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Urethral duplication in male children: A study of 12 cases☆,☆☆

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ABSTRACT

Purpose: To present our experience in urethral duplication focusing on detailed surgical management.
Methods: We retrospectively reviewed the records of 12 male patients treated for urethral duplication between 2005 and 2017. Evaluations included ultrasound, retrograde urethrography, cystoscopy, and voiding cystourethrography.
Results: The age at presentation ranged from birth to 11 years. All 12 cases were classified using the Effmann classification. Case 1–4 patients with type I underwent excision of the dorsal accessory urethra by stripping technique. In case 5 patient(type IA) with two adjacent apical urethras, the septum was opened to form a single channel. Case 6 patient with type IB underwent visual internal urethrotomy near bulbous urethra to combine urethra into one channel. Five patients classified as type II (one with a type IIA1, and four with type IIA2 urethras). Urethral duplication was incidentally found during epispadias repair in case 7 patient with type IIA1, which was corrected by ventral plication, and excision of the dorsal epispadial urethra with stripping technique just below pubic bone. Case 8 patient with type IIA2 also required dorsal urethral excision with stripping technique. The two Y-type patients (case 10, 11) underwent urethrourethrostomy with a single-stage buccal mucosa tube graft, followed by repetitive surgeries owing to urethral stricture. One type III patient presented with penile inflammation and suprapubic pain, and underwent excision of both the dorsal urethra and nonfunctional anterior bladder.
Conclusions: Urethral duplication requires individualized surgical approaches based on the anatomical and functional characteristics. Because prognosis is variable depending on type and accompanied anomalies, these should be taken into account when planning a comprehensive workup and surgical management.
Level of evidence: Level IV.

Urethral duplication is a very rare congenital anomaly, but it is occasionally encountered by urologists. Although urethral duplication has been reported primarily in males, it can also occur in females, usually combined with bladder duplication [1,2]. As the anatomical patterns observed can be quite diverse, a patient's symptoms and appropriate treatments also vary. Patients can be either asymptomatic or symptomatic; common clinical findings are incontinence, urinary obstruction, recurrent urinary infection, and a double urinary stream. The diagnosis and classification of a double urethra should be based on detailed evaluations, including cystoscopy and urethrography.

Several classifications of urethral duplication have been described to distinguish between the different types and to define an appropriate plan of management [3,4]; Effmann's classification scheme is regarded as the current standard [5]. Despite the different classifications of urethral duplication, a standardized treatment for a particular classification cannot be applied because of the variety of different anatomical configurations that exist, necessitating customized surgical correction [1]. Surgical treatments for each classification type have been presented in several previous reports [1,3,6], but detailed descriptions of these surgical approaches are insufficient.

Herein, we report our experiences with 12 diverse cases of urethral duplication. We detail the patients' conditions and our surgical technique to contribute to the management of urethral duplication.

1. Materials and methods

We managed 12 cases of urethral duplication from 2005 to 2017, and retrospectively analyzed the electronic medical records of these cases after approval of the institutional review board (4-2018-1169). All 12 cases were classified using Effmann's classification (Supplemental Fig. 1). Clinical characteristics, such as the presentation type of the urethral duplication, clinical symptoms, associated anomalies, surgical treatment, and complications after surgical correction, were identified and summarized. Retrograde urethrography was performed in all 12 cases, and ultrasonography, intravenous pyelography, and magnetic

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resonance imaging (MRI) were performed as needed. All patients were treated by one of two pediatric urologists (S.W.H. or S.W.K.).

2. Results

Surgical treatment was carried out for 11 of 12 patients. The median age at the time of surgery was 33 months (interquartile range: 12.75–-99.25 months). The median follow-up period was 24.5 months (IQR: 15.75–29.75 months). Table 1 summarizes the details of the 12 cases.

2.1. Type I urethral duplication

Six cases were classified as a type I urethral duplication; of these, five cases (cases 1–5) were type IA, and case 6 was type IB. The specific combined abnormalities were hypospadias in one case (case 2), chordee in two cases (cases 1 and 3), and a concealed penis in one case (case 5). Cases 1–4 (Supplemental Fig. 2) all underwent excision of the accessory urethra. Case 5 (Supplemental Fig. 3A) is a patient in whom the urethral duplication was incidentally found during penoplasty for the repair of a concealed penis. In this case, a thin septum was present between the ventral and dorsal accessory urethras. The dorsal accessory urethra was blind-ended and measured 1.5 cm on cystoscopy. Treatment involved the formation of a single urethral channel by urethrourethrostomy, involving a septal incision from the point of the meatus.

