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Case report

Abordagem diagnóstica e terapêutica da válvula de uretra posterior: relato de caso

Diagnostic and therapeutic approach of the posterior urethral valve: case report

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Resumo

Objetivo: realizar a descrição de um relato de caso de paciente com quadro clínico de válvula uretral posterior. **Materiais e Método:** o estudo foi elaborado por meio da coleta de informações presentes em relatórios, laudos, exames complementares e alta hospitalar em posse do responsável pelo paciente. Para a confecção da discussão, foram realizadas buscas, na plataforma PUBMED, de artigos abordando o tratamento de casos clínicos similares. Foram utilizados os descritores em português e inglês: *child*, *urethral obstruction* e *congenital abnormalities*. Para a seleção dos artigos, foram utilizados os critérios de inclusão: artigos disponíveis na íntegra, relatos de casos com tratamento com acompanhamento por, pelo menos, três meses. Os critérios de exclusão foram: artigos envolvendo adultos e conteúdo abordando apenas a fisiopatologia da doença. **Relato de caso:** gestante com 28 semanas e seis dias realizou exame ecográfico obstétrico que identificou pieloectasia renal e hidronefrose bilaterais em feto masculino. Optou-se por tratamento conservador até o pós-natal. O paciente nasceu a termo sem intercorrência, mas apresentou infecção do trato urinário (ITU) aos 12 meses, confirmada por exames laboratoriais. A ultrassonografia indicou válvula de uretra posterior, após confirmada por uretrocistografia miccional e retrógrada (UCM). Realizou-se ablação endoscópica primária de urgência. Após 15 dias, houve ITU e a repetição de UCM identificou alterações remanescentes, indicando necessidade de nova cirurgia. **Conclusão:** o diagnóstico precoce em período pós-natal promove impactos positivos na vida do paciente, visto que evita danos renais permanentes na criança.
Palavras-chave: Criança. Obstrução Uretral. Anormalidades Congênitas.

Abstract

Objective: describing case studies of a patient with a clinical presentation of posterior urethral valve. **Materials and Methods:** there was compiled through the collection of information from reports, medical records, complementary exams, and hospital discharge documents in the possession of the patient's guardian. For the discussion, a literature search was conducted on the PUBMED platform using articles addressing the treatment of similar clinical cases. The descriptors used in Portuguese and English were: child, urethral obstruction, and congenital abnormalities. Inclusion criteria for article selection were: full-text articles, case studies with treatment and follow-up for at least 3 months. The exclusion criteria were: articles involving adults and content focusing solely on the pathophysiology of the disease. **Case Report:** a pregnant woman at 28 weeks and 6 days underwent a routine obstetric ultrasound, which identified renal pyelectasis and bilateral hydronephrosis in a male fetus. Conservative treatment was opted for until the postnatal period. The patient was born at term without complications but presented with a urinary tract infection (UTI) at 12 months, confirmed by laboratory tests. Ultrasound indicated a posterior urethral valve, which was subsequently confirmed by voiding cystourethrogram (VCUG). An emergency primary endoscopic ablation was performed. After 15 days, another UTI occurred, and a repeat VCUG identified remaining abnormalities, indicating the need for another surgery. **Conclusion:** early diagnosis in the postnatal period has positive impacts on the patient's life as it prevents permanent renal damage in the child.
Keywords: Child. Urethral Obstruction. Congenital Abnormalities.

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Introduction

The posterior urethral valve (PUV) is a rare congenital abnormality that affects the male urinary system. It is usually diagnosed in male infants during the neonatal phase or late in young children when symptoms are manifeste[1,2]. It is the most common cause of congenital urethral obstruction in children, with an incidence of 1:3000 to 1:8000 live births, and 2 to 8 new cases in pediatric urology reference units per year[3].

This condition is characterized by the presence of an anomalous membrane located in the posterior urethra, which causes obstruction of urine flow. Obstruction caused by PUV can lead to several complications in the urinary system, including hydronephrosis, bladder dilation, vesicoureteral reflux renal insufficiency[1,4,5].

Posterior urethral valves continue to be an important cause of morbidity, evolving into chronic kidney disease (CKD) in approximately 30% of cases, with terminal CKD in about 25% at puberty[6].

Diagnosis at the initial stage of the disease and effective treatment of PUV are essential to prevent permanent kidney damage and bring improvement in patient's quality of life[7]. Treatment usually involves surgical intervention to remove or relieve the obstruction of the urethra and correct structural abnormalities.

