Posterior urethral valves in children. Part II

Curajos B.

"Nicolae Testemitanu" State University of Medicine and Pharmacy

Abstract

Valvele uretrei posterioare la copii

Autorul prezintă experiența proprie confruntată cu datele literaturii de specialitate în diagnosticul și tratamentul valvei de uretră posterioară (VUP) la copii. În ultimii 15 ani, în Centrul Național Științifico-practic de Chirurgie Pediatrică Natalia Gheorghiu au fost diagnosticați și tratați 25 de pacienți cu VUP, initial fiind stability diagnosticul de stenoză congenitală a uretrei (290 copii). Severitatea și gradul de obstructive au depins de configurația membrane obstructive. Toți copiii au beneficiat de ablația valvei fiind folosit uretroromul original propus de autor.
Autorul conchide că toți pacienții cu VUP supuși tratamentului chirurgical necesită monitorizare pe termen lung până la adolescent, iar prognosticul depinde de modificările suprastenotice și de gradul de disfuncție a vezicii urinare la adresare.

Cuvinte cheie: valve uretrale, diagnostic, ablație, copii

Abstract

The author presents his own experience confronted with data from the literature in the diagnosis and treatment of posterior urethral valve (VUP) in children. During the last 15 years, in CNȘPCP "Natalia Gheorghiu" 25 patients with urethral valves were diagnosed and treated, initially for which the diagnosis of congenital stenosis of the urethra was established (290 patients). The severity and degree of obstruction depended on the configuration of the obstructive membrane. All children benefited from valve ablation using the original urethrotome proposed by the author, which allowed good results.
The author concludes that all patients with VUP undergoing surgical treatment require long-term monitoring until adolescence, and the prognosis depends on suprastenotic changes and the degree of bladder dysfunction at the address.

Keywords: urethral valves, diagnostic, excision, childrens

Correspondence to: e-mail: boris curajos@gmail.com

Introduction

Posterior urethral valves (VUP) are defined as a congenital malformation characterized by the presence of semicircular folds of the posterior urethral mucosa, which creates an obstacle to urine evacuation with serious repercussions on the upper urinary tract [27]. VUP is the leading cause of congenital postvesical obstruction of urinary flow in male children, being a significant cause of morbidity, mortality and persistent kidney damage in infants and children of various ages [23].

The incidence of VUP is 1: 5000 - 1: 8000 boys or 1: 25000 live births [25, 26], this malformation representing about 10% of all urinary obstructions diagnosed prenatally [4].

VUP were first described in 1717 by Morgagni, later Langenbeck (1802) reported the presence of valve-like folds during the dissection of corpses. In 1832, Velpeau described the presence of folds in the posterior urethra as houses resemble valves and mentioned that they can cause obstruction when a catheter passes. In 1840, Budd presented the case of a 16-year-old sailor who died, the necropsy found excessive dilation of the ureters similar to the size of an intestinal loop, a dilated and thickened bladder, and in the upper wall of the membranous urethra was observed a fold similar to venous valves. In 1847, Bednar presented the case of a 12-day-old premature newborn, who died after 5 days of urinary retention, at necropsy it was discovered that the lower end of the verumontan was divided into two concave folds facing the bladder, the kidneys being atrophic and hydronephrotic. Later, several authors described the presence of posterior urethral valves, including: Godart (1854), Picard (1855), Jarjavay (1856), etc. The first scientific description of VUPs, their embryology and the role of these valves in urinary tract pathology was presented by Tolmatschew in 1870 [11, 14].

Various disorders associated with congenital urethral obstruction have been reported internationally, including: Cobb collar (1968), Moorman ring (1972), congenital obstructive membrane of the posterior urethra (COPUM), the differences between these concepts of disease remaining unclear [22]. In the early 1990s, Dewan et al. considered the term valve to be incorrect because this condition reflects obstruction in the posterior urethra of a single membrane, proposing the term COPUM. These authors were of the opinion that the type I and III valves after Young are identical, being confused during endoscopy, claiming that there are only 2 distinct entities, with different embryological origin, that cause obstruction of the posterior urethra: COPUM and Cobb collar [18, 22].

