بسم الله الرحمن الرحيم

PEDIATRICS

PEDIATRICS SURGICAL CONDITIONS

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GASTROESOPHAGEAL REFLUX (GER)

Etiology & epidemiology

GER is defined as the effortless retrograde movement of gastric contents upward into the esophagus or ororpharynx. In infancy, GER is not always an abnormality. Physiologic GER ("spitting up") is normal in infants younger than 8 to 12 months old. Nearly half of all infants are reported to spit up at 2 months of age. Infants who regurgitate stomach contents met the criteria for physiologic GER so long as they maintain adequate nutrition & have no signs of respiratory or peptic complications. Factors involved in GER include liquid diet; horizontal body position; short, narrow esophagus; small, noncompliant stomach; frequent, relatively large volume feedings; & an immature lower esophageal sphincter (LES). As infants grow, they spend more time upright, eat more solid foods, develop a longer & larger diameter esophagus, have a larger & more complaint stomach, & experience lower caloric needs per unit of body weight. As a result, most infants stop spitting up by 9 to 12 months of age.

Clinical manifestations

The presence of GER is easy to observe in infants who spits up. In some cases, especially in older children, the refluxate is kept down by reswallowing, but GER may be suspected by associated symptoms, such as heartburn, cough, dysphagia (trouble swallowing), wheezing, aspiration pneumonia, hoarse voice, failure to thrive, & recurrent otitis media or sinusitis. In severe cases of esophagitis, there may be laboratory evidence of anemia & hypoalbuminemia secondary to esophageal bleeding & inflammation.

Pathologic GER is diagnosed after 18 months of age or if there are complications, such as

esophagitis, respiratory symptoms, or failure to thrive, in younger infants. In older children, normal protective mechanisms against GER include antegrade esophageal motility, tonic contraction of the LES, & the geometry of the gastroesophageal junction. Abnormalities that cause GER in older children & adults include reduced tone of the LES, transient relaxation of the LES, esophagitis (which impairs esophageal motility), increased intra-abdominal pressure, cough, respiratory difficulty (asthma or cystic fibrosis), & hiatal hernia.
When esophagitis develops as a result of acid reflux, esophageal motility & LES function are impaired further, creating a cycle of reflux of esophageal injury.

Diagnosis

A clinical diagnosis is often sufficient in children with classic frequent effortless vomiting or regurgitation without evidence of GIT obstruction & no complications. Diagnostic studies are indicated if there are persistent symptoms or complications or if other symptoms suggests the possibility of GER in the absence of regurgitation. A child with recurrent pneumonia, chronic cough, or apneic spells without overt emesis may have occult GER. A barium upper GI series helps to rule out gastric outlet obstruction, malrotation, or other anatomic contributors to GER. Because of the brief nature of the examination, a negative barium study does not rule out GER (50% sensitivity). The most reliable test for the diagnosis of GER is the "24-hour esophageal pH probe monitoring" by using a pH electrode placed transnasally into the distal esophagus, with continuous recording of esophageal pH. Data typically are gathered for 24 hours, following which the number & temporal pattern of acid reflux events are analyzed. A similar test, which does not require the presence of acid in the stomach, is esophageal impedance monitoring, which records the migration of electrolyte-rich gastric fluid in the esophagus. Endoscopy is useful to rule out esophagitis, esophageal stricture, & anatomic abnormalties (suggestive but not diagnostic for reflux).

Treatment

In otherwise healthy young infants ("well-nourished, happy spitters"), no treatment is necessary, other than a towel on the shoulder of the caretaker. Other conservative therapies include thickened feedings (alleviate GER 40-50% of infants, especially with elevation of the head 30° in prone position), small & more frequent feedings, avoidance of tobacco smoke &

alcohol, & positional therapy (elevation of the head). For infants with complications of GER, pharmacologic therapy with a proton-pump inhibitor should be offered. Lesser benefits are obtained with H2 receptor antagonists. Prokinetic drugs, such as metocloporomide, occasionally may be helpful by enhancing gastric emptying & increasing LES tone, but are seldom very effective. When severe symptoms & complications (as refractory esophagitis, apnea, & failure to thrive) persist despite medication, or if life-threatening aspiration is present, surgical intervention may be required. Fundoplication procedures, such as the Nissen operation, are designed to enhance the antireflux anatomy of the LES. In children with a severe neurologic defect who cannot tolerate oral or gastric tube feedings, placement of a feeding jejunostomy should be considered as an alternative to fundoplication.