In this case studies, the history, clinical examination, diagnostic methods, therapeutic approach and follow-up of a patient with posterior urethral valve are presented. In addition, the challenges faced during diagnosis, surgical intervention and clinical outcomes are highlighted.

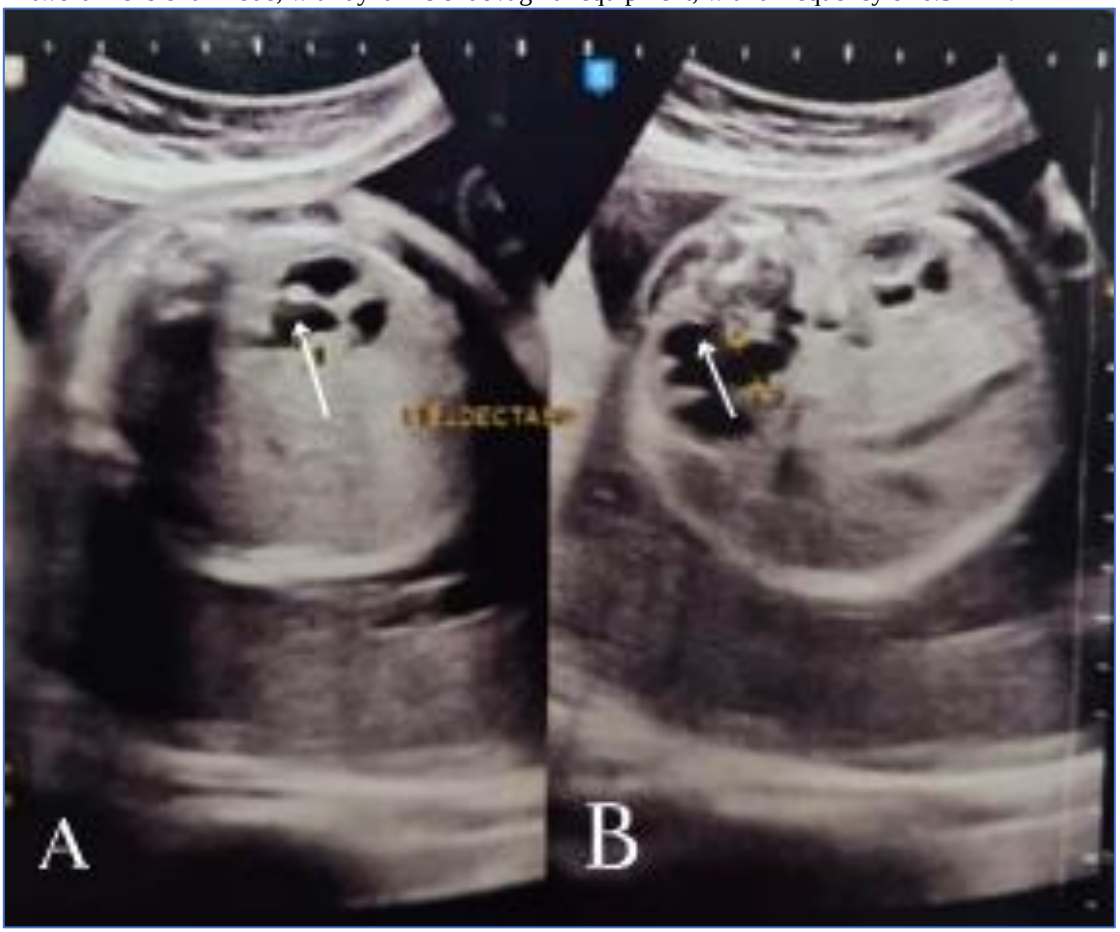
The study was approved by the institution's Research Ethics Committee under CAAE: 75090623.0.0000.5141.

