Management of female congenital short patulous urethra with urethral tapering and pubovaginal sling: A report of two cases

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INTRODUCTION

Congenital short patulous urethra in females is a rare condition and has been previously reported in association with epispadias, hypospadias, and urogenital sinus anomalies.[1] MRKH syndrome is a type of mullerian duct malformation characterized by congenital absence or hypoplasia of the uterus and the upper two-thirds of the vagina in phenotypically and genotypically normal females with functional ovaries. These patients may also have associated urological or skeletal abnormalities. Urethral coitus is reported as a cause of SUI in these patients, but primary short patulous urethra causing SUI has been reported in only two cases.[2]

Uterus didelphys with longitudinal vaginal septum is a fusion defect of mullerian ducts with usual presentation of dyspareunia. Short patulous urethra causing SUI is a very rare presentation.

We report our experience of surgical management of two rare cases of short patulous urethra causing SUI.

CASE REPORTS

Case 1

A 14-year-old girl presented with complaints of cyclical lower abdominal pain every month for the last 1 year (primary amenorrhea) along with SUI while walking, coughing, and laughing (Grade 2 as per the Ingelman–Sundberg–Stamey classification scale) since childhood. She used to void normally (150–200 ml) and remained dry during night time.

Physical examination revealed normal secondary female sexual characteristics. Perineal examination revealed normal labia majora, labia minora, and clitoris with a single opening in the vulva, with urinary leakage on suprapubic compression [Figure 1a].

Her karyotype was 46 XX and the hormonal profile was normal. Magnetic resonance imaging (MRI) showed a septated cystic structure in the left adnexa with a normal right ovary. Uterus was bicornuate with hematometra. Lower uterine segment including the cervix and vagina was absent. Micturating cystourethrogram (MCU) was
suggestive of normal capacity bladder with wide patulous bladder neck and urethra [Figure 1b].

Cystometrogram (CMG) (occlusion of bladder neck by balloon catheter after determining leak pressures) revealed a stable bladder (normal compliance without any detrusor overactivity DO) with a Valsalva leak point pressure (VLPP) of <20 cmH₂O, suggestive of intrinsic sphincter deficiency (ISD). Cystourethroscopy confirmed single opening in vulva to be dilated patulous urethra (minimal coaptation) with normal bladder capacity. Bilateral ureteric orifices were situated at normal location.

After confirming the diagnosis of MRKH syndrome with SUI, the patient was planned for surgical repair with a realistic aim of creating vagina for intercourse, restoring continence, and restoring normal cyclical menstruation (since uterus was present).

Excisional tapering of urethra (wedge shaped on ventral aspect with the apex up to the bladder neck) with pubovaginal sling (rectus sheath) placement with excision of the left rudimentary horn of the uterus and the left tubo-ovarian mass along with sigmoid vaginoplasty was performed [Figure 2a–c]. The lower end of the uterus was sutured to the upper end of sigmoid neovagina over a 14 Fr Malecot catheter which was left in place for 6 weeks to ensure canalization. Postoperatively, the patient was continent and was menstruating monthly. The neovagina was pink and healthy with mild stenosis at the introitus which stabilized after monthly dilatations performed in the first 6 months [Figure 2d]. The patient was dry and menstruating normally with normal neovagina on subsequent follow-up at 1 year. Postoperative Urodynamic study (UDS) was suggestive of a stable bladder (normal compliance without DO or leakage) with a capacity of 300 cc. Voiding phase was also normal with insignificant residual urine.

**Case 2**

A 12-year-old girl presented with leakage of urine while at rest, walking, or coughing (Grade 3 SUI) since birth. She used to void small volumes of urine (approximately 100 ml) six to eight times in the day and remained wet during the night. She had normal regular monthly menstruations. Physical examination revealed normal secondary female sexual characteristics. Perineal examination revealed normal labia majora, labia minora, clitoris, and a septated vagina with a wide patulous urethra with urinary leakage on suprapubic compression [Figure 3a]. Her MCU was suggestive of wide patulous urethra and bladder neck with good bladder capacity [Figure 3b]. MRI scan [Figure 3c and d] was suggestive of normal bilateral ovaries with uterus didelphys, double cervix, and a septated vagina (longitudinal septum). CMG revealed stable bladder of capacity around 400 cc (normal compliance without any DO) with VLPP of <20 cmH₂O suggestive of ISD.

