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CLINICAL MANIFESTATIONS OF PRUNE-BELLY DISEASE, ONE OF THE UROLOGICAL DISEASES THAT REQUIRES TREATMENT IN CHILDHOOD

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Annotation. Prune-Belly syndrome, also known as Eagle-Barrett syndrome, is a rare disorder characterized by partial or complete absence of the stomach (abdominal) muscles, failure of both testes to descend into the scrotum (bilateral cryptorchidism), and/or urinary tract malformations. The urinary malformations may include abnormal widening (dilation) of the tubes that bring urine to the bladder (ureters), accumulation of urine in the ureters (hydroureter) and the kidneys (hydronephrosis), and/or backflow of urine from the bladder into the ureters (vesicoureteral reflux).

Keywords: urinary system, kidney, bladder, ureter, testes, prostate.

Introduction

Prune-belly syndrome (PBS) is a syndrome with a broad spectrum of severity. It is a constellation of anomalies. The three major aspects are abdominal musculature deficiency, bilateral intra-abdominal testes, and an anomalous urinary tract. The urinary tract is characterized by variable degrees of hydronephrosis, renal dysplasia, dilated tortuous ureters, an enlarged bladder, and a dilated prostatic urethra. Associated anomalies involving the respiratory tract, gastrointestinal tract, cardiac system, and musculoskeletal systems also occur. There is a broad spectrum of severity of the syndrome, with some children who do not survive the newborn period and others who are minimally affected. The single most important determinant of survival is usually the severity of the urinary tract anomaly, and in particular, the degree of renal dysplasia. Osler’s vivid description of the abdominal wall of an infant with the characteristic findings led to the term “prune-belly syndrome.”However, Frolich first described the characteristic abdominal wall in 1839, and the full triad of anomalies was described by Parker in 1895. Other names that have been applied to this syndrome include triad syndrome, Eagle Barrett syndrome, and abdominal musculature syndrome. The incidence of PBS has been reported to be between 1 in 29,000 and 1 in 40,000 live births, similar to that of bladder exstrophy, with 95% of cases occurring in males.7 Females with PBS have the abdominal wall deficiency and urinary tract dysmorphism without any gonadal anomaly. A higher incidence is noted among twins, blacks, and children born to younger mothers. The incidence appears to be declining in recent years, possibly due to prenatal diagnosis and a decision to terminate the pregnancy.

Embryology

There have been several theories as to the embryogenesis of PBS. However, there is no experimental model that can be used to test these theories, so the exact mechanism remains elusive. The four principal theories are early in utero posterior urethral obstruction resulting in severe dilation of the urinary tract and possible fetal ascites and oligohydramnios; a primary defect in the lateral plate mesoderm, which is the precursor of the ureters, bladder, prostate, urethra, and gubernaculum; an intrinsic defect of the urinary tract leading to ureteral dilation and fetal ascites; and a yolk sac defect. None of these theories has universal acceptance, and there is some overlap among them.

Clinical features of Prune-Belly syndrome (Genitourinary anomalies)

Kidneys. The spectrum of renal abnormalities extends from normal renal parenchyma to dysplasia. The more severely dysplastic kidneys are generally associated with bladder outlet obstruction in which there has not been decompression

through a patent urachus. Dysplasia is present in 50% of cases; however, it may vary in degree and laterality. Renal dysplasia in PBS of the Potter type II and IV varieties are seen. The Potter type II variety with few nephrons and parenchymal disorganization is more indicative of a renal mesenchymal defect, whereas the Potter type IV with cortical and tubular cysts is associated with outlet obstruction. The renal collecting system is characteristically dilated, often to a severe degree. The degree of dilation, however, does not correlate with the degree of renal dysplasia. Calyceal morphology may be well preserved even in the face of massively dilated ureters and renal pelves. Ureteropelvic junction obstruction can occur on a primary or secondary basis; however, nonobstructive hydronephrosis is the rule. It is renal infection, rather than obstruction, that poses the greatest risk to renal function.

Ureters. The ureters are typically dilated, tortuous, and redundant. The proximal (upper) portions of the ureters are usually less abnormal than the distal segments, although massive dilation and stenosis can occur at any level. Typically, histologic sectioning demonstrates a lack of smooth muscle cells and an increase in fibrous connective tissue. There are more normal-appearing smooth muscle cells in the proximal segments. This fact is critical when ureteral reconstruction is undertaken. The ratio of collagen to smooth muscle cells in prune-belly ureters is elevated, especially in refluxing ureters. A decreased number of thick and thin myofibrils seen on ultrastructural examination is thought to contribute to the poor peristalsis. Vesicoureteral reflux is present in 75% of children with PBS. Obstruction is not common but has been reported both at the ureteropelvic and the ureterovesical junctions. These large ureters may have ineffective peristalsis because of poor ureteral wall coaptation. The ureteral conduction wave reaches a reduced smooth muscle cell population that has poor contractile potential because of reduced myofibrils, often separated by patches of collagen; the resulting bolus of urine reaches more dilated ureteral segments as it progresses towards the bladder. This can be seen fluoroscopically as ineffective peristalsis, resulting in upper tract stasis that may lead to infection.