Case report

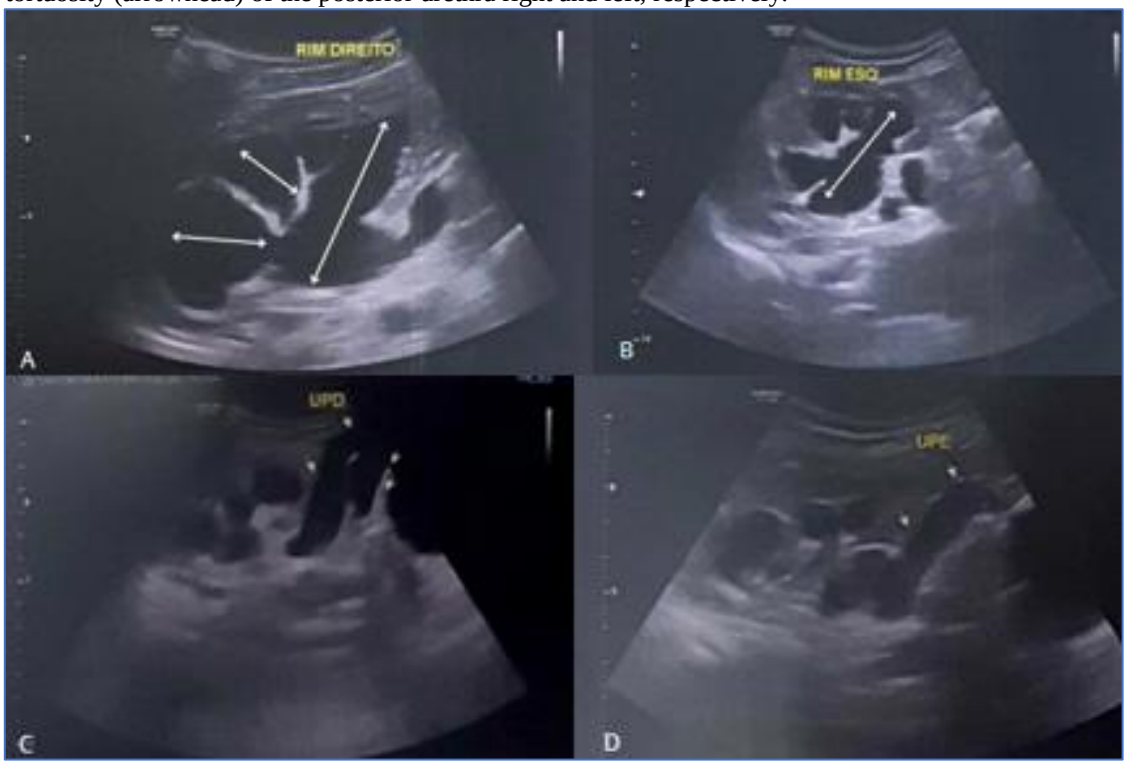
Pregnant woman with 28 weeks and 6 days performed routine obstetric ultrasound, which identified bilateral renal pyeloctasia and hydronephrosis (Figures 1A and 1B) in male fetus. At that time, it was decided to carry out conservative treatment until the postnatal period.

The patient was born at term by delivery via transvaginal without complications, weighing 3600g, measuring 49 cm and Apgar 9/10. At 1 year of age, he presented a urinary tract infection (UTI) confirmed by laboratory tests and considerable weight loss. He underwent an ultrasound of the kidneys and urinary tract, which showed the permanence of renal pyeloectasia and hydronephrosis, suggesting the presence of posterior urethral valve (Figures 2A, 2B, 2C and 2D).

Figures 1A and 1B. Obstetric ultrasound showing bilateral renal pyeloectasia. The exam was performed in two-dimensional mode, with dynamic endovaginal equipment, with a frequency of 6.5MHz.



