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"POSTERIOR URETHRAL VALVES" EMBRYOLOGICAL BASIS AND ITS CLINICAL SIGNIFICANCE

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ABSTRACT
A lot of controversy continues regarding the development of human penile urethra. Posterior urethral valves are congenital disorder and can only seen in male infants. It caused by failure of regression of the mesonephric duct. Most of cases PUV's a sporadic and only occur in males, evidence suggests that they can be found in siblings or twins in a family. If PUV's are not diagnosed and treated early they can cause damage in the ureters, urethra, bladder and kidney, constraints lung developments cause of low quantity of amniotic fluid. PUV's are commonly diagnosed prior to birth or at birth when a male infant is evaluated for antennal hydronephrosis.

Introduction

The male urethra is a slender fibro muscular tube that steers urine and semen from the bladder and ejaculatory ducts, respectively, to the exterior of the body. The male urethra is a distinct structure, composed of a mixed series of sections, prostatic, membranous, and spongy.

PUV's was first described in 1515 and afterwards observed at autopsies. In 1802, the first description for PUV was scripted and presented in an article on lithotomy. The first report was in British journals is discovered in the Lancet, in which Dr Budd reported a PUV's in a 16-year old schoolboy who died because of renal failure. PUV's remains to be a important cause of sickness, mortality and ongoing renal damage in infants and children. The incidence of PUV's is estimated to be 1 in 5000 to 8000 male births, but it may be more common for some fetal death. The incidence of this disorder in the African population is yet unknown. High bladder outlet blockade throughout gestation period leads to harshly compromised renal function secondary to renal dysplasia in many children with PUV. Treatment of PUV remains a medical challenge, requiring active supervision from infancy to adulthood to prevent progressive renal dysfunction and deterioration of the upper and lower urinary tracts.

Posterior urethral valve is a genital congenital disorder. It is an obstructive developmental anomaly of male newborns in urethra and genitourinary system. A posterior urethral valve is a barricading membrane in the posterior end of the male urethra as a result of abnormal in utero development. This membrane affects urine flow cause urine flows back to becomes full and and the amniotic fluid decreases and less amniotic fluid means major problems in development of structures like lungs. This anomaly is the most common cause of urinary bladder outlet obstruction in male neonates. The disorder differs in degree, with minor cases followed, more severe cases. Severe cases can have renal and respiratory failure from lung witch are very fatal if not detected early. Underdevelopment of lungs and renal structures is result of low amniotic fluid volumes. This neonate will require intensive care and close nursing. The incidence of this anomaly one in 8000 babies.

Incidence

PUV's still continues to be a major cause of morbidity, mortality and ongoing renal damage in infants and children. The incidence of PUV is estimated to be 1 to 5000 or 8000 male births. The incidence of this disorder in the African population is not yet known. Elevated bladder outlet obstruction throughout gestation period leads to severely compromised renal function secondary to renal dysplasia in many offspring with PUV,s. Treatment of PUV remains a clinical challenge, requiring active management from early infancy to adulthood in order to avoid continuation to renal failure and degradation of the upper and lower urinary tracts.

Bladder and Urethra normal ontogenesis

The urinary bladders to the openings of ejaculatory ducts are derived from caudal part of vesicourethral canal. The posterior wall is derived from absorbed mesonephric ducts. The rest of prostatic urethra and membranous urethra are derived from the pelvic part of definitive urogenital sinus. The Penile part of the urethra is derived from the phallic part of definitive urogenital sinus .The most terminal part is derived the ectoderm.

During the normal development urethra and bladder 4th to 7th week. The cloaca will divide into two parts the urogenital sinus anteriorly and the anal canal posteriorly. In-between this parts the uro-rectal septum is found this is a layer of mesoderm between the two parts separating them, primitive anal canal and the urogenital sinus. The tip of the septum will form the perineal body.

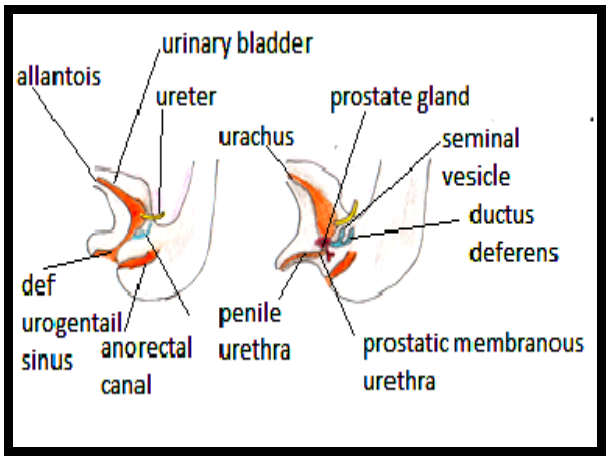

Fig 1. Diagrammatic representation of the development of male external genitalia.

