Annals of Pediatric Surgery

CASE REPORTS

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# Scaphoid megalourethra associated with congenital urethrocutaneous fistula: a case report of a rare association

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## Abstract

**Background** Congenital megalourethra is a rare male genital malformation, with less than 100 cases reported in literature. Urethrocutaneous fistula, in turn, has only 65 cases described, which what makes the association between these two anomalies an even rarer condition.

**Case presentation** The patient was male and born at 37 weeks of gestational age. At birth, the testicles were impalpable, and he had a penile malformation compatible with scaphoid megalourethra associated with congenital urethrocutaneous fistula, with urine output exclusively through a small orifice in the ventral region of the penis. When the child completed 7 months and 22 days, underwent the first surgery, laparoscopically. The first Fowler-Stephens was performed, and the gonadal vessels were ligated, with reconstruction of the penis. Nine months later, the second Fowler-Stephens procedure was performed, with bilateral orchidopexy and penoplasty to improve penis aesthetics. The urethra region was not approached. The postoperative period of the second surgery progressed uneventfully. Nowadays, the patient presents adequate urinary stream.

**Conclusion** The scaphoid megalourethra associated with congenital urethrocutaneous fistula is an extremely rare pathology, with very few cases reported in the literature to date. The obstetric USG is important to increasing the chances of early diagnosis and treatment.

**Keywords** Congenital megalourethra, Urethroplasty, Urethrocutaneous fistula

## Background

Congenital megalourethra is a male genital malformation of the urinary system, characterized by dilatation of the urethra [1–3]. It is often associated with other genitourinary anomalies and is classified as scaphoid or fusiform [3–5]. Megalourethra was first described in 1955 by Tom E. Nesbitt, who reported the case of a 1-month-old child with a large dilatation of the body of the penis that, upon manual compression, released urine through the urethral meatus [1] So far, less than 100 cases have been reported in literature [6, 7]. The urethrocutaneous fistula is also a very rare pathology, with only 65 cases described [8].

The diagnosis of congenital megalourethra can be made by clinical examination and retrograde urethrocystography, which shows the dilatation of the anterior portion of the urethra, also differentiating the scaphoid and fusiform types. Urinary tract ultrasonography (USG) is utilized to complement the diagnosis and detect possible

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concomitant renal malformations [4] The definitive treatment involves urethroplasty surgery [9]

Congenital urethrocutaneous fistula, when associated with megalourethra, is an even rarer condition. The diagnosis also can be made through clinical examination or retrograde urethrocystography, and the treatment is exclusively surgical too [10] This case report presents an association of these two urinary tract anomalies, highlighting its main characteristics and emphasizing the importance of early diagnosis and suitable treatment.

## Case presentation

The patient was male, Brazilian, born on March 08, 2019, by cesarean delivery, in Maringá, state of Paraná, at 37 weeks of gestational age, weighed 3.480 kg, with 52 cm of stature, and had an Apgar score of 8/10. At birth, the testicles were impalpable, and he had a penile malformation compatible with scaphoid megalourethra associated with congenital urethrocutaneous fistula, with urine output exclusively through a small orifice in the ventral region of the penis (Figs. 1 and 2). During prenatal care, a morphological obstetric USG image acquired at 33 weeks of gestational age suggested omphalocele, bilateral hydronephrosis, and altered nuchal fold, at which point he started follow-up with the pediatric surgery team. At a second obstetric USG examination, at 35 weeks, an undefined alteration was observed in the distal portion of the penis, besides oligohydramnios, bilateral hydronephrosis (worse on the right side), and dilatation of cerebral ventricles. The omphalocele was not identified in this last examination. He was submitted to noninvasive prenatal testing (NIPT), and the result showed no alterations in chromosomes 13, 18, 21, and XY. He also presented right pneumothorax identified by respiratory difficulty after birth, requiring drainage of the hemithorax, intubation, and admission to the intensive care unit, where he remained under invasive mechanical ventilation for 56 days. He was discharged 70 days after birth.