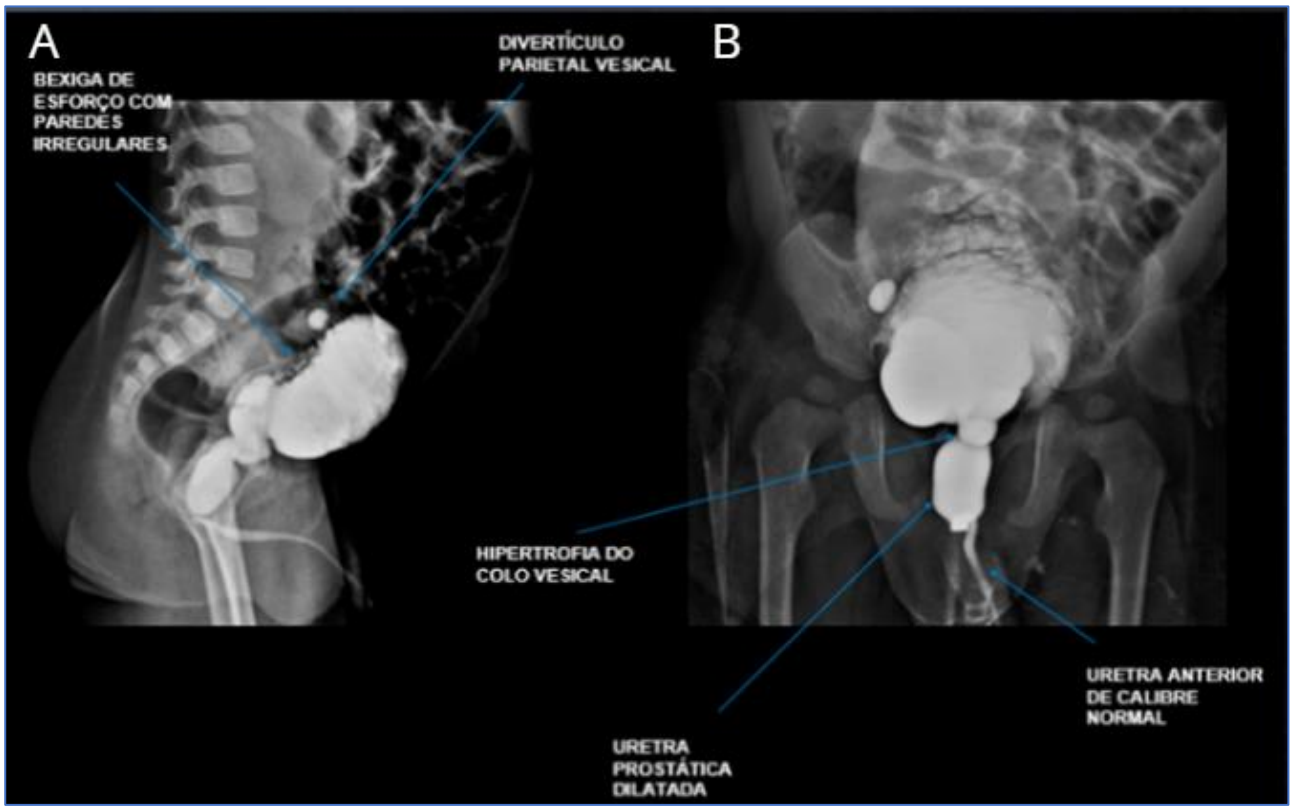
Figures 2A, 2B, 2C and 2D. Ultrasonography of kidneys and urinary tract. Examination performed with convex transducer at the frequency of 2.0 to 5.0 MHz. 2A and 2B show the presence of pronounced bilateral pyelocalyceal dilatation (double arrows), with right accentuation. 2C and 2D show dilatation and tortuosity (arrowhead) of the posterior urethra right and left, respectively.



In sequence, a retrograde mictional urethrocystography (VUCG) was requested, which could not be performed due to the non-exposure of the glans of the penis. Within a month, the urinary tract infection cases became recurrent, with history of hospitalizations and repeated use of antibiotic therapy.

