Original Article

Chordee without hypospadias: Operative classification and its management

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Abstract

Context: Developing countries.

Aims: To propose a operative classification of Chordee without hypospadias (CWH) with its management.

Settings and Design: Tertiary referral centre; Retrospective study from January 2000 to January 2011.

Materials and Methods: Total 26 patients were classified peroperatively into sixtypes (A: Cutaneous chordee→ Degloving skin and dartos (1/26); B: Fibrous chordee→ chordectomy (4/26);C: Corporocavernosalchordee→ Corporoplasty ± Urethral mobilization (4/26); D: Urethral tethering with Hypoplastic urethra→ Urethral mobilization ± urethral reconstruction because of hypoplastic urethra (14/26); E: Congenital short urethra→ excision of urethra from the meatus and urethroplasty (2/26); and F: Complex chordee→ Degloving ± Corporoplasty ± urethroplasty (1/26 patients).The follow-up over 6 months to 9 years were analyzed.

Statistical Analysis: SPSS soft ware version 17.0 for Windows.

Results: The mean age of surgery was 5.33 ± 0.11 years. The success rate defined on uroflowmetry and voiding cystourethrography was 65.6%. The coronal urethra-cutaneous fistula developed in 26.9% (7/26) {including 7.7% (3/26) of associated metal stenosis}. The urethral stricture developed in 3.8% (1/26).

Conclusions: CWH needs stepwise surgical management. The operative classification may help in better understanding and management of this difficult entity. Meticulous tissue handling and urethroplasty is needed for good and promising results.

Key Words: Chordee without hypospadias, congenital short urethra, hypospadias, hypospadias sine hypospadias

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INTRODUCTION

The term chordee means "curvature".[1] The term chordee without hypospadias (CWH) is used when the meatus is located at the tip of the glans penis, yet prepuce is distorted and, ventral penile curvature is associated with abnormalities of the fascial tissues, corpus spongiosum, or both [Figure 1].[1-4] Isolated CWH is a rare entity, which comprises 4-10% of all congenital chordee.[2] The CWH or Hypospadias sine hypospadias cripple is an iatrogenic anomaly that results from the failure of surgical repairs.[4-6]

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The entity was first described by Siever in 1962, since then, various classifications were advocated for it.[7] The most widely accepted one is modified Devine *et al.* classification.[1-5] According to which cutaneous chordee corresponds to type III; fibrous

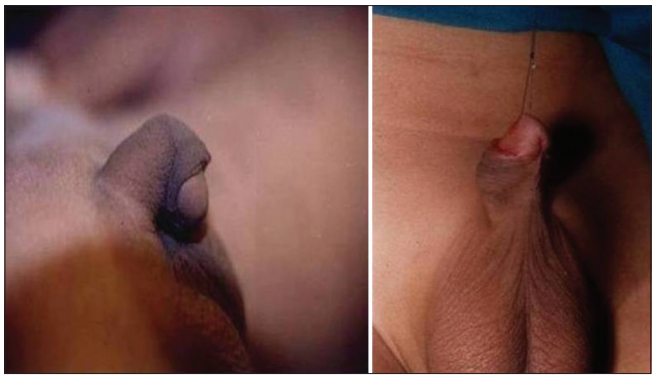


Figure 1: Preoperative photograph showing (a and b) severe chordee with congenital short urethra

chordee, to type II; corporocavernosal chordee, to type IV; and congenital short urethra (CSU), to types I and V.[1-5] On extensive study of the literature, authors believe that all these classifications were confusing and not covering all aspects of the entity. Hence, there is still need for development of easy and understandable classification covering all the aspects of the entity. The authors have proposed a simplified operative classification according to the steps of surgery.

MATERIALS AND METHODS

Data of all patients having CWH was retrospectively reviewed at the Department of Pediatric Surgery of the University Hospital from January 2000 to January 2011. We have excluded cases of Hypospadias sine hypospadias cripple. The ethical approval was taken from ethical committee of the university. Informed and written consent was taken from the parents. All patients were operated by single surgeon (second author). The data was analyzed using SPSS 17.0 version for Windows. Continuous variables were expressed as mean values with 2 standard deviation, median and ranges (minimum to maximum), as well as in percentages.

