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# Urethral duplication associated with complex chordee: a narrative review of literature and report of a case

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## Abstract

**Background:** Urethral duplication (UD) is reportedly rare. It is rarer in females. Knowledge on this anomaly comes from isolated report of cases. The aim of this review is to summarize information available on this anomaly thereby revealing gaps in knowledge, and to appropriately situate a recently managed case.

**Methods:** Publications on UD in English language from 2001 to 2021 were searched for in the literature. Of importance were age at presentation, class of UD, nature of associated penile deformity and other structural anomalies. Available information was used to synthesize opinions after descriptive analyses using SPSS<sup>®</sup> version 21 (IBM Co., Armonk, NY, USA). In addition, a recently managed case of UD was reported and appropriately situated in the discourse.

**Result:** In all, 115 articles met the inclusion criteria. Majority (75.7%) were individual case reports, while the rest were retrospective review of case series. These provided a total of 269 reported cases. Of this lot, 38 cases were excluded because they either had no Effmann's class or were not described well enough for an Effmann's class to be assigned. Ultimately, 231 cases formed the basis for this review. Male to female ratio was 12:1. Types III and IIB had more females. Types IIA 2 (26.0%), IIA 2 "Y" (26.4%) and IA (22.5%) were frequently reported. About 61.8% males and 68.4% females had no associated defects. Isolated dorsal chordee was prevalent (7.1%), especially among type IA (16.7%) UD. Reported in 10.4%, vesicoureteric reflux may not be attributable always to high pressure voiding.

A boy who presented at the age of 18 years with Effmann type IA UD and an associated complex chordee of the penile shaft was reported. The complex nature of the chordee adds to the challenge of explaining associated penile defects in UD.

**Conclusion:** UD is rare, but reported from all parts of the globe. There are a number of associated defects involving the external genitalia and other organs reported in UD in the male. The mechanisms of these defects are yet to be fully understood.

**Keywords:** Complex chordee, Urethral duplication, Effmann classification, Urethrography, Urethral excision, Case report

## 1 Background

Duplication of the urethra is a rare congenital anomaly of the lower urinary tract. It is rare in males and rarer in females [1]. Reported cases of urethral duplication (UD) in the literature so far is put at less than three hundred cases [2, 3]. Though the defect in embryogenesis resulting

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in this anomalous urethral development may not be fully understood yet, some theories have been put forward to explain the observed variants of this anomaly [4]. Usually the duplicate urethrae are sagittal in relation to each other, and the ventral urethra tends to be the dominant urethra in terms of function.

Currently, Effmann classification appears to be more commonly applied in discourses on urethral duplication [5, 6], whereas the aetiological factors for UD are not clearly understood yet, UD has been reported to be associated with a number of other structural defects in the physical appearance of the penis and/or scrotum [5, 7], in the lower and upper urinary tract [8], in the anorectum [9], and in remote organs such as the heart and vertebral column.

In many cases of UD in male, the external genitalia appear morphologically normal. However, reported morphological defects in the male external genitalia include isolated dorsal chordee and isolated ventral chordee which are usually explained as consequent upon possible disproportionate growth or tethering of tissues of the penis in the vicinity of the duplicate urethra [10–12]. More grotesque malformations such as bifid scrotum and penoscrotal transformation have also been reported in association with UD and may not be easily explained. Similarly, a combination of dorsal chordee, lateral chordee and torsion of the penile shaft, described as a complex chordee suggests that there may be other mechanisms acting to bring about the observed appearance of the external genitalia in UD.

The aim of this study is to perform a narrative review of the literature on UD, especially UD in males, with the purpose of summarizing reported associated deformities of the penis and other organs of the body, thereby revealing gaps in knowledge. A recent case of UD presenting in adulthood with complex chordee is presented and appropriately situated in the discourse.

## 2 Methods

The available literature was searched between November 2021 and January 2022 for journal publications in English language on urethral duplications from year 2001 to year 2021. Searches were done in PUBMED, MEDLINE, AJOL and Google Scholar using the MeSH terms “urethral duplication,” “penile deformities,” “chordee,” and “urethrocutaneous fistula.” In addition, the references of relevant publications from primary searches were reviewed to the point of saturation.

From the identified relevant publications, the year of publication, age at presentation, class of UD, nature of associated penile deformity, and other associated structural anomalies were extracted. Cases of urethral

triplication, penile or glans duplication, and cases of caudal duplication syndrome were not included.