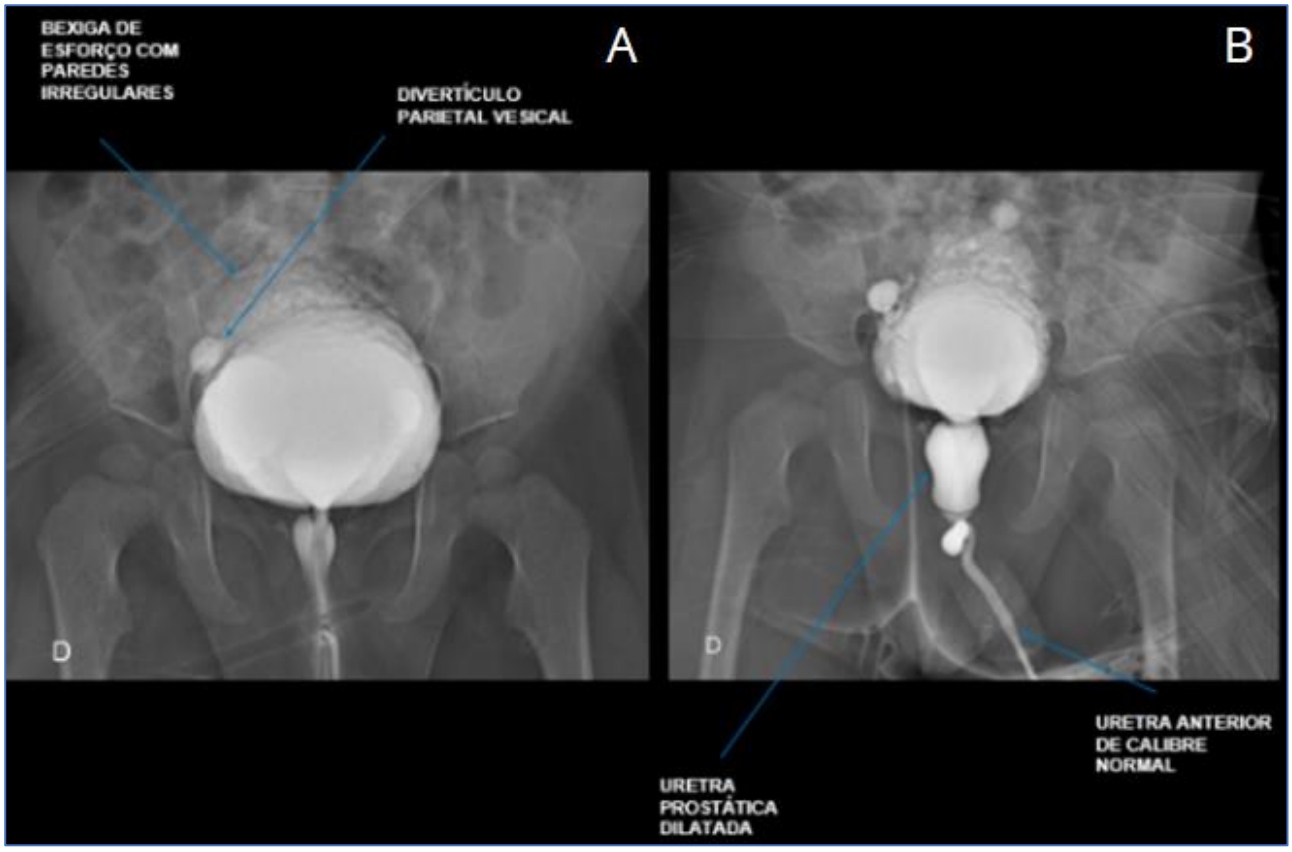
Subsequently, VCUCG was performed confirming the diagnosis of posterior urethral valve (Figures 3A and 3B). Therefore, an emergency surgical approach was performed using the primary endoscopic ablation technique. After 15 days postoperative, after spontaneous fall of the vesical delay probe, he presented sudden fever and UTI confirmed and subsequently treated.

Figures 3A and 3B. Diagnostic voiding cystourethrography demonstrating alterations resulting from the posterior urethral valve.



The VCUG examination was repeated, identifying residual alterations of the posterior urethral valve (Figures 4A and 4B) compatible with the diagnosis of a straining bladder. A second surgical approach was recommended by the specialist.

Figures 4A and 4B. Postoperative voiding cystourethrography showing alterations remaining from the PUV.



Discussion

Regarding the diagnosis, the gold standard method for PUV diagnosis is the retrograde urinary urethrocystography, which should be performed in the postnatal period, immediately when the clinical conditions allow. However, the diagnostic suspicion is made through obstetric ultrasound, usually performed in the 2nd trimester, between the 20th and 24th weeks of pregnancy. In newborns with a suspected prenatal diagnosis of posterior urethral valve, ultrasonography should be performed soon after birthing.

In the VCUG, dilated and elongated posterior urethra, increased and irregular bladder and ureters, hypertrophied vesical neck, little distal flow, and vesicouteral reflux in about half of the patients[9-11].

In addition to the tests performed in this case, other tests such as renal scintigraphy and urodynamic studies may be complementary in the postnatal diagnosis of PUV. Renal scintilography, when indicated, should be performed after 14 days of life and in premature infants after 60 or 90 days of life. This provides information on renal excretion, glomerular filtration and proximal tubular function of the kidneys. These tests are indicated in clinical control after initial or definitive treatment[3,10].

As the diagnosis can only be confirmed after birth, the newborn may present signs and symptoms resulting from the change until the therapeutic approach is performed. Urinary infections, fever, urine retention, septicaemia, weight loss, growth impairment and hydroelectrolyte disorders are common signs and symptoms in neonates with untreated PUV[12]. In the case presented, the patient continued to present such signs and symptoms even after treatment.

There are no indications for prenatal intervention. However, some authors advocate intervention in cases where posterior urethral valves were diagnosed in the second trimester with evidence of oligohydronic gravinium[13].

Among the differential diagnoses, the anterior urethral valve and the urethral diverticula should be considered, which have a frequency eight times lower than that of the posterior urethral valves[14].

