PEDIATRIC SURGERY HANDBOOK

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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 is: 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 amplifies 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 require 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. Newborn 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 heal 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. Infant 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 adult’s levels. C3, C4, C5 complex, factor B, and properdin concentration is 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 inmaturation 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 impose 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, nor-inflamed, positive skin test, excision is curative, chemotx is of no value. (2) Cat-Scratch adenitis- caused by *A. Fellis*, 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 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 stepladder 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 chromosomic 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.

G. Parotid Mass

A parotid mass in a child creates great concern and should be managed promptly since a high percentage of cases will harbor a malignant tumor. The more common benign parotid tumor in children includes hemangioma, pleomorphic adenoma and lymphangiomas. Infants with a rapidly enlarging violaceous or soft tumor in the parotid region harbor a hemangioendothelioma. Hemangioendothelioma is the most common parotid gland tumor of childhood. They seldom need excision as spontaneous regression is the norm. An asymptomatic, slow growing solid mass is the most common presentation in older children. Tenderness is associated with an infectious process. Diagnosis includes the use of Doppler ultrasound, CT-Scan, MRI and fine needle aspiration (FNA) biopsy. Salivary gland carcinoma is rare in children. Most common histological type is mucoepidermoid either as a primary neoplasm or secondary malignancy after neck irradiation. Management consists of superficial or total parotidectomy extent which is selected during the surgical procedure depending on deep gland or fascial nerve involvement. The tumor must be widely excised. Recurrence is managed with postoperative irradiation for high or intermediate grade malignancies. Rhabdomyosarcoma arising in the parotid gland area is another locally invasive aggressive tumor that presents early with swelling and symptoms of seventh nerve deficit. This tumor needs surgical excision followed by chemotherapy and irradiation.

Acute parotitis is a self-limiting disease most commonly associated with mumps (epidemic parotitis) in children. Other times the parotitis is associated with bacterial infection progressing to frank suppuration. Recurrent parotitis, also known as juvenile recurrent parotitis, is characterized by a cyclic swelling of the parotid glands associated with discomfort and/or pain in the absence of external inflammatory changes during a period of several years. The condition mainly affects children between the ages of three and six, males being more commonly affected. The symptoms peak in the first year of school and usually begin to subside after puberty. Retrograde infection induced by the mumps virus and upper respiratory infection play a major role in the etiology of recurrent parotitis. Sialography demonstrates sialectasia. Children with recurrent parotid swelling needs to be screened for underlying systemic immune disorders such as Sjögren’s syndrome. With

time the recurrent episodes reduce salivary flow, while increasing the chloride, sodium, copper, albumin, IgA and lactoferrin concentration. Etiology of juvenile recurrent parotitis is a combination of congenital malformation of portions of the salivary ducts and a set-in infection. Treatment is conservative.

H. Hashimoto Thyroiditis

Hashimoto thyroiditis (HT) is a chronic lymphocytic autoimmune thyroiditis seen with some frequency in adolescent females and children. Most common cause of asymptomatic enlargement of the thyroid gland in children in iodine-sufficient geographic regions. Thyroid cell damage in HT is caused by antithyroid antibody-dependent cell-mediated direct toxicity linked to deficiency in antigen-specific suppressor T lymphocytes. The gland shows lymphocyte infiltration with follicular cell hyperplasia. Thyroid antibodies are elevated. Radionuclear scans show absent uptake. Initially the child develops elevated thyroid hormones (T3 and T4) followed by symptomatic hypothyroidism. Following the hypothyroid phase there is final recovery in most patients. Indications for surgery in HT include: 1- firm enlargement of the gland causing tracheal compression with dyspnea, hoarseness or swallowing difficulties, 2- failure to respond to suppressive therapy and development of symptomatic hyperthyroid goiter, and 3- development and enlargement of a solitary thyroid nodule. The incidence of malignancy in HT is low. Differentiating a hyperplastic follicular cell nodule from a follicular neoplasm is very difficult using fine needle aspiration biopsy. Patient with malignant nodules in Hashimoto glands are most commonly papillary, females, low frequency of extrathyroidal invasion and nodal metastasis with absent distal metastasis. It is believed the lymphocytic infiltration of HT causes a form of immune reaction to control tumor growth and proliferation.

I. Parotid Hemangioma

Parotid hemangioma (or hemangioendothelioma) is by far the most common tumor of the parotid gland seen in infants and children. Initially the infant presents with non-tender swelling of the cheek during the first weeks of life. The swelling is generally confined to the superficial lobe of the parotid gland, but it can involve the masseter muscle. With capillary and bluish involvement of the skin and subcutaneous tissue the diagnosis is easier to establish. MRI is the investigation of choice because of picture quality, definition of soft tissues and lack of exposure to ionizing radiation. MRI allows a definite diagnosis to be made without any invasive procedure being required. When in doubt a fine-needle biopsy will establish the histologic nature of the mass. US with Doppler imaging (lobular internal structure, fine echogenic internal septations, mildly lobulated contour and extremely high vascularity), and labeled red cell scintigraphy (well-defined area of intense activity) can also sustain the diagnosis of parotid hemangioma. Management is conservative since most lesions involute spontaneously. During involution ulceration and calcification can occur. Medical management (intralesional injection of steroids, systemic steroids or interferon) is given when the tumor is large, deforming, ulcerated, or involves nearby structures with functional consequences. The overall response rate is very high.

J. Congenital Tracheal Stenosis

Congenital tracheal stenosis (CTS) is a rare condition seen immediately after birth or in early infancy that is uniformly life-threatening. Infants with CTS presents with stridor, respiratory distress, recurrent pulmonary infections or failure to thrive. Inflammation of the mucosa or mucous accumulation can easily obstruct the already stenotic airway. The stenosis includes a short or long segment of circular cartilaginous ring. Diagnosis is established using bronchoscopy, MRI (assessment of vascular structures and relation to the stenosis), or CT-scan (good anatomic delineation of the airway). Each individual malformation is studied using rigid tracheo-broncho- esophagoscopy. Associated cardiac defects should rule out with echocardiogram. Management of CTS is surgical. Selection of the type of treatment depends on the patient's clinical status and the anatomic pattern of the stenosis. Resection of a short stenosis with anastomosis can be possible with a length that does not exceed half of the trachea. For longer stenosis the most useful technique consists of enlargement tracheoplasty with cartilaginous or a pericardial graft or the more recent and slide-tracheoplasty. The latter technique is preferable because it preserves native tracheal tissue with fewer postop complications. For CTS one should always look for other associated thoracic malformations, such as a pulmonary sling which may compromise the results of the surgical correction of the tracheal stenosis. Best prognosis is obtained with simultaneous correction of the respiratory and cardiovascular malformation. Overall survival of these children is 75%.

III. OBSTRUCTIVE PROBLEMS

LOGICAL APPROACH TO NEONATAL INTESTINAL OBSTRUCTION

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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 structures 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, hypospadia, 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.

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 populations 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. Barium swallows 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 procedure 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 consists 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 consists 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 consists of a careful gastrografin enema after the baby is well-hydrated. Gastrografin 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 classify 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. It's 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 unprepped 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 consists 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 a 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 approach 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 females' patient with imperforate anus (IA) is vestibular fistula, followed by vaginal fistulas. In more than 90% of females 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 accompany 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 frequents episodes of urinary tract infections shows 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, twining, 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 suspect 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 water soluble contrast colon enema. Hydrostatic reduction of the intussusception with the contrast material is successful in approximately 50% of cases. To be successful, the water soluble material must reflux into the terminal ileum. The surgeon should be notified before an attempt at hydrostatic reduction. 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 and appendectomy is added as cecopexy and avoidance of future diagnostic problems of a RLQ incision. 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 accumulate and acutely distend 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. Urinalysis 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 and 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.

In the event of right lower abdominal pain caused by appendicitis, almost 15% of children will present with a right lower quadrant mass effect. The mass can be a phlegmon with a central inflamed appendix which appears after four to five days from the onset of abdominal pain accounting for 50% of appendiceal masses. Almost 20% of these masses will contain a frank abscess cavity. Periappendiceal masses are the result of perforated appendicitis. Due to the increase use of abdominal CT in the setting of abdominal pain more cases of periappendiceal phlegmon/abscess are discovered with this diagnostic modality. CT is reliable in distinguishing periappendiceal abscesses from phegmons and its use can be extended into percutaneously draining the abscess cavity. US with color Doppler demonstrate a hyperemic periappendiceal or pelvic fluid collection and periappendiceal soft-tissue hyperemia. Management of a periappendiceal abscess depends on the clinical condition of the child. Nontoxic patient with mild peritoneal signs warrants non-operative management with intravenous fluids and antibiotics until the acute process subsides. Rising heart rate, continued spiking fever, worsening peritoneal signs, intestinal obstruction or enlargement of the mass may require urgent surgery. Patients undergoing initial nonoperative management have a lower rate of complications. Six to eight weeks after resolution of the inflammatory process interval appendectomy is recommended to avoid recurrent abdominal pain. Laparoscopic appendectomy can be performed safely and effectively in such interval cases.

Mesenteric lymphadenitis is the condition most commonly mimicking acute appendicitis resulting in a high rate of negative appendectomies in children. Mesenteric adenitis is frequently associated with an upper respiratory infection. Clinical presentation includes fever, leukocytosis and low abdominal pain. Mesenteric adenitis can be the result of a viral or bacterial infection. Viruses implicated include Epstein-Barr; Adenovirus type 3, influenza B and Coxsackie B. Bacteria associated with mesenteric adenitis includes hemolytic streptococci, Yersinia and Salmonella species. The diagnosis of mesenteric adenitis is principally one of

exclusion. CT-Scan can help decide whether the child has mesenteric adenitis when the lymph nodes aggregates can be clearly seen. Otherwise, since it can be very difficult to distinguish appendicitis from mesenteric adenitis the diagnosis is establish at surgery. Laparoscopy can also be useful to differentiate appendicitis from mesenteric adenitis. After surgery the postoperative course of children with mesenteric adenitis is usually uneventful and recovery is rapid.

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, and 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 arises 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 (fascial 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 produces 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 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 have 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 can be associated with Hirschsprung’s disease so a rectal biopsy is in order. 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. Fecal Incontinence

Fecal incontinence (FI) in children is usually the result of a congenital (imperforate anus, Hirschsprung's disease and myelomeningocele) or acquired (trauma, pelvic tumor resection and spinal cord injury) condition. FI causes a psychological, developmental and social stress of great magnitude. Assessment of the problems includes the use of anal tonometry, electromyography and pudendal nerve terminal motor latencies. Rectal emptying and angulation can be evaluated with defecogram, MRI or scintigraphic proctography. The management of FI is complex. For HD biopsychosocial treatment consisting of explanation, extinction of fear and avoidance behavior, learning new defecation behavior, learning an adequate straining technique, and generalization toward daily life have been effective. Biofeedback is a harmless and inexpensive treatment coordinating pelvic floor muscle contraction with the sensation of rectal filling but has brought partial success in children with FI. Other less effective forms of treatment are dynamic graciloplasty and sacral nerve stimulation. Functional constipation associated with fecal incontinence and the presence of segmental dilatation of the sigmoid or rectum responds poorly to medical management. The mega-bowel lacks an adequate anorectal reflex, shows decrease propulsive function causing a functional obstruction. Segmental resection of the dilated segment has brought successful relief of symptoms. Antegrade enemas by way of an appendicocecostomy or cecostomy tube can achieve total colonic evacuation and socially controlled continence in children with anorectal malformations and caudal agenesis in a high percentage of cases.

Q. Inflammatory Bowel Disease

1. Crohn's Disease

Crohn's (terminal ileitis) is a chronic, transmural inflammatory bowel disease most frequently involving the terminal ileum and proximal colon that adversely affect growth and sexual maturation in children. Incidence is growing and etiology is undetermined. Diarrhea, abdominal pain, failure to thrive and weight loss is the most frequent clinical feature. Diagnosis is established by colonoscopy or imaging studies (CT-Scan). Initial management is medical and consists of Azulfidine or 5-amino salicylic acid preparations, local and systemic steroids, metronidazole, immunosuppressives, and enteral and/or parenteral nutrition. Indication for surgery is limited to complications of the disease process and includes failure of medical therapy, perforation, abscess, severe malabsorption and growth retardation, persistent bowel obstruction, fistulas (entero-enteric and entero-urinary) and strictures. Surgery can be accomplished using limited resection and anastomosis or stricturoplasty. Best long-term results after surgery occurs in children with disease confine to the small bowel and ileocecal region. Diffuse ileocolonic involvement (Panenteritis), preoperative use of 6-MP, and colonic involvement is associated with early relapse. Early relapse after surgery is also seen after failure of medical therapy independent of disease location as the sole indication for surgery and in children undergoing resection within one year of the onset of symptoms. 2.

2. Ulcerative Colitis

Ulcerative colitis (UC) is a chronic debilitating inflammatory disease of the bowel affecting primarily the mucosa and to a lesser extent to the adjacent submucosa. UC affects mainly the rectum and colon causing delayed growth and development. In some children the terminal ileum is affected. Cause of UC remains unknown. Peak incidence is between 1.5 and 17 years (mean 11 years). UC originates as an acute inflammation of the crypts (cryptitis) developing tissue reaction of chronicity. More than 90% children with UC have moderate to severe disease. Clinically, the child with UC develops bloody diarrhea, abdominal cramps, anemia, fever, tachycardia, hypoalbuminemia and weight loss. Colonoscopy is diagnostic. Medical management includes restriction of milk protein from diet, parenteral nutrition, steroids, sulfasalazine, metronidazole, 6-mercaptopurine, cyclosporine and tacrolimus. UC can be cured by surgical resection of the colon. Indications for surgery in UC include inability to attain growth and development under medical therapy, fulminant disease refractory to medical therapy, extensive rectal bleeding, perforation and toxic megacolon. Surgical management consists of total proctocolectomy and ileal pouch anal anastomosis. The J-pouch is the simplest to construct. Retention of mucosa above the dentate line after surgery produces recurrent

inflammatory disease and high risk of developing carcinoma. A low risk of bladder dysfunction and impotence due to damage to pelvic nerves is associated with proctocolectomy. Postop complications are associated with duration of the disease and length/dosage of medication (steroids). Long-term function after surgery is good in more than 90% of children with high patient satisfaction.

R. Intractable Constipation & Encopresis

Constipation is a common abdominal symptom in childhood. In the majority of cases no cause is identified and the condition is labeled as idiopathic. More than 90% of children with idiopathic constipation respond to medical treatment (bulk diet, laxatives and enemas). Less than 10% develops intractable constipation. Intractable constipation, not associated to Hirschsprung's disease, neuromuscular disease or repaired anorectal malformations, that fails to respond to aggressive medical management is one of the most difficult conditions to manage in children. Children have duration of symptoms for a period beyond five years. Intractable constipation produces progressive fecal retention, fecal incontinence, distension of the rectum and sigmoid colon with loss of rectal sensory and motor function. Encopresis ensues when fecal soiling results from the retained fecal material. Idiopathic constipation is associated with a thickened internal anal sphincter. Colonic manometry helps differentiate causes of intractable constipation in childhood showing the length of the abnormal colonic involved segment. Surgical management for intractable constipation can consist of internal myectomy, placement of cecostomy or left-colon tubes for antegrade enema cleansing, or resection of the disease colonic segment when there is severe stasis and luminal dilatation. Outcomes have thrown mixed results.

Encopresis refers to the involuntary loss of formed, semiformed, or liquid stools into the child's underwear in the presence of constipation Solid fecal material accumulated in the distal rectum unable to be discharged appropriately produces seepage of more proximally fecal fluid which escapes unconsciously into the cloths of the child. It's a very difficult social and physical problem to manage satisfactorily in the child. Encopresis is a complex abnormal motility disorder, requiring a multidisciplinary approach. The most common causes associated with encopresis consist of slow transit functional constipation, Hirschsprung's disease and anorectal malformations. Severely constipated children with encopresis in whom outpatient management has failed frequently require several days of hospitalization, as well as conventional treatments involving cathartics and enemas. A balanced electrolyte solution of the nonabsorbable polymer polyethylene glycol (Golytely) offers a safe and efficient method for clearing the intestine in such cases. Children with encopresis have normal functioning internal sphincter and can acquire normal bowel control using biofeedback therapy to correct the abnormal defecation dynamics. A continent appendicostomy (Malone procedure) is a promising treatment that completely cleanses the colon, increases the child's autonomy, and decreases the chance of soiling in intractable cases of encopresis with pseudo-incontinence.

S. Short Bowel Syndrome

Short bowel syndrome is a very serious gastrointestinal disorder characterized by the absence of significant length of bowel capable of normal digestion and absorption. It is estimated that more than 70% of small bowel length must be lost to develop a short bowel syndrome. The three most common causes of short bowel syndrome in the pediatric age are necrotizing enterocolitis, midgut volvulus and gastroschisis. Intestinal adaptation can occur when the neonate is left with more than thirty (30) centimeters of small bowel with an intact ileo-cecal valve. Though the prospect of bowel transplant continues to develop better forms of avoiding acute rejection, still the median survival after transplantation is short (mean of 15 months). Recently, a novel experimental procedure has attained the attention of surgeons managing this devastating disease complication. The operation is termed serial transverse enteroplasty (STEP) procedure. After short bowel ensues the process of adaptation includes mucosal hyperplasia and bowel dilatation. The STEP procedure is based on the anatomic principle that the blood supply to the bowel comes from the mesenteric border traversing along the perpendicular long axis of the bowel. Multiple stapler lines are placed perpendicularly alternating the direction of the stapler creating a channel of bowel smaller in diameter and longer in length than the original bowel. Advantages of STEP: easy to do, no anastomosis needed, does not result in intestinal obstruction, mesentery is not jeopardized, the length is almost double, the tapering is customizable, and can be performed in sequence after a successful Bianchi procedure. STEP could become the lengthening bowel

procedure for short bowel syndrome.

