

Familial Urethral Stricture, Five Adult Patients Overview

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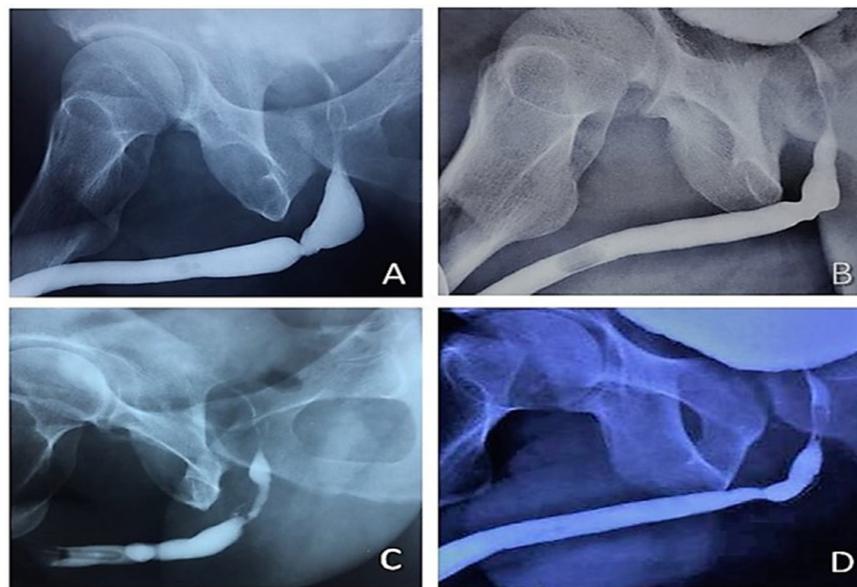
Congenital stricture, specifically with manifestation in adulthood is extremely a rare cause of urethral stricture and is not associated with known etiologies. It was first described by Cobb et al., and to our knowledge only 5 families were reported in English literatures to have familial urethral stricture.

We report two families with urethral stricture including five male patients referred to our tertiary reconstructive urology department during 1994 to 2017. The age and severity of symptoms at presentation are variable; as are the surgical interventions required. There are no phylogenetic, familiar or racial relationship between the two families described.

Keywords: urethral stricture; bulbar stricture; urethroplasty; Cobb's urethral stenosis

INTRODUCTION

The term urethral stricture mainly refers to an anterior urethral disease occurring for the most part secondary to pelvic trauma, inflammatory conditions, iatrogenic or surgical interventions including urethral instrumentation.^(1, 2) Familial stricture, specifically with presentation in adulthood, is an extremely rare cause of urethral stricture. This phenomenon was described by Cobb et al. (1968), who proposed that the obstacle forms from partial fusion of the anterior and posterior urethra. The fusion arises from incomplete opening of the urogenital membrane during the embryonic phase of development.⁽³⁾ We report two families with familial urethral stricture, referred to our tertiary reconstructive urology department during 1994 to 2017. To the best of us knowledge, only 5 families have been reported in the literature.⁽⁴⁻⁷⁾



A. Cobb's bulbar stricture
B. RUG of 1year post operation follow up of case A
C. Cobb's bulbar stricture (post traumatic catheterization)
D. RUG of 20 months post-DVIU follow up of case C

Figure 1. Retrograde Urethrography of patients.

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CASE REPORT

Series 1

Three brothers presented, all with complaints of difficult voiding. The youngest brother's obstructive urinary symptoms started 25 years ago (at 27 year old age), but he did not have any intervention until he referred to our clinic due to severity of symptoms. Retrograde urethrogram (RUG) plus, simultaneous voiding cystogram showed a small stricture in the mid bulbar urethra. (**Figure A**)

He underwent Direct Vision Internal Urethrotomy (DVIU); and there was no symptom recurrence during one year follow up. (**Figure B**)

The second brother (55-years-old) underwent two urethral dilatations in the last 5 to 7 years, also due to mid-bulbar urethral stricture. The oldest brother (65-year-old) presented to the clinic 9 years ago following an unsuccessful urethral catheterization after an acute urinary retention. RUG showed bulbar urethral stricture. (**Figure C**) DVIU was done and there was no recurrence of stricture. (**Figure D**) All three brothers of this family underwent close follow up postoperatively without any self-catheterization order and so far, they are happy with their micturition.⁽⁸⁾

Series 2

The second series was a family with two brothers. The older brother, (56 years old), underwent DVIU in 1984 for a web-shaped mid-bulbar stricture without any known etiology. He had history of 10 DVIU procedures within 15 years due to his refusal to consent to open surgery. He underwent end-to-end bulbar urethroplasty in 2001. The younger brother (47-year-old) had a web-shaped bulbar stricture which initially presented at 31 years old.

He underwent DVIU every 2 years before he was referred to our hospital in 2006 for open surgery and an end-to-end urethroplasty revealed a 2 cm bulbar stricture. Although he was asymptomatic for about 1 year after the surgery, but his bulbar urethra progressively narrowed and he had repeat urethral dilatations in 2008, 2010 and 2014.

DISCUSSION

The overwhelming majority of congenital urethral strictures are referred to as a variety of posterior urethral valve (PUV) disorders. Familial PUVs are rarely reported.⁽⁹⁾ The stenosis in the above cases is called Cobb's Collar, which is a membranous stricture of the bulbar urethra distal to the external urethral sphincter. The location differentiates this issue from PUV^(10,11). Although presentation in adulthood suggests a trauma or inflammation history, the site of strictures in the above cases exactly matches the definition of Cobb's. The similarity of symptoms, age of incidence and response to the treatment in the above individuals suggests a familial disease.⁽⁵⁾ There are no phylogenetic or familiar relationship between the two families described.

Moreover, since other reported cases have a world-wide distribution, racial factors do not seem to be involved in these presentations.

The age and severity of symptoms at presentation are variable⁽⁴⁾; as are the surgical intervention required. As seen in our cases, differs from a minimally invasive DVIU to urethroplasty and multiple urethral dilatations.

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