Original Article

Bladder dysfunction in pelvic urinary tract obstructions Congenital posterior urethral stenosis in boys. Part I

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Abstract

Disfuncția vezicii urinare în obstrucțiile tractului urinar pelvin
Stenoza congenitală uretrală posterioară la băieți. Partea I

Stenoza congenitală a uretrei posterioare se întâlnește frecvent și duce la tulburarea întregului tract urinar - uretra suprastenotică, colul vezicii urinare, vezica urinară, ureterohidronefroză, mai frecvent bilateral. Având origine congenitală, această malformație rezultă din dezvoltarea incompletă a membranei urogenitale - limita pasajului uretral posterior și anterior și spasmul muscular al tractului urinar pelvin.
Diagnosticul include ultrasonografie, urografie, cistouretrografie micțională, renoscintigrafie dinamică, cistometrie, urofluometrie radionuclidică, uretroscopie, calibrarea uretrei cu buj cu olivă, examen neurologic, miografie musculară perineală. A fost elaborată clasificarea clinico-radiologică a acestui tip de stenoze, fiind descrise diferite grade, care necesită un tratament diferențiat în funcție de fazele clinic-radiologice. Stenoza neurogenă se dezvoltă la copiii mai mici cu un tablou clinic mai sever comparativ cel forma congenitală.
Rezecția transuretrală a peretelui anterior al uretrei stenotice este metoda de elecție în tratamentul stenozei uretrale.

Cuvinte cheie: afecțiune renală cronică, valvă uretrală, diagnostic imagistic, ablația valvei

Abstract

Congenital posterior urethral stenosis is common and leads to disorder of the entire urinary tract - suprastenotic urethra, bladder neck, bladder, ureterohydronephrosis, more often bilaterally.
It can be congenital as a result of incomplete development of the urogenital membrane - the limit of the posterior and anterior urethral passage and the muscular spasm of the pelvic urinary tract.
Diagnosis includes ultrasound, urography, micturating cystourethrography, dynamic renoscintigraphy, cystometry, radionuclide uroflowmetry, urethroscopy, calibration of the urethra with olivary bougies, neurological examination, perineal muscle myography. The clinical-radiological classification of stenosis was developed, which is more didactic. There are different degrees of stenosis as suprastenotic resonance that requires a differentiated treatment depending on the clinical radiological phases. Neurogenic stenosis develops in younger children with a more severe clinical picture than the congenital one.
Transurethral resection of the anterior wall of the stenotic urethra is the method of choice in the treatment of stenosis.

Keywords: chronic kidney disease, congenital obstructive uropathy, urethral valve, imaging diagnosis, valve ablation

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Introduction

Restoration of the upper urinary tract by antireflux surgery is performed after removing the obstruction, carrying out intravesical surgery; in neurogenic stenosis spasmolytics are administered, neurological observation and treatment is required [5, 6].

Obstructive bladder dysfunction is a urodynamic disorder of the urinary tract. An in-depth study of congenital posterior urethral stenosis performed by Stefens F. (1963), Sow (1968), and especially Moormon J. (1976), etc., was based on the interpretation of the etiology, clinic and diagnosis of obstruction. This is of paramount importance in establishing the tactics, the use of either one or another method of treatment.

There are still a number of unresolved questions so far, regarding early diagnosis, therapeutic tactics, prognosis of complications, consequences. There is lack of data on the clinical-anatomical picture of congenital stenosis in the literature, concerning the degree of stenosis and changes in the suprastenotic sectors of the urinary system. Moorman (1977) described the radiological phases of congenital stenosis of the bulbar urethra, considering only changes in the upper urinary tract. However, Moorman did not provide data on changes revealed by cystourethrography, which depend on the degree of stenosis and the compensatory capacities of the suprastenotic segment of the urethra, bladder neck, etc. The reciprocal relationship between the timely diagnosis and differentiated surgical correction according to the case help control the pathological process [3].

Purpose of the paper: to elaborate a complex of diagnostic tests and differentiated surgical management for the correction of congenital stenosis of the posterior urethral spasm and its consequences in children.

If the disorders are persistent, especially if they are associated with urinary tract infections, it is necessary to determine the anatomical and neurological causes [4].

The work is based on the results of clinical observation, methods and results of complex surgical treatment of a group of 265 boys with congenital urethral stenosis, the age ranging in newborns, infants and 15-year old adolescents, treated in the Department of Urology and Neonatal Surgery of the Republican Clinical Hospital „E. Coțaga" and HCF IM and C during 1970-2018.

Patients were examined pre-, intra- and postoperatively according to a pre-established protocol. The "Patient Observation Sheet" and the "Patient Outpatient Sheet" were used as sources of information. It hould be mentioned that the only selection criterion was the diagnosis of infravesical obstructive syndrome. A thorough urological examination was performed in newborns with dilation of the upper urinary tract detected intrauterinely.

If there are 3 cases of correctly diagnosed urinary tract infection in girls and 2 cases in boys, it is necessary to carry out a thorough examination (many years of experience) of urodynamics in order to reveal the factors involved in filling, storing, transporting and evacuation of a part of the lower urinary tract. In some cases ultrasound, micturating cystourethrography, i/v urography, dynamic renoscintigraphy, and radionuclide cystourethrography with the examination of bladder and urethra urodynamics as well as cystometry are carried out.