The surgery was performed under general anaesthesia. A glanular stay suture (polypropylene 5-0) was taken to minimize tissue handling, which was also used to secure the urethral stent at end of surgery. Adrenalin with 1% lidocaine (1:100,000) was infiltrated along the proposed site of incision and into the glans deep to urethral plate to facilitate hydrodissection and hemostasis. The authors did not prefer tourniquet application for hemostasis as it can conceal the proximal limits of penis. A circum-coronal incision was made 3 mm below the corona. According to authors' classification, patients were categorized intraoperatively into six types [Table 1]. In cutaneous chordee (type A), dysgenic and inelastic tissue in the dartos and, to an extent, Buck's fascia was present (the corpus spongiosum was normal). After degloving by dissection of skin and dartos

Table 1: Operative classification

Type	Nomenclature	Correction required
A	Cutaneous chordee	Degloving by dissection of skin and dartos up to penopubic angle dorsally and just below penoscrotal angle ventrally
B	Fibrous chordee	Excision of fibrous tissue lateral urethral plate known as chordectomy
C	Corporocavernosal chordee	After degloving, chordectomy and full urethral mobilization, corpora needs dorsal plication/ventral grafting/complete corporo-spongial disassembly
D	Urethral tethering associated with hypoplastic urethra	If degloving, chordectomy and full urethral mobilization (corporocavernosal chordee excluded) left otherwise straight penis with normal length but hypoplastic urethra. The urethra has deficient spongiosum on ventral aspect or completely dysgenic urethral epithelium and spongiosum. The urethra needs lay open/excision with reconstruction
E	Congenital short urethra	Normally developed urethra but short in length (true congenital short urethra), which needs urethral reconstruction
F	Complex chordee	Association of fibrous chordee with or without corporocavernosal chordee, and/or congenital short urethra. The entity needs degloving with chordectomy±corporoplasty± urethroplasty

up to penopubic angle dorsally and just below penoscrotal angle ventrally, cutaneous chordee was released. Thereafter, a tourniquet was transiently applied at the base of penis and artificial penile erection performed by intracorporeal normal saline instillation by 26 gauze needle. If chordee was present, fibrous tissue derived from Buck's and dartos fascia lying deep and lateral to the partially abnormally developed corpus spongiosum (type B: Fibrous chordee) was excised (chordectomy). Now, artificial erection was again induced, if curvature still persisted, whole corpus spongiosum was mobilized from glans to penoscrotal junction along whole length of corpora cavernosa and decision was taken for corporoplasty alone or with urethroplasty.

In corporocavernosal chordee (type C), corpus spongiosum and fascial layers normally developed, and after full urethral mobilization, shortness/inelasticity of the tunica albuginea of the corporal bodies was the cause of chordee. In adequate length penis with minor bending (<30°), we performed dorsal plication (excised) or excising ellipses of dorsal tunica albuginea (dorsal corporoplasty). If curvature still persisted, the urethral plate was transected proximally and deficient urethra was reconstructed in same stage. In short length penis, corpora needed either ventral lengthening procedure via incision at transverse axis of corpora and patching the gap with autologus graft, or dorsal corporoplasty via transverse corporotomy with longitudinal closure. The artificial erection was repeated to confirm the absence of persistent chordee. The authors' preference for urethroplasty was mostly single stage until deficient urethra extending too proximally or penile length is

too short (penile girth/penile length < 10th percentile for age).

If degloving and chordectomy left otherwise straight penis with normal length but hypoplastic urethra, that is papery thin urethra through which white line of silastic catheter can be well visualized with necked eyes (type D), the urethra was laid open (if spongiosa was dysgenic on ventral aspect only) or excised (spongiosa poorly developed on dorsal as well as ventral aspect) up to normally developed spongiosum. For dysgenic ventral urethra, the glans wings were elevated with taking care not to divide the spongiosum insertion on the glans. The glanular groove should be about 12-14 mm in < 10 years of age and 25 mm in teenagers, if less than this a dorsal vertical incision was made until the width of glanular groove is adequate for the meatus. Now about 1 cm wide transverse inner preputial onlay island flap (OIF) based on dorsal dartos was rotated ventral and suture to spongiosa over urethral stent.