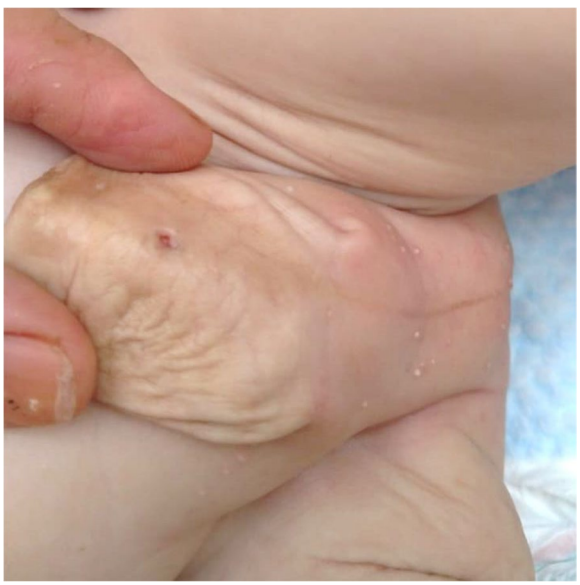

**Fig. 1** Penile malformation compatible with megalourethra and urethrocutaneous fistula with an orifice in the ventral region through which urine drained

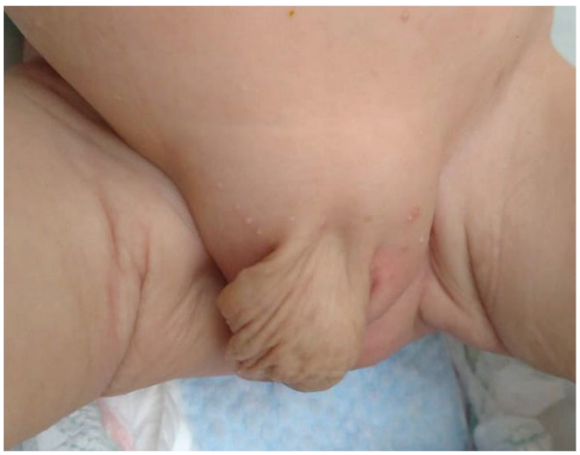

**Fig. 2** Scaphoid megalourethra

At 3 months of age, he underwent USG of the kidneys and urinary tract. The results showed that the right kidney's cortex thickness and echogenicity were preserved. There were signs of mild renal pelvis dilatation, measuring about 6×8 mm. Similarly, the left kidney also showed preserved cortex thickness and echogenicity, and there were signs of mild dilatation of the renal pelvis measuring about 6×5 mm. The renal scintigraphy with DMSA, performed on June 21, 2019, detected a mean tubular quantification of 50% in both kidneys and an absolute tubular quantification of 40% in both kidneys, with no renal scarring. In addition, karyotyping showed 46 XY, with translocation of chromosomes 07 and 18.

At 7 months and 22 days, the child underwent the first surgery, laparoscopically. The first Fowler-Stephens was performed, and the gonadal vessels were ligated, with reconstruction of the penis (Figs. 3, 4, and 5). The patient remained with a bladder catheter for 10 days, presenting a strong jet from the end of the penis after its removal.

After 8 months, the second Fowler-Stephens procedure was performed, with bilateral orchidopexy and penoplasty to improve penis aesthetics. The urethra region was not approached (Fig. 6). The postoperative period of the second surgery progressed uneventfully (Fig. 7). A new USG of the kidneys and urinary tract, realized after 13 days of the second surgery, showed minimal ectasia of the left renal pelvis, and other portions appeared normal.