If the urethra cannot be examined by the cystourethrography, in girls the urethra is calibrated with normal diameter bougies according to age, in boys with olivary bougies, to exclude meatal stenosis in girls or posterior membranous urethra in boys.

Congenital posterior urethral stenosis (CPUS) is often confused with form III valves, the treatment of which differs. Frequently, open perineal urethrotomy does not detect any mucosal folds in the urethral lumen that can lead to obstruction.

There were attempts to introduce new terms and new methods of treatment. Thus, Dewan and Ransley (1992) proposed a new term "Obstructive posterior urethral membrane". Courajos B.M. (2019) - Congenital posterior urethral stenosis, incomplete development of the urogenital membrane - the limit of the posterior urethral passage to the anterior one, Cobb`s collar (Moorman`s ring) -1975. Muscle spasm of the pelvic urinary tract.

Material analysis

Based on the data presented as well as the clinical examination data, it was made an attempt to demonstrate the mechanism of evolution of suprastenotic changes of the entire urinary system, the latter being dependent.

However, suprastenotic disorders in urethral obstruction are manifested by urodynamic disorders and infection association. In urethral obstruction, the more marked congenital stricture is, the more forced urination is. In most cases this fact is overlooked by both parents and children. As a reaction to stricture, the urethra dilates, the urine flow becomes turbulent, the retrograde parietal movement occurs, causing urethritis and cystitis. The bladder infection penetrates the upper urinary tract, thus causing chronic pyelonephritis. The bladder becomes hypertrophied, the infravesical pressure is increased, manifesting clinically by pollakiuria and enuresis. This represents the urination left in the first stage of its evolution. The striated sphincter does not intervene at all and involuntary urination occurs diurnally. Decompensation of the bladder and bladder neck subsequently causes forced urination, with residual urine and *ischuria paradoxa.*

Particular attention is paid to vesicoureteral reflux, which can accompany chronic cystitis, hyper- and hypotonia of the bladder. This causes chronic pyelonephritis and dilation of the upper urinary tract. It should be mentioned that in case of bilateral vesico-renal reflux, the suprastenotic disorders of the urethra, bladder neck and upper bladder are not so marked even if stenosis has a pronounced character.

It was found that in the suprastenotic segment of the posterior urethra during urination, the tension increases and the urethra dilates, initially the posterior wall, then the entire posterior urethra does. The retrograde parietal turbulent movement in the urethral lumen favors urethral dilation, bringing about urethritis, prostatitis or cystitis. The turbulent urine flow can only be observed in isotope uroflowmetry.

The upper urinary tract undergoes the same changes as the bladder, namely, lesions that can be detected sonographically, urographically and by radionuclide scanning. The ureter, the renal pelvis and the calyx system become hypertrophied, trying to overcome the vesicoureteric hypertension, then become hypercontracted, producing ureterohydronephrosis that distorts the structure of the renal parenchyma.

The longer the duration and degree of hypertension, the more impaired the kidney function is, and it does not depend only on the child`s age. This condition can be temporarily remedied by the presence of an extrarenal pelvis, acting as an expansion vessel. Impaired kidney function occurs as a consequence of the two mechanisms - ischemia and infection. It should be mentioned that the initial kidney injuries are reversible, and advanced injuries do not have the chance to recover.

The ureters undergo morphological changes that represent the pathophysiological mechanisms of high resonance of the posterior urethral stricture (fig. 1).

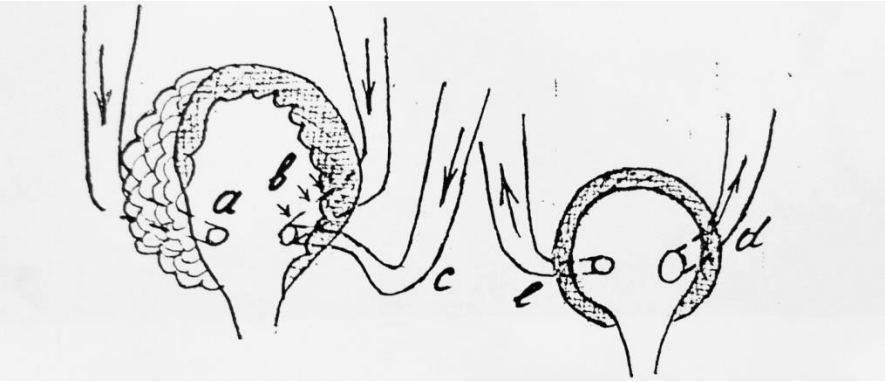

Fig. 1. Mechanism of urethral disorders in infravesical obstruction:
a - compression of intramural ureters by hypertrophy, the detrusor spasm (initial mechanism);
b - difficult ureteral discharge, caused by intravesical hypertension following urine stagnation in the bladder;
c - fishhook ureters - bladder dilation and trigone lifting cause the ascent of the terminal part of the ureters, which angles at the vas deferens crossing

Retrograde reflux is due to:

- meatal insufficiency in intravesical hypertension;
- insufficiency of the vesicoureteral junction in the phase of bladder distension and shortening of the intramural part of the ureter in chronic cystitis;

Clinical manifestations of CPUS

1. The clinical symptoms of CPUS depend on the degree of obstruction, the child's age and the time of seeking medical care.

