

MODERN METHOD OF RECONSTRUCTIVE URETHEROPLASTY IN GIRLS WITH EPISPADIA

O.B. Loran MD, Prof.², E.L. Vichnevsky MD¹, Prof., A.A. Demidov PhD¹

¹Urology and Neurourology Department of Moscow Scientific-Research Institute of Paediatrics and Children Surgery, Russia

²Chamber of Urology of Russian Medical Academy of Postgraduate Education, Moscow

Abstract

10 girls with epispadias aged 7–12 have been operated in our clinic since 2007. Urethroplasty as the method of Otto-Dranicin was the first stage of surgical reconstruction of lower urinary tracts. In all cases urethral anatomy was formed. In 8 patients the process of urine transportation through the lower urinary tracts has changed cardinally. Children began to store 150–300 ml of urine in every position, to hold urine up to 3 hours at rest, on strain totally evacuate urine with normal stream from the bladder. Total urine loss decreased in 10–15 times in 24 hours and was not more than 20–40 ml. The main clinical characteristics of urination disturbances after the operation was stress urine incontinence that appeared only with sudden and sharp increase of intraabdominal pressure (coughing, quick walking and so on). Urethroplasty according to Otto-Dranicin is very effective in epispadias in girls, both anatomically and functionally, and we consider it as the most effective surgical correction of this malformation.

Keywords

epispadias, girls, urethroplasty.

INTRODUCTION

Epispadias is the one of the rare malformations in children. Traditionally it is considered to be met in 1: 100000 of newborns, while the proportion of males and females is 1: 3,5. That's why epispadias is registered nearly in one girl out of 350000 children. Every year 10–11 such patients are born in Russian Federation. All the above said data underline that this malformation is extremely rare in females.

Urine incontinence and absence of voluntary urination are the most clinically vivid expressions of epispadias. That is the reason for parents to go to the doctor with their child. During the latest 150 years all the doctors working on that problem tried to find the most effective procedure to restore the full functioning of urinary bladder in the regimen «filling/evacuating». Numerous publications show that a lot of surgeons work hard, correctly formulate the

targets and invent different new methods of treatment, but still they tried to find, without a great number of evidences, the main mechanism in that complicated disease structure.

If the doctors were oriented on using only one method of operation, the results of malformation correction could not always be prognosticated, those operations were often a failure and that problem in children surgery was not solved.

We should state that the efficiency of time-proved surgical methods of treatment urinary incontinence in children was not more than 50%. Half of the children were operated several times. For example, in Savchenko clinic operations after Derzhavin and Yong-Dees were effective in 86,9% and 61,9% of cases, respectively, and 42% of patients were operated twice.

At first, all the unsatisfactory results of the operations were explained as not having been followed the authors recommendations of their fulfillment. Later on another reason of it was found. Epispadias was often followed by overactive bladder as well as by myelodysplasia with the disturbance of lower pelvic muscles innervation. While studying this malformation it was stated that the degree of diastasis of pelvic bones was the integral index of structural-functional deep changes in the lower urinary tract in girls with such malformation. The degree of diastasis correlates well with the severity of malformations, with urinary bladder dysfunction, with disturbances of pelvic muscles innervation, while the results of classical operations were unsatisfactory.

Surgeons of the Department of Urology and Neurourology of Moscow Scientific Research Institute of Paediatrics and Children Surgery have been working on the problem of epispadias for 40 years. During this period of time operations, mainly plasties of urinary bladder neck out of the urinary bladder triangle muscles were made in more than 450 children of both sexes. But the results of treatment were not as effective as expected. That is why while studying the disturbances of urination pathophysiology we began to understand the necessity of cardinal changing the traditional methods of epispadias treatment. First of all, it should have several stages. The first stage is to restore the urethra, the second – to think of sphincteroplasty, the third – to abolish the residual disturbances of urination with the help of noninvasive medical technologies and of pharmaco-therapy of urinary bladder dysfunction.

This paper is dedicated to the first stage of epispadias surgical treatment in girls – that is to the results of reconstructive urethroplasty.

MATERIALS AND METHODS

During the latest 4 years we were studying epispadias in 10 girls aged 7–10. «Urine incontinence» in horizontal and vertical postures was the main complain and that incontinence grew with the increasing of abdominal pressure.

The malformation of the lower urinary tract development was stated on the bases of physical examination and special methods of investigation – excretory urography, cystography, cystoscopy, uroflowmetry and electromyography.