Classification

In 1913, Young H.H. and coauthor. published the presentation of a 20-month-old child with successfully treated VUP, and in 1919, trying to unify several developmental theories, described the famous work with the classification of these valves into three different types, including: Type I (90-95 %) - the presence of a ridge, which continues with the vera montanum and which protrudes anteriorly; Type II - retraction of the mucosa, which takes place proximal between the vera montanum and the bladder neck, behaving obstructively; Type III - presents as a diaphragm with a distal opening of the vera montanum, generating an obstruction on the entire circumference of the urethra. Subsequently, this classification was challenged [12, 21].

Although they believed that Tolmatchew's theory explained the development of type 1 valves, and Bazy's theory explained the appearance of type 3 valves, they favored the VUP development theory proposed by Watson (1918), believing that none of the proposed theories explained valves. type 2 [18].

Lately, more and more papers are using the classification proposed by Douglas Stephens, based on findings from urinary cystourethrography and endoscopic examination, according to which there are types 1, 3 and 4 of the posterior urethral valves and congenital bulbar urethral narrowing. Type 1 is defined as a valve structure, which connects directly from the vera montanum or lower ridge; type 3 is a ring-like structure located at the junction of the membranous and bulbar urethra, and a bent urethra associated with the prominence of the anterior wall of the posterior urethra has been classified as type 4. In cases where a ring-like structure has been located in the bulbar urethra, at the junction of the membranous and bulbar urethra is considered a congenital narrowing of the bulbar urethra [17].

In the opinion of some authors, a useful classification would be the one proposed by Hendren (1971), resumed by Caione et al., Who distinguish from the anatomical point of view and the clinical consequences two categories of VUP: "gentle shape" - 2 folds of mucosa with distal origin of vero montanum, forming two folds that partially obstruct the posterior urethra; „Severe form" - 2 wider folds of mucosa, with distal origin of vera montanum, but which join anteriorly, considerably narrowing the urethral lumen [27].

Embryology

The development of the male urethra begins with the expansion of the urogenital sinus cavity on the surface of the genital tubercle during the 6th week of gestation. Subsequently, this groove, which is an endodermal derivative, becomes a solid cell plate, which eventually tubules proximal-distally to form the phallic urethra, at week 14 the male urethra is fully developed [18]. The origin of the urethral plaque and its role in urethral development to date remains a topic of discussion, with some authors arguing that this plaque is the only precursor of the glandular urethra, with the rest of the

penile urethra developing from the urogenital sinus, while others claim that the urethral plaque it is the precursor of the entire penile urethra [16, 19].

VUP embryology is insufficiently understood, several theories have been proposed over the years. The hypothesis of an anomaly in the regression of the Wolf and Muler channel, reaffirmed by Stephens, seems the most accepted. It is claimed that the abnormal, too anterior insertion of the distal orifice of the Wolf canal and the migration anomalies, which occur much more sagittally than laterally, are the origin of the cup-shaped folds at the level of vero montanum and constitute type 1 valves [27, 28].

Clinic and diagnosis

VUP is associated with several morbidities, and the pathology is characterized by a spectrum of serious urological and renal consequences (hydronephrosis, urinary tract infection, urinary incontinence, sepsis, chronic kidney disease), in some cases fatal even in the prenatal or neonatal period [1, 4]. At the same time, in some cases, the condition remains asymptomatic, manifesting later with subtle signs and symptoms [13]. Some studies indicate that 20-65% of VUP patients will develop chronic kidney disease and 8-21% will progress to the final stage of kidney disease during childhood [8].

During the last 15 years, in CNȘPCP "Natalia Gheorghiu" 25 patients with urethral valves were diagnosed and treated, initially for which the diagnosis of congenital stenosis of the urethra was established (290 patients). The severity and degree of obstruction depended on the configuration of the obstructive membrane, the clinical symptoms being dominated by: fever, vomiting, palpable kidneys, bladder, dysuria, hematuria. Pollenuria, urinary incontinence, renal failure, urosepsis, stature-weight and psychomotor retardation have been associated with several young children.

VUP embryology is insufficiently understood, several theories have been proposed over the years. The hypothesis of an anomaly in the regression of the Wolf and Muler channel, reaffirmed by Stephens, seems the most accepted. It is claimed that the abnormal, too anterior insertion of the distal orifice of the Wolf canal and the migration anomalies, which occur much more sagittally than laterally, are the origin of the cup-shaped folds at the level of vero montanum and constitute type 1 valves [27, 28].