Congenital urethral stenosis is an abnormal embryological development, being a spasm of the external urethral sphincter with fibrosis, which at birth has a symptomatology defined by the degree of stenosis and suprastenotic resonance of the urinary tract.

In newborns and infants, the clinical picture is severe, being characterized by general signs, such as fever, anemia, rickets, digestive disorders (anorexia, nausea, vomiting, diarrhea), which lead to acute dehydration, the neurological picture being more severe. Urinary disorders are present, such as weak urine flow, urinary retention, bladder distension, etc. On palpation, the kidneys are usually enlarged and painful, although on CUGM the bladder is virtually unchanged.

In children and adolescents, the clinical picture is characterized by urological signs, mainly micturition disorders, depending on the degree of obstruction. Dysuria, pollakiuria and nocturnal urination are the most common, subsequently followed by diurnal urinary incontinence. The urinary flow continuity as well as the caliber and projection force are reduced. In severe cases, urination is painful and requires effort, which can only be achieved by compressing the hypogastric region. Finally, detrusor hypotonia develops with false urinary incontinence (overfull urination) and ischuria paradoxa.

The evolution of these cases is sometimes torpid, other times the urinary signs remain constant for years. Getting used to them, the child seeks medical assistance in the advanced stages of the disease, when, anticipated by imperative urination. Thus, acute urinary retention develops, accompanied by abdominal pain, febrile seizures, sometimes with severe renal failure.

Urethroscopy - stenosis of the distal part of the posterior urethra is detected as well as an annular prolapse in the urethral lumen with a centric or eccentric foramen. Depending on the obstruction severity, the posterior urethral dilation is observed, the dimensional increase of the verumontanum, sometimes with edema, and the bladder neck being opened. In the bladder there are columns, trabeculae and pseudodiverticula.

A difference in supra- and substenotic transit can be revealed on radionuclide uroflowmetry. This test allows exclusively the recording of the turbulent urine flow in the supra-stenotic segment.

Clinical-radiological classification of CPUS

Summarizing the symptomatic evolution of CPUS depending on the pathophysiological substrate, four clinical-radiological phases have been highlighted;

Phase I - compensated

Phase II - undercompensated

Phase III - decompensated

Phase IV - refluxing ureterohydronephrosis

The highlighted phases are strictly didactic, as they cannot be delimited from each other. They have been classified as such, because there are different degrees of stenosis as suprastenotic resonance requiring a differentiated treatment.

Depending on the clinical-radiological phases, micturating cystourethrography and urographic examination found the following (fig. 2).

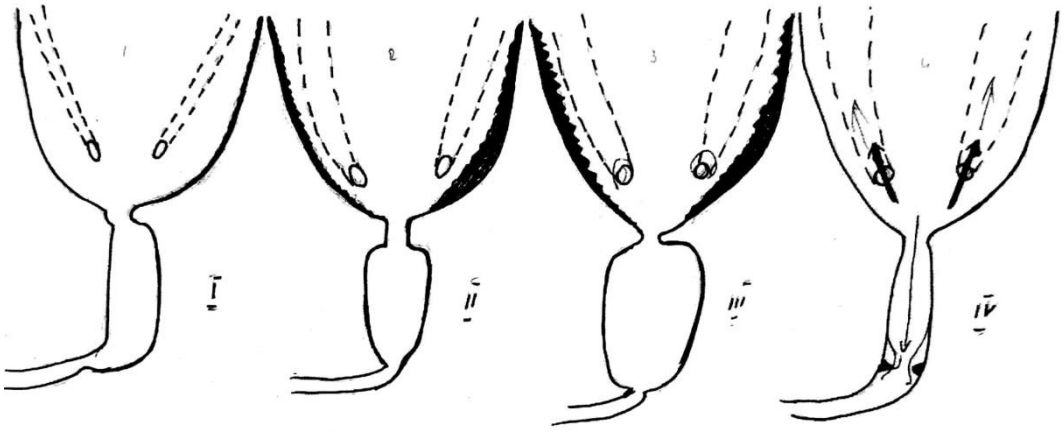

Fig. 2. Clinical-radiological phases of congenital posterior urethral stenosis. Scheme

Phase I - annular stenosis of the distal portion of the posterior urethra with insignificant suprastenotic urethral dilation on account of the posterior wall. In the region of the bladder neck there is a prominence in the lumen of the posterior wall. The bladder is slightly deformed with trabeculae on the posterior wall (fig. 3). Ureters and kidneys do not have any marked disorders (75 patients).

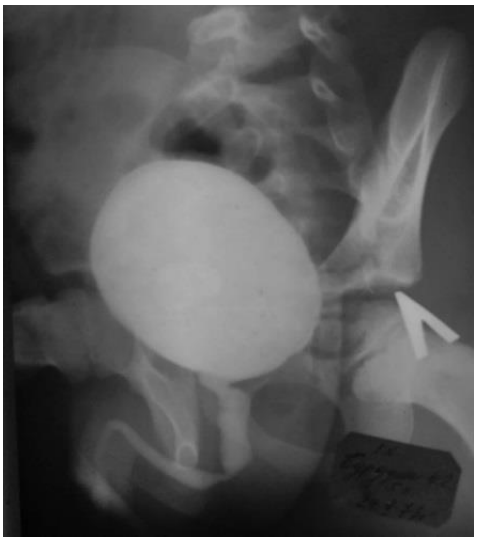

Fig. 3. Child C., 6 years; Micturating cystourethrography - posterior urethral stenosis, phase I

Phase II - annular stenosis of the distal posterior urethra, with marked dilation of the suprastenotic urethra. The bladder neck is narrowed and elongated. The bladder is deformed, with trabeculae on all walls, dilation of the upper urinary tract (46 patients).

