Çoc. Cer. Derg. 2021;35(3):155-8
doi:10.5222/JTAPS.2021.59329

Surgical Approach to a Late-Diagnosed Coexistence of Megalourethra, Double Vagina and Uterus Didelphys Presenting with Total Urinary Incontinence in an Adolescent Girl

Adolesan Kız Hastada Total Üriner Inkontinansla Seyreden Geç Tanı Almış Megaloüretra, Çift Vajina ve Uterus Didelfis Birlikteliğine Cerrahi Yaklaşım

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ABSTRACT

Mullerian duct anomalies can sometimes accompany urinary malformations and they can be diagnosed with different symptoms in late childhood. We aimed to present a thirteen year-old adolescent girl with megalourethra, double vagina and uterus didelphys who presented with total urinary incontinence. Her urogenital examination revealed a large orifice of megalourethra in the middle and two symmetric orifices of bifid vagina on both sides. An amorphous bladder with a large urethra was detected at cystoscopic examination. Urethral plication, resection of vaginal septum and reconstruction were performed. Congenital urological malformations associated with mullerian anomalies can be diagnosed with urinary symptoms in adolescence. Combined corrective surgery can be performed safely in a single session.

Keywords: *adolescent, double vagina, megalourethra, urinary incontinence, uterus didelphys*

ÖZ

Mülleriyen anomaliler üriner malformasyonlara eşlik edebilir ve geç çocukluk döneminde farklı semptomlarla tanı alabilir. Megaloüretra, çift vajina ve uterus didelfis birlikteliği olan ve total üriner inkontinans ile tanı alan 13 yaşında adolesan kız hastaya cerrahi yaklaşımı sunmayı amaçladık. Ürogenital muayenede, ortada megaloüretraya ait geniş orifis ve bu orifisin her iki yanında simetrik olarak yerleşmiş bifid vajinaya ait orifisler saptandı. Sistoskopik incelemede geniş bir üretra ve amorf bir mesane görüldü. Üretral plikasyon, vajinal septum rezeksiyonu ve vajinal rekonstruksiyon uygulandı.Müllerian anomalilere eşlik eden konjenital ürolojik maformasyonlar, adolesan dönemde üriner semptomlar ile tanı alabilir. Tek seansta kombine cerrahi düzeltme güvenle uygulanabilir.

Anahtar kelimeler: *adolesan, çift vajina, megaloüretra, uterus didelfis, üriner inkontinans*

Received/Geliş: 08.03.2021
Accepted/Kabul: 02.06.2021
First Publication: 14.09.2021

Cite as: Türedi Sezer B, Yılmaz MU, Parlak A, Sğlam S, Utangaç MM, Balkan E, Kılıç N. Surgical approach to a late-diagnosed coexistence of megalourethra, double vagina and uterus didelphys presenting with total urinary incontinence in an adolescent girl. Çoc. Cer. Derg. 2021;35(3):155-8.

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Introduction

Congenital anomalies of urinary tract can be seen in a wide range. Urethral anomalies are the most common types of lower urinary tract anomalies and they are more common in male patients. Dilation of urethra can be seen secondary to penile or urethral surgery in boys. Few cases with congenital dilation of urethra in boys have been reported and the pathology was thought to be developed by absence or hypoplasia of corpus spongiosa and cavernosa [(1)]. Urethral anomalies can also be associated with Mullerian duct anomalies in girls [(2)]. Congenital urethral dilation of an infant was first described as megalourethra by Nesbitt in 1955 [(3)]. Several cases have been reported about megalourethra from then but few of them were about female megalourethra. Diagnosis of this rare anomaly is difficult without specific symptoms. These girls with both urinary and Mullerian duct anomalies can present with recurrent urinary tract infections, dyspareunia and urinary incontinence during coitus. We aimed to present the surgical technique which we performed to an adolescent girl with total urinary incontinence who had a megalourethra, uterus didelphys and double vagina.