In all 25 cases, intrauterine ultrasound was performed, which revealed: bladder distinguished by hypertrophied walls (>3-4 mm), ureters and renal cavities dilated bilaterally, sometimes you could see the dilated urethra. The presence of focal renal parenchymal cystic formations, found in 6 cases, indicated renal dysplasia. At the same time, the presence of ascites and oligohydroamnios was diagnosed (13 cases).

In children of various ages, along with ultrasound, we used micturition cystourethrography, dynamic renoscintigraphy, which allowed us to assess renal function and highlight evacuation disorders, as well as cystoscopy for direct visualization of the valve positioned posteriorly. The early diagnosis in the prenatal period of this malformation is due to the presence of hydroureteronephrosis found on ultrasound examination, subsequently confirmed postnatal by micturition cystourethrography (fig. 1), which remains a method of choice in the diagnosis of VUP. In some cases, the diagnosis of VUP is established quite late: in children of various ages, in adolescence or even in adults [1, 24].

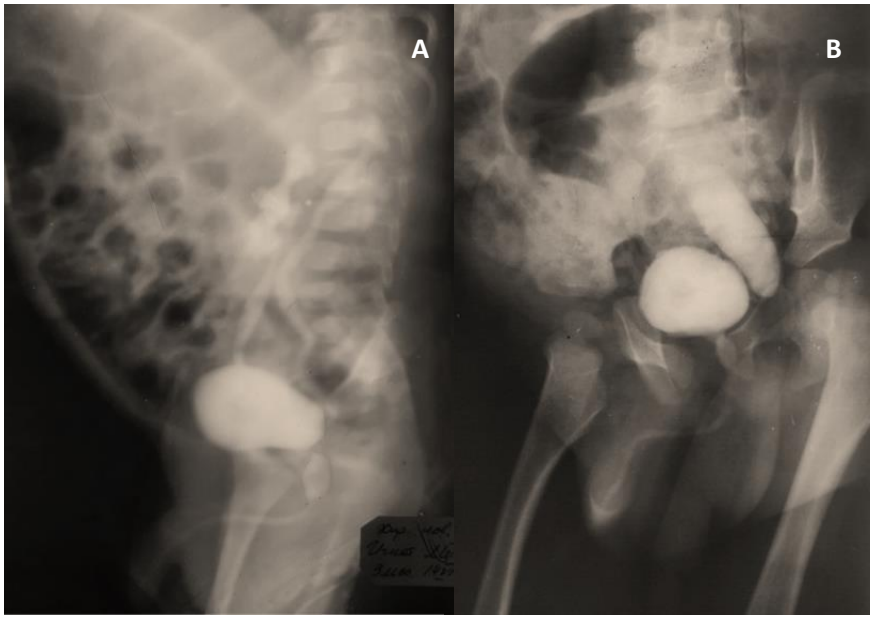

Fig. 1. Urinary cystourethrography: appearance of the posterior urethral valve diagnosed in the newborn (A) and infant (B)

Differential diagnosis includes: neurogenic bladder, Prune-Belly syndrome, bladder neck pathology, congenital stenosis of the posterior urethra, vesicoureteral reflux, primary megaureter, obstructive ectopic implantation of the ureters, stenosis or atresia of the ureters [3, 5, 15].

Cystography and cystocopy are essential in clarifying the diagnosis, more often confusing congenital stenosis with type III VUP, which on cystography presents as obstruction in the upper third of the posterior urethra with dilation of the proximal prostatic urethra, the bladder still having the appearance of "bladder the fight". To take into account the clinical signs present, more pronounced in VUP compared to stenosis, and urinary tract infection is characterized by severe evolution, often ending with renal failure. Usually, children with stenoses with urological accusations are addressed after the age of 1 year with urinary disorders [7, 9]. Urinary cystourethrography indicates obstruction of the distal part of the membranous urethra, dilation of the suprastenotic posterior urethra, bladder with pseudodiverticles, located more frequently on the posterior wall. Some authors describe a picture similar to micturition cystourethrography as stenosis, others as VUP. Until 1975, I also supported the idea promoted in the literature that the obstruction in the distal portion of the posterior urethra is type III valve after Young.

The open surgeries that we performed countless times due to urethral valves forced us to give up this method: during these operations we did not find such valves, mucosal folds, but we found small sectors of sclerotic urethra on a path of 0.2-0.4 cm in the form of a rigid ring.

