

The exstrophy–epispadias complex: is aesthetic appearance important?

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OBJECTIVE

To show the relevance of cosmetic appearance in the adequate treatment of patients with exstrophy–epispadias complex (EEC), and to indicate that surgery by experienced teams can improve the long-term treatment forecast and the patient's body image.

PATIENTS AND METHODS

From 1978 to 2002, 71 patients diagnosed with EEC were treated in the authors' institution; 24 (aged 2–23 years) were selected to undergo different surgical procedures. The criterion for surgery considered interviews conducted by the psychology team with the parents and children. The plastic surgery and paediatric urology teams carried out the procedures jointly; the follow-up was 0.33–7 years.

RESULTS

Five female patients and six male had abdominoplasty to treat multiple scars; eight had intermittent catheterization conduits repositioned from the right iliac fossa to the umbilicus. Six female patients had plastic surgery of the external genitalia and three had a broad mobilization of the urogenital sinus. Thirteen male patients had a small penis and had the corpora cavernosa fully mobilized and the penis reconstructed. Five female patients and one male had anterior osteotomy. One patient with no left testis had it replaced and one patient with uterine prolapse had the uterus fixed to the posterior abdominal wall. Six patients had a second procedure, in two because the outcome of the initial operation was poor and in the others to complement the initial treatment. In all but one patient there was an improvement in the objective criteria, e.g. school absences,

difficulty in establishing long-lasting social relationships and refusal to participate in sports activities. However, none of the patients would attempt sexual intercourse.

CONCLUSIONS

Body image, self-esteem, sexuality, sexual function and fertility are deemed crucial by adolescents; in patients with EEC customised surgical procedures can give a satisfactory aesthetic outcome, and be a further reason for adequately following occasional urinary complications and renal function, to avoid loss to follow-up.

KEYWORDS

bladder exstrophy, epispadias, cosmetic appearance.

INTRODUCTION

Children with the exstrophy–epispadias complex (EEC) pose a great challenge to the multidisciplinary teams caring for them. These children almost invariably undergo many surgical procedures, the outcome of which sometimes fails to fully resolve serious problems, e.g. achieving urinary continence and fashioning external genitalia capable of adequate sexual intercourse. The primary aims are to close the bladder plate, and then reconstruct the external genitalia and fashion mechanisms capable of rendering such children continent, while preserving their upper urinary tract function [1].

The body image and emotional development of these patients can be seriously compromised by both their genital deformity and reconstructive surgery. When assessing their results the medical teams invariably consider the initial severity of the

malformation and current treatment possibilities, failing to consider that as these children become aware of their body image they may consider themselves disabled, with sexually inadequate genitalia. The long-term medical follow-up, with several annual visits, and the inevitable comparison with their peers and possibly 'normal' siblings, usually cause their opinion of the treatment outcome to differ from that of their doctors, particularly when the latter report a 'good outcome'. Thus we report a series of patients with EEC who had different surgical procedures, aiming to assess the cosmetic aspects of the malformation.

PATIENTS AND METHODS

Between 1978 and 2002, 71 patients diagnosed with EEC were treated at the authors' institution; 24 (eight female and 16 male, age 2–23 years), who the psychology

team had been following regularly, were selected after an interview in which the patients or their family members claimed to be deeply interested in having their body appearance changed cosmetically, and were assigned to different surgical procedures. The interview considered variables such as school absence, difficulty in establishing long-lasting social relationships, systematic refusal to participate in sports activities with other children, or any other situation in which the child would be forced to undress in front of peers [2]. After surgery all the patients were again interviewed every 6 months, and had a bimonthly psychological follow-up to assess the effect of surgery on all the variables evaluated.

Because of the good outcome in the first patients we decided to also use surgery in nine patients in whom an aesthetic change indicated that a cosmetic problem was inevitable in the future. The plastic surgery

FIG. 1. A patient undergoing abdominoplasty, repositioning of the urinary catheterization conduit and plastic surgery of the external genitalia; **a,b**, before and **c,d** after surgery.



and paediatric urology teams jointly performed the procedures.

The most common cosmetic problems were an absent or too low umbilicus, abdominal scars and stomas requiring interventions with flap rotation and repositioning, hairless mons venus, causing the girls to feel embarrassed about undressing in front of peers, bifid clitoris (the correction of which was never fully possible), anteriorization of the introitus of the vagina (requiring extensive mobilization of the urogenital sinus), full or partial uterine prolapse and, in boys, a short and broad penis with a pronounced dorsal curve. The patients were followed for 0.33–7 years.

RESULTS

All patients had normal renal function and were already continent when they underwent surgery. Five females and six males had abdominoplasty to treat multiple scars; one of the boys had two abdominoplasties and two patients, who had previously had other cosmetic procedures, were re-operated and had another abdominoplasty. All these patients and their parents claimed to be satisfied with the outcome.

Eight patients had intermittent-catheterization conduits on the right iliac

fossa and were dissatisfied with their aesthetic appearance. The conduits were repositioned in a new umbilicus, fashioned according to the technique described by Sumfest and Mitchell [3] (Fig. 1). One of such patient, because of a poor surgical outcome, had further surgery 3 years later.

Six female patients had plastic surgery of the external genitalia and three had broad mobilization of the urogenital sinus (Figs 2 and 3). Although they have had no sexual relationship yet the results were considered satisfactory both by the patients and their parents.

Thirteen male patients had a small penis that also had a marked dorsal chordee; the corpora cavernosa was fully mobilized [4–6] and the penis was reconstructed. In two patients the penis was covered with a perineal region flap (Fig. 4) and in three with a skin graft from the inguinal region (Fig. 5). One of the patients having the corpora cavernosa fully mobilized had complete scar retraction and required further surgery. As in the girls, the result was deemed satisfactory by both the patients and their parents.

Five female patients and one male had the pubic symphysis widened and had anterior osteotomy with their iliac bone borders externally fixed. One patient with no left testis

had it replaced and one patient with uterine prolapse had the uterus fixed to the posterior abdominal wall (Fig. 2).

Six patients had a second procedure, in two because the outcome of the initial operation was poor, and in the others to complement the initial treatment.

In all but one patient there was an improvement in the objective variables, e.g. school absences, difficulty in establishing long-lasting social relationships and refusal to take part in sports activities. However, none of the patients reported wishing to attempt sexual relationships; the patients' data are summarized in Table 1.

DISCUSSION

The EEC is an uncommon malformation involving disorders in the lower abdominal wall, pelvic floor and anatomical disorders of the external genitalia and hip [1,7]. Changes in neural innervation, collagen and smooth muscle of the bladder wall were also reported recently [8]. The usual treatment involves a staged approach, starting by closing the bladder plate, with a bilateral iliac osteotomy in the first 48 h of life, if possible [1,8].

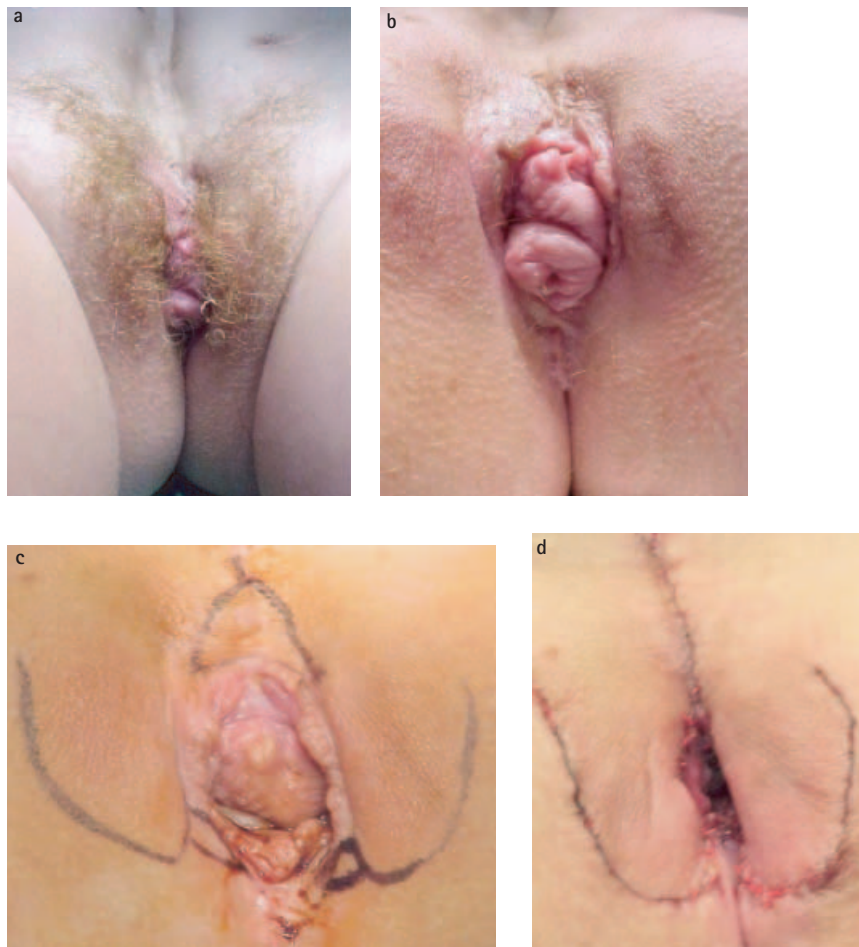


FIG. 2.

A patient undergoing abdominoplasty, mobilization of the urogenital sinus, plastic surgery of the external genitalia and uterine fixation; **a,b**, before and **c,d** after surgery.

The requirement for pelvic osteotomy was considered controversial until it is confirmed that there is at least a 12° rotation of the posterior pelvic segment, an $\approx 18^\circ$ rotation of the anterior segment, and a significant acetabular retroversion. Some authors even recommend that double osteotomy, anterior and posterior, be used, as complications from omitting the osteotomy might imply dire consequences for the child's future development [8–11]. Thus in the first phase it is recommended that the neonate should be assessed with CT to determine the appropriate course of the osteotomy.

Later treatment is directed to render the patient continent, which often implies creating urinary reservoirs using different intestinal segments, and constructing conduits for intermittent catheterization [6,12]. Many such children develop recurrent UTIs, often worsened by ubiquitous VUR or caused by an inadequate bladder incapable of storing urine at low pressure. Although the

external genitalia are already treated in the initial surgery, it is not unusual to find a shortened phallus in boys (Fig. 4) and a bizarre appearance of the external genitalia in girls (Fig. 2), with widening of the pubic symphysis.

The follow-up by the present psychology team showed that after the treatment of patients with EEC has achieved its objectives of urinary continence, protection of the upper urinary tract and reconstructing the external genitalia, it is crucial that attention is given to the cosmetic appearance. In significantly many patients this aspect has a major effect, particularly when they become teenagers. As noted by Gearhart *et al.* [1] it is almost redundant to say that currently, with easy access to the Internet, the physician's decision should consider the relatives' opinion and particularly that of the patients. The treatment possibilities currently available to these children should consider their subsequent quality of life [2,13,14].

When girls enter puberty, even when they have had iliac osteotomies whose initial result is deemed adequate, there can be new widening of the pubic symphysis. This gives the external genitalia an inadequate cosmetic appearance, as it almost always augments the median scar, limiting hair growth in the central part of the mount of Venus (Fig. 2). The external rotation of the hip leads to an inadequate positioning of the urogenital sinus, weakness of the pelvic muscles and occasionally uterine prolapse, as in one of the present patients [15]. In such cases full mobilization of the urogenital sinus, with a possible new iliac osteotomy when required, repositions the internal genital organs into the abdominal cavity, giving a better cosmetic result and more potential for adequate and satisfactory sexual intercourse.

The aesthetic aspects, often also compromised by hypertrophic scars, can have dramatic effects on an adolescent. In these cases surgical procedures, even when

FIG. 3. A patient undergoing abdominoplasty and plastic surgery of the external genitalia; *a,b*, before and *c,d* after surgery.