Case Presentation

A thirteen year-old adolescent girl referred to our clinic with total urinary incontinence. She never had a dry period since age of gaining toilet habit and she was wet all day and night. She had been using diapers all time. Many different anticholinergic treatments were used but none have been successful. She had recurrent urinary tract infections requiring antibiotic therapies. In detailed urinary tract ultrasonography both kidneys were normal and there were no pelvicalyceal or ureteral dilation. Urogenital examination revealed a large orifice in the middle and smaller two symmetric orifices at both sides of bifid vagina and sacral examination was normal. Lumbosacral graphy was normal and urodynamic evaluation showed a hypocompliant bladder with low capacity and detrusor instability. Pelvic magnetic resonance imaging which was planned for probable Mullerian duct anomalies revealed bifid vagina and uterus didelphys. Cystoscopic and genitoscopic examinations under general anesthesia were planned. A megalourethra and bifid vaginal orifices were seen at examination and cystoscopy revealed an amorphous bladder with a dilated urethra (Figure 1). Bilateral uterine cervices were seen at genitoscopic examination of both sided vaginal orifices. Urethral plication and vaginal reconstruction were planned. After mobilization of posterior wall of urethra, plication with tailoring was performed for megalourethra over an 10 F Foley catheter and vaginal septum was divided by 55 mm linear stapler (Ethicon®) (Figure 2). Neovaginal lumen had an adequate calibration and a small sterile gauze was inserted in. There were no intraoperative and postoperative complications. Patient was continent. Nocturnal enuresis regressed with behavioral therapy and anticholinergic agents. Her urogenital examination was normal 2 months after operation (Figure 3). Control cystoscopy and vaginoscopy at postoperative 6th month revealed a normal bladder, good calibrated urethra and vaginal lumen. Nine months follow-up was uneventful.

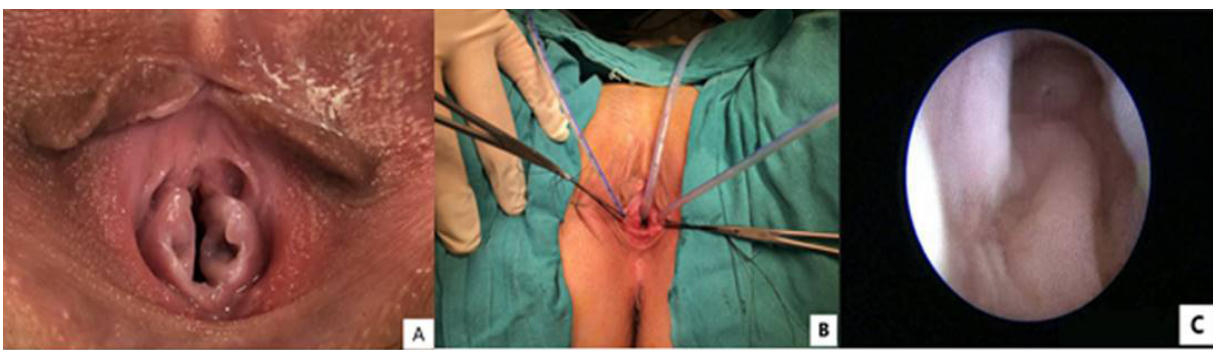

Figure 1. (A) Genital examination revealed a megalourethra in the middle and two vaginal orifices on both sides (B) View of catheterisated orifices (C) Large urethra and bladder neck seen in cystoscopy.

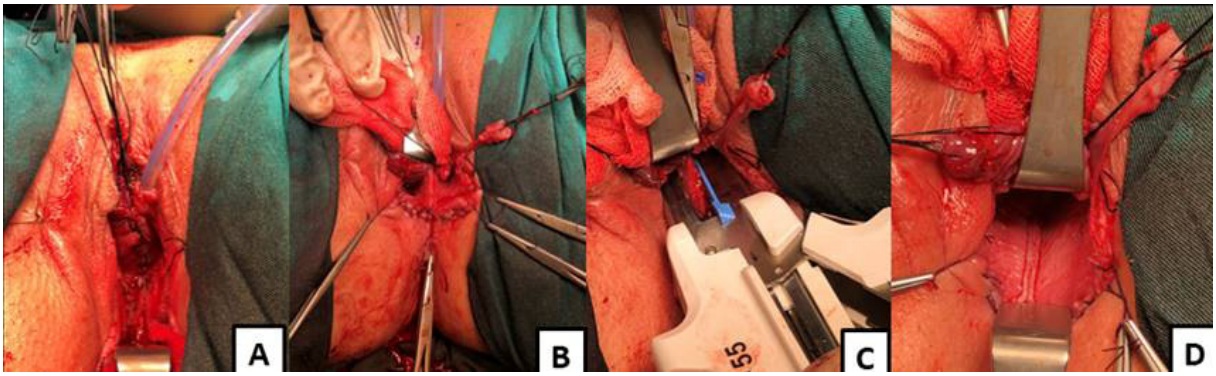

Figure 2. (A) Urethral plication with tailoring on a 10 F catheter (B) Reconstructed vaginal walls and vaginal septum (C) Resection of vaginal septum with linear stapler (D) View of vaginal lumen and orifice after resection.