In addition, cases that are not classified in their publications, or could not be classified by the reviewing team because of limited imaging descriptions of the anatomy in the publications were captured as unknown class, and were excluded from further interrogation in the review. Reports of UD in females were not also included in further analysis because a key feature of interest in this study, nature of associated penile deformities, is not applicable to females. The report of the index case was appropriately situated in the established context and discussed.

The Health Research Ethics Committee of University of Nigeria Teaching Hospital Ituku-Ozalla approved of this study, and written consent was obtained from the index patient to publish this case report.

## 2.1 Evidence synthesis

Seven hundred and seventy-one journal articles emerged through the searches, but 115 of this lot met the inclusion criteria, among which were 87 case reports and 28 retrospective review of cases. The 115 journal articles yielded 269 cases of UD from which 38 cases were excluded because they were not given an Effmann class, or were not described well enough for an Effmann class to be assigned. Further analyses in this narrative review are based on these 231 cases. The extracted information was fed into IBM SPSS® version 21 (IBM Co., Armonk, NY, USA) and descriptive statistical analyses done.

## 3 Results

Table 1 shows the frequency distribution of the urethral duplications according to Effmann classification. In addition, the table shows the oldest age at diagnosis in each

**Table 1** Frequency distribution of the reported Effmann classes of urethral duplication, the oldest age at presentation for each class, and the female proportion of each class

Effmann Class	Frequency (Proportion of total)	Oldest Age at Diagnosis	Number (Proportion) of Females in class
Type IA	52 (22.5%)	54yrs	4 (7.7%)
Type IB	8 (3.5%)	20yrs	0
Type IIA 1	25 (10.8%)	56yrs	4 (16.0%)
Type IIA 1 “Y”	2 (0.9%)	11yrs	0
Type IIA 2	60 (26.0%)	35yrs	3 (5.0%)
Type IIA 2 “Y”	61 (26.4%)	30yrs	3 (4.9%)
Type IIB	11 (4.8%)	58yrs	2 (18.2%)
Type III	12 (5.2%)	52yrs	3 (25.0%)
Total	231		19 (8.2%)

class as well as the proportion of females in the various classes.

All the urethral duplications were sagittal in disposition except for 1 (1.9%) of type IA, 1 (12.5%) of type IB, 2 (18.2%) of type IIB and 3 (25.0%) of type III. About 61.8% (131) in males and 68.4% (13) in females had no associated defects in the external genitalia, or in any other organ.

In males, 38 (17.9%) presented with associated defects in the external genitalia. The frequency distribution of the reported defects in the external genitalia in males is shown in Table 2.

In addition, 1 (1.8%) isolated right lateral torsion of the penile shaft was reported among type IIA 2 duplications. Megalourethra was reported in 1 (1.7%) of type IIA 2 “Y” and in another 1 (11.1%) of type IIB duplications. A combination of dorsal chordee and penile shaft torsion to the left was reported in 1 (2.1%) of type IA duplications.

Continuing, 57 (24.7%) of males had defects in other organs beyond the external genitalia: vesicoureteric reflux [*n* = 22 (10.4%)], anomalies of the kidney such ectopic kidney, horse-shoe kidney and solitary kidney [*n* = 19 (9.0%)], and anorectal anomalies [*n* = 10 (4.7%)] were the common defects in other structures reported in association with UD in the males. The dataset with all parameters of interest is available at Mendeley Data, V1, <https://doi.org/10.17632/8jy398xzhm.1>.

### 3.1 Report of the index case

An 18-year-old boy presented to the outpatient clinic with complaint of abnormal shape of his penis from birth. His penis was noticed from birth to be bent upward and to the left. This deformity worsened at puberty creating increasing anxiety for the boy and his mother. After his circumcision as a neonate, an opening was noticed at the dorsum of the penis just proximal to the corona. Urine did not pass through the opening and

no discharge of any fluid was noticed either. He did not experience any symptoms of urinary tract infections. His scrotum and testes appeared normal, and he had no bowel symptoms.