The therapeutic approach to postnatal patients with PUV aims to solve the obstruction of the urinary tract and prevent complications arising from the disease. The cystoscopic valve ablation is the current initial treatment of choice for posterior urethral valves[13,15]. In stable patients, born to term, the surgical procedure begins with calibration and dilation of the urethra by means of plastic urethral catheter. Cauterization of the valves should be completed in the posterior and anterior region of the urethra to ensure complete relief from clogging[15].

As the patient frequently presented with urinary infection, it was necessary to investigate PUV through the VCUG, which gave the definitive diagnosis, allowing the therapeutic approach. VCUG should be performed without hesitation to diagnose when there is suspicion of urethral lesion, at least by abnormal ultrasound findings.

Table 1 presents reports of similar cases found in the literature.

Table 1: Studies of similar cases found in literature.

Authors	Year	N. of patients	Signs and Symptoms	Treatment	Outcome
Tucci Jr. S et al.[20]	2003	1	Drip diuresis, without mictional jet, having been observed bulging of the base of the penis during urination, evolved with septicemic picture, diagnosing urinary infection and pionefrose to the right.	Submitted to exploratory lombotomy, right upper polar nephrectomy was performed with ureterectomy followed by vesicostomy. After 28 months, endoscopic transurethral fulguration of the anterior urethral valve and concomitant closure of the vesicostomy were performed.	Showed good evolution in the postoperative period. Normalization of the lower urinary tract in the examination of urinary urethrocystography. Ultrasonography revealed spontaneous reduction of the diverticulum in the urethra and persisted with slight pyelocaliceal dilation.

Kiliś-Pstrusińska K et al.[23]	2013	1	Previously healthy 6-week-old boy presented fever, vomiting and oliguria persisting for several hours. The abdomen was distended, tense and sensitive to palpation.	Treatment was started with cefotaxime, vancomycin, dopamine and furosemide. Due to the worsening of the patient's condition, he was qualified for an emergency laparotomy. After stabilization, the cystoscopy revealed urethral structures that could correspond to the posterior urethral rupture. Valve ablation was performed.	Spontaneous renal rupture was retrospectively diagnosed due to posterior urethral valves. At 20 months of follow-up, the ultrasonographic examinations revealed slight and persistent dilation of both pelvic-caliteal systems. The history of urinary tract infection was negative.
Alencar VCS, Duarte CS, Pacheco GA.[19]	2022	1	Weak and intermittent urine jet, urinary leakage and recurrent urinary infections.	Oxybutynin as an anticholinergic, endoscopic evaluation of the urethra, through cystoscopy, which visualized many trabeculations in the bladder, however, there are anatomical obstruction points.	Improvement of micturition urgency, loss in the intervals of urination and reduction of urinary frequency.
Nishio H. et al.[21]	2022	1	Effort to urinate	A transurethral incision was performed in the valvar lesion at 12 hours with a cold scalpel. A Foley catheter was inserted and the operation was completed.	Two years after the operation, his effort to urinate disappeared and there was no recurrence of the urofluxometry findings.
Ikhuoriah T et al.[22]	2023	1	Urinary retention and recurrent fevers since childhood. The physical examination revealed still slight abdominal distension and suprapubic sensitivity.	The patient underwent a valvotomy.	Medical follow-up was neglected after hospital discharge.

The postoperative period can last from a few days to weeks depending on the type of abnormality associated with PUV. Most patients have successful valve ablation after a single approach. However, among patients undergoing surgical treatment, about one third will need a new surgery to achieve the desired result[16,17], as in the case studies mentioned above.

In the postoperative follow-up, the patient should be submitted to a new urethrocystography within one or two months after the first procedure, to identify and treat any remaining abnormalities[10,16].

Another surgical technique is urinary bypass through the vesicostomy. This surgical method is not advantageous when compared to the primary endoscopic ablation technique, because when compared for postoperative complications, the mortality rate and renal function preservation did not show superior results. Vesicostomy is indicated in the group of patients with severe infectious disease,

despite the efficient endoscopic ablation and clearing surgery, not corresponding to the case addressed, where the endoscopic technique was the most advantageous[18].

Conclusion

It is therefore deduced that early diagnosis of PUV, through retrograde urinary urethrocystography imaging in the postnatal period, has positive impacts on the patient's life, since early diagnosis prevents permanent kidney damage.

Despite the possibility of identifying changes in the prenatal period through USG of 2nd trimester, interventions considered safe are performed after birth, since there are no indications of prenatal approach.

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