

## Management of Congenital Urethral Strictures In Infants. Case Series

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**Purpose:** Infra-vesical obstruction is uncommon in infants and generally due to urethral valves. Congenital urethral strictures (CUS), instead, defined as a concentric narrowing of the urethral lumen, are exceedingly rare in infants.

**Materials and Methods:** We reviewed our experience with 7 patients treated at our institution for CUS <age of 1-year. The study is a retrospective review of 7 patients treated for CUS diagnosed <age 1-year during a 10-year period.

**Result:** In a single patient, the urethral stricture was an isolated condition, 3 had a Prune Belly Syndrome (PBS) and the remaining 3 had an Ano-Rectal Malformation (ARM). Four patients had upper tract dilatation detected on prenatal ultrasound. Five patients had upper tract dilatation on postnatal ultrasound. Five patients had impaired renal function at diagnosis and 3 required renal transplantation eventually. On micturating cystourethrography, all strictures were located in the anterior urethra and 4 cases had associated vesicoureteral reflux. In all cases, but one urinating via a patent urachus, initial management included insertion of a supra-pubic catheter. Subsequently, the CUS could be treated by dilatation or endoscopic incision in the 3 patients with Prune belly syndrome, whereas 3 of the remaining 4 required a formal urethroplasty.

**Conclusion:** Diagnosis and treatment of CUS in infants and children remain difficult to standardize. At presentation, urinary diversion is key to avoid progressive renal damage in infants that can already have an impaired renal function. Anterior strictures in patients with PBS are likely to be fixed with progressive dilatation. In other patients, instead, urethroplasty should be considered. A formal vesicostomy or, if possible, an urethrostomy can allow temporizing final surgery. A major problem we experienced in the treatment of CUS is that the small endoscopic instruments required in this age group make urethral instrumentation more difficult and less effective than in older children and adults.

**Keywords:** anorectal malformation; congenital urethral stricture; Prune-Belly Syndrome; urethral dilatation; urethroplasty; urethrotomy

## INTRODUCTION

Infra-vesical obstruction is uncommon in infants. Posterior urethral valves followed in frequency by anterior urethra valves and congenital urethral diverticula, are well-known causes of infra-vesical obstruction in this age group and endoscopic treatment is generally considered a viable option under these circumstances<sup>(1,2)</sup>. Congenital urethral strictures (CUS), instead, defined as a concentric narrowing of the urethral lumen, are exceedingly rare in infants. Only few retrospective case series are available in the literature, and this determines a lack of consensus on the ideal management<sup>(2-5)</sup>. Regarding the latter, the main problem is whether urethral dilatation might suffice, if an endoscopic treatment should be pursued, or finally whether a primary surgical repair should be favoured given the difficulties with endoscopic urethral instrumentation in infants and the known limited effectiveness of dilatation and endoscopic urethrotomy in fixing urethral stricture in older children and adults (although the latter are different in

nature as generally acquired after urethral surgery or trauma)<sup>(3,6,7)</sup>

In this study, we reviewed our experience with 7 patients treated at our institution for urethral stricture diagnosed before age one year.

## MATERIAL AND METHODS

### Study population

We retrospectively interrogated the institutional database for urethral stricture in the period 2005-2015. Data were collected regarding antenatal history, presentation, diagnostic work-up (including radiological assessment of the stricture, assessment of upper urinary tract status and renal function), treatment, and outcome.

### Inclusion and exclusion criteria

Inclusion criteria were age at the diagnosis under 1 year, available radiological or endoscopic demonstration of the stricture, and a minimum follow up of 12 months. Exclusion criteria were evidence of urethral valves or meatal stricture, and previous history of trauma, inflam-

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**Table 1.** Patients' characteristics and management of the stricture