Case 6 (Supplemental Fig. 3B) had primary symptoms of dysuria and a weak urine stream. A ventral blind-ended urethra was observed during cystoscopy that originated from the bulbous region. A urethrourethrostomy was performed via an endoscopic septal incision between the two urethras.

2.2. Type II urethral duplication

Case 7 (Fig. 1) was classified as type IIA1 with two meatuses and two independent, noncommunicating urethras originating from the bladder. The dorsal urethra had an epispadial urethral opening. Initially, an epispadias repair was attempted, but the ventral urethra was incidentally found on a retrograde urethrogram. The dorsal urethra was stripped to avoid injuring the nerve and was then dissected proximally to just below the pubic bone and ligated at that point. An additional ventral plication of the penis was required to correct a dorsal curvature.

Case 8 was classified as type IIA2 with a dorsal urethral opening in the penopubic region. A dorsal urethral excision using the stripping technique was performed owing to recurrent inflammation.

Case 9 was classified as a type IIA2 with a two apical meatuses, and the dorsal urethra was found to be separate from the verumontanum. The ventral urethra appeared hypoplastic on cystoscopy. This patient had vesicoureteral reflux and underwent surgical treatment. Surgical repair of the dorsal urethra was not performed.

Two cases were classified as type IIA2 Y-type with an anorectal malformation. In both patients, urine flow was observed in the ventral urethra near the anus. In case 10, the patient was also diagnosed with an imperforate anus, and correction of the urethra was performed after a Pena procedure and colostomy repair. The ventral urethra was extensively mobilized to the perineal–scrotal junction; the distal end of the ventral and proximal dorsal urethras was connected via single-stage urethroplasty using tubularized buccal mucosa. In case 11 (Fig. 2), a low type anorectal malformation was corrected using the Pena procedure; also single-stage urethroplasty with a buccal mucosa tube graft was performed for Y-type duplication. In both cases, owing to a stricture in the neourethra, several additional procedures, including visual internal urethrotomy and urethroplasty (end-to-end anastomosis and buccal mucosa tube graft), were required after the initial surgery.

2.3. Type III urethral duplication

Case 12 (Fig. 3) was classified as a type III urethral duplication. Symptoms included pain in the penis and pus from the midpenile shaft emanating through a dorsal urethral opening. A preoperative MRI confirmed that an anterior accessory bladder was present. The dorsal urethra originated from the anterior bladder. Intravenous pyelography confirmed that both ureters were inserted into the posterior bladder. The anterior bladder was excised via a transpubic approach using a Pfannenstiel incision. The dorsal urethra was then excised from the penis to the lower side of the pubic bone.

Table 1
Twelve cases of urethral duplication.

Case No.	Age	Presentation	Type	Other abnormalities	Management
1	2Y5m	Dorsal accessory urethra Dorsal penile root opening	IA	Penile ventral curvature Chordee	Excision of accessory urethra Chordectomy
2	11Y6m	Dorsal accessory urethra Double apical	IA	Hypospadias	Excision of accessory urethra, TIP
3	9Y11m	Dorsal accessory urethra Double apical	IA	Chordee	Excision of accessory urethra, Chordectomy
4	12Y2m	Dorsal accessory urethra, Dorsal penile root opening	IA	None	Excision of accessory urethra
5	1Y2m	Dorsal accessory urethra Double apical	IA	Concealed penis	Urethrourethrostomy Penoplasty
6	3Y10m	Proximal accessory urethra	IB	None	VIU
7	1Y	Epispadial dorsal accessory urethra	IIA1	Epispadias	Excision of accessory urethra
8	7Y11m	Dorsal accessory urethra Penopubic opening	IIA2	None	Excision of accessory urethra
9	8 m	Dorsal accessory urethra, Double apical	IIA2	Horseshoe kidney VUR	Vesicostomy Ureteroneocystostomy
10	3Y1m	Y-type duplication	IIA2	VACTER syndrome	Urethroplasty with BM VIU
11	2Y4m	Y-type duplication	IIA2	Anorectal malformation	Urethroplasty with BM Open urethrotomy
12	9Y4m	Dorsal accessory urethra, Dorsal midpenile opening	III	None	Excision of accessory urethra and bladder

NO, number; VUR, vesicoureteral reflux; TIP, tubularized incised plate; BM, buccal mucosa; VIU, Visual internal urethrotomy.

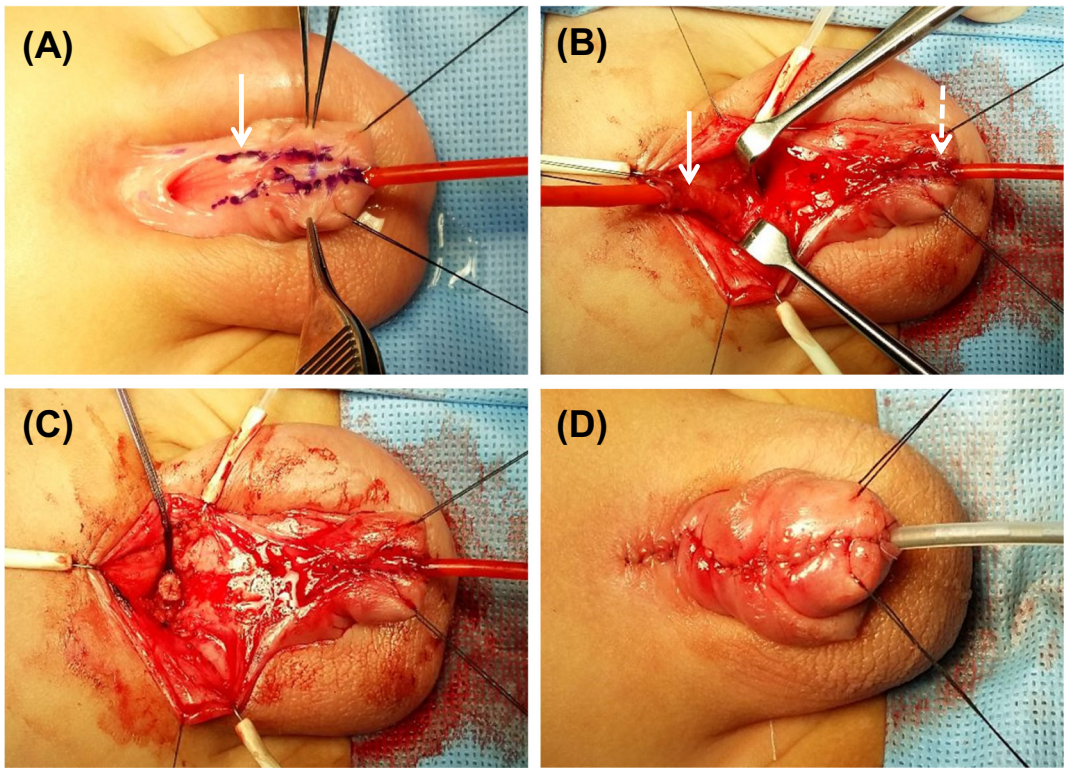

Fig. 1. Case 7 (type IIA). Steps for the excision of the dorsal urethra by stripping. (A) External anatomy of the duplication with a urethral catheter in the ventral urethra (solid arrow, dorsal epispadial urethra). (B) Separation of the two urethras by dissection of the dorsal epispadial urethra (solid arrow, dorsal epispadial urethra; dotted arrow, ventral urethra). (C) Ligation of the dorsal urethra beneath the pubic bone. (D) Surgical outcome after excision of the dorsal urethra by stripping.

2.4. Postoperative voiding conditions

Cases 1–5, 12 had no preoperative or postoperative voiding symptoms. Case 6 had a weak urine stream and dysuria prior to surgery. Before surgical correction, the maximum urinary flow rate (Qmax) was 4 mL/s, and the flow curve plateaued. Two months after the endoscopic septal incision, the Qmax increased to 17 mL/s, and the flow curve became bell-shaped, indicating improvements in voiding symptoms.

Cases 7 and 8 had no difficulties in voiding before or after surgery. After the dorsal urethral excision, the Qmax values were 17 mL/s and 10.4 mL/s, respectively, and both patients' flow curves were bell-shaped.

Cases 10 and 11 underwent several procedures owing to urethral stenosis and were evaluated 25 and 23 months later after last procedures, respectively. Qmax values and postvoiding residual (PVR) volumes were 5.7 mL/s and 12 mL (case 10), and 4.1 mL/s and 0 mL (case 11). While both patients had low Qmax values and small PVR volumes, neither patient complained of voiding symptoms after the last procedure.

3. Discussion

The origin of urethral duplication during embryogenesis is unclear, and numerous theories have been postulated as the anatomy of the

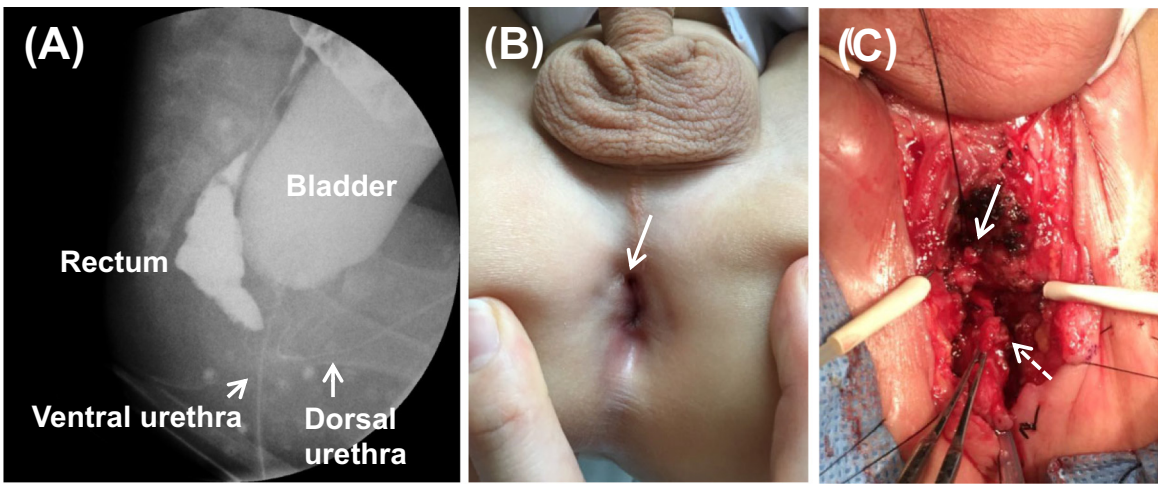

Fig. 2. Case 11 (type IIA, Y-type duplication). (A) Simultaneous cystography and administration of a barium enema show that there was no fistula between the rectum and urethra. (B) The white arrow indicates that the opening of the ventral urethra was at 12 o'clock from the anus. (C) An intraoperative image, showing the dissection of the ventral urethra (solid arrow, dorsal urethra; dotted arrow, ventral urethra).

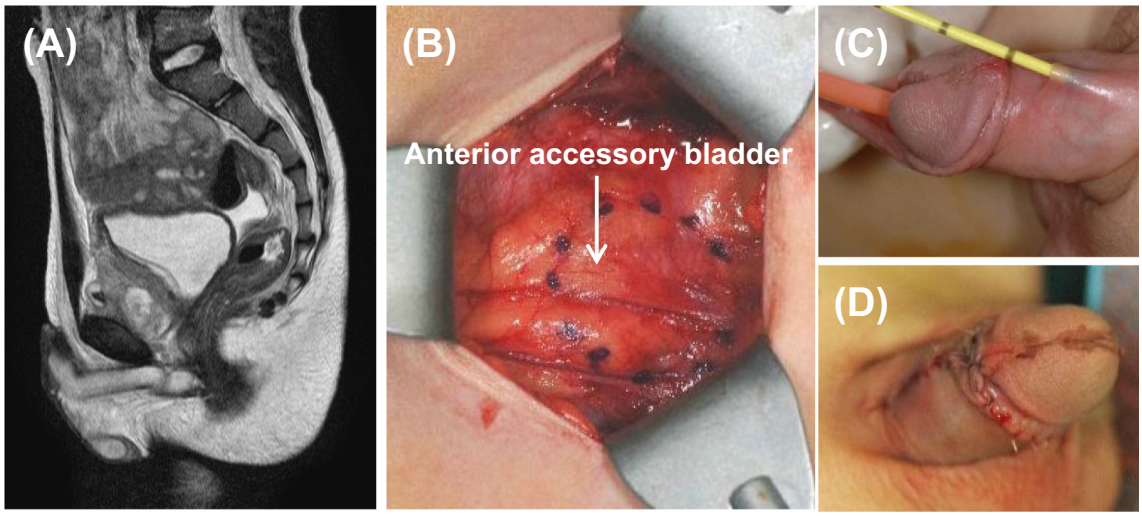

Fig. 3. Case 12 (type III). (A) A magnetic resonance image showing the presence of an anterior accessory bladder. (B) An intraoperative image showing the use of the transpubic approach. (C) External anatomy of the opening of the dorsal urethra. (D) Surgical outcome after the excision of the anterior bladder and dorsal urethra.

malformation varies widely [7,8]. However, to our knowledge, no single theory explains all of the various types of anomalies. Owing to the wide spectrum of anatomical deformities, it is difficult to apply a formalized treatment according to each type of urethral duplication. Thus, detailed information about individual surgical treatments is essential for their clinical application. In this report, we describe various surgical approaches based on their anatomical and functional characteristics in detail.

Duplication of the urethra can be either asymptomatic or present with various symptoms that range from inflammation to a double urine steam. The surgical treatment for urethral duplication is usually indicated if there is a bothersome symptom or cosmetic problem [1,3]. In case 9, surgical correction of the urethra was not required because the dorsal accessory urethra on the apex of the glans was nearly obliterated without any sign of inflammation of infection. The remaining 11 cases underwent various surgical treatments. In cases 1–4, 7, and 8, the dorsal accessory urethra was excised using the stripping technique. A type I duplication has an obliterated, nonfunctional dorsal urethra that is not connected to the ventral urethra, so there is no difficulty in excising the dorsal urethra using the stripping technique. In cases 7 and 8, surgery was performed to resolve a double stream and cosmetic problems. In both cases, a functional, ventral urethra, which included the verumontanum and sphincter, was present, and thus dorsal urethra excision was performed. Ablation of the dorsal urethra can be considered if the only purpose is to address its patency. However, owing to concerns about corporeal thrombosis or fibrosis, this method is no longer in use [4].

Surgical approaches should be carefully considered in cases of a type II A urethral duplication because the dorsal urethra will have different points of proximal insertion from the posterior urethra to the bladder. The use of either a transpubic approach or stripping is customary for dorsal urethral excision [1–3,7]. We stripped the dorsal accessory urethra, taking care not to damage the external sphincter or neurovascular bundle, and ligated the urethra underneath the pubic bone as proximally as possible. The use of postoperative cystourethrography demonstrated that the dorsal urethra could be successfully and completely excised, even in a type II A1 duplication with insertion of the dorsal urethra into the bladder. We believe that for dorsal urethral excision, the use of stripping techniques to minimize dissection ranges yields satisfactory results for both cosmetic and functional outcomes.

Case 7 was originally scheduled for surgery to correct epispadias, but another functional urethra was incidentally observed under the epispadial urethra. Cases of urethral duplication combined with epispadias have been reported occasionally; in such cases, embryological development is believed to be associated with an exstrophy–epispadias complex [9]. There are also reports of an accessory urethra found after bladder exstrophy repair [10]. Even though it is rarely reported, a careful physical examination of the urethra is required in patients with an exstrophy–epispadias complex to confirm a urethral duplication.

Cases of duplication in which one of the urethral meatuses is located in the anal or perineal region, called a Y-type duplication, are known to be associated with abnormal development of urorectal septum, and are most likely the result of ischemia during embryogenesis or impairments in the growth of the dorsoinferior wall of the urogenital sinus [7,8,11].

