Feminizing genitoplasty in congenital adrenal hyperplasia: the value of urogenital sinus mobilization

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Background/purpose **Congenital adrenal hyperplasia is a common cause of ambiguous genitalia in female individuals. These patients require feminizing surgery aiming at reconstruction of feminine external genitalia with normal function. Total urogenital mobilization was developed to avoid dissection in the common wall between the vagina and urethra. This study aims at evaluating the outcome of feminizing genitoplasty after the use of urogenital mobilization.**

Patient and methods **Fourteen female patients with congenital adrenal hyperplasia were managed during the period from July 2007 to April 2011. They were assessed clinically according to the Prader score. The common channel anatomy was studied by a flush retrograde genitogram. Clitoroplasty, vaginoplasty, and labioplasty were performed. The common sinus was managed by urogenital mobilization. Postoperative cosmetic outcome was evaluated according to the criteria described by Creighton and colleagues. Follow-up included clinical assessment of the urethral and vaginal openings and clinical evaluation of the continence for urine.**

Results **A genitogram has a sensitivity of 64.3% in estimating the length of the common channel. The length of common channel is not related to the degree of virilization. Good cosmetic outcome was reported in 71.4% of cases. All postoperative complications were minor and managed by simple maneuvers. All patients had good urinary control after urogenital mobilization.**

Conclusion **Urogenital sinus mobilization is a valuable tool in the early one-stage feminizing surgery with few technical problems, good cosmetic outcome, low incidence of complications, and good urinary continence. *Ann Pediatr Surg* 8:111–115 © 2012 Annals of Pediatric Surgery.**

Annals of Pediatric Surgery 2012, **8**:111–115

Keywords: congenital adrenal hyperplasia, feminizing genitoplasty, urogenital sinus mobilization

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Received 3 September 2011 **accepted** 16 July 2012

Introduction

Congenital adrenal hyperplasia (CAH) is an autosomal recessive disorder with an incidence of 1/10 000–1/15 000 live births [1]. It results from inherited defects in one of the five enzymatic steps required for the biosynthesis of cortisol from cholesterol [2]. The virilizing forms (simple virilizing or salt-wasting types) are characterized by impaired cortisol biosynthesis that leads to the accumulation of steroid intermediates proximal to the deficient enzyme [3]. Newborn girls with classical virilizing CAH present clinically with variable degrees of genital ambiguity in the form of clitoromegaly, labial fusion, fusion of the vagina and urethra into a common urogenital sinus, and lack of palpable gonads [4].

CAH is responsible for about one-half of patients presenting with ambiguous genitalia, and these children should be given a female sex assignment and undergo feminine reconstruction because they have the potential for both normal sexual function and fertility [5]. The aims of feminizing surgery are to create female-appearing external genitalia, to provide the benefit a normal psychosexual development, and to create a functional vagina, to allow menstruation and sexual activity [6].

Total urogenital mobilization (TUM) is an operative technique developed by Peña [7] for the treatment of cloacal anomalies to avoid the difficult dissection through the common wall between the vagina and the urethra, hence avoiding the related complications. With the use of the perineal approach, the application of this technique had been extended to conditions other than anorectal malformations, especially intersex conditions [8]. Partial urogenital mobilization utilizes the surgical principles of TUM, but the dissection of the common channel is limited to being distal to the pubourethral ligament [9].

The purpose of this study is to evaluate the outcome of feminizing genitoplasty in correction of genital abnormalities associated with CAH after the use of urogenital sinus mobilization as a part of the operation.

Patients and methods

This study included 14 female patients with CAH managed in the Pediatric Surgery Unit, Mansoura University Children Hospital, during the period from July 2007 to April 2011. Thorough genital examination was performed for all patients to assess the degree of virilization according to the Prader scoring system [10]. Diagnostic workup included estimating the level of blood electