Urology Case Reports 58 (2025) 102924

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Pediatrics

A case report of hypospadias combined with polyorchidism

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ARTICLE INFO

Keywords:
Hypospadias
Polyorchids
Duckett method
Orchiopexy
Cryptorchids
Testicular atrophy

ABSTRACT

Hypospadias is a common congenital malformation of the reproductive system; however, to date, no cases of hypospadias complicated by polyorchidism have been reported in the literature. This article reports a case of a 1-year-old boy presenting with hypospadias, who was also diagnosed with polyorchidism combined with cryptorchidism. Preoperative Doppler ultrasound revealed two testes on the right side, and an atrophic testis on the left. After surgical treatment, the patient recovered well and was discharged. Hypospadias combined with polyorchidism is highly prone to misdiagnosis or missed diagnoses. This case provides insight into the recognition, diagnosis, and treatment of this condition.

1. Introduction

Hypospadias is the second most common congenital malformation of the reproductive system after cryptorchidism.[1] Currently, standardized diagnostic and treatment protocols for hypospadias exist, with the most common associated conditions being inguinal hernia, hydrocele, and incomplete testicular descent, each occurring in approximately 9% of cases.[2] However, there have been no reported cases of hypospadias combined with polyorchidism, and standardized diagnostic and treatment protocols for this condition have not yet been established. Moreover, the treatment of polyorchidism remains controversial and is often determined by various factors, such as testicular location, reproductive potential, testicular size, and patient age.[3] In this case, the occurrence of left-sided testicular atrophy is extremely rare. We will now discuss this case in conjunction with clinical data and relevant literature.

2. Case description

The patient is a male child aged 1 year and 5 months. He was admitted to the hospital on April 10, 2024, due to an abnormal urethral opening observed since infancy. Physical examination revealed an underdeveloped penis with the urethral opening located at the base of the ventral side near the scrotum, a markedly chordee and a split scrotal raphe (Fig. 1A). No palpable testis was found in the left scrotum or inguinal region, and two peanut-sized testicular-like masses that could slide into the scrotum were detected in the right inguinal area. The patient was not treated in other hospital. The patient had no other family history. Auxiliary examinations: Anti-Müllerian hormone (AMH) level was 22.96 ng/ml, estradiol 5.00 pg/ml, luteinizing hormone (LH) 0.275 mIU/ml, progesterone 0.167 ng/ml, and testosterone <0.03 ng/ml. Chromosome analysis indicated a 46XY karyotype. Ultrasound examination showed two testicular-like echo areas in the right spermatic cord measuring 7.4 × 4.3 mm and 11.6 × 5.0 mm, and a solid hypoechoic nodule of approximately 5.6 × 4.3 mm in the left inguinal canal (Fig. 1B and C). Color Doppler Flow Imaging (CDFI) revealed blood flow signals within the right testis. The child also presented with a mild left-sided spermatic sheath effusion, and based on the test results, we suspected that the child had Müllerian duct persistence syndrome. The preoperative pelvic floor ultrasound did not reveal any Müllerian duct remnants, and the patient's family declined the suggestion for an MRI for further evaluation. Therefore, we performed laparoscopic exploratory surgery to investigate Müllerian duct remnants and simultaneously manage the

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https://doi.org/10.1016/j.eucr.2024.102924
Received 21 October 2024; Received in revised form 18 December 2024; Accepted 23 December 2024
Available online 24 December 2024

spermatic sheath effusion. On April 29, 2024, hypospadias repair (Duckett) + pedicled axial flap grafting + change to repair of chordee + bilateral orchiopexy + laparoscopic ligation of the bilateral tunica vaginalis was performed under general anesthesia. Intraoperative laparoscopy showed no significant remnants of the Müllerian duct in the abdominal cavity, and bilateral tunica vaginalis were found to be patent. At laparoscopy, the left tunica vaginalis revealed remnants of the vas deferens, and the spermatic vessels were markedly atrophied; the right vas deferens and spermatic vessels appeared normal. Squeezed the inguinal region bilaterally to avoid inadvertent ligation to testicular tissue. After ligation of the bilateral tunica vaginalis, the right scrotum was incised and exploration of the inguinal region failed to reveal any other abnormal tissue, revealing two soy-sized testes in the right tunica vaginalis cavity connected in tandem by the tail of the epididymis (Fig. 2A). The left scrotal tunica vaginalis was incised, and hydrocele fluid was drained. The same examination of the inguinal region did not reveal any other abnormal tissue. A rice-sized atrophic testicular-epididymal-like tissue observed in the left scrotum was excised (Fig. 2B). The right spermatic cord was freed, and one testis was placed into the left scrotal cavity and secured to the dartos, while the other was fixed to the right scrotal dartos. Subsequently, a midline incision was made along the urethral plate for release, and a dorsal plication suture was placed to correct the chordee. A pedicled transverse flap from the dorsal penile skin was harvested and sutured to the defect in the midline of the proximal urethra, and the flap was inverted and sutured to form the urethra (Fig. 2C–F). Finally, the corpora cavernosa were wrapped around the new urethra and sutured in place, each layer closed sequentially, and the penis was compressed and bandaged. Postoperative pathological examination of the atrophic testicular-like tissue on the left side suggested fibrous connective tissue with benign glandular structures. The discharge diagnosis: (I) Hypospadias, (II) Polyorchidism, (III) Left testicular atrophy, (IV) Bilateral patent sheath, (V) cryptorchidism. Before discharge, the urethral surgical area healed well, and the patient was able to urinate normally after the catheter was removed (Fig. 3). Four months after surgery, follow-up showed good recovery of the surgical area (Fig. 4A). Ultrasound re-examination indicated that both testes were in ideal positions, with visible blood flow signals within both testes (Fig. 4B and C). All study methods were performed in accordance with relevant guidelines and regulations. This study was approved by the XXX Ethics Committee. Informed consent was obtained by the parents for all procedures and for the publication of this case.

3. Discussion

Hypospadias is the second most common congenital disorder of the reproductive system after cryptorchidism, with an incidence of 3–9 per 1,000, and its prevalence has been increasing in recent years in China.[4,5] Hypospadias is caused by endocrine defects, disorders during embryonic sexual differentiation, and other factors that result in incomplete fusion of the urethral groove, leading to varying types of hypospadias.[6,7] However, the exact mechanisms remain unclear. Current research has shown that androgen receptor abnormalities, genetic mutations, endocrine disorders, maternal placental factors, and environmental influences all contribute to the multifactorial aetiology of hypospadias.[8,9] The main clinical manifestations of hypospadias are an ectopic urethral meatus, choree and hooded prepuce. The position of the ectopic urethral opening is the primary basis for classification and an important indicator of severity.[10] In clinical practice, to clarify the relationship between the urethral opening position and the severity of hypospadias, Duckett et al.[10] classified the urethral opening after penile straightening into distal, midshaft, and proximal types. Orkiszewski et al.[11,12] proposed a more detailed classification based on the position of the external urethral opening: (I) distal types, including glandular, coronal, and subcoronal types; (II) midshaft type; and (III) proximal types, including penoscrotal, scrotal, and perineal types. In this case, after repair of chordee during surgery, the urethral meatus was located at the base of the penis and was classified as a penoscrotal type of proximal hypospadias. Cryptorchidism and inguinal hernia occur in 9 % of hypospadias cases, with a higher incidence of cryptorchidism in posterior hypospadias (32 %), compared to 6 % in middle hypospadias and 5 % in anterior hypospadias. Inguinal hernia occurs in 17 % of posterior hypospadias, 8 % of middle hypospadias, and 7 % of anterior hypospadias. In this case, the patient was found to have cryptorchidism during the physical examination upon admission, a common complication of posterior hypospadias. During surgery, the patient's chordee was repaired by incising the dorsal deep fascia and suturing the tunica albuginea, followed by tubularized urethroplasty using a transverse

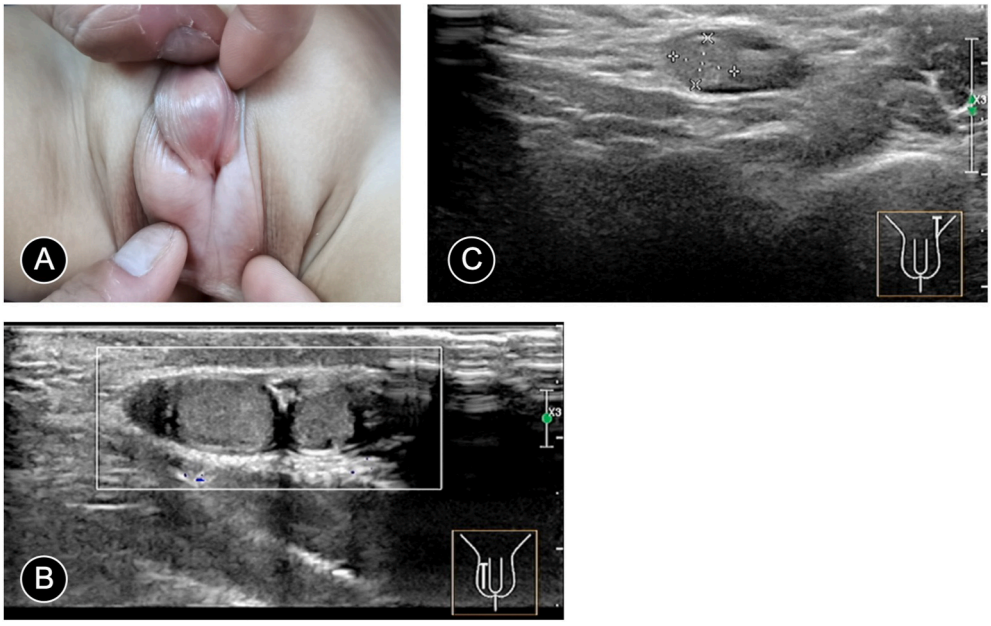

Fig. 1. Physical examination and preoperative scrotal ultrasound of a child with hypospadias and polyorchidism: A. Preoperative condition of hypospadias in the patient. B. Two testes in the right scrotum. C. One testis in the left scrotum.

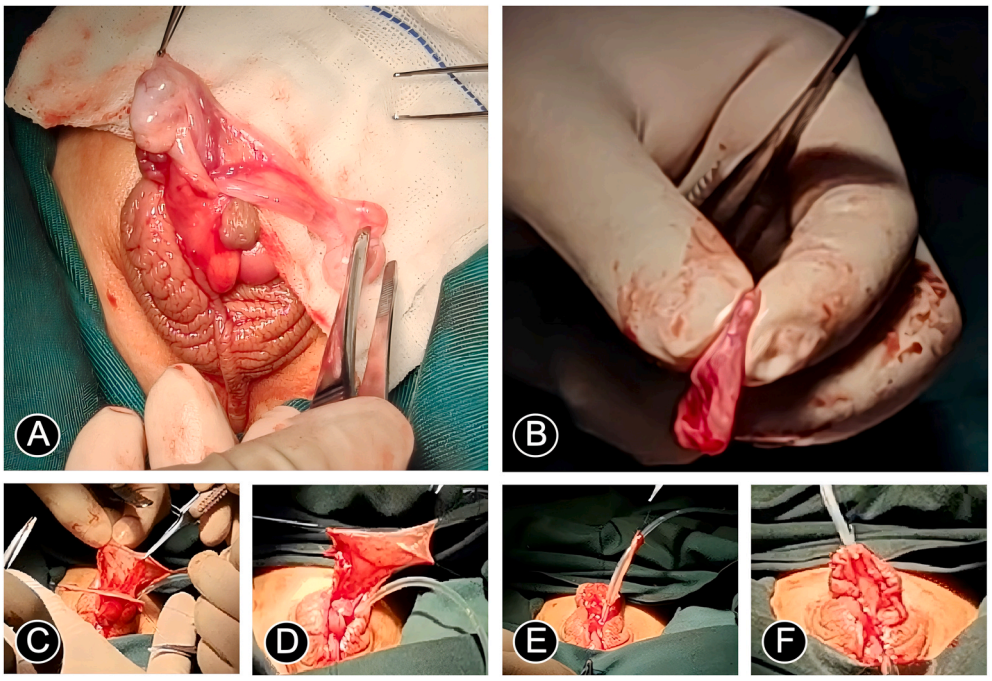

Fig. 2. Intraoperative findings in a child with hypospadias and polyorchidism. A. Two tandem testes on the right side during surgery. B. Atrophic testis and epididymal tissue on the left side during surgery. C-F. Harvesting of the dorsal penile flap and inverting sutures to form a new urethra during the Duckett procedure.