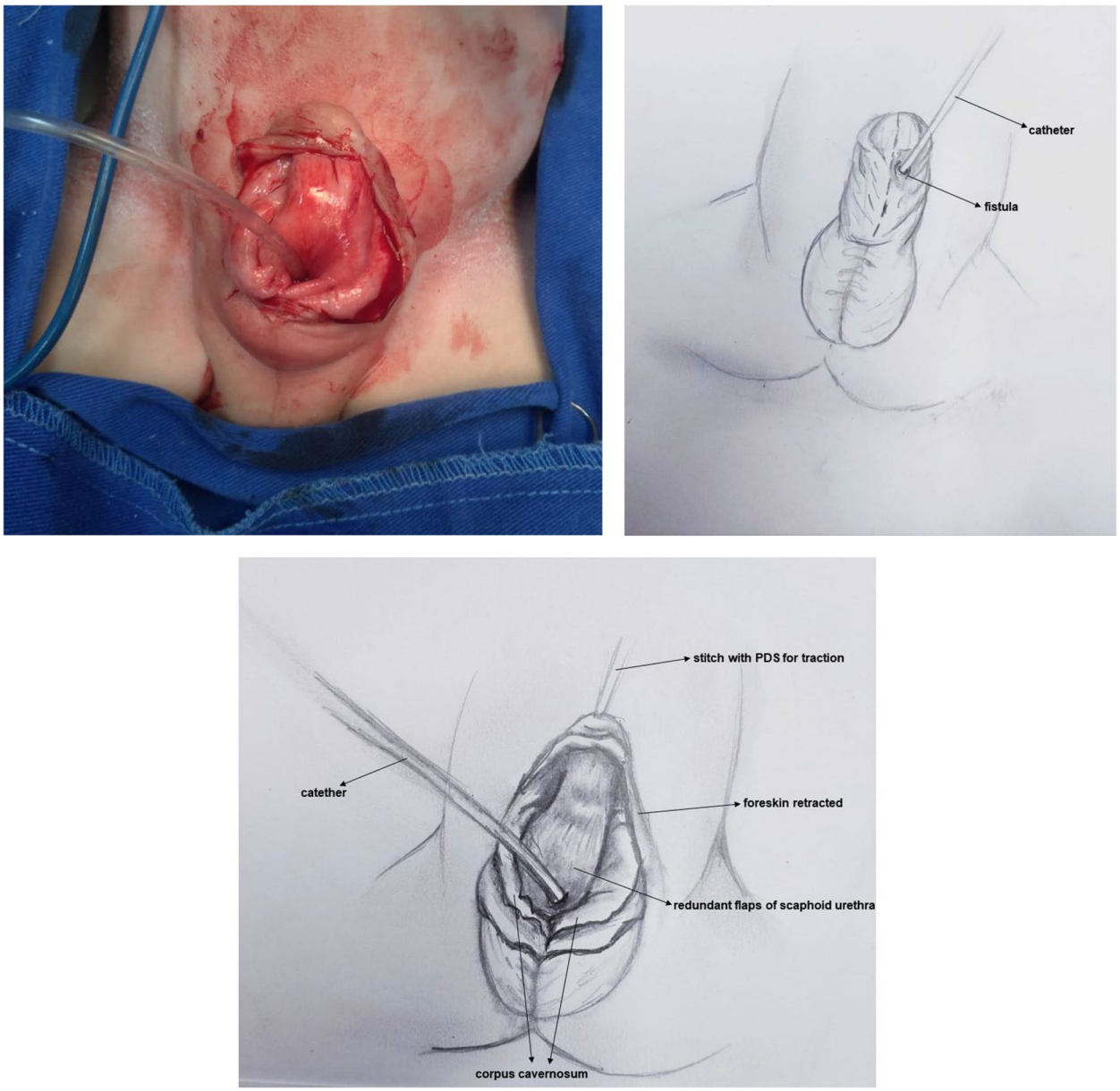


**Fig. 3** Urethral catheter of 6 French (Fr) was inserted into the urethra and returned urine. Degloving the penis and a scaphoid dilatation of urethra were noted. Diagram of the operating technique. PDS: polydioxanone

Currently, the patient presents adequate urinary stream, and the results of previous post-op follow-up visits were satisfactory.

