CE 3.5 HOURS Continuing Education

Prune Belly Syndrome

A Focused Physical Assessment

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ABSTRACT

Prune belly syndrome, a rare congenital anomaly, exists almost exclusively in males and consists of genital and urinary abnormalities with partial or complete absence of abdominal wall musculature. The syndrome, caused by urethral obstruction early in development, is the result of massive bladder distention and urinary ascites, leading to degeneration of the abdominal wall musculature and failure of testicular descent. The impaired elimination of urine from the bladder leads to oligohydramnios, pulmonary hypoplasia, and Potter's facies. The syndrome has a broad spectrum of affected anatomy with different levels of severity. The exact etiology of prune belly syndrome is unknown, although several embryologic theories attempt to explain the anomaly. With advances in prenatal screening techniques, the diagnosis and possible treatment of prune belly syndrome can occur before birth, although controversy exists on the proper management of prune belly syndrome. This article explores the theories behind the pathophysiology and embryology of prune belly syndrome and its multisystemic effects on the newborn infant. Specific attention is paid to presentation, clinical features, head-to-toe physical assessment, family support, and nursing care of infants with prune belly syndrome.

KEY WORDS: congenital absence of abdominal musculature, deficiency of abdominal musculature, Eagle-Barrett syndrome, prune belly syndrome, triad syndrome

The most common cause of perinatal and infant mortality in the United States is congenital anomalies. Advances in prenatal screening techniques have increased earlier diagnosis of congenital anomalies. Obstructive uropathy, an obstruction in the urinary tract, includes obstructions such as ureteropelvic junction (UPJ), ureterovesical junction (UVJ), posterior urethral valves (PUV), vesicoureteral reflux (VUR), urethral atresia/stricture, duplicating collecting system, multicystic dysplastic kidney, and prune belly syndrome. Obstructions can occur at different levels of the urinary tract, resulting in hydronephrosis (dilation of the urinary tract), and ultimately can cause renal parenchymal damage. The level of severity depends on the duration and area of obstruction.[1] This article focuses on prune belly syndrome and discusses the underlying obstruction.

The underlying etiology of the urinary tract obstruction and the degree of pulmonary hypoplasia influence the survival and outcome of infants with prune belly syndrome. Management of these infants depends on a functioning urinary tract system and the extent of associated symptoms. Understanding the pathophysiology provides a foundation for nursing care of infants with prune belly syndrome and provides accurate information to parents for making decisions about treatment of their infant. Although rare, anomalies such as prune belly syndrome are often difficult to manage because of their complexity.[2] Prune belly syndrome, also known as Eagle-Barrett syndrome, comprises a triad of anomalies that include abdominal wall flaccidity, urologic abnormalities, and bilateral cryptorchidism. The characteristic wrinkled, prune-like abdomen of the infant gives the syndrome its name. In addition, cardiovascular, respiratory, orthopedic, and gastrointestinal anomalies can accompany prune belly syndrome.[3]

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INCIDENCE

The incidence of prune belly syndrome is between 1 in 29,000 to 1 in 40,000 live male births.[1] Males are affected 20 times more often than females, with approximately 3% of all cases occurring in females. Infants born to younger mothers are at greater risk for having prune belly syndrome.[4,5] The incidence of prune belly syndrome in twins is 4 times greater than that found in single births, at 12.2 per 100,000 live births.[6]

ETIOLOGY OF PRUNE BELLY SYNDROME

The exact etiology of prune belly syndrome is unknown; however, several theories exist to explain the syndrome.[7] To understand the theories behind prune belly syndrome, an understanding of normal genitourinary development is essential. Development of the urogenital system is divided into the urinary and reproductive systems. During the third week of embryonic development, a process called *gastrulation* occurs. Gastrulation is the beginning of embryonic body formation by the development of 3 specific germ layers (ectoderm, mesoderm, and endoderm). These 3 germ layers will then give rise to specific organs and tissues. One of the germ layers, the mesoderm, will give rise to urinary and reproductive organs. The urinary system begins development at approximately 3 weeks' gestation and occurs before the genital system is evident, although, embryological development between these 2 systems is closely related. Four components comprise the urinary system:[8,9]

- kidneys–primary role is urine excretion
- ureters–primary role is transport of urine from the kidneys to the bladder
- bladder–primary role is storage of urine until elimination
- urethra–primary role is to carry urine from the bladder to be eliminated

The kidneys develop first through a series of 3 consecutive steps. The first kidneys, called pronephroi, appear between weeks 3 and 4 of gestation. They are essentially nonfunctioning kidneys represented by a cluster of cells in the neck region and usually disappear by the end of the fourth week of gestation. Next, mesonephroi develop around the fourth week of gestation and serve as a temporary excretion organ until the permanent kidneys develop. The mesonephroi begin to disappear by 8 weeks of gestation. Finally, the metanephroi develop by the fifth week of gestation and become the functioning permanent kidneys by approximately 9 weeks' gestation. Initially, the kidneys are located in the pelvis; as the fetus grows, they gradually ascend to their proper location in the abdomen.[8]

The bladder and urethra develop following a division of the cloaca by the urorectal septum between 5 to 6 weeks of gestation. The anterior part becomes the primitive urogenital sinus, and the posterior part becomes the anorectal canal. The urogenital sinus has 3 parts. The vesicle part will form the bladder; the pelvic part will form the urethra in females and the urethra in the bladder neck and prostatic urethra in males; and the phallic part will eventually grow toward the genital tubercle.[8] In infants, the urinary bladder is located in the abdomen, entering the pelvis at about 6 years of age.[8]