A more in-depth study of cystourethrographies performed in patients with congenital stenosis and posterior urethral valves identified identical radiological signs for both pathologies, including: obstruction, proximal urethral obstruction dilation, narrowing and lengthening or dilation and dilation and shortening of the bladder neck, enlargement and change in the shape of the bladder, trabeculae and pseudodiverticles on the bladder wall. On cystography, the irregular bladder is observed, the side walls with clear, wavy contours, with prominences. On the lateral cystogram, the posterior wall has a trabecular appearance. At a more pronounced obstruction, at the same time with age, the capacity of the bladder, trabecularity and pseudodiverticles of the bladder walls also advance. Proximal to the obstruction, the prostatic urethra is dilated, the degree of dilation depending on the type and degree of obstruction.

Anatomically, VUPs are located along the proximal segment of the posterior urethra (prostate), in the region of the spermatic colic, upper or lower than it, and congenital stenosis affects the distal segment of the posterior urethra, which is why cystourethrography pathologies will manifest differently.

In micturition cystourethrography, in cases of VUP, the posterior urethra is dilated in the prostate portion, up to the level of the middle third, while in cases of stenosis - throughout, up to the level of the passage in the anterior portion (bulbar). In case of stenosis, the urethra is abruptly interrupted at the level of the obstacle in the form of a ring, while in the case of VUP the dilated urethra is observed above the obstacle, gradually narrows in the form of a funnel and almost constantly a diaphragm is present in the urethral lumen. stretched out. The distal urethra of stenosis can often be slightly dilated, which is never seen in the case of the valve [2, 28].

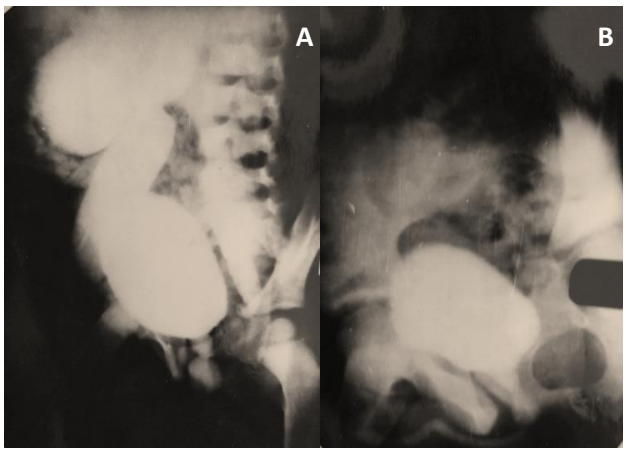

Fig. 2. Patient C., 3 years. Urinary cystourethrography: posterior urethral stenosis, suspicion of VUP, bilateral vesico-renal reflux gr. IV-V on the right, gr. IV on the left (A); B - radiological appearance over 10 after transperineal plasty of the urethra (intraoperatively VUP was not detected, being found urethral stenosis)

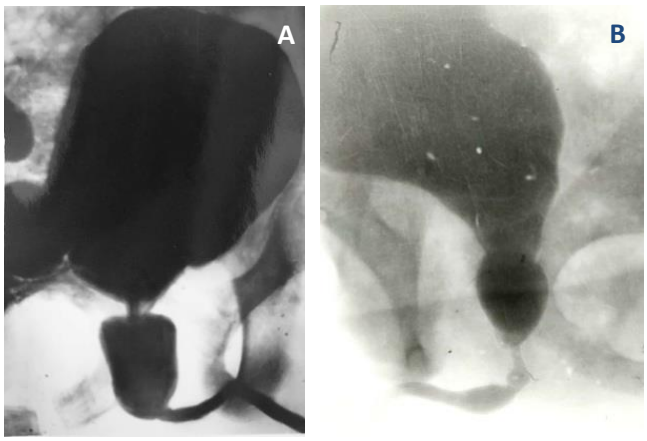

Fig. 3. A – Patient A., 4 years with congenital stenosis of the posterior urethra. B – Patient K., 6 years: posterior urethral valve

Evaluation of urodynamics is the only investigation, which allows to establish with certainty the diagnosis of overactive bladder, determining the direction of therapeutic conduct and timely monitoring of treatment outcomes [2, 28]. If the urodynamic changes are not obvious, the initial drug treatment is not necessary, but micturition education is used with the inclusion of behavioral psychological treatment, which aims to normalize bladder function and prevent functional disorders. In cases of obtaining negative results, it is possible to resort to applications of stimulation of presacral roots.