## Discussion

The etiology of megalourethra is still not clearly understood [4, 5]. It is believed that a dysgenesis of the mesoderm occurs during the 7th week of embryonic development, which leads to the absence of the corpus spongiosum in the anterior region of the penis and, in some cases, partial or total absence of the corpora cavernosa. The penile glands and the navicular fossa are not affected [4, 11]. Due to the lack of these structures, a balloon-shaped dilated anterior urethra is formed, leaving the body of the penis dilated and deviated in its ventral portion [11].

Two types of congenital megalourethra have been described in the medical literature: scaphoid, which is the most common and consists of deficiency or exclusive absence of the corpus spongiosum, and fusiform, which

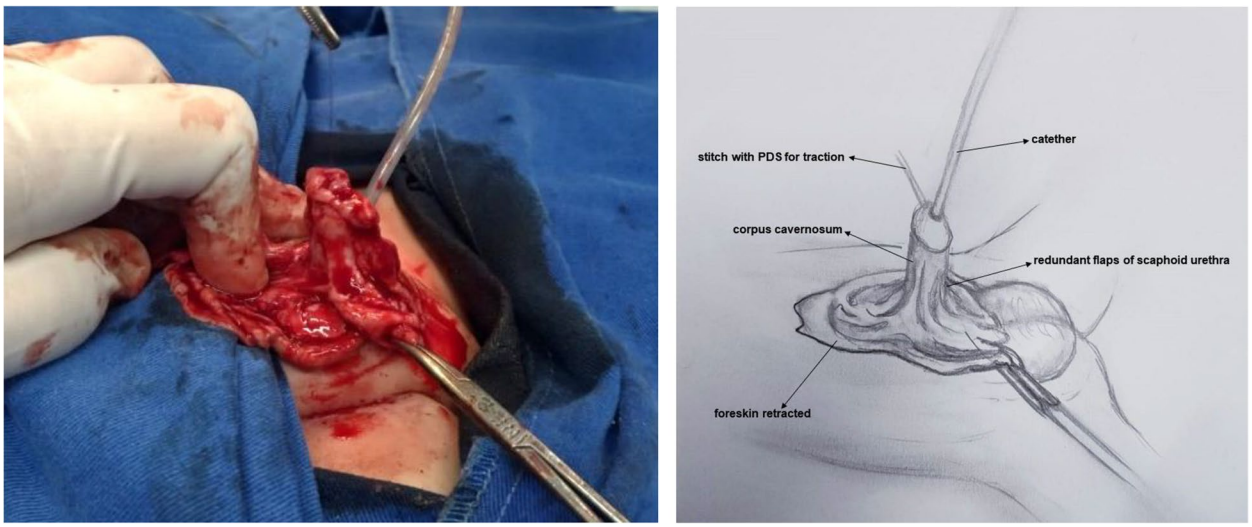


**Fig. 4** Posterior view after degloving showing the corpus cavernosum, foreskin retracted, and the redundant flaps of scaphoid urethra. Diagram of the operating technique