An important factor in fetal development and growth is amniotic fluid. At first, amniotic fluid comes from maternal tissue and fluid that moves across the amniochorionic membrane from decidua parietalis through diffusion. Soon after, fluid moves through the chorionic plate from the blood of the placenta also by diffusion. Amniotic fluid is also comprised of fluid secreted by the fetal respiratory tract, known as fetal lung fluid. Urine production begins between 9 and 10 weeks of gestation and will contribute to the amniotic fluid volume by 11 weeks' gestation.[8,9] At 20 weeks' gestation, fetal urine production is approximately 0.1 mL/minute and gradually increases to 1 mL/minute by 40 weeks' gestation.[10] During the first and second trimesters, electrolytes and osmolality in fetal urine reflect maternal blood. After the third trimester, sodium and chloride absorption results from a more hypotonic urine and amniotic fluid.[11] Consequently, if an obstruction or arrest occurs during renal development, urine production is low, resulting in low amniotic fluid levels (oligohydramnios). After 20 weeks' gestation, urine excretion by the kidneys is responsible for approximately 90% of amniotic fluid volume.[1]

Lack of urine production or excretion leads to oligohydramnios, a decreased amount, or a lack of, amniotic fluid. This leads to a number of findings in the infant. Fetal deformation caused by compression of the abdominal wall may cause characteristic facial features and contraction of extremities. Adequate levels of amniotic fluid are also fundamental for normal pulmonary development. Infants with prune belly syndrome who have not undergone antenatal medical interventions to divert urine may present with pulmonary hypoplasia, depending on the degree of urinary obstruction.[12]

Complications associated with prune belly syndrome originate during embryonic and fetal development. Infants with prune belly syndrome who have urethral obstructions can have a patent urachus, which allows elimination of urine from the body.[1] Normally, the urachus develops from the urogenital sinus with the superior portion of the sinus developing into the bladder and urachus and the inferior portion of the sinus developing into the urethra. During the fourth to fifth months of gestation, the urachus obliterates. If the urachus fails to obliterate, a fibrous cord that stretches from the bladder to the umbilicus exists and allows urine to escape through the umbilical cord.

Other complications that can originate during embryonic development include renal agenesis, horseshoe kidney, duplications of the urinary tract, ectopic ureter, and cystic kidney disease.[8]

One of the earliest theories regarding prune belly syndrome dates to 1903. The theory proposes that an in utero bladder obstruction leads to bladder distention and failure of the abdominal wall to develop between the sixth and eighth weeks of gestation. More current obstructive theories focus on an early urethral obstruction that causes massive bladder distention and urinary ascites. The ultimate result is degeneration of the abdominal wall musculature and prevention of testicular descent.[1] Rupture of the obstructing membrane of the urethra often occurs before birth; at birth, the urethra can range from patent to stenotic.[13] The bladder will have increased wall thickness, eventually leading to bladder dysfunction. The ureters will be dilated, tortuous, and have thickened walls. The kidneys may be dysplastic, have tubular defects, or display hydronephrosis.[1]

The most convincing theory encompasses genitourinary tract, testes, and abdominal wall musculature problems occurring as a direct result of a disturbance early in embryogenesis.[7] This theory, known as the theory of mesodermal arrest, includes an aberration of mesenchymal development between the sixth and tenth weeks of gestation, as opposed to a mechanical obstruction, as the etiology. Because of the abundance of fibrous tissue, connective tissue, collagen, and scattered smooth musculature along the urinary tract in infants with prune belly syndrome, a mesodermal etiology is highly supported because there is no evidence of obstruction in these findings.[14]

Whether prune belly syndrome has an obstructive or a mesodermal arrest etiology, the consequences of urinary flow impairment affect urinary tract development. Initially, there is dilatation of renal pelvis and calyces, then histologic damage to the renal parenchyma, which causes changes in renal function and abnormal renal growth and development. The result is tubular dilatation and apoptosis, interstitial inflammation, and fibrosis.[7,14]

Mapping of genetic disorders and malformations during the last decade has merged with mapping of the isolation of sex-influenced traits that are determined by genes.[7] Eleven cases of familial prune belly syndrome have been reported. The exact mode of inheritance of prune belly syndrome is unknown, although it has been hypothesized that a genetic basis exists. With prune belly syndrome, the most likely mode of transmission is a sex-linked inheritance.[15]

If prune belly syndrome has an autosomal recessive mode of inheritance, the syndrome would appear equally in males and females. Because most reported cases of familial prune belly syndrome have occurred in males, a sex-influenced mode of inheritance is suspected. A reported case of prune belly syndrome was found in one male with an early urethral obstruction that had a large interstitial deletion located on the long arm of chromosome 6 [del 6 (q15-q2.33)]. Twenty-eight percent of all familial cases of prune belly syndrome were female, as compared with 5% in nonfamilial forms. One model of this sex-influenced syndrome would display a male with prune belly syndrome in which one recessive allele is expressed. Consequently, it would take 2 affected alleles for a female to display prune belly syndrome.[15]

Antenatal Evaluation

With advancements in sonography equipment, a diagnosis of prune belly syndrome can occur as early as 12 weeks' gestation.[12] Fetal kidneys can be visualized on ultrasound between 10 to 12 weeks of gestation. However, the exact renal size, parenchymal architecture, collecting systems architecture, and the fullness and function of the bladder are most accurately assessed between 16 and 18 weeks of gestation. Currently, there is no concurrence on the size of normal fetal renal pelvis diameter (RPD); however, an RPD of greater than 5 mm is thought to be abnormal at 16 to 18 weeks' gestation. If an RPD of greater than 5 mm is discovered by antenatal ultrasound, close postnatal evaluation is merited.[1,16]