T. Gardner's & Turcot Syndrome

Gardner's syndrome refers to a group of children born with familial adenomatous (multiple) polyposis and significant extracolonic manifestations. Familial adenomatous polyposis is an autosomal dominant disorder originating from a germline alteration of the adenomatous polyposis coli gene in the long arm of chromosome 5. The most significant extracolonic manifestation of Gardner's syndrome consists of soft tissue (desmoid) tumors and osteomas. Bowel cancer develops in one-third of patients with Gardner's syndrome from malignant degeneration of the adenomatous polyps. Desmoid tumors are typically benign but locally aggressive slow-growing tumors that surround and compress adjacent vascular structures and viscera. Affected children are asymptomatic until they manifest rectal bleeding or multiple soft/hard tissue tumors. Osteomas appear in the mandible causing dental abnormalities (odontomas, cementomas, cysts, supernumerary teeth). Surveillance by colonoscopy is imperative in affected family members. Development of a subcutaneous fibroma single or multiple that recurs as a desmoid tumor is a sentinel event identifying children with Gardner's syndrome. Management of Gardner's syndrome consists of excision of the soft/hard tissue tumor and removal of the affected colon (proctocolectomy) with preservation of the sphincteric muscle mechanism. Sulindac has been reported to produce drug-induced complete regression of colonic adenomas in Gardner's syndrome.

Turcot (glioma-polyposis) syndrome refers to the presence of multiple adenomatous polyposis coli associated with glioblastoma multiforme, medulloblastoma, or glioma developing during the pediatric teens. The polyposis in Turcot is associated with a low number of polyps, large polyps over three cm in diameter, and complication by colonic cancer occurring during the second or third decades of life. Turcot syndrome is determined by an autosomal gene with pleiotropic effect and variable expressivity. Children with Turcot syndrome can develop multiple regions of congenital hypertrophy of the retinal pigment epithelium with areas of surrounding hypopigmentation in the fundi of both eyes, a fact which can help in the diagnosis. Two types of Turcot syndrome have been identified: Type I, also known as true Turcot syndrome (autosomal recessive) with less than 100 intestinal polyps, large size and apt to transform to the malignant tumor. Brain tumor is mainly diagnosed as glioblastoma or astrocytoma and mismatch repair genes might be involved. Type II with FAP-associated type (autosomal dominant) predisposing to medulloblastoma. Management in both cases is surgical.

U. Esophageal Foreign Bodies

The superior esophagus is the narrowest portion of the alimentary tract of children and the most common site for lodge foreign bodies. Due to the nature of infants and toddlers to place objects in their mouth, especially a coin, this represent the most common foreign body identified within the proximal esophagus. The child will develop cough, stridor, choking, drooling, pain and inability to swallow with a lodge esophageal foreign body. Complications secondary to the esophageal foreign body itself include erosion/perforation, stricture, migration, mediastinitis and airway complications. Since aspiration and perforation are immediate complications, the impacted foreign body mandates urgent surgical attention. A simple chest film will delineate the position of the lodge coin. With other type of non-opaque foreign bodies an esophagogram will be needed to help visualized the position and type of obstruction. Rigid esophagoscopy under general anesthesia or flexible esophagoscopy under sedation is the procedure of choice to remove the foreign body, though Foley balloon extraction under fluoroscopic control is an acceptable method of coin extraction with minimal morbidity. Other times the foreign body can be pushed toward the stomach using esophageal bougienage. Children younger than one years, those with a widened tracheoesophageal interface, not a smooth object or more than one week after ingestion seems to be at highest risk for esophageal edema, failure of balloon extraction and complications.

V. Ogilvie Syndrome

Acute colonic pseudo-obstruction also known as Ogilvie≪s syndrome is a massive colonic dilatation associated with signs and symptoms of colonic obstruction without an evident mechanical cause. Ogive syndrome is observed predominantly in the elderly population with few cases reported in children. Predisposing factors for Ogilvie≪s syndrome in children includes postoperative state, trauma, infections,

Sickle cell disease, cardiac diseases and chemotherapy for malignancy. Symptoms include constipation, abdominal pain, nausea, vomiting and abdominal distension. Diagnosis is suggested in flat simple abdominal films. Findings on CT-Scan are diagnostic showing massive colonic dilatation with diameters of eight to 12 cm and without evidence of overt mechanical obstruction. If left untreated, this dilatation can lead to colonic perforation and peritonitis in 10% of children with high mortality rates. Initial management consists of nasogastric decompression, bowel rest, hydration, electrolyte correction, along with discontinuation of drugs affecting bowel motility. If symptoms fail to improve with initial management then rectal tube or colonoscopy decompression is utilized. Neostigmine, an acethycholinesterase inhibitor which increases parasympathetic tone, has been found to be very effective in managing patients with Ogilvie syndrome. Neostigmine is slowly titrated in increments up to a total of 0.05mg/kg of weight. Also, oral erythromycin therapy has been used to manage this condition. Surgery will be needed if the child develops perforation or signs of bowel ischemia.

W. Magnet Ingestion

Most foreign body ingestion in infant and children passes through the gastrointestinal tract without causing significant sequelae. Surgical intervention is generally required if an object becomes lodged in the gastrointestinal tract or if the material has a harmful effect such as the corrosive effect of batteries. With rare-earth magnets present in many small toys, the situation can be very different. A single magnet ingestion is innocuous and is expected to pass through the GI tract. Unfortunately, a misdiagnosis and misconception that a solitary magnet has been ingested when in fact they are two or more joined together may lead to a delay in diagnosis and subsequent severe and possibly preventable complications. The ingestion of multiple magnets can cause bowel obstruction, volvulus, perforation or internal bowel fistula formation owning to pressure necrosis from magnet attraction. Pressure necrosis and fistula formation can be a gradual process resulting in minimal physical examination findings. In many of the toys the magnets are embedded in plastic parts that are easily detachable. If in the stomach, the magnet should be removed endoscopically. If the history, clinical findings and imaging are suggestive of multiple magnetic ingestion early intervention using laparoscopy or open surgery is indicated to prevent serious life-threatening complications. It is imperative health authorities give more information to parents and physicians about the potential risk of small magnetic toys in children.

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 consists 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 consists 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 have 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.

4. Paraesophageal Hernias

Paraesophageal hernia (PEH), a rare entity in children, occurs when the stomach protrudes laterally through the esophageal hiatus toward the chest while the gastro-esophageal junction stays in anatomic position. Though most cases remain asymptomatic, PEH can cause upper bowel obstruction, gastroesophageal reflux, gastric volvulus, bleeding and perforation. Most PEH in children are acquired resulting after fundoplication. A small group of children are born with the PEH. Groups of patients with a higher incidence of developing PEH after fundoplication includes infants under the age of one, neurologic impaired children and surgical patients where a crural repair is not done after fundoplication. PEH is linked to gagging before an antireflux procedure in children. Diagnosis of PEH is confirmed during a barium swallow and upper gastrointestinal series. A small PEH can be managed non-operatively if the child is asymptomatic. With the presence of symptoms or enlargement of the hernia operative repair must be done. A transabdominal approach is preferred for reducing the stomach and crural repair of the diaphragm. The use of mesh hiatal reinforcement is recommended for patients undergoing reoperation for PEH and recurrent gastroesophageal reflux or if the diaphragmatic crura is thought to require reinforcement at the time of the original surgery.

5. Traumatic Diaphragmatic Hernia

Motor vehicle trauma is the leading cause of an acquired diaphragmatic hernia in a child and adult. The traumatic event can either be penetrating directly injuring the diaphragm, or most commonly blunt abdominal causing a sharp increase in intraabdominal pressure with rupture of the diaphragmatic muscle. The more medial and lateral fibers of the posterior diaphragm arising from the lumbocostal arch and the vertebrocostal trigone are the weakest points of rupture. The posterolateral portion is virtually always the area that ruptures with trauma. Diaphragmatic injuries are difficult to diagnose preoperatively and can be missed easily. Traumatic diaphragmatic hernia should be suspected on the basis of an abnormal chest radiograph in the trauma victim with multiple injuries. If diaphragmatic injury is suspected, ultrasound or CT Scan investigation must be performed. Most cases involve the left diaphragm due to the buttressing effect of the liver. The incidence of bowel strangulation is high in traumatic diaphragmatic hernias. In the acute setting, transabdominal repair after palpation of both hemidiaphragms is the procedure of choice because of the high incidence of associated trauma. Injury severity score and hemorrhagic shock upon admission strongly influence the outcome. Delayed presentation can be repaired through the chest.

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 have been reported.

3. Cystic Adenomatoid Malformation

Congenital cystic adenomatoid malformation is a lung bud lesion characterize 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 follow 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 consist 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. Pneumothorax

Pneumothorax is the presence of air in the pleural cavity. Results from either a tear in the visceral or parietal pleura. Pneumothorax can be spontaneous (primary or secondary), or acquire. The most common cause of primary spontaneous pneumothorax is rupture of an apical subpleural bleb of the lung, usually a thin

adolescent male who suddenly develops chest pain and shortness of breath. Secondary spontaneous pneumothorax occurs after hyaline membrane disease, meconium aspiration, cystic fibrosis, or AIDS. Acquired pneumothorax is more common than spontaneous usually the result of blunt or penetrating trauma, iatrogenic after central line placement, thoracentesis, lung biopsy, barotrauma from mechanical ventilation and laparoscopic procedures. Diagnosis of pneumothorax is done with simple chest films. Complex cystic lung conditions will need chest CT scan for diagnosis. The purpose of management is to evacuate the air in the pleura and expand adequately the lung. Small pneumothorax (less than 20%) can be managed with observation and oxygen therapy. Tube thoracostomy is recommended for pneumothorax larger than 20%. The tube is removed when the lung has expanded completely and the air leak is no longer present for at least 24 hours. Surgical treatment is indicated using video assisted thoracic surgery (VATS) when air leaks continues for more than 72 hours, there is incomplete lung expansion or pneumothorax recurs after adequate management.

D. Inguinal hernias, Hydroceles, Undescended & Ectopic Testis

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 then 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 this 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 makes 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 premature 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/pubic 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.

Inguinal hernias are the most common surgical pathology seen in infants and children. A lump in the inguinal canal area of a newborn or infant female is most probably an ovarian incarcerated inguinal hernia. On very rare occasion the lump is a testis in a child with testicular feminization syndrome. Clinically the irreducible ovarian lump is usually asymptomatic, movable and non-tender mass within the labia majora. Ultrasound can determine the nature of the gonad present. The main problem with an ovarian hernia in infants is the incidence of ovarian torsion associated before repair. Ovaries trapped within inguinal hernias undergo torsion far more commonly than ovaries and tubes in the normal pelvic position increasing the chances of infarction. Torsion can occur at any time after diagnosis of the hernia. The incarcerated ovarian pedicle is narrowed and lengthened within the defect and the internal ring serves as a fixed point around which a twist can occur. The risk of torsion and infarction creates the view that ovarian hernias should be repaired at the earliest elective opportunity if they can be reduced manually. Children with edema, tenderness or skin discoloration in the inguinal area should be repaired immediately. Early recognition and management of this condition reduce the risk of gonadal infarction. During repair surgeons must be aware that in 20% of girls with inguinal hernia, the fallopian tubes occasionally with the ovary or uterus comprise the wall of the hernial sac (sliding component).

Inguinal hernias continue to be the most common congenital pathology in children needing surgical repair early in life. Approximately 1-3% of children have an inguinal hernia. The incidence is higher in premature babies (3-5%). Almost all inguinal hernias in children are the indirect type (99%). The few direct hernias in children are the result of previous surgery or inguinal floor disruption. Management of inguinal hernias in infants and children is straightforward: outpatient surgery after diagnosis for most cases. The procedure consists of high ligation of the hernial sac. Incidence of developing a recurrent inguinal hernia is around 0.8%. Most recurrences occur two years after the initial surgery. Several factors play a role in increasing hernia recurrence. These are: 1) Missed sac or inadequate ligation of the indirect sac. 2) Children operated for incarcerated inguinal hernias since tissue is more friable and edematous at the time of surgery. 3) Infection of the wound after hernia repair predisposing to tissue breakdown and a higher recurrence rate. 4) Connective tissue disorders (Hurler, Ehlers-Danlos, etc.). 5) Growth failure and poor nutrition. 6) Prematurity has been identified as a comorbid factor in hernia recurrence. 7) Children hernia repair done by non-pediatric surgeons is also a risk factor for recurrence. 8) Conditions causing increase intra-abdominal pressure (VP shunts, posterior urethral valves, bladder exstrophy repair, weight lifting and respiratory conditions) are also related to higher rate of recurrence in children. Repair of the recurrent hernia is done through the inguinal scar or using laparoscopy. The sac is mobilized and ligated. Laparoscopic repair of recurrent inguinal hernia has the advantage of passing through a virgin field reducing damage to vas or vessels and allowing inspection of the area with direct purse string repair of the defect.

Incarceration and strangulation are the most dreaded complications of inguinal hernias in children. Incarceration refers to viscera (bowel, ovaries, and bladder) that protrudes through the inguinal defect and cannot return back to its anatomical position without manipulation or surgery. Bowel incarceration in infants with inguinal hernia is a notable cause of intestinal obstruction in this age group. Strangulation is the ischemic effect caused on the trapped viscera by the incarcerated defect. Incarceration occurs in almost one-third of inguinal hernias. It is more common in children less than one year of age and males. With prolonged incarceration there can also occur testicular infarction. In infant girls the normal anatomy is altered when an ovary is trapped in a hernia sac, and these changes make torsion more likely. This risk warrants treating the asymptomatic irreducible ovary as any other incarcerated hernia with urgency. Incarceration increases the rate of complications, is seen in a younger population of children and increases hospital stay. Children with incarcerated hernia should have a trial of manual reduction followed by prompt repair within the next five to seven days to avoid re-incarceration.

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. Abdomino-scrotal hydrocele (ASH), also known as "hydrocele en bissac", is a very rare condition seen in infants consisting of a collection of fluid in the tunica vaginalis extending through the inguinal canal into the abdominal cavity. Clinically, the child with an inguino-scrotal hydrocele has an abdominal mass of variable size and firm consistency characterized by increase in tension of the hydrocele when squeezing the abdominal mass and vice versa. The abdominal compartment can be retroperitoneal or properitoneal. An increasing pressure within the hydrocele is transmitted above the deep inguinal ring because of the inexpansible musculofascial covering of the inguinal canal. The diagnosis of an abdomino-scrotal hydrocele is made with the help of ultrasound or MRI showing the fluid filled cavities in both the abdominal (pelvic) and scrotal compartments in communication. ASH should be differentiated from other cystic tumors of the abdominal cavity such as hydronephrosis, bladder diverticulum, mesenteric cysts and lymphangiomas. ASH has been found to cause

obstructive uropathy, reduced blood supply to the testis or hemorrhage. Total excision of the abdomino-scrotal hydrocele through an inguinal approach is the proposed treatment of choice.

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% occurring 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. Retractile testis needs 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 testes that did not developed, suffered an ischemic intrauterine event, and in performing first stage Steven-Fowler technique.

Whenever a child is born with an empty scrotum, the physical examination should include a diligent palpable search for the undescended testis in the inguinal, femoral, perineal or medial thigh areas. Testes palpable in areas away from the normal descent from the retroperitoneum to the scrotum are termed ectopic testis. An ectopic testis is caused by misallocation of the ipsilateral genito-femoral nerve controlled stimulation causing the gubernaculum to migrate to the wrong site because the chemotactic signal is arising from this wrong place. Testes palpable in the inguinal canal or found intra-abdominally are termed undescended. Compared with undescended testes, ectopic testes are extremely rare found most commonly in the perineal ipsilateral area. Other sites include the femoral canal, suprapubic region (at base of the penis), medial thigh, preperitoneal, umbilical, contralateral scrotum or associated with gastroschisis. The perineal testis is particularly subject to trauma. Management is orchiopexy as soon as the diagnosis is established. The most effective route of approach for repair is inguinal allowing replacement of the testis into the corresponding hemiscrotum without difficulty. Other surgeons use a low scrotal approach due to the low incidence of concomitant hernia. Because of the histopathologic features involved, prognosis is better than that associated with cryptorchidism.

E. Mediastinal Cysts, Thymoma & Myasthenia Gravis

The differential diagnosis of an anterior mediastinal mass in a child includes in order of increased frequency a lymphoma, teratoma, thymoma or suprasternal goiter. Histopathologic analysis of an anterior mediastinal mass of unknown origin is essential for treatment decision. An anterior mediastinal mass can risk airway compromised. General anesthesia should be avoided in children with tracheal cross sectional area or peak expiratory flow rate less than 50% of predicted for age and sex, or severe narrowing and occlusion of a main stem bronchi. Diagnosis can be established in most patients by open biopsy or image-guided core needle biopsy under local anesthesia. Other cases might need aspiration of pleural effusion or bone marrow

biopsy for diagnosis. Children that present with symptoms of dyspnea at rest, orthopnea, respiratory distress, or stridor are at risk of respiratory collapse. In severe cases when symptoms of respiratory collapse are present and the diagnosis cannot be obtained, a short course of steroid therapy can reduce the size of the mass and improve the child clinically. The Chamberlain procedure provides excellent access to the antero-superior mediastinum for biopsy of obscure mediastinal mass lesions in childhood with a low rate of complications. Should general anesthesia be needed, spontaneous ventilation is preferred.