It is quite enough for an experienced surgeon to make only physical examination so that to disclose the malformation, but using special methods has another target. With the help of special methods we can diagnose concomitant malformations of kidney and urinary tracts, can estimate the condition of the urinary bladder and ureteropelvic junction, the degree of pelvic bones diaphysis, the length of urethra, the condition of sphincter apparatus and the innervation of pelvic muscles fundus. One can meet some difficulties while identifying this malformation in girls. There are some diagnostic characters typical for «women epispadias»: clitoris cleft and genital lips segregation, subsymphisal situs and funneling of the outer hole of urethra. Clinical picture is composed of the involuntary urination with strain (changing of the body position, coughing and so on). Cystoplasty showed the wide open neck of the urinary bladder. The urethra is short, its length is not more than 0,5–1,5 sm. Positive intraurethral pressure is decreased or is not registered at all. Diaphysis of pelvic bones can be seen. In some

cases systometry gives the results of hyperreflected detrusor, and electromyography shows the disturbances of segmental somatic innervation.

The above described picture is composed on the basis of the so called «women epispadias», which has been examined for many years and in which all the individual characteristics of this malformation have been included.

The girls in our study group had all the external signs of typical epispadias. In the supine and sitting position the capacity of the bladder varied from 30 to 200 ml. While changing the position into vertical urine incontinence emerged and increased with coughing and walking. The length of the urethra was 5–13 mm (normal 23–32 mm). Diaphysis of pelvic bones was 18,1–38,7 mm (normal 7–8 mm). In two cases cystography showed bladder-ureteral reflux of the 2d degree on both sides; in one of these two cases the reflux was complicated with returning pielonephritis. Electromyography showed the disturbances of the somatic innervation in 4 girls.

After the diagnosis of «epispadias» was stated the girls were operated on, the first stage of the operation – urethroplasty according to D.O. Ott. This method was described in 1894 for restoring urethra in women. In the same year Dranicin published the article about successful use of it in girls of 9 years old with total epispadias. The same operation to correct epispadias with good results was used in women by Atabekov. This operation is simple, quite effective and helps to solve the main problem of the first stage – to restore «urethral anatomy». After all this variant of urethroplasty in women epispadias should be called «the operation of Ott – Dranicin».

The essence of this operation is as follows (see Figures 1–5).



Fig. 1. Total epispadias in a girl

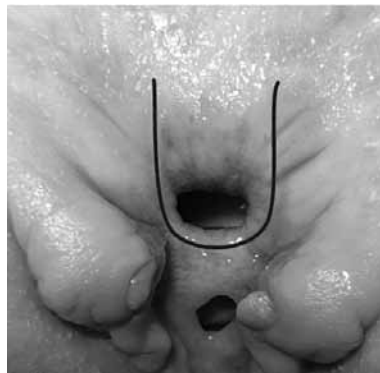


Fig. 2. The shape of incision



Fig. 3. Urethral tube grafting



Fig. 4. Tubularisation on the catheter



Fig. 5. The end of urethroplasty

After general anesthesia we made U-shaped incision of the skin and subcutaneous fat, the lower part of it is in between urethra and vagina (Figure 2). The back wall of the urethra is separated from the front wall of the vagina as far as 4–7 mm. This manipulation is necessary to make the connection between the proprius urethra and muco-cutaneous urethral tube easy. The urethral tube and the outer hole of the urethra are formed on the FOLEY catheter №10 with a thread PDS 5/0 (Figure 4). The front vagina wall and the surrounding tissues close the anastomosis between the proprius urethra and the urethral tube with the help of additional sutures (Figure 5). The urethral tube formed in such a way «is closed» with 2 suture layers out of the zones of U-shaped incision.

After the operation antibiotics were prescribed for prophylaxis of wound and urinary infection. Urinary bladder has been catheter drained for 7–8 days. The child was to get up and walk on the 3rd day after the operation.

RESULTS

With the help of Ott–Dranic operation all the girls got the restored urethra. It was lengthened in 2–2,5 times, the outer hole was front and above shifted.

Nevertheless, 4 girls have got urethral fistulas. In 2 cases they were abolished with the help of sutures and repeated urethroplasty.

«Urethral anatomy» restoration in the following 3 years of investigation was accompanied by serious changers of urine transportation along the lower urinary tracts:

1. The premium stage of micturition – the storage of urine – was restored. Children began to store 150–300 ml of urine in every position.
2. They could hold store and hold urine up to 3 hours at rest.

3. The desire to micturate appeared after the bladder was filled; voluntary urination had normal stream without residual urine.
4. The main characters of urine incontinence have been changed: incontinence emerged only when sudden and sharp increase of intraabdominal pressure was registered (coughing, quick walking and so on). «General urine loss» decreased and was not higher than 20–40 ml – that was in 10–15 times less during the 24 hours.

The problem of the second stage – sphincteroplasty – is being solved individually. Proper limits of time were not defined. The main criteria are: physical development of a child and the condition of tissues where the intervention is to be done. Our experience showed that children «were ready» for this or that sphincteroplasty in 6–12 months after the first stage of operative intervention in order to treat epispadias.