A distended bladder, distended ureters, large kidneys, extremity abnormalities, cryptorchidism, and oligohydramnios may indicate prune belly syndrome on antenatal ultrasound.[12] Key factors in a prenatal diagnosis include the age of gestation at the time of diagnosis, ultrasound results of the renal parenchyma, and amniotic fluid volumes.[16] The initial assessment and diagnosis of an infant with prune belly syndrome is influenced by whether prune belly syndrome was suspected in the antenatal period. If prune belly syndrome is suspected, the antenatal assessment should include serial ultrasounds. The presence of abnormal amniotic fluid levels, timing of the rupture of amniotic membranes, determination of maternal medications, and determination of maternal use of tobacco, recreational drugs, and/or alcohol are also essential to the antenatal assessment.[4,5]

The technological advances that have occurred in routine antenatal ultrasounds have played a fundamental role in the diagnosis and management of antenatal abnormalities such as urinary tract obstructions. Potential benefits of relieving urinary tract obstruction in the antenatal period include improved growth, improved pulmonary development, improved renal and bladder function, and fetal survival. Some fetal interventions include vesicoamniotic shunts, open fetal surgery, fetoscopic surgery, and early delivery.[17]

Vesicoamniotic shunting, with percutaneous placed catheters, is useful in some lower urinary tract obstructions such as those that occur with prune belly syndrome. Controversy exists on whether intrauterine

shunt therapy actually improves the survival rate of infants with urinary obstructions compared to no treatment in the antenatal period.[4,17] These shunts are recommended based on serial evaluations of fetal urine chemistries such as sodium, chloride, osmolarity, microglobulin, and total protein. Fetuses with high urine sodium and osmolarity values have the poorest prognosis.[17] Intrauterine shunt placement works by preventing bladder distention, thus preserving renal function. A double pigtail catheter is used as the shunt. It is placed in the bladder and leads to the amniotic cavity to drain fetal urine, prevent bladder distention, and establish normal amniotic fluid levels. After placement, weekly ultrasounds are performed to monitor urinary tract dilation and amniotic fluid levels.[17]

Pulmonary function preservation occurs by allowing urine to drain into the amniotic fluid; creating adequate amounts of amniotic fluid and preventing pulmonary hypoplasia. In 2001, a 20-year-old woman was reported to have undergone 5 surgeries to place fetal urinary shunts to decompress the fetus' bladder and control amniotic fluid levels. At birth, the fetus presented with characteristics of prune belly syndrome, a decompressed bladder, and no pulmonary abnormalities.[12]

The criteria for intrauterine surgical intervention of prune belly syndrome includes second or third trimester gestation, oligohydramnios, megacystis, advanced hydronephrosis, normal karyotype, and an encouraging urinary index (urine sodium less than 100 mg/dL and urine osmolality less than 210 mOsm).[12] Complications of shunt placement include dislodgement of the shunt, urinary ascites, the initiation of premature labor, and chorioamnionitis. A major drawback of intrauterine intervention includes the lack of standardization and indications for shunt placement, lack of randomized trails demonstrating clear benefit, as well as a limited number of institutions that perform the procedure.[4]

With an antenatal diagnosis or suspicion of urinary tract obstruction, the maternal and fetal medical team has an obligation to ensure delivery of the infant occurs at a hospital that is equipped with pediatric urology and neonatology services. In the event a mother with a prenatal diagnosis of prune belly syndrome is admitted to a community hospital, immediate arrangements should be made for maternal transport to a hospital that provides pediatric specialty services. Delivery complications can include preterm labor and chorioamnionitis. The enlarged bladder can lead to fetal entrapment during vaginal delivery. The associated pulmonary hypoplasia in infants with prune belly syndrome can lead to pneumothoraces. The complex management of an infant with pulmonary hypoplasia will most likely need advanced support such as high-frequency ventilation and possibly inhaled nitric oxide (iNO). An experienced obstetric and neonatal team should be present at delivery.[18]

PHYSICAL ASSESSMENT

The characteristic presentation of prune belly syndrome prompts a complete head-to-toe physical assessment and diagnostic evaluation (Table 1). A focused physical assessment of a newborn infant, combined with maternal history, establishes risk factors, detects anomalies, aids in the development of differential diagnosis, creates a baseline assessment, and initiates the need for specialty referrals. The pediatric urologist can guide the healthcare team through the proper steps to make a definitive diagnosis of prune belly syndrome.[14]

In assessment of an infant with prune belly syndrome, perform a thorough head-to-toe assessment. Observe the head for characteristics of Potter's facies caused by oligohydramnios. These include mandibular micrognathia; wide set eyes; flattened palpebral fissures; prominent epicanthus; flattened nasal bridge; large, low-set ears lacking cartilage; and skeletal deformities (Figures 1A-D).[5,19] The classic Potter's facies, caused by oligohydramnios, may not be present if vesicoamniotic shunts have been placed in utero to facilitate adequate amniotic fluid volumes.[17]

Next, examine the chest and abdomen, noting size, shape, and contour.[20] If pulmonary hypoplasia is present, the chest will appear bell-shaped and the infant will display signs of respiratory distress, such as tachypnea, nasal flaring, retractions, grunting, and oxygen desaturation. The abdomen of an infant with prune belly syndrome will appear "prune-like," lacking abdominal musculature, with visible bowel loops and visible margins of the spleen and liver (Figures 2, 3). In severe cases of prune belly syndrome, the complete absence of abdominal wall musculature allows the entire abdominal wall to be lifted and palpated.[14]

Next, auscultate heart sounds and note for the presence and quality of audible murmurs.[21] Ten percent of all infants with prune belly syndrome will have cardiac abnormalities, such as atrial septal defects, patent ductus arteriosus, ventricular septal defects, and tetralogy of Fallot.[1] An echocardiogram can confirm the presence of an underlying cardiac defect.[1,14,22] Then, progress to the abdomen for auscultation. Auscultation of the abdomen should be done before percussion and palpation because of interference with normal bowel sounds.[20] Infants with prune belly syndrome will have normal bowel sounds in all 4 quadrants of the abdomen.[14]