Three portions of the urogenital sinus are notable. The uppermost and largest part is the urinary bladder. Originally the bladder is constant with the allantois, but when the lumen of the allantois is obliterated, a fibrous cord, the urachus, remains and connects the apex of the bladder with the umbilicus in the adults, median umbilical ligament. The pelvic section of the urogenital sinus is a narrow channel which in the male will give rise to the prostatic and membranous parts of the urethra. We will be concentrating phallic section of the urogenital sinus. It is flattened from either side and as the genital tubercle grows, this part of the sinus will be pulled ventrally (differs greatly between the two sexes).

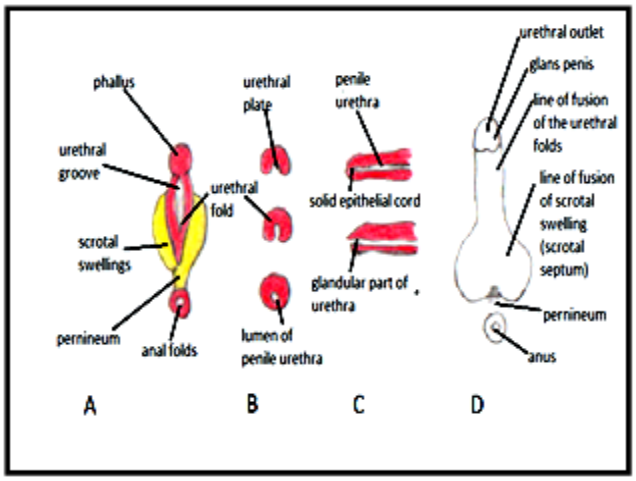

Fig 2. Diagrammatic representation of the development of male external genitalia.

Ontogenesis of Abnormal male urethra

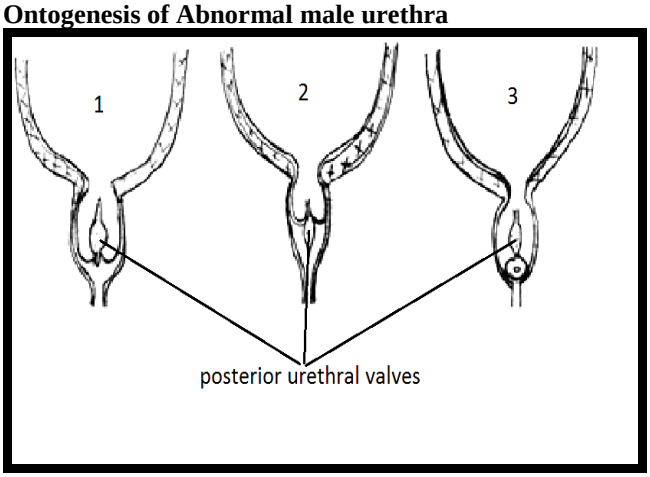

Fig 3. ***Diagrammatic representation of three types of PUV's.***

For males at the 9 ninth week of gestational age, under the influence of testosterone produced by Leydig cells, the genital tubercle and the genital swellings lengthens, they enlarge and rotate posteriorly, they begin to fuse from posterior to anterior. The genital tubercle elongates, on the ventral surface on either side of the evolving channel, two sets of tissue folds develop, the urethral groove. The closest medial endodermal folds will join in the ventral midline to form the male urethra and the lateral folds will fuse developing the urethra, penile shaft and skin. The Glans penis develops develops genital tubercle, body of the penis develops from urethral folds and the scrotum develops from genital swellings. At the 13th week the urethra is almost fully developed.

Type 1: Most common, occurs when the two mucosal folds lengthen anteroinferiorly from bottom of verumonatum and fuse anteriorly at lower level. Type 2: Rare it no longer considered as a valve but a variant the folds extend along posterolateral urethral wall from ureteric orifice to verumontanum. Type 3: Circular diaphragm with central opening in membranous urethra. It is found below the verumontanum and occurs due because of the abnormal canalization of urogenital membrane.

