A title page

1. The category of the paper

Case report

2. The title of the paper

ANTERIOR URETHRAL VALVES IN CHILDREN: A POSSIBLE ASSOCIATION BETWEEN ANTERIOR URETHRAL VALVES AND COWPER'S DUCT CYST

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8. A short running title

Anterior urethral valves in children

Abstract

Aims: To elucidate the pathophysiology of congenital obstruction of the anterior urethra and to investigate the association between anterior urethral valves and syringocele.

Methods: Three boys with congenital obstruction of the anterior urethra diagnosed at our department between 1997 and 2004 were analyzed retrospectively.

Results: All of the three boys had congenital obstruction in bulbar urethra. The presenting symptom and age of each patient were varied. Whereas continuity between Cowper's duct and the diverticulum was not demonstrated radiographically in all boys, it was speculated endoscopically in all.

Conclusion: Our series has suggested a possible association between anterior urethral valves (diverticulum) and syringocele. These congenital anomalies of the anterior urethra should be considered in the differential diagnosis of obstructive lesions of the urinary tract.

Key words

anterior urethral valves, syringocele, Cowper’s duct cyst, children

Introduction

Congenital obstruction of the anterior urethra is a rare deformity in boys. Whereas proximal obstruction is caused mainly by posterior urethral valves, the causes of distal obstruction are varied and less well recognized. Distal obstructions can be caused by anterior urethral valve (AUV), a diverticulum, cystic dilation of the ducts of Cowper's gland (syringocele), or valvular obstruction of the fossa navicularis. The diagnosis of these uncommon anomalies is clinically and radiographically difficult because they can produce the same symptoms and present similar radiographic findings. Moreover, their anatomical interpretation is unclear. A recent report has suggested that AUV and diverticulum formation may result from the distal lip of a ruptured Cowper's syringocele, acting as an obstructing valve leaflet.[1] In the present study, to elucidate the pathophysiology of congenital obstruction of the anterior urethra and to investigate the association between AUV and syringocele, we retrospectively evaluated three cases of congenital obstruction of the anterior urethra.

Case Report

Three boys (aged 6 months to 9 years) with congenital obstruction of the anterior urethra diagnosed at our department between 1997 and 2004 were analyzed retrospectively. The presenting symptom and age of each patient were varied (**Table 1**). Voiding cystourethrography (V-CUG) with pressure flow study suggested anterior urethral obstruction. The degree of the obstruction was varied, however thought to be adequately large to cause lower urinary tract symptoms. V-CUG showed bladder deformity in all and vesicoureteral reflux (VUR) in Case 2. and 3. Pressure flow study demonstrated ideopathic detrusor overactivity during storage phase in all cases. Small functional bladder capacity and hypercontractility of detrusor were found during voiding phase. Post-voided residual urine was detected in Case 2. Urethrocystoscopic examination suggested Cowper's duct and the diverticulum in bulbar urethra in all. Our series suggested a possible association between AUV (diverticulum) and syringocele.

Case 1

A 9-year-old boy had had refractory urge incontinence and nocturnal enuresis. Transabdominal ultrasound showed bladder wall thickness. V-CUG demonstrated AUV accompanied by a diverticulum in the penoscrotal urethra (**Fig. 1**). Urethrocystoscopy showed the ostium on the ventral aspect of the bulbar urethra. We speculated that it may be an ostium of a ruptured syringocele. A dilated prostatic

urethra, hypertrophic bladder neck and trabeculated bladder were noted. The distal and proximal wall of the syringocele was electroresected. Six months later, internal urethrotomy was required because of iatrogenic stricture of the more distal urethra confirmed by postoperative V-CUG. The patient became free of symptoms 29 months later.

Case 2

An 8-year-old boy was further evaluated for refractory urge incontinence and enuresis. Transabdominal ultrasound showed bladder wall thickness and post-voided residual urine. V-CUG showed AUV in the penoscrotal urethra and concomitant unilateral grade 4 VUR (**Fig. 2**). Radioisotope renal scintigraphy showed left renal scarrings. Urethrocystoscopy disclosed membranes on both side of the bulbar urethra. We speculated that it may be the remnant of a cyst wall. It was observed endoscopically that the distal portion of the membranes gradually distended by running normal saline solution and that the distended diverticulum-like swelling finally compressed the interior urethra upon. The distal portion of the membranes was resected sharply. After 24 months of close follow-up, although the patient became continent during the daytime with occasional enuresis, unilateral ureteric re-implantation was performed because of uncontrolled febrile urinary tract infection (UTI). Four years after the endoscopic correction, the patient had no urinary symptoms

and no sign of recurrent UTIs.