For resected urethra, reconstruction was performed by tubularised vascular flaps either via Duckett's tube, that is transverse prepucial island flap based on dorsal dartos flap (TIPF-DD) or Asopa skin tube. A Y-shape incision was made on the glans, the centre of which was at the site of neomeatus. Upper two limbs of Y was 0.5 mm while long vertical limb extended down to whole length of glans penis to coronary sulcus. Thus, three flaps were raised at glans and core of tissue was excised to incorporate neourethra. A 1.5-cm wide rectangular flap of inner prepuce was tubularised on urethral stent and anasotomosed proximally to meatus after adequate spatulation, and stitched proximally and dorsally at glans. The mobilized glans wings were rotated medially to cover the neourethra. The suture line of neourethral tube was placed toward corpora.

The Asopa tube was reconstructed from inner surface of foreskin keeping the common blood supply for skin and neourethra. The prepuce was divided longitudinally in two parts (right larger than left). The right sided inner prepuce was tubularized on urethral stent, rotated ventrally and anastomosed to proximal urethra. The glans was split in midline. The neourethra was placed within glanular bed, sutured with glans and covered by glans flap. The ventral surface was covered by right prepucial flap, while left flap was also rotated from opposite side and sutured on ventral aspect of penis.

If after urethral mobilization and transection of urethra distally, on artificial erection, there was no corporocaevernosal curvature, we confirmed that CSU was causing the chordee. In other words straightening of the penis required resection of urethra and excision of fibrous tissue (type E). The CSU was also reconstructed by TIPF-DD or Asopa Skin tube as described above.

There may be cases of complex chordee (type F: Association of fibrous chordee ± corporocavernosal chordee ± CSU) needed chordectomy ± corporoplasty ± urethroplasty.

After appropriate urethroplasty, barrier to neo urethral tube was provided by double breasting of penile dartos flap in distal penile urethoplasty. If there was hypoplastic urethra or short urethra need urethral reconstruction up to base of penis mid scrotal septal dartos flap was used for augmentation of neourethral tube. The glanuloplasy was performed and Byars' skin flaps used for the coverage of penile shaft skin.

Diverting urethral stent was placed for 10-12 days in all except patients didn't need urethroplasty (3 days). Postoperatively, 0.2 mg/kg oxybutenin given thrice in a day for 2 days, followed by once at bedtime for 10 days. Immediate and follow-up results over 6 months to 9 years were recorded. Postoperatively, the success of surgery was confirmed by physical examination, voiding cystourethrography, and uroflowmetery.

RESULTS

Over 11 years of period, total 26 patients of CWH were operated. The mean age at surgery was 5.33 ± 0.11 years. Patient's characteristics and their management are shown in Table 2. In 65.38% (17/26) patients, tethered urethra was found, which needed complete urethral mobilization and division. The hypoplastic urethra was present in

Table 2: Categorization of patients' and their management

Type	Mean age(years)	*N* (%)	Associated anomalies	Procedure performed	Management of complications	Recurrent complications
A	4	1 (3.8)	-	Degloving of skin and dartos	-	-
B	4.33±0.01	4 (15.4)		Chordectomy	-	-
C	5.53±0.11	4 (15.3)	-	• MDP=2 • Ventral dermal graft=1 • Corporo-spongeal disassembly=1	-	-
D	4.67±0.67	14 (53.8)	Partial Peno-scrotal transposition=2 Penile torsion=4	• OIF=2 • TIPF-DD=7 • Asopa's tube urethroplasty=5	• Meatotomy=3 • UCF repair=5 • Redo-urethroplasty for stricture=1	Recurrent UCF=1
E	5.30±0.11	2 (7.7)	-	TIPF-DD=2	• UCF repair=1	-
F	4	1 (3.8)	-	MDP+TIPF-DD urethroplasty	-	-

CSU: Congenital short urethra; MDP: mid dorsal plication, OIF: Onlay island flap, TIPF-DD: Tubularised transverse inner preputial island flap based on dorsal dartos; UCF: Urethrocutaneous fistula

53.8% (14/26) patients, which was laid open or excised until normal urethra encountered [Figure 2a]. The OIF urethroplasty was performed in 2, TIPF-DD in 7, and Asopa's preputial skin tube urethroplasty in 5 patients [Figure 2b and c]. Two cases of CSU were managed by TIPF-DD while one patient needed corporoplasty with urethroplasty.