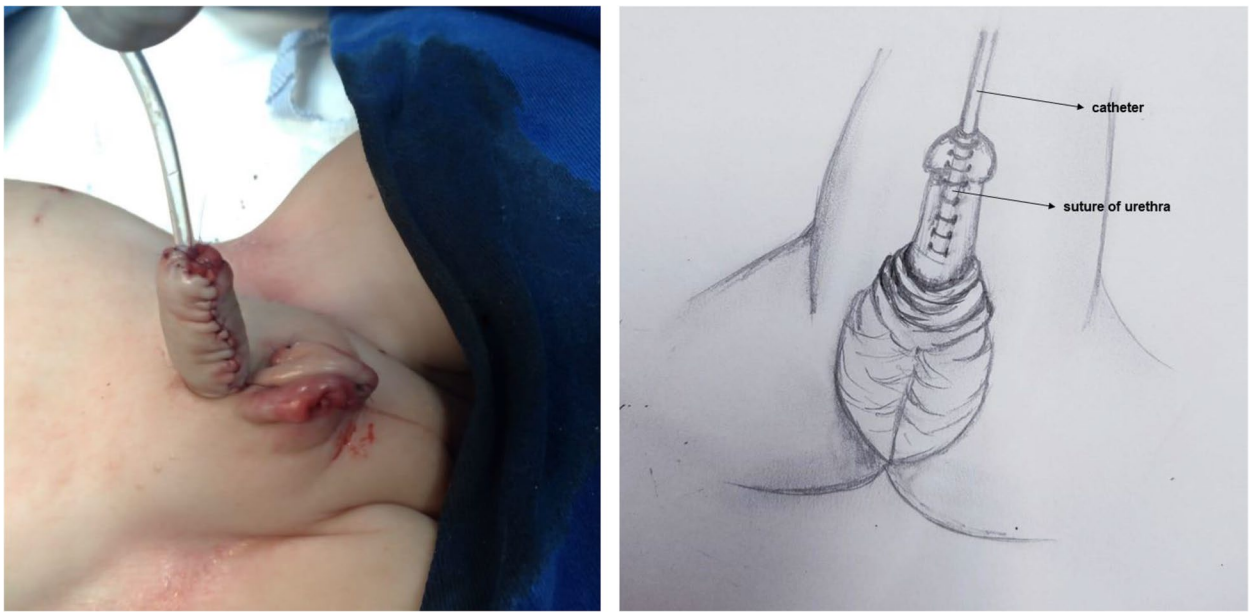


**Fig. 5** Final aspect of the first penile reconstruction surgery. Reduction urethroplasty was done with 6.0 PDS in three stages over 10-Fr urethral catheter. We did an initial scrotum reconstruction (both testis were impalpable). Diagram of the operating technique

is less prevalent and associated with a combined malformation of the corpora cavernosa and corpus spongiosum [3, 12]. The severity of the disease varies according to the association with other urinary tract malformations or deformities, and the fusiform megalourethra is considered the most severe form, as it is more frequently associated with other congenital malformations [5, 13].

Cases of congenital megalourethra linked to renal dysplasia and hypoplasia, posterior urethral valve, ureteral duplicity, vesicoureteral reflux, hydronephrosis, hypospadias, Prune-Belly syndrome, VACTERL syndrome, cryptorchidism, and anorectal malformations have been described [6, 14, 15]. The associated malformations are present in 80% of scaphoid cases and 100% of fusiform cases [6].

The diagnosis of congenital megalourethra can be made either prenatally or postnatally [6]. Benacerraf first identified the malformation in the prenatal period,

