International Journal of Medicine Research
ISSN: 2455-7404; Impact Factor: RJIF 5.42
Received: 04-05-2019; Accepted: 07-06-2019
www.medicinesjournal.com
Volume 4; Issue 3; July 2019; Page No. 57-62

Genitoplasty in the Exstrophy Epispadias Complex (EEC) a retrospective study about 75 cases

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Abstract

Purpose: The aim of our study is to focus on recent techniques of genitoplasty and to analyse the long term results and complications.

Material and Methods: We retrospectively reviewed the files of 75 patients who underwent Modified cantwell Ransley epispadias repair for from 2003 to 2018. kelly procedure was performed in 17 cases.

Results: Particularly focusing on postoperative anatomical and functional complications; 16% of patients had urethral fistulas; 15% had spraying micturition; short appearance penile occured in 20%; Some other patients presented horizontal penile in 22%, inaesthetic appearence 18%, penile curvatum 5%, imperfect meatus 45%, and skin excess 27%.

Conclusion: The recent techniques of genital repair seem to have satisfactory results on the cosmetic aspect of the genitals, although none of these techniques is free from the occurrence of complications. the Surgical techniques continue to advance in the treatment of this urological congenital anomaly.

Keywords: epispadias- genitoplasty - complications

Introduction

The exstrophy-epispadias complex (EEC) is a rare congenital malformation with a wide spectrum of clinical features.

The incidence of this entity which has been estimated torange between 1 in 10,000 and 1 in 50,000 live births is rare, but it presents a significant physical, functional, social, sexual and psychological burden for the patients and their families [1].

The treatment of the epispadias has always been a real challenge in pediatric urology and urology [2].

Improved surgical techniques have led to successful functional and cosmetic reconstruction in the majority of children [3].

Although there is some controversy about the technique and the timing of the reconstruction, the preservation of the upper part and continence are possible with most treatment regimens [4].

Male adolescents consider genitals with a bizarre appearance with a small enlarged penis to be a more important psychosocial problem than incontinence, and therefore every effort should be made to restore the penis to normal [5].

The repair of epispadias remains a challenging operation even in expert hands. The incidence is rare and repair should stay in specialized hands to provide the patient the best possible outcome. Different techniques and modifications to these repairs have been developed by various centers. Each group has their unique experience and feels passionate about their approach. Controversies in surgical approach are common in major birth defects and reflect the intense research and dedication that individual groups devote to the subject. It is a healthy debate that promises to advance the understanding of the disease and improve the surgical outcome of patients [6].

Materiels and Methods

Data were collected retrospectively from the medical records of a consecutive series.

All of 75 patients underwent epispadias repair at our center during the period 2003 to 2018, using the Modified Cantwell Ransey technique.

Kelly procedure was performed in 17 patients All of them were operated by the same personal surgeon.

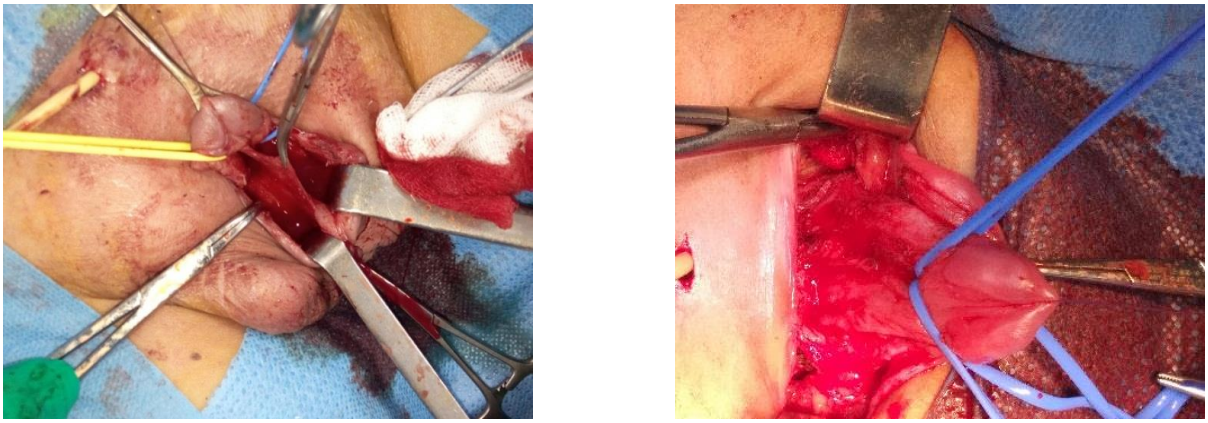
Modified Cantwell Ransley technique

A circumferential incision was performed under the corona, followed by complete degloving of the penile skin preserving the submucosal tissue attached to the urethral plate.

The mesentery of the urethral plate is approached ventrally in the middle of the penile shaft, and it is dissected off the corporeal bodies. (Figure 1)

The dissection of the urethral plate is advanced down to the penile base and distally to the level of the tip of the penile glans where the glanular wings are dissected.

leaving the distal most 0.5 cm attached to the glans. Care must be taken to preserve both the laterally placed urethral blood supply and the neurovascular bundles (NVB) (Figure 2).



a b

Fig 1: dissection the urethral plate from the corpora

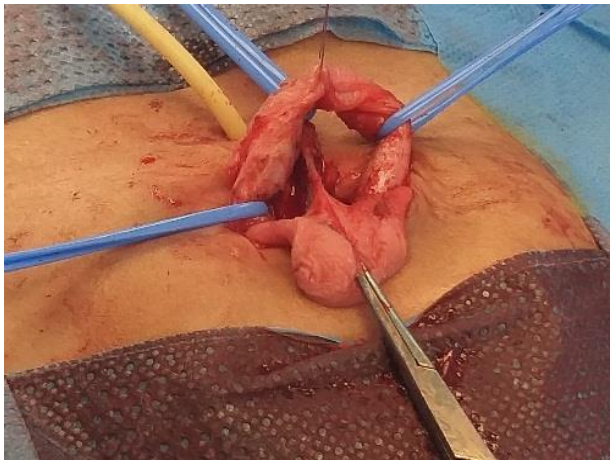


Fig 2: Completing dissection with leaving the distal 0.5 cm attached to the glans

Significant penile lengthening is achieved by completely detaching the insertion of the corpora cavernosa from the ischiopubic rami: kelly procedure.

The periosteum is incised in an elliptical fashion where the corpora cavernosa joined the inferior pubic ramus. We then elevate the periosteum off the bone in an attempt to gain greater mobility of the corpora cavernosa.

A limited dissection inside Alcock's canal is also performed (Figure 3 a and b)

Fig 3a: The periosteum is incised in an elliptical fashion where the corpora cavernosa joined the inferior pubic ramus.

Fig 3b: The periosteum and the attached corpora cavenosa are elevated off the bone in an attempt to gain greater mobility of the corpora cavernosa

Fig 3c: Lengthening the penile after kelly procedure

The urethra is tubularized over a urethral catheter with a running absorbable suture (Figure 4).

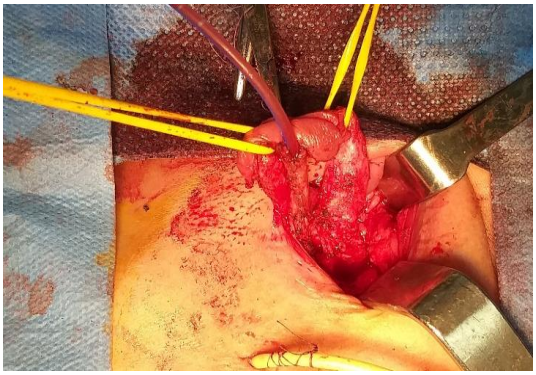


Fig 4: The urethra is tubularized over a urethral catheter

The glanular urethra and the meatus were fashioned with the IPGAM(incorporated glanuloplasty advancement meatal) maneuver, which involved a deep vertical incision in the distal urethral plate followed by transverse closure with 3 absorbable sutures creating a ventral urethral meatus.
The reconstructed urethra is then mobilized ventrally under the anti clockwise rotated
corporeal bodies (Figure 5).
The corporeal bodies are brought together, with serial stitches (4 or 5), without caverno-cavernosal anastomosis (Figure 6).

