Pyloric Stenosis

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Hypertrophic pyloric stenosis is the most common cause of metabolic alkalosis in infancy. The incidence of pyloric stenosis ranges from 1 in 250 to 1,000, depending on geographic location, and is reportedly on the rise. Boys are affected four to eight times more often than girls, and there is a Caucasian predilection. In addition, sometimes there is a clustering in families. Pyloric stenosis is the most common reason for abdominal surgery in the first 6 months of life.

Although the precise etiology of pyloric stenosis remains unknown, there is some evidence that it may be an acquired condition, rather than a congenital disorder, as previously thought. In this condition, gastric outlet obstruction results from hypertrophy of the pyloric muscle, edema of the pyloric canal, and spasm of the antropyloric muscle, which leads to vomiting, dehydration, and metabolic alkalosis.

Clinical manifestations of pyloric stenosis begin at a mean age of 3 weeks after birth, but they may occur at any time between birth and 5 months of age. The onset of clinical symptoms is heralded by regurgitation of feeds and progresses to the classic nonbilious vomiting, which often is projectile. Physical examination may reveal a dehydrated or wasted infant who is an avid sucker. An olive-like pyloric mass may be palpated in the upper abdomen in up to 90% of patients. When palpated, a pyloric mass is pathognomonic for hypertrophic pyloric stenosis. To enhance the sensitivity of the physical examination, it may be helpful to empty the stomach by using a nasogastric or orogastric tube and palpate the abdomen with the infant held in a prone position; comforting the infant to prevent crying also helps.

Imaging procedures are reserved for instances when pyloric stenosis is highly suspected, but the infant does not have a palpable pyloric mass. Although upper gastrointestinal contrast radiography was used in the past, ultrasonography is the current study of choice. On ultrasonography, a pyloric muscle thickness of greater than 4 mm and a pyloric length of greater than 16 mm have a diagnostic sensitivity and specificity of 89% and 100%, respectively.

Patients who have pyloric stenosis typically present with hypochloremic metabolic alkalosis. Although the serum potassium level may be normal or low, there often is total body potassium depletion. The observed metabolic alkalosis is a result of two related but independent processes: loss of acid and retention of bicarbonate. Initially, vomiting of gastric contents results in excessive loss of hydrogen chloride and potassium chloride, which leads to a metabolic alkalosis. Under normal circumstances, carbonic acid (H_2CO_3) in the villi of the stomach dissociates into H^+ and HCO_3^-. The hydrogen ions cross the luminal membrane of the enterocyte and enter the stomach, from where they are transported to the duodenum. The entry of acid into the duodenum stimulates the secretion of an equal amount of pancreatic HCO_3^-. This normal stimulus is absent in pyloric stenosis because of the mechanical obstruction, and the diminished secretion of pancreatic bicarbonate into the gastrointestinal tract contributes further to the metabolic alkalosis created by stomach acid lost through vomiting. Moreover, as intravascular volume decreases with dehydration, the concentration of HCO_3^- in the plasma increases, resulting in what is known as a contraction alkalosis. The loss of potassium during vomiting also may contribute to the development of metabolic alkalosis. As plasma potassium concentration falls, potassium moves out of the cells to replete extracellular stores, and electroneutrality is maintained by hydrogen moving into the cells. This shift causes paradoxic extracellular alkalosis with intracellular acidosis.

The kidneys also play a role in the metabolic alkalosis seen with pyloric stenosis. To maintain volume in the face of fluid losses from vomiting, the kidneys reabsorb bicarbonate in the distal tubules despite alkalosis. If the excess bicarbonate is excreted in the urine, it obligates sodium loss to maintain electroneutrality, and because water follows sodium, further volume is lost. In addition, decreased chloride delivery to the macula densa of the kidneys results in renin release and secondary hyperaldosteronism, leading to increased distal hydrogen secretion and the paradoxic finding of an acid urine in the presence of alkalemia. Finally, in response to hypokalemia, the distal tubules of the kidneys reabsorb potassium in exchange for hydrogen, resulting in further loss of acid.

Management of pyloric stenosis includes meticulous correction of fluids and electrolytes, followed by corrective surgery. Appropriate medical management reverses the volume contraction and metabolic derangements. Pyloromyotomy, which is curative for gastric outlet obstruction, involves incision of the serosa and the underlying mass through to the mucosal layer. The prognosis following surgery is excellent, but it depends on a prompt diagnosis of pyloric stenosis prior to the development of severe metabolic derangements.