DISCUSSION

In spite of numerous publications on that theme there are a lot of topics waiting to be discussed. Classification is one of them. The most comprised is the classification of epispadias made by Savchenko and Derzhavin (1976). However its practical value in choosing the method of treatment is not quite enough.

We suppose it will be right to cite the words of Derzhavin V.M. that clinically we should divide epispadias in two forms: with incontinence and without incontinence and «knowing that this division is very relative we still can not help recognizing that very characteristic being the definition for both the attitude of a patient to her disease and the tactics of a surgeon».

It is necessary to note that most girls and women, if they do not have incontinence, even do not suppose they have got epispadias. Low degrees of malformations simply are not being diagnosed and they are not included into statistics. There are some problems with the forms accompanying urine incontinence. Previously this complicated patophysiologic condition was not differentiated in forms. Nowadays we know imperative, stress, combined urine incontinence and paradoxal ishuria. All of them are met in epispadias and can serve as a criterion of for treatment choice. We had the evidence that the imperative incontinence, which had been by mistake regarded as a failure of sphincter apparatus reconstruction, was with ease treated pharmasutically. In these cases the question of the repeated operation was not on the agenda.

One should always be very careful regarding the results of the epispadias operation when formally the incontinence preserved. Before you decide to make the repeated intervention, you should classify the variant and all the reasons of incontinence. In some cases less invasive technics (paraurethral infusion of volume-forming agents) and pharmacotherapy might influence the total result of the treatment.

Before surgical treatment of epispadias the surgeon should solve the difficult task – he should choose the algorithm of correction of a very complicated malformation. What will he begin with? Urethroplasty or sphincteroplasty? Our experience showed that it is advisable, first of all, to restore the integrity of lower urinary tracts, that means to do only urethroplasty.

The surgeon should be experienced enough to choose the proper method of operation. As far as «women epispadias» is concerned we may say that the operation according to Ott–Dranicin, developed in

1893, is still popular among surgeons. Besides restoring the anatomy of urethra, this operation helps to restore voluntary urination. We can evidence that not only sphincter apparatus of urinary bladder, but urethra as well take part in the most complicated mechanism of urine holding.

While operating urethra, especially correcting its inherent malformation, you should remember and bare in mind one more problem – spontaneous fistula forming. Fistulas are the acute angle of urethral surgery. To solve this problem or to lessen its severity we should use a complex of methods. They are: prophylaxis of wound infection, use of miniature instruments to treat the tissues carefully and of course use of modern suture materials.

Summing up we can conclude that surgical treatment of such rare malformations as epispadias in girls should be carried in special surgical clinics, where this problem is being worked at.

in brief...

PRENATAL DIAGNOSIS OF AORTIC ARCH ABNORMALITIES

M.N. BARTAGOVA,
E.D. BESPALOVA, R.M. GASANOVA,
I.I. PITIRIMOV, A.I. TUMENEVA

OBJECTIVE. To assess the added value of 3/4-dimensional echocardiography in the diagnosis of aortic arch abnormalities in fetuses.

MATERIALS AND METHODS. 26 patients were examined between 20 and 36 weeks of gestation/ All fetuses (n=26) in the following groups were included in the study:

- 1) normal heart and great vessels (n=20),
- 2) isolated aortic arch abnormalities (n=6): coarctation of aorta (n=5) and interrupted aortic arch, Type A (n=1).

Volume data sets of 26 fetuses, examining by 2D US, were ac-

quired by 3D and cine 4D using spatiotemporal image correlation (STIC) software. Various additional rendering tools were applied. Color and power Doppler were added to the volumes acquired. A retrospective offline analysis was performed. Neonatal echocardiography and pathological examination were performed to verify the prenatal diagnosis.

RESULTS. 1 group of fetuses (normal heart and great vessels, n=20): In 18 cases 3/4D ultrasound volume did not add to the information in the 2D loop. Normality of the heart and great vessel was demonstrated by 2D and 3/4D too. In 2 cases obesity and a fetus with the spine toward the maternal abdomen were associated with unsatisfactory screening examination by 2D also 3/4D. 2 group of fetuses (aortic arch abnormalities, n=6): 3/4D ultrasound had added value in achieving

or enhancing diagnosis in 2/6 of diagnosed cases and a definitive diagnosis was made only after 4D examination. This was 1 coarctation of aorta and 1 interrupted aortic arch, Type A. In 4/6 cases (4-coarctation of aorta) 2D and 3/4D ultrasound examination were equivalent. The resolution of three/four-dimensional systems is generally less good than that of high-quality two-dimensional systems.

DISCUSSION. 3/4D features may work to extend the benefits of prenatal diagnosis of aortic arch abnormalities. However, no single module is sufficiently accurate for the diagnosis of aortic arch abnormalities, each case requires different and appropriate module of visualization. 3/4D addition enhances precision of diagnosis by providing planes and data that «flesh out» the 2D ultrasound examination.