Case 3

A 6-month-old baby was further evaluated for left acute pyelonephritis with hydroureteronephrosis. Ultrasound showed bladder wall thickness. V-CUG provided a radiographic diagnosis of AUV, bladder deformity and unilateral grade 5 VUR (**Fig. 3**). Urethroscopy revealed a cavity with a widely patulous orifice on the ventral aspect distal to the external sphincter. We speculated that it may be the diverticulum of a perforate syringocele. The wide orifice was resected sharply. Six months after the endoscopic correction, V-CUG demonstrated improvement of the bladder deformity and persistent unilateral grade 3 VUR. While the VUR persisted, UTI was well managed without clinical symptoms with prophylactic antibiotics and propiverine hydrochloride.

Discussion

Congenital obstruction is much less common in the distal urethra than in the proximal urethra. However, it may also appear as an underlying etiologic factor that is responsible for a wide spectrum of lower urinary tract symptoms. Whereas the forms of congenital obstruction can vary, AUV – usually accompanied by diverticulum – is the most common.[2] However, the anatomical interpretation of AUV and diverticulum is unclear,[3] and it remains controversial whether the primary lesion is truly a valve or a diverticulum.[1] It is considered that the basic difference is in the contiguity between the anomaly and the corpus spongiosum.[4] Syringocele is also regarded as one of the anterior urethral obstructions, creating cystic dilatation of the duct of the bulbourethral (Cowper's) gland.[5] The distal margin of the AUV or the distal portion of the syringocele can cause infravesical obstruction. The obstruction is generally minor, and not all cases require treatment. Rarely, however, the anomaly may be sufficiently large to cause various lower urinary tract symptoms, essentially depending on the severity of the outlet obstruction.[2] The clinical presentations are varied, and can include palpable abdominal masses (distended bladder, bladder rupture, or ascites), hydroureteronephrosis, renal insufficiency or respiratory distress resulting from pulmonary hypoplasia in the prenatal and newborn, and afebrile/febrile UTIs or voiding dysfunction such as incontinence and slow stream in young boys.

The diagnosis is generally made by V-CUG. The confirmed diagnosis is made

by urethrocystoscopy. In our series, whereas all patients were radiographically diagnosed as AUV, urethroscopy speculated a Cowper's syringocele. Recently a common association between syringoceles and Cobb's collar, suggesting a common origin, has been noted.[6] Our results support the suggestion that AUV and diverticulum formation may result from the distal lip of a ruptured syringocele,[1] but the association between syringocele and Cobb's collar was not confirmed.

The main therapeutic option is commonly made by urethroscopy. Postoperative V-CUG may show an unchanged diverticulum.[2] However, the diverticulum-like defect is rarely obstructive and generally does not require further surgical correction. Complications of urethroscopic surgery are various, including persistent urethral dilation, urethral stricture and urethra-cutaneous fistula.

While various degrees of vesical dysfunction can be caused by posterior urethral valves, there are few reports of vesical dysfunction caused by anterior urethral obstructions.[7] In our series, preoperative videourodynamic demonstrated various types of vesical dysfunction, such as small functional bladder capacity, idiopathic detrusor overactivity, hypercontractility, bladder deformity and VUR. In general, VUR resolves spontaneously after correction.[2] However, if a patient with AUV or syringocele has a thick-walled, poorly compliant bladder and VUR, careful postoperative management of the lower and upper urinary tract must be necessary.

The incidence of cystic abnormalities of Cowper's duct ranges from 1.5% on

V-CUG[6] to 2.3% when revealed by autopsy.[8] Among 61 boys with refractory voiding dysfunction consulting our department, the incidence of AUV or syringocele was 3.3%.[9] The conditions may be overlooked due to unfamiliarity[10] or difficulty with clinical diagnosis. Anterior urethral obstruction in boys may be more common than currently believed.

In conclusion, our series has speculated a possible association between AUV (diverticulum) and syringocele. These anomalies of the anterior urethra should be considered in the differential diagnosis of obstructive lesions of the urinary tract.

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Figure legends

Fig. 1-a

V-CUG showed anterior urethral valves accompanied by a diverticulum in the penoscrotal urethra (indicated by the solid arrow).

Fig. 1-b, c

The ostium of a ruptured Cowper's duct in the bulbar urethra (solid arrow). The wall of the syringocele was electroresected.

Fig. 1-d

Postoperative V-CUG showed iatrogenic stricture of the more distal urethra (dashed arrow).

Fig. 2-a

Preoperative image of anterior urethral valves and left vesicoureteral reflux (indicated by the solid arrow).