The glansplasty is completed,

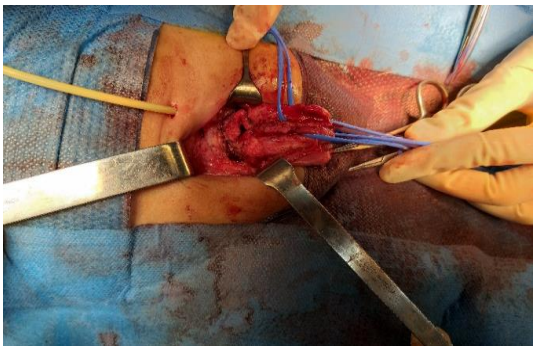


Fig 5: Anti clockwise rotated corporeal bodies

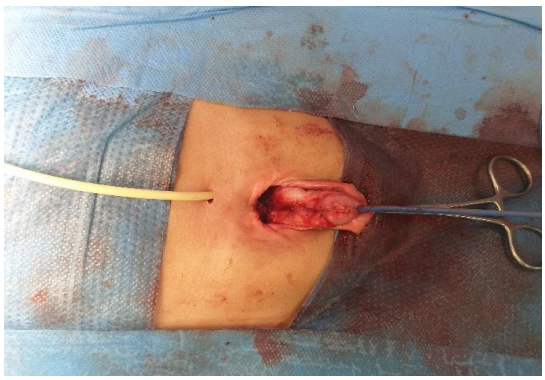


Fig 6: The corporeal bodies are brought together and glansplasty is completed

Skin coverage is achieved with ventral penile skin using reverse Byars flaps (Figure 7) and a longitudinal dorsal suture line closed by interrupted absorbable sutures.

Fig 7: Skin coverage is achieved with ventral penile skin using reverse Byars flaps

Fig 8: The final appearence after surgery.

Results
All of patients underwent epispadias repair during the study period, using the Modified Cantwell Ransley. kelly procedure is associated in 17 patients.
At a mean follow up of 5 years, 70% of patients achieved a good cosmetic, anatomical and functional result (Figure 9);
The complications are dominated by urethral fistulas at the level of the penis and imperfect meatus.fistulas were encountered in 16%; Imperfected meatus were presented in 45%, who needed meatal revision.
The complication rate was also slightly higher in the postpubertal (60%) than in the prepubertal (40%) patients;
5% of patients had a persistant dorsal curvatum;
the short appearance penile was occured in 20% of patients,including those who undewent kelly procedure; most of them continues to complain that the penis is unacceptably short, and asked for penile increasing length in the postpubertal.
15% presented spraying micturition; and 12% had dysuria; 5% presented urethral diverticulum, due to misalignment of the neourethra.
Three cases presented lithiasis, operated succesfully.

Cosmetic outcomes
78 % of the boys had a penis that was inclined downward (Figure 10), horizontally in 22%; skin excess was finded in 27% cases. And 18 % of patients presented inaesthetic appearance of penile, (Table 1)

Fig 9: satisfactory cosmetic and anatomical result in bladder plasty procedure

Fig 10: inclined downward penis

Table 1: Results of our study

Results	Rates
Fistulas	16%
Imperfect meatus	45%
Dorsal curvatum	5%
Spraying micturition	15%
Dysuria	12%
Urethral diverticulum	5%
lithiasis	5%
Cosmetic outcomes	
Inclined downward penis	78%
Horizontal penis	22%
Short appearance penis	20%
Inaesthetic appearance	18%
Skin excess	27%

Discussion

Epispadias has been considered as the least severe defect of the exstrophy-epispadias complex(EEC); however, the treatment of this anomaly is far from easy and trivial. The key concerns that need to be addressed during reconstruction
of penis and urethra, so as to ensure a functionally and cosmetically acceptable penis are correction of dorsal chordee, creation of a straight urethra, glandular
reconstruction, maintenance of erectile function, creation of urinary continence, satisfactory appearance and penile skin cover [7].