Percussion is an assessment technique that involves striking one part of the body surface sharply to produce a certain sound to determine where underlying organs are located. Percussion of the abdomen is a useful technique to determine the size of the bladder. Percussion is performed by placing the nondominant hand's middle index finger over the area to be assessed. Then strike the finger sharply with the mid-

TABLE 1. Assessment and Findings by Systems[14]

Physical Assessment	
System	**Findings**
Potter's facies	• Mandibular micrognathia • Wide set eyes • Flattened palpebral fissures • Prominent epicanthus • Flattened nasal bridge • Large, low-set ears, lacking cartilage
Chest	• Bell shape • Respiratory distress
Pulmonary	• Respiratory distress (retractions, nasal flaring, grunting, oxygen desaturation) • Bell shaped thorax
Abdomen	• Normal to prune-like in appearance • Visible bowel loops and margins of the liver and spleen
Cardiac	• Murmur
Bladder	• Distended
Urachus	• Patent urachus
Musculoskeletal	• Skin dimples over joints • Scoliosis • Congenital torticollis • Lower extremity malformations: clubfeet, congenital dislocated hips
Genitalia	• Cryptorchidism in males • Pseudohermaphroditism in females

dle finger of the other hand. First, start by percussion of all 4 quadrants of the abdomen to determine the overall tympany and dullness. Tympany, a drum-like sound, will be heard over organs that contain air, such as the stomach. Solid mass organs or organs filled with fluid, such the bladder, will produce dull sounds. To determine if bladder distention is present, start percussion at the symphysis pubis. The bladder is normally located 1 to 4 cm above the symphysis pubis and will produce a dull sound upon percussion. Percuss upward until a tympany sound is audible; this locates the top of the bladder. If the bladder is greater than 4 cm from the symphysis, it is considered distended.[23] An infant with prune belly syndrome will have a large, distended bladder.[14]

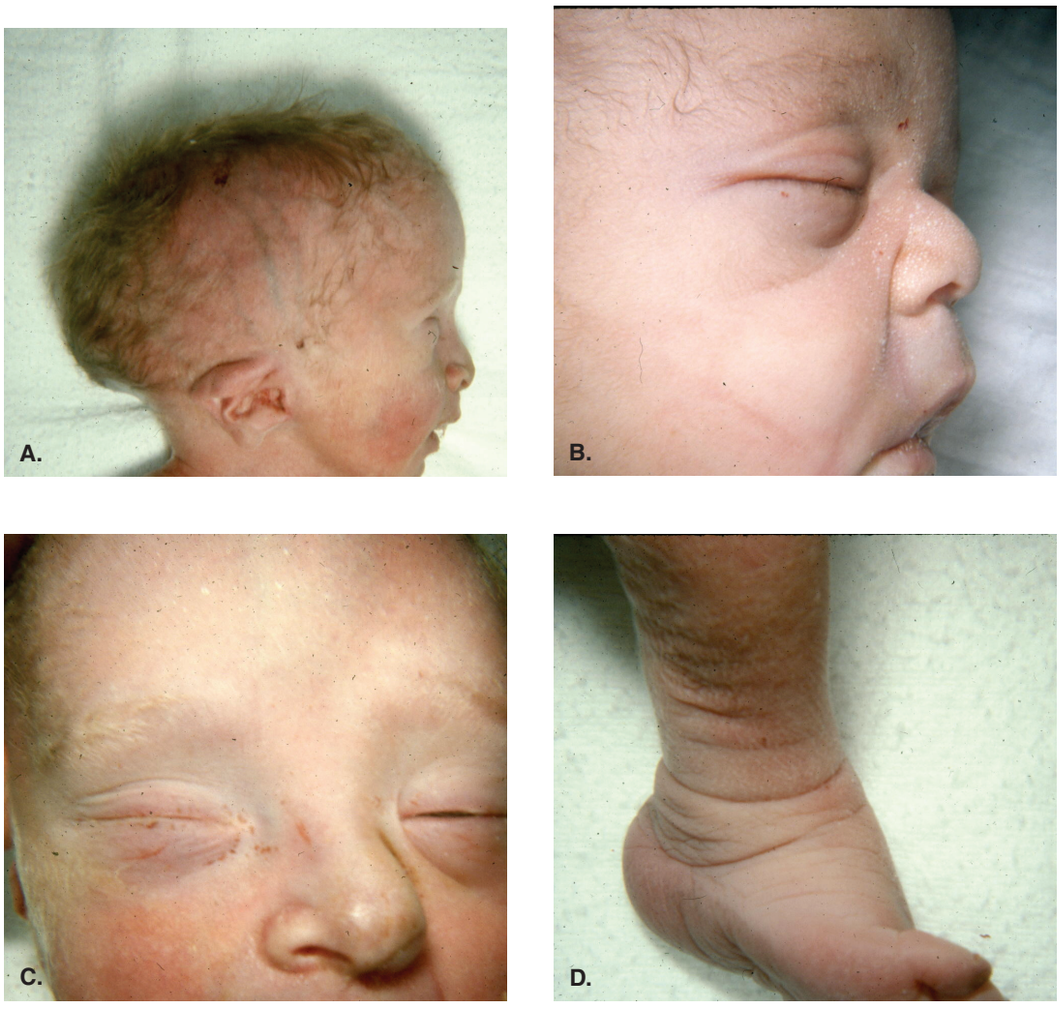
Palpation of the abdomen is best done before air enters the gastrointestinal tract, usually within the first 8 hours after birth. Progression from light to deep palpation of the abdomen in all 4 quadrants locates masses.[21] If possible, palpate the abdomen with the neonate's knees slightly flexed. This will promote abdominal muscle relaxation. A pacifier may make the assessment easier by keeping the infant calm and from crying during the examination. Warming hands by placing them under warm water can also make the examination more comfortable and less stressful for the neonate.[22]