Bladder. The bladder usually appears massively enlarged, with a pseudodiverticulum at the urachus. The urachus is patent at birth in 25% to 30% of children. Histologically, the bladder has an increased ratio of collagen to muscle fibers in the absence of obstruction. The wall is smooth, unlike that seen in obstructed bladders. The pelvic distribution of ganglion cells has been shown to be normal. Smooth muscle hypertrophy is seen in the obstructed prune bladder. As noted by Williams and Burkholder, Stephens had demonstrated that the trigone is splayed, with the ureteral orifices displaced laterally and superiorly, possibly contributing to the high incidence of reflux. On voiding, the bladder neck opens widely into a dilated prostatic urethra. Urodynamic assessment usually shows normal compliance; however, there is a delayed first sensation to void and a very large capacity. The ability to empty the bladder is variable, with some emptying well and others carrying a significant postvoid residual. This variability is thought to be based on a relative outlet obstruction and a variable ability of the bladder to generate sufficient pressure with a detrusor contraction. If the relative outflow resistance prevents effective bladder emptying, the term unbalanced voiding is used. Despite these limitations, about 50% of PBS patients void spontaneously with normal voiding pressures, normal flow rates, and low post-void residuals.

Prostate and Accessory Sex Organs. The dilation of the posterior urethra is a result of prostatic hypoplasia, probably caused by abnormal mesenchymal-epithelial development. Histologically, there are few prostatic cellular elements, with a reduction of both epithelial and smooth muscle cells and an increase in connective tissue cells. Various obstructive lesions of the distal posterior urethra have been described, such as urethral atresia, valves, urethral stenosis, urethral membrane, and urethral diverticulum, and are thought to occur in 20% of cases. Stephens described an angulation of the urethra during voiding, which he referred to as a type IV valve, that results from lack of prostatic parenchymal tissue. Prostatic hypoplasia, the etiology of which is controversial, is thought to be a factor in the ejaculatory failure of PBS patients. The vas and seminal vesicles are often atretic, although either may be dilated or thickened. The epididymis may be poorly attached to the testis, as is seen commonly in abdominal undescended testes. There also may be lack of continuity between the efferent ductules and the rete testis. Ejaculation usually occurs in a retrograde fashion because of the incompetent bladder neck.

Testes. The most typical finding is bilateral intra-abdominal testes lying over the iliac vessels. Although mechanical forces such as a distended bladder and intra-abdominal pressure have been implicated in maldescent of the testes, the fact that some patients with the typical urinary tract and abdominal musculature anomalies (termed pseudo-prune patients) may have descended testes raises some doubt as to a purely mechanical etiology. Pak and colleagues compared the histology of the testis in PBS patients with that of non–prune-belly intra-abdominal testis as well as age-matched controls. They found no difference in germ cell counts, spermatogonia, or Leydig cells between PBS

and non-PBS patients with intra-abdominal testes. However, germ cell counts in PBS patients younger than 1 year of age were similar to those of age-matched controls, implying that the environmental state of the abdomen is a major factor in their later spermatogenic potential. This mirrors the finding by Nunn and Stephens of normal germinal epithelium in fetal and newborn PBS testes. Alternatively, Orvis and associates noted a decreased number of spermatogonia and Leydig cell hyperplasia in fetal PBS testes, implying an intrinsic testicular abnormality. Azoospermia was found in adult PBS patients, and no patient with PBS has been reported to have fathered a child. More recently, Ross and associates documented paternity in three adults with classic PBS, which was achieved by sperm retrieval techniques and intracytoplasmic sperm injection (ICSI). The infertility is thought to be caused by a combination of testicular histologic abnormalities, structural defects of the ducts, and prostatic abnormalities. Three cases of testis tumor have been reported. Massad and coworkers described histologic testicular patterns similar to intratubular germ cell neoplasia in three infants. Although the risk of malignancy may be relatively low considering the lack of germinal epithelium, it is clear that placement of the testis in the scrotum and long-term follow-up are necessary to potentially reduce the risk of testicular malignancy and enhance detection.

Conclusion, some of the cases of this disease related to the genitourinary system have been listed above. In addition, there are also abdominal wall anomalies, cardiac, pulmonary and orthopedic manifestations of this disease. The diversity of clinical manifestations requires the knowledge of a number of specialists to treat the disease.

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