There were no intraoperative complications. In immediate postoperative period, wound infection rate was 23.0% (6/26), which was managed conservatively. In mean follow-up of 24 ± 0.34 months (ranged from 6 months to 9 years), coronal urethra-cutaneous fistula (UCF) developed in 26.9% (7/26) and urethral stricture in 3.8% (1/26) [Table 3]. The submeatal stenosis was associated with UCF in 7.7% (3/26) of patients [Figure 3]. The success rate (defined by normal bell shaped curve on uroflowmetery and normal calibre urethra on voiding cystourethrogram) was 65.4% (17/26 patients). On 6 months of conservative treatment, five UCF failed to close spontaneously, which needed simple closure. One redourethroplasty done for stricture urethra. Recurrent UCF was developed in one patient.

DISCUSSION

Devine *et al.* proposed a classification for CWH and divided it into three types.[8] Krammer *et al.* added corporal disproportion as type IV.[9] The widely accepted classification is modified Devine *et al.* classification.[1-5] Donnahoo *et al.* gave another classification for CWH.[2] The authors believe that all these proposed classifications were confusing in some aspects. As in previous classifications CSU was categorized in both I and V, there were no description of hypoplastic urethra, and no description of complex chordee where curvature attributed by more than one factors. Further, the definite sequence was missing in all these classifications. The authors have proposed a simplified classification according to the steps of surgery as shown in the algorithm [Flow chart 1]. The authors also added a new type F in the classification, which is not mentioned in any of the previous classifications. If the surgery is performed meticulously according to the steps described, there will be no issue of residual chordee. In all types of authors' classification except types A and B, mobilization of urethra may be needed to decide whether the curvature was due to urethra or not?[1,5,10-13]

The age of presentation was from 5 to 10 years, which signify that the entity was not noticed by any of parents up to 5 years of age. This highlighted the need of awareness of the entity by medical practitioner. The urethral tethering is not always the cause of penile curvature, and many cases can be managed by the urethra-preserving procedures.[2,3] If CWH is associated with an abnormal meatus, it should be treated as concealed hypospadias, and entire dysplastic urethra should be reconstructed.[2] If the meatus is normal, peroperative decision must be taken for either urethral preservation or reconstruction, whichever is needed as per the situation.[2,11] Similarly, for dysgenic urethral tethering, options described in literature are division with reconstruction of urethra, mobilization ofthe anterior urethra with excision of the underlying fibrous tissue,

Table 3: Complications

Complications	OIF (*N*=2)	TPIF-DD (*N*=10)	Asopas' urethroplasty (*N*=5)	Total number of patients
Coronal UCF	2	3	2	7
Meatalstenosis	1	1	1	3
Urethral stricture	1	0	0	1
Wound infection	3	3	0	6
Urethral diverticulum	0	0	0	0
Residual/recurrent chordee	0	0	0	0
Skin necrosis	0	0	0	0
Glanular tilt	0	0	0	0
Glans dehiscence	0	0	0	0

OIF: Onlay island flap, TIPF-DD: Tubularised transverse inner preputial island flap based on dorsal dartos, UCF: Urethrocutaneous fistula

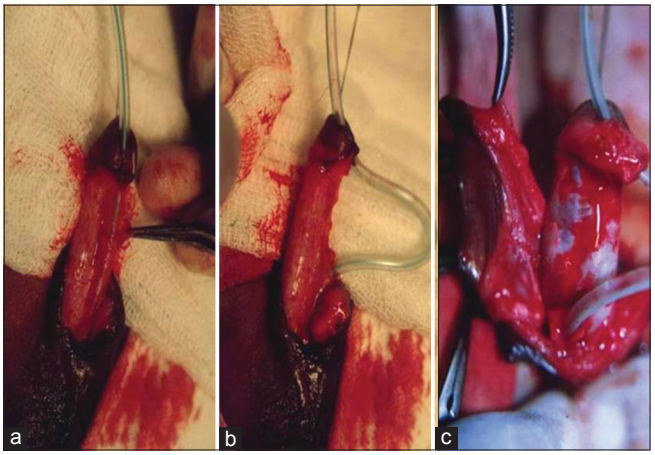


Figure 2: Peroperative photograph showing (a) Distal hypoplastic urethra, (b) Deficient spongiosum after excision of dysgenic hypoplastic urethra, (c) Asopa's tube urethroplasty