Pt	Associated condition	Presentation	Upper tract status at presentation	Renal function	Urinary tract diversion at presentation	Stricture location	Stricture length	Age at (mos)	Treatment of Stricture		Additional surgery
									1st treatment PADUA	2nd treatment	
1 (MZ)	PBS	Prenatal detection of Bil HUN, poor stream at birth	Bilateral HUN (No VUR)	Renal Failure (RTx)	Vesicostomy at birth	Penile	> 1 cm	10	up to 12 Fr		
2(GF)	PBS	Prenatal detection of Bil HUN, poor stream at birth	Bilateral HUN (No VUR)	Renal Failure (RTx)	Vesicostomy h at birt	Bulbar	< 1 cm	1	EU (cold knife)		
3 (DC)	None (Prenatally detected bil HUN)	Prenatal detection of Bil HUN Urosepsis at 3 weeks of age	Bilateral HUN (VUR Rt)	Mild Chronic renal failure (Estimated GFR 70 ml/min/1,73m <sup>2</sup> )	Epicistostomy tube then scrotal Urethroscopy	Penile	>1 cm	1	Urethroplasty		
4 (EB)	ARM (fistula with bladder -neck)	Impossibility to pass a catheter at colostomy (2 days of life)	Bilateral HUN (VUR Bil)	Normal renal function	Epicistostomy tube	Bulbar	<1 cm	5	EU (cold knife)	EU (Laser)	Urethroplasty
5 (FZ)	PBS	Voiding via a patent urachus	Bilateral HUN (VUR Rt)	Renal Failure (RTx)	None	Penile	>1 cm	2	PADUA up to 12 Fr		
6 (JT)	ARM	Poor urinary stream and impossibility to pass a catheter		Normal renal function	Epicistostomy tube	Bulbar	<1 cm	6	PADUA up to 10 Fr	EU (cold knife)	
7 (AL)	ARM	Poor urinary stream and impossibility to pass a catheter		Normal renal function	Vesicostomy	Penile	>1 cm	1	Urethroplasty		

**Abbreviations:** PADUA, progressive augmentation by dilating the urethra anterior; PBS, Prune-Belly Syndrome; ARM, Anorectal Malformation; VUR, vesicoureteral reflux; HUN, hydro-ureteronephrosis; rTX, renal transplantation; EU, endoscopic urethrotomy

mation, urethral instrumentation or surgery.

**Procedures:** Management was individualized based on the preferences of the attending surgeon and included initial diversion, if deemed appropriate, followed by a combination of urethral dilatation, endoscopic internal urethrotomy (EU) under direct vision, or formal urethroplasty. Dilatations were performed as Progressive Dilatation of the Urethra Anterior (PADUA procedure), which include forceless urethral insertion of catheters increasing in size over few weeks (8). Catheters were substituted over a guide-wire, sometimes in the outpatient clinic.

**Evaluation**

For the purpose of the present study, strictures were classified as penile or bulbar, and as short (< 1cm) or long (> 1cm). Success of treatment was defined as absence of urinary symptoms at follow-up, and radiological or endoscopic documentation of resolution of the stricture.

**Statistical Analysis**

Only descriptive statistic was used.

**RESULTS**

Seven patients met the inclusion criteria for study (**Table 1**).

In a single patient, the urethral stricture was an isolated condition, 3 had a Prune Belly Syndrome (PBS) and the remaining 3 had Ano-Rectal Malformation (ARM).

In no patient the stricture was diagnosed prenatally but 4 had upper tract dilatation detected on prenatal ultrasound. Four patients (2 PBS and 2 ARM) presented with urinary retention and poor stream. One patient with PBS was urinating only via a patent urachus, whereas in the last patient with ARM the stricture was detected for the impossibility to pass a catheter during surgery for creation of a colostomy at 24 hours of life. The single patient with an isolated stricture presented at 1 month of life with an urosepsis while he was followed conservatively for bilateral hidroureteronephrosis (HUN) elsewhere.

On ultrasound, 5 patients had evidence of bilateral HUN. The patient with the isolated stricture had also ultrasound evidence of penile cysts. Five patients had impaired renal function at diagnosis and 3, all PBS, have required renal transplantation so far.



**Figure 1.** MCUG performed via a suprapubic catheter. The white arrow shows a long stenosis involving the anterior urethra, with a proximal (posterior) rosary-like dilation. The white star points to a radiopaque marker located on the basis of the penis.

In all cases, but the one urinating via the patent urachus, initial management included percutaneous supra-pubic diversion of the bladder. A combination of retrograde urethrography and micturition cystourethrography (MCUG) was used to confirm the diagnosis and determine the extension of the stricture in 7 out of 8 cases. All strictures were located in the anterior urethra (**Figure 1**). On MCUG, a dilated proximal urethra was present in all patients, and 4 cases had associated vesicoureteral reflux.