PYLORIC STENOSIS

Etiology & epidemiology

Pyloric stenosis is an acquired condition caused by hypertrophy & spasm of the pyloric muscle, resulting in gastric outlet obstruction. It occurs in 6 to 8 per 1000 live births & has a 5:1 male predominance & is more common in first-borne children. Its cause is unknown, but it seems that a deficiency in inhibitory neuronal signals, mediated by nitric oxide, is likely.

Clinical manifestations

Infants with pyloric stenosis typically begin vomiting during the first month of life, but onset of symptoms may be delayed. The emesis becomes increasingly more frequent & forceful as time passes. Vomiting in pyloric stenosis differs from spitting up because of its extremely forceful & often projectile nature. The vomited material is of gastric content (milk) & never contains bile because the gastric outlet obstruction is proximal to the duodenum. This feature differentiates pyloric stenosis from most other obstructive lesions of early childhood. It may become brownish or visibly bloodstained due either to an accompanying gastritis or to rupture of capillaries in the gastric mucosa from frequent vomiting. Affected infants are ravenously hungry early in the course of illness, but become more lethargic with increasing malnutrition & dehydration. The stomach becomes massively enlarged with retained food &

secretions, & gastric peristaltic waves are often visible in the left upper quadrant traveling towards the right. A hypertrophied pylorus (the “olive”) may be palpated just to the right of the midline, in the right hypochondrium. This is called “feeding test” which is essential to confirm the diagnosis. As the illness progresses, very little of each feeding is able to pass through the pylorus, & the child becomes progressively thinner & more dehydrated.

Diagnosis

Repetitive vomiting of purely gastric contents results in loss of hydrochloric acid; the classic laboratory finding is a hypochloremic hypokalemic metabolic alkalosis with elevated BUN secondary to dehydration. Jaundice with unconjugated hypebilirubinemia also occurs. Plain abdominal x-rays typically show a huge stomach & diminished or absent gas in the intestine. Ultrasound examination of the pylorus shows marked elongation & thickening of the pylorus. A barium upper GIT series also may be obtained whenever doubt about the diagnosis exists; this shows a “string sign” caused by barium moving through an elongated , constricted pyloric channel.

Treatment

Treatment of pyloric stenosis includes IV fluid & electrolyte resuscitation followed by surgical pyloromyotomy. Before surgery, dehydration & hypochloremic alkalosis must be corrected , generally with an initial normal saline fluid bolus followed by infusions of half-normal saline containing 5% dextrose & potassium chloride when urine output is observed. For pyloromyotomy, a small incision is made, usually directly over the pylorus or at the umbilicus, & the pyloric muscle is incised longitudinally to release the constriction. Care is taken not to cut into the mucosa itself.

DUODENAL ATRESIA

Etiology

Duodenal atresia is thought to arise from failure to recanalize the lumen after the solid phase of intestinal development during the 4^{th} & 5^{th} wk of gestation. The incidence of duodenal atresia is 1 in 10000 births & accounts for 25-40% of all intestinal atresias. Half the

patients are born prematurely. Duodenal atresia may take several forms, of which the membranous form is most common with obstruction occurring distal to the ampulla of Vater in the majority of patients. Down syndrome occurs in 20-30% of patients with duodenal atresia. Other congenital anomalies that are associated with duodenal atresia include malrotation (20%), esophageal atresia (10-20%), congenital heart disease (10-15%), & anorectal & renal anomalies (5%).

Clinical manifestation

The hallmark of duodenal atresia is bilious vomiting with abdominal distention, which is usually noted in the 1st day of life. Peristaltic waves may be visualized early in the disease process. A history of polyhydramnios is present in 50% of the pregnancies & is caused by a failure of absorption of amniotic fluid in the distal intestine. Jaundice is present in one third of the infants.