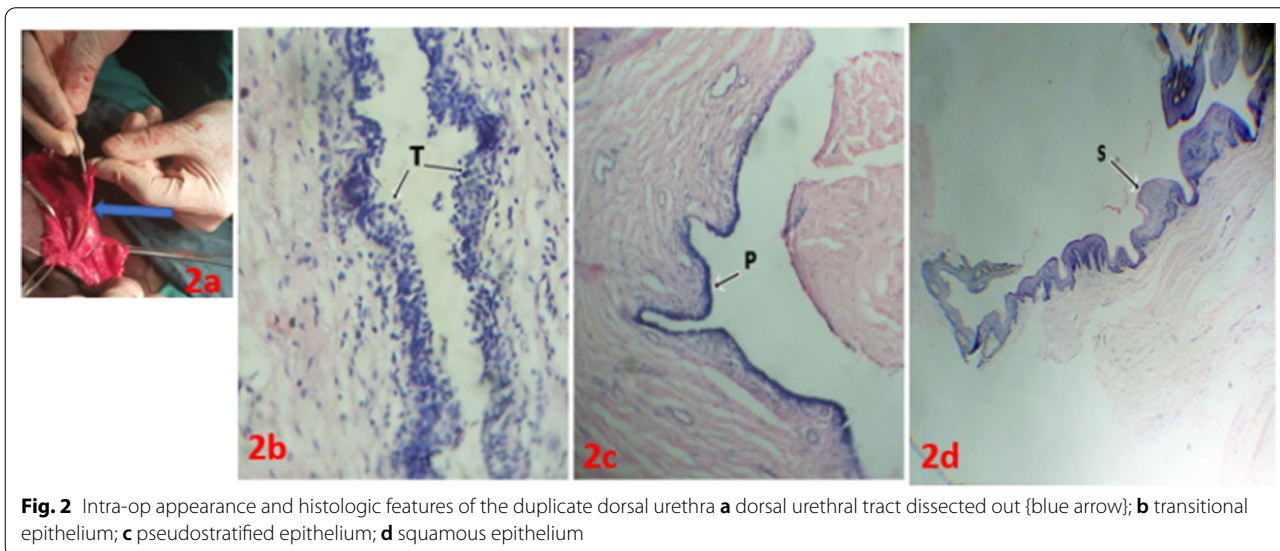
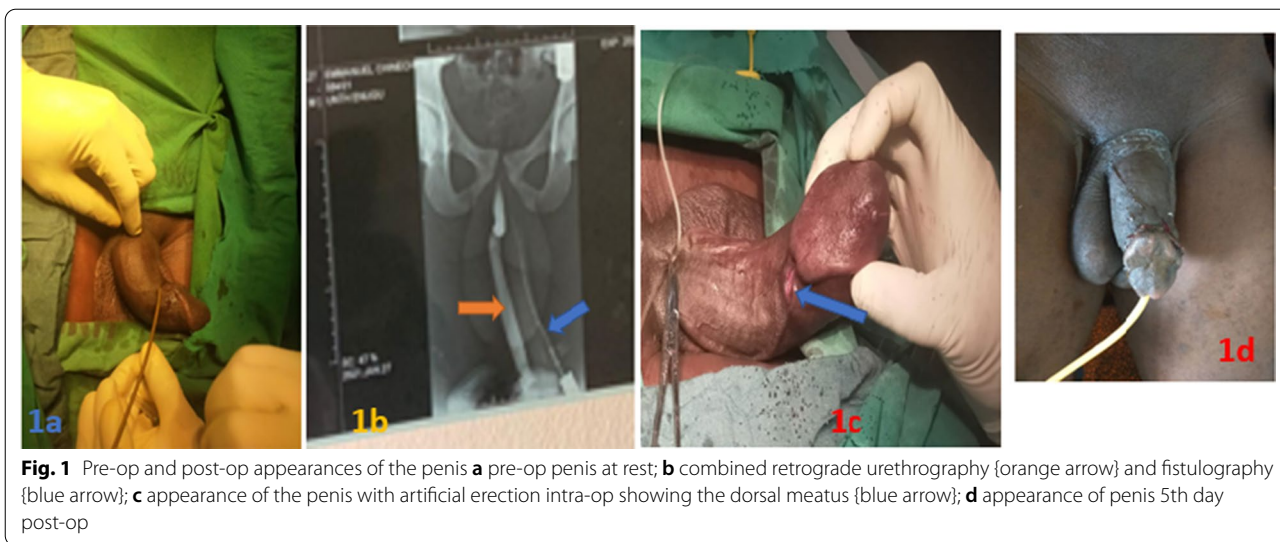
Upon physical examination, he had an appropriate size penis for his age. The penis at rest was as shown in Fig. 1a with a normally situated urethral meatus at the summit of a conical glans penis. Another opening was seen at the dorsum of the penile shaft just proximal to the coronal sulcus. Further evaluation showed normal abdominal sonogram, retrograde urethrocytogram, voiding cystourethrogram and intravenous urogram. Fistulography revealed a tract along the dorsum of the deviated penile shaft from the dorsal meatus in the sub-coronal location to a blind end at the root of the penis (Fig. 1b). Urethrocytostomy through the orthotopic meatus confirmed adequate urethra with obvious verumontanum, normal bladder neck and bladder. With methylene-blue dye instillation into the dorsal tract, there was no communication between the dorsal tract and the orthotopic urethra or urinary bladder.