In some cases, pathophysiological changes in the bladder may develop after VUP ablation, which may be referred to as Valve Bladder Syndrome, first described by Mitchell M.E. (1982) [6]. This syndrome includes disorder of the bladder storage function that develops as a result of overcontractility associated with emptying disorders caused by bladder neck hypertrophy, which can result in ureterohydronephrosis and renal failure. Along with the dilation of the urinary tract, after the ablation of the valve in almost 50% of cases urinary incontinence develops, usually at a critical age of 5-6 years. In patients with ureterohydronephrosis without valves, bladder disorders were not found. I assume that these changes can be categorized into stages of the disease, which are present until ablation, becoming much more expressed after the age of 5-6 years. In some cases, in children with infravesical obstruction can be detected some various neurological disorders, found by us in 7 cases of the study group [10].

I would mention that the neurogenic dysfunction of the bladder, detected after the age of 4-5 years, can be present in younger children, but it is not always established due to an obvious symptomatology. Later, the children become more sociable, they alone can appreciate the urination disorders, the complications become more obvious, the parents addressing themselves with concrete accusations, which at the beginning they did not pay due attention to. The compensatory efforts made by the body to synchronize urinary propulsion reach a phase of exhaustion, followed by the onset of mild symptoms of urodynamic disorders, with chronic kidney disease, recurrence of urinary tract infection, caused by the consequences of neurogenic obstruction, intravesical hypertension, in the bladder and upper urinary tract, with the development of a dilated ureter, vesico-renal reflux.

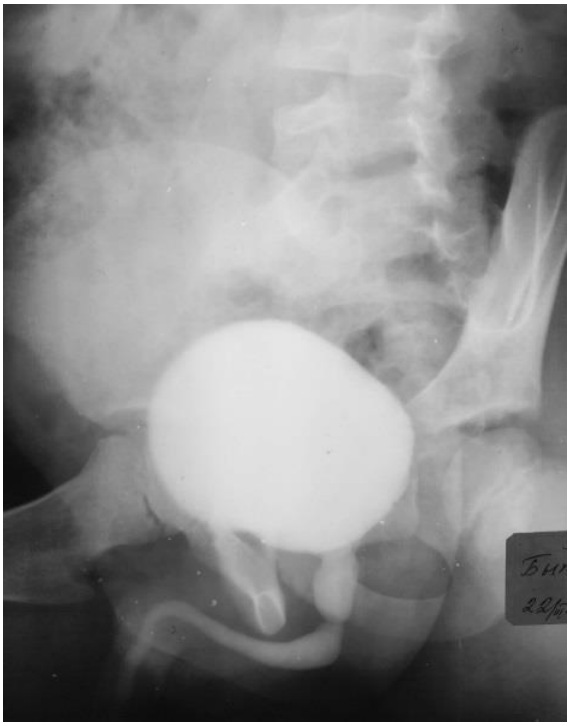

Fig. 5. Patient N., 3 years. Urinary cystourethrography: VUP; the prostate segment of the urethra is clearly dilated

Fig. 6. Patient V., 4 years. Urinary cystourethrography: stenosis of the posterior urethra. Posterior urethra dilated proximally by stenosis of the bulbar segment

Treatment

In the case of the newborn with VUP, it is recommended: to ensure the bladder drainage (gastric tube 5,6,8 Fr., suprapubic catheter); ensuring the venous approach; monitoring of renal function (serum creatinine value); ensuring the hydro-electrolytic balance (monitoring of transient polyuria, correction of metabolic acidosis; prophylaxis / treatment of UTI; endoscopic resection of VUP / temporary urine derivations [27].

Endoscopic valve ablation is the postnatal treatment of choice for VUP, aiming to resolve the obstruction, maintain renal function and achieve adequate urinary flow. However, lower urinary tract dysfunction is a common finding after VUP ablation in children [24]. Although after the correction of the valve and neurogenic dysfunction the kidney function improves, in some cases postoperatively the reflux on the contralateral side can develop. In cases where urethral obstruction predominates, signs of diurnal urination are associated with enuresis. The child tries to prevent this by restraint maneuvers, by voluntary contraction of the pelvic floor, urine is eliminated in small amounts so drink less fluid. Subsequently, muscle spasm, present in the first months of life, is associated with urinary tract infection and urinary tract dilation.