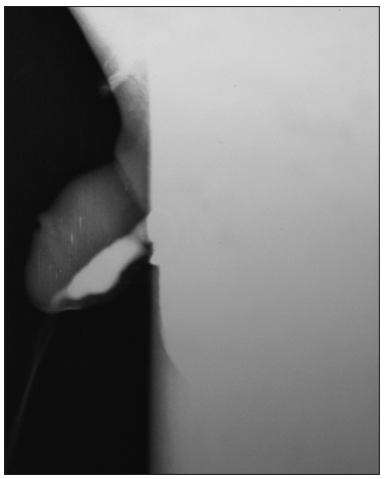
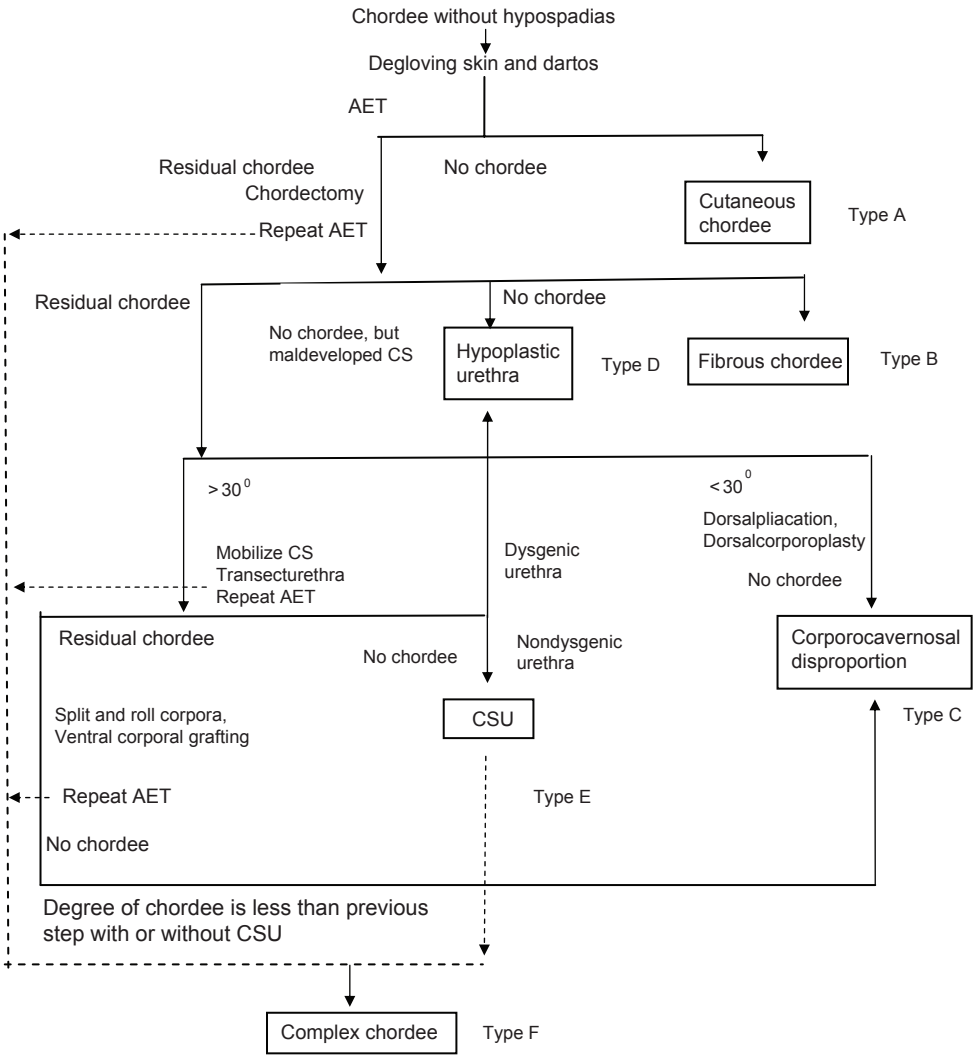