Fig. 2-b

Radioisotope renal scintigraphy showed left renal scarrings.

Fig. 2-c

Urethrocystoscopy disclosed membranes (solid arrow) on both side of the bulbar urethra which appeared to be the remnant of a cyst wall (a perforate syringocele).

Fig. 2-d

It was confirmed that the distended diverticulum-like swelling compressed the interior urethra.

Fig. 3-a

V-CUG showed anterior urethral valves (indicated by the solid arrow), bladder deformity and unilateral grade five vesicoureteral reflux.

Fig. 3-b

Radioisotope renal scintigraphy.

Fig. 3-c

Urethroscopy revealed a cavity on the ventral aspect distal to the external sphincter, with a widely patulous orifice of a perforate syringocele (solid arrow).

Fig. 3-d

Postoperative V-CUG demonstrated improvement of bladder deformity and persistent grade three left reflux.

Table legend

Table 1

Clinical details of all patients with congenital obstruction of the anterior urethra

TU; transurethral incision or electroresection

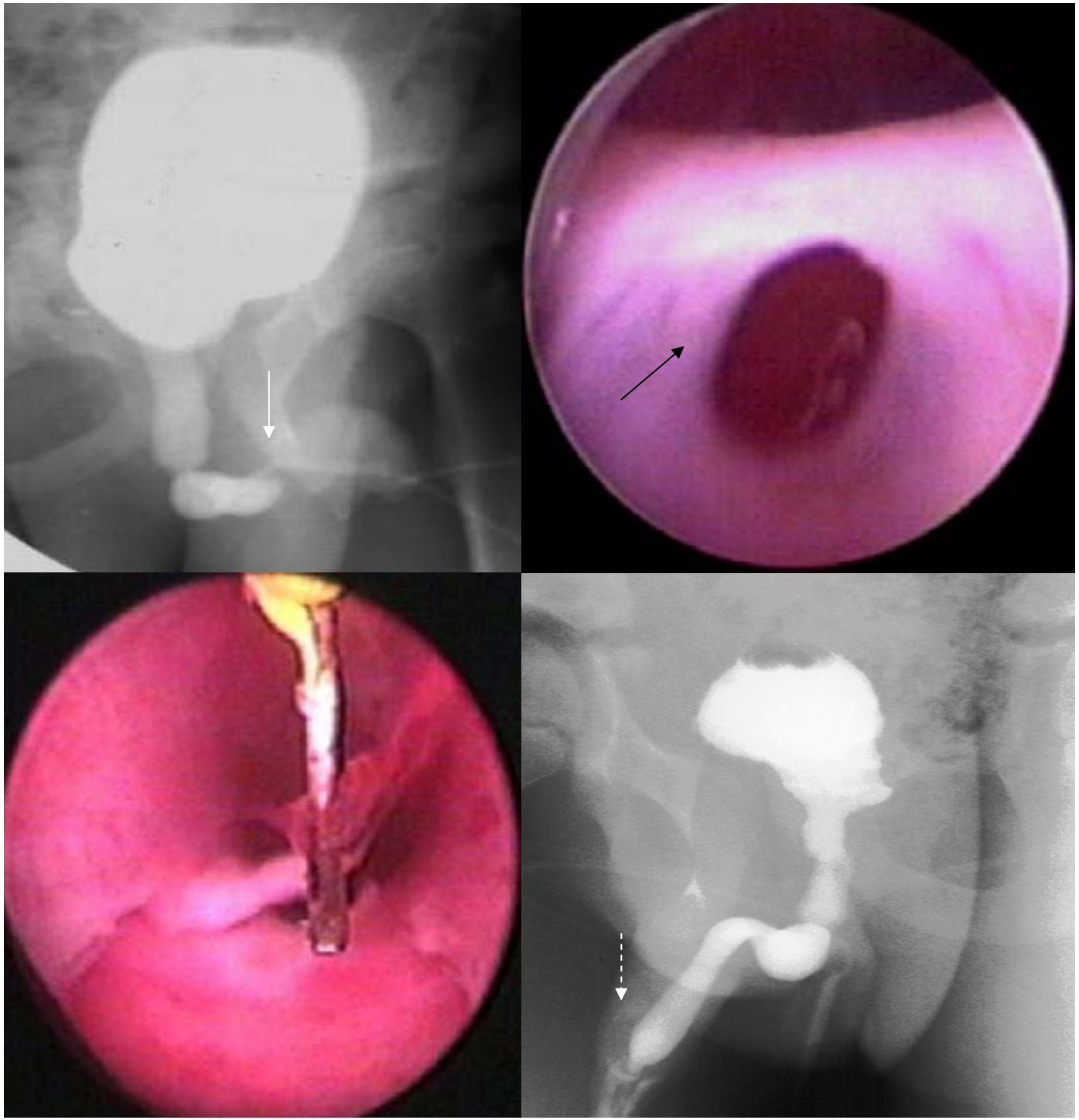

Fig. 1 a b / c d

Case 1: 9 yr

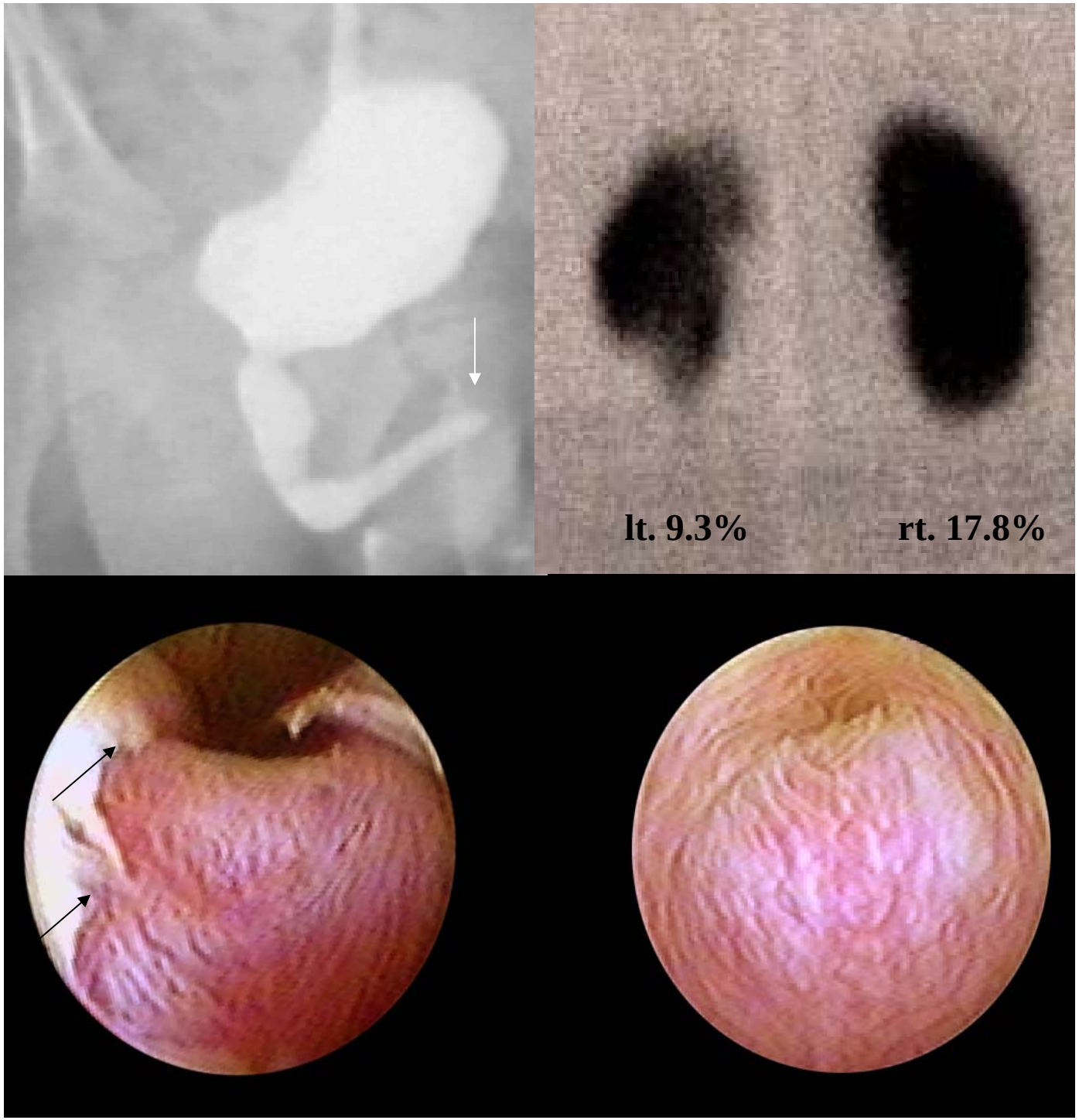

Fig. 2 a b / c d

Case 2: 8 yr

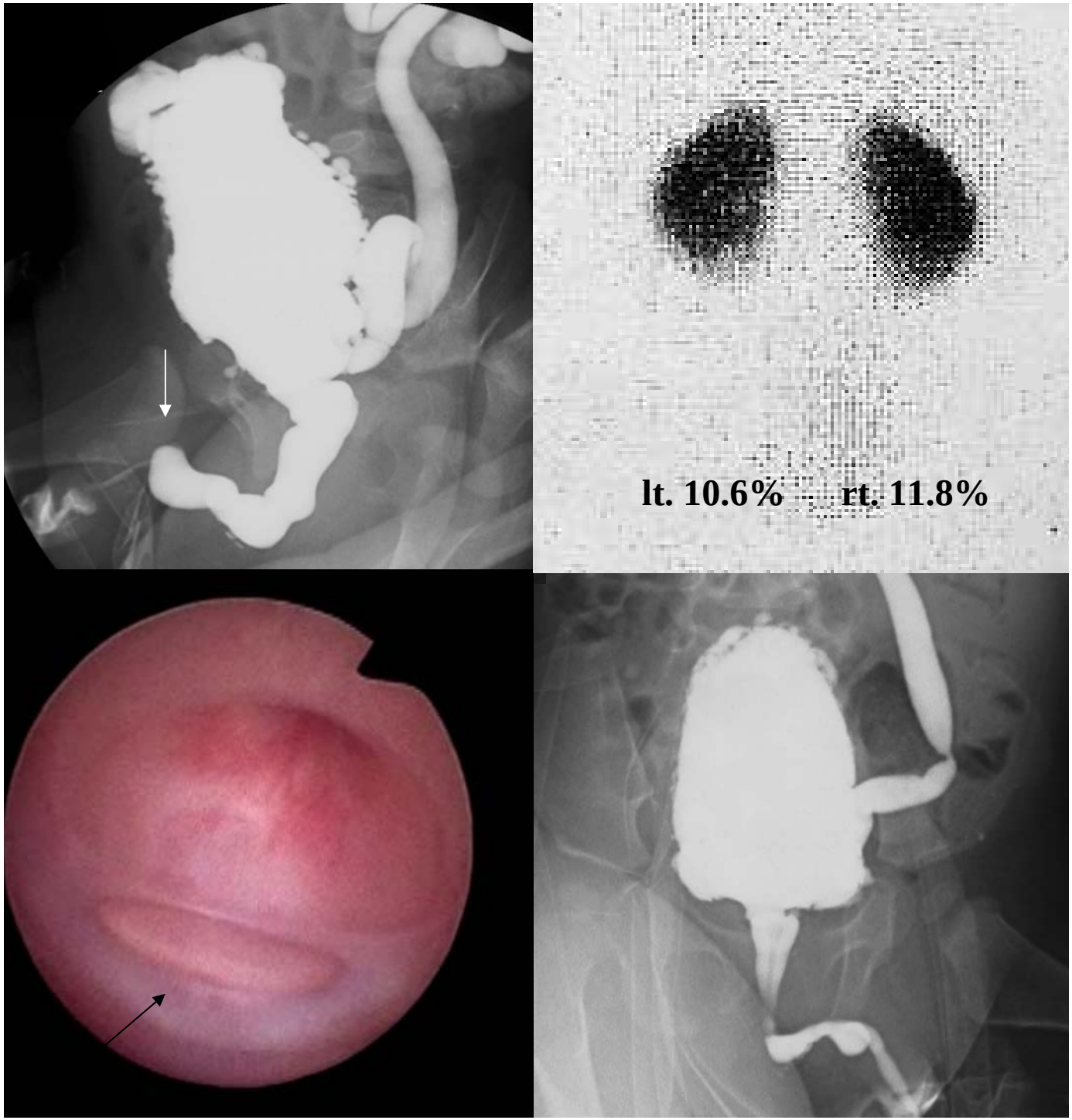

Fig. 3 a b c d

Case 3: 6 M

Table 1

Case No. (Age)	Clinical presentation	History of UTI	Enuresis	Radiographic diagnosis Position	Treatment	Outcome
1 (9 yr)	Refractory urge incontinence, urgency and enuresis	None	Every night	AUV with diverticulum in the penoscrotal urethra	TU	Cure of incontinence, but internal urethrotomy was required due to iatrogenic urethral stricture
2 (8yr)	Refractory urge incontinence, slow stream and nocturia	Fever of unkown orgin	Every night	AUV in the penoscrotal urethra	TU	Cure of incontinence, but anti VUR surgery was underwent due to remaining left VUR, caused febrile UTI
3 (6 M)	Febrile UTI and left hydroureteronephrosis	Febrile UTI	Unknown	AUV in the penoscrotal urethra	TU	While VUR persisted, UTIs were well managed without clinical symptoms.

TU: transurethral incision or electroresection