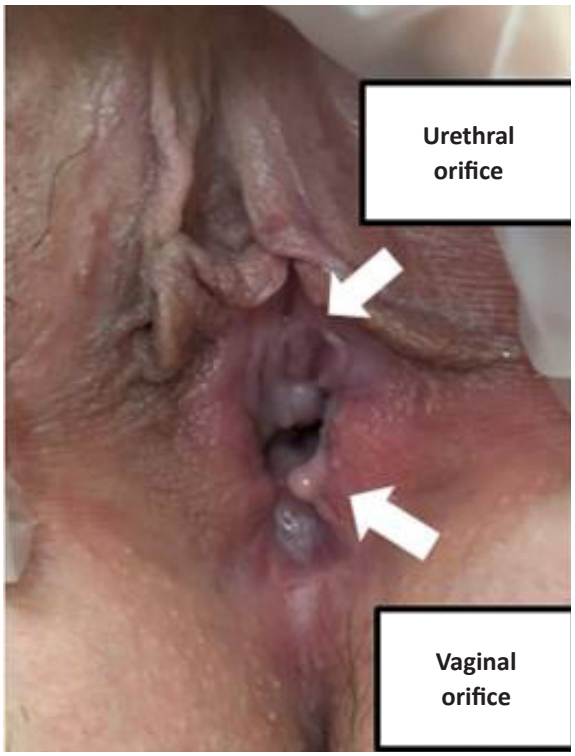

Figure 3. Perineal view at postoperative 2 months (showed with arrows).

Discussion

Dilation of urethra secondary to distal obstruction is a common entity. Secondary urethral dilation due to obstruction may develop especially after penile or urethral surgeries. Idiopathic dilation is rare and its etiology is unknown.

Congenital male urethral dilation which was named as megalourethra, was first decribed in 1955 by Nesbitt in a male infant [3]. Several case reports about megalourethra in boys have been published afterwards. Savanelli et al. reported a case with congenital megalourethra with urethral duplication and imperforate anus [4]. Ozcan et al reported a case with congenital megalourethra which was presented with an inguinal cystic mass in 2014 [5].

There are a few reports about female urethral dilation. It can be seen as a result of urethral coitus in the patients with unrecognized genital anomalies. Sharlflaghdas et al. reported a 38 year-old woman with Mayer-Rokitansky-Küster-Hauser Syndrome (MRKHS) who had experienced urethral coitus for many years when she was admitted with urinary incontinence and dyspareunia [6]. It is not common in adolescent patients and there are only a few reported cases in the literature. Ryckman et al reported two cases of megalourethra secondary to urethal coitus in adolescents [7]. One adolescent had vaginal agenesis and VACTERL (Vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula,renal anomalies and limb abnormalities) association and the other had complete oblique vaginal septum and non communicating-functioning uterine horn. Our patient was virgin and had no history of sexual activity.

Congenital urethral anomalies can be seen with Mullerian duct anomalies in female patients. Stuppler et al. reported a female patient with congenital megalourethra which had other additional anomalies [8]. They reported that the patient had urgency and nocturnal incontinence. She had uterus didelphys, complete duplication of the vagina and megalourethra.

Complete resection of vaginal septum and urethral plication were done. Several etiological factors may play a role in congenital urethral dilation such as hypertonicity of muscular fibers in urethra resulting in the development of a functional disorder, neurogenic dysfunction of the urethra and bladder, forced bladder emptying and dysfunction and myogenic deficiency of helicoidal muscle fibers [8]. Since our patient had no other dysplastic syndrome, possible cause was thought to be myogenic deficiency of helicoidal muscles [8]. To our knowledge our case is the only case which is completely similar to that case.

Conclusion

Congenital urethral dilation is commonly seen in male patients. There are reported cases of female patients with Mullerian duct anomalies who were diagnosed with megalourethra due to urethral coitus. Urinary incontinence and dyspareunia may be the diagnostic symptoms for these patients. Congenital megalourethra can be seen in female patients with Mullerian duct anomalies such as uterus didelphys, complete vaginal septum, vaginal agenesis or MRKH syndrome. Diagnosed coexistence of congenital megalourethra and other genital anomalies should be kept in mind for adolescent female patients admitted with urinary incontinence who do not have a history of sexual activity.

Conflict of Interest: The authors declare no conflict of interest.

Funding: None.

Informed Consent: Informed consent was obtained from parents for the publication of this case report.

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