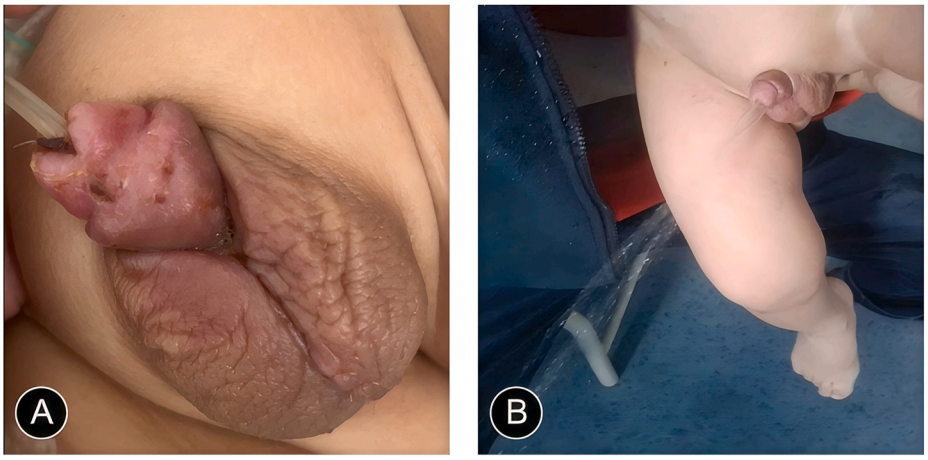

Fig. 3. Postoperative recovery in a child with hypospadias and polyorchidism. A. Postoperative surgical site of the penis and scrotum. B. Voiding status after catheter removal before discharge.

preputial island flap (Duckett procedure).[13,14]

Hypospadias is easy to diagnose, but polyorchidism, being rare and lacking specific symptoms and clinical manifestations, is often underdiagnosed or misdiagnosed by clinicians. While hypospadias and its repair are well-documented in the literature, this case is unique due to the presence of polyorchidism. The exact cause of polyorchidism remains unknown, but the most widely accepted hypothesis is that the primordial testis begins to develop from the primordial germinal ridge to the mesonephric ducts around the sixth week of embryonic life.[15] Around 8 weeks, the primordial testis undergoes morphological changes, and the epididymis and vas deferens develop from the Wolffian ducts.[16] Longitudinal duplication of the germinal crests causes the testes to separate, while transverse splitting results in various combinations of testes, epididymis, and vas deferens, leading to polyorchidism.[16] Different criteria are used for classification based on anatomical positions and reproductive functions. The most widely accepted system is Bergholz's classification of reproductive potential, based on Leung's classification. This system divides the types into type A, which has reproductive function, and type B, which lacks reproductive function. Type A includes subtypes A3 (without separate epididymis and vas deferens) and A4 (with separate vas deferens but without separate epididymis). Type B is further subdivided into B1 (with separate epididymis) and B2 (without epididymis).[16,17] This case has an independent epididymis and a common vas deferens, and is potentially reproductive and is type A2. In most cases, ultrasound and MRI are the primary diagnostic tools, in addition to physical examinations. If ultrasound results are inconclusive, MRI can be used to further confirm the diagnosis. For scrotal abnormalities, MRI is the most reliable and sensitive imaging technique. In this reported case, the patient presented with hypospadias, and polyorchidism was not detected during the physical examination but was found through color Doppler ultrasound, which revealed duplicated testes on the right side. Additionally, preoperative tests such as AMH, LH, and chromosomal analysis can be used to further clarify the diagnosis.[18] Since the patient presented with

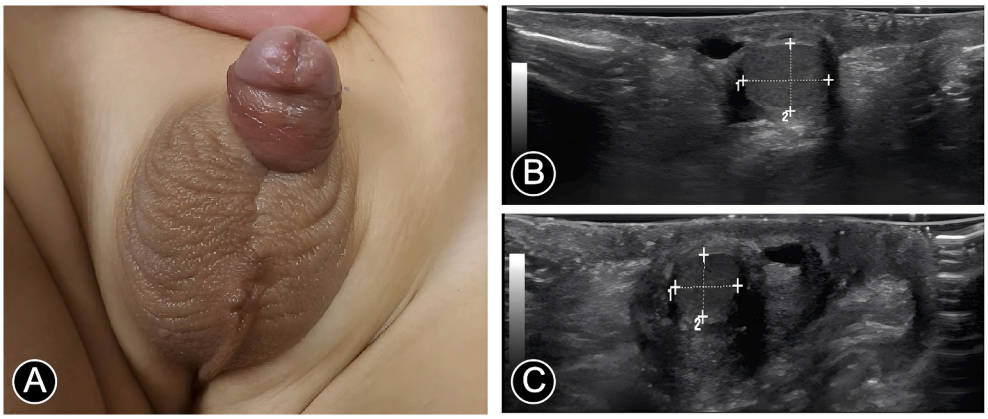

Fig. 4. Four-month postoperative follow-up in a child with hypospadias and polyorchidism. A. Surgical site of the penis and scrotum at 4 months postoperatively. B. Ultrasound of the left scrotum at 4-month follow-up. C. Ultrasound of the right scrotum at 4-month follow-up.