Despite improvement in techniques, no single approach has been considered the gold standard and epispadias repair remains a surgical challenge [8].

Epispadias surgery may be quite complex and requires an experienced team. Male epispadias patients are best served when treated at a center staffed by surgeons who have extensive surgical experience in managing epispadias cases [9].

Important for reconstruction of the penis in EEC is the unique presence of two completely separated corpora cavernosa without any vascular anastomosis and the completely isolated neurovascular bundles running on the corporal outside. Regardless of the type of epispadias repair, the four following key issues must be addressed to ensure a functional and cosmetically acceptable penis [10]

1. Correction of dorsal chordee
2. Urethral reconstruction for micturition and semen
3. Transport
4. Glandular reconstruction
5. Penile skin closure.

Modified Cantwell-Ransley technique

The most popular modern approach for surgical repair of epispadias is the Cantwell Ransley technique and its modifications [11].

Despite recent advancements in epispadias reconstruction techniques, complication rates are still significant [8].

Surer *et al*., Reporting on their 10 years' experience with the modified Cantwell–Ransley epispadias repair in 93 males (79 had classic bladder exstrophy and 14 had complete epispadias) had a 23% urethrocutaneous fistula rate in the immediate postoperative period and 19% at 3 months. The success rates following epispadias repairs vary depending on the degree of epispadias, age of the child/ patient, experience of the surgeon and the presence of previously operated tissues [7].

Similarly, Baird *et al.*, evaluated and updated the long term results of using this technique on 129 boys (97 had classic bladder exstrophy and 32 complete epispadias). Urethrocutaneous fistulae were noted in 16% and 33% after primary and repeat urethral repair, respectively [7]. in our study, the fistula rate is 20%, Is similar to those reported in literature.

Outcomes are significantly impacted by the quality and quantity of the penile skin and urethral plate available [12].

Several remedies have been suggested so as to reduce the fistula rate, including the use of tunica vaginalis pedicled wrap primarily along with the repair [13].

Pippi Salle *et al*., described a technical modification using a ventral penile skin flap, which facilitated dorsal skin closure, improved cosmesis and eliminated chordee [14].

In the 11 cases operated by them, all patients had an uneventful course after surgery and no patient developed recurrence of chordee or fistula.

Frequently, the authors use intramuscular and topical testosterone and they believe to reduce the incidence of fistula formation and other complications [11, 12].
A series of 75 boys [5]; (60 with bladder exstrophy and 15 with isolated epispadias) underwent repair of Cantwell-Ransley epispadias over a period of 6 years.
Primary repair was performed in 58 boys (45 with exstrophy and 13 with epispadias), and a secondary repair was performed after a non-surgical closure. successful in 17 boys.
All patients had a horizontal or standing pendulum penis, during a mean follow-up of 28 months. The incidence of urethrocutaneous fistulas in the immediate postoperative state was 21% and 15% at 3 months. Two patients developed urethral stricture at the proximal anastomosis and 4 patients had minor cutaneous separation of the dorsal closure of the penile skin [5].
In our study, 20% of patients had horizontal penis, and 5% had recurvated penis.

Most of them had inclined downward penile
Dysuria occured in 10% of patients, 5% developped urethral diverticulum.
Subsequently; The staged approach to bladder exstrophy reconstruction includes the bladder neck reconstruction, is usually performed at 4 to 5 years of age or when age is appropriate for toilet training and bladder of adequate capacity [15].
Since a few years, in our institution, we started to modify our attitude, bladder neck narrowing is performed during epispadias repair.
More than 10 patients underwent this procedure and results seem to be satisfactory.they acquired partially dryness (less than 20 minutes), and may increase bladder capacity and improve Bladder neck reconstruction continence results.

