OnLine *Pediatric Surgery* HANDBOOK

for Residents and Medical Students...

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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.

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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 100cc/kg/24 hrs for the first 10 kg of weight. As a rule of thumb, the daily fluid requirements can be approximated too:

prematures 120-150cc/kg/24 hrs

neonates (term) 100cc/kg/24 hrs

Infants >10kg 1000cc+ 50cc/kg/24 hrs.

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

(1) urinary water losses,

(2) gastrointestinal losses,

(3) insensible water losses, and

(4) surgical losses (drains).

Blood Volumes estimates of help during surgical blood loss are:

premature 85-100 cc/kg,

term 85 cc/kg,

and infant 70-80 cc/kg.

The degree of dehydration can be measured by clinical parameters such as: body weight, tissue turgor, state of peripheral circulation, depression of fontanelle, dryness of the mouth and urine output. Intravenous nutrition is one of the major advances in neonatal surgery and will be required when it is obvious that the period of starvation will go beyond five days. Oral feeding is the best method and breast is best source. Newborn infants requires 100-200 calories/kg/day for normal growth. This is increased during stress, cold, infection, surgery and trauma. Minimum daily requirement are 2-3 gm/kg of protein, 10-15 gm/kg of carbohydrate and small amount of essential fatty acids.

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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.

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2. Metabolic and Host Defenses

Handling of the breakdown products of hemoglobin is also a difficult task for the premature infant. The ability of the immature liver to conjugate bilirubin is reduced, the life span of the red blood cell is short, and the bilirubin load presented to the circulation via the enterohepatic route is increased. "Physiologic" jaundice is, therefore, higher in the preterm infant and persists for a longer period. Unfortunately, the immature brain has an increased susceptibility to the neurotoxic effects of high levels of unconjugated bilirubin, and kernicterus can develop in the preterm baby at a relatively low level of bilirubin. Other problems affecting the baby include the rapid development of hypoglycemia (35 mg%), hypocalcemia and hypothermia. Newborns have a poorly developed gluconeogenesis system, and depends on glycolysis from liver glycogen stores (depleted 2-3 hrs after birth) and enteral nutrition. Immature infants can develop hyperglycemia from reduced insulin response to glucose causing intraventricular hemorrhage and glycosuria. The preterm and surgical neonate is more prone to hypocalcemia due to reduced stores, renal immaturity, and relative hypoparathyroidism (high fetal calcium levels). Symptoms are jitteriness and seizures with increase muscle tone. Calcium maintenance is 50 mg/kg/day. Human beings are homeothermic organisms because of thermoregulation. This equilibrium is maintained by a delicate balance between heat produced and 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 adults levels. C3,C4,C5 complex, factor B, and properdin concentration are also low in comparison to the adult. IgM, since it does not pass the placenta, is absent.

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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 cathecolamines, 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 aminoacids 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.

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II. 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.

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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.

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C. Thyroglossal Duct Cysts

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

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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.

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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.

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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. series - 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 series 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.

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A. Esophageal Atresia

Esophageal atresia (EA) with distal tracheo-esophageal fistula (TEF) is the most common congenital anomaly of the esophagus, followed by EA without TEF also known as pure esophageal atresia and pure TEF. Incidence is one in every 2500 live births. 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 remians to be proven. Polyhydramnios is most commonly seen in pure EA. EA causes excessive salivation, choking, coughing, regurgitation with first feed and inability to pass a feeding tube into the stomach. Contrast studies are rarely needed and of potential disaster (aspiration). Correct dehydration, acid-base disturbances, respiratory distress and decompress proximal esophageal pouch (Reploge tube). Evaluate for associated conditions such as VACTERL association. Correct dehydration, acid-base disturbances, respiratory distress and decompress proximal esophageal pouch (Reploge 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) is undertaken for those babies with adequate arterial blood gases, adequate weight (>1200 gm) and no significant associated anomalies. Delayed repair (gastrostomy first) for all other patients. Repair consists of muscle-sparing thoracotomy, closure of TEF and primary anastomosis. Esophagogram is done 7-10 days after repair. Most important predictors of outcome: birth weight, severity of pulmonary dysfuntion, and presence of major congenital cardiac disease. Complications after surgery: anastomotic leak, stricture, gastroesophageal reflux, tracheomalacia and recurrent TEF. Increase survival is associated with improvements in perioperative care, meticulous surgical technique and aggressive treatment of associated anomalies.
Congenital isolated tracheo-esophageal fistula (TEF) occurs as 4-6% of the disorders of the esophagus bringing problems during early diagnosis and management. More than H-type is N-type, due to the obliquity of the fistula from trachea (carina or main bronchi) to esophageal side (see the figure) anatomically at the level of the neck root (C7-T1). Pressure changes between both structure can cause entrance of air into the esophagus, or esophageal content into the trachea. Thus, the clinical manifestation that we must be aware for early diagnosis are: cyanosis, coughing and choking with feedings, recurrent chest infections, persistent gastrointestinal distension with air, and hypersalivation. Diagnosis is confirmed with a well-done esophagogram, or video-esophagogram (high success rates, establish level of the TEF). Barium in the trachea could be caused by aspiration during the procedure. Upon radiologic doubt bronchoscopy should be the next diagnostic step. Any delay in surgery is generally due to delay in diagnosis rather than delay in presentation. Management consists of surgical closure of the TEF through a right cervical approach. Hint: a small guide-wire threaded through the fistula during bronchoscopy may be of some help. Working in the tracheo-esophageal groove can cause injury to the recurrent laryngeal nerve with vocal cord paralysis. Recurrence after closure is rare.aggressive treatment of associated anomalies.
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. Gastro-Duodenal Anomalies

B.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 radioluscent 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-obtructive 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.

B.2 Pyloric Stenosis

Is an abnormality of the pyloric musculature (hypertrophy) causing gastric outlet obstruction in early infancy. The incidence is 3 per 1000 live births. The etiology is unknown, but pylorospasm to formula protein cause a work hypertrophy of the muscle. Diagnostic characteristics are: non-bilious projectile vomiting classically 3-6 weeks of age, palpable pyloric muscle "olive", contrast studies are not necessary when the pyloric muscle is palpated, enlarged width and length in ultrasonography. The treatment consist in correction of hypochloremic alkalosis and state of dehydration and performing a Fredet-Ramstedt modified pyloromyotomy. Post-operative management consist of: 50% will have one to several episodes of vomiting, usually can feed and go home in 24-36 hours, initial feeds start 8-12 hours after surgery.

B.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.
Congenital partial obstruction of the duodenum can be either intrinsic (membrane, web or pure) or extrinsic (Ladd's bands, annular pancreas). A significant group (25-33%) is born with Down's syndrome. This does not entail a higher risk of early mortality unless associated with cardiac malformations. Other associated conditions are malrotation (midgut volvulus is rare due to absent bowel distension and peristalsis), biliary tract anomalies and Meckel's diverticulum. The diagnosis is suggested in utero by the double-bubble image on ultrasound. Vomiting is the most frequent presenting symptom. UGIS is diagnostic, showing a dilated stomach and first duodenal portion with scanty passage of contrast material distally. Management varies accordingly to the type of stenosis: Ladd's bands are lysed. Pure stenosis is opened longitudinally and closed transversely (Heineke-Mickulicz). Membranous stenosis is resected. Successful endoscopic membranectomy of duodenal stenosis has been reported.
Duodeno-duodenostomy is the procedure of choice for annular pancreas. Diaphragms can rarely be double. Anastomotic malfunction requiring prolonged intravenous nutrition and hospitalization has prompted development of a diamond shape larger stoma. Tapering or plication of the dilated duodenum is another effective method of improving disturbed transit. Other complications after surgery are megaduodenum with blind loop syndrome, biliary reflux, cholestatic jaundice, delayed transit and bowel obstruction. Early mortality is associated to prematurity and associated malformations. Long-term follow-up is warranted to identify late problems.
The diagnostic characteristics are: bilious vomiting, history of polyhydramnios in mother, KUB with classic "Double-bubble" appearance, a microcolon in barium enema study or malrotation.
Treatment consist in: (1) duodeno-duodenostomy bypass for atresias, annular pancreas, and some stenosis. (2) duodenoplasty for webs, and stenosis, and (3) lysis of ladd's bands and Ladd's procedure for malrotation. Associated anomalies are: Down's syndrome (20-30%), VACTERL syndrome, CNS anomalies and cardiac anomalies.

C. Malrotation and Volvulus

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.