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.

The thymus remains quite prominent in the anterior mediastinum during the first year of life causing discrepancy between a normal and hyperplastic gland. Involution occurs in response to stress and sepsis. Rebound hyperplasia after involution can be seen after cardiac surgery, major burns and chemotherapy. Thymoma is the most common neoplastic tumor found in the thymus of children and adults. There is a close relationship between myasthenia gravis and thymoma. Most thymic tumors in children are benign, share a low rate of association with myasthenia gravis and a favorable prognosis. Thymomas are considered malignant on the basis of macroscopic and microscopic capsular invasiveness. The most significant predictors of long-term survival of thymoma include complete excision, stage I disease, and lymphocytic histology. Management of thymoma entails surgical resection through a median sternotomy. To increase survival a policy of aggressive, complete surgical resection of all thymomas is advice. Thymoma behaves as a rather indolent tumor, with most deaths from causes unrelated to thymoma or its direct treatment. Chemotherapy is reserved for patients with refractory or metastatic disease. Thymomas are moderately radiosensitive but radiation therapy is not an attractive option for children due to side-effects on developing organs.

Children constitute 10% of all cases of Myasthenia Gravis (MG) with three individual forms identified: neonatal, genetic or juvenile. The neonatal phase is transient, associated with a newborn whose mothers have MG and the baby recovers completely after several days or weeks. Genetic MG is not associated to a parent with MG with symptoms confined to ptosis and almost no weakness. The juvenile phase of MG is similar to the adult phase occurring after the age of ten. Symptoms include fluctuating weakness and fatigue in the ocular (diplopia), facial (ptosis), bulbar or limb muscles, weakness, fatigability, ptosis and diplopia. The child develops motor weakness, preservation of sensation, coordination and deep tendon reflex. MG is an autoimmune disease in which there is loss of acetylcholine receptors at the neuromuscular junction. Thymic enlargement occurs in patients with MG. MG is best managed: 1) enhancing neuromuscular transmission with cholinesterase inhibitors though the effect is partial with time; 2) using immune suppression with steroids, azathioprine or cyclophosphamide; 3) with short term immune therapy including plasma exchange or intravenous immune globulin; 4) removal of the thymus (thymectomy) if its enlarged or the child has increase medication requirements.

F. Umbilical Hernias & Granulomas

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 promote 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.

Persistent umbilical swelling and discharge during the neonatal period is of serious concern to both parents and physicians. Among umbilical swelling, the umbilical granuloma is one of the most commonly seen condition in the pediatric practice. The normal granuloma, a common inflammatory reaction to the resolving umbilical stump of a newborn should disappear by the 2nd to 3rd week of life after proper hygiene. Persistent beyond this time will need some type of therapy. Umbilical granuloma is managed with 75% Silver nitrate stick application. Silver nitrate is not innocuous and when apply liberally can cause a minor burn of the periumbilical skin area of the baby. Caution must be observed while applying Silver nitrate; careful drying the umbilical exudate to prevent periumbilical spillage, and discussion with parent that burns may occur but apparently are not serious. Whenever Silver nitrate therapy fails and discharge persists, or contains urine or fecal material, the physician should suspect that the child has either a patent urachus or omphalomesenteric duct remnant as both conditions resemble the common umbilical granuloma seen in general practice. Ultrasound studies of the periumbilical area looking for a cyst, masses or fixed bowel loops can help determine the presence of such congenital remnants. Management of the persistent umbilical granuloma is surgical with double ligature, cauterization of the base or formal umbilical exploration.

G. Omphalocele and Gastroschisis

The three most common abdominal wall defects in newborns are umbilical hernia, gastroschisis and omphalocele. 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. G. Epigastric Hernias

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.

A giant omphalocele is defined as a defect larger than 10 cm in length that harbors the liver. Prenatally diagnosed giant omphaloceles will need cesarean section as route of birth to avoid fetal liver rupture. Management of giant omphalocele has a high morbidity and mortality due to the defect size, visceroabdominal disproportion, and the associated congenital and genetic malformations. The large size of the defect and small abdominal cavity creates a situation where primary closure is almost impossible unless some sort of stage reduction is tailored. Pulmonary hypoplasia, genetic defects and cardiac malformation are the source of early mortality in these babies. The pendulum of management of giant omphalocele has moved toward a more conservative initial management using topical coverage creams to create granulation tissue and skin on top of the membrane followed by repair of the ventral hernia much later in life when the medical condition of the child permits. Silver sulfadiazine (Silvadene) provides a moist wound healing environment that promotes epithelization and simultaneously minimizes the risk of invasive infection including antifungal coverage. Silver toxicity, though rare can include seizures, peripheral neuropathy, ocular pathology, nephrotic syndrome, raised liver enzymes, leukopenia and algiria. For smaller size defects the use of silo, tissue expanders, biologic mesh, vacuum-assisted closure or component separation technique closure is indicated. Giant omphalocele is associated with deficits in developmental achievements in most of the affected infants ranging from mild to profound delays.

H. Epigastric Hernias

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.

I. Recurrent Inguinal Hernias

Inguinal hernias continue to be the most common congenital pathology in children needing surgical repair early in life. Approximately 1-3% of children have an inguinal hernia. The incidence is higher in premature babies (3-5%). Almost all inguinal hernias in children are the indirect type (99%). The few direct hernias in children are the result of previous surgery or inguinal floor disruption. Management of inguinal hernias in infants and children is straightforward: outpatient surgery after diagnosis for most cases. The procedure consists of high ligation of the hernial sac. Incidence of developing a recurrent inguinal hernia is around 0.8%. Most recurrences occur two years after the initial surgery. Several factors play a role in increasing hernia recurrence. These are: 1) Missed sac or inadequate ligation of the indirect sac. 2) Children operated for incarcerated inguinal hernias since tissue is more friable and edematous at the time of surgery. 3) Infection of the wound after hernia repair predisposing to tissue breakdown and a higher recurrence rate. 4) Connective tissue disorders (Hurler, Ehlers-Danlos, etc.). 5) Growth failure and poor nutrition. 6) Prematurity has been identified as a comorbid factor in hernia recurrence. 7) Children hernia repair done by non-pediatric surgeons is also a risk factor for recurrence. 8) Conditions causing increase intra-abdominal pressure (VP shunts, posterior urethral valves, bladder exstrophy repair, weight lifting and respiratory conditions) are also

related to higher rate of recurrence in children. Repair of the recurrent hernia is done through the inguinal scar or using laparoscopy. The sac is mobilized and ligated. Laparoscopic repair of recurrent inguinal hernia has the advantage of passing through a virgin field reducing damage to vas or vessels and allowing inspection of the area with direct purse string repair of the defect.

J. Acute Scrotum & Epididymitis

The term acute scrotum refers to signs and symptoms associated with local inflammation of the scrotum that appears suddenly and usually is not associated with trauma. Is a common urological emergency. Such signs and symptoms include scrotal pain, swelling, redness and heat. The most common causes of acute scrotum in children constitute testicular torsion, appendix testis torsion, epididymitis, orchitis and pyocele. Testicular torsion, a surgical emergency, occurs in 15% of all cases and is the most important condition to diagnose and manage early in order to avoid testicular loss, fertility problems and medico-legal issues. Testicular loss commences past the twelve hours of initiation of symptoms. Beyond twenty-four hours of symptoms testicular loss in the norm. This is the main reason why in the absence of ancillary studies surgeons immediately explore the acute scrotum. The two most commonly used preoperative studies are testicular scan and color Doppler ultrasound. Testicular scans reliable show whether the testes have vascular flow or not, but are difficult to be obtained in the middle of the night. Doppler ultrasounds are operator dependant and when done by experienced physician can help reduce the number of emergency operations and hospitalization days. Clinical judgment by the surgeon is probably the most important factor in assessing testicular salvage. In the face of doubt the next step in management is immediate surgical scrotal exploration.

Idiopathic scrotal edema is a very uncommon cause of acute scrotal swelling, but considered the commonest cause of the 'acute scrotum' in prepubertal boys. Idiopathic scrotal swelling is characterized by edema and erythema of the scrotal wall, is usually bilateral and can sometimes involve the shaft of the penis. The swelling and erythema can extend into the abdominal wall and perineum. Children affected with acute scrotal swelling are four to six years in age with symptoms present for less than 24 hours at the time of initial medical evaluation. The cause of the swelling is usually not identified but can be associated to reaction to an allergen, bug bite, contact dermatitis or angioneurotic edema. Leukocytosis is absent, urinalysis is usually normal and urine culture is sterile. Peripheral eosinophilia is present in some patients. A connection with trauma, periurethral disease, or streptococcal disease appears unlikely. The differential diagnosis includes torsion of the testis or one of the testicular appendages, hydrocele, varicocele, trauma, tumor, idiopathic scrotal edema, and Henoch-Schönlein purpura. Color Duplex ultrasound of the scrotum will show increase testicular blood flow and thickening of skin and muscle of the scrotum. Exploration is required when a normal testis cannot absolutely be identified. Swelling usually resolves within two to five days. Management consists of bed rest, reassurance, and oral histamine.

Acute inflammation of the epididymis is an infectious process which usually occurs during adolescent years, very rarely during prepubertal ages. The infectious process is caused by a distal urethral obstruction, ectopic ureter entering the seminal vesicles or epididymis, or after instrumentation. Bacterial or viral organisms are involved in the infectious process. Epididymitis seems to be more common than acute testicular torsion. Early clinical manifestations of epididymitis include scrotal edema, pain, erythema, tenderness with an associated reactive hydrocele. The epididymis turns elongated and exquisitely tender to palpation. The differential diagnosis includes testicular torsion, torsion of the testicular appendage or idiopathic scrotal edema. The urinalysis will demonstrate pyuria with bacteriuria. Leukocytosis is also identified. Color Doppler ultrasound or testicular scans can determine rapidly if we are dealing with torsion due to reduced or absent testicular blood flow in need of urgent surgery. In the event of doubt or absence of imaging studies the diagnosis of an acute scrotum requires scrotal exploration. Management of epididymitis includes intravenous antibiotics, pain medication, scrotal support and bed rest. Further renal ultrasound and excretory urography are needed after the episode subsides to determine a congenital urologic anomaly.

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_2O, 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 it's 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 consists 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 requires 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 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 ir 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 consists 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.

(c) 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 are 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 99mTc 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 syndromes 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 majorities 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. Treatments depend 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.

E. Peptic Ulcer Disease

With the advent of more powerful acid inhibiting drugs (H2 antagonists) and proton pump inhibitors, surgery for refractory peptic ulcer disease (PUD) is becoming something of the past. We now manage rare complications of PUD such as bleeding, perforation and obstruction. PUD is either primary or secondary in origin. Secondary PUD are more common and primarily seen in infants the result of an associated condition such as prematurity, steroids, burns, trauma, immune deficiency and brain tumors. Primary PUD occurs more commonly in older children and adolescents the result of an imbalance between acid secretion and gastric mucosal protection. Helicobacter pylori infestation plays a crucial role in the genesis of gastritis and primary duodenal ulcer formation. Successful management of PUD disease encompasses eradication of Helicobacter infection. Initial management of PUD includes medication to reduce acid production (omeprazole, ranitidine, cimetidine) combined with agents that improve mucosal defense (sucralfate). Bleeding PUD manifests itself with either melena or hematochezia. The bleeding site must be visualized during upper endoscopy. Most cases will stop with prompt medical management or endoscopic diathermy. Beyond 50% blood volume loss in a short period of time (8-24 hours) is an indication for surgery. Infants benefit from duodenotomy and bleeding ulcer ligation, while older children will need more definite ulcer surgery (vagotomy and pyloroplasty). Perforated PUD is the manifestation of a coexisting illness. Simple surgical closure with an omental patch is sufficient. Obstruction produces chronic vomiting due to pyloric scarring.

VI- PANCREATIC, HEPATIC, BILIARY, SPLENIC and ADRENAL 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 is 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 papillas are stenotic. Once chronic pancreatitis is established, ductal drainage or resection may be necessary.

B- Pancreatitis

1. Acute 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.

2. Chronic Pancreatitis

Chronic pancreatitis is an unusual condition seen in children. In contrast to adults, chronic pancreatitis has significant lifelong morbidity. Most common etiology of chronic pancreatitis in children consists of trauma, cystic fibrosis, hyperparathyroidism, hyperlipidemia, aminoaciduria, hereditary, congenital ductal anomalies and choledochal cyst. Main symptoms are recurrent abdominal (epigastric) pain associated with nausea and vomiting. A transient elevation of amylase and lipase is associated with the pain. Once the diagnosis of chronic pancreatitis is suspected, the anatomy of the pancreatic duct should be defined using ERCP or MRCP (less invasive). In chronic pancreatitis, the pancreatic duct is frequently dilated with one or more obstructive lesions. Ongoing attacks of pancreatitis may lead to diabetes, steatorrhea and pancreatic stones. Early endoscopic sphincterotomy can slow the progressive nature of the disease and has been suggested as initial choice of treatment. With continued symptoms, lateral pancreaticojejunostomy (Puestow procedure) in dilated and obstructed ductal anatomy has been found to decrease significantly the pancreatitis episodes, relieve the recurring abdominal pain and arrest the progression of exocrine and endocrine insufficiency. Recently, the Frey procedure adding pancreatic head coring to the pancreaticojejunostomy has provided pain relieve in more than 85% of children with chronic pancreatitis.

C. 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.

D. 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, Combs' 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 are 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.

E. 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 (fascial 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.

F. Choledochal Cyst & Choledochocele

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.

In 1985, a new variant of choledochal cyst known as forme fruste was described in the pediatric literature. Forme fruste choledochal cyst (FFCC) is characterized by minimal dilatation of the extrahepatic bile duct which does not grow with time. The normal diameter of the common bile duct in children ranges between two and 6 mm. FFCC is associated with a diameter above six mm and below 10 mm. Most patients with FFCC have a long common channel, in which the common bile duct-pancreatic duct junction is away from the duodenal papilla, with partial obstruction of the terminal common bile duct. FFCC is associated with fever, jaundice, abdominal pain, recurrent pancreatitis and altered liver function tests. Histologically FFCC demonstrates thickened fibrous connective tissue, absent muscular layer with flattened, ulcerated and dysplastic mucosa. Diagnosis is established with ultrasound. ERCP or better yet MRCP can help delineate the anatomy and presence of a long common pancreaticobiliary channel in FFCC. Management consists of cyst excision and Roux-en-y hepaticojejunostomy. Due to the small size the anastomosis is technically difficult and should be performed carefully to avoid stricture and postoperative cholangitis. To maintain ductal anastomosis patency it is imperative that diseased ductal tissue not be incorporated in the anastomosis, the circumstance most likely responsible for the high incidence of anastomotic stricture in choledochal cyst past drainage operations.

Choledochocele is an extremely rare variant of choledochal cysts classified as type III by Alonso-Lej. The cystic dilatation occurs in the distal portion of the common bile duct most commonly in an intrapancreatic fashion protruding toward the duodenum. Patients with choledochocele can develop intermittent colic abdominal pain, obstructive jaundice and recurrent bouts of pancreatitis. Nearly half of the patients have previously undergone cholecystectomy. Diagnosis is suggested by ultrasonography and confirmed with HIDA scan, MRCP or ERCP. In the most common variety the ampulla opens into the choledochocele which in turns communicates with the duodenum via another small opening. Cyst distension explains the episodes of colicky pain. Adenocarcinoma has been reported arising from choledochocele, though some researchers believe choledochocele does not share the strong premalignant potential of the more common types of choledochal cysts. The mucosa lining of choledochocele is usually duodenal, arguing that the cyst is a duodenal duplication. Successful endoscopic management (papillotomy) of choledochocele has been reported when the lesion is small. Surgical management entails excision of the duodenal luminal portion of the cyst leaving the medial portion containing the ampulla intact. Prognosis after surgery is good.

G. Cholelithiasis, Choledocholithiasis, Gallbladder Hydrops & Gallstone Ileus

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 disease 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 ducts 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.

Cholelithiasis is a frequent complication of Sickle Cell Disease (SCD) in children. Prevalence of developing pigmented stones can range from thirty to 50%. Even the prevalence of developing common bile duct stones in children with SCD is higher than the general population. Complications related to cholelithiasis include biliary colic, pancreatitis, cholecystitis and obstructive jaundice. Children with SCD should be screened for cholelithiasis since the age of eight years using abdominal ultrasound. Once cholelithiasis is identified removal of the sick gallbladder should be accomplished regardless if the child is asymptomatic or not. Preoperative transfusion reduces the morbidity of the surgical procedure. Elective laparoscopic cholecystectomy should be the gold standard in children with SCD and asymptomatic cholelithiasis to prevent the potential complications of biliary colic, acute cholecystitis, and choledocholithiasis, which lead to major risks, discomfort, and longer hospital stay. A laparoscopic cholecystectomy result in a shorter hospital stay with fewer postoperative complications than open operation in patients with sickle cell disease and is the procedure of choice in the treatment of cholelithiasis in such patients.

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.