Phase III – marked annular stenosis with exaggerated suprastenotic dilation. Dilated and short bladder neck. Enlarged bladder with trabeculae and pseudodiverticula. Refluxing megaureter on the right (fig. 4).

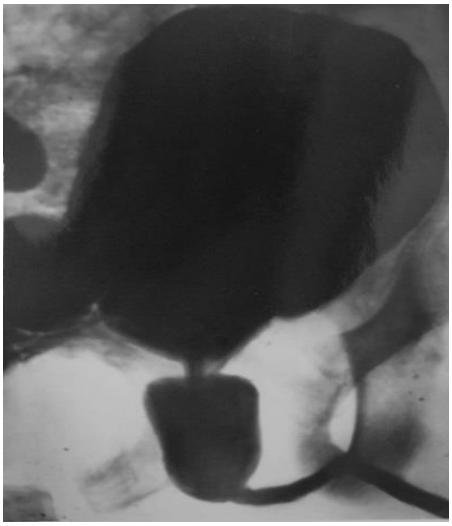

Fig. 4. Micturating cystourethrography - posterior urethral stenosis, phase III

Phase IV – It should be mentioned that in case of bilateral vesico-renal reflux the suprastenotic changes of the urethra, bladder neck and bladder are not marked, even in significant stenosis. This is due to urine flow which encounters an increased hypertension, obstructively, more easily returns to the ureters and renal pelvis. The bladder in this case does not look like a fight bladder.

In newborns and infants there are changes highlighting stenosis of the membranous urethra, suprastenotic dilation of the posterior urethra; the bladder and bladder neck are without essential changes. In these patients, it is necessary to calibrate the urethra with olivary bougies, which either confirm or not stenosis.

The principles of treatment of CPUS depending on the clinical and radiological phases

Treatment of patients with congenital stenosis of the posterior urethra is difficult, because once the obstacle (stenosis) is removed, it is necessary to correct the resonance of suprastenotic urinary tract - bladder disorders, vesico-renal reflux, ureterohydronephrosis, chronic urinary tract infection and kidney failure even in infants.

Considering the pathophysiology of suprastenotic disorders of the urinary system and the changes that take place in the bladder neck in urethral obstruction, after the `80s of the 20th century, no intervention on the bladder neck was performed. According to the literature, in about a quarter of patients, vesico-renal reflux gr. I-II disappeared during the first year after removing the obstruction.

Vesico-renal reflux should be closely monitored. If there is no spontaneous improvement over an adequate period of time after surgery, careful assessment of the renal function is required. Nephroureterectomy of the refluxing unit may be considered when no function is marked. If the function is maintained, urethral reimplantation can be individually considered.

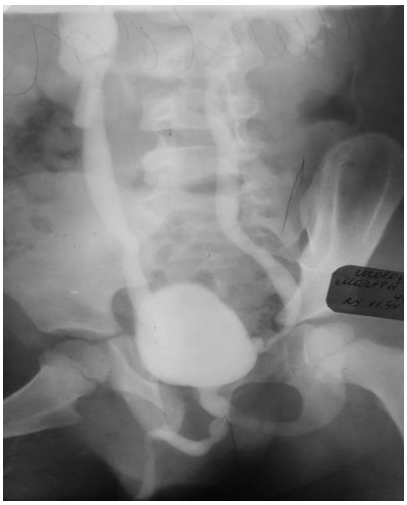

Fig. 5. Posterior urethral stenosis. Bilateral reflux. The bladder has no signs of "fight bladder"

Over several years the treatment of urethral stenosis entailed different procedures and methods. In the `70s of the 20th century, the dilation of the strictured segment was performed with metallic bougies under anesthesia.

The vast majority of the results of these procedures were unsatisfactory and sometimes stricture progression was recorded. Later, dilation with a mechanical urethral dilator or blind urethrotomy along with *commissurotomy* was performed.

Because the treatment results were not satisfying, in more severe cases of stricture, transperineal plastic surgery was performed, with a longitudinal incision and transverse suturing of the anterior wall of the stenotic or transperineal segment, namely, Holtsov procedure – resection of the stenotic segment and termino-terminal anastomosis.

Although these treatment methods have good results, in recent years they haven`t been used because they are laborious surgeries with anesthesia lasting 1.5-2 hours and a postoperative hospitalization period lasting for 10-12 days. During transperineal surgery, there is the risk of injuring the external urethral sphincter and perineal nerves, which can lead to urinary incontinence and impotence. In recent years, obstruction has been removed with an original urethrotome that allows us to resect transurethrally the anterior wall of the stenotic segment of the posterior urethra with good results (fig. 6).