D. 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.

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E. 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 mucoproteint content, the result of decreased pancreatic enzyme activity and prolonged small bowel intestinal transit time. Meconium Ileus is classified into two types: (1) Simple meconium ileus: The distal small bowel (10-30 cm of distal ileum) is relatively small, measuring less than 2 cm in diameter and contains concretions of gray, inspissated meconium with the consistency of thick glue or putty. It is often beaklike in appearance, conforming to the shape of the contained pellets. Proximally, the mid-ileum is large, measuring up to 7 cm in diameter. It is greatly distended by a mass of extremely thick, tenacious, dark green or tarry meconium. The unused small colon (microcolon) contains a small amount of inspissated mucus or grayish meconium. (2) Complicated meconium ileus: usually occurs during the prenatal period associated to volvulus, atresias, gangrene, perforation or peritonitis. A cystic mass or atresia of the bowel may occur. The degree of obstruction varies, may be cured in mild cases by rectal irrigations. Failure to pass meconium, abdominal distension and vomiting are seen in more severe cases. The diagnosis is suspected with findings of: multiple loops of dilated small bowel and coarse granular "soap-bubble" appearance on plain abdominal films. Some cases may show calcifications in the peritoneum (Meconium peritonitis). The Sweat Test is diagnostic of cystic fibrosis (value over 60 meq/L of sweat sodium or chloride are diagnostic). This test is not useful in infant during first weeks of life. Therapy is either: (1) Nonoperative- should be tried first. It consist of a careful gastrograffin enema after the baby is well-hydrated. Gastrograffin 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-Koop, 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.

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F. 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. This 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 consist of resection of the majority of aganglionic bowel except for the most distal rectum, the mucosa and submucosa of this rectum is excised and the normally innervated proximal bowel is pulled through the seromuscular coat of retained rectum and suture immediately above the dentate line. Recently a laparoscopic procedure without colostomy use is being done earlier in life with promising results.

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F1. Total Colonic Aganglionosis

by: Peter Sacher, MD *

Total colonic aganglionosis (TCA) is found in approx. 2 to 13% of patients with Hirschsprung's disease (2). There are three critical phases for patients with TCA (15).

The first period comprises the time from birth until correct diagnosis. Patients with TCA present with a large variety of symptoms. Several authors have outlined the diagnostic problems in patients with TCA (2,4,18). Atypical symptoms may lead to excessively delayed diagnosis. Festen et al. report a delay in diagnosis up to 160 days after birth (4). Patients present with either ileus or symptoms as in typical Hirschsprung's disease but additionally with recurrent vomiting. In patients presenting with ileus, diagnosis may be delayed for several weeks because causative factors like volvulus or meconium ileus do not primarily warrant investigations for aganglionosis. Furthermore, TCA may be associated with other anomalies of the gastrointestinal tract. Only a few reports of TCA associated with small bowel atresia and volvulus can be found (3, 7). Lally et al. report anastomotic failure following repair of ileal atresia due to underlying extensive aganglionosis (11). In cases of midgut volvulus without malrotation, aganglionosis has to be ruled out. Stringer et al. recently published a series of seven full-term infants with meconium ileus due to extensive intestinal aganglionosis (18). Neonatal appendicitis, a very rare disease, may be the leading symptom of TCA. Therefore, rectal biopsies are mandatory in those cases. Ratta et al. state that lack of awareness of the condition may lead to delay in diagnosis and inappropriate treatment (15). Additional to the diagnostic problems due to atypical and heterogenic symptoms, histochemical examination of rectal biopsies may prove negative or equivocal because increased acetylcholinesterase activity may not be present in TCA (5,10,12). Furthermore, there is no typical radiographic pattern (13,17). Plain abdominal radiographs usually suggests low bowel obstruction whereas barium enema usually does not show pathognomonic features.

If no mechanical obstruction is found at laparotomy in neonates presenting with ileus, it is suggested to resect the appendix to rule out TCA. If rectal mucosal biopsies are negative or equivocal, biopsies should be repeated or a formal sphincterectomy for thorough analysis is done.

The second period lasts from the raising of stoma to its closure, including the definite surgical procedure. Failure to thrive and excessive fluid losses have been reported in patients with ileostomies (2). Post-ileostomy complications, however, have been eliminated after the importance of oral sodium supplementation to maintain the enteral co-transport system has been realized (16). In the series of Cass & Myers, ileostomy dysfunction was common with a 20% prolapse rate and 25% rate of persistent excessive losses (2). Interestingly, right transversostomies may show a good function even in cases of TCA. Therefore, frozen section biopsies are mandatory when raising a stoma.

The definitive surgical procedure has been debated (2,5,8,9,15). Colonic patch graft procedures were the first proposals for surgical management of TCA (14). The rational behind were to use the distinctive resorptive function of part of the aganglionic colon (6). Use of the right colon has the theoretical advantage of improved water resorption. However the colon patch procedures have significant complications, e.g., enterocolitis, ulceration of the aganglionic pouch, perforations and extreme dilatation. Multiple modifications of the technique have been reported but none were superior (1,8,9,18). Actually, a modified Duhamel's pull-through procedure seems favorable in the treatment of TCA (2,15).

The third critical phase begins with closure of the stoma. Complications in this period are predominantly recurrent episodes of sub-ileus and diarrhea or nocturnal incontinence. The cause for sub-ileus is a raised tone in the residual sphincter. Repeated manual anal dilatations may be mandatory. The treatment of diarrhea may be managed by diets and/or Loperamide often in large doses. Side effects of large doses of Loperamide are mental irritability and dyskinesia.

Significantly better survival of the patients with TCA nowadays is mainly attributed to more accurate diagnosis and improved management of infants with ileostomies.

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IMPERFORATE ANUS

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Imperforate anus (IA) is a congenital anomaly in which the natural anal opening is absent. Diagnosis of IA is usually made shortly after birth on routine physical examination. The incidence of IA is approximately 1 in 4000-5000 live births and it is more common in males. Its etiology is unknown and it runs equally through all racial, cultural and socio-economic groups. There is preliminary evidence (> 5 case reports) of the existence of autosomal inheritance (both dominant and recessive) in patients with anorectal malformations.

IA is classified as either "high" or "low" depending on the termination of the distal rectum. When the rectum ends above the levator muscles the malformations are classified as high, and when the rectum ends below the levator muscles the malformations are classified as low. High lesions are more frequent in males, low ones in females. Determination of the level of the lesion (by abdominal x-ray or perineal ultrasound) is critical for appropriate management. Children who have IA may also have other congenital anomalies. The acronym VACTERL describes the associated problems that infants with IA may have: Vertebral defects, Anal atresia, Cardiac anomalies, Tracheoesophageal fistula, Esophageal atresia, Renal anomalies, and Limb anomalies.

Repair of low IA is relatively simple and is usually treated with perineal anoplasty; however, repair of high IA is more complex. Patients are initially given a temporary colostomy and time is given to allow the child to grow. A pull-through operation is completed at a later date. Independent of the level of the lesion, the goal of the surgery is the creation of adequate nerve and muscle structures around the rectum and anus to provide the child with the capacity for bowel control.

MALES: 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.

FEMALES: 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 this cases. Perineal fistulas can be managed with cutback without colostomy during the neonatal period.

The most important prognostic characteristic is the severity of the IA. Patients with low IA have a good probability of having normal stool patterns. Patients with high IA report more problems such as fecal incontinence and constipation. For patients who cannot maintain normal bowel function, the use of a special diet, underpants liners, enemas and drugs have ameliorated their lives. Long-term follow up (with both qualitative and quantitative quality of life considerations) of these patients is very important. In addition, the use of anal endosonography and/or manometry can be used as objective measurements of anorectal pressure and sphincter function.

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H. 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 consist of surgical excision avoiding massive loss of normal bowel and removing all bowel suspect of harboring ectopic gastric mucosa.

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

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

The traditional method of diagnosing and managing ileo-colic intussusception is barium enema contrast reduction. In China where this is the most common surgical emergency in childhood, pneumatic reduction has been used for more than 25 years. A recent tendency toward this approach is seen in recent years in Occident. This consist of rectal insufflation of oxygen at a flow rate of 2 L/min, controlling pressure by adjusting the height of the mercury column, and using maximal pressures of 80 mm Hg. Small bowel aeration is a sign of complete reduction. Series are successful in 70-90% of cases. Gas enema reduction is very 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 flouroscopy time), less costs and less morbidity in cases of perforations.

J. APPENDICITIS

By: Antonio Carlos Sansevero Martins, MD *

INTRODUCTION

The most common cause of emergency laparotomies in all age groups, acute appendicitis remains a diagnostic challenge, with high perforation rates at laparotomy, and, curiously, high negative laparotomy rates. With current standard approach and use of antimicrobial medication the overall mortality is almost zero, and major complications are gradually declining.

ETIOLOGY

Appendiceal lumen obstruction by a fecalith, lymphoid hyperplasia, carcinoid tumor, anomalous vessels, or intestinal parasites as Ascaris lumbricoides leads to acute increase in intraluminal pressure with consequent ischemia and mucosal damage. In association with bacterial proliferation this may lead to gangrene and perforation.

DIAGNOSIS

Early diagnosis is the key for success in management of appendicitis, and it is based solely upon history and physical findings. Image and laboratory studies are of relative value.

Signs and Symptoms

Abdominal pain is the chief complaint of appendicitis. It is usually periumbilical at onset, colicky due to lumen obstruction and distension (visceral referred pain), and in a few hours shifts its location to the RLQ changing to a continuous and intense pattern, explained by parietal irritation caused by the inflamed appendix exudate. Special concern must be given to patients with initial periumbilical pain who localize it into uncommon sites. One must always remember that the localized pain in appendicitis is directly related to where it lays, so we can have flank or back pain in retrocecal appendicitis, dysuria in retro vesical cases, etc. Vomiting, generally after the pain, is usually present in appendicitis. Bowel habits are generally unchanged in early cases, but diarrhea can be present initially in up to 10% of cases, usually associated to pelvic appendicitis.

