SCAPHOIDAL MEGALOURETHRA: A RARE CASE PRESENTATION

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ABSTRACT

Objective: *To propose reduction urethroplasty as a technique for urethral reconstruction for scaphoidal megalourethra.* ***Case(s) Presentation:*** *A male child with the age of one year and four months was known to suffer from megalourethra when dorsumcision was done. The procedure was done starting with marking sutures at the penile gland. Disclose the penile skin as proximal as possible. When disclosed, identify the urethra and dilation was seen so as a finger could be inserted for around 2 centimeters. Urethra was incised, then excise the excess urethral tissue, suturing was done based on the 6F Folley catheter to the point where the urethra dilates. After Folley catheter was inserted, inversion suture was done on the urethra with PDS 6.0 and glanulopasty with PDS 6.0. After evaluation, urethral lumen was found not narrowed, skin was sutured with PDS 6.0. Cystostomy was inserted to divert urine and maintained until one week post-operation.* ***Discussion:*** *Cosmetic and functional results were good and satisfactory. Patient could urinate with no hindrance. Post-operation complication was not found. However, erectile function was difficult to obtain in pediatric patients.* ***Conclusion:*** *Megalourethra is a rare case that needs appropriate diagnosing and management to prevent complication. Operation technique of reduction urethroplasty was done as the chosen management for scaphoidal megalourethra. Furthermore, more in depth research with better methodology was needed to determine the outcome of patients with megalourethra.*

Keywords: *Scaphoidal megalourethra, reduction urethroplasty, urethral reconstruction.*

ABSTRAK

Tujuan: *Mengusulkan uretroplasti reduksi sebagai teknik rekonstruksi uretra untuk scaphoidal megalourethra.* ***Presentasi Kasus:*** *Anak laki-laki berusia satu tahun dan empat bulan diketahui menderita megalourethra ketika akan menjalani dorsumsisi. Prosedur ini dimulai dengan membuat jahitan di glans penis, lalu kulit penis dibuat tertutup se-proksimal mungkin. Setelah tertutup, uretra diidentifikasi dan dilebarkan dengan jari hingga dapat dimasukkan sekitar 2 cm. Uretra lalu diinsisi, kemudian jaringan uretra yang berlebih dieksisi, penjahitan dilakukan dari awal dimana kateter folley 6F berada hingga di titik dimana uretra melebar. Setelah kateter folley dimasukkan, jahitan inversi dilakukan pada uretra dengan PDS 6.0 dan granuloplasty dengan PDS 6.0. Setelah dievaluasi, lumen uretra ditemukan tidak menyempit, kulit dijahit dengan PDS 6.0. Sistostomi dipasang untuk diversi urin dan dipertahankan hingga satu minggu post-operasi.* ***Diskusi:*** *Hasil kosmetik dan fungsional dianggap memuaskan. Pasien dapat buang air kecil tanpa hambatan. Komplikasi pascaoperasi tidak ditemukan. Namun, fungsi ereksi sulit didapatkan pada pasien anak.* ***Simpulan:*** *Megalourethra adalah kasus langka yang membutuhkan diagnosis dan tatalaksana yang tepat untuk mencegah komplikasi. Teknik operasi uretroplasti reduksi dilakukan sebagai tatalaksana yang dipilih dalam penanganan megalourethra scaphoid. Penelitian yang lebih mendalam dengan metodologi yang lebih baik diperlukan untuk menentukan hasil pasien dengan megalourethra.*

Kata Kunci: *Scaphoidal megalourethra, uretroplasti reduksi, rekonstruksi uretra.*

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INTRODUCTION

Megalourethra is a congenital anomaly with the characteristic of dilatation of the urethra. According to a reference by Promsonthi et al. in 2010, only 28 cases of megalourethra especially diagnosed prenatally from 20 articles collected.[1] Cheng et al. also suggested that only 47 cases of megalourethra were reported until 2016. This rare case is divided into two types, scaphoidal and fusiform. Each type of megalourethra has a different representation as well as management. In this case,

we present a case of a male child diagnosed with megalourethra after dorsumcision.

CASE(S) PRESENTATION

A male child with the age of one year and four months came to Anak Bunda Harapan Kita Hospital with a diagnosis of megalourethra obtained by dorsumcision. Parents admitted the patient did not complain when admitted. Patient was born full term, delivered at a hospital via caesarean section with weight of 3.400 grams, and length of 51 cm. History of immunization and growth and development within normal limit. There was no history of other congenital disorders. From retrograde urethrography, dilation of the pendulous urethra for 2 cm was found, normal posterior urethra, the contrast was found filling the vesica, VUR was not found. From urethrocystoscopy, dilation of pendulous urethral was found, whereas bulbous urethra to the bladder neck looked normal, normal sphincter, normal verumontanum, and normal vesical mucosa. The conclusion of the urethroscopy is congenital scaphoid type megalourethra for 2 cm. The patient then underwent urethral reconstruction operation with reduction urethroplasty technique.