Laparoscopic cholecystectomy (LC) has replaced the open procedure as the treatment of choice in gallbladder disorders. With the lap technique an increase two- to threefold in the incidence (0.5%) of bile duct injuries (BDI) has also occurred. Inappropriate anatomy identification and aberrant situations are the most common cause of BDI. BDI can be classified into transection, lacerations, leaks or strictures. Disease complexity, increasing age, male gender and admission to a teaching hospital are associated with an increase risk of injury. Classic injuries involve clipping & dividing the CBD for the cystic duct. Other BDI may result from stenting injury, thermal injury; lose cystic duct clip or an accessory "Luschka" duct. Such injuries cause two principal clinical manifestations: 1) bile leaks with pain and secondary bile peritonitis, and 2) biliary obstruction due to partial or complete hepatic or common duct ligation (jaundice) or late stricture formation. US, HIDA scan or CT-Scan followed by ERCP or PTC may help diagnosed and define the exact location and nature of the BDI. Complete proximal obstruction may need transhepatic biliary catheters in each radical duct to relieve obstruction. A bile leak may need percutaneous drainage for sepsis control. Cystic or accessory

ducts leaks can be dealt with endoscopic ampullary stenting or re-laparoscopic ligation if identified early. The most effective means of reconstructing a major BDI is using roux-en-y hepaticojejunostomy after inflammation and adhesions of the initial insult have subsided. Intraoperative cholangiography appears to protect against BDI by preventing misidentification of anatomy. Conversion to open cholecystectomy is essential in avoiding these injuries when the anatomy remains uncertain.

The finding of gallstones in neonates and infants is no longer rare. With the widespread use of abdominal sonography in a growing population of prematures TPN-fed survivals more cases of cholelithiasis in infants are reported. Stone formation increases with the duration of TPN. Other lithogenic factors are prolonged fasting, ileal resection, cystic fibrosis, polycythemia, multiple blood transfusions and inflammatory bowel disease. Approximately 80% of gallstones in children are not due to hemolytic disease. Clinically, the symptomatic infant with gallstone can develop colicky abdominal pain, postprandial nausea/vomiting, fatty-food intolerance and failure to thrive. Severe complications are seen in almost one-third of the infants with gallstones including jaundice by common bile duct obstruction, pancreatitis and perforation with bile peritonitis. Surgery is the treatment of choice for infants presenting with symptomatic cholelithiasis. Resolution of gallstones is seen in most asymptomatic infants between six months and one year of oral intake justifying a period of observation. Ursodeoxycholic acid is not effective in the treatment of pigmented stones. Laparoscopic cholecystectomy is recommended for the asymptomatic child younger than three years of age when echogenic shadows have been present for at least 12 months following resumption of oral feedings or when the gallstones are radiopaque (calcified).

A polypoid lesion identified in the gallbladder of a child is a very rare event. It represents an elevated lesion of the mucosal surface of the gallbladder which in most instances causes parental concern. Fortunately, most polypoid lesions identified in gallbladders are benign (90%). Histologically they are adenomatous, hyperplastic, gastric heterotopia or cholesterol polyps. The prevalence of such polyps is greater among males and obese children. Ultrasonography is the image method of choice in diagnosing gallbladders polyps in children and adults. They are seen as pedunculated or sessile echogenic lesions attached to the gallbladder wall protruding toward the lumen and fixed in changed of posture. Gallbladder polyps can be associated with acalculous cholecystitis. Lesions smaller than 10 mm do not progress to malignancy or development of stones, and none produces symptoms or complications of biliary disease. Surgical management of gallbladder polyps is indicated when the size of the polypoid lesion is above 10 mm in diameter, when associated with gallstones and when the child has consistent biliary symptoms. Treatment consists of laparoscopic cholecystectomy. Asymptomatic small polyps (< 10 mm) should be maintained under ultrasonographic surveillance.

Common bile ducts stones (choledocholithiasis) are usually secondary stones from the gallbladder that migrate through the cystic duct causing acute or chronic biliary tract obstruction (obstructive jaundice), pancreatitis or cholangitis. Most common bile duct stones in children pass spontaneously without significant complications. Rarely, children can also develop primary common bile ducts stones. Up to 15% of children harboring gallstones can harbor asymptomatic common bile duct stones. Recurrent right upper quadrant pain is the most common clinical presentation. Most children have idiopathic gallstones. Ultrasound is the first study which will reveal whether the child has common bile duct dilatation with the presence of stones. This is accompanied with measurement of liver function tests and pancreatic enzyme. Once dilatation of the common bile duct or stones is suspected, an MRCP is indicated. With good sonographic evidence of common bile ducts stones or a positive MRCP an ERCP should be performed to accomplish endoscopic sphincterotomy with extraction of the biliary duct stones. This is followed in the next few days with laparoscopic removal of the gallbladder. The endoscopic approach to choledocholithiasis produces less morbidity, less possibility of long term common bile duct stenosis, and permits preop visualization of the biliary tree. If the endoscopic stone extraction cannot be accomplished with ERCP, then open or laparoscopic common bile duct exploration is indicated.

Acute distension of the gallbladder in the absence of stones, bacteria, or congenital malformations is known as gallbladder hydrops. This condition has a vascular origin such as transient arteritis or ischemia. In older children hydrops will present with fever, nausea, vomiting, right upper quadrant mass and abdominal

tenderness, while neonates and infants might show only a palpable mass. Gallbladder hydrops in neonates or infants is caused by systemic sepsis, while Kawasaki disease (mucocutaneous lymph node syndrome), scarlet fever, leptospirosis and trauma are the most common cause of hydrops in older children. The diagnosis of hydrops is established using abdominal ultrasound demonstrating normal biliary ducts and a distended gallbladder without calculi or congenital malformation. The treatment of hydrops is conservative. Management consists of systemic antibiotics and early enteral feeding to stimulate gallbladder function and decompression. Should pain and distension persists, open or percutaneous cholecystostomy may be helpful depending on the medical condition of the child.

Gallstone ileus is a very rare disorder characterized by mechanical obstruction of the gastrointestinal tract due to incipient impacted gallstones that passes through a bilio-enteric fistula. Impaction followed by obstruction can occur at the ileum, duodenum or stomach (Bouveret's syndrome). Diagnosis is usually delayed due to lack of specific signs of biliary disease. The classic triad of Rigler (small bowel obstruction, ectopic gallstones and air in the biliary tree) is visualized on abdominal plain films in only one-thirds of cases. Age ranges from 13 to 87 years with most cases seen in older patients. Most bilio-enteric fistulas are cholecystoduodenal type, with a few choledochoduodenal. Work-up includes ultrasound, upper gastrointestinal series with water soluble contrast medium and contrast enhanced computed tomography (CT). Preoperative diagnosis of gallstone ileus significantly increases by combining plain film and US findings. Management consists initially of simple enterotomy (enterolithotomy) which can be done laparoscopically assisted. This is followed by takedown of the bilioenteric fistula and cholecystectomy in a later stage procedure if the medical condition of the patient permits and he continues symptomatic. Some reports encourage enterolithotomy, repair of the fistula and cholecystectomy in one procedure. Other workers report that enterolithotomy alone is adequate treatment in the elderly, and subsequent cholecystectomy is not mandatory. Early diagnosis and treatment improve the outcome.

H. 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.

I. Splenic Cyst & Abscess, 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.

Splenic abscess is an uncommon event identified during the pediatric age. Nevertheless, is a potentially fatal disorder if not diagnosed and managed in a timely fashion. Children harboring a splenic abscess present with fever, leukocytosis and left upper quadrant abdominal pain. Simple chest films could be associated with a left pleural effusion or basal atelectasis. Most children with splenic abscess have an

associated predisposing medical condition such as sickle cell disease, immune deficiency (HIV), leukemia, aplastic anemia, perforated bowel, typhoid fever, endocarditis, otitis media, appendicitis or trauma. Staphylococci, Salmonella and Escherichia coli are the most common etiologic agents in single abscess, while Candida species predominates in multiple splenic abscess. Many children are septic before the diagnosis is made. Blood cultures are seldom positive. Diagnosis is made with abdominal ultrasound or CT-Scan. Initial management incorporating the strategy of preserving the spleen consists of CT-guided percutaneous drainage and antibiotics. This combined approach is effective in two-thirds of cases. If the child does not improve, splenectomy follows. Children with immune deficiency and splenic abscess have rapid resolution of symptoms with immediate splenectomy. Some cases with splenic abscess will completely resolve with antibiotics alone.

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 heterogenous 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 Combs' 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.

Splenectomy impairs the immune response to bacterial infections. Such impaired immunologic functions include: formation of antibodies, deficiency of opsonization, lower IgM levels, deficiency in bacterial clearing and tuftsin deficient phagocytosis. Overwhelming post-splenectomy infection (OPSI) refers to a constellation of fast-developing symptoms (high fever, hypotension, rigor, bacteremia, and leucocytosis) that leads to death in patients that have undergo removal of the spleen. Mortality rates after OPSI is established are 50%. When obtainable, blood cultures grow encapsulated organisms (pneumococcus, meningococcus, hemophilus, etc.). The vulnerability of OPSI is greatest within the first two years after the splenectomy, and it persists throughout life. The clinical appearance of OPSI can go from a mild event to death from sepsis with pulmonary complications as the most common morbidity. OPSI is more commonly identified after spleen removal for Hodgkin and trauma. Immunization against pneumococcus, H. Influenza and meningococcus should be given to all children who undergo splenectomy since these are the most common organisms associated with OPSI. In the elective situation the vaccine should be given two weeks prior to removal of the spleen. In setting of trauma it should be given as soon as possible, Though several studies have found better functional antibody responses with delayed (14-day) vaccination in the setting of trauma we will continue to administer the vaccine as soon as possible until well-randomized trials are done.

It is estimated that 10% of the general population carries an accessory spleen. Accessory spleens are situated on the hilum of the spleen, splenic artery, pancreas, splenocolic ligament, greater omentum, mesenterium, adnexal region and scrotum. Trauma, torsion and hematologic hemolytic conditions affect an accessory spleen. A careful search should be made for accessory spleens, as they should be removed at the time of primary splenectomy to avoid a second operation later in life. Torsion with infarction of an accessory spleen must be considered as a rare cause of acute abdominal pain in childhood. Accessory splenic torsion causes acute diffuse or localized (left upper quadrant) abdominal pain sometimes indistinguishable from that caused by acute appendicitis or intussusception. Most affected children develop an intraperitoneal inflammatory mass. Preoperative diagnostic imaging is unable to point to the diagnosis. Ultrasound shows a round, hypoechoic, solid mass. CT Scan demonstrates a low-density mass with peripheral enhancement after intravenous contrast medium. MRI can be helpful in the differential diagnosis of infarction by suggesting hemorrhagic necrosis on the T2-weighted images. Diagnosis is corroborated during laparoscopy or laparotomy. Accessory splenectomy is curative.

Removal of the spleen in children is considered necessary in some hemolytic diseases and trauma. Hemolytic disease where splenectomy is therapeutic includes idiopathic thrombocytopenic purpura, congenital spherocytosis and Sickle cell disease. Removal of the spleen with its attendant immunologic shortness can cause overwhelming sepsis. Overwhelming postsplenectomy infection is a fulminant process that carries a poor prognosis. The risk of serious infection is highest among young children, in immunologically compromised individuals and in the first few years after splenectomy. Current guidelines for children undergoing elective splenectomy include immunization for pneumococcus, meningococcus and hemophilus at least two weeks prior to the procedure. After emergency splenectomy the immunization protocol is the same, except that the amnestic response of the host is lower. Another guideline for splenectomized children includes the use of continuous antibiotic prophylaxis (penicillin) until the age of twelve years. Compliance with this antibiotic regimen is poor. It is recommended that parents be advised to bring the postsplenectomy child to the hospital anytime an illness or fever develops that might require an immediate loading dose of an appropriate antibiotic.

Splenic artery embolization as an alternative for splenectomy is a minimally invasive technique that has been utilized for the past 25 years. The procedure is done under local anesthesia, sedation or general anesthesia. Partial splenic artery embolization has been used in cases of thalassemia major to reduce transfusion requirements with variable results. Also in children with secondary hypersplenism or massive spleens due to portal hypertension, variceal bleeding (portal vein thrombosis, biliary atresia and biliary

cirrhosis) or myelodysplastic disorders. The leukopenia and thrombocytopenia is corrected temporarily and variceal hemorrhage is ameliorated in most cases. Objective is to embolize at least two-third of the splenic circulation. After splenic artery embolization the child develops prolonged fever, pain in the left hypochondrium, leukocytosis and ileus. If the embolization causes total shutdown of the arterial circulation the child might develop a subcapsular hematoma with effusion, abscess formation or need for open splenectomy. Embolization allows for safe surgical splenectomy. Long term follow-up shows evidence of partial splenic regeneration. Partial splenic embolization is a useful method for reducing serum bilirubin concentrations in patients with hypersplenism following the Kasai procedure for biliary atresia, has also been found safe and effective minimally invasive treatment for patients with bleeding from a blocked distal splenorenal shunt, and as therapy for post-traumatic splenic artery pseudoaneurysm.

J. Adrenal Incidentaloma

With the advent of potent imaging studies during the eighties a group of adult patients was found with incidentally discovered adrenal masses, hence the term coined of adrenal Incidentaloma. At the time, masses below a size of three centimeters were observed with follow-up studies for spontaneous regression. Most cases resulted in benign non-functioning adenomas which disappeared with time. The situation in children is different. A mass identified in the adrenal gland is cause for concern. In infancy and childhood the most common adrenal mass is the neuroblastoma, a malignant neural crest tumor. Initial diagnosis of an adrenal mass in a child is made with Ultrasound, which is also used to document regression of uncomplicated neonatal adrenal hemorrhage. Further radiological assessment of an adrenal incidentaloma in a child should include CT-Scan and MRI. MRI can accurately distinguish adrenal adenomas from adenocarcinoma, pheochromocytoma and neuroblastomas. Endocrine tests evaluating pituitary-adrenal function (urinary excretion of 17-hydroxycorticosteroids, 17-ketosteroids and catecholamines, plasma concentrations of ACTH, cortisol, DHEAS, androstenedione and testosterone, dexamethasone suppression test and corticotrophin-releasing hormone stimulation test) should be part of the work-up. Should biochemical studies revealed no hormonal related disease (Cushing, hyperaldosteronism, pheochromocytoma, etc.) a histological diagnosis should be obtained by either CT-guided fine needle biopsy or surgical resection. In the event of no diagnosis, adrenal tumor resection should be done.

K. Budd- Chiari Syndrome

Obstruction to the hepatic venous outflow tract is commonly known as Budd-Chiari Syndrome. The Budd-Chiari syndrome (BCS) in children can be the result of a congenital or acquired web in the inferior vena cava, a thrombotic, inflammatory, neoplastic process or an hypercoagulable state (antithrombin 3 deficiency). Hepatic venous outflow obstruction produces hepatic dysfunction producing abdominal pain, ascites, jaundice, hepatosplenomegaly, portal hypertension and cirrhosis. The factors that influence management of the BCS include the state of hepatic dysfunction, type of presentation (acute or chronic), how much venous occlusion is present and the presence of collateral circulation. Pulsed Doppler ultrasound, venography and liver biopsy are very helpful in diagnosis. Management of BCS in children has included use of anticoagulation, thrombolytic therapy, angioplasty with or without stenting, transjugular intrahepatic portosystemic shunts and surgical portosystemic shunts. This last choice has fewer options in the face of liver transplantation and does not improve survival. The combination of thrombolytic therapy and balloon angioplasty is the best option in the acute setting of BCS or during the first four weeks after development of the syndrome. Late or chronic presentation with established hepatic cirrhosis and portal hypertension sequelae is best managed with liver transplantation. Early diagnosis offers the best possible chance of cure.

L. Gallstone Ileus

Gallstone ileus is a very rare disorder characterized by mechanical obstruction of the gastrointestinal tract due to incipient impacted gallstones that passes through a bilio-enteric fistula. Impaction followed by obstruction can occur at the ileum, duodenum or stomach (Bouveret's syndrome). Diagnosis is usually delayed due to lack of specific signs of biliary disease. The classic triad of Rigler (small bowel obstruction, ectopic gallstones and air in the biliary tree) is visualized on abdominal plain films in only one-thirds of cases. Age ranges from 13 to 87 years with most cases seen in older patients. Most bilio-enteric fistulas are cholecystoduodenal type, with a few choledochoduodenal. Work-up includes ultrasound, upper

gastrointestinal series with water soluble contrast medium and contrast enhanced computed tomography (CT). Preoperative diagnosis of gallstone ileus significantly increases by combining plain film and US findings. Management consists initially of simple enterotomy (enterolithotomy) which can be done laparoscopically assisted. This is followed by takedown of the bilioenteric fistula and cholecystectomy in a later stage procedure if the medical condition of the patient permits and he continues symptomatic. Some reports encourage enterolithotomy, repair of the fistula and cholecystectomy in one procedure. Other workers report that enterolithotomy alone is adequate treatment in the elderly, and subsequent cholecystectomy is not mandatory. Early diagnosis and treatment improve the outcome.