Long-term treatment with anticholinergic medication (oxybutynin 0.1-0.15 mg / kg 2 times / day) can contribute to the development of constipation, increased bladder residue, dry mucous membranes, sometimes aggression

Endoscopic treatment, especially in cases of reflux, did not positively influence the improvement of functional results. For this reason, it is recommended to initially remove the obstruction, then to resort to non-implantation or the simultaneous performance of both procedures. We performed the valve ablation with the urethoma proposed by us, which turned out to be quite safe. Removal of the valve can be performed transvesically with or without sectioning of the pubic symphysis (Gross), transsymphysis with the opening of the posterior urethra (Millin) or with the help of a hard probe with perineal destruction, endoscopic resection or retrograde bladder, balloon catheter crushing. We note that the original urethrotome proposed allows the ablation of the valve practically without complications in 4-5 min. Potential complications may include persistence of the valve and scarring of the resection, in some cases a significant increase in urinary flow.

In severe cases, surgical treatment includes a prior derivation of urine by cystostomy or transcutaneous nephrostomy. After 4-6 weeks, valve excision was used, using one of the suprapubic, perineal or endoscopic resection. In cases of reflux, the ureters were reimplanted, using the anti-reflux procedure after Cohen, Leadbetter Politano, which allowed the shortening and narrowing of the dilated ureters. Nephrectomy was necessary in one kidney with marked hydronephrotic atrophy, the other kidney having a satisfactory function. In the postoperative period, treatment with uroseptics and periodic control is indicated, which will include urine culture, origography, cystography, ultrasound, renoscintigraphy. Assessment of serum urea and creatinine levels is a true prognostic indicator in the postoperative evolution of VUP [24].

Conclusions:

- Posterior urethral valves are considered as one of the most common causes of infravesical obstruction, probably in the top of the valves are included SCUP (organic and neurogenic form) as type IIIa of the valves.
- Stenosis is located in the distal part of the posterior urethra with dilation of the urethra throughout; in cases of VUP the obstruction is located in the proximal, prostatic part of the urethra with dilation of the urethra in the proximal segment.
- Suprastenotic urinary tract disorders are identical. From our experience we found that the clinical evolution in VUP is comparatively more serious, it is highlighted earlier in newborns and infants, while in cases of stenosis - more frequently after 1 year.
- Also due to the inclusion of SCUP as valves, neurological signs appear in these patients at 5-6 years of age. According to our data, they are also present in newborns and infants, not being pronounced, we do not diagnose them, we do not treat them.
- These neurological signs with age worsen, the child at 5-6 years becomes more sociable, can alone appreciate urinary disorders, on examination the complications are obvious.
- The treatment of choice in VUP is transurethral resection.
- All patients with VUP need long-term monitoring until adolescence. The prognosis depends on the suprastenotic changes and the degree of bladder dysfunction at the address.