TABLE 1 Patients with exstrophy-epispadias complex undergoing plastic surgery

No./sex	Surgery	Age at surgery, years	Reoperation	Age at reoperation, years
1/F	UGS + O	3		
2/F	GE + O	5		
3/F	GE	6		
4/F	Ab + GE + R	10		
5/F	Ab + GE + O + R	11		
6/F	Ab + GE + O + R + Ut	13	GE	16
7/F	Ab + GE + O + U + UGS	14		
8/F	Ab + U + UGS	15		
9/M	P	5		
10/M	P + SG	5		
11/M	P + R + U	6		
12/M	P + Ur	6		
13/M	P + SF	8		
14/M	R	8	Ab + P	13
15/M	P + SF	11		
16/M	P	12		
17/M	P + R + T + U	15	P	16
18/M	P + O + Ur	16		
19/M	Ab + U + Ur	16		
20/M	P + R + U	16	P + SG	21
21/M	P	17	Ab + P + R	20
22/M	Ab + P + R + SG + U	19	Ab + U	22
23/M	Ab + U	19		
24/M	P	20		

Ab, abdominoplasty; SG, skin graft; GE, external genitalia reconstruction; T, testicular prosthesis; O, anterior osteotomy; U, umbilicoplasty; P, penile reconstruction; UGS, urogenital sinus, total mobilization; R, repositioning of catheterization conduit; Ur, urethroplasty; SF, skin flap; Ut, uterus fixation.

relatively complex, if performed by multidisciplinary teams, can change the patient's behaviour and even be associated with improving the compliance with the clinical follow-up, which will probably be lifelong.

Male patients almost always present with a short penis, usually wider than expected for their age, caused by corpora cavernosa that are too short and usually rotated, and with a major dorsal curvature [1,4,5]. In such cases a new broad and full mobilization of the corpora cavernosa usually implies an absence of adequate skin to cover them and, as in the girls, another iliac osteotomy may create conditions for the re-approximation of the symphysis. Placing skin grafts to cover the penis (by the plastic surgery team, and therefore used to the special care required in such surgery) gave surprisingly good results. In all cases there was a significant gain in penile length, with major changes to the boys' body image. Such aesthetic aspects have not been adequately emphasized previously, and thus when asking boys about their penile appearance the inadequate phallus should be considered as possibly causing greater dissatisfaction than urinary incontinence [16,17].

To adolescents, their body image, self-esteem, sexuality, sexual function and fertility are



FIG. 4.

A patient undergoing full mobilization of the corpora cavernosa and skin graft interposition, all views during surgery.

crucially important [1,4,13,15,18]. We consider that the important advances in technique have made children with EEC more demanding about their body image, experiencing the same distress and anxiety as their unaffected counterparts, and probably with normal or even aroused libido from the embarrassment of exposing their genitals to others [5]. In such cases customised surgical procedures may lead to an aesthetically satisfactory reconstruction, and further motivate them to comply with the monitoring of possible renal and urinary complications, avoiding loss to follow-up.

Thus a rational approach for patients with EEC is initial surgery in the first days of life and iliac osteotomy regardless of the age of the child. Even with the first 48 h of life osteotomy causes the possible posterior retraction of the pubic rami to be much less marked, thus preventing the corpora