Physical Examination

Simple examination can predict appendicitis when we see the child walking, bent over his right hip, and stepping carefully to avoid minimum shaking. The apprehensive little patient lies on the examining table in a supine position, with a slight flexion of the right leg. When asked to show the pain site, he points out with a finger. Abdominal examination should be done gently, divided in quadrants, and beginning distant from the site of complaint. It always helps if the patient is asked to help in the examination, the surgeon palpating over his hand, to reduce voluntary guarding. Point tenderness and/or appendiceal masses at RLQ are diagnostic, but unfortunately, they are not present in a great deal of cases. Peritoneal irritation signs can be assessed without performing the classic, Blumberg maneuver, just by soft right heel percussion, or gentle Mc Burney's point finger percussion. In advanced cases with "wooden abdomen" and clear infectious signs, the diagnosis is obvious.

Image and Laboratory

Image resources are poor in diagnosing appendicitis. Plain abdominal films can show a fecalith (20%), a dilated and fixed ileal loop (sentinel loop) and indirect signs such as scoliosis, psoas line and/or pre-peritoneal line blurring. Ultrasound, very good to assess collections, has gained popularity, as far as more experience has been achieved, and modern devices have been developed. In some series, its accuracy has been proved in 90% of cases, but it is still an operator-dependent exam, with a fail in at least 10 to 15% of negative findings, even with color Doppler scans. No lab test can confirm the diagnosis of appendicitis. Dueholm et al could predict 100% normal appendices when WBC and C-reactive protein were normal, but no inverse relation could be proved. The urinalysis is important to assess possible infection or calculi migration. Massive pyuria usually preannounces urinary infection,

but discrete findings can be due to appendicitis.

TREATMENT

Surgical Management

Standard appendectomy through a Babcock-Davis muscle splitting approach is a safe, quick and efficient technique to manage appendicitis. The stump can be inverted, but nowadays simple ligation with electrocauterization is the procedure of choice. In complicated cases, with perforation and peritonitis, exhaustive cavity lavage with saline should be accomplished. No improvement is shown by cavity antibiotic irrigation. Drainage is controversial and we do not use drains for appendicitis. Abdominal wall is approximated with Smith-Jones sutures. Primary skin closure is performed even in advanced cases. Laparoscopic appendectomy has gained force in the '90's as a minimal invasive technique, easy visualization of the structures even in difficult sites, e.g., retrocecal appendices, and accurate suction of purulent material and irrigation of the cavity with very low postoperative complication rates (less than 1.5%). Operative time, costs, intraoperative complications such as a bleeding, stump leakage and visceral puncture are some limitations to be reduced with time and experience. A subgroup of children (obese, female, athletic) benefit from the use of this surgical technique.

In patients with appendiceal mass or abscess, with no signs of peritonitis or systemic sepsis "cooling the process" with antibiotic therapy, followed by interval appendectomy (operating on the patient on an elective basis free from inflammation) can be done with relative security.

Antibiotic Therapy

Prophylactic antibiotic is given one hour before surgery and continued for 24 hours in uncomplicated appendicitis, and must be continued for at least five to seven days, in cases of a complication (perforation, peritonitis, abscess). Triple combination of ampicillin (200 mg/Kg/day), gentamicin(5 mg/Kg/day) and clindamycin (40 mg/Kg/day) has shown the best results in treating infection and preventing septic complications.

COMPLICATIONS

Major complications as intra abdominal abscesses, small bowel obstruction and sepsis, have declined to an overall rate of 7%, in well controlled studies. Minor ones such as wound infection to a rate of less than 2%.

THE "NORMAL APPENDIX"

It has been common knowledge that a diagnostic error of 10 to 15% could be acceptable when compared with the risks of appendicitis, and some series stills carry this unacceptable rate, with no decrease in the complication rates, compared with other series. In the first 24 hours, close attention, and short period reexaminations, added to critical interpretation of image and lab studies will help decrease unnecessary surgery with no increase in complication rates.

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K.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.

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L.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 dymotility and decreased acidity. Management can consist of mechanical or pulsating jet of water fragmentation via the endoscope, operative extraction, shock-wave lithotripsy (ESWL) with subsequent evacuation, or dissolution by oral ingestion of proteolytic enzymes (papain, acetylcysteine, cellulase). With ESWL the shock wave pressure needed is less than half used for urolithiasis cases.

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M. Neuronal Intestinal Dysplasia

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 propia and muscularis mucosa, elevation of acetylcholinesterase in parasympathetic fiber of lamina propia 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.

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IV. HERNIAS AND ADBOMINAL WALL DEFECTS

A. Diaphragmatic Hernias

A.1 Congenital Diaphragmatic Hernia (Bochdalek)

The most common congenital diaphragmatic hernia (CDH) is that which occurs through the postero-lateral defect of Bochdalek. It is caused by failure of the pleuroperitoneal membrane to develop adequately and close before the intestines returning to the abdomen at the tenth week of gestation. The intestines then enter the pleural cavity and cause poor lung development leading to pulmonary hypoplasia (a reduced number of alveoli per area of lung tissue). This defect is postero-lateral in the diaphragm and may vary in size. Stomach, liver or spleen may be partly in chest as well. Frequency is 1:2000 live births and the natural history in prenatally diagnosed CDH is that 60% will die. The clinical presentation is that the newborn becomes rapidly cyanotic, acidotic, and has poor ventilation. Major findings relate to the degree of pulmonary maldevelopment. Chest films will show intestines in the chest. Left sided hernias are more common than right (90% on left). Placement of a radiopaque nasogastric tube may show the tube coiled in the lower left chest. Higher risk factors are: early appearance of symptoms in life, prematurity and associated anomalies. Treatment consist of rapid intubation and ventilation with use of muscle relaxants, placement of a nasogastric tube to prevent gaseous distension of the intestines and preoperative stabilization of arterial blood gases and acid-base status. Surgery can be undertaken when one of the following objectives are met: (1) blood gases normalize with no significant changes between preductal and postductal samples, (2) echocardiogram demonstrate reduce pulmonary pressure and pulmonary peripheral resistance.

Operative management consist of abdominal approach, closure of hernia by primary repair or use of mesh, and correction of malrotation. Postoperative management is very difficult. Due to hypoplastic lungs, there is frequently pulmonary hypertension leading to right-to-left shunting and progressive hypoxemia, hypercarbia, and acidosis that worsens the pulmonary hypertension. The use of chest tubes may cause overstretching of the already hypoplastic alveoli causing: increase pulmonary hypertension, reduce functional residual capacity and reduce lung compliance. Postoperatively, the infant should be kept paralyzed and ventilated and only very slowly weaned from the ventilator. The severity of pulmonary

hypoplasia, both ipsilaterally and contralaterally, is the main determinant of outcome. ECMO (extracorporeal membrane oxygenator) has come to reduce somewhat the mortality of this condition.

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

Delayed presentation beyond the neonatal period is rare, estimated to occur in 4-6% of cases. Infants and children will present with either respiratory or gastrointestinal symptoms such as: chronic respiratory tract infection, vomiting, intermittent intestinal obstruction, and feeding difficulty. Occasionally the child is asymptomatic. The small size of the defect protected by either the spleen or the liver and the presence of a hernial sac may delay the intestinal herniation into the chest. A rise intrabdominal 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.

A.2 Morgagni Hernias

First described in 1769, 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 Cantrells' 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.

A.3 Esophageal Hernias

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.

PROCESUS VAGINALIS REMNANTS

B.1 Inguinal Hernias

A hernia is defined as a protrusion of a portion of an organ or tissue through an abnormal opening. For groin (inguinal or femoral) hernias, this protrusion is into a hernial sac. Whether or not the mere presence of a hernial sac (or processus vaginalis) constitutes a hernia is debated. Inguinal hernias in children are almost exclusively indirect type. Those rare instances of direct inguinal hernia are caused by previous surgery and floor disruption. An indirect inguinal hernia protrudes through the internal inguinal ring, within the cremaster fascia, extending down the spermatic cord for varying distances. The direct hernia protrudes through the posterior wall of the inguinal canal, i.e., medial to deep inferior epigastric vessels, destroying or stretching the transversalis fascia. The embryology of indirect inguinal hernia is as follows: the duct descending to the testicle is a small offshoot of the great peritoneal sac in the lower abdomen. During the third month of gestation, the processus vaginalis extends down toward the scrotum and follows the chorda gubernaculum that extends from the testicle or the retroperitoneum to the scrotum. During the seventh month, the testicle descend 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 observation 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 prematures above the 60 wk. of postconceptual age. The very low birth weight infant with symptomatic hernia can benefit from epidural anesthesia.

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

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

Approximately 1% of females with inguinal hernias will have the testicular feminization syndrome. Testicular feminization syndrome (TFS) is a genetic form of male pseudohermaphroditism (patient who is genetically 46 XY but has deficient masculinization of external genitalia) caused by complete or partial resistance of end organs to the peripheral effects of androgens. This androgenic insensitivity is caused by a mutation of the gene for androgenic receptor inherited as an X-linked recessive trait. In the complete form the external genitalia appear to be female with a rudimentary vagina, absent uterus and ovaries. The infant may present with inguinal hernias that at surgery may contain testes. Axillary/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. This 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.