M. Portal Hypertension

Portal hypertension (PH) in children is caused by increased portal venous flow from such conditions as hemangiomas or hepatic arterioportal fistulas, or by increase resistance to flow from conditions such as portal vein thrombosis, liver cirrhosis, congenital fibrosis, biliary atresia, neonatal hepatitis or hepatic vein thrombosis. In children, extrahepatic obstruction due to portal vein thrombosis is the most common cause. Most common presentation of PH is upper gastrointestinal bleeding from esophageal or gastric varices, followed by splenomegaly with hypersplenism. Diagnostic studies include liver function tests, upper endoscopy, color Doppler US, splenoportography and MRI. Initial management of PH can entail the use of vasoactive beta blockers such as propanolol or somatostatin. Bleeding varices can be managed with banding or sclerotherapy. Children with favorable liver function, but unfavorable anatomy and continuous variceal bleeding can benefit from a devascularization procedure. Those with unfavorable liver function and bleeding can benefit from transjugular intrahepatic portosystemic shunt (TIPS), though shunt thrombosis is a problem the smaller the kid. Children with favorable anatomy can benefit from a distal splenorenal (or splenoadrenal) shunt, or a makeshift shunt such as the Rex shunt between the inferior mesenteric vein and a branch of the portal vein high in the hepatic hilum using autologous vein graft. Liver transplantation is the treatment of choice for children with PH complicating end-stage liver cirrhosis.

N. Cholelithiasis in Sickle Cell Disease

Cholelithiasis is a frequent complication of Sickle Cell Disease (SCD) in children. Prevalence of developing pigmented stones can range from thirty to 50%. Even the prevalence of developing common bile duct stones in children with SCD is higher than the general population. Complications related to cholelithiasis include biliary colic, pancreatitis, cholecystitis and obstructive jaundice. Children with SCD should be screened for cholelithiasis since the age of eight years using abdominal ultrasound. Once cholelithiasis is identified removal of the sick gallbladder should be accomplished regardless if the child is asymptomatic or not. Preoperative transfusion reduces the morbidity of the surgical procedure. Elective laparoscopic cholecystectomy should be the gold standard in children with SCD and asymptomatic cholelithiasis to prevent the potential complications of biliary colic, acute cholecystitis, and choledocholithiasis, which lead to major risks, discomfort, and longer hospital stay. Laparoscopic cholecystectomy results in a shorter hospital stay with fewer postoperative complications than open operation in patients with sickle cell disease and is the procedure of choice in the treatment of cholelithiasis in such patients.

O. Alagille Syndrome

Alagille syndrome is an autosomal dominant disorder characterized by paucity of intrahepatic bile ducts, typical facies, congenital cardiac defects (pulmonary stenosis), posterior embryotoxon of the eye and butterfly vertebra arch defect. Due to paucity of bile ducts the baby presents with inefficient bile excretion causing intrahepatic cholestasis, direct hyperbilirubinemia and hypercholesterinemia. The clinical picture is sometimes indistinguishable from biliary atresia needing cholangiogram and liver biopsy for diagnosis. The vast majority of patients present with jaundice and failure to thrive or cardiovascular symptoms before six months of age. Mutation in the JAG-1 gene of chromosome 20p12 is responsible for more than 90% of cases. There is no cure for Alagille syndrome. The most disturbing manifestation includes pruritus and xanthomas. Most children with Alagille syndrome can be managed conservatively with choleretics (ursodeoxycholic acid), nutrition optimization, fat-soluble vitamin supplementation and medication for pruritus (cholestyramine, rifampin, naltrexone). Surgery is aimed at reducing the pruritus consist of partial external biliary diversion creating a conduit between gallbladder and skin using jejunum, performing an ileal exclusion end to side

ileocolostomy between proximal ileum and right colon or partial internal biliary diversion between gallbladder and ascending colon with jejunum. These procedures do not improve growth and does not prevent progression of disease toward liver failure. Liver transplant is used for liver failure. Mortality is approximately 10%, with vascular accidents, cardiac disease, and liver disease accounting for most deaths.

P. Peritoneovenous Shunt

Peritoneovenous shunt (PVS), also known as Leveen or Denver shunt, is a shunt utilized to manage medically intractable ascites in adults and children. These shunts allow ascitic fluid to flow down a pressure gradient from the peritoneal cavity to the venous circulation and have a valve mechanism that prevents backflow of blood if the venous pressure rises above the intraabdominal pressure. The advantage of the Denver shunt is that the valve chamber lies in the subcutaneous tissue and therefore can be manually compressed to relieve blockage and promote flow. The shunt can also be flushed percutaneously if necessary. When peritoneal pressure is 3 cm higher than CV pressure the valve opens. PVS can be placed surgically, laparoscopically-assisted or percutaneously. Persistent ascites is rare in children, carries a significant morbidity and is a difficult management problem owing to the massive abdominal distension. Etiology is often related to previous surgery, congenital malformation of lymphatic channels, or idiopathic. Other causes include inflammatory, neoplastic, traumatic, mechanical obstruction or nonaccidental injury. Conservative and symptomatic management is usually the mainstay of treatment while surgery is indicated when conservative therapy fails. Certain complications have been described in association with the procedure of placing the PVS. One of the major concerns is diversion of large amounts of fluid into the central circulation, potentially contributing to fluid overload. In anticipation of this potential complication, diuretic therapy is initiated that seemed to avoid this problem. Other reported complications include shunt blockage and leaks, venous thrombosis, disseminated intravascular coagulation, infection, and air embolus during insertion. The PVS is an effective alternative in management for intractable ascites in children.

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 consists 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.

Congenial 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 treatments are question waiting answer in the near future.

Opsoclonus-myoclonus-ataxia (OMA) syndrome is a rare paraneoplastic or paraviral neurologic syndrome commonly associated with neuroblastoma. Less than 3% of children with neuroblastoma develop OMA syndrome. Clinically the child with OMA syndrome develops an acute onset of rapid chaotic eye movements, myoclonic jerking of the limbs and extremity and ataxia. OMA is believed to be an immune-mediated disorder due to the detection of antineural, antineurofilament and anti-Hu antibodies. The child with OMA syndrome caused by neuroblastoma has a favorable staged disease due to rapid work-up and findings

that the patient's immune response to the tumor limits the metastatic and growth potential of the tumor. Rarely do these tumors shows n-MYC gene amplification, a poor prognostic finding. Children with neuroblastoma and OMA have an excellent survival. There is no correlation with duration of symptoms and late neurologic outcome. Most children respond to treatment of acute symptoms with steroids or ACTH. Late neurologic sequelae (delay in motor function, speech and cognition) of OMA children can be drastic and affect quality of life. Children with advanced stage disease require more intensive chemotherapy and have better outcomes with regard to late neurological sequelae. The higher the immune response limits the spread of disease but increases the neurologic sequelae.

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, covert 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 maturate 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.

Sacrococcygeal 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 consists 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.

Mediastinal teratomas are rare tumors that originate in the anterior mediastinum and comprise almost one-fifth of all mediastinal masses. Most grow to large size before causing symptoms. Mediastinal teratoma can appear in any age of the child. Primary symptom is respiratory distress caused by airway compression, followed by feeding problems related to dyspnea, coughing, wheezing and chest pain. Most mediastinal teratomas are mature and benign. Teratomas arise from pluripotent cells and are composed of a wide diversity of tissues originating from three germ layers ectoderm, mesoderm and endoderm. Besides an anterior mediastinal mass, plain chest films can show calcifications. CT Scan is the study of choice to demonstrate the extent of the tumor and its relationship with other structures. As with any other suspected teratoma preoperative alpha fetoprotein and human chorionic gonadotropin markers levels should be obtained. Teratomas are classified as mature, immature and malignant. Mature teratomas are predominantly cystic, while malignant teratomas are mainly solid lesions. Immature teratomas have immature tissue with mature elements. Surgical excision through a median sternotomy is the treatment of choice for mediastinal teratomas. Adjuvant chemotherapy is used in immature and malignant teratoma to increase survival.

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.

Sertoli-Leydig cell ovarian tumors are rare androgen producing tumors causing masculinization in

most girls. A few are nonfunctional tumors. Sertoli-Leydig cell tumors used to be called arrhenoblastoma or androblastomas. One-third of all Sertoli-Leydig cell tumors (SLCT) occur in children. Most SLCT are unilateral. Histologic diagnosis depends on the presence of heterologous endodermal and mesenchymal components. The androgenic effect of the tumor causes accelerated somatic growth and amenorrhea in prepubertal girls. Postpubertal girls develop irregular menstrual cycles, hirsutism and masculinization. Most affected children usually present with a pelvic mass. Testosterone and alpha-fetoprotein produced by the tumor are used as genetic tumor markers. Diagnosis is usually done by ultrasound or CT-Scan in association with the masculinizing clinical picture. Management consists of unilateral salpingo-oophorectomy. Poorly differentiated tumors might need adjuvant chemotherapy and radiotherapy. Prognosis correlates most meaningfully with the stage and degree of differentiation of the tumor. High-stage tumors are all clinically malignant.

G. Thyroid Nodules & Multinodular Goiter

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.

Non-toxic multinodular goiter (MNG) refers to multinodular enlargement of the thyroid gland without overt hormone output. MNG is rare in children affecting primordially adolescent kids. The etiology of pediatric MNG appears multifactorial including autoimmune and familial factors (familial form has increased incidence of malignancy). Children presents with asymptomatic progressive nodular enlargement of both lobes of the thyroid gland. Work-up should include neck ultrasonography, thyroid scintigraphy, thyroid hormone levels, assessment of autoantibodies (antimicrosomal, antithyroid), aspiration cytology and histological examination. In populations with iodine deficiency, multinodular goiter is endemic. MNG follows an initial phase of hyperplastic goiter or results from the generation of several individual nodules. Alterations of the stromal and vascular tissues as well as the occurrence of somatic mutations are contributing factors. Histological examination of removed affected glands shows multiple adenomas with areas of epithelial hyperplasia, hemorrhage, and calcification. MNG has an 8% potential for malignant transformation in the form of papillary carcinoma, mostly increased in familial cases, those that have received cervical irradiation and presence of cervical adenopathies. Indications for surgery in non-toxic MNG includes compression symptoms such as painful or difficulty in swallowing, breathing discomfort, suspicion of carcinoma or cosmetic. Total thyroidectomy seems to be the most effective surgical procedure with lower morbidity than subtotal thyroidectomy.

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.

I. Gastrointestinal Stromal Tumor

Gastrointestinal stromal tumor (GIST), previously known as gastric leiomyoblastoma, is an uncommon nonepithelial mesenchymal kit-positive (CD117 antigen) tumor of the gastrointestinal tract. GIST are the most common mesenchymal tumors of the gastrointestinal tract. Cell of origin is the interstitial cell of Cajal. The frequency of malignant GIST is 20-30% of the frequencies of all soft-tissue sarcomas, but small benign tumors often found incidentally during unrelated surgery or autopsy are more common. GIST occurs in children, young adults or on a familial basis. Most involved children are girls with symptoms of abdominal pain and anemia. CT-Scan or MRI suggests the diagnosis. Most GIST appears in the stomach (submucosal mass), followed by the intestine and rarely the colon. Metastasis occurs to the liver. Large tumors (> 5 cm) with high mitotic activity are associated with bad prognosis. Management consists of complete surgical resection with prophylactic omentectomy to reduce the recurrence of GIST. GIST have lower survival rate and more resistance to chemotherapy.

J. Osteochondromas

Osteochondroma is the most common benign bone exostosis found in children. Osteochondroma most frequently arise sporadically and as a solitary lesion, but may also arise associated with hereditary multiple exostosis. Hereditary multiple exostosis is an autosomal dominant disorder in which the clinical hallmark is the growth of bony protuberances from long bones causing a variety of orthopedic deformities. In hereditary multiple osteochondromas the prevalence is one in 50,000 individuals. Ten percent of affected children have no family history of multiple exostosis. Median age at the time of diagnosis is three years. Most cases present with an obvious deformity of the forearm, followed by an inequality in the lengths of the limbs, an angular deformity of the knee, or a deformity of the ankle. Symptomatics complications of osteochondroma consists of pain, fracture, osseous deformity limiting range of motion, vascular injury, neurological compromise, bursa formation and malignant transformation (chondrosarcoma). MRI is the ideal imaging modality in the diagnostic evaluation of symptomatic complications of osteochondromas and often avoids the need for further imaging. Spontaneous resolution of a solitary osteochondroma is rare. Management of symptomatic osteochondromas is surgical excision. Surgical complications associated with excision consist of peroneal neurapraxias, arterial laceration, compartment syndrome and fibular fracture. The surgical risk for the management of osteochondromas is low.

K. Juvenile Secretory Carcinoma

Carcinoma of the breast in a child is a rare pathologic entity, constitutes less than 1% of all breast lesions in this age group. Juvenile secretory carcinoma is an uncommon malignant tumor that can develop in the breasts of both sexes with a mean age of occurrence at 10 years. It's a slow growing tumor that can recur locally and metastasize to the ipsilateral axillary lymph nodes. Juvenile secretory carcinoma component are devoid of estrogen and progesterone receptors. The child develops a painless mass often near the areola in the breast early in life. The lesion is well demarcated and unencapsulated invading the adjacent tissue. Immunohistochemical staining for alpha lactalbumin is present. When in doubt fine needle aspiration biopsy (cytology) can provide a preoperative diagnosis of the lesion. In all cases, the samples are cellular and feature diffuse, prominent, intracytoplasmic vacuoles and secretion in malignant cells with occasional signet-ring like forms. Management consists of simple mastectomy (wide local excision) with sentinel axillary lymph node biopsy, especially in males cases were metastasis to the axilla are more common. Biological behavior seems to be similarly favorable in both sexes.

L. Vascular Access

Central venous access in children is a necessity for drawing blood, administering blood products and chemotherapeutic agents, and providing parenteral nutrition. Access through the various sites such as the

internal and external jugular veins, subclavian and saphenous vein can be plague of complications. Immediate complications at the time of the procedure includes failure to achieve successful access, pleural laceration with pneumothorax development, laceration of the vein with hemothorax, shock, and extravascular placement of the catheter leading to infection, airway compression and pericardial tamponade. Unless you do the procedure fluoroscopically, it is imperative to obtain a chest film immediately after central venous access to confirm adequate position of the catheter and check for the above mentioned complications. Complications associated with long term vascular access include infection (local or systemic bacteremia), occlusion of the catheter, and chronic erosion of the catheter through the wall of the vessel with extravasation. Deep venous thrombosis can occur due to nidus deposition of fibrin. Catheter breakage with embolization is another complication of long standing access.

Implantable central venous catheters constitute a necessity for the management of long term intravenous nutrition and chemotherapy. Implantable central venous access devices placed via the subclavian vein may become obstructed by thrombosis, impingement against a vein wall, or compressed between the clavicle and first rib. Compression of the catheter between the clavicle and first rib is known as pinch-off syndrome (POS). Beside obstruction, pinch-off syndrome can cause fragmentation, fracture or rupture of the catheter causing embolization of the released fragment of tubing. Mechanical friction against the catheter has been well established as the mechanism for most fractures. POS is characterized by intermittent catheter malfunction in conjunction with radiologic evidence of catheter compression. Warning signs of POS include difficulty withdrawing blood samples and resistance to infusion of IV fluids. Catheter transection with migration of the catheter into the heart or pulmonary artery may be accompanied by the sudden onset of chest pain, palpitations, and arrhythmias. Electron microscopic scanning tends to prove that the catheter's rupture is caused by a fatigue process. Treatment of POS is removal of the catheter. If the tip of the catheter has embolized, it can usually be retrieved percutaneously with a transvenous snare. POS can be prevented by using the internal jugular vein for access rather than the subclavian vein.

Hemoports catheters play a vital role in providing continuous central venous access for such therapy as parenteral alimentation, long-term antibiotics and cancer chemotherapy in children and adults. The tip of the hemoport catheter should lie within the superior vena cava or right atrial junction during placement. Placement can be done through the external or internal jugular vein or using the subclavian vein with the port usually lying infraclavicularly in the anterior chest area. Very rarely fragmentation with embolization of the port catheter can occur specially during removal of the port. Incidence of catheter fractures is 0.1%. Fracture is suspected if the catheter offers resistance to removal and/or the length removed is too short. The fragmented retained catheter can cause endocarditis, thrombosis, pulmonary abscess, dysrhythmia or sudden death. Causes of fracture include manufacturing defect, mechanical trauma, excessive hydrostatic pressure when flushing or infusing, material degradation, stress due to constant motion, deposition of fibrin, clot or calcium within the catheter, or pinching between the clavicle and the first rib. The fragmented catheter should be differentiated from a calcified “ghost” cast by CT-Scan. The fragmented catheter can stay within the vascular vessel, or embolized into the right heart or pulmonary arteries. Management should consist of percutaneous endovascular retrieval by an invasive cardiologist or radiologist.

M. Diaphragmatic Tumors

Tumors arising from the muscle or elements of the diaphragm are very rare in occurrence. The small published series has shown that the incidence is similar between boys and girls along with left or right involvement. Most primary tumors arising from the diaphragm are malignant, with rhabdomyosarcoma the most commonly encountered followed by sarcomas, yolk sac tumors and extraosseous Ewing sarcoma. Lymphangiomas and hemangiomas are the most common benign tumors found in the diaphragm. The clinical presentation in children varies, with predominantly chest symptoms (chest pain, shortness of breath, cough, chest asymmetry or hemothorax). Identifying the site of origin of the tumor to the diaphragm is difficult even after using CT, MRI and ultrasound. Exploratory laparotomy with biopsy is the best tool to assign location to the tumor. Management of primary diaphragmatic tumors encompasses wide local resection with reconstruction, chemotherapy and radiotherapy. To obtain cure, a tumor free resection margin must be obtain initially or after chemotherapy shrinkage of the tumor. Reconstruction of the diaphragm at the time of

resection can be accomplished with a muscle flap or prosthetic graft (PTFE or Gore-Tex).