The abdomen of an infant with prune belly syndrome will feel flaccid, lacking muscular tone.[14] To assess the liver, palpate below the costal margin along the midclavicular line. When the infant inspires, the edge of the liver can be felt along the midclavicular line 1 to 3 cm below the right costal margin. The edges of the liver should be smooth, firm, and sharp.[21] When the liver is felt more than 3 cm below the right costal margin, hepatomegaly is suspected. However, more of the liver borders may be palpable in infants with prune belly syndrome without a true hepatomegaly etiology.[24]

To locate the spleen, start palpation above the iliac crest on the left. The spleen of an infant is rarely palpable; however, palpation of the spleen 1 to 2 cm below the left costal margin during the first few weeks after birth is usually a benign finding.[22,24] Infants with prune belly syndrome may have a palpable spleen.[14]

The kidneys are located lower in the abdomen and usually felt in the infant with deep palpation. To palpate the kidneys, start on one side and place one hand behind the infant's back. Use the other hand

FIGURES 1A–D.

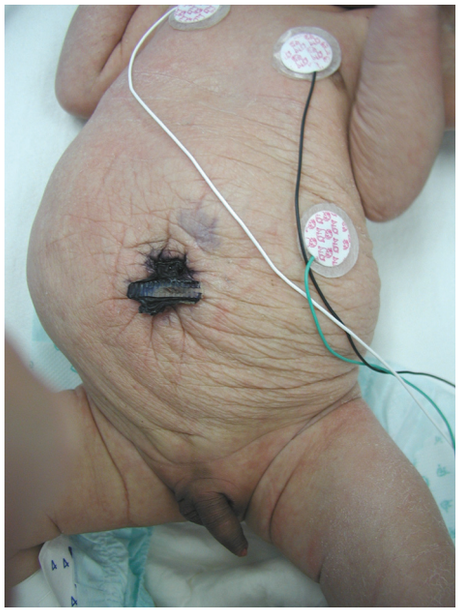


Potter's facies. Characteristics of Potter's facies caused by oligohydramnios. Notice the low-set ears that lack cartilage (A.), flattened palpebral fissures (B.), flat and deformed nasal bridge (C.), and foot deformity (D.). (Used with permission from David A. Clark, MD, Albany Medical Center.)

to start palpation in the lower quadrant and work toward the upper quadrant. Palpation should progress from light to deep.[22] The right kidney is positioned slightly lower than the left kidney. Upon examination, the kidney should feel like a large ripe olive. The normal kidney is approximately 4 to 5 cm, as measured from the upper to lower poles.[20,22] The kidneys of an infant with prune belly syndrome will be larger than normal.[14]

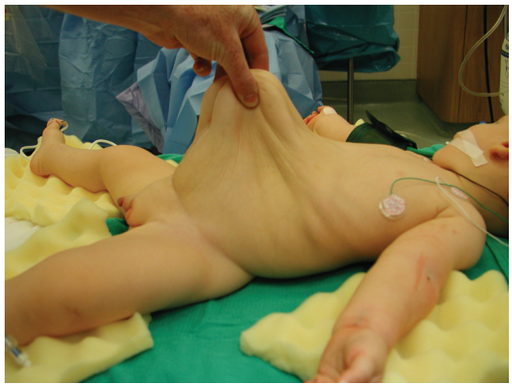
To palpate the bladder, begin at the umbilicus and progress downward until the upper part of the bladder is located. Normally, the bladder is located 1 to 4 cm above the pubic symphysis.[20] A distended bladder will be located centrally in the lower part of the abdomen and feel firm with a dome-shaped structure. The bladder may be diffusely large in an infant with prune belly syndrome.[14] If the infant has undergone vesicoamniotic shunt placement in utero, the shunts may be visible on the abdomen at the time of delivery. Check the umbilical cord, counting the number of vessels. Normally the umbilical cord will contain 2 arteries and 1 vein. A clear discharge from a normal appearing umbilical cord may indicate the presence of a patent urachus. To confirm a patent urachus, a specific gravity test can be preformed on the clear discharge to confirm that the substance is urine. If the infant voids through a normal urethral opening, the quality of the stream of urine should be noted.[21]

FIGURE 2.



Prune-like abdomen. Characteristic appearance of the wrinkled prune-like abdomen of a newborn infant with prune belly syndrome. Notice the bilateral cryptorchidism. (Photo courtesy of John S. Wiener, MD, Division of Urologic Surgery, Duke University Medical Center.)

FIGURE 3.



Flaccid abdomen. Notice the flaccid abdomen of this infant with prune belly syndrome. (Photo courtesy of John S. Wiener, MD, Division of Urologic Surgery, Duke University Medical Center.)

Other associated gastrointestinal abnormalities can include[8,14]:

- Malrotation–midgut anomaly, intestines abnormally rotate or become twisted during embryologic development
- Atresia–failure of the intestinal tract to completely form, causing absence of a normal opening
- Stenosis–narrowing of the center passage within the intestines
- Volvulus–intestines or stomach lack normal attachments to the body, causing the intestines to become twisted, resulting in a lack of blood supply

Clinical symptoms of these abnormalities include bilious vomiting, abdominal distention, abdominal tenderness, failure to pass meconium, rectal bleeding, and signs of shock and sepsis.[9]

Visual assessment of orthopedic anomalies should also occur during the entire examination. Of infants with prune belly syndrome, 50% will have anomalies that are associated with oligohydramnios. Anomalies include[14,21,25]:

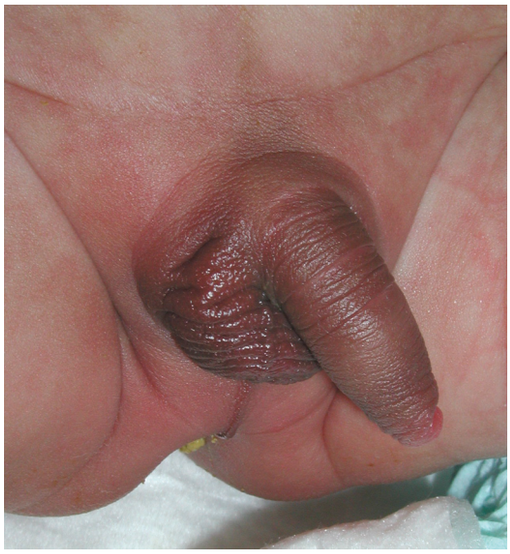
- Clubfeet–complex foot deformity, present at birth
- Congenital hip dysplasia–abnormal development of hip joint, present at birth
- Scoliosis–failure of vertebral formation, segmentation, or a variation of both causing the spine to curve from laterally
- Metatarsus adductus–structural foot deformity in which the forefoot is turned inward, caused by intrauterine positioning
- Vertical talus–disorder of the foot comprised of a rigid rockerbottom or flat foot
- Congenital torticollis–unilateral, spasmodic contraction of neck muscles
- Contractures–an immobilized joint caused by lack of movement in utero

Assessment of the genitourinary system always reveals bilateral cryptorchidism in males (Figures 2, 4) and can reveal pseudohermaphroditism in females.[14] A true undescended testicle occurs when the normal path of testicular descend is interrupted. Empty, hypoplastic scrotal sacs will be found in males with bilateral cryptorchidism.[21] External structures that cannot be determined as male or female in origin are often found in females with prune belly syndrome. The pseudohermaphrodite may present with an abnormally located urethral meatus, clitoral hypertrophy, palpable gonad, inseparably fused labia, or other abnormal dimples or openings.[21] Karyotyping will confirm a male or female genetic composition.[7]

Diagnostic Evaluation

After examination, radiography use evaluates the chest and abdomen for further underlying abnormalities (Table 2). A renal ultrasound assesses urinary

FIGURE 4.



Bilateral cryptorchidism. Bilateral cryptorchidism, which occurs in males with prune belly syndrome. (Photo courtesy of John S. Wiener, MD, Division of Urologic Surgery, Duke University Medical Center.)

tract, bladder, and kidney structures. A definitive diagnosis of prune belly syndrome is made with a comprehensive clinical assessment, radiography, and ultrasound results. On ultrasound, prune belly syndrome will be evident by bladder distention, urethra distention, hydroureter, and hydronephrosis.[14]

Because of the position and makeup of the ureters and kidneys, the infant with prune belly syndrome may exhibit hydronephrosis caused by the reflux of urine in the ureters as well as renal dysplasia and tubular defects.[1] The ureters and bladder of infants with prune belly syndrome are large and dilated (Figure 5). Controversy exists on whether or not to obtain a voiding cystourethrogram (VCUG) for a diagnosis of hydronephrosis. A VCUG evaluates the prostatic-membranous urethra and the bladder and detects the presence of a patent urachus. In addition, a VCUG detects the extent of vesicoureteral reflux, bladder size, and urethral stenosis (Figure 6). Some urologists think a VCUG in patients with urinary obstructions should be avoided because the procedure may be more damaging than beneficial to infants who have severe urinary reflux and retention.[14] During a VCUG, bacteria could potentially be introduced and cause a urinary tract infection because of the stasis of urine within the bladder. Other urologists argue that a VCUG is necessary for the evaluation of reflux, the extent of reflux, and bladder size.[14] On a VCUG, prune belly syndrome will be evident by bladder distention without complete emptying, vesicoureteral reflux, dilated posterior urethra, urethral obstruction, posterior urethral values, urethral diverticulum, and bladder diverticulum. Prophylactic antibiotics should be started if a diagnosis of reflux is made to reduce the risk of pyelonephritis. Some literature supports discontinuing antibiotics if the VCUG and renal ultrasounds are normal; however, most pediatric urologists agree that prophylaxis should be started with or without reflux because of the presence of urine stasis.[1] Surgical management of reflux is indicated only for repeated urinary tract infections.[1]

For institutions with access to nuclear radiology, a technetium Tc 99m dimethylsuccinic acid (DMSA) renal scan is a useful diagnostic tool. The scan uses cortical agents to evaluate kidney appearance and function; however, it is an unpredictable tool for the detection of urinary tract obstructions in cases with poor renal function in combination with urethral dilatation and tortuosity. Computed tomography (CT) scans may be a useful diagnostic tool for diagnosing prune belly syndrome (Table 1).[14]

Immediate Care Issues and Nursing Implications

There are key challenges in caring for an infant with prune belly syndrome. Complex care issues include respiratory and fluid assessment. Infants with urinary tract obstructions who have low amniotic fluid volumes are at risk for pulmonary hypoplasia. Acute clinical signs of pulmonary hypoplasia include inability to ventilate properly, requirement of high pressures to ventilate the lungs, and lungs that feel stiff during ventilation. Pulmonary air leak risks increase in infants with pulmonary hypoplasia. Signs of a pulmonary air leak include sudden deterioration of clinical condition, oxygen desaturation, decreased breath sounds on the affected side, hypotension, a shift of the mediastinum (detected by a shift of the point of maximal cardiac impulse on auscultation only with tension pneumothoraces), and mottling of the skin. To confirm a pneumothorax without having to wait for a chest x-ray, transillumination of the chest wall can confirm the diagnosis. If symptomatic and the clinical status has deteriorated, needle aspiration to remove the air should occur immediately. Otherwise, placement of a thoracostomy tube in the anterior chest wall may be necessary to remove the air.[9,14]