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Comment: The pediatric literature frequently comments that pyloric stenosis is more common among first-born boys. The evidence, I think, is clear that far more boys are affected than girls, but what about this first-born business? We would expect to find more affected first-borns for every disease, not just pyloric stenosis, simply because there are more first-borns than second-borns than third-borns, and so on. Is anyone aware of data that suggest pyloric stenosis really occurs at a higher rate among first- than later-borns?

We also should note that jaundice is a fairly common finding among infants who have pyloric stenosis. They frequently have a modest indirect hyperbilirubinemia, possibly from inadequate glucuronyltransferase activity, that resolves with the resumption of normal feeding after surgery.

Henry M. Adam, MD
Editor, In Brief

IN BRIEF

Delayed Puberty

The Biological Aspects of Puberty. Kulin HE, Muller J. *Pediatr Rev.* 1996;17:75–86

Investigation of Delayed Puberty. Albanese A, Stanhope R. *Clin Endocrinol.* 1995;43: 105–11

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Early Morning Plasma Testosterone is an Accurate Predictor of Imminent Pubertal Development in Prepubertal Boys. Wu FCW, Brown DC, Butler GE, Stirling HF, Kelnar CJH. *J Clin Endocrinol Metab.* 1990;70:26–31

Delayed puberty describes a condition in youth who fail to demonstrate signs of sexual development by a certain chronologic age. As with any developmental characteristic that occurs along a spectrum, suspicion for "potential pathology" is set arbitrarily. Age of puberty may be affected by both physiologic and socioeconomic factors and can vary widely. Delayed puberty is present when no signs of sexual maturity (Sexual Maturity Rating Stage I) are apparent. In the United States, most authors use a cutoff of 2 standard deviations from the mean with corresponding ages of 13 years in girls and 14 years in boys. Using these criteria, 2.5% of the normal population will be classified as "delayed." Once the child's chronologic age surpasses the age standard, the physician's job becomes to determine whether delayed puberty is based on constitutional factors or an underlying organic disease.

Because the list of diseases that can cause delayed puberty is long, the clinician must be vigilant in the search for any clues that might suggest an underlying primary disorder. These include physical findings or historical details that point to any systemic or chronic disorders. Diseases as diverse as inflammatory bowel disease, thyroid disease, chronic renal or heart disease, or even asthma can cause pubertal delay. The neurologic component of the physical examination is particularly important to elicit any findings that might indicate a central nervous system tumor. Additionally, family history or physical abnormalities that suggest genetic syndromes such as Kallmann, Prader-Willi, Noonan, or Turner syndrome should be considered initially.

Many authors recommend initial assessment of gonadotropin levels (usually urine or serum luteinizing hormone [LH] or follicle-stimulating hormone [FSH]) in a patient who presents with no signs of sexual maturity as defined previously and no physical or historical clues to the delay. If the levels are high (hypergonadotropic hypogonadism), primary gonadal failure should be investigated with the assistance of a pediatric endocrinologist. If they are low, as is the case in most cases of delayed puberty seen by the general clinician, either constitutional delay or hypogonadotropic hypogonadism (from many potential causes) is the diagnosis. Although such findings may be reassuring from the perspective of an overall favorable clinical outcome for most patients who present with delayed puberty, the sobering fact remains that currently there are no endocrinologic tests in wide clinical use that can discriminate between these two causes.

Recent studies offer hope that in the future it will be possible to predict impending sexual development routinely by using more sensitive immunoassays. In boys, for example, an 8:00 AM serum testosterone level appears to offer a relatively sensitive and specific prediction of future sexual development. In one investigation, a level greater than 0.7 nmol/L predicted an increase in testicular size to more than 4 mL within 1 year in 77% and 15 months in 100% of boys; only 12.5% of boys who had levels less than 0.7 nmol/L progressed to a testicular size of greater than 4 mL within 1 year.

Currently, however, it seems reasonable to adopt a conservative strategy of watchful waiting in patients who have delayed puberty, low LH/FSH levels, and no significant findings on physical examination and family history. Clues to patients who might have constitutional delay include slow growth in childhood with normal growth velocity, thin body habitus, and a family history of constitutional delay ("late bloomers"). Those who have hypogonadotropic delay usually grow normally in childhood but do not manifest the usual growth spurt of adolescence.

If development does not progress within 6 months or other abnormal findings become apparent, further step-wise investigation with the assistance of a pediatric endocrinologist is warranted. This may include radiographic imaging to determine bone age (which is more closely correlated to sexual development

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