N. Gonadoblastoma

Gonadoblastoma is a sex cord gonadal tumor that contains both germ cell and sex cord stromal elements. It occurs almost exclusively in sexually abnormally individuals with gonadal dysgenesis and Y-containing cells, while other cases occur in children with mixed gonadal dysgenesis (mosaic 45XO/46XY). The combination of the Y chromosome with a dysgenetic gonad is all that is needed for a gonadoblastoma or dysgerminoma to develop. The tumor is usually quite small and calcifications are common. Almost 40% of all gonadoblastomas are bilateral. The germ cell component may outgrow the stromal elements and result in the formation of a dysgerminoma. Most cases will appear in young female adults with history of primary amenorrhea during teenage years and virilization. Management of gonadoblastoma consists of removal of both dysgenetic gonads irrespective of the bilaterality of the lesion. Because these tumors occur in up to 50% of patients with gonadal dysgenesis early bilateral prophylactic gonadectomy should be performed. Gonadoblastomas can exhibit either benign or malignant features, though most cases are benign tumors that have a good prognosis after excision. Gonadectomy can either be done open or laparoscopically. With the presence of malignant germ cell elements, chemotherapy will be needed. Other children at risk to develop gonadoblastoma later in life include those with Turners and androgen insensitivity syndrome.

O. Pancreatic Carcinoma & Frantz Tumor

Tumors of the pancreas can arise from exocrine or endocrine cells. Adenocarcinoma of the pancreas is an extremely rare tumor found during the pediatric age. Most adenocarcinomas of the pancreas are non-islet cell lesions of ductal origin with more than 70% located in the head of the pancreas. Others are acinar cell carcinomas and nonfunctional islet cell carcinomas. More than half the cases are females with a mean age of nine years (range three months to 18 years). As in adults this is a very aggressive malignant neoplasm with early metastatic spread to lymph nodes and liver. While the majority of pancreatic tumors are exocrine lesions of ductal origin, acinar cell tumors are more commonly observed in children and associated with a better prognosis. Children with pancreatic ductal carcinoma presents with abdominal and back pain, vomiting, obstructive jaundice, a palpable mass and weight loss. Diagnostic imaging should consist of ultrasound, CT-Scan and MRI to look for chances of surgical resectability. Genetic markers such as CEA, C19-9, and alpha fetoprotein should be obtained. Whenever feasible and in the event of no metastatic disease management should consist of surgical resection of the tumor. Body and tail tumors can be dealt with distal pancreatectomy, while head of pancreas tumors will need pancreaticoduodenectomy (Whipple procedure). The response rate with chemotherapy and/or radiotherapy is very poor. The prognosis with metastatic disease is dismal.

Papillary cystic tumor of the pancreas, also known as Frantz tumor (FT) since 1959, occurs predominantly in girls and young women (mean age 21 years). Abdominal pain and a slowly growing incidentally found epigastric mass is the most common complains, associated at times with weight loss, anorexia and vomiting. FT is well-encapsulated, shows solid and hemorrhagic patterns, contain PAS-positive cytoplasmic or prozymogen granules as seen in acinar cell tumors and behaves as a low-grade malignancy. CT scans suggest the diagnosis (thick capsule, calcifications, mixed solid and cystic patterns, grows toward the outside of the pancreas). Differential diagnosis includes traumatic pseudocysts, serous and mucinous cystadenomas of the pancreas. Immunohistochemically the tumor is positive for alpha-1-antitrypsin while negative for insulin and glucagon. Complete removal is the treatment of choice for tumor arising in any part of the pancreas. FT is frequently amenable to local resection and has a good long-term survival rate after excision. Metastasis (liver) or local recurrence occurs in 10% of cases. Older age at diagnosis or recurrence disease increases the malignant biological behavior of the tumor. Radiotherapy and, or chemotherapy are of no use for its treatment.

P. Genital Tumors

Tumors occurring in the vulva and external vaginal orifice are rare to find in female children. In the area of the vulva the most common tumor is either and hemangioma, lymphangioma, lipomas, neurofibromatosis and vulvar intraepithelial neoplasia. Hemangiomas resolve with conservative therapy, while lymphangiomas or other type of tumor will require surgical excision. Vulva intraepithelial neoplasia are

associated with cases of sexual abuse and human papilloma viral infections. In the external vaginal orifice bleeding hemangiomas have been previously reported in children. Cavernous hemangiomas with brisk and continuous bleeding will require some form of therapy such as cryosurgery, excision or steroid injection. Interferon has also been used effectively. It is always important to study with imaging (MRI) the extension of the perineal hemangioma. Another lesion of importance in this privilege area is the vaginal rhabdomyosarcoma which presents with protrusion and a bleeding mass. Management consists of a biopsy, chemotherapy followed by surgery if necessary.

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 & Torsion

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.

Ovarian torsion is a true surgical emergency mostly affecting woman during their first three decades of life. Ovarian torsion results from partial or complete twisting of the ovarian pedicle on its axis causing vascular compromise, congestion and hemorrhagic infarction. Clinically, most children present with an abrupt onset of low abdominal pain, low-grade fever, nausea and vomiting sometimes mimicking symptoms of acute appendicitis. Mean age is 12 years. Symptoms may be recurrent. Pain is localized toward the affected ovary. Most ovarian torsion are associated with a concomitant ovarian cystic or solid mass. Neonates with ovarian torsion are usually diagnosed during prenatal ultrasound studies when a cystic mass is identified. Ultrasound is the imaging study of choice. An enlarged solid ovarian mass with peripheral cysts noted at US suggests the diagnosis of torsion and should be followed by diagnostic laparoscopy or exploration. A prepubertal child will have a complex mass. CT Scan demonstrates a heterogenous, retrovesical mass. Color Doppler US sometimes confirms the absence of ovarian blood flow. Management is established at exploration. Removal of a mass carrying a twisted ovary is standard therapy. Untwisting of torsed adnexa and observation in case of absence of a mass (normal ovaries) are accepted modes of treatment. Most common encountered morbidity is postoperative fever. Contralateral oophoropexy in cases of torsion of a normal adnexum is advice.

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/pubic 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 hypospadia. 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 advice 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 antimüllerian 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 testes in MDS are at risk of malignant degeneration.

H. Bleeding Nipple

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.

I. Breast Fibroadenoma, Cysts & Papillomas

Fibroadenoma is a common benign tumor found in the breast of adolescent girls. It is also considered the most common discrete solid mass found within the adolescent breast tissue. Most girls harboring a fibroadenoma have between thirteen and 16 years of age, the tumor is slow growing, tends to develop in the upper outer quadrant and is more common in African-American race. Though females may develop breast masses early in life, the risk of malignancy is extremely low. The tumor is usually solitary, with an average diameter of two to 4 cm, characterized by rich cellular stroma and prominent glandular epithelium. At physical exam the mass feels like a well-circumscribed movable nodule. Fibroadenomas may be related to an exaggerated local response to the estrogenic effects of puberty. Mammography, due to the inherent radiation

risk and dense fibrous tissue, is not recommended for routine screening or routine imaging of breast masses in adolescents. Alternatively, sonography of the breast is diagnostic on most cases. The tumor looks well-circumscribe, hyperechoic and homogenous on ultrasound. Ten percent of cases harbor a giant juvenile fibroadenoma, a large lesion that distorts the normal breast architecture eroding through the skin and areolar complex. Management of fibroadenoma could be observation or cryoablation. Growing, symptomatic or anxiety-producing masses should be managed with excision through a periareolar incision to preserve cosmesis.

Juvenile or giant fibroadenoma of the breast is a benign lesion that can obtain a large formidable proportional size during breast development in female adolescent patients. Most cases in children are seen between the ages of 10 and 15 years. The tumor is solitary in most affected children with a diameter of 4-6 centimeters. Multiple and bilateral involvement has been reported in a few cases. Differential diagnosis includes cystosarcoma phyllodes, benign virginal hypertrophy (juvenile gigantomastia) or rhabdomyosarcoma. FNA or Tru-cut needle biopsy can establish a precise histological diagnosis. Growth is so fast that it can cause non-tender cellulitis of the skin by way of stretching. Microscopically the tumor is characterized by a rich cellular stroma and a prominent glandular epithelium. Juvenile adenofibromas regardless of size should be excised so as to preserve as much breast tissue as possible. Management options include local excision with reconstruction, reduction mammoplasty, or simple mastectomy with reconstruction.

Breast cysts are very common in the adolescent female, while not so common in males. Rapid cyst growth causes pain and a palpable mass in the breast that brings the child to seek medical help. Other times breast cysts get infested and are managed as a breast abscess with antibiotics and drainage. In males solitary, large male breast cysts are extremely rare. They occur mainly in children up to the age of seven years and should be removed under surgery. Whenever a breast mass develops the next step in diagnosis is an ultrasound-mammography studies. This will corroborate the cystic or solid nature of the cyst along with its size. Radiation exposure in children using mammography is not necessary or warrant. Management of breast cysts in females consists of observation or aspiration of the cyst with cytological exam. The overwhelming majorities of breast cyst in children are benign and will go spontaneously with time. Follow-up breast ultrasound of the child for persistent of the mass or bloodstained aspirate should be done.

Breast juvenile papilloma in children is a rare benign lesion featuring atypical papillary duct hyperplasia and numerous cysts. They manifest clinically as a localized, multinodular mass that is usually interpreted as a juvenile fibroadenoma. Most cases occur in females, though some cases in males have been reported. Mean age of diagnosis occurs during the late adolescent years. Left breast is affected slightly more often than the right. Patterns of menarche, marital history, parity, and use of birth control pills are not exceptional for women in this age group. No instance is found of maternal use of estrogens during pregnancy. Family history of breast carcinoma is seen in one-thirds of all cases of papillomatosis. Juvenile secretory carcinoma can be associated with papillomatosis. Breast ultrasonography will show an ill-defined, inhomogeneous mass with numerous small, hypoechoic areas, but cannot differentiate a fibroadenoma from papilloma. Excisional biopsy through a periareolar incision will establish the diagnosis. Should a secretory carcinoma be found wide local excision is warranted. Due to the precancerous nature of papillomatosis, long-term yearly follow-up is recommended.

J. Supernumerary Nipple

Supernumerary (or accessory) nipple, also known as polythelia, is a congenital developmental abnormality that occurs most commonly over the anterior aspect of the trunk in the pathway of the embryonic milk line extending from the axilla to pubic region. Supernumerary nipple shows all the histologic components observed in the normal nipple including epidermal thickening, pilosebaceous structures, smooth muscles and mammary glands. Incidence of supernumerary nipple is 25 per 1000 live births with a higher prevalence for the left side and male gender. The accessory nipple is a cosmetic defect with the potential to give rise to a neoplasm since any disease process that involves anatomically normal breasts may affect aberrantly located breasts or nipples as well. Due to its atypical appearance and ectopic location, diagnosis of the anomaly may require a high index of suspicion along with histologic verification. Once thought to have an association with renal defects, the presence of supernumerary nipple in asymptomatic children is not an indication to do

additional diagnostic studies of the urinary tract. Supernumerary nipple and ectopic breast are different entities. Supernumerary nipples can be identified at birth, whereas ectopic breast tissue becomes noticeable only after hormonal stimulation, usually during puberty, pregnancy or lactation. Management of supernumerary nipple consists of excision for diagnosis, treatment of symptoms, or cosmesis.

K. Vaginal Conditions

Transverse vaginal septum is a congenital condition of females that can block the passage of vaginal secretions causing primary amenorrhea, hematocolpos and cyclic pelvic pain. Different to imperforate hymen, in a transverse vaginal septum you find a rim of nonbulging hymenal tissue on the vestibular floor with an intravaginal bulging membrane. The septum can be found in the upper, middle or lower vagina varying in thickness. Most common location of the septum is the upper vagina. Histologically, the diagnosis of transverse vaginal septum is made due to the presence of müllerian duct (mesodermal origin) tissue in the septum. Transverse vaginal septum is a defect of vertical fusion during embryogenesis of the vagina. The estimated incidence is one per 30,000 to 84,000 women. It is sometimes associated with genitourinary tract, gastrointestinal tract, musculoskeletal, and cardiac malformations. Physical exam and pelvic ultrasound are diagnostic. Surgical resection is the treatment of choice. The mucosa on either side of the blockage should be mobilized for approximation with interrupted sutures, while the underlying fibrous septum should be excised. Postoperative dilation may be necessary to prevent restenosis and dyspareunia. Patients with a complete transverse septum in the middle or upper vagina are less likely to conceive than patients with a septum in the lower vagina. Prompt diagnosis and surgical correction to drain accumulated blood may preserve fertility possibly through the prevention of endometriosis.

A septate vagina is another congenital condition that occurs from failure of longitudinal fusion of the lower müllerian ducts leaving a variable amount of an asymptomatic longitudinal vaginal septum in the child. Two vaginal canals are created by the septum completely or partially. As time passes the septate vagina goes unnoticed until the adolescent child start to use tampons or engage in sexual activity. The affected patient might complain of menstrual leakage with the use of tampons since menstruation will continue to egress from the other vaginal canal not occluded by the tampon. This is the case with complete duplication of the müllerian system. Reassurance and vaginal hygiene is all that is required in these cases. Physical exams including endoscopy along with pelvic ultrasound are diagnostic. The identification of a duplicated cervix and a vaginal septum is consistent with several uterine malformations, which leads to frequent misdiagnosis and errors in management. On the other hand when the defect is an isolated partial vaginal septum the patient will complain of dyspareunia during sexual intercourse. Management in these cases consists of excision of the thick septa (septectomy) maintaining good hemostasis.

Vaginal bleeding in the pre-menstrual female infant is cause for concern both medically and socially. Differential diagnosis of vaginal bleeding in this age group includes estrogen stimulation, vulvovaginitis, tumors of the lower genital tract, ovarian tumors, foreign bodies, or trauma. Transplacental estrogen stimulation can cause self-limited vaginal bleeding in newborns during the first two weeks of life. Vulvovaginitis is the most common gynecological infection in children caused by ascending enteric organisms due to poor hygiene and is managed with systemic antibiotics. Condyloma can cause painless vaginal bleeding. Tumors of the genital tract associated with vaginal bleeding include hemangiomas of the vulva, arteriovenous malformation of the uterus, rhabdomyosarcoma botryoid (the most common malignant tumor of the low genital tract in young females), endodermal sinus tumors of the vagina and endometrial carcinomas. Functional ovarian or adrenal tumors that produce estrogen can be associated with sexual precocity and vaginal bleeding. Foreign bodies in the vagina of a small girl produce local inflammation resulting in a foul smelling discharge which can be serosanguineous. The debris (foreign body) is often wads of toilet paper. Redundant urethral mucosa may prolapse through the urethral meatus and present as a friable polypoid lesion. Finally genital injury is a major cause of vaginal bleeding including those associated with child sexual abuse.

L. Pelvic Inflammatory Disease

Acute pelvic inflammatory disease (PID) is a major gynecologic health problem in the USA, afflicting more than 1 million women each year. PID continues to be a common diagnosis among adolescent girls

presenting with low abdominal pain. Adolescents have a higher rate of diagnosis of PID than any other age group. PID is an ascending polymicrobial infection affecting the upper genital tract. Risk factors associated to PID include young age, age at first intercourse, multiple sex partners, the presence of bacterial vaginosis, vaginal douching, the use of an intrauterine contraceptive device, and a history of a sexually transmitted disease. Classic symptoms of pain, fever, and a history of high-risk sexual behavior, is easily diagnosed with a high degree of specificity in PID. Unfortunate, most females with PID demonstrate atypical symptoms which sometimes mimic appendicitis discovering the disease during the appendectomy. Abnormal vaginal discharge full of neutrophils is an indicator of PID, along with a positive vaginal culture for Chlamydia or Gonorrhea. Management of PID entails the use of broad-spectrum antibiotics, which represent the cornerstone of therapy and must adequately cover the polymicrobial spectrum of pathogens implicated in this infection, which includes Neisseria gonorrhoeae, Chlamydia trachomatis, and specific cervicovaginal anaerobic and aerobic bacteria. Sequelae associated with PID includes infertility, ectopic pregnancy, and chronic pelvic pain syndromes. The sexual partner of the affected patient should also be treated.

M. Acquired Undescended Testis

Undescended testes (UT) are recently categorized into congenital and acquired forms. In the congenital form the child is born with an undescended testis, while the acquired form is characterized for later development in life of cryptorchidism. The acquired form of undescended testes comprises the high rate of orchidopexies performed later in life in children. Is a condition in which a previously fully descended testis can no longer be manipulated into a stable scrotal position. Acquired UT are mostly situated in a superficial inguinal pouch, of normal size, with a normal attachment of the gubernaculum, and in half of the cases associated with an open processus vaginalis They are not associated with epididymal deformities or abnormal attachment of the gubernaculum. Acquired UT can be secondary to failure of natural elongation of cord structures in proportion to body growth due to complete disappearance of the processus vaginalis. In contrast the congenital variety of UT is proximal to the external spermatic ring (intracanalicular), a complete hernial sac is present, and is associated with epididymal deformities and abnormal attachment of the gubernaculum. Acquired UT has also been reported in children with cerebral palsy due to spasticity of the cremasteric muscle. Both forms of UT should be managed by orchiopexy to avoid the adverse effects on germ cell development and fertility potential. For congenital UT orchiopexy is recommended at 6-12 months of age. Postoperative results are better for the acquired form compared with the congenital variety.

