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<http://www.pediatricurologycasereports.com>**Anterior urethral valves without diverticulum, a rare cause of infravesical obstruction and vesicoureteral reflux in children: Report of two cases and literature review****Jyoti Bothra, Charu Tiwari, Gursev Sandlas, Hemanshi Shah***Department of Pediatric Surgery, TNMC & BYL Nair Hospital, Mumbai Central, Mumbai, Maharashtra, India***ABSTRACT**

Congenital anterior urethral valve is a rare condition causing significant obstructive uropathy in pediatric age group. It is much rarer than posterior urethral valve. However, the clinical course is similar. We present two cases of anterior urethral valves in children.

Key Words: Urethral valves, anterior urethra; children.

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Introduction

Congenital anterior urethral valve is a rare cause of obstructive uropathy [1,2]. The diagnosis of AUV is difficult and can be missed; this situation may lead to a spectrum of complications ranging from simple urethral dilatation to bilateral hydronephrosis resulting in end-stage renal disease (ESRD) [3,4]. A well performed radiological investigation is imperative for diagnosis [2,5]. Although much rarer than posterior urethral valves, they follow the same clinical course and an early diagnosis and treatment can prevent renal morbidity in these patients. We report two such cases with different ages of

presentation. A brief review of the literature is also presented.

Case 1

The patient presented to us at 8 years with history of poor stream and dribbling of urine. He was investigated elsewhere and diagnosed as bilateral grade V vesicoureteral reflux. Two cystoscopies at the age of 9 months and 2 years were reported as cystitis with dilated ureteric orifices and a normal urethra. Patient was on chemoprophylaxis. He had persistence of symptoms and a deteriorating renal function on renal scans. A repeat voiding cystourethrography at 8 years of age showed a thin membrane like shadow in mid penile urethra suggesting presence of an anterior urethral valve [Fig. 1]. Transpenile micturating cystourethrosonography [Fig. 2] showed a semilunar valve in proximal penile urethra and a sacculum with obstruction to the stream of

urine. Patient underwent a cystoscopic fulguration of the anterior urethral valves. On a follow up, transpenile sonography and a check cystoscopy at 6 weeks no residual valve or obstruction to flow was seen. The child is on regular follow up with down gradation of reflux.



Fig. 1. Voiding cystourethrogram of the first patient showing a thin membrane like shadow in mid penile urethra suggesting anterior urethral valve.



Fig. 2. Preoperative transpenile micturating cystourethrosonography showing a semilunar valve-like structure at membranous urethra with dilated posterior urethra causing obstruction to the urine stream.

Case 2

A 2 month old child was referred with complaints of excessive crying during micturition. Voiding cystourethrogram [Fig. 3] delineated a negative shadow in the mid penile region suggesting anterior urethral valve. At cystoscopy, anterior urethral valves were confirmed and fulgurated. A check cystoscopy at 6 weeks showed no residual obstructing valves.



Fig. 3. Voiding cystourethrogram of the second patient showing a negative shadow in the mid penile region suggesting anterior urethral valve.

Discussion

Congenital anterior urethral valve is a rare condition causing significant obstructive uropathy. The first description of the condition was reported by Watts in 1906 [6]. Although the occurrence of the condition is ten to fifteen times less than the posterior urethral valves, the obstruction pattern can vary.

Anatomically the anterior urethral valves may be present anywhere distal to the membranous urethra. 40% are found in bulbar urethra, 30%

in penoscrotal junction and 30% in penile urethra [7]. Various theories have been put forth for the embryology of the valves. Incomplete formation of the ventral corpus spongiosum or imbalance of tissue growth, congenital cystic dilatation of periurethral gland or an abortive attempt at urethral duplication is few of these theories [2,8].

The clinical presentation varies depending on the age and degree of obstruction. Some patients may have subtle symptoms while others may develop urinary incontinence, retention, weak urinary stream, strangury, post micturation dribbling, urinary tract infections or urosepsis. The genitourinary tract may show changes of mild urethral dilatation or bilateral hydronephrosis due to reflux with renal function deterioration as seen in reported cases. One third of these patients have an associated diverticulum [8]. Firlit [9] had classified the obstructive changes accompanying the anterior urethral valves depending on the degree of urethral dilatation, presence of a diverticulum and grade of upper tract dilatation.

Antenatal diagnosis of anterior urethral valves is rare and if done is in patients with gross bilateral hydronephrosis or severe megaureters and/or mega cystitis. The investigation of choice is a voiding cystourethrography. It is imperative to obtain and evaluate the voiding phase with the oblique view to avoid missing the diagnosis [10]. Transpenile ultrasound is complementary to confirm the diagnosis [11]. The rest of the investigations include the serum renal chemistries, urine routine microscopy and culture and renal scans.

The aim of the treatment of AUV is to excise the urethral valve and to re-establish the continuity of the urethra [2]. Historically the management of urethral valves was open resection and reconstruction of diverticulum

[12]. With the advent of newer technology as smaller cystoscopes are available, today, endoscopic fulguration is the standard treatment. A diverticulectomy and reconstruction of urethra is rarely required in cases with large associated diverticulum [13, 14]. The efficacy of prenatal intervention awaits further evaluation, though it is believed that pulmonary maturation is successfully attained and the exacerbation of the renal problems is prevented [13-15].

The prognosis of patients with AUV is usually good [12]. Routh et al. [16] had noted in their study that the prognosis of this condition was associated with the renal chemistry. Patients with high pre-operative creatinine levels and high grade vesicoureteric reflux and infections had a 25 fold increase in poor renal outcome post treatment also. Lower grade reflux is known to improve after fulguration; however, higher grades may persist and require further intervention [12]. This highlights the importance of long-term follow-up [12].

Conclusion

Anterior urethral valves are a rare congenital disorder affecting boys. A high degree of suspicion accompanied with good radiological diagnosis can lead to an early and precise diagnosis of anterior urethral valves in boys.

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