B.2 Hydroceles

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 resolved 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.

C. Undescended Testis

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 undescended testis found in 0.28% of males can be palpable (80%; most at inguinal canal), or non-palpable (20%). Testes that can be manually brought to the scrotum are retractile and need no further treatment. Parents should know the objectives, indications and limitations of an orchiopexy: that the testis could not exist (testicular vanishing syndrome), even after descend can atrophy, that it cannot be fixed and removal is a therapeutic possibility. To improve spermatogenesis (producing an adequate number of spermatozoids) surgery should be done before the age of two. Electron microscopy has confirmed an arrest in spermatogenesis (reduced number of spermatogonias and tubular diameter) in undescended testis after the first two years of life. Other reasons to pex are: a higher incidence of malignancy, trauma and torsion, and future cosmetic and psychological problems in the child. The management is surgical; hormonal (Human Chorionic Gonadotropin) treatment has brought conflicting results except bilateral cases. Surgery is limited by the length of the testicular artery. Palpable testes have a better prognosis than non-palpable. Laparoscopy can be of help in non-palpable testis avoiding exploration of the absent testis.

D. Umbilical Hernias

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An umbilical hernia is a small defect in the abdominal fascial wall in which fluid or abdominal contents protrude through the umbilical ring. The presence of a bulge within the umbilicus is readily palpable and becomes more apparent when the infant cries or during defecation. The actual size of the umbilical hernia is measured by physical examination of the defect in the rectus abdominis muscle, and not by the size of the umbilical bulge. The size of the fascial defect can vary from the width of a fingertip to several centimetres.Embryologically, the cause of an umbilical hernia is related to the incomplete contraction of the umbilical ring. The herniation of the umbilicus is a result of the growing alimentary tract that is unable to fit within the abdominal cavity. Umbilical hernias are more prevalent in females than in males and are more often seen in patients with African heritage. The increased frequency of umbilical hernias has also been attributed to premature babies, twins and infants with long umbilical cords. There is also a frequent association with disorders of mucopolysaccharide metabolism, especially Hurler's Syndrome (gargoylism).Most umbilical hernias are asymptomatic; the decision to repair the umbilical hernia in the first years of life is largely cosmetic and is often performed because of parental request, not because of pain or dysfunction. In the past, some parents use to tape a coin over the umbilical bulge, however, manual compression does not have an effect on the fascial defect.Treatment of umbilical hernia is observation. Most umbilical hernias spontaneously close by age two, with 90% closed by age three and 95% closed by age five. However, surgical repair is recommended if the hernia has not closed by the age of five.If a large defects (> 2cm) remains after the age of 2, spontaneously repair in unlikely and may be closed surgically. The incidence of incarceration (trapped intestinal loop) is rare, even in larger defects. Females should especially have their umbilical hernia corrected before pregnancy because of the associated increased intra-abdominal pressure that could lead to complications. The procedure is simple and incidence of complication such as infection is extremely rare. The repair is usually done as outpatient surgery under general anesthetic.

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E. Omphalocele

The three most common abdominal wall defect 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.

F. Gastroschisis

Gastroschisis is a congenital evisceration of part of the abdominal content through an anterior abdominal wall defect found to the right of the umbilicus. The protruding gut is foreshortened, matted, thickened and covered with a peel. In a few babies (4 to 23%) an intestinal atresia (IA) further complicates the pathology. IA complicating gastroschisis may be single or multiple and may involve the small or large bowel. The IA might be the result of pressure on the bowel from the edge of the defect (pinching effect) or an intrauterine vascular accident. Rarely, the orifice may be extremely narrow leading to gangrene or complete midgut atresia. In either case the morbidity and mortality of the child is duplicated with the presence of an IA. Management remains controversial. Alternatives depend on the type of closure of the abdominal defect and the severity of the affected bowel. With primary fascial closure and good-looking bowel primary anastomosis is justified. Placement of a silo calls for delayed resection performing a second look operation at a later stage to save intestinal length. Angry looking dilated bowel prompts for proximal diversion, but the higher the enterostomy the greater the problems of fluid losses, electrolyte imbalances, skin excoriation, sepsis and malnutrition. Closure of the defect and resection with anastomosis two to four weeks later brings good results. Success or failure is related to the length of remaining bowel more than the specific method used.

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

B.1 Necrotizing Enterocolitis (NEC)

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

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

C. Upper GI Bleeding (Older Children)

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

1. Esophagus

(a) Varices- usually presents as severe upper gastrointestinal bleeding in a 2-3 year old who has previously been healthy except for problems in the neonatal period. This is a result of extrahepatic portal obstruction (portal vein thrombosis most commonly), with resulting varices. The bleeding may occur after a period of upper respiratory symptoms and coughing. Management is initially conservative with sedation and bedrest; surgery 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 consist of antacids, frequent small feeds, occasionally medications and if not rapidly improved, then surgical intervention with a fundoplication of the stomach.

(c) 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 hemorraghe, 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. Older GI Bleeding (Older Child)

D.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.

D.2 Meckel's Diverticulum

Meckel's diverticulum (MD), the pathologic structure resulting from persistence of the embryonic vitelline duct (yolk stalk), is the most prevalent congenital anomaly of the GI tract. MD can be the cause of: gastrointestinal bleeding (most common complication), obstruction, inflammation and umbilical discharge in children and 50% occur within the first two years of life. Diagnosis depends on clinical presentation. Rectal bleeding from MD is painless, minimal, recurrent, and can be identified using 99mTc- pertechnetate scan; contrasts studies are unreliable. Persistent bleeding requires arteriography or laparotomy if the scan is negative. Obstruction secondary to intussusception, herniation or volvulus presents with findings of fulminant, acute small bowel obstruction, is diagnosed by clinical findings and contrast enema studies. The MD is seldom diagnosed preop. Diverticulitis or perforation is clinically indistinguishable from appendicitis. Mucosal polyps or fecal umbilical discharge can be caused by MD. Overall, complications of Meckel's are managed by simple diverticulectomy or resection with anastomosis. Laparoscopy can confirm the diagnosis and allow resection of symptomatic cases. Removal of asymptomatic Meckel's identified incidentally should be considered if upon palpation there is questionable heterotopic (gastric or pancreatic) mucosa (thick and firm consistency) present.

D.3 Juvenile Polyps

Childhood polyps are usually juvenile (80%). Histology features a cluster of mucoid lobes surrounded by flattened mucussecreting glandular cells (mucous retention polyp), no malignant potential. Commonly seen in children age 310 with a peak at age 56. As a rule only one polyp is present, but occasionally there are two or three almost always confined to the rectal area (within the reach of the finger). Most common complaint is bright painless rectal bleeding. Occasionally the polyp may prolapse through the rectum. Diagnosis is by barium enema, rectal exam, or endoscopy. Removal by endoscopy is the treatment of choice. Rarely colotomy and excision are required.

VI. PANCREATIC AND BILIARY DISORDERS

A. Pancreatitis

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.

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B. Biliary Atresia

Persistent conjugated hyperbilirubinemia (greater than 20% of total or 1.5 mg%) should be urgently evaluated. Initial evaluation should include a well-taken history and physical exam, partial and total bilirubin determination, type and blood group, Coomb's test, reticulocyte cell count and a peripheral smear. Cholestasis means a reduction in bile flow in the liver, which depends on the biliary excretion of the conjugated portion. Reduce flow causes retention of biliary lipoproteins that stimulates hypercholesterolemia causing progressive damage to the hepatic cell, fibrosis, cirrhosis and altered liver function tests. Biliary Atresia (BA) is the most common cause of persistently direct (conjugated) hyperbilirubinemia in the first three months of life. It is characterized by progressive inflammatory obliteration of the extrahepatic bile ducts, an estimated incidence of one in 15,000 live births, and predominance of female patients. The disease is the result of an acquired inflammatory process with gradual degeneration of the epithelium of the extrahepatic biliary ducts causing luminal obliteration, cholestasis, and biliary cirrhosis. The timing of the insult after birth suggests a viral etiology obtained transplacentally. Almost 20% of patients have associated anomalies such as: polysplenia, malrotation, situs inversus, preduodenal portal vein and absent inferior vena cava. Histopathology is distinguished by an inflammatory process in several dynamic stages with progressive destruction, scar formation, and chronic granulation tissue of bile ducts. Physiologic jaundice of the newborn is a common, benign, and self-limiting condition. In BA the patient develops insidious jaundice by the second week of life. The baby looks active, not acutely ill and progressively develops acholic stools, choluria and hepatomegaly. Non-surgical source of cholestasis shows a sick, low weight infant who is jaundiced since birth. The diagnostic evaluation of the cholestatic infant should include a series of lab tests that can exclude perinatal infectious (TORCH titers, hepatitis profile), metabolic (alpha-1-antitrypsin levels), systemic and hereditary causes. Total bilirubin in BA babies is around 6-10 mg%, with 50-80% conjugated. Liver function tests are nonspecific. Lipoprotein-X levels greater than 300 mg% and Gamma Glutamyl Transpeptidase (GGT) above 200 units% suggest the diagnosis. The presence of the yellow bilirubin pigment in the aspirate of duodenal content excludes the diagnosis of BA. Ultrasound study of the abdomen should be the first diagnostic imaging study done to cholestatic infants to evaluate the presence of a gallbladder, identify intra or extrahepatic bile ducts dilatation, and liver parenchyma echogenicity. The postprandial contraction of the gallbladder eliminates the possibility of BA even when nuclear studies are positive. Nuclear studies of bilio-enteric excretion (DISIDA) after pre-stimulation of the microsomal hepatic system with phenobarbital for 3-5 days is the diagnostic imaging test of choice. The presence of the radio-isotope in the GI tract excludes the diagnosis of BA. Percutaneous liver biopsy should be the next diagnostic step. The mini-laparotomy is the final diagnostic alternative. Those infant with radiographic evidence of patent extrahepatic biliary tract has no BA. Medical management of BA is uniformly fatal. Kasai portoenterostomy has decreased the mortality of BA during the last 30 years. Kasai procedure consists of removing the obliterated extrahepatic biliary system, and anastomosing the most proximal part to a bowel segment. Almost three-fourth of patients will develop portal hypertension in spite of adequate postoperative bile flow. They will manifest esophageal varices, hypersplenism, and ascites. Hepatic transplantation is reserved for those patients with failed portoenterostomy, progressive liver failure or late-referral to surgery.