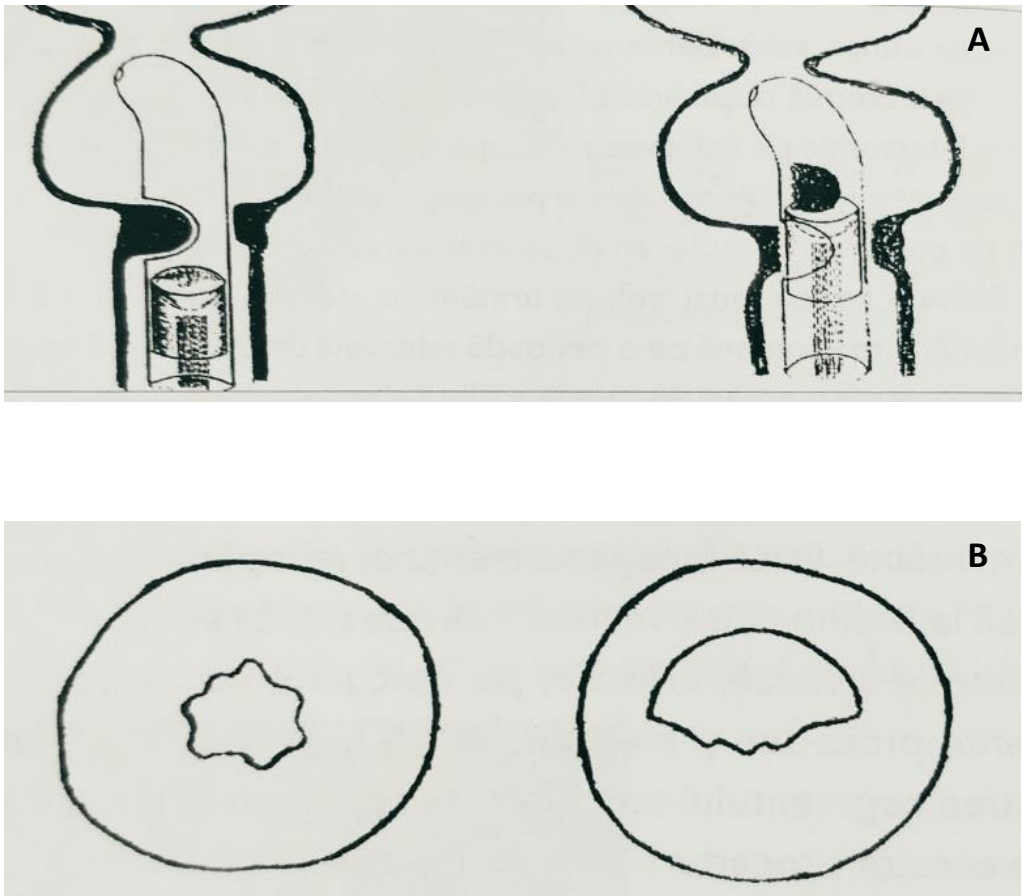

Fig. 6. A - Resection of the stenotic segment with an original urethrotome: before and after resection
B - Stenotic segment before and after resection: before and after resection

Macroscopically, the removed material has whitish tissue of hard elastic consistency with areas of fibrosis, the mucosa being pale pink and well differentiated. Microscopically, the urethral mucosa is lined with multilayered epithelium, in some places with proliferative phenomena in the submucosa, vast connective tissue with edema and infiltration, as well as dispersed lymphocytosis is revealed. In some sections, ectopy of the multilayered epithelium in the submucosa is determined. The muscle tunic is almost completely replaced by the connective tissue, which shows atrophied muscle fibers, being hypertrophied here and there.

Blood and lymphatic vessels show lesions manifested by mucoid and fibrinoid intumescence, in some places with hyalinization phenomena, deformation of thickened and sclerosed walls. Lymphatic vessels are in the form of deformed caverns. There is vast lymphocytosis. Nerve fibers are irregularly thickened and deformed. Analysis of histological results confirms the congenital genesis of obstruction.

Based on experience of over 20 years, the principles of treatment in disease phases have been developed, because patients in different phases require differentiated treatment.

Other diseases with infravesical obstruction are treated depending on the clinical picture of urodynamic disorders, urinary tract infection and neurological disorders.

In phase I – compensatory stage, changes in the suprastenotic urinary tract are not marked. In most cases it is necessary only to remove the obstruction, less often to balance the bladder tone and administration of uroseptics for a short time.

If urodynamic changes with signs of overactive bladder are not obvious, no drug treatment is required. Initially, urination reeducation should be carried out (behavioral urotherapy aimed at normalizing bladder function and preventing functional disorders). If the desired effect is not obtained, the therapy of stimulation of the presacral nerve roots is applied. Treatment with anticholinergic medication also has some side effects, such as: constipation, dryness of the mucous membranes, increased residual urine (Oxybutynin 0.1 - 0.15mg / kg)

In phase II - subcompensatory, the resonance of suprastenotic urinary tract is more pronounced, often with a marked urinary tract infection. Initially, cystostomy is performed, then the obstruction is removed, then the muscular tone of the bladder should be balanced with a longer treatment of the urinary tract infection. If after 6 months - 1 year there is no clinical improvement, the infection, vesico-renal reflux, and ureterohydronephrosis are present, the reconstruction of the upper urinary tract should be performed (antireflux surgery, ureteroplasty, pelvic plasty, etc.), after which the conservative treatment is prescribed.