All patients underwent endoscopic assessment. In 3 cases (2 PBS, 1 ARM), a 3 Fr urethral catheter could be passed into the bladder and an attempt to a PADUA was elected. The percutaneous drainage was converted in a formal vesicostomy in 2 such cases. The PADUA was successful in the 2 PBS patients whereas the patients with ARM went on with a cold knife EU that was successful. EU was performed as primary procedure in two patients (1 PBS, 1 ARM). A cold knife incision was successful in the patient with PBS whereas EU failed in the other patient despite two attempts, one with cold knife and the other with holmium laser. In the remaining two patients (1 isolated stricture and 1 ARM), the stricture was considered unsuitable to attempt an endoscopic management. Therefore, a scrotal urethrostomy and a formal vesicostomy were performed, respectively, in the prospect to perform a primary urethroplasty on an elective basis. Formal urethroplasty was successful in both these patients as well as in the one with ARM where two EU failed. Overall, none of the 3 patients with PBS required a formal urethroplasty vs. 3 of the 4 remaining patients.

## DISCUSSION

In children, urethral strictures, defined as a concentric narrowing of the urethral lumen, generally follow perineal traumas or urethral surgery, such as hypospadias

repair or surgery for anorectal malformation<sup>(2,3,4)</sup>, and occur after the first year of life. In infants, instead, infra-vesical obstruction is most commonly due to urethral valves whereas congenital urethral strictures are exceedingly rare. We identified 7 patients undergoing treatment for congenital urethral strictures at our centre over a 10-year period. These patients are peculiar in many respects. To begin with, 6 out of 7 had associated conditions including PBS and ARM, 3 each. The associated underlying condition seems to have important implications both to explain the development of the stricture and for its management. It is indeed of note that the stricture could be fixed by progressive urethral dilatation or by EU, irrespective of its length, in all the 3 patients with PBS whereas 3 of the other 4 patients required a formal urethroplasty. Stumme was the first suggesting that all the features of the PBS might be due to an in utero bladder outlet obstruction distal to the prostatic urethra. This obstruction would be typical transient and should disappear before birth<sup>(9)</sup>. We assume, therefore, that our PBS patients had some degrees of developmental delay of the urethral lumen, but not an abnormal urethral wall, which can explain the good response to dilatation or EU in contrast to the stricture in the other patients<sup>(9)</sup>. The other major group of patients included children with high ARM. Also in these patients, the presence of urethral stricture is not surprising as it might be part of the developmental defect leading to the formation of a rectal-urinary fistula. In keeping with previous reports, we noticed that under these circumstances, dilation with or without urethrotomy is fraught with a high failure rate<sup>(10,11)</sup>. This would be consistent with a more significant abnormality of the urethra. An additional problem peculiar to this group is that the presence of a recto-urethral fistula can complicate the diagnostic workup. Radiographic appearance of the urethra can be altered by inadequate passage of contrast in the urethra distally to the fistula opening. Moreover, in one patient, the anomaly was discovered due to the impossibility to pass a catheter at the time of colostomy opening during the neonatal period before the urinary stream could be properly assessed and any radiological study of the urethra planned. Still, this is a sign difficult to interpret since urethral catheterization in ARM patients may be demanding per se due to the presence of a recto-urethra fistula. Anyway, our series shows that this can be the first sign of a primary disorder of the urethra.

Regardless of the nature of the stricture and the associated condition, unless the patient has developed a pop off mechanism such as our patient with a persistent urachus, the first step in the management of these patients should be, in our opinion, the achievement of urinary diversion to preserve renal function. Placement of a percutaneous epicystostomy tube is the option of choice at the outset, particularly considering that most of these patients present without a suspicion of a urethral stricture and can be symptomatic with acute urinary tract infection or also urosepsis. This diversion also allows checking safely bladder emptying after treatment of the stricture by intermittent closure of the tube. The drawback is that tube blockage or dislodgment is common in infants, therefore in a few cases conversion to a formal vesicostomy can be advantageous if the stricture tends to recur or to temporize final surgery. In patients with penile stricture a urethrostomy allows to bypass the

stricture while preserving bladder cycling. If this kind of diversion is elected the final repair can be easily postponed to an older age. However, diversion of the urinary flow proximally to the stricture may lower the success of dilatation and endoscopic incision of the stricture, as the urethra is no longer distended by the urinary flow during healing process and this might cause stricture recurrence. We selected this diversion in one patient and he was indeed scheduled for subsequent urethroplasty. It is noteworthy, anyway, that despite an early diversion 4 of our 7 cases in present series presented with renal failure, and 3 required renal transplantation eventually. This clearly depends from the degree of renal damage developed prenatally already.