Intraoperatively, artificial erection revealed the appearance of the penis during erection (Fig. 1c). The penis on complete degloving revealed that the dorsal urethral tract was running beneath the Buck’s fascia. The tract was dissected out completely with the aid of a sinus probe, and the complex chordee corrected using a combination of tissue-releasing incisions, Heineke-Mikulicz and Nesbit curvature correcting manoeuvres [13, 14]. Histological examination of the excised tract (Fig. 2a) revealed an epithelium-lined tissue with transitional, pseudostratified and squamous epithelia at different sections (Fig. 2b–d).

The post-op period was uneventful and on the 5th day post-surgery, the penis had a satisfactory outlook (Fig. 1d). At the last follow-up visit, 12 months post-surgery, he reported satisfactory erection.

**Table 2** Frequency distribution of reported defects of the external genitalia according to Effmann class in males

Effmann class	Isolated dorsal chordee (n = 15)	Isolated ventral chordee (n = 4)	Isolated bifid scrotum (n = 3)	Penoscrotal transposition (n = 8)
Type IA (n = 48)	8 (16.7%)	1 (2.1%)	0	0
Type IB (n = 8)	1 (12.5%)	0	0	0
Type IIA 1 (n = 21)	2 (9.5%)	1 (4.8%)	0	0
Type IIA 1 “Y” (n = 2)	0	0	1 (50.0%)	0
Type IIA 2 (n = 57)	3 (5.3%)	2 (3.5%)	0	1 (1.8%)
Type IIA 2 “Y” (n = 58)	0	0	2 (3.4%)	7 (12.1%)
Type IIB (n = 9)	0	0	0	0
Type III (n = 9)	1 (11.1%)	0	0	0
Total (n = 212)	15 (7.1%)	4 (1.9%)	3 (1.4%)	8 (3.8%)



#### 4 Discussion

Urethral duplication is observably a rare disorder on the assumption that most if not all cases are reported. It is rarer in females with a male to female ratio of 12:1 from this review (Table 1). Almost all that is known about the condition come from case reports and from retrospective review of cases. Could more robust studies in terms of tissue analyses, or the like be conducted on this subject matter? This narrative review reveals that reports have come from all parts of the world and that greater proportions of reported cases in males do not have symptoms attributable to the urethral defect as is observed in this index case. In addition, associated defects of the genitalia or other organs are not common. So, it is possible

that a good number of cases of UD may go undiagnosed and unreported. Beyond abnormal urine stream due to a functional duplicate urethra, challenges with cosmesis and sexual activities as in the index case (Figs. 1a and c) are reported where there are defects of the external genitalia [15, 16]. Other reasons for presentation, or other clinical findings are dependent on associated defects.

Many reports suggest that Effmann type I duplication, especially the type IA is the commonest variant [6], but this review suggests that the type IIA 2 duplication as well as the type IIA 2 “Y” variant may be as common (Table 1). The retrospective reviews of cases by Wani et al. [17] in India, Guglielmetti et al. [18] in Switzerland and AbouZeid et al. [19] in Egypt suggest



also that type IIA 2 and its “Y” variant are commonly encountered. However, it is possible that because type IIA 2 duplication and type IIA 2 “Y” variant are more challenging anomalies in terms of symptoms, cosmesis and surgical correction, they usually will not pass unattended to, or unreported, thereby increasing the frequency of such reports. On the other hand, except where they are diligently searched for, many Effmann type I UD may pass unnoticed and unreported resulting in type I being relatively less commonly reported.

In line with existing literature, this review shows that duplication in the sagittal plane is by far the commoner [2]. In Effmann type III variant, however, duplication in coronal plane may constitute a reasonable proportion (about 25.0% from this review). Our patient presented with type IA duplication in the sagittal plane (Fig. 1b).