Cystourethroscopy was suggestive of short patulous urethra with wide bladder neck. Bladder capacity was normal with normally situated bilateral ureteric orifices.

Excisional tapering of the urethra (wedge shaped on dorsal aspect with the apex up to bladder neck) with rectus sheath pubovaginal sling placement was performed [Figure 4a].
Incision for the placement of pubovaginal sling was not made on the vaginal wall. Instead, we entered the same suburethral space by incising the triradiate septum of introital mucosa at the confluence of both the hemivagina and external urethral orifice [Figure 4b and c]. Postoperatively, the patient improved drastically and was completely dry on the follow-up at 3 months.

**DISCUSSION**

In the case of SUI and ISD, pubovaginal slings are more effective than the retropubic colposuspensions, with outcomes comparable to those reported with the midurethral slings. However, in cases of short patulous urethra with severe urinary incontinence, the sole dependence on midurethral slings for continence is unjustified. The artificial urinary sphincter provides a long-term cure but is associated with considerable morbidity and failure within 5–10 years, whereas the clinical results of bolking agents are not sustained beyond the first year.

In patients with congenital severe SUI due to dilated, short patulous urethra with a defective sphincter mechanism, the goal of urethral reconstructive surgery should be directed to achieving adequate urethral coaptation and bladder neck support. Pubovaginal sling using autologous tissue is considered the gold standard procedure for the treatment of urinary incontinence in the cases of defective sphincter mechanism in females.[3] None of our patients had a previous history of sexual intercourse. Both the patients had a short patulous urethra with defective sphincter mechanism, so pubovaginal sling was also placed along with urethral tapering.

Dilated patulous urethra with wide caliber can be tapered or plicated over a 12 or 14 Fr Foleys catheter so as to achieve urethral mucosal coaptation, in turn helping the continence mechanism. Good results have been reported with urethral tapering[4] and urethral plication[5] for patients with urinary incontinence and dilated patulous urethra due to urethral coitus, but this may not result in complete continence as reported by Khattar et al.[4] It is not advisable to rely only on urethral tapering in a congenital patulous urethra where the bulk of the submucosal tissue and the presence of periurethral muscles is already questionable; hence, we chose to address the bladder neck mechanism also along with the improvement in coaptation.

Perimeatal–semicircular incision for urethral dissection to enable tapering can be made on the ventral or the dorsal aspect. Ventral incision is preferred in patients planned simultaneously for neovaginal formation as performed in our first case.[4] A dorsal incision in the second case was an automatic choice as she had a normal menstrual function and capacious hemivagina, so we intended not to disturb the genital tract. A modified incision for pubovaginal sling placement avoided incising the vaginal septum.

Both of our patients were treated with urethral tapering and pubovaginal sling (rectus sheath) placement at bladder neck with good postoperative results. Repair of continence mechanism and neovaginal creation can be performed in a single stage surgery with good results.

**CONCLUSION**

Management of SUI with short patulous urethra as part of developmental anomaly can be performed with urethral plication and pubovaginal sling placement at bladder neck with good results.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**REFERENCES**

1. Rajamaheswari N, Agarwal S, Chhikara AB, Seethalakshmi K. Anterior bladder flap neo urethra as treatment for stress urinary incontinence due to developmental urogenital anomaly. Urol Ann 2013;5:215-7.
2. Caione P, Silveri M, Capitanucci ML, Capozza N, De Gennaro M. Urinary incontinence in Müllerian duct anomalies. Panminerva Med 1995;37:14-7.
3. Rosenblum N, Nitti VW. Female urethral reconstruction. Urol Clin North Am 2011;38:55-64.
4. Khattar N, Dorairajan LN, Kumar S, Soundararaghavan S, Pal BC. Single-stage correction of urinary incontinence and vaginoplasty in a case of urethral coitus with vaginal agenesis. Urol Int 2008;81:364-6.
5. Sakinci M, Kokcu A, Malatyalioglu E. Satisfactory urethral coitus in a patient with vaginal stenosis: Case report. Int Urogynecol J 2012;23:237-9.

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