Diagnosis

The diagnosis is suggested by the presence of a "double-bubble sign" on plain abdominal radiographs. The appearance is caused by a distended & gas-filled stomach & proximal duodenum. Contrast studies are usually not necessary & may be associated with aspiration if attempted, but these studies may occasionally be needed to exclude malrotation & volvulus because intestinal infarction may occur within 6-12 hr if the volvulus is not relieved. Prenatal diagnosis of duodenal atresia is readily detected by fetal ultrasonography.

Treatment

The initial treatment of infants with duodenal atresia includes nasogastric or orogastric decompression with IV fluid replacement. Echocardiogram & radiology of the chest & spine should be performed to evaluate for associated anomalies. Approximately one third of infants with duodenal atresia have associated life-threatening congenital anomalies. Definitive surgical correction of duodenal atresia is usually postponed to evaluate & treat these life-threatening anomalies. The usual surgical repair of duodenal atresia is duodenoduodenostomy. A gastrostomy tube may be placed to drain the stomach & protect the airway. Intravenous

nutritional support or a trnsanastomotic jejunal tube is needed until infants start to feed orally. The prognosis is primarily dependent on the presence of associated anomalies.

INTUSSUSCEPTION

Etiology & epidemiology

Intussusception is the “telescoping” of a segment of proximal bowel (the intussusceptum) into downstream bowel (the intussuscipiens). Most cases occur in infants 1 to 2 years old. In infants younger than 2 years old, nearly all cases are idiopathic. Viral-induced lymphoid hyperplasia may produce a lead point in these children. In older children, the proportion of intussusception caused by a pathologic lead point increases. In young children, ileocolonic intussusception is common; the ileum invaginates into the colon, beginning at or near the ileocecal valve. When pathologic lead points are present, the intussesception may be ileoileal, jejunojejunal. There is a slight male predominance.

Clinical manifestations

An infant with intussusception has sudden onset of crampy abdominal pain; the infant’s knees draw up, & the infant cries out & exhibits pallor with a colicky pattern occurring every 15 to 20 minutes. Feeding are refused. As the intussusception progresses, & obstruction becomes prolonged, bilious vomiting becomes prominent, & the dilated, fatigued intestine generates less pressure & less pain. As the intussuscepted bowel moves further & further into the downstream intestine by its native motility, the mesentery is pulled with it & becomes stretched & compressed. The venous outflow from the intussusceptum is obstructed, leading to edema, weeping of fluid, & congestion with bleeding. Third space fluid losses & “currant jelly” stools result. Another unexpected feature of intussusception is lethargy. Between episodes of pain, the infant is glassy-eyed & groggy & appears to have been sedated. A sausage-shaped mass caused by the swollen, intussuscepted bowel may be palpated in the right upper quadrant or epigastrium.

Diagnosis

The diagnosis depends on the direct demonstration of bowel-within-bowel. A simple &

direct way of showing this is by abdominal ultrasound. If the ultrasound is positive, or if good visualization has not been achieved, a pneumatic or contrast enema under fluoroscopy is indicated. This is the most direct & potentially useful way to show & treat intussusception. Air & barium can show the intussusception quickly &, when administered with controlled pressure, usually can reduce it completely. The success rate of pneumatic reduction is probably a bit higher than hydrostatic reduction with barium & approaches 90% if done when symptoms have been present for less than 24 hours. The pneumatic enema has the additional advantage over barium of not preventing subsequent radiologic studies, such as upper GI series or CT scan. Nonoperative reduction should not be attempted if the patient is unstable or has evidence of perforation or peritonitis.

Treatment

Therapy must begin with placement of an IV catheter & NG tube. Before radiologic intervention is attempted, the child must have adequate fluid resuscitation to correct the often severe dehydration caused by vomiting & third space losses. Ultrasound may be performed before the fluid resuscitation is complete. Surgical consultation should be obtained early because the surgeon often prefers to be present during nonoperative reduction. If pneumatic or hydrostatic reduction is successful, the child should be admitted to the hospital for overnight observation of possible recurrence (risk is 5% to 10%). If reduction is not complete, emergency surgery is required. The surgeon attempts gentle manual reduction, but may need to resect the involved bowel after failed radiologic reduction because of severe edema, perforation, a pathologic lead point (polyp, Meckel diverticulum), or necrosis.