In phase III - decompensation, refluxing phase, the treatment is aimed at achieving three goals. If the general condition of the patient allows, the obstruction is removed, the treatment of bladder tone balancing is performed, the treatment of the urinary infection is carried out, and in 6 months - 1 year the upper urinary tract plastic surgery should be performed with a prolonged conservative treatment.

In phase IV - with obvious dilation of the urinary tract, the patient's condition is severe with exacerbation of the urinary tract infection, a permanent probe is applied to the bladder (preferably a catheter smaller than the urethral lumen), cystostomy and percutaneous nephrostomy, after which the condition quickly improves and the obstruction is removed. Ureteral interventions are delayed until the patient is stabilized for 2-3 months.

Subsequently, intensive detoxification therapy and antibiotic therapy are performed, and uroseptics are administered. After 10-14 days, when the child's condition improves, cystourethrography is performed along with bladder filling through the stoma, antibiotics and diuretics being added to the contrast medium. It is possible to remove the obstruction 2-3 weeks after admission. Prolonged conservative treatment is prescribed, and plastic surgery of the suprastenotic urinary tract is performed after 6 months - 1 year with the conservative treatment – anti-reflux surgery. Surgery for neurogenic stenosis includes ureteral resection, even if the distal ureter is not spasmed, there is also obstruction of the intramural segment with intravesical neoimplantation. Ureterostomy is an additional burden for the patient, physician and parents.

Neither ureterostomy, nor probing, nor vesicostomy were applied to any patient. It is necessary to monitor the functional dynamics of the bladder, ureters, and kidneys. Sometimes cholinolytics, physiotherapy and phytotherapy should be administered.

If there is marked dilation of the ureters and renal pelvis, as well as megaureterohydronephrosis with impaired kidney function, antireflux surgery should be performed. If the kidney function is unilaterally disrupted, with the function preserved less than 25-30%, nephroureterectomy should be performed and ureter removed, to prevent reflux into the ureteral stump.

The X-rays of 105 newborns and infants with reflux megaureter were selected and studied from the archive data. There were radiological signs of posterior urethral obstruction in 38 patients, due to posterior urethral valve. Of 7 children with obstruction, neurological disorders were detected in 5 patients.

In phase IV - patients are usually hospitalized with exacerbation of the urinary tract infection. In parallel with antibacterial treatment and detoxification, the urethro-bladder probe or cystostomy is applied. In 7-10 days, after the condition improves, voiding cystourethrography should be performed where the urethra is practically

unchanged, but bilateral vesico-renal reflux is present, sometimes very pronounced or having an advanced degree. The bladder does not look like a *fight bladder*.

If the condition allows, the antireflux surgery should be performed, preferably transvesically (to capitalize on the increased intravesical pressure - Cohen, Leadbetter - Politano, *Bischoff* methods, etc.) on the most affected part of the kidney (after urography, scintigraphy). After 3-6 months, CUM should be repeated, where usually posterior urethral stenosis is clearly manifested and the obstruction is removed and antireflux surgery on the opposite side.