The operation in this case was started from marking suture of the penile gland. Then degloving of the penile skin as proximal as possible. After degloved, identification of the urethra was done, and dilation was found until a finger could be inserted for around 2 cm. Then, urethra was incised and tailored according to Folley catheter 6Fr guidance until the dilated urethra. After the Folley catheter was inserted, inversion suturing was done at the tailored urethra with PDS 6.0 and glanuloplasty with PDS 6.0. After evaluating the urethral lumen and found no narrowing, skin suture was done with PDS 6.0. Cystostomy was made to divert urine and maintained until one-week post-operation.

Patient follow up post reduction urethroplasty was done periodically and long term. This is needed to determine the erectile function and fertility of the patient. The patient revisited with good condition. The patient could urinate with no hindrance. Post-operation complication was not found. However, the erectile function could not be determined because the patient was considered pediatric.

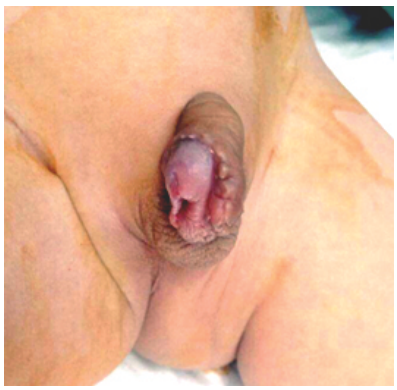


Figure 1. Pre-operation

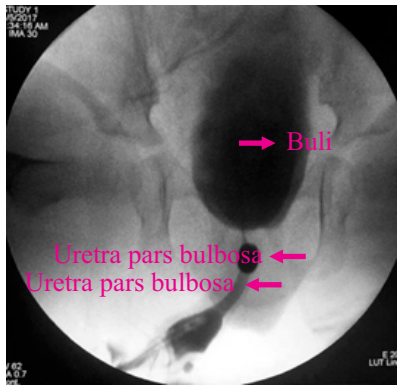
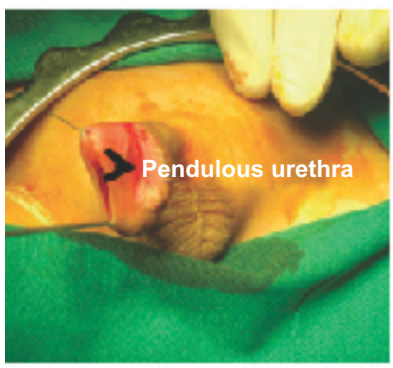
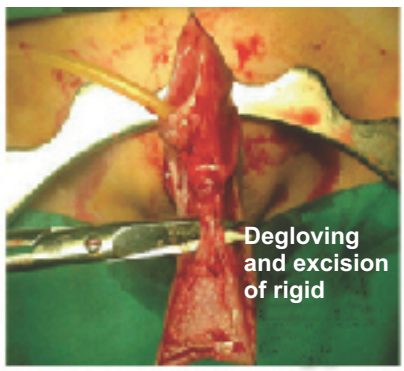


Figure 2. Urethrography



(a)



(b)

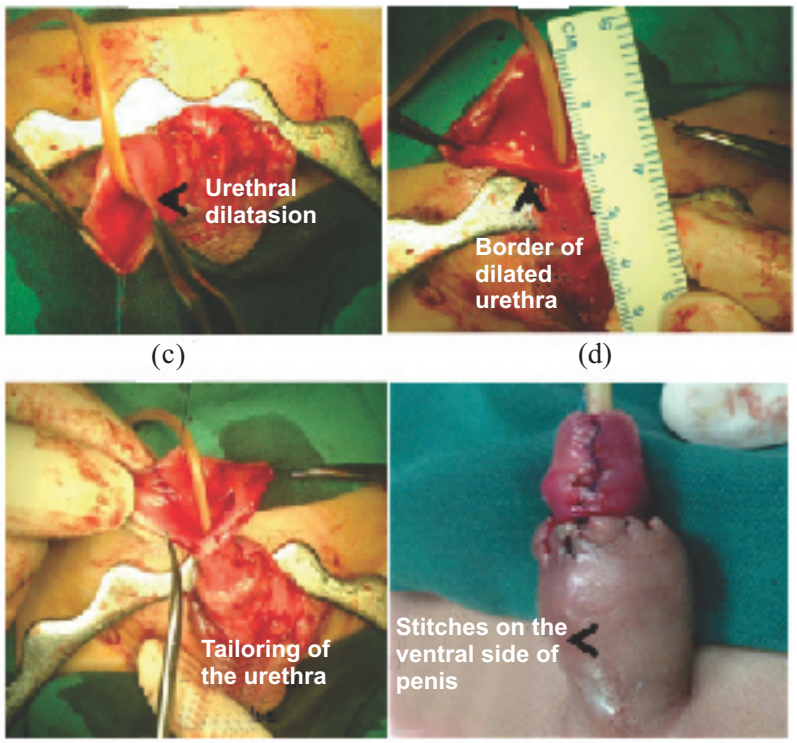
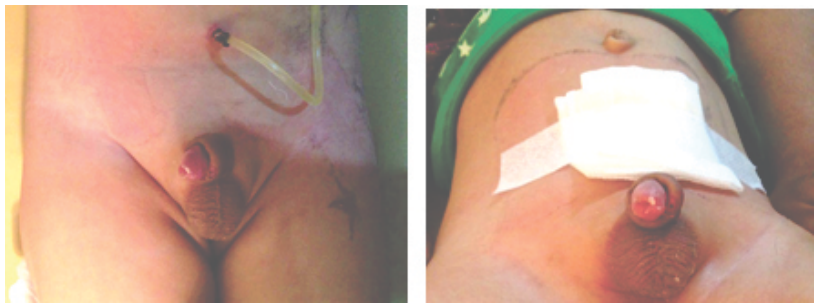