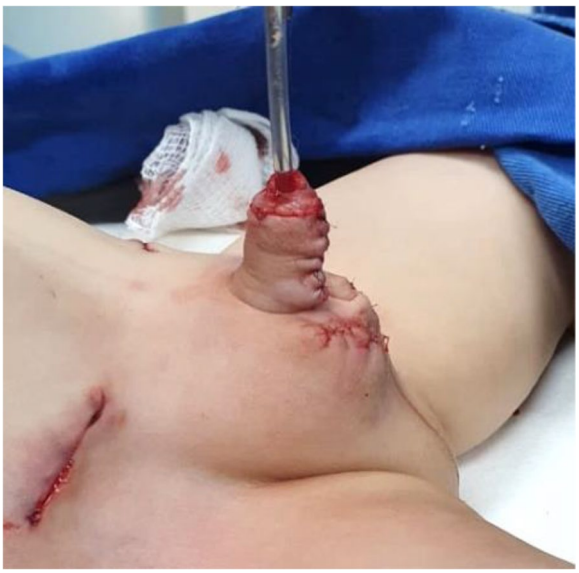

**Fig. 6** Final aspect of the second Fowler-Stephens procedure, with bilateral orchidopexy and penoplasty

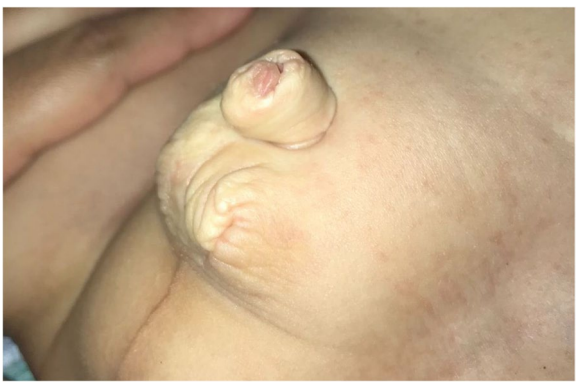

**Fig. 7** Current postoperative appearance

through morphological obstetric USG [6]. The earliest prenatal diagnosis ever recorded was made at 13 weeks of gestational age in a fetus with multiple malformations, but most cases are detected only in the second trimester [16]. Additionally, fusiform megalourethra is more easily diagnosed in the prenatal period, by obstetric USG, than scaphoid [6].

In the postnatal period, the diagnosis can be made by clinical examination and retrograde urethrocystography, which helps differentiate the scaphoid and fusiform types. Urinary tract USG complements the diagnosis and aids detection of potential concomitant renal malformations [4, 6, 9].

The patient presented in this case report had already shown signs suggestive of megalourethra at obstetric USG before birth, demonstrating the relevance of this test for early diagnosis. Additionally, postnatal USG made it possible to identify renal alterations such as hydronephrosis. The patient does not realized retrograde urethrocystography, which would help to see a possible anterior urethral valve associated in some cases.

The definitive treatment to correct congenital megalourethra is surgical, namely, urethroplasty in cases of scaphoid megalourethra [9, 17]. In cases of fusiform megalourethra, besides urethroplasty, the placement of a penile prosthesis in adulthood can be considered [4]. The technique for surgical correction of scaphoid megalourethra was published by Nesbit and Baum more than 50 years ago, and this procedure continues to be used today, with modifications [6, 9].

The etiology of congenital urethrocutaneous fistula is not completely elucidated too. Champbell states that the fistula represents an embryonic urethral defect caused by a distal congenital obstruction. The diagnosis can be clinical, based on urine output through the orifice of the ventral region of the penis, or retrograde urethrocystography [10]. Treatment of both megalourethra and urethrocutaneous fistula is exclusively surgical, by urethroplasty, which follow the same basis as the hypospadias technique [1, 7, 10, 18].

In the case presented here, the patient did not undergo retrograde urethrocystography. However, it was possible to confirm the diagnosis of scaphoid megalourethra intraoperatively, since the corpus spongiosum was absent while the corpora cavernosa were present. Regarding the diagnosis of urethrocutaneous fistula, it was confirmed by physical examination. It is worth mentioning that the penile reconstruction was technically difficult, due to the large dilatation of the urethra and the reduced size of the glans, which can be observed in the figures presented above.

## Conclusion

The scaphoid megalourethra associated with congenital urethrocutaneous fistula is an extremely rare pathology, with very few cases reported in the literature to date. Surgical treatment is necessary in all cases.

Even though it is a rare and complex disease requiring difficult surgical treatment, the postoperative outcome was satisfactory. Currently, the patient has adequate urinary stream and has not presented urinary tract complications in follow-up visits.

Based on this case report, it is expected that physicians recognize the main characteristics related to megalourethra and urethrocutaneous fistula, with special attention to findings suggestive of the malformation on an obstetric USG, increasing the chances of early diagnosis and treatment. This may allow better prognosis for patients with this pathology.

**Abbreviations**
USG Ultrasonography
NIPT Noninvasive prenatal testing

**Acknowledgements**
Not applicable.

**Authors' contributions**
All authors read and approved the final manuscript.

**Funding**
No funding or grant support.

**Availability of data and materials**
All data generated or analyzed during this study are included in this published article.

## Declarations

**Ethics approval and consent to participate**
The present study was approved by the ethics committee of the State University of Maringá.

**Consent of publication**
Informed written consent was obtained from the parents of the patient for publication of this report and the accompanying images.

**Competing interests**
The authors declare that they have no competing interests.

Received: 11 September 2022 Accepted: 3 October 2023
Published online: 19 October 2023

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## Publisher's Note