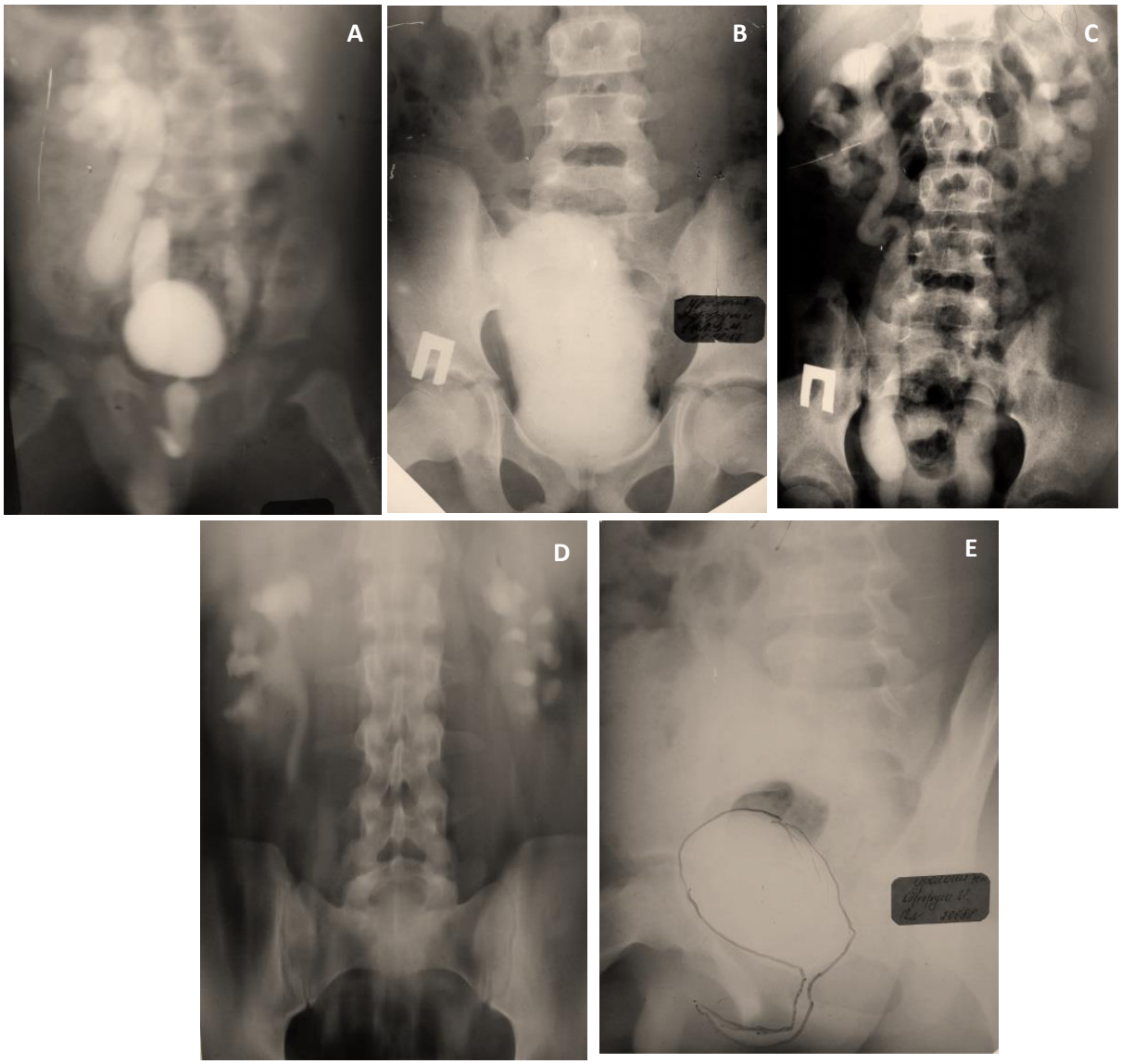

Fig. 7. A - Child C., 11 months; Cystourethrography - posterior urethral stenosis, undercompensated form. Bilateral vesico-renal reflux, marked megaureter on the right; B - The same child, 11 years Cystourethrography - marked urine retention, very large bladder with marked trabeculae; C - The same child urography: marked bilateral megauretero-hydronephrosis; D - 1 year after obstruction removal; urography - preserved renal function, dilation of the left renal cavities with nephrosclerosis signs; E - Cystourethrography - bladder with clear outline, normal size, permeable urethra

According to the literature, on urethrotomy there are recurrences of strictures, the recovery rate of subsequent urethrotomy decreasing by 10%, being almost zero for the following procedures (Heynes, 1998); this being a palliative treatment, often burdened by traumatic and infectious complications, progressively aggravating urethral lesions in length and depth [1].

The urethral stent or augmented anastomosis is one of the solutions. Urethrotomy originally excises the sclerotic wall and perforation is excluded because the tube is at the level of the urethral wall.

Antireflux surgery is performed no later than 6-12 months after removing the obstruction and only after normalization of micturition, suppression of bladder disorders, treatment of recurrent urinary tract infection and reduction of intravesical pressure as much as possible. Intravesical antireflux surgeries (Cohen, Leadbetter - Pollitano) are recommended to capitalize on the intravesical pressure and to form a stable antireflux procedure.

If urinary tract infection develops slowly, without exacerbation, patients with reflux are monitored for a longer time (1-2 years).

the intrauterine muscle spasm.

In one case the distal part of the posterior urethra is affected and the external urethral sphincter, in another case (neurogenic) along with the damage to the urethra, the muscles of the juxtavesical ureter (Waldeyer`s segment) are also affected, pelvic urinary tract muscles are hypertrophied, and there is increased intravesical pressure. In neurogenic stenotic urethra, children are restless, can not stand the urinary catheter. The physiological contracture of skeletal muscles maintains for a long time, strong urine stream (when urinating the stream reaches the face). In congenital stenosis, children bear calmly the urinary catheter. The urination is free and more frequent, sometimes at older age children have diurnal urinary incontinence.

In case of antireflux surgery, if the obstruction was not removed, and reflux was not detected on examination, ureterohydronephrosis maintains.

If urethral obstruction is removed, the overlying urinary tract returns to normal, in other cases, after removing the urethral obstruction, the urinary tract urodynamics is disturbed, although the vesico-renal reflux is not recorded, the *ureterovesical* segment is removed, renal parenchymal fibrosis and urinary tract,