After urinary diversion, treatment options for anterior urethral strictures include wire-guided dilation, direct vision internal urethrotomy, and open surgery. The latter includes stricture excision and direct anastomosis, or augmentation urethroplasty with interposition of a graft as an onlay or inlay in the narrowed urethral segment<sup>(3-5)</sup>. Reportedly, urethral dilatation has the lowest success rate, ranging from 20% to 55%. It is considered suitable only for short and mild strictures. Moreover, repeated attempts are discouraged and change to other strategies is recommended after the first or the second failure. As mentioned above, in our experience urethral dilatation worked in PBS patients, perhaps due to the peculiar nature of the stricture in these patients. It should be noted that we performed the dilatation using the PADUA technique (8). This procedure was first described in the late 80s specifically for anterior urethral hypoplasia. The principle is to avoid rapid dilation. Urethral dilatation should occur passively. Stent size is progressively increased at one-week intervals until an adequate caliber, of at least 8 Fr, is achieved. In a few cases, we managed to change the catheter over a guide in an outpatient setting. The final caliber was steadily achieved after a median of 4 weeks. We consider this approach easier than balloon dilation, which requires the child to undergo repeated sections under fluoroscopy. Furthermore, the success rate of balloon dilatation appears unsatisfactory<sup>(8-9)</sup>.

In contrast, formal urethroplasty, with a success rate ranging from 80% to 95 %, is reportedly the most effective treatment option for urethral strictures in children<sup>(2,3,8,9)</sup>. Urethroplasty, however, is also the most invasive option and carries a specific morbidity. In our opinion, it remains the option if less invasive manoeuvres fail or if the strictures cannot be negotiated at all during initial endoscopy.

Direct vision internal urethrotomy stays somewhere in between urethral dilatation and formal urethroplasty and this is generally recommended as primary treatment for short (less than 1 cm) urethral strictures. Nevertheless, a notable technical problem related to the endoscopic treatment of urethral stricture in infants is that urethral instrumentation can be difficult with the small endoscopic instruments required in this age group and not as effective as with the instruments used in older children and adults. For instance, our subjective impression was that the small cold knives available for the 8 Fr and 9.5 Fr cystoscopes are generally ineffective in obtaining a satisfactory incision of a stricture. Of course, this is likely to improve with the development of new technologies. The holmium laser seems to allow for a more effective incision, and the smaller fibres easily

fit in endoscopes as small as 7.5 Fr. Though, we used the holmium laser in one case and despite the incision looked deep and net, the stricture recurred and required a formal urethroplasty eventually.

Given the lack of solid evidence, we generally individualized the treatment based on patient characteristics. Moreover, the treatment of the stricture has to be put in the contest of the other surgeries required, such as in ARM patients. In general, once an adequate urinary diversion is ensured, unless in cases of very long and severe strictures that we scheduled for urethroplasty from the outset, we think that a stepwise approach moving from dilatation, to endoscopic incision to formal urethroplasty might be the most reasonable strategy. This, however, can be quite a long process requiring months to be accomplished.

## CONCLUSIONS

Diagnosis and treatment of CUS in infants and children remains difficult to standardize. The paucity of cases in the literature does not allow an agreed flowchart and treatment has to be tailored to each single patient. Urinary diversion should be achieved at presentation to avoid progressive renal damage in infants that can already have an impaired renal function. Following management should be tailored based on the location and length of the stricture, and the associated condition. In our opinion a stepwise approach should be favoured. Anterior strictures in patients with PBS are likely to be fixed with progressive dilatation irrespective of their length, whereas this treatment modality is unlikely to be effective in other patients. In the latter, an endoscopic urethrotomy can be attempted, but if it fails urethroplasty should be considered. Placement of a formal vesicostomy or, if possible, a urethrostomy depending on the location of the stricture, allows temporizing this surgery until after one year of age.

## CONFLICT OF INTEREST

There are no conflicts of interest to be disclosed.

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