REFERENCES

1. Alsaywid B.S., Mohammed A.F., Jbril S.M., Bahashwan M., Mukharesh L., Al Khashan M. Renal outcome among children with posterior urethral valve: when to worry? Urol. Ann. 2021. 13(1):30-5.
2. Androulakakis P.A. Miogenic bladder decompensation in boys with posterior uretral valves may be secondary to bladder neck obstruction BJU Int. 2005. 1:140-3.
3. Bernardes L.S., Aksnes G., Saada J., Masse V., Elie C. et al., Keyhole sign: how specific is it for the diagnosis of posterior urethral valves? Ultrasound Obstet. Gynecol. 2009. 34(4):419-23.
4. Bilgutay A.N., Roth D.R., Gonzales E.T., Janzen N., Zhang W. et al. Posteior urethral valves: rosk gactors for progression to renal failure. J. Pediatr. Urol. 2016. 12(3):179-
5. Bulucea D., Georgmaneanu M. Manual de pediatrie. 2000. Vol. .II. 241 p.
6. Chan E.P., Wang P.Z.T., Dave S. Valve bladder syndrome associated with posterior urethral valves: natural history, work-up, and management. Curr. Bladder Dysfunct. Rep. 2020. 15:76-82. https://doi.org/10.1007/s11884-020-00577-1
7. Ciofu E.P., Ciofu C. Tratat de pediatrie Ed.1. 2001. pp.726-8.
8. Coquillette M., Lee R.S., Pagni S.E., Cataltepe S., Stein D.R. Renal outcomes of neonates with early presentation of posterior urethral valves: a 10-year single center experience. J. Perinatol. 2020. 40(1):112-7.
9. Curajos B. Diagnosticul și tratamentul chirurgical diferenciat al stenozei congenitale a uretrei posterioare și consecințelor ei la copii. Autoref. tezei de doctor hab. în șt. med. Chișinău. 1997. 28 p.
10. Curajos B. Compendiu de urologie pediatrică. Chișinău. 2018. p.202-6.
11. Dewan P.A. Congenital posterior urethral obstruction: the historical perspective. Pediatr. Surg. Int. 1997. 12:86-94.
12. Heikkilä J. Posterior urethral valves. Early features and renal outcome. Academ. Diss. Helsinki 2015. 69 p.
13. Herbs K.W., Tomlinson P., Lockwood G., Mosha M.H., D'Alessandri-Silva C. Survival and kidney outcomes og children with an early diagnosis of posterior urethral valves. Clin. J. Am. Soc. Nephrol. 2019. 14(11):1572-80.
14. Hodges S.J., Patel B., McLorie G., Atala A. Posterior urethral valves. Sci. World J. 2009. 9:1119-26.
15. Hydrycks A., Delatte L. Prenatal diagnosis of congenital renal and urinary tract malformation. Facts View. Vis. Obgyn. 2011. 3(3):165-74.
16. Hynes P.J., Fraher J.P. The development of the male genitourinary system: II. The origin and formation of the urethral plate. Brit. Assoc. Plast. Surg. 2004. 57:112-21.
17. Kihara T., Nakai H., Mori K., Sato R., Kihara S., Yasuda K. Variety of congenital urethral lesions in boys with lower urinary tract symptoms and the results of endoscopic treatment. Int. J. Urol. 2008. 15(3):235-40.
18. Krishnan A., De Souza A., Konijeti R., Baskin L.S. The anatomy and embryology of posterior uretral valves. J. Urol. 2006. 175:1214-20.
19. Levin T.L., Han B., Little B.P. Congenital anomalies of the male urethra. Pediatr. Radiol. 2007. 37:851-62.
20. Manzoni C., Valentini A.L. Posterior urethral valves. Rays. 2002. 27(2):131-4.
21. Mohamed A.O., Eradi B., Owen A., Rajimwal A. Rare associations with posterior urethral valves. Hindawi Case Rep. Urol. 2021. Art. ID 6647692. 4 p. https://doi.org/10.1155/2021/6647692.
22. Nakamura S., Hyuga T., Kawai S., Kubo T., Nakai H. The endoscopic morphological features of congenital posterior urethral obstructions in boys with refractory daytime urinary incontinence and nocturnal enuresis. Eur. J. Pediatr. Surg. 2016. 26(4):368-75.
23. Nasir A.A., Ameh E.E., Abdur-rahman L.O., Adeniran J.O., Abraham M.K. Posterior urethral valve. Word J. Pediatr. 2011. 7(3):205-16.
24. Shields L.B.E., White J.T., Mogamed A.Z., Peppas D.S., Rosenberg E. Delayed presentation of urethral valves: a diagnosis that should be suspected despite a normal prenatal ultrasound. Glob. Pediatr. Health. 2020. 7:2333794X20958918. doi: 10.1177/2333794X20958918.
25. Talabi A.O., Sowande O.A., Etonyeaku A.C., Salako A.A., Adejuyigbe O. Posterior urethral valves in children: pattern of presentation and outcome of initial treatment in lle-Ife, Nigeria. Niger. J. Surg. 2015. 21(2):151-6.
26. Tambo F.F.M., Tolefac P.N., Ngowe M.N., Minkande J.Z. et al. Posterior urethral valves: 10 years audit of epidemiologic, diagnostic and therapeutic aspects in Yaoundé gynaeco-obstetric and paediatric hospital. BMC Urology. 2018. 18:46. https://doi.org/10.1186/s12894-018-0364-1.
27. Țîrlea S., Ionescu S. Considerente privind diagnosticul și tratamentul primar al valvelor de uretră posterioară. Chirurgia. 2012. 107:218-25.
28. Гурская А.С. Диагностика и лечение новорождннных и детей раннего возраста с клапанами задней уретры. Андрол. и Генит. Хир. 2014. 15(2):44-7.

Conflicts of interest: *authors have no conflict of interest to declare*