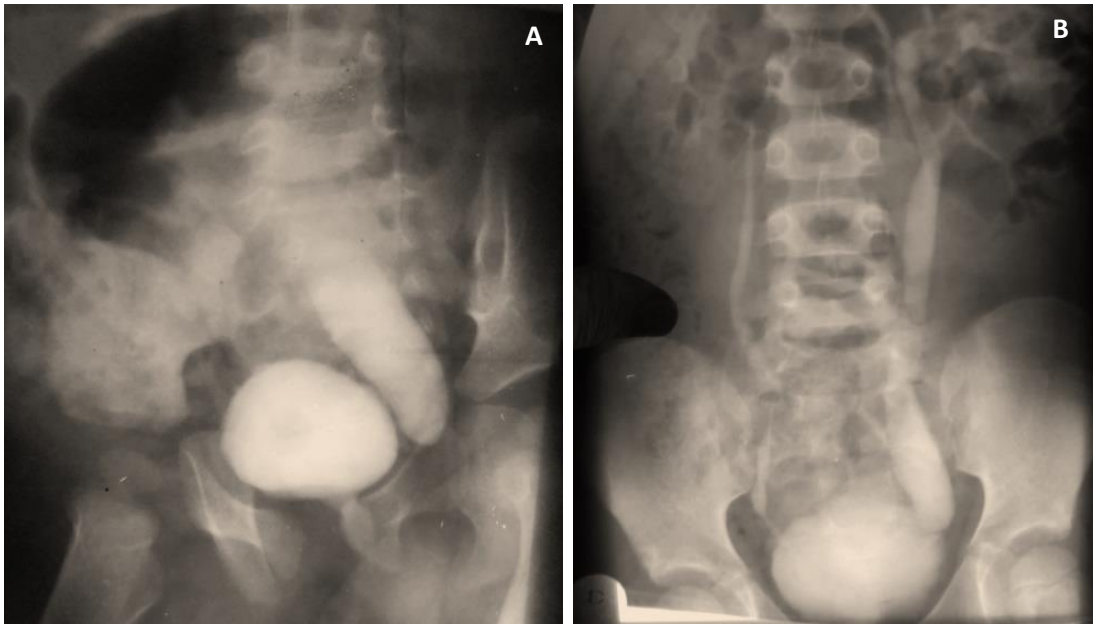

Fig. 8. Child C., 3 months. A – Micturating cystourethrography: bilateral vesico-renal reflux, megaureter on the left. Posterior urethral stenosis; B – urography after surgery: bilateral antireflux, infravesical obstruction was not removed – bilateral ureteral dilation.

Posterior urethral stenosis in some boys may be a consequence of muscle spasm in the intrauterine period with subsequent fibrosis.

It cannot be said that infravesical obstruction is absolutely a congenital organic or neurogenic abnormality. In both organic and neurogenic obstruction there is fibrosis of the urinary tract muscles as a result of

infection persist, but with less frequent exacerbations. On scintigraphy, there are sometimes marked evacuation disorders, caused by the spasm of bladder muscles, obstructing the intramural ureter as well as increasing the intravesical pressure, which is indicative of the persistence of the obstruction in the pelvic urinary tract.

There are also neurological signs after removing the

organic obstruction, which persist and require additional treatment. According to the literature, 5-6 years after removing the obstruction, neurological signs appear, which are likely present from birth but have not been detected. Thus, of 7 newborns and infants with megaureter, neurological manifestations were detected in 5 of them. Posterior urethral stenosis is probably not only an abnormality of organic development, but also a component of prolonged intrauterine muscle spasm with fibrosis of this segment [2]. Regardless of how advanced the urethral obstruction is, the section is maintained. Obliteration of the urethra or distal ureter in the *ureterovesical* segment is not common. This accounts for the fact that CPUS is not only an abnormality such as congenital abnormality of the rectum, namely fistula and atresia, but also represents a prolonged spasm causing its fibrosis. Both forms require surgical resolution.

Conclusions:

1. Annular stenosis of the distal segment of the posterior urethra which is a congenital abnormality or malformation caused by a prolonged intrauterine muscle spasm with fibrosis of the external urethral and intrauterine sphincter cannot be diagnosed and its evolution cannot be influenced, thus requiring surgical treatment.
2. The complications of stenosis are typical and atypical determined by age, degree of obstruction and ability to compensate for suprastenotic urinary tract. Signs of congenital stenosis in patients older than 3 years are urinary disorders, such as pollakiuria, urinary incontinence, difficult urination, abdominal pain, lumbar pain, and recurrent urinary tract infections. In children under 3 years the disease develops severely with difficult urination, bladder distension, leukocytosis, leukocyturia, proteinuria (urinary tract infections, sometimes severe infections).
3. To establish the diagnosis some diagnostic tests are used, such as micturating cystourethrography, urethroscopy, urethral calibration with olivary bougies (it is superior to radiological examination in children up to 3 years).
4. There are 4 phases from the evolutionary, clinical and radiological perspectives. Phase I is frequently characterized by pollakiuria and enuresis; phases II and III are marked by difficult urination, nocturnal / diurnal urinary incontinence, recurrent urinary tract infections with suprastenotic urinary tract dilation; phase IV is characterized by frequent refluxing ureterohydronephrosis and recurrent urinary tract infections.
5. The treatment of choice is partial transurethral excision of the anterior stenotic wall, satisfactory results up to 100% being recorded in the compensated phase and 47-70% in the decompensated phase.
6. Functional changes in the suprastenotic urinary tract are reversible and regress after surgical treatment, requiring only conservative *maintenance* treatment. Advanced changes, such as ureterohydronephrosis, vesicular-renal reflux grade III-IV, will require surgical correction but not earlier than 6-12 months, after normalization of urination and detrusor function.
7. Initially, the suprastenotic urethra, bladder neck and bladder are restored postoperatively. In the early stages of the disease, after surgery, the upper urinary tract recovers definitively, but the dilation is persistent in the decompensation phase, which is idicative of the presence of general dysplasia of the pelvic urinary tract.
8. The patient should be monitored pre- and postoperatively by a pediatric neurologist.

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