The respiratory status of intubated infants with prune belly syndrome is critical because any respiratory infection can pose a serious risk for these infants. In addition, the lack of abdominal muscles means there are no accessory breathing muscles, placing the patient at high risk for respiratory infections. The nurse and respiratory therapist are key individuals for assessing the respiratory status of these infants on a continued basis. Bedside assessments should include

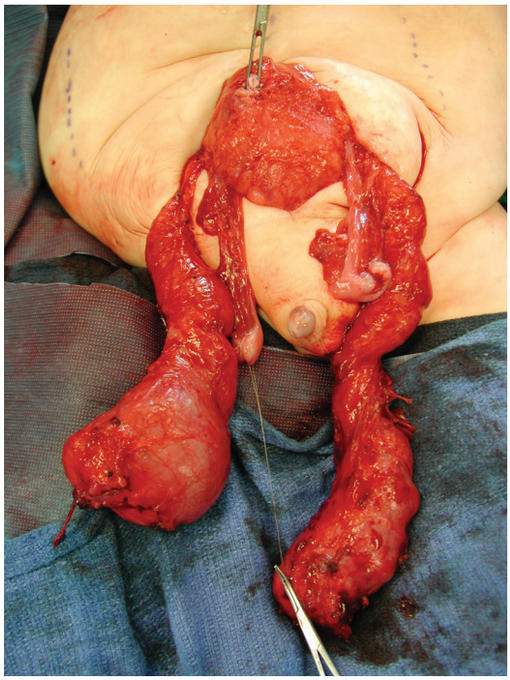
TABLE 2. Diagnostic Findings[14]

Radiographic Findings	
Abdomen	• Diffusely distention flanks • Mass-like areas that represent hydronephrotic urinary tract
Chest	• Hypoplastic lungs • Radiolucent lung fields • Flared lower ribs • Improper alignment of vertebral column
Kidneys	• Dilated • Clubbed • Dysplastic calyces
Ureters	• Markedly dilated and twisted • Tortuous
Bladder	• Vertical with a triangle base
Urethra	• Urachal remnant at dome • Wide, long, and posterior with utricular remnant • Posterior urethra triangle in appearance • Testes located in the abdomen or inguinal canal
Cryptorchidism	• Testicular hypoplasia
Ultrasound Findings	
Kidneys	• Renal dysplasia • Dilated renal pelvis • Diffuse, hyperechoic parenchyma • Small parenchymal cysts • Clubbed dysplastic calyces
Ureters	• Posterior urethral valves (PUV) • Tortuous ureters • Dilated with thickened, fibrous walls
Bladder	• Hourglass shaped bladder in the presence of a patent urachus • Large with thickened walls
CT Findings	
Abdomen	• Broad and floppy • Absence of abdominal musculature
Ureters and Bladder	• Dilated
Kidneys	• Hydronephrosis

monitoring of ventilator settings, oxygen requirements, blood gases, and vital signs. Adequate ventilation pressures should be established without placing undue pressure on the abdomen.[14]

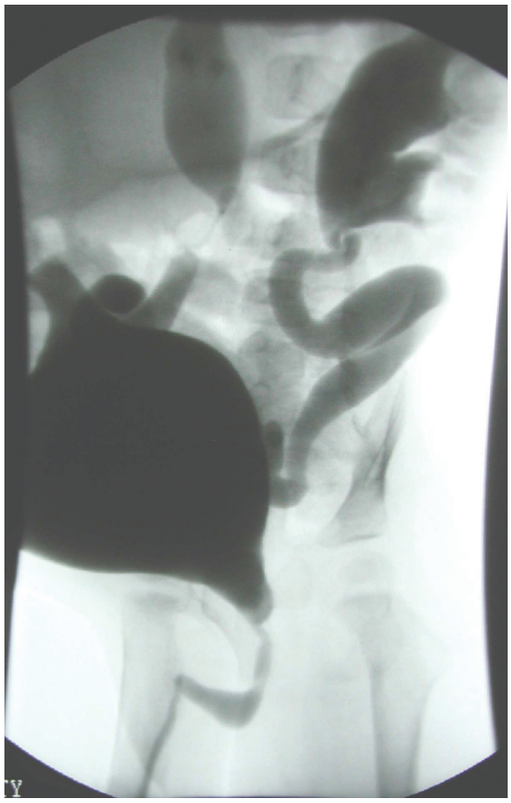
Fluid and electrolyte balance, as well as nutritional needs, are important for infants with prune belly syndrome. Infants with urinary obstruction are prone to fluid and electrolyte imbalances. Because of decreased sodium absorption in the collecting tubules, infants with prune belly syndrome waste sodium. Sodium supplementation is often required. Serum electrolytes, including a blood urea nitrogen (BUN) and creatinine (Cr), require close monitoring during the first week of life. A rising BUN and Cr suggests poor renal functioning. The main goal of treatment of prune belly syndrome is the preservation of existing renal function. Other electrolyte imbalances include hyperkalemia. Daily monitoring of strict intake and output (including any flush solutions) and weight are vital to assess alterations in fluid balance. Assessment of renal function includes monitoring of laboratory values (Table 3).[1,24,26] Parenteral and enteral nutritional should include low protein intake to reduce the workload of the kidneys.[24] Adequate nutritional needs are also required to support the body's response to infection. Infants with renal insufficiency, as sometimes

FIGURE 5.



Megaureters. An intraoperative dissection revealing a large bladder and megaureters in an infant with prune belly syndrome. (Photo courtesy of John S. Wiener, MD, Division of Urologic Surgery, Duke University Medical Center.)

FIGURE 6.



Urinary reflux. A radiographic image of a VCUG representing extensive reflux of urine from the bladder into the ureters. (Photo courtesy of John S. Wiener, MD, Division of Urologic Surgery, Duke University Medical Center.)

