Journal of Neonatal Surgery 2018; 7:34
DOI: 10.21699/jns.v7i3.765

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CASE REPORT

Congenital Megalourethra

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How to cite: Sarin YK, Jain N, Kumar P. Congenital megalourethra. J Neonatal Surg. 2018;7:34.

ABSTRACT

Congenital megalourethra is a rare mesenchymal anomaly of the male urethra. A sparse literature is available for its early surgical management. We present here an antenatally diagnosed case of congenital megalourethra, who underwent reduction urethroplasty at 18 days of life. To the best of our knowledge, this is the youngest case reported hitherto.

Key words: Congenital; Megalourethra; Reduction urethroplasty

INTRODUCTION

Megalourethra is a rare congenital anomaly of the male anterior urethra characterized by a non-obstructive dilatation of the penile urethra [1]. The entity is characterized by severe dilatation of the penile urethra due to poor development of the corpus spongiosum and/or corpus cavernosum.

Prenatal diagnosis of congenital megalourethra has been reported and may be associated with pulmonary hypoplasia and renal insufficiency leading to fetal demise. The associated congenital anomalies in such cases may contribute to comorbidities [2]. Postnatally, the untreated condition may be associated with morbidity of recurrent urinary tract infection, stone formation, obstructive uropathies, and renal insufficiency, so surgical intervention should be undertaken as soon as possible [3].

We report a neonate with congenital megalourethra who presented with urosepsis in whom a reduction urethroplasty was performed by a novel technique in the very first few days of life.

CASE REPORT

A 2-day-old newborn presented with an antenatal history of bilateral hydroureteronephrosis along with complaints of weak stream and ballooning of the penis during voiding and post-voiding dribbling since birth. On examination, there was a globular swelling on the ventral aspect of shaft of the penis that ballooned markedly during voiding (Figure 1a). On compression of the swelling, urine dribbled from the normally-placed meatus. Both testes were palpable in scrotum.

Antenatal ultrasound done during third trimester showed bilateral hydroureteronephrosis and oligohydramnios. Post-natal ultrasound was done on day 1 of life that showed bilateral moderate hydronephrosis (RK - 4.9 cm × 2.3 cm, APPD - 1.2 cm and LK - 4.2 cm × 2.2 cm, and APPD - 2 cm) with thinned out renal parenchyma, grossly dilated both ureters, and posterior urethra was dilated. Micturating cystourethrogram (MCU) found marked fusiform dilatation of the anterior urethra with right-sided grade V vesicoureteral reflux (VUR). Renal function tests (RFT) showed raised serum creatinine (1.3 mg/dL). The neonate was catheterized, and serial RFT was done. Serum creatinine normalized after 5 days to 0.7 mg/dL.

On day 18th of life, a penile incision along the ventral penile raphe was given and different layers of urethra were laid open. No formal degloving was done (Figure 1b). The redundant urethra was excised, and the urethra was reconstructed over 8 Fr infant feeding tube using 6–0 polyglactin suture (Figure 1c). The redundant flayed corpus spongiosum was approximated as second layer (Figure 1d). Protective

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Submitted: 04-04-2018 Accepted: 05-04-2018
Conflict of interest: None Source of Support: Nil

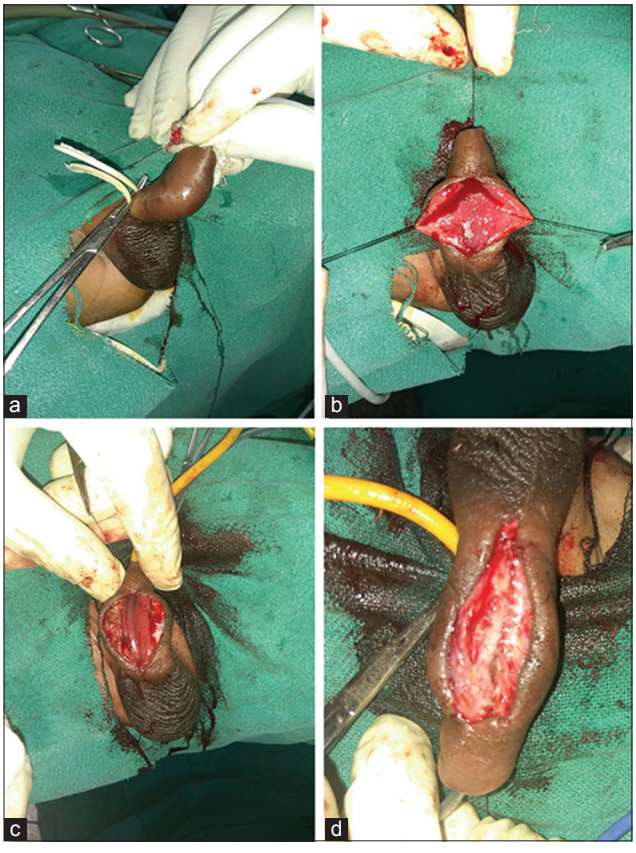

Figure 1: (a) Ballooned congenital megalourethra. (b) Ventral midline incision without degloving. (c) Redundant urethra excision. Cover of corpora spongiosum and dartos

diversion suprapubic cystostomy was done. The child was allowed to micturate normally after 10 days of surgery; he passed urine in good stream. He is doing well on follow-up after 3 months. A repeat MCU has shown downgrading of right VUR from Grade V to Grade II.

DISCUSSION

Benacerraf et al. [4] in 1989 were the first to report this condition prenatally. Various associated congenital anomalies include renal dysplasia–hypoplasia, bilateral hydronephrosis, bilateral hydroureter, vesicoureteric reflux, megacystis, Prune-Belly syndrome, urethral duplication hypospadias, posterior urethral valves, undescended testes, VATER (vertebral, anal atresia, tracheoesophageal fistula, and renal anomalies) and VACTERL (with limb deformities) [5,6].

The differential diagnosis of congenital megalourethra includes congenital anterior urethral diverticulum, anterior urethral valve (AUV), Cowper's syringocele, and congenital urethral stricture. The MCU helps in the differentiation of the same [7]. In AUV, proximal end of dilated urethra forms an obtuse angle with ventral floor and it is acute in case of diverticulum. Cowper's syringocele occurs due to dilatation of the main draining duct, which gets filled at time of micturition.

The management may include single or staged urethroplasty. Nesbitt first described a longitudinal reduction urethroplasty that was somewhat similar to that performed in our case [8]. A staged procedure may be needful in severe variety (fusiform usually), in which first marsupialization of megaurethra is done ventrally to prevent stasis, this is followed by closure of defect when the child grows older. Heaton and colleagues described a technique of urethral plication for some cases of scaphoid megalourethra [9]. The degloving of the penis was not done by us, as suggested in previous procedures. Vesicostomy, performed as the initial diversion in staged procedures in many such patients previously, is now known to cause urodynamic abnormalities and should ideally be avoided.

Most of the cases reported in literature underwent urethroplasty at an older age, except one reported by Obara et al. who performed reduction urethroplasty in a 1-month-old boy [10]. We present here case of youngest neonate. The redundant spongiosum and large dartos tissue availability helped to give two layered safe covers to the urethroplasty suture line.

In conclusion, we performed single stage reduction urethroplasty without penile degloving in neonatal period with good outcome.

Author's contribution

All authors contributed equally in concept, design, literature review, drafting the manuscript, and approval of the final manuscript.

Consent statement

Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

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