Figure 3: Follow-up voiding cystourethrogram showing meatal stenosis



Flow chart 1: Algorithm for the management and classification of chordee without hypospadias

or alternatively preservation of all the layers around the local dysgenic distal urethra.[2,4,12] But few authors proposed that, if the meatus is normal, urethra should not be divided to correct ventral curvature of penis, instead transposition graft may be used to bridge the gap.[1-3]

We performed single stage surgery for CWH, as patients may not turn up for further surgery, which is a common situation in our setup. We also believe that even if there is a higher chance of secondary procedures, approximately two-third of vascular tissue is preserved for subsequent procedures.[6] The only pre-requisite for a one-stage repair is the appropriate dorsal hood foreskin for a preputial-based island flap and adequate penile length (suggested by girth/length > 10thpercentile for age).[6] But, as there is scant literature on single or multi staged surgery for CWH, it is difficult to comment upon the suitability of one or other technique. A prospective study in this regard may answer it.

The CSU is believed to be exceeding rare entity. Devine saw only two cases in his experience.[2,8] In CSU there is lack of development of the epithelium, corpus spongiosum, buck's, and dartos fascias.[8,9] We also encountered only three case of CSU (two in type E and one in type F) over 11 years of period. We had unusually high rate of dysgenic tethered urethra (65.38%, 17/26 patients), which needed urethral reconstruction. In these patients, complication rate was 52.9% (9/17), which was comparable to Donnahoo *et al.* type I reconstructed urethra (50%).[2] We had higher UCF rate 26.9% (7/26) compared to 5-8% in recent series.[3-10] Development of fistula depends on several factor, including opposing suture lines of neourethra and of skin closure, distal obstruction because of metal stenosis, urethral stricture, turbulent urine flow (because of diverticulum), impaired local vascularity, wound infections, epithleal tracking and use of barrier layers.[14-16] In our study high rate of wound infection, associated meatal stenosis were responsible for higher fistula rate.[14,15] Further, the size of fistula was (>2 mm), and all were coronal thus they usually fails to close spontaneously.[14,15]

It was proved by Shanker *et al.* (2002) that recurrence of UCF does not depend on the type of urethroplasty initially performed.[16,17] Timing of UCF repair was also not suppose to be responsible for fistula recurrence, as in literature it is well mentioned that resolution of tissue edema and inflammation takes maximum of 6 months.[14-16]

The overall success rate in our patients (65.6%) with single stage operation being comparable to other authors (Scuderi *et al.*, Yun-MunTang); while lower than Donnahoo *et al.*, (92%).[2,3,18] It was probably due to higher proportion of dysgenic urethra (65.38%) in our study compared to 7% in Donnahoo's study. Additionally, Donnahoo *et al.*, excluded hypoplastic urethra from their study assuming it to be a variant of hypospadias; while we encountered 53.8% of hypoplastic urethra during repair of CWH.[2,3,18] Unusually high rate of hypoplastic urethra may be merely a co-incidence or population bias the authors' couldn't definitely comment the reason behind. But, in view of ongoing debate whether hypoplastic urethra should consider as a variant of hypospadias, if the meatus of dysplastic urethra is located at the tip of glans? The authors' opinion is to include it in type D of authors' operative CWH classification.

In this study the authors have focused on the management and classification of CWH and dysplastic urethra/fibrous chordee were identified on peroperative gross anatomical findings. Although, the histological diagnosis not going to change the management. But, the same can be done for the patohogenesis of CWH.

To conclude, because urethral tethering is not always the cause of CWH, a step-wise approach is the best way to manage it. Most of these cases have hypoplastic urethra, which makes the management of this entity difficult. Hence, meticulous tissue handling and urethral reconstruction may give good and promising results. The simplified operative classification proposed by authors may help in better management of this entity.

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