hypospadias, other complications without clinical features, such as polyorchidism, are easily overlooked. Polyorchidism has no obvious clinical symptoms, and no cases of hypospadias combined with polyorchidism have been reported in the literature. Although the patient in this case was diagnosed with polyorchidism through ultrasound and preoperative serological tests, clinicians should maintain divergent thinking to screen for other conditions to avoid missed or incorrect diagnoses.

There is still controversy regarding the treatment of polyorchidism. The choice of the optimal treatment plan must not only preserve reproductive potential but also take into account the high risk of testicular torsion and malignancy. Additionally, treatment choices must also consider factors such as compliance with follow-up, parental preferences, and cosmetic outcomes.[19] According to the literature, approximately 50%–65 % of supernumerary testes have reproductive potential, which is why conservative treatment is often adopted for intra-scrotal supernumerary testes in China.[20] In this case, two testes were found in the right scrotal tunica vaginalis, while a rice-sized atrophied testicle and epididymal-like tissue were found on the left side. Therefore, the left side was excised, and the two right-sided testes were freed and fixed separately on the left and right tunica dartos.

The patient has since been successfully discharged. Hypospadias combined with polyorchidism is extremely rare, and this case provides valuable reference information for clinicians regarding diagnosis and treatment. For cases of polyorchidism, which is easily missed or misdiagnosed, ultrasound should be conducted if abnormalities are found during physical examination or routine admission checks to rule out polyorchidism. However, whether there is a connection between hypospadias and polyorchidism remains unclear, as no studies have indicated a relationship. Unfortunately, the child's family was unable to undergo genetic testing due to financial constraints. Additionally, the patients left testicle and epididymal-like tissue were atrophied, and two right-sided testes were preserved and fixed with orchidopexy. This is rare in the literature on polyorchidism, as most case reports describe patients with normally presenting and functioning testes bilaterally. We informed the child's family in advance about the potential findings on preoperative imaging and the possible intraoperative encounters. Only after obtaining their consent did we proceed with the removal of the left atrophied testis and epididymal tissue. However, long-term follow-up is essential to ensure that affected children maintain reproductive function into adulthood and to enhance awareness, diagnosis, and treatment of the condition.

4. Conclusion

Hypospadias combined with polyorchidism is highly prone to misdiagnosis or missed diagnoses, and this case provides insight into the recognition, diagnosis, and treatment of this condition.

CRediT authorship contribution statement

Wentao Yu: Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Software, Resources, Project administration, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Wei Gong:** Resources, Investigation, Data curation. **Fuhan Zhao:** Resources, Investigation, Formal analysis, Data curation, Conceptualization. **Guan Zhang:** Validation, Supervision, Software, Project administration, Methodology, Investigation, Data curation, Conceptualization. **Zhenyu Liu:** Writing – review & editing, Visualization, Validation, Software, Resources, Methodology, Funding acquisition, Formal analysis, Data curation, Conceptualization. **Tiejun Pan:** Writing – review & editing, Validation, Supervision, Software, Resources, Methodology, Investigation, Data curation, Conceptualization.

Data availability statement

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

Ethics committee approval

This study was approved by the Central Theater General Hospital of Chinese People's Liberation Army Ethics Committee (date: 10.23.2024, number: 2024119–01). Informed consent was obtained by the parents for all procedures and for the publication of this case. The identities of the patients have been anonymized to protect their privacy. All study methods were performed in accordance with relevant guidelines and regulations.

Consent for publication

Written consent for publication was obtained from the parents. There are no identifying images or other personal or clinical details of the patients that compromise anonymity in this manuscript.

Funding statement

This work was supported by the Natural Science Foundation of Hubei Province (No. 2023AFB498 to Zhenyu Liu), Natural Science Foundation Exploration Plan (Morning Light Plan) of Wuhan City (No.2024040801020363 to Zhenyu Liu) and Postdoctoral Scientific Research Foundation, General Hospital of Central Theater Command (No. 20230102KY39 to Zhenyu Liu).

Declaration of competing interest

The authors have no potential conflicts of interest to disclose.

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