As we already know vast majority of cases of PUV,s are sporadic and rare examples of PUVs occurring in a family have been reported .The male penis develops from different parts of the urogenital sinus .The prostatic part develops from the mesonephric duct, the membranous parts develops from the pelvic part of DUGS ,The penile part from phallic part of DUGS and the terminal part develops from surface ectoderm. In this article we we will focus on the the prostatic part of the penis because this is the site where posterior urethral valves develop. During the early embryogenesis, the utmost caudal end of the mesonephric duct is absorbed into the primitive cloaca at the place of the future verumontanum in the posterior urethra. Failure for regression of mesonephric duct will leave behind a valve like membrane from Wolffian duct and it intern called PUV. In fit males, the remnants of this development are the posterior urethral folds, called plicae colliculi. Posterior urethral valves result from the formation of a thick, valve-like membrane from a tissue of Wolffian duct origin (failure of regression of the mesonephric duct) that courses obliquely from the verumontanum to the most distal portion of the prostatic urethra. This is thought to occur in early gestation (5-7 weeks). The valve is a diaphragm with a central pinhole, however as it is more rigid along its line of fusion it gradually distends and becomes distended into a bilobed sail-like or windsock-like structure.

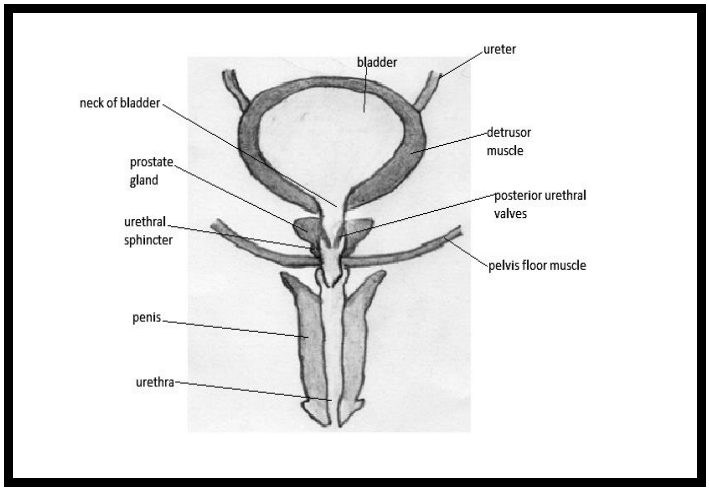

Fig 4. Diagrammatic representation of posterior urethral valves in a penis.

Histological studies suggest that PUV's are developed at approximately 4 weeks gestation, as the mesonephric duct joins with the developing cloaca.

Discussion

There is still much more to be learned about PUV's. The embryology and pathology aspect of urethral development remains a field of active study. Simple questions concerning the birth of the disorder and its classification remain unanswered and still a mystery. It is so very uncommon for the research of an unusual clinical entity to raise more questions than solutions and, in fact, it is a testament to the broad research that has been executed over the past century that the traditional explanations and classification of PUV have been questioned, altered, and clarified. Hopefully, the future will continue to produce new revelations. PUVs are the most common cause of obstruction in neonates, when obstruction can be overcome by detrusor contraction it may remain silent until later life. The exact age of presentation is not known and varies greatly. PUVs usually detected in infants are more severe than in adults. Symptoms leading to the diagnosis include irritative symptoms of the lower urinary tract, recurrent urinary infections, obstructive symptoms and rarely, ejaculation diseases, gross haematuria, and renal insufficiency

Renal treatment for patients present with bilateral renal dysplasia at birth, in the past, if patients did not die of associated pulmonary insufficiency, they died cause of progressive renal insufficiency. Advances in peritoneal dialysis have made it possible for some to be treated successfully from birth. About one third of patients with PUVs eventually progress to end-stage renal disease (ESRD) and will in turn require dialysis or transplantation. Progression of ESRD is speeded at the time of puberty as a result of the increased metabolic workload placed on the kidneys. Bladder management to all male children with antenatal hydronephrosis should undertake voiding cystourethrography (VCUG) shortly after birth to exclude PUV. While awaiting the study results, place a 5- or 8-French urethral catheter to allow for bladder drainage. If valves are confirmed. Valves can be incised within the first few days of birth. However, the newborn urethra may be too tiny to accommodate available equipment. In these persons, a vesicostomy can be performed as a temporary answer until urethral growth has been adequate to allow transurethral incision. Urinary drainage by feeding tube in early days of infancy, followed by valve ablation is the best treatment in PUV, and urinary diversion improves the outcome. Voiding cystourethrogram (VCUG) is still the best imaging modality for documenting PUV. The factors like renal dysplasia and UTI have their role in final outcome.

Conclusion

Management of posterior urethral valves is still a clinical challenge in pediatric urology. There is still much to know about obstructive bladder physiology. The long-term outcome depends on the degree of renal damage, upper tract changes and bladder dysfunction. The ultimate goal of management is to maximize renal function, maintain normal bladder function, minimize morbidity and prevent iatrogenic problems.

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