic mass, anteriorly compress bladder and fluid-debris level), voiding cystogram, and sinoscopy can establish the cause of HMC. Vaginal

decompression by catheter placement, endoscopic septotomy or vaginostomy is done initially followed later by opening of septum or vaginal pull-through.

## **IX. PRENATAL CONGENITAL MALFORMATIONS**

Poor survival with neuroblastomas, diaphragmatic hernias and necrotizing enterocolitis requires efforts during the next few years to reduce mortality rates. These areas will require extensive investigation as to etiology, unique characteristics and better management.

### **A. Fetal Surgery**

Certain lesions such as hydrocephalus, hydronephrosis and diaphragmatic hernias may benefit from intrauterine correction.

### **B. Fetal Intestinal Obstruction**

The fetal gastrointestinal tract (foregut, midgut and hindgut) undergoes ventral folding between 24-28 days' gestation. By the 5-6th wk the stomach rotates to the right and the duodenum occludes by cell proliferation. Recanalization of the duodenum occurs around the 8th wk. The midgut rotation takes place during the 6-11th wk and the final peritoneal closure by 10th wk. The fetal GI tract begins ingestion and absorption of amniotic fluid by the 14th wk. This fluid contributes to 17% effective nutrition; proximally obstructed gut can cause growth retardation. Fetal intestinal obstruction is caused by: failure of recanalization (duodenal atresia), vascular accidents (intestinal atresias), intrauterine volvulus, intussusception, or intraluminal obstruction (meconium ileus). Esophageal obstruction causes polyhydramnios, absent visible stomach and is related to tracheo-esophageal anomalies. Duodenal obstruction seen as two anechoic cystic masses is associated to aneuploidy (trisomy 21) and polyhydramnios. Jejuno-ileal obstruction produces dilated anechoic (fluid-filled) serpentine masses and bowel diameter of 1-2 cm. Large bowel obstruction is most often caused by meconium ileus, Hirschsprung's disease or imperforate anus. The colon assumes a large diameter and the meconium is seen echogenic during sonography. In general the method of delivery is not changed by the intrauterine diagnosis of intestinal obstruction. Timing can be affected if there is evidence of worsening intestinal ischemia (early delivery recommended after fetal lung maturity).

### **C. Fetal Abdominal Wall Defect**

Most common abdominal wall defects (AWD) are gastroschisis, omphalocele and hernia of the umbilical cord. Referral to tertiary centers with available neonatal intensive care is necessary in prenatally diagnosed cases. Changing the route of delivery does not affect outcome for either defect. Omphalocele has a high incidence of associated anomalies (cardiac, neurogenic, genitourinary, skeletal, chromosomal syndromes) that are the cornerstones of mortality. Detailed search for associated anomalies, fetal echocardiogram and karyotyping should be performed always. Cesarean section is justified in large omphaloceles (> 5 cm) to avoid liver damage, sac rupture and dystocia. Gastroschisis prenatal US appearance depends on gestational age and condition of extruded bowel. Fetal karyotyping testing is less important. Intestinal atresia complicates the defect, the result of an intrauterine vascular accident. Intestinal obstruction due to atresia or luminal constriction may cause polyhydramnios, fetal growth retardation and preterm labor, findings that can be monitored with serial US. No benefit has been found in recommending routine c-section for most cases of gastroschisis. Preterm deliveries by c-section have been found to prevent bowel damage in fetus with progressive bowel dilatation and thickening, a finding that has not been corroborated by others. Abnormal US appearance of fetal bowel is associated with more bowel edema, longer operative time and a higher incidence of postoperative complications.

## **X. SUGGESTED READING**

### **Specific Readings**

- 1- Lugo-Vicente HL, Pediatric Surgery Update, Monthly Newsletter, San Juan, Puerto Rico. Vol 1-15, 1993, 1994, 1995, 1996, 1997, 1998, 1999, 2000.
- 2- Coran AG: Vascular Access and Infusion Therapy. Seminars in Pediatric Surgery 1(3): 173-241, 1992
- 3- Altman RP: Liver and Biliary Tract Diseases. Seminars in Pediatric Surgery 1(2):97-171, 1992

4- Gans SL, Grosfeld Jay L: Current Concepts of Pediatric Oncology. Seminars in Pediatric Surgery 2(1):1-82, 1993

5- Lugo-Vicente HL. Colangiopatías Infantiles. En: José Ma. Valoria Villamartín, editor. Cirugía Pediátrica. Madrid: Diaz de Santos, 1994; pags. 210-220

### General Reading

1- Welch KJ, Randolph JG, Ravitch MM, O'Neill JA, Rowe MI. Pediatric Surgery. 4th edition. Chicago. Year Book Medical Publishers. 1986

2- Ashcraft KW, Holder TM. Pediatric Surgery. 2nd edition. Philadelphia. W.B. Saunders Co. 1993

3- Grosfeld JL. Common Problems in Pediatric Surgery. 1st edition. St Louis. Mosby Year Book. 1991

4- Ashcraft KW, Holder TM. Pediatric Esophageal Surgery. 1st edition. Orlando. Grune & Stratton, Inc. 1986

5- Seeds JW, Azizkhan RG. Congenital Malformations: Antenatal Diagnosis, Perinatal Management and Counseling. 1st edition. Maryland. Aspen Publishers, Inc. 1990

6- Puri P (ed): Congenital Diaphragmatic Hernia. Mod Probl Paediatr. Basel, Karger, Vol 24, 1989

7- Pediatric Surgery Update <<http://home.coqui.net/titolugo/>> © 1993-2000

\*\*\*\*\*

### IMAGES

The following images can be obtained at URL: <<http://home.coqui.net/titolugo/diag.htm>>

1- KUB of **prepyloric antral membrane**

2- Esophagogram of **Achalasia**

3- US and HIDA of **choledochal cyst**

4- **Gastroschisis**

5- **Omphalocele**

6- CT Scan of **gastric duplication cyst**

7- Barium enema of **left hypoplastic colon syndrome**

8- Physical exam findings and CT Scan of **imperforate hymen**

9- KUB and operative findings of **Meconium Ileus**

10- KUB of a **duodenal stenosis**

\*\*\*\*\*

\* Address reprints requests to:

**Humberto L. Lugo-Vicente, MD**

**P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico, 00922-0426.**

**Tel (787)-786-3495 Fax (787)-720-6103**

**e-mail: titolugo@coqui.net**

**Internet: <http://home.coqui.net/titolugo>**

**Private Practice: Suite 309, Edificio Médico Santa Cruz,**

**Santa Cruz # 73, Bayamón, Puerto Rico 00961**

\*\*\*\*\*

Revised: April 2002  
PSU © 1996-2002