IX. SKIN & MISCELLANEOUS DISORDERS

A. Pilonidal Sinus Disease

Pilonidal sinus disease (PSD) is the result of a chronic inflammatory reaction of the subcutaneous tissue of the low back from midline skin orifices that ingest hair shafts in hirsute buttocks. PSD arises from hair follicles in the skin, appears after puberty and affects mostly males' adolescents. Male sex, adolescence and a familial disposition are associated with the development of PSD. Minor local trauma ('Jeep Disease' in military personnel) and overweight are the most important factors for development of symptomatic PSD. Primary closure after excision of PSD has frequently been complicated by wound breakdowns. Sepsis and hematoma formation are the main causes of wound breakdowns. PSD is best treated in the acute situation with follicle removal and lateral drainage. The method is suited to outpatient management and gives minimal disability and good long-term control. In the chronic situation conservative therapy (meticulous hair control by natal cleft shaving, perineal hygiene, and limited lateral incision/drainage for abscess) effectively controls PSD in the nonoperative outpatient setting while promoting near-normal work status preferred over excisional operations.

B. Ingrown Toe Nails

Onychocryptosis is the medical term use to describe ingrown toe nails (ITN). Congenital hyperplasia of the nail bed leads to this condition in infants and children. Adolescent and athletes are susceptible to develop ITN. Other conditions associated with ingrown nails are hallux valgus, claw toes, gout, diabetes, arthritis, or fungus infections of the nails. Initial management should be conservative inserting cotton wool pledgets moistened with an antiseptic under the ingrowing nail edge along with oral antibiotics if there are

signs of infection. Insertion of the pledget under anesthesia improves the results and chronicity does not adversely affect the outcome though the procedure is time consuming. Other alternatives are segmental total nail avulsion, nail edge excision, nail matrix phenol cauterization or wedge resections. Total nail avulsion and nail edge excision carries the highest recurrence rate. Nail matrix phenolization is an effective short procedure that can be done under the presence of an infection with a low recurrence rate. Radical wedge resection is simple, has a low recurrence rate, leaves patients with an intact, pain-free, cosmetically acceptable nail-bearing toe, and permits the wearing of normal shoes within a short period. In children conservative treatment is more promising than in adults. If unsuccessful the next reliable surgical approach is wedge resection. Attentions to hygiene and always cutting nails transversely are important preventing factors.

C. Ganglion Cysts

Ganglion cysts are synovial cysts that appear in the wrist or foot after minor trauma or stress. Most occur in the dorsal wrist area. Presence of a colorless to pale-yellow gelatinous material in the aspirate is pathognomonic of ganglion cysts. FNA smears are monotonous showing abundant mucoid material, single cells resembling histiocytes, a few tight clusters of cells, some collagen fibers, and some red blood cells with altered shapes. Serial microscopic studies have shown evidence of a one way valve like system between the affected joint and the ganglion. Diagnosis is physical, though ultrasound findings (cysts with a mean diameter of 1.4 cm and projection into the joint or tendon) are of help. Management consists of excision. Recurrence rates are high ranging between 10 and 35%. Ganglion cysts in the volar aspect of the wrist have a higher incidence of postoperative complications (nerve and radial artery damage). Intralesional injection of hyaluronidase has been found a safe, fast, well accepted and cost-effective alternative to surgical excision.

D. Anal Warts

Warts in the perianal region of prepubertal infant or child most commonly are the result of human papilloma virus (HPV) infestation. Main clinical manifestations of anal warts are cauliflower-like Condylomata Acuminata that usually involves moist surfaces, keratotic and smooth papular warts usually on dry surfaces, and subclinical flat warts that can be found on any mucosal or cutaneous surface. Mode of transmission can occur from an infected maternal birth canal (perinatally), by autoinoculation or heteroinoculation from common hand warts, through sexual abuse and possibly indirect transmission via fomites. Often, the mode of transmission is unknown. HPV-DNA typing is a useful technique that helps identify the genital types involved (6 or 11, 16 or 18) alerting the physician to proceed with a careful assessment for sexual abuse. Predisposing factors for anal warts include social problems, lack of hygiene, promiscuity, diabetes and ammoniacal erythema. The appearance of the papilloma is diagnostic. Management includes cytotoxic agents (podophyllin, podophyllotoxin and fluorouracil), destructive procedures (scissor excision, cryotherapy, electrocautery, and laser photocoagulation) and recently topical interferon hydrogel. In case of transmission by sexual abuse child protection is warranted.

E. Gynecomastia

Gynecomastia refers to abnormal breast enlargement in males. In children, gynecomastia can be classified as simple pubertal, pathological, general obesity and pectoral muscle hypertrophy. Most cases of gynecomastia are simple pubertal associated to a transient or permanent disturbance in steroid hormone physiology occurring when the male breast is exposed to a decreased ratio of androgen to estrogen. Pubertal gynecomastia can be managed non-operatively since breast enlargement start one year after the onset of puberty and subside two years later. Pathological gynecomastia is associated with drug use (steroids, digitalis, spironolactone, and marijuana), chronic liver disease or malignancy (Leydig cell tumor of the testis). General obesity is associated with fat deposition surrounding breast tissue that lends itself to weight reduction. Diagnosis is by history and physical exam. Routine endocrine work-up is not cost effective. Persistent pain, uncertain diagnosis and cosmetic reasons (embarrassment or distress) are the major reasons for operation. Subcutaneous mastectomy through a periareolar incision gives the best cosmetic results. Common late postoperative sequelae are inverted areolae, hypopigmentation and hypoesthesia of the areolar region.

F. Bites & Stings

It is estimated that more than 1.5 million bites occur yearly in the United States. Most of them within

the home setting in children younger than five years of age. When managing bites in children three general concepts must be observed: 1) Determine if the attacking animal has injected venom into its victim. For this to occur the history must provide the exact identification of the animal and type of venom. Once identified an specific antivenom can be utilized. 2) All bites wounds are considered contaminated wounds. This includes rabies and tetanus. Prophylaxis in either case should be provided. Furthermore the bite can produce significant tissue loss and destruction. Initial management should include thorough mechanical cleansing and debridement along with systemic antibiotics. 3) An assessment should be made of the long-term outlook for both disfigurement and loss of function of the wound. Wounds of the face do fairly better with primary closure no matter the inciting initial agent. Significant loss of tissue might need conservative management followed by reconstruction when adequate granulation tissue appears. Ischemic extremity from compartment syndrome might need fasciotomy. Fortunately we don't see snake bites in Puerto Rico. Few cases of spider bites has been reported. Dog bites (pit bull) are the most common bite seen in the emergency room affecting the extremity or face of young infants. Cat bites or scratch can produce a febrile condition associated with lymphadenopathy. Human bites carry the risk of HIV and hepatitis infestation.

The insects that inflict more venous stings than any other in children are the bees and ants. Stings from bees and wasps produce a local tissue reaction with a wheal and flair. Symptoms develop within twenty minutes of the sting and include urticaria, syncope and respiratory distress. Most serious sequelae is anaphylaxis which occur when the child has been previously inoculated. More than 500 stings are needed to cause death in a child. Management is local and systemic. The venom can be removed if the event has less than 20 minutes. Cold compresses will reduce pain associated with the sting and baking soda helps with the itching. Systemic support includes airway control, alpha agonists medication, inhaled beta agonist for bronchospasm and calcium for muscle spasms. Best prophylaxis is reducing exposure. Fire ants' sting can produce edema, pruritus, erythema, pain and burning with a characteristic wheal. Wound is cleaned with soap and water. Rarely systemic management is needed.

G. Perianal Abscess

Perianal abscess is a not so rare condition seen almost exclusively in infants less than two years of age. Most cases are seen in males' infants. The infant presents with a history of increasing irritability, fever, erythema and induration of the perianal skin. In a period of 48 to 72 hours the area becomes fluctuant. Oral antibiotics are ineffective in controlling the infectious process. It is theorized that a perianal abscess arises from a developmental anomaly in the deep crypts of Morgagni which trap bacterias initiating a cryptitis that proceed to a perianal abscess. This abscess may open or not to become later a fistula in ano. Gut derived organisms are isolated from most cases of perianal abscess. Most abscess are located laterally equally divided between right and left. Perianal abscesses in children are best treated by incision, drainage and systemic antibiotics. A proportion of patients with perianal abscess later develop a fistula in ano. This fact has led some researchers to propose that primary treatment of perianal abscess in childhood involve a careful search for a coexisting fistula and treatment of this by fistulotomy. Long term recurrence is very rare. Recurrence should prompt a search for associated disorders such as Crohn's, immunodeficiency and autoimmune neutropenia.

H. Keloids

Excessive scarring is deposition of collagen out of the range of normal after surgery. Two types of excessive scarring seen in children and adults are hypertrophic scars and keloids. Hypertrophic scars are slight elevation of the scar restricted to the boundaries of the original wound, regress with time and rarely cause functional impairment. Hypertrophic scars rarely need to be removed. Keloids extend beyond the borders of the original wound, cause functional impairment, contain mast cells that release histamine and produce pruritus. Familial cases have been seen. Keloids do not regress over time and recur after surgical excision. Keloids contain large thick collagen fibers composed of numerous fibrils closely packed together. In contrast hypertrophic scars exhibit modular structures in which fibroblastic cells, small vessels, and fine, randomly organized collagen fibers are present. Earlobe piercing is the main etiological factor of earlobe keloids. Keloids removal is indicated in cases of functional impairment of a joint or for cosmetic reasons. Excision with intraoperative local injection of triamcinolone or alpha-2b interferon has produced some

therapeutic advantage in severe cases. Early single postoperative fraction radiotherapy claims effectiveness in reducing keloid recurrences.

I. Felons

A felon is defined as a painful fusiform abscess (swelling) in the distal fat pad of the phalanx with increased tenderness over the flexor aspect and erythema (redness and cellulitis) distributed uniformly along the entire flexor aspect of the pulp space. Felon is very different from a paronychia. Paronychia is a more localized painful swelling on the extensor aspect of the distal digit located at the proximal extent of the sulcus. Initial management consists of antibiotics, warm soaks and elevation of the affected extremity. If the child does not improve rapidly with medical treatment, surgical incision and drainage are indicated to avoid involvement of the bony distal phalanx due to rapid extension of the infections process through synovial-lined spaces. Felons are managed with digital anesthetic block and incision along the ulnar aspect of the digit to produce drainage of the closed space infection. The incision is made along the ulnar aspect of the phalanx to avoid the possibility of a painful scar on the radial side of the phalanx which is the pinching side of the distal phalanx. Cultures are routinely taken during surgery. Staphylococcus aureus is a common offending organism. Penrose drain for 48 hours is recommended.

J. Latex Allergy

Allergy to natural rubber latex is an increasing common condition in both children and health care workers. Almost 20 to 40% of children with spina bifida are allergic to latex. Other affected persons are health care workers, latex industry workers, immune compromised individuals, children with bladder exstrophy, anorectal anomalies, and persons with positive risk factors such as multiple surgical procedures. Chemical additives in latex gloves can cause an irritant or allergic contact dermatitis. Latex proteins are responsible for most of the immediate IgE-mediated hypersensitivity allergic reactions. Symptoms range from rhinitis, conjunctivitis and urticaria to intraoperative anaphylaxis and death. Skin prick testing with natural rubber latex and glove tests are safe diagnostic procedure. In children with spina bifida significant and independent risk factors identified for latex sensitization are multiple interventions and higher levels of total serum IgE. The only currently available treatment is complete avoidance of latex. For children with known history of latex allergy premedication with antihistamines and steroids is in order.

K. Nevus Sebaceous

Nevus sebaceous of Jadassohn is a congenital hamartomatous skin lesion occurring mostly on the scalp, face and neck. At physical exam the lesion is well circumscribe with a yellow-orange smooth plaque appearance. Borders become irregular with puberty and hormonal changes. Malignant transformation has been reported in 10% of cases occurring only after puberty. The nevus sebaceous can transform into a basal or squamous cell carcinoma. The lesion will not go away spontaneously. It is uncommon for malignancy to develop in a sebaceous nevus before puberty. Due to the risk of malignant transformation and the difficulty in follow-up of these children with time, early complete excision for prophylaxis is recommended in cases of nevus sebaceous. Excision must encompassed clean deep and lateral margins of resection to be effective. Large lesion will benefit from use of tissue expanders.

L. Polydactylism

Polydactylism (accessory finger or toe) is one of the most common congenital anomaly of hands or feet in children. The prevalence of polydactylism is almost two cases for every each 1000 born alive newborns. Familial occurrence is associated in almost 20% of cases with variable gene penetrance. Polydactylism of the hand is more common than in the foot. In the hand the ulnar accessory finger predominates. A few cases undergo traumatic intrauterine amputation, a condition known as rudimentary polydactylism. Different genes are involved in the pathomorphogenesis of postaxial polydactylism. Polydactyly may be preaxial (medial, thumb side) or tibial (hallux-side), postaxial (lateral) or ulnar (side of the little finger or toe), and central (middle fingers or toes). The duplication may appear at the distal and medial phalanges or at the whole digit. Surgical amputation of the affected finger or toe is indicated for cosmetic reasons or for functional disturbances in wearing shoes respectively. Careful clinical and radiographic evaluation should be made prior to treatment to achieve good functional and cosmetic results. Polydactylism can be diagnosed prenatally and when isolated is associated with good perinatal outcome. Primary suture ligation of

accessory digits in infancy can be associated with later development of neuroma in the stump. Identification and high transection of the accessory digital nerve is essential in the treatment of pedunculated supernumerary digits.

M. Giant Pigmented Nevus

Giant Pigmented Nevus (GPN) refers to a congenital melanocytic nevi characterized by a diameter of 20 cm or greater in adulthood, or a lesion occupying greater than 2% of the body surface area of an infant or child. Generally, melanocytic nevi is solitary, but smaller satellite lesions may be present within a GPN. The mode of inheritance of GPN is probably multifactorial with a 2:1 female predominance. The lesion is typically brown pigmented, varies in size and shape, contains hair and can be found anywhere on the face, neck, trunk or extremity of the child. GNP extensively involving the extremities can result in reduced growth of the affected limb. GPN has a higher incidence of malignant transformation (melanoma formation) due to the increase number of nevus cells at risk for such transformation. Lifetime incidence of malignant transformation is four to 6%. This capacity of GPN to develop into malignant melanoma is the main reason for opting for prophylactic surgical excision as treatment of choice. Tissues expansion is the primary modality for excision and reconstruction in the face and scalp since it can be started early in life and repeated as required. For the trunk excision with abdominoplasty or skin graft is preferred. In the extremity excision and skin grafting is best option. Dermabrasion or laser therapy cannot ensure complete removal of nevus cells.

N. Chronic Granulomatous Disease of Childhood

Chronic Granulomatous Disease (CGD) is a very rare inherited primary immunodeficiency disease of childhood. Due to defective respiratory burst, phagocytic cells of children with CGD are not able to kill certain bacterias and fungi despite normal chemotaxis and phagocytosis. The proper functioning of the NADPH oxidase of the phagocytic cell is impaired. Children with CGD are rendered susceptible to infection by a group of catalase positive microorganisms due to the inability of phagocytic cells to reduce molecular oxygen and create reactive oxygen metabolites necessary for normal intracellular killing. Catalase positive organisms affecting children with CGD include staph aureus, Escherichia Coli, Serratia, Salmonella, Candida and Aspergillus. Children with CGD present clinically with recurrent abscess involving skin, soft tissue (most common site), lymph nodes, lung, bone and liver that responds poorly to antibiotics needing surgical drainage. Palisading granulomas with central necrosis can be seen in histologic samples. Though four different types of CGD have been described, most cases are X-linked defects. Diagnosis of CGD is made by showing the inability of neutrophils from the patient to undergo respiratory burst after phagocytosis (NBT Test). Fungal infections account for most deaths. Children with CGD should receive long-term Trimethoprim-Sulfamethoxazole prophylaxis. Gamma interferon reduces the number of infection requiring hospitalizations.

O. Congenital Melanocytic Nevus

Skin nevus found in infants and children can be a source of concern to both parents and physician dealing with pediatric patients. Congenital melanocytic nevus (CMN) is a characteristic pigmented nevus with mild raised borders and a strong skin discoloration caused by melanin deposits. Incidence of CMN is 0.2% with more than 90% small nevi (< 1.5 cm). Congenital melanocytic nevus may cause cosmetic defects and represent a risk of malignant transformation, namely melanoma. The incidence of developing melanoma is associated with location and size of the CMN; those identified in the face and neck or covering large areas of the body (larger than 5 cm) has a higher incidence of malignant transformation. Not always there is a correlation between pre and postoperative diagnosis after prophylactic excision of a nevi. Digital videomicroscopy using polarize light can help increase the diagnostic yield of CMN. A high index of suspicion for cutaneous melanoma is needed by clinicians assessing melanocytic lesions in children and adolescents for early diagnosis taking into consideration change in color, growth rate, and bleeding, For large lesions and those in exposed areas of the body, surgical excision is the treatment of choice. Medium and small lesions can be managed with Ruby laser treatment. Ruby laser treatment does not result in scarring, mutilation, or functional impairment.