Figure 3. (a) Marking suture of the penile gland (b) Degloving of the penile skin (c) Identification of urethral dilation around 2 cm (d-e) Urethra was incised and tailored according to Folley catheter guide number 6 (f) Suture was done using PDS 6.0 at the tailored urethra and PDS 6.0 for granuloplasty.



Figures 4. Evaluation one-week post-operation.

DISCUSSION

Megalourethra is a congenital deformity of the urinary tract with the characteristic of the absence of penile erectile tissue that causes functional obstruction due to urinal stasis. This disorder has a rare prevalence, but must be diagnosed and managed accurately to prevent complications.[3] The cause of this disorder has not yet known but it was suggested by the failure of migration, differentiation or development of mesenchyme of the phallus. Another theory stated that there was a delay or failure of granular urethral canalization that relates to the failure of development of corpus spongiosum and corpus cavernous located around the urethra.[4]

Megalourethra was divided into two types: scaphoid and fusiform. For the scaphoid type, abnormality involves only the urethra and corpus spongiosum so that when urinating, the urethra dilates and forms a scaphoid. The second type is fusiform in which corpus spongiosum and corpus

cavernosum both were not formed in which the urethra dilates and looks fusiformed (coiled).[5] Megalourethra is usually related to other congenital disorder, 80% in scaphoid type, and 100% in fusiform type. Other genitourinary disorders that could happen are renal dysplasia, hydronephrosis, hydroureter, vesicourethral reflux, hypospadia, and prune-bell syndrome. Furthermore, other congenital disorders that could happen are VATER (vertebral, anal atresia, tracheoesophageal fistula, renal anomalies) and VACTERI (vertebral, anal atresia, tracheoesophageal fistula, renal, and limb deformities). Hence thorough examinations are needed.[4]

Initial presentation is frequently found during the first days or weeks of life, even though some reports antenatal diagnosis. The chief complaint often found in the patient with megalourethra is phallus enlargement that is seen during urination. The most important examination for this case is ultrasonography to determine the kidneys and vesical condition; to ensure diagnosis, intravenous urogram (IVP) or voiding cysto-urethrogram (VCUG) are used.[3,5] From these examinations, dilation of the anterior urethra is found with or without vesicourethral reflux.[3]

Management for this condition greatly depends on the clinical condition and the presence of other related abnormalities. Urethral definite management is highly suggested early in life to avoid recurrent urinary tract infection. Megalourethral management is different in scaphoid and fusiform. Management for megalourethra can be within one or two phases. One phase operation in scaphoid type megalourethra was reported the first time by Nesbitt in 1955 with reduction urethroplasty longi-tudinally.[5,6] Other techniques were also presented by Heaton et al. using plication technique of the urethra. Heaton reported that this technique has less complication and can be used in various megalourethra cases.[7] This case uses the same technique with the same base as Nesbitt's. The key of the revision success in urology is the presence of supporting corpus tissue. Surgical management for this case is reported to have high success rate in fixing external appearance and urination function.5 In one of the cases reported, there is a chance of erectile dysfunction in adult life, though there was no cohort data available.

CONCLUSION

Megalourethra is a rare case that needs appropriate diagnosing and management to avoid complication. Reduction urethroplasty can be used as the chosen management for scaphoidal megalourethra. Furthermore, in depth researches are required with better methodology to determine the outcome of patients with megalourethra.

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