HIRSCHSPRUNG DISEASE

Etiology & pathology

Hirschsprung disease is a motility defect caused by failure of ganglion cell precursors to migrate into the distal bowel during fetal life. The aganglionic distal segment does not exhibit normal motility & is functionally obstructed secondary to spasm. In 75% of cases, the involved segment is limited to the rectosegmoid, but total colonic involvement is seen in 8% . Rarely, long segment of small bowel also are aganglionic. “Ultrashort ” segment involves

only a few centimeters of distal rectum.

Clinical manifestations

Hirschsprung disease typically presents in the newborn period with failure to pass meconium by 24 hours of age. About 95% normal infants pass stool spontaneously by this age; 95% of infants with Hirschsprung disease do not. Symptoms of distal bowel obstruction occur with distention & bilious vomiting. If the diagnosis is not made quickly, enterocolitis can result, associated with a high rate of mortality.

Diagnosis

Diagnosis is based on examination & one or more diagnostic studies. Abdominal distention is present in most cases. Digital rectal examination reveals an empty rectum that clenches around the examiner's finger, giving the impression of an elongated sphincter. When the finger is withdrawn, a powerful gush of retained stool is often expelled. A deep rectal biopsy specimen obtained surgically or by using a suction biopsy instrument is required for diagnosis. When no ganglion cells are shown in the submucosal plexus, accompanied by nerve trunk hyperplasia, the diagnosis is certain. Barium enema & anorectal manometry may be used before biopsy; but false-negative & false-positive results can occur.

Treatment

Treatment is surgical. When the bowel is markedly distended or inflamed, an initial colostomy usually is performed above the aganglionic segment, followed weeks later by one of several definitive repair procedures. The transanal pull-through excises the aganglionic bowel & creates a primary colorectal anastomosis without laprotomy. This procedure can be considered in patients with uncomplicated involvement limited to the rectosigmoid region.

HYPOSPADIAS

Hypospadias occurs in approximately 1-2 out of every 1000 live births. The urethral meatus is located ventrally & proximal to its normal position, an abnormality resulting from a failure of the urethral folds to fuse completely over the urethral groove. The ventral foreskin is also lacking, & the dorsal portion gives the appearance of hood. Severe hypospadias with undedescended testes is a variant of ambiguous genitalia, & the underlying etiologies include congenital adrenal hyperplasia with musculinazation of females or an androgen insensitivity syndrome. Other urinary tract anomalies do not occur often in association with hypospadias.. Hypospadias may occur alone, but it is usually associated with a chordee (a fixed ventral curvature of the penile shaft). Rarely, the urethra opens onto the perineum. In this circumstances, the chordee is extreme, & the scrotum is bifid and sometimes extends to the dorsal base of the penis. When assessing hypospadias , it is useful to describe where the urethral meatus appears (i.e., glandular, distal shaft, proximal shaft, or perineal) & also the degree & location of chordee. The mildest & most common form of hypospadias is distal hypospadias which occurs in the subcoronal or glandular area in about 80-85% of cases. About 10-15% of cases occur in the penile shaft. Only 5-10% occur in the severe penoscrotal or perineal location. A number of associated genitourinary abnormalities have been described with hypospadias, including meatal stenosis, inguinal hernias, and undescended testes (10%). The incidence of these abnormalties rises sharply with the more severe degree of hypospadias. In the mild forms, which account for most cases, radiography or endoscopy of the urinary tract is unnecessary. Males with hypospadias should not be circumcised, because the foreskin may be necessary for later surgical repair which is optimally done at 6-12 months of age (before 18 months of age).

PHIMOSIS

is a narrowing of the distal foreskin, which prevents its retraction over the glans of the penis. In newborns, retraction is difficult because of normal adhesions that gradually self-resolve. It is rarely symptomatic, & parents should be reassured that loosening of the

prepuce usually occurs during puberty. Treatment, if needed, is the application of topical steroids. If the narrowing is severe, gentle stretching often results in improvement. Chronic inflammation or scarring can cause true phimosis with persistent narrowing & may require circumcision.

PARAPHIMOSIS

is incarceration of a retracted foreskin behind the glans. It occurs when the retracted foreskin is not repositioned. Progressive edema results, which, if uncorrected, can lead to ischemic breakdown. Local anesthesia, ice, & manual reduction usually correct the problem, but if these are unsuccessful, surgical reduction is necessary.