P. Hyperhydrosis

Primary idiopathic hyperhydrosis is the occurrence of perspiration in excess of that required for body cooling. Secondary hyperhydrosis can be the result of conditions such as hyperthyroidism, severe obesity,

pheochromocytoma or anxiety. Primary hyperhydrosis can involve the hands, axillae, or trunk. Sweat is secreted by eccrine glands innervated by sympathetic cholinergic fibers. Eccrine glands consist of tubules which extend deeply in the dermis becoming coiled and surrounded by myoepithelial cells. The majority of children seek help due to palmar hyperhidrosis. The condition can be socially debilitating causing psychological and emotional upset. Conservative management consists of antiperspirants, iontophoresis and systemic medication (anticholinergics). If the condition does not improve with conservative measures, surgical sympathectomy is indicated. The aim of surgery is to disrupt the sympathetic supply to palms by destroying relevant ganglions in the upper thoracic sympathetic chain (T2 and T3). The sympathectomy can be done supraclavicular, limited posterior thoracotomy, transaxillary thoracotomy or using thoracoscopy. Results are excellent. The most serious and uncommon complication is Horner's syndrome.

Q. Preauricular Tags

Preauricular skin or chondrocutaneous branchial remnant is not a rare finding in infants. Preauricular tags occur in five of every 1000 live births. They are congenital malformations that should be taken care for aesthetic reasons. Most preauricular tags are from branchial origin and contain cartilage. Excision is indicated after the age of six months under general endotracheal anesthesia as an ambulatory procedure. Cosmetic results are very good. Less than 5% of all preauricular tags are associated with other malformations of the ear/face region, most of them conductive type hearing impairment. Physical examination under such circumstances provides the first clue of a hearing disorder. Possibility increases if there is familial history of hearing impairment. When in doubt, audiometry is indicated. Another association with preauricular tag is the presence of a urinary tract anomaly which occurs in 8% of all cases. Such renal anomaly includes hydronephrosis caused by ureteropelvic obstruction or vesicoureteric reflux, and horse-shoe kidney. Some workers recommend that urinary tract ultrasonography be conducted in the routine assessment of infants with preauricular tags and multiple congenital anomalies. Specific disorders associated with tags that increase the yield of finding a renal disorder includes CHARGE association, Townes-Brocks syndrome, branchio-oto-renal syndrome, Nager syndrome, and diabetic embryopathy. Routine renal ultrasonography is not necessary in the evaluation of children with isolated preauricular tags.

R. Stings

The insects that inflict more venous stings than any other in children are the bees and ants. Stings from bees and wasps produce a local tissue reaction with a wheal and flair. Symptoms develop within twenty minutes of the sting and include urticaria, syncope and respiratory distress. Most serious sequelae is anaphylaxis which occur when the child has been previously inoculated. More than 500 stings are needed to cause death in a child. Management is local and systemic. The venom can be removed if the event has less than 20 minutes. Cold compresses will reduce pain associated with the sting and baking soda helps with the itching. Systemic support includes airway control, alpha agonists medication, inhaled beta agonist for bronchospasm and calcium for muscle spasms. Best prophylaxis is reducing exposure. Fire ants≪ sting can produce edema, pruritus, erythema, pain and burning with a characteristic wheal. Wound is cleaned with soap and water. Rarely systemic management is needed.

S. Molluscum Contagiosum

Molluscum contagiosum is a pox viral infection affecting primarily the skin of infants, children and adults. It causes firm discrete pearly papules that measure between one and four millimeters in diameter. The papules have a characteristic central umbilication with a caseous type of material containing virus-laden cells. One-third of children have symptoms from, or secondary reactions to the infection, including pruritus, erythema and, occasionally, inflammation and pain. Molluscum contagiosum can occur singly or in clusters anywhere on the body, though the trunk is more commonly affected. Spread is usual by direct contact, with genital involvement suggesting the possibility of sexual abuse in the young child. The virus produces a number of substances that block immune response formation in the infected host. Molluscum contagiosum is a benign and self limited disease with most cases resolving within six months to one year irrespective of therapy, though patients with weakened immune systems have increased difficulty in clearing lesions. A single, most effective treatment for either infection has not been defined. Conventional methods attempt to nonspecifically destroy infected tissue. Immunocompetent children can be managed with imiquimod, retinoids,

and alpha-hydroxy acids. Surgical management, if undertaken, includes curettage of the central plug, cryosurgery and/or electrodesiccation.

T. Ehlers-Danlos Syndrome

Ehlers-Danlos syndrome (EDS), also known as cutis hyperelastica, is a group of inherited connective tissue disorders caused by a defect in the synthesis of collagen. Depending on the individual mutation the severity of the syndrome can vary from mild to severe. Several types of EDS have been categorized with type 4 considered the most dreadful considering the high propensity of these individuals to develop life-threatening arterial and digestive complications. EDS type 4 is an autosomic dominant defect (missense mutation in the COL3A1 gene) characterize by a fascial acrogeria appearance (large eyes, small chin, thin nose, lobeless ears), small stature with slim build, and thin pale translucent skin. Children with EDS have poor wound healing, hypermobile joints, clotting anomalies, spontaneous pneumothorax and recurrent hernias. Among the catastrophic events associated with type 4 EDS we can find arterial dissection or tear caused by deterioration of congenital fragile tissue leading to hematoma, false aneurysm and intracavitary bleeding. They are responsible for the majority of deaths. The next set of complications are spontaneous and recurrent perforation of the colon associated with a significant risk of leakage after anastomosis and spontaneous perforation or bleeding of the uterus. There is no cure for EDS and management is supportive.

U. Metal Allergy

Jewelry, dental and surgical implants from craniofacial, orthopedic, neurosurgical and pediatric surgery physicians can lead to metal allergy in children. As many as 13% of patients are sensitive to nickel, cobalt or chromium. Metal allergy from nickel is the most common contact allergy in the United States and Europe. The classical symptom of dermatitis caused by nickel is a rash in the earlobes, periumbilical region or wrist resulting from contact with costume jewelry, buttons and zipper. Metal allergy is a typical delayed type IV hypersensitivity reaction caused by T-lymphocytes reaction. CD8 and CD4 cells cause cytotoxic and inflammatory response to the metal. Children with metal allergy usually elicit a past history of atopy including allergic rhinitis, asthma, eczema and urticarial rash. Metal allergies are frequently misdiagnosed as surgical infections. Symptoms of inflammation such as pain, warmth, erythema and swelling can be seen over the implant, including pericarditis and pleural effusion in those in a thoracic position. As a screening measure to determine if a child can or might develop metal allergy to an implant the following should be evaluated: 1- history of allergy to jewelry, orthodontic braces, metal buttons on clothing and food. 2- History of previous atopy and eczema. If any of the above indications are found, a dermal patch test should be performed. This patch test contains 23 allergens and allergen mixes that cause up to 80% of allergic contact dermatitis cases. Should the child be found to have metal allergy implants of titanium should be considered, since titanium does not produce allergic reactions but are more expensive.

X. LAPAROSCOPY/THORACOSCOPY IN PEDIATRIC SURGERY

A. Thoracoscopic Lung Biopsy

The first minimally invasive procedure reported using thoracoscopic techniques was a lung biopsy. Lung biopsy is generally utilized to provide answers for causes of interstitial lung disease in both the immunocompetent and immunocompromised child, lung residual masses after chemotherapy and evaluation of new suspicious nodules found in oncology patients. Interstitial lung disease is defined as diffuse interstitial findings on chest x-ray or CT scan. Interstitial disease may be bilateral or unilateral and may be more prominent in one lobe over another. Posterior pleural biopsies are performed with the patient almost prone and anterior lesions are performed with the patient almost supine. Thus, positioning takes advantage of gravity to allow the lung to fall away from the lesion when the lung is collapsed. Instead of single lung ventilation children can benefit from undergoing contralateral lung ventilation using ipsilateral bronchial blockers or Fogarty balloon catheters. Use of CO2 insufflation creates a pneumothorax and further collapses the ipsilateral lung. Pressures of 4-6 mm should be utilized. The lung biopsy can be performed with endoloop or endoscopic staplers. Image-guided percutaneous needle biopsy is preferred for pulmonary nodules. Thoracoscopy reduces pain, shortens hospital stay and is more pleasing cosmetically.

B. Thoracoscopy CDH Repair

Congenital diaphragmatic hernias (CDH) can be surgically managed using either laparoscopic or thoracoscopic minimally invasive technique. Either Bochdalek and Morgagni hernias have lent themselves to repair using these minimally invasive approach. The ideal child for thoracoscopic repair would be those that have delayed presentation beyond the neonatal period since the presence of pulmonary hypoplasia and pulmonary hypertension is minimal or none. The child should be in stable cardiovascular and respiratory status before surgery. The procedure is performed under general anesthesia without single lung ventilation with an epidural thoracic catheter. Reduction of the hernia occurs using one optical trocar, two operating trocars and a pleural insufflation pressure of carbon dioxide between five and 8 mmHg. The hernia defect is repaired using non-absorbable interrupted sutures or mesh with absence of significant diaphragm. Conversion to open occur with difficulty in reducing the hernia toward the abdominal cavity, herniation of liver and intolerance of insufflation. Reduction of the defect is easier in babies that have a hernial sac. Without a sac the bowel is gently pushed down into the abdominal cavity. Using the laparoscopic approach the spleen and bowel are difficult to reduce into the peritoneal cavity and the working space is very restricted. The thoracoscopic technique causes minimal trauma, results in good respiratory function, excellent cosmetic results and promotes early recovery. End tidal CO2 is significantly elevated during repair.

C. Laparoscopic Rectopexy

Full-thickness rectal prolapse is a relatively common and distressing condition in children that fortunately is usually self-limiting in the life of the affected child and family. Peak incidence is between one and three years of age. Medical management improves more than 80% of all cases of rectal prolapse. Patients are managed for the underlying cause of the prolapse such as chronic constipation or acute diarrhea, while cystic fibrosis is rule-out. Failed medical therapy and recurrent prolapse will be managed with surgery. Before surgical intervention the rectum should be evaluated with imaging studies (Barium Enema) and flexible endoscopy. Therapeutic surgical procedures include submucosal injection of sclerosant, Thiersch cerclage, open abdominal rectopexy with or without sigmoid resection and perineal rectopexy. Rectopexy alone for megarectum or megasigmoid is not appropriate and will eventually need resection of the dilated bowel. With the use of minimally invasive techniques, laparoscopic suture rectopexy namely, full posterior mobilization and fixation to the sacrum, has emerged as an alternative surgical management for recurrent rectal prolapse in children. No preoperative bowel preparation is required while the procedure can be done ambulatory with a low morbidity, low recurrence rate and excellent cosmetic results. Laparoscopic rectopexy has also been found to improve constipation and fecal incontinence.

D. Single Site Laparoscopic Surgery

As technological surgical advances continue to occur in the field of laparoscopic surgery in search of the scarless operation, single site umbilical laparoscopic procedures have emerged. A single multi-port with several holes is introduced through the navel, instead of several small incisions. The working space in children is small and space is needed for instrument triangulation and retraction. Nevertheless, single site umbilical laparoscopic surgery is performed in children for such procedures as cholecystectomy, appendectomy, intussusception reduction, splenectomy, nephrectomy, inguinal hernia repair, fundoplication, gastrostomy, orchiopexy and pyloromyotomy. Due to lack of triangulation roticulating and curved instruments have been developed for single port procedures. Most roticulating instruments are disposable and need a learning curve for safe use. Alternatives to roticulating instruments are rigid bent instruments which can be reused and are low cost. The shaft of the telescope comes longer to avoid clashing with the surgeons hand. Retraction can be achieved with the used of magnets. This is achieved with magnetic intracorporeal graspers and an extracorporeal magnet that is manipulated over the abdominal wall to adjust and control the instruments. Laparoscopic surgery in children is moving toward achieving this goal of scarless surgery.

E. Laparoscopy for Ventriculoperitoneal Shunts

Ventriculoperitoneal (VP) shunt is the standard treatment for hydrocephalus in children and adults. Malfunctioning VP shunts causing increase intracranial pressure needs surgical revision. Such malfunctioning can occur due to obstruction from peritoneal adhesion, multiple infections, ascites and pseudocysts formation, or mechanical causes such as catheter fracture, disconnection, migration and misplacement. VP shunt failure is common with 25-40% occurring within the first year, and 50% by the second year. Laparoscopy has an

important role in initial VP shunt placement and later revisions. Laparoscopy reduce the trauma to the abdominal wall decreasing adhesion formation and optimizing visualization during placement. With VP shunt failure laparoscopy can be both diagnostic and therapeutic. While visualizing the entire abdominal cavity the causes of failure can be identified, lysis of adhesions can be performed, retrieval of disconnected shunt can be accomplished, flow of CSF fluid can be observed and proper placement of the catheter can be obtained The technique is safe with a very low morbidity rate. The use of laparoscopy assisted VP shunt revision is advocated for patients with multiple previous shunt revisions, prior abdominal surgery, previous intraperitoneal infections, broken devices, obesity or CSF pseudocysts.

F. Magnet-assisted Laparoscopic Surgery

Magnets have been introduced in the armamentarium of laparoscopic surgery to recapture the triangulation that is affordable by conventional laparoscopy while decreasing the number and size of the abdominal incisions utilized. Specialized magnetic grasper are inserted into the peritoneal cavity through the port cannula and attached to intraabdominal organs. These magnets are controlled by another external magnet placed on top of the abdominal wall. The magnet grasper moves to provide further traction on an organ without the additional need of another port. With the magnet you can retract the liver, stomach, lung tissue, gallbladder providing traction and facilitating exposure. Magnet-assisted laparoscopy is safe and effective means of reducing the number and size of abdominal incisions while improving exposure, triangulation, and the ergonomics of the procedure.

G. Trocar Injury

Injury by a trocar during a laparoscopic or thoracoscopic procedure is a very serious complication in surgery. The two most serious complications most likely to result in death caused during entrance of a trocar are hemorrhage due to vessel injury and infection due to bowel injury. The rate of trocar related complication is less than 3%. The average incidence of trocar-related vascular injury is 0.1%. Major vessel injury is almost invariably operator error. Most vascular, bowel and local hemorrhage injury are caused during the initial trocar insertion. There is some blind force exertion that causes the blunt/sharp injury to the major vessel. The vessels most frequently involved are the aorta, the iliac arteries, the mesenteric vessels, and the vena cava. Force require to insert reusable trocars is twice that for disposable trocars. Shielded trocars might provide a margin of safety. Trocar use requires considerable training, practice, skill, manual dexterity, adequate muscular strength, knowledge of the associated risks, and careful patient selection. In addition to laparoscopist-related issues (trocar insertion technique, patient selection, injury recognition and effective intervention), the lack of standard device designs, a lack of proven-effective fail-safe features, and failure of patients to report symptoms in a timely manner may also contribute to morbidity and mortality. Open (Hasson) entrance or optical access trocars are recommended for patients with prior abdominal surgery, small children, patients with lower abdomen skin cannot be adequately stabilized for safe insertion or Veress needle.

H. Laparoscopic Excision Choledochal Cyst

The management of choledochal cyst entails complete excision of the cyst and reconstruction using roux-en-y hepaticojejunostomy in most cases or hepaticoduodenostomy. During the last ten years there has been a tendency for the procedure to be performed laparoscopically. Laparoscopic excision of choledochal cysts with reconstruction is feasible and can be performed safely with a low intraoperative complication rate. The laparoscopic technique includes excision of the gallbladder and cyst, followed by a Roux-en-Y anastomosis constructed after exteriorization of the small bowel via an infraumbilical trocar incision or intracorporeally. After repositioning of the bowel, an end-to-side hepaticojejunostomy is carried out laparoscopically. Dissection close to the cyst wall is mandatory to prevent injury to the hepatic vessels, especially the portal vein. Hepaticoduodenostomy is s simple procedure than hepaticojejunostomy, with shorter operating time reducing the risk of postop adhesions, can be performed totally laparoscopically giving superior cosmetic results, but carries a higher incidence of postop cholangitis and bile-reflux gastritis. Laparoscopy presents a longer operation duration than laparotomy and requires more instruments. Less blood loss and chances of transfusion is seen in the laparoscopic group due to the improved accuracy provided by the magnified view. Cases without severe infection, without common hepatic duct or left/right duct strictures, and those without a cyst deeply embedded in the pancreas can be left undrained. In neonates

the laparoscopic procedure curtails further complications of the cyst and reverses the derangement of liver function associated with the choledochal cyst.

I. Laparoscopic Peritoneal Cannula Placement

Peritoneal dialysis is preferred to hemodialysis to manage chronic renal failure in children because it has a lower incidence of serious complications, is more cost-effective and improves patient nutrition and independence. The peritoneal cannula for such purposes can be place using an open conventional surgical technique, percutaneously or laparoscopically. Laparoscopic placement of peritoneal dialysis catheters or revision allows complete visualization of the peritoneal cavity placing the catheter under direct vision, preferably in the pelvis. Suturing the catheter tip into the pelvis is associated with a low rate of catheter migration. In addition a more complete omentectomy can be performed and lysis of adhesions can be accomplished to increase the peritoneal absorptive surface. Omentectomy is beneficial during placement of peritoneal dialysis catheters to prevent blockage of the catheter. Laparoscopy can allow for the rescue of block catheters in cases of revisions due to malfunction. Others procedures that can be done concomitantly using the laparoscopic technique include repair of inguinal hernias, gastrostomy tube placement, kidney biopsy and cholecystectomy. Peritoneal dialysis is delayed as long as possible to allow for healing of the incision to prevent leakage. Other recognize benefits of the laparoscopic technique include better cosmetic results, less postoperative pain and shorter hospital stay.

XI. 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, hydroureteronephrosis 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).

XII. SUGGESTED READING

Specific Readings

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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

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XIV. 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**

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"A hundred years from now,
It will no matter my bank account
Or the house I used to lived,
Live will be different
since I was important
In the life of a child".