Our two cases of Y-type duplication had an orthotopic dorsal urethra, with a functional ventral urethra located in the perineal or anal area. The ventral urethra usually had a functional sphincter and verumontanum, whereas the dorsal urethra was hypoplastic, similar to previous reports [12,13].

Surgical correction of Y-type duplication requires a procedure linking the gab of the proximal end of the ventral urethra and the distal end of the dorsal urethra, which is challenging procedure, because complications such as neourethra dehiscence and stenosis can occur. In our study, a significant number of surgical procedures were required for each patient with a Y-type duplication because of complications. Several surgical techniques including scrotal flap, tubularized preputial island flap and buccal mucosa graft have been introduced to bridge the urethral gab [1,4,14]. Because of complications, such as a hairy urethra and poor surgical outcomes, urethroplasty using a tubularized preputial island flap or buccal mucosa graft was considered a better option [2]. We performed single-stage urethroplasty with a buccal mucosa tube graft to bridge the gap between the proximal end of the ventral urethra and the distal end of the dorsal urethra. We believe that minimizing the length of the neourethra constructed from tubularized preputial island flap or buccal mucosa grafts can reduce complications by transferring the majority of the ventral urethra in the perineal area to the perineal–scrotal junction via extensive mobilization. However, several additional procedures, including visual internal urethrotomy and urethroplasty, are often required, regardless of the initial surgical technique performed; such a necessity for additional procedures is probably owing to the hypoplastic nature of the dorsal urethra as a result of ischemia of embryological origin [8]. A Y-type duplication is associated with many congenital anomalies, such as VACTERL complex (vertebral anomalies, ventricular septal defect, anal atresia, tracheoesophageal fistula, radial dysplasia, renal anomalies, and cleft lip/palate) and cloacal exstrophy [15]. Therefore, long-term outcomes for patients with a Y-type duplication emphasize the need for careful planning because complex-associated anomalies can affect prognosis and require multiple procedures for reconstruction [16].

Urethral duplication accompanied by bladder duplication in the coronal or sagittal plane is rare [17]. Case 12 was classified as a type III sagittal duplication. Woodhouse and Williams noted that bladder duplication more commonly occurs in the coronal plane [8]. Coronal duplication typically results in two hemitrigones, each with the insertion of an ipsilateral ureter. Conversely, a sagittal bladder duplication is extremely rare; when present, it is described as a nonfunctional anterior bladder. In the case of a type III sagittal duplication (case 12), excision of the nonfunctional anterior bladder and urethra was performed via a combination of transpubic excision and stripping of the urethra. In this case, the anterior bladder is often a source of infection, and thus, excision of the anterior bladder must be executed [17]. Recently, anterior accessory bladder excision using a laparoscopic technique was reported [18].

Summarizing the surgical techniques used in this study, in cases with accessory urethra, dorsal accessory urethra can be simply excised with stripping technique. If there is only a thin septum between the two urethras and stripping technique is unable to be applied, septal incision can be used for treatment.

Even if the dorsal urethra communicates with the ventral urethra or bladder, satisfactory results can be obtained by excising the dorsal urethra using the stripping technique. In cases of a Y-duplication, it is believed that shortening the length of the neourethra by displacing the ventral urethra toward the perineal–scrotal junction via extensive mobilization reduces complications from urethroplasty. In cases of sagittal duplication accompanied by bladder duplication, we can choose a bidirectional approach to remove the bladder through a Pfannenstiel incision and excise the urethra using the stripping technique.

Our study has a limited number of cases and lacks the objective outcomes of the patients owing to its retrospective nature.

However, our study includes most types of urethral duplications, introduces surgical techniques described the literature, and describes our surgical technique according to the anatomical and functional characteristics of the duplications in detail.

4. Conclusion

Depending on the anatomical configuration of the urethral duplication, a variety of surgical procedures can be applied. In cases of a dorsal accessory urethra, a transpubic approach is usually unnecessary, and satisfactory results can be obtained by excision of the urethra beneath the pubic bone using the stripping technique. In cases of a Y-type duplication, additional procedures to prevent or address complications may be required, and a careful investigation of functional outcomes should be performed after urethroplasty.

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jpedsurg.2019.12.012.

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