Kelly procedure
Male adolescents consider the odd appearing genitalia with a short widened penis to be a greater psychosocial problem than incontinence [16].
The most common deficiencies of the penis that the patients complain of the short appearance of the penis; because of the wide separation of the crural attachments, the prominent dorsal chordee, and the shortened urethral groove [17].
The Kelly procedure for patients with bladder exstrophy and epispadias is a technically demanding surgery that requires the surgeon to pay a great deal of attention to the penile neurovascular supply. The operation can be unforgiving, causing significant and irreversible morbidity with even minor vascular compromise [18].
The operation is based on the use of the hidden portion of the corpora cavernosa to provide additional length to the visible penile shaft. The corpora, with their vital structures, are mobilized and displaced forward and medially to lie together in the midline. Sufficient length of nerves and blood vessels is achieved by freeing the neurovascular pedicle from the fascial tunnel back to the ischial spine [19].
Detaching the corporal cavernosa from the pubic bones at the subperiosteal level allows for a safe distance to be maintained from the pudendal nerve at all times. It illustrates its relationship to the corpora cavernosa at the superior medial aspect, at which it is at greatest risk for injury. If a surgeon performs the dissection inferiorly and laterally, the corpora cavernosa can be safely detached from the ischiopubic ramus, and injury to the pudendal vessels and nerve can be avoided [18].
Some length can be gained by dissection from the body of the pubis, however it may not be useful to continue dissection posterior to the junction with the inferior pubic rami. Kelly achieved good lengthening in 9/11 children with complete detachment of the corpora from the inferior pubic rami, however, follow-up was not reported.
Such a procedure could lead to damage to the blood supply reaching the corpora through Alcock's canal [17].
All techniques of epispadias repair have in common that they require a very meticulous dissection in the anatomical layers using magnification glass in order to maintain the blood and nerve supply of the individual structures to avoid erectile dysfunction and corporal atrophy [20].
Berrettini *et al*. were previously able to show that with careful deep dissection of the corpora cavernosa, irreversible impairment of erectile function could bereduced, and even prevented [21].
In their series, parents of children who underwent the procedure reported that their children continued to have erections after surgery in all cases.
They did, however, report that two out of their nine patients experienced ischemic injuries to the glans, with glans loss following the Kelly procedure in those cases. Given this fact, that over 22% of the patients had some sort of vascular injury to the glans, it is not unreasonable to infer that the vascular pedicle may be tenuous and that patients may be at increased risk for developing significant vascular compromise during certain aspects of the radical soft tissue dissection.
The highest risk is during the dissection that is usually performed at the base of the epispadiac penis, it is for this reason that many pediatric urologists, even experienced ones, have hesitated in offering this surgery, which can augment the length of the penis.
In our serie, 17 patients underwent Kelly procedure,
No glans ischemic complication.
Patients in our serie, who underwent penile lengthening achieved adequate lengthening, compared to others. Traction on the corporal cavernosa has made us gain length, but once released, the penis seems to regain its anatomical memory.
Undoubtedly, Kelly procedure remains an attractive technique theoretically, but we don’t feel this procedure have impressive final results.
So we noticed that despite the obvious lengthening of the penis intraoperatively, this gain in length does not remain stable after corrective surgery.

Conclusion
The epispadias is a severe abnormality, especially when associated with bladder exstrophy. The Modified Cantwell-Ransley technique with kelly procedure allows reconstruction with good cosmetic, anatomical and functional results in most patients.
However, postoperative complications that are sometimes serious may develop. Although failure can occur even in the most experienced hands, the Choice of surgical technique may play a role in the overall outcome; however, the choice of an experienced surgeon over an occasional one may have a bigger impact.

Acknowledegemnts
I would like to express my special thanks of gratitude to my Professor Mohamed Amine
Bouhafs for their able guidance and support.
I thank all authors for theit efforts and contributions.

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