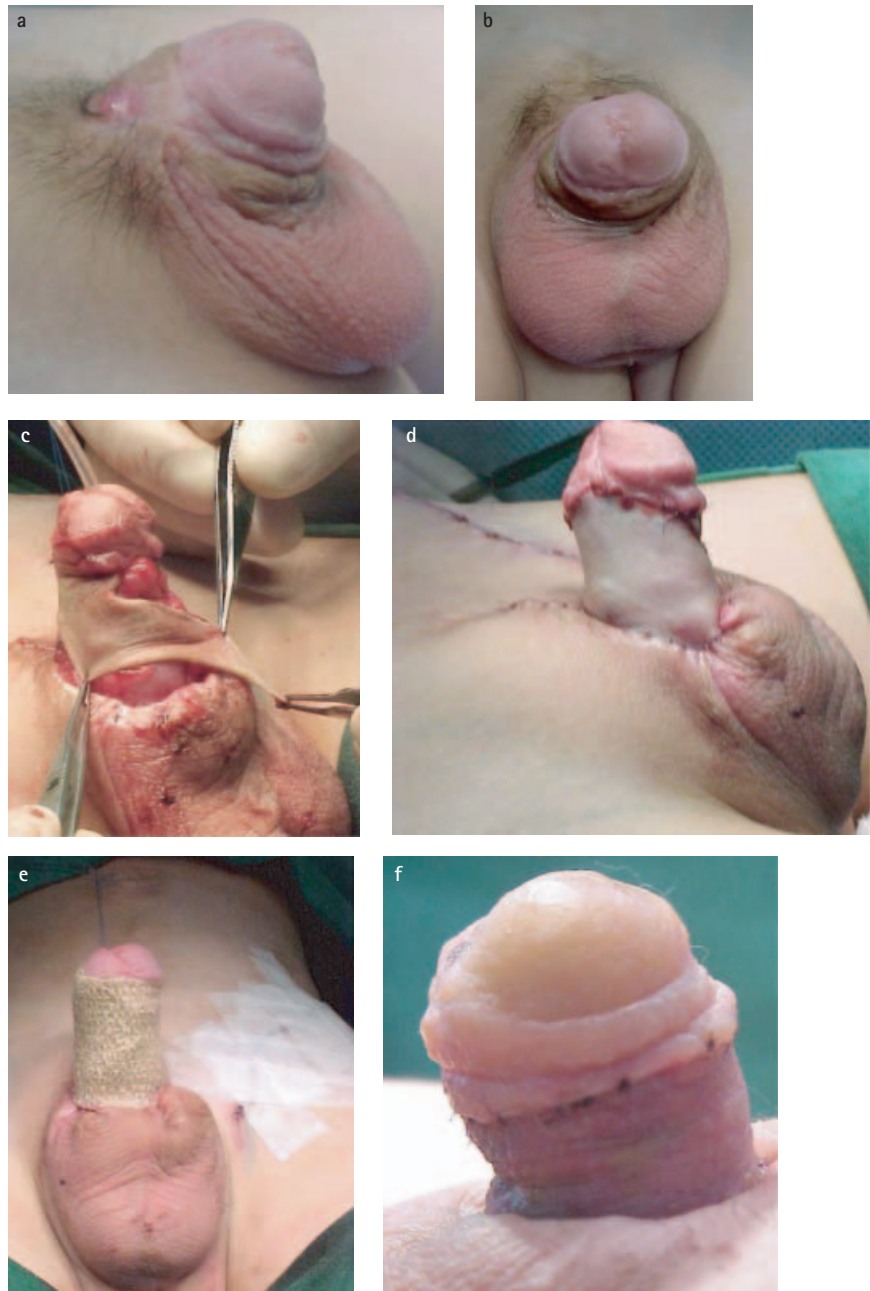
cavernosa from sinking into the pubic fat in boys, and a bifid labia majora in girls. An osteotomy also provides greater stability for abdominal wall synthesis, preventing the formation of an inadequate umbilicus and reducing the risk of dehiscence, which usually implies loss of the vesical plate, particularly when it is poorly developed. The penis should be fully mobilized in the initial surgery, which has often implied obtaining a short urethra, not usually exceeding the base of the penis. We chose to maintain the urethra at the base of the penis and to treat it a few months later as though it were a proximal hypospadias, often using foreskin or oral mucosal grafts. We have not mobilized the urogenital sinus in the initial surgery in girls, but make initial attempts to improve the aesthetic aspect of their external genitalia and of the clitoris.

Urinary continence should be attained before school age, with renal function carefully

monitored. Many children will subsequently require construction of continent urinary diversions, which implies the use of intermittent catheterization and care to avoid the formation of stones and resulting UTIs. Perhaps because of the irregularity of the reconstructed urethra in boys and the increased sensitivity in the urethra of girls we have been unable to persuade children who use intermittent catheterization to do so urethrally; in almost all cases a catheterization conduit had to be constructed.

The multidisciplinary team presents all these aspects of treatment to the families, as they see the children at least bimonthly, emphasizing a regular follow-up as the most suitable way of treating such a complex disease. From the start of treatment the families should have psychological support, because present surgical techniques enable

FIG. 5. Full mobilization of the corpora cavernosa and use of inguinal skin to cover the penis: **a,b**, before, **c,d** during and **e,f**, 2 months after surgery.



renal function and urinary continence to be adequately preserved, and families should be given the chance to openly discuss the inevitable frustrations that these children have with an aesthetically inadequate body.

There is as yet no optimum treatment for a malformation that involves the abdomen, hip, perineum and genitalia. The next major advance might be tissue engineering and cellular therapeutic

techniques, to provide a full reconstruction of both genitourinary organs and musculoskeletal tissues.

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CONFLICT OF INTEREST

None declared.

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Abbreviations: EEC, exstrophy-epispadias complex.