TABLE 3. Laboratory Tests and Interpretation for Assessing Renal Function[1,14,26]

Laboratory Test	Value Interpretation
Specific gravity	Indirect measurement of urine osmolarity, reflects renal capacity to concentrate or dilute urine and absorb or excrete sodium; normal values range from 1.002 to 1.012
Protein excretion	Evaluates the kidney's ability to process protein
Hematuria	Evaluates intrinsic renal damage or clotting abnormalities
Serum urea nitrogen	Indicator of renal function; suspect renal insufficiency if values of greater than 20 mg/dL or a rise of 5 mg/dL per day are observed
Glomerular filtration rate (GFR)	A rate at which blood passes through the renal capillaries and plasma is filtered through the glomerular capillary walls per unit of time; estimates the number of functioning nephrons
Serum and urine electrolytes	Evaluates fluid and electrolyte management and assesses renal tubular function

observed in prune belly syndrome, can exhibit signs of failure to thrive.[1]

Preservation of renal function is aided with the use of prophylactic antibiotics to prevent infection. The urinary tract is a major source of infection in infants with prune belly syndrome because of the stasis of urine and potential for reflux; therefore, the use of prophylactic antibiotics is warranted.[14] A urinary tract infection may be asymptomatic, or the infant may display nonspecific signs, such as poor feeding, irritability, and lethargy.[9] Urine cultures, urinalysis, electrolyte values, and serum Cr and BUN values can identify metabolic abnormalities and urinary tract infections early.[14]

Surgical Management

For more than 25 years, surgical treatment for a dilated urinary tract in infants with prune belly syndrome has been debated. Renal deterioration could occur because of infection from the stasis of urine by the poorly functioning ureters. Nevertheless, approximately 50% of infants with prune belly syndrome require only nominal surgical intervention. Even though a urachus may be patent initially, bladder drainage can be unpredictable because the urachus usually closes within the first few weeks of life. The best surgical intervention for a urethral obstruction is an urethrotomy.[14] Repeated soft dilation of the urethra also can manage urethral stenosis or atresia and eventually produce a normal caliber urethra. When urinary infection has occurred from stasis of urine within the bladder, a cutaneous vesicostomy can be valuable. In a vesicostomy, the dome of the bladder is brought to the surface of the skin below the navel. Ligation of a patent urachus can also be preformed at the time of the vesicostomy. Several surgical procedures exist for the treatment of a functioning obstruction caused by posterior urethral valves. The valves are best managed by performing a transurethral resection.[14] One method of correction is performed by endoscopic incision at the site of the posterior urethral valves to perform a resection of the valves.[14]

Because of the potential risk of testicular malignancy, the testicles should be placed in the scrotum (orchiopexy).[14] Orchiopexy may be delayed until the time of urinary tract reconstruction if repair is anticipated before 1 year of age. Abdominal wall laxity in infants with prune belly syndrome can improve with time; and abdominal wall reconstruction can be performed to improve urinary tract emptying and respiratory and bowel functioning. Various surgical options exist for abdominal wall reconstruction, but discussions of such options are beyond the scope of this article. This stage of surgical correction usually occurs when the infant is between 8 and 12 months of age. During abdominal wall reconstruction, the most critical factor to monitor is airway pressure. If the abdominal wall is too tight, spontaneous breathing after surgery may be hindered. Airway pressures should be noted before surgery and maintenance of the same airway pressures during and after surgery should be the goal.[14,27]

Outcomes

Mortality depends on the function of the urinary tract system and the degree of pulmonary hypoplasia. If the urinary tract system is in poor condition with decreased urethral peristalsis and weak expulsion of urine through the ureters, infection and stone formation caused by stasis of urine can result. In these cases, renal failure develops from renal dysplasia and from renal scarring secondary to infections. In the 1970s, approximately half of the patients with prune belly syndrome died early in infancy. Today, the survival rate has improved in patients who survive the neonatal period, with approximately 25% to 30% of these patients experiencing chronic renal failure and end-stage renal failure.[14,28] In one study, infants who underwent in utero vesicoamniotic shunts had a overall 1 year survival rate of 91%.[26] Acceptable renal and bladder function occurred in most of these patients. Approximately one third of these surviving infants ultimately required peritoneal dialysis and/or renal transplantation. There are no increased complications in infants with prune belly syndrome who require peritoneal dialysis as compared with infant's without prune belly syndrome who require peritoneal dialysis.[26] Appropriate initial evaluation and treatment often can avoid or delay the need for peritoneal dialysis and/or renal transplantation.[14]

Family Care Issues

When prune belly syndrome is diagnosed prenatally, parents can be provided with a wealth of information and guidance regarding the anticipated care of their infant. After the infant is admitted to the neonatal intensive care unit (NICU) with a confirmed diagnosis of prune belly syndrome, daily updates on the infant's progress and condition facilitate effective communication and build trust between the family and the entire healthcare team. Postdischarge needs and assessments must not be ignored. When social challenges are identified, the help of the unit's social worker should be enlisted. The NICU social worker must assess the family's home environment, availability of adequate supplies, and telephone and heat access and coordinate follow-up care after discharge from the hospital. By understanding the pathophysiology and theories behind prune belly syndrome as well as general treatment regimens, the bedside nurse can provide optimal care for the newborn and provide accurate information and support to the parents.[29]

CONCLUSION

Because of the rarity of prune belly syndrome, little consistent and evidence-based data exists on the proper management of this condition. The underlying etiology of obstruction and pulmonary hypoplasia influence the survival and outcome of infants with prune belly syndrome. From a research perspective, no reports have been published comparing surgical and nonsurgical treatment options.[3] However, it is concluded that management of prune belly syndrome depends on a functioning urinary tract system and the extent of associated symptoms. The timing and selection of the management plan is highly individualized. With the standardization of diagnosis and treatment, a more research-based plan of care can be devised, enabling an accurate assessment of the efficacy and role of prenatal therapy and postnatal interventions.[14]

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