With respect to associated defects of the external genitalia, dorsal chordee of varying severity is commoner among the type IA duplications from this review (Table 2). This may be because the ambient tissue of the dorsum of the penile shaft tends to be hypoplastic just as the dorsal non-orthotopic duplicate urethra is usually atretic and non-functional. It is possible to explain this association in line with the theory of dysplastic penile tissue in close proximity to poorly developing urethral tissue [20]. In line with the meeting report by Stadler and colleagues, further studies on this possibility concept are necessary [21]. Montag and Palmer noted from a review of penile curvatures that penile chordee could be anchored at the depth of the skin, dartos fascia, Buck’s fascia, or cavernous tissue [13]. Unilateral or bilateral corporal disproportion of varying extents will determine the attendant degree of dorsal and lateral deviations of the penile shaft. Our patient presented with a complex chordee that is at the depth of the Buck’s fascia manifest as a combination of dorsal and left lateral deviations as well as left torsion of about 80° (Fig. 1c). Such a combination of penile shaft curvatures and torsion adds to the challenge of explaining penile deformities in UD. Similarly, hypoplasia of ventral penile tissue resulting in penoscrotal transposition, bifid scrotum or ventral chordee is commoner with the “Y” type duplication possibly because of the concurrent non-orthotopic ventral disposition of the dorsal duplicate urethra to the ventral penile shaft, and the ventral duplicate urethra to the anal, perineal, or scrotal area.

In correcting significant chordee associated with UD, it is therefore appropriate to deglove the penile shaft so as to tackle the chordee at the appropriate tissue plane [13, 22]. That is the strategy deployed in correcting the complex chordee in our patient, the outcome of which is acceptable to all concerned (Fig. 1d).

Our patient does not have any other structural anomaly beyond the disfigured external genitalia as is seen with some other reported cases [2, 22–24]. From the scoping done, only about 24% of the reported cases of urethral duplication in males had other anomalies beyond the external genitalia. The prevalent anomalies reported are vesicoureteric reflux of varying grades and renal defects of differing categories [17, 18]. The reasons for the associations are not clear. Where present, vesicoureteric reflux does not appear to be due always to infravesical obstruction as many of these patients do not have high pressure voiding [17, 25]. Anorectal anomalies are seen more with the class “Y” duplications possibly because of the proximity of the ventral duplicate urethra to the anorectal/perineal region in this class of UD. Considering that some of these associated anomalies could be life-threatening, or organ-threatening, and it is therefore pertinent that they are sought for and managed appropriately [18, 23].

## 5 Limitations

It is quite possible that some cases of urethral duplication are not recognized, or are not reported, a situation that will definitely affect the frequency distribution of cases as reported in this narrative review. There exists a chance also that a case reported as case report by one set of authors may find its way into a retrospective review of cases managed by another set of authors. Where this was suspected during the scoping because of similarity in authorship, author affiliation and year of presentation of the case of interest, the case report was excluded.

## 6 Conclusions

Urethral duplication is a rare disorder that has been reported from all the regions of the world. It is rarer in females. It has been identified at birth, childhood as well as early and late adulthood incidentally, or as a result of abnormal urine stream, poor cosmesis, or features of associated anomalies. In addition, there are quite a number of issues regarding UD that have not been researched on deeply. For instance, there is limited evidence in literature that the nature of the association between urethral duplication and chordee or penile deformities as well as the difference in the nature of tissue interactions during development between ambient nonurethral tissue and urethral tissue in the contexts of orthotopic and non-orthotopic urethral location. The presence of complex chordee in association with Effmann type IA urethral duplication as reported in our index case adds to the complexity in explaining the association between chordee and urethral duplication.

**Abbreviation**

UD: Urethral duplication.

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**Author contributions**

IN contributed to evaluation and treatment of index patient, study design, literature search, scoping review and data acquisition, and drafting the manuscript; WCO contributed to evaluation and treatment of index patient, literature search, scoping review and data acquisition; FIU contribute to evaluation and treatment of index patient, study design, literature search, scoping review and data acquisition, and critical review of the draft manuscript; EON contributed to evaluation and treatment of index patient, study design, literature search, and drafting the manuscript; CAI contributed to evaluation and treatment of index patient, study design, literature search, scoping review and data acquisition; INA contributed to evaluation and treatment of index patient, study design, scoping review and data acquisition; CAN contributed to study design, literature search, scoping review and data acquisition, and critical review of the draft manuscript; ACO contributed to evaluation and treatment of index patient, study design, literature search, scoping review and data acquisition. All authors read and approved the final manuscript.

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**Availability of data and materials**

Data supporting this study are available at Mendeley Data (Nnabugwu, Ikenna; Nnabugwu, Chinwe (2022), "Scoping review of literature in urethral duplication", Mendeley Data, V1, <https://doi.org/10.17632/8jy398xzhm.1>.

**Declarations****Ethics approval and consent to participate**

The Health Research Ethics Committee of University of Nigeria Teaching Hospital Ituku-Ozalla approved of this study.

**Consent for publication**

Consent for publication was obtained from the index patient.

**Competing interests**

The authors declare no competing interests.

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