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C. Choledochal Cyst

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

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D. Cholelithiasis

With the increase use of sonography in the work-up of abdominal pain, cholelithiasis is diagnosed more frequent in children. Gallstones occur as consequence of loss of solubility of bile constituents. Two types are recognize: 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 intrabdominal 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. 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.

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E. 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.

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VII. TUMORS

A. Wilms Tumor & Genetics

Wilms tumor (WT) is the most common intra-abdominal malignant tumor in children affecting more than 400 children annually in the USA. It has a peak incidence at 3.5 years of age. WT present as a large abdominal or flank mass with abdominal pain, asymptomatic hematuria, and occasionally fever. Other presentations include malaise, weight loss, anemia, left varicocele (obstructed left renal vein), and hypertension. Initial evaluation consists of: Abdominal films, Ultrasound, IVP, urinalysis, Chest-X-rays and Computed Tomography. The presence of a solid, intrarenal mass causing intrinsic distortion of the calyceal collecting system is virtually diagnostic of Wilms tumor. Doppler sonography of the renal vein and inferior vena cava can exclude venous tumor involvement. Metastasis occurs most commonly to lungs and occasionally to liver. Operation is both for treatment and staging to determine further therapy.

Following NWTSG recommendation's primary nephrectomy is done for all but the largest unilateral tumors and further adjuvant therapy is based on the surgical and pathological findings. Important surgical caveats consist of using a generous transverse incision, performing a radical nephrectomy, exploring the contralateral kidney, avoiding tumor spillage, and sampling suspicious lymph nodes. Nodes are biopsied to determine extent of disease. WT staging by NWTSG consists of:

Stage I- tumor limited to kidney and completely resected.
Stage II- tumor extends beyond the kidney but is completely excised.
Stage III- residual non-hematogenous tumor confined to the abdomen.
Stage IV- hematogenous metastasis.
Stage V- bilateral tumors.

Further treatment with chemotherapy or radiotherapy depends on staging and histology (favorable vs. non-favorable) of the tumor. Non-favorable histologic characteristics are: anaplasia (three times enlarged nucleus, hyperchromatism, mitosis), sarcomatous or rhabdoid degeneration. The success in managing WT has been remarkable the result of stratification, registry and study from the NWTSG. Disease-free survival is 95% for Stage I and approximately 80% for all patients. Prognosis is poor for those children with lymph nodes, lung and liver metastasis.

WT occurs either sporadic (95%), familial (1-2%) or associated with a syndrome (2%). Such syndromes predisposing to WT are WAGR (Wilms, aniridia, genitourinary malformation and mental retardation), Beckwith-Wiedemann Syndrome (gigantism, macroglossia, pancreas cell hyperplasia, BWS), and Denys-Drash Syndrome (male pseudohermaphrodite, nephropathy and Wilms tumor, DDS). They tend to occur in younger patients. Sporadic WT is associated in 10% of cases with isolated hemihypertrophy or genitourinary

malformations such as hypospadia, cryptorchidism and renal fusion. Bilateral synchronous kidney tumors are seen in 5-10% of cases. Routine abdominal ultrasound screening every six months up to the age of eight years is recommended for children at high risk for developing WT such as the above-mentioned syndromes.

It was originally thought that WT developed after the two-hit mutational model developed for retinoblastoma: When the first mutation occurs before the union the sperm and egg (constitutional or germline mutation) the tumor is heritable and individuals are at risk for multiple tumors. Nonhereditary WT develops as the result of two-postzygotic mutations (somatic) in a single cell. The two-event hypothesis predicts that susceptible individuals such as familial cases, those with multifocal disease and those with a congenital anomaly have a lower median age at diagnosis than sporadic cases. It is now believed that several genes' mutations are involved in the overall WT pathogenesis.

Loss of whole portions of a chromosome is called loss of heterozygosity (LOH), a mechanism believed to inactivate a tumor-suppressor gene. WT has been found 50% of the time to contain LOH at two genetic loci: 11p13 and 11p15. WT will develop in 30% of WAGR children. Children with the WAGR association shows a deletion in the short arm of chromosome 11 band 13 (11p13) but a normal 11p15 region. Up to a third of sporadic WT have changes in the distal part of chromosome 11, a region that includes band p13. The region of the deletion has been named the WT1 gene, a tumor suppressor gene that also forms a complex with another known tumor-suppressor, p53. WT1 gene express a regulated transcription factor of the zinc-finger family proteins restricted to the genitourinary system, spleen, dorsal mesentery of the intestines, muscles, central nervous system (CNS) and mesothelium. WT1 is deleted in all WAGR syndromes cases. The important association of WT1 mutation and WAGR syndrome with intralobar nephrogenic rests immediately suggest that WT1 expression be necessary for the normal differentiation of nephroblasts. Only five to 10% of sporadic WT have thus far been shown to harbor WT1 mutations. Inactivation of WT1 only affects organs that express this gene such as the kidney and specific cells of the gonads (Sertoli cells of the testis and granulosa cells of the ovary). WT1 has been shown to cause the Denys-Drash syndrome. Most of the mutations described in DDS patients are dominant missense mutations.

A small subset of BWS has a 11p15 duplication or deletion. The region 11p15 has been designated WT2 gene and is telomeric of WT1. Beckwith- Wiedemann form of WT is also associated with IGF2, an embryonal growth-inducing gene. This might prove that two independent loci may be involved in tumor formation. Candidates genes include insulin-like growth factor II gene (IGFII) and the tumor suppressor gene H19. A substantial fraction of WT (without LOH at the DNA level) has been found to have altered imprints with resultants over expression of IGFII and loss of expression of the tumor suppressor H19. IGFII may be operating like an oncogene by perpetuating nephroblast and may account for the perilobar rests observed in BWS patients.

A gene for a familial form (FWT1) of the tumor has also been identified in chromosome 17q. There also might be a gene predisposing to Wilms tumor at chromosome 7p, where constitutional translocations have been described. Mutation in p53 is associated with tumor progression, anaplasia and poor prognosis. Most WT are probably caused by somatic mutations in one or more of the increasing number of WT genes identified.

A few chromosomal regions have seen identified for its role in tumor progression. LOH at chromosome 16q and chromosome 1p has been implicated in progression to a more malignant or aggressive type Wilms' tumor with adverse outcome. This occurs in approximately 20% of patients with WT. These children have a relapse rate three times higher and a mortality rate twelve times higher than WT without LOH at chromosome 1p. p53 is also associated with the so called anaplastic unfavorable histology.

Patients with WT and a diploid DNA content (indicating low proliferation) have been found to have an excellent prognosis. Hyperdiploidy (high mitotic activity) is a poor prognostic feature of Wilms tumor, rhabdomyosarcoma and Osteosarcoma.

Nephrogenic rests are precursor lesions of WT. Two types are recognized: perilobar nephrogenic rests (PLNR) limited to the lobar periphery, and intralobar nephrogenic rests (ILNR) within the lobe, renal sinus or wall of the pelvocaliceal system. The strong association between ILNR, aniridia and Denys-Drash syndrome where the WT1 zinc finger gene has been implicated suggests that this locus might be linked to the pathogenesis of ILNR. Also the association between BWS and some cases of hemihypertrophy with abnormalities of more distant loci on chromosome 11p raises the possibility that the putative WT2 gene might be more closely linked to PLNR.

An advantage of genetic testing is that children with sporadic aniridia, hemihypertrophy or the above discussed syndromes known to be at high risk for developing WT can undergo screening of the germline DNA. This might identify if they harbor the mutation and need closer surveillance for tumor development.

B. Neuroblastoma & Genetics

Neuroblastoma (NB) is the most common extracranial solid tumor in infants. More than 500 new cases are diagnosed annually in the United States. Most neuroblastomas (75%) arise in the retroperitoneum (adrenal gland and paraspinal ganglia), 20% in the posterior mediastinum, and 5% in the neck or pelvis. NB is a solid, highly vascular tumor with a friable pseudocapsule. Most children present with an abdominal mass, and one-fourth have hypertension. Other have: Horner's syndrome, Panda's eyes, anemia, dancing eyes or vaso-intestinal syndrome. Diagnosis is confirmed with the use of simple X-rays (stipple calcifications), Ultrasound, and CT-Scan. Work-up should include: bone marrow, bone scan, myelogram (if there is evidence of intraspinal extension), and plasma/urine tumor markers level: vanillylmandelic acid (VMA), homovanillic acid (HVA) and dopamine (DOPA).

Management of NB depends on the stage of disease at diagnosis. Localized tumors are best managed with surgical therapy. Partially resected or unresectable cases need chemotherapy a/o radiotherapy after establishing a histologic diagnosis. Independent variables determining prognosis are age at diagnosis and stage of disease. The Evans classification for NB staging comprised:

Stage I - tumor confined to an organ of origin.
Stage II - tumor extending beyond an organ of origin, but not crossing the midline. Ipsilateral lymph nodes may be involved.
Stage III - tumor extending beyond midline. Bilateral lymph nodes may be involved.
Stage IV - remote disease involving skeleton, bone marrow, soft tissue or distant lymph nodes.
Stage IVS - same as stage I or II with presence of disease in liver, skin or bone marrow.

Young children with stage I/II have a better outcome. A poor outcome is characteristic of higher stages, older patients and those with bone cortex metastasis. Other prognostic variables are: the site of primary tumor development, maturity of tumor, presence of positive lymph nodes, high levels of ferritin, neuron-specific enolase, and diploid DNA.

Neuroblastoma is a malignant tumor of the postganglionic sympathetic system that develops from the neural crest: sympathetic ganglion cells and adrenal gland. In vitro three cell types have been identified:

1- neuroblastic (N-type) cells that are tumorigenic. These cells are responsible for producing cathecolamines and vasoactive substances which help in diagnosis and follow-up therapy.
2- the Schwannian or substrate-adherent (S-type) cells that are non-tumorigenic, and the
3- intermediate I-cells.

NB can behave seemingly benignly and undergo spontaneous regression, mature into a benign ganglioneuroma or most commonly progress to kill its host. This disparate behavior is a manifestation that we are dealing with related tumors with differently genetic and biological features associated with a spectrum of clinical behaviors.

Conclusive associations with environmental factors have not been proved in NB. Hereditary factors are important in NB since a few cases exhibit predisposition following a dominant pattern of inheritance. The most characteristic cytogenetic abnormality of neuroblastoma is deletion of the short arm of chromosome 1 in locus 36 (1p36) occurring in 50 to 70% of primary diploid tumors. LOH of the short arm of chromosome 1 is also associated with an unfavorable outcome, suggesting that a tumor suppression gene may be found in this region. The common region of deletion or LOH resides at the distal end of the short arm of chromosome 1 from 1p36.2 to 1p36.3. Loss or inactivation of a gene at this site is critical for progression of neuroblastoma. A few candidate genes from this site have been mapped. LOH in chromosome 14 long arm (14q) has also been identified in 25-50% or neuroblastoma cells studied but no clinical behavior has been identified with this finding. Gain of chromosome 17 is associated with more aggressive tumors.

Another consistent chromosomal aberration identified in 25 to 30% of NB cells is the presence of double-minute chromosomes, small fragments of chromatin containing multiple copies of the oncogene N-myc produced by amplification. N-myc protooncogene is found on chromosome 2p and its activation results in tumor formation. The amplified N-myc sequence is found on extrachromosomal double minutes (DM) or on homogeneous staining regions (HSR) involving different chromosomes in neuroblastoma (N-type) cell lines. N-myc amplification is strongly associated with advance stages of disease, rapid tumor progression and poor outcome independent of the stage of the tumor or the age of the patient. NB tumors associated with N-myc amplification needs aggressive therapy. N-myc amplification associated with deletion of 1p is correlated with a poor outcome. Deletion of the long arm of chromosome 1 (1q-) is also a poor prognostic sign.

Though most NB cells are diploid, a good number of them are hyperdiploid or triploid. Hyperdiploidy is a good prognostic feature of NB and embryonal rhabdomyosarcoma in infants, while diploid tumors at any age and hyperdiploid in older patients carry a worse prognosis requiring more intensive treatment.

Neuroblast cells needs nerve growth factor (NGF) for proper differentiation. NB tumor cells do not respond to NGF or do not express the receptor. This receptor consists of three transmembrane tyrosine kinase receptors (TRK-A, TRK-B, and TRK-C), known together as the TRK receptor. TRK-A is detected in 90% of NB cells and correlates inversely with N-myc expression. High TRK-A levels correlate strongly with improved survival and plays a role in the propensity for tumors to regress or differentiate into a more benign nature. TRK-B is associated with more matured tumors and TRK-C with lower stage tumors. Alteration in the NGF receptor function or expression promotes tumorigenesis. In conclusion, high levels of TRK expression are associated with better prognosis, earlier stage, lower patient age and lack of N-myc expression.

Neuroblastomas in newborns, cystic tumors, bilateral tumors in infants, and infants less than one year of age with neuroblastoma stage IV-S can undergo neuronal cell differentiation with spontaneous regression. It is thought that high level of TRK-A found in this cases might explain differentiation and regression as high level of this glycoprotein is associated with a favorable prognosis. Regression might be associated with non-affected tumor cell apoptosis.

Other biological markers associated with NB are the multidrug resistance-related protein (MRP) gene, telomerase activity and bcl-2 gene activity. MRP shows a strong correlation with an advanced clinical stages and poor prognosis. High telomerase activity is associated with poor prognosis and high N-myc amplification. The bcl-2 gene produces a protein that prevents neuronal cell death (apoptosis) and promotes tumor progression. Bcl-2 expression is associated with a poor outcome. Apoptosis in NB may result in tumor progression.

The RET proto-oncogene is a protein tyrosine kinase gene (Ret protein) expressed in the cells derived from the neural crest. The activation of RET involves a chromosomal inversion of the long arm of chromosome 10 that juxtaposes the tyrosine kinase encoding domain of RET to the amino terminal sequences of at least three unrelated genes. Germline mutations in the RET gene have been associated with neuroblastoma, pheochromocytoma, multiple endocrine neoplasia (MEN) 2, familial medullary thyroid carcinoma (MTC), radiation-induced thyroid papillary carcinoma, and recently Hirschsprung's disease. RET analysis is a suitable method to detect asymptomatic children with MEN at risk to develop MTC allowing us to consider thyroidectomy at a very early stage of neoplasm development (C-cell hyperplasia) or prophylactically.

High levels of neuron specific enolase and serum ferritin levels are associated with a poor prognosis in NB. Nm-23 and ganglioside GD2 are still other tumor markers associated with poor outcome, active disease and tumor progression.

C. Rhabdomyosarcoma & Genetics

Rhabdomyosarcoma (RMS), the most common soft tissue sarcoma in infants and children, represents about 5-15% of all malignant solid lesions. It has a peak incidence before the age of five years, and a second surge during early adolescence. Head, neck and pelvic malignancies are more prevalent in infancy and early childhood, while trunk, extremity and paratesticular sites are largely a disease of adolescents. RMS arises from a primitive cell type and occurs in mesenchymal tissue at almost any body site excluding brain and bone. The predominant histologic type in infants and small children is embryonal rhabdomyosarcoma, occurring in the head and neck, genitourinary tract and retroperitoneum. Embryonal RMS is associated with a favorable prognosis. Botryoid RMS is a subtype of the embryonal variety, which ordinarily extends into body cavities such as bladder, nasopharynx, vagina, or bile duct. The alveolar cell type, named for a superficial similarity to the pulmonary alveoli, is the most common form found on the muscle masses of the trunk and extremities, and is seen more frequently in older children and young adults. Alveolar RMS is associated with a poor prognosis. This unfavorable prognosis is the result of early and wide dissemination, bones marrow involvement and poor response to chemotherapy.

Clinical findings, diagnostic evaluation and therapy depend upon location of the primary tumor and are beyond the scope of this review. Head and neck RMS are most common and occur in the orbit, nasopharynx, paranasal sinuses, cheek, neck, middle ear, and larynx. 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 gross tumor as possible. The trend in management is more chemotherapy with conservative surgical therapy. Survival has depended on primary site, stage of disease, and treatment given.

Most RMS occurs sporadically. Approximately 5% are associated to syndromes such as Beckwith-Wiedemann with LOH at the 11p15 locus, Li-Fraumeni, the neurofibromatosis-NF1 gene located on 17q11, Basal Cell Nevus, and the Fetal Alcohol syndrome. Other risk factors in the development of RMS include maternal use of marijuana and cocaine, exposure to radiation, and maternal history of stillbirth.

Alveolar and embryonal RMS are the most genetically studied sarcomas in children. The expression of a number of human paired box-containing (PAX) genes has been correlated with various types of RMS. In alveolar RMS novel fusion genes encoding chimeric fusion proteins have been identified. The most consistent genetic mutation identified in more than 70% of alveolar RMS is translocation of chromosomes 2 and 13, t(2;13)q35-37;q14). The PAX3 loci in chromosome 2 fuses to the FKHR (fork head in rhabdomyosarcoma) domain of chromosome 13 creating a powerful chimeric PAX3-FKHR gene that encodes an 836 amino acid fusion protein. This information is obtained using reverse transcriptase PCR assays of alveolar RMS or by protein immunoprecipitation with PAX3 and FKHR antisera. The PAX3-FKHR protein is an active transcription factor. The t(2;13) activates the oncogenic potential of PAX3 by dysregulating or exaggerating its normal function in the myogenic lineage and affecting the cellular activities of growth, differentiation and apoptosis. Another of the reported translocation is t(1;13)(p36;q14) involving chromosome 1 and 13 in 10% of alveolar RMS. In this variant, Chromosome 1 locus encoding PAX7 fused to FKHR in chromosome 13 resulting in another chimeric transcript PAX7-FKHR. PAX7-FKHR tumors tend to occur in younger patients, are more often in the extremity, are more often localized lesions and are associated with significantly longer event-free survival. Still, a small subset of alveolar RMS does not contain either fusion mutation. The PAX3-FKHR and the variant PAX7-FKHR fusions are associated with distinct clinical phenotypes. Identification of fusion gene status by PCR is a useful diagnostic tool in differentiating RMS from other round cell tumors.

Embryonal RMS contains frequent allelic loss on chromosome 11 (11p15), a genetic feature specific for this type of tumor. Allelic loss is manifested by the absence of one of the two signals in the tumor cells indicating a genetic event such as a chromosome loss, deletion, or mitotic recombination that eliminates one allele and the surrounding chromosomal region. The smallest affected region has been localized to chromosomal region 11p15.5. The presence of a consistent region of allelic loss is often indicative of the presence of a tumor suppressor gene inactivated in the associated malignancy. The mechanism for inactivation of tumor suppressor genes is postulated to be a two-hit scenario in which both copies of the gene are sequentially inactivated: a small point mutation inactivates one of the two alleles, preferably the maternal side allele, and the allelic loss event inactivate the second allele (the paternal allele). This leads to over expression of the insulin-like growth factor II gene known to play a role in the development of embryonal tumors.

Other alterations associated with embryonal RMS are distinct patterns of chromosomal gains (chromosomes 2,7,8,12,13,17,18, and 19) in contrast with alveolar RMS which shows genomic amplification of chromosomal region 12q13-15 in 50% of cases. Notably, these distinct changes predominantly involved chromosomes 2, 12, and 13 in both subtypes. Additionally embryonal RMS cases shows mutations of members of the RAS gene family, a second proto-oncogene. Both tumors share alterations in the p53 gene at the germline level contributing to increase susceptibility to other tumors characteristics of the Li-Fraumeni syndrome. There is also greater over expression of c-myc in alveolar RMS when compared with embryonal RMS. All this favors a multi-step origin of RMS tumors generating phenotypic changes of growth autonomy, abnormal differentiation and motility.

The Li-Fraumeni familial cancer syndrome is manifested by increased susceptibility of affected relatives to develop a diverse set of malignancies during early childhood. The major features of the syndrome include breast cancer, osteosarcoma, rhabdomyosarcomas of soft tissue, glioblastoma, leukemia and adrenal cortical carcinoma. More than one-half of the cancers overall and nearly one-third of the breast cancers were diagnosed before 30 years of age. Among females, breast cancer is the most common. Germline mutations within a defined region of the p53 gene have been found in families with the Li-Fraumeni syndrome. Persistence of the mutation in the germline suggests a defect in DNA repair in the family member first affected. Asymptomatic carriers of p53 germline mutation needs closed evaluation and follow-up for early detection and treatment in case neoplasia develops.

D. Hepatic Tumors: Hepatoblastoma

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.

E. Sacrococcygeal Teratoma

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 consist of tissue from the three germ cell layers. SCT is classified as: mature, immature, or malignant (endodermal sinus) and produces alpha-feto protein (AFP). Prenatal sonographic diagnostic severity criteria are: tumor size greater than the biparietal diameter of the fetus, rapid tumor growth, development of placentomegaly, polyhydramnios and hydrops. Large tumors should benefit from cesarean section to avoid dystocia or tumor rupture. Management consist of total tumor resection with coccyx (recurrence is associated with leaving coccyx in place). Every recurrence of SCT should be regarded as potentially malignant. Malignant or immature SCT with elevated AFP after surgical resection will benefit from adjuvant chemotherapy. Survival is 95% for mature/immature tumors, but less than 80% for malignant cases. Follow-up should consist of (1) meticulous physical exam every 3-6 months for first three years, (2) serial AFP determination, (3) fecal/urodynamic functional studies. Long term F/U has found a 40% incidence of fecal and urinary impairment associated to either tumor compression of pelvic structures or surgical trauma.

F. Ovarian Tumors

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

G. Thyroid Nodules

The need to differentiate malignant from benign thyroid nodules is the most challenging predicament in management.Present diagnostic work-up consists of ultrasonography (US), radionuclear scans (RNS) and fine-needle aspiration biopsy (FNAB). After reviewing our ten-year experience with twenty-four pediatric thyroid nodules we found nineteen benign and five malignant lesions. Benign nodules were soft, movable, solitary and non-tender. Malignant nodules were found during late adolecence, characterized by localized tenderness, a multigandular appearance and fixation to adjacent tissues. US and RNS were of limited utility since malignancy was identified among cystic and hot nodules respectively. Suppressive thyroid hormone therapy was useless in the few cases tried. FNAB in eighteen cases did not limit the number of thyroid resections. It showed that the probability that a malignant nodule had suspicious or frankly malignant cytology was 60%. The specificity was 90%. This is the result of a higher number of patients with follicular cell cytology in the aspirate. No attempts should be made to differentiate follicular adenoma from carcinoma since capsular and vascular invasion cannot be adequately assessed by FNAB.The physical exam findings, persistence of the nodule, progressive growth and cosmetic appearance were the main indications for surgery. FNAB is a safe procedure that plays a minor role in the decision to withhold surgery. Its greatest strength is to anticipate in case of malignancy that a more radical procedure is probably needed. FNAB, US and RNS should not replace clinical judgement or suspicion as the most important determinants in management.

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

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

H. Burkitt's Lymphoma

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

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VIII. GYNECOLOGIC 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.

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B. Ovarian Cysts

Ovarian cysts in fetus and infants are usually follicular in nature and less than 2 cm in size. They are commonly diagnosed between the 28th and 39th wk. of gestation by sonography. Hypotheses on etiology are: (1) Excessive fetal gonadotropic activity, (2) enzymatic abnormalities of the theca interna, and (3) abnormal stimulation by the mother HCG. Obstetric management consists on observation and vaginal delivery. After birth, diagnostic assessment and management will depend on the size and sonographic characteristics of the cyst. Simple anechoic cysts, and those less than 5 cm in size can be observed for spontaneous resolution. Cyst with fluid debris, clot, septated or solid (complex nature), and larger than 5 cm should undergo surgical excision due to the higher incidence of torsion, perforation and hemorrhage associated to them. Percutaneous aspiration of large simple cysts with follow-up sonography is a well-accepted therapy, preserving surgery for recurrent or complicated cases. Surgical therapy is either cystectomy or oophorectomy that can result in loss of normal ovarian tissue.

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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.

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IX. PRENATAL CONGENITAL MALFORMATIONS

A. Fetal Surgery

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

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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).

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C. Prenatal CCAM

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.

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X. PEDIATRIC LAPAROSCOPY

A. Physiology of the Pneumotperitoneum

The concept behind minimally invasive surgery is that the size of the wound has a direct correlation with the metabolic and endocrine response to surgical trauma. The greater the cutting of fascia, muscle and nerve the higher the catecholamine and catabolic response of the body to surgical trauma.

A potential working space during video-laparoscopic abdominal procedures in children is established with the help of a carbon dioxide pneumoperitoneum. The most popular technique used in children for developing a pneumoperitoneum is the open (Hasson) technique, usually in children less than two years of age. Closed or percutaneous (Veress

needle) technique is mostly practice in older children and adolescents. Insufflation by either technique will cause an increase in intrabdominal pressure (IAP). Studies during congenital abdominal wall defects closure such as gastroschisis and omphalocele has shown that the rise in IAP may cause decrease venous return, decrease renal perfusion, low splanchnic flow, and increased airway pressures. In addition, abdominal distension causes pulmonary function abnormalities such as decreased functional residual capacity, basilar alveolar collapse, and intrapulmonary shunting of deoxygenated blood. The cardiac afterload will increase, an effect that may be magnified by hypovolemia.

Hypotension during the establishment of the pneumoperitoneum is a very feared complication. It could be the result of vascular injury, arrhythmia, insufflating too much carbon dioxide, impending heart failure, gas embolism or the development of a pneumothorax. We generally insufflate a three-kilogram baby with ten millimeters of mercury of intra-abdominal pressure and a 70-kilogram child with a maximum of fifteen mm of Hg.

Increase awareness of the intrinsic effects carbon dioxide insufflation may cause in the child abdominal cavity is necessary. Carbon dioxide is absorbed by the diaphragmatic surfaces and cause hypercapnia, respiratory acidosis, and pooling of blood in vessels with decrease cardiac output. This effect is usually controlled by the anesthesiologist increasing minute ventilation by 10% to 20% to maintain normocapnia. Increase dead space or decrease functional residual capacity caused by the Tredelenberg position and administration of volatile anesthetic agents can increment this problem. High risk children where this effect can be potentiate further are those with pre-existent cardio-respiratory conditions causing increase dead space, decrease pulmonary compliance and increase pulmonary artery pressure and resistance. It is estimated that carbon dioxide accumulates primarily in blood and alveoli due to the decrease muscular components to buffer the excess absorbed gas present in children. After the procedure, the combination of residual carbon dioxide in the diaphragmatic surface and water forms carbonic acid that upon absorbtion by the lymphatics produces referred shoulder pain. There is always a small risk of ventricular dysrhythmia with insufflation of carbon dioxide in children.

Some contraindications for performing laparoscopy during the pediatric age are: history of severe cardio-pulmonary conditions, uncorrectable coagulopathy, prematurity, distended abdomen with air or ascites, and multiple abdominal scars from previous operative procedures.

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B. Laparoscopic Cholecystectomy

Laparoscopic Cholecystectomy (LC) has become the procedure of choice for the removal of the disease gallbladder of children. The benefit of this procedure is obvious: safe, effective, and well tolerated. It produces a short hospital stay, early return to activity and reduced hospital bills. Several technical differences between the pediatric and adult patient are: lower intrabdominal 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, i.e., bile duct injury or leak. Three 5 mm ports and one 10-mm umbilical port are 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 using a Kumar clamp is advised by some workers to avoid iatrogenic common bile duct (CBD) injuries during dissection due to anomalous anatomy, and the best method to detect CBD stones. Treatment of CBD stones may consist of:

1- endoscopic sphincterotomy followed by LC,
2- open (conventional) 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.

San Pablo Medical Center performed 4439 cholecystectomies from January 1990 to July 1995; 83 (1.8%) of them in children. LC was found superior to the open conventional procedure reducing the operating time, length of stay, diet resumption, and use of pain medication. The child is more pleased with his cosmetic results and activities are more promptly established. We also found that CBD stones can be managed safely with simultaneous endoscopic papillotomy and costs of LC are further reduced employing re-usable equipment and selective cholangiographic indications.

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C. Laparoscopic Appendectomy

Semm, a gynecologist, is credited with inventing laparoscopic appendectomy in 1982. With the arrival of video-endoscopic procedures the role of laparoscopic appendectomy in the management of acute appendicitis in children has been studied and compared with the conventional open appendectomy. General advantages of laparoscopic appendectomy identified are: ease and rapid localization of the appendix, ability to explore and lavage the entire abdominal cavity, decrease incidence of wound infection, less cutaneous scarring, more pleasing cosmetically, and a rapid return of intestinal function and full activity. There is certainly some advantage in doing laparoscopic appendectomy in the obese child, teenage female with unclear etiology of symptoms, for athletes, children with chronic right lower quadrant abdominal pain, and cases requiring interval appendectomy. Disadvantages are: expensive instrumentation, time-consuming and tedious credentialing, and the major benefit is in the postop period.

Analyzing the results of several series that compare laparoscopic vs. conventional appendectomy in the management of acute appendicitis we can conclude that laparoscopy produces no difference with open appendectomy in respect to operating room complications and postoperative morbidity, has a longer operating and anesthesia time, higher hospital costs, a shorter length of stay, less postop pain, less pain medication requirement, and shorter convalescence. One series warned that complicated cases of appendicitis done by laparoscopy could increase the postoperative infectious rate requiring readmission. Otherwise, they all favored laparoscopic appendectomy in the management of appendicitis.

Still, unresolved issues in my mind are: Does laparoscopic appendectomy reduce postoperative adhesions? , Is it necessary to remove a normal looking appendix during a negative diagnostic laparoscopy performed for acute abdominal pain? , Will the increase intrabdominal pressure alter the diaphragmatic lymphatic translocation of bacteria favoring higher septic rates in complicated cases? Experimental evidence in animal models favors higher rates of systemic sepsis after sequential development of pneumoperitoneum.

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D. Laparoscopy for the Undescended Testis

The undescended testis identified in 0.28% of males can be palpable (80%) or non-palpable (20%). It is difficult to determine either location or absence of the non-palpable undescended testis by clinical examination. Imaging studies (Ultrasound, CT Scan, Magnetic resonance, gonadal venography) are not reliable in proving its absence. Diagnostic laparoscopy is reliable in finding the non-palpable undescended testis or proving its absence. Furthermore it can be combine to provide surgical management. After reviewing several series, with non-palpable undescended testes managed by laparoscopy the following three findings were identified:

1- The testis is present; in either an intra-abdominal (38%) or inguinal position (12%). Intrabdominal testes can be managed by first stage laparoscopic internal spermatic vessel clipping and cutting (Stephen-Fowler's), followed by second stage vas-based standard orchiopexy six to nine months later. Inguinal testes are managed by standard inguinal orchiopexy.

2- The testis is absent (vanishing testicular syndrome) as proven by blind ending vas and testicular vessels (36%). These children are spare an exploration. If the vas and vessels exit the internal ring, inguinal exploration is indicated to remove any testicular remnant as histologic evidence, although I have found useful removing the testicular remnant by the laparoscopic approach. The presence of a patent processus vaginalis may suggest a distal viable testis.

3- The testis is hypoplastic, atretic, or atrophic (26%), in which case is removed laparoscopically.

Exact anatomical localization of the testis by laparoscopy simplifies accurate planning of operative repair; therefore, is an effective and safe adjunct in the management of the cryptorchid testis.

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E. Groin Laparoscopy

The issue of contralateral exploration in the pediatric inguinal hernia patient has been hotly debated. Proponents of routine contralateral exploration cite the high percentage of contralateral hernia a/o potential hernia found at exploration, the avoidance of the cost of another hospitalization, psychological trauma and anxiety to the child and parents over a second operation, and the added risk of anesthesia of a second procedure. Most pediatrics surgeons habitually explore the contralateral side. They disagree in opinions about exploration depending upon the primary site of inguinal hernia, age, sex and the use of herniography or some intra-operative technique to check the contralateral side.

Recently the use of groin laparoscopy permits visualization of the contralateral side. The technique consists of opening the hernial sac, introducing a 5.5-mm reusable port, establishing a pneumoperitoneum, and viewing with an angle laparoscope the contralateral internal inguinal ring to decide the existence of a hernia, which is repaired if present. Requires no additional incision, avoids risk of vas deferens injury in boys, is rapid, safe and reliable for evaluating the opposite groin in the pediatric patient with unilateralinguinal hernia. Children less than two years of age have a higher yield of positive contralateral findings.

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F. Laparoscopic Splenectomy

Laparoscopic splenectomy is another safe and technically feasible video-endoscopic procedures in children. Indications are usually hematological disorders such as Idiopathic thrombocytopenic purpura, spherocytosis, and Hodgkin's staging. Technical considerations of the procedure are based on anatomical facts such as the variability in the splenic blood supply, the ligaments anchoring the organ and the size of the diseased spleen. Generally the avascular splenophrenic and colic ligaments are cauterized, the short gastric and hilar

vessels are individually ligated with metallic clips or gastrointestinal staplers, and the spleen is placed in a plastic bag, fracture or morzelized until it is removed through the navel.

Comparing the laparoscopic procedure with the conventional splenectomy, the advantages are: improved exposure, decreased pain, improved pulmonary function, shortened hospitalization, more rapid return to normal activities and excellent cosmetic appearance. Disadvantages are longer operating time, higher costs and the need to open 5-20% of cases due to technical uncontrolled hemorrhage, such as bleeding from the splenic artery.

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G. Laparoscopic Fundoplication

Fundoplication for the management of symptomatic gastroesophageal reflux (GER) is another procedure that has evolved recently taking advantage of minimally invasive technique. Indications for performing either the open or laparoscopic fundoplication is the same, namely: life threatening GER (asthma, cyanotic spells), chronic aspiration syndromes, chronic vomiting with failure to thrive, and reflux induced esophageal stricture. Studies comparing the open versus the laparoscopic technique in the pediatric age have found a reduced mean hospital and postoperative stay with laparoscopy. The lap procedure seems similar to the open regarding efficacy and complication rates. Costs are not excessive, they are even lower if we take into consideration the shorter length of stay. Lower rate of adhesions, pulmonary and wound complications are another benefit of the lap technique suggested. Percutaneous laparoscopic gastrostomy can be done concomitantly for those neurologically impeded children refer with feeding problems and GER.

Whether to do a complete (Nissen) or partial (Toupee, Thal, or Boix-Ochoa) wrap relies on the experience of the surgeon with the open procedure. He should continue to do whatever procedure he used to perform using open surgery. Long-terms results of complications or recurrence of GER after laparoscopic fundoplication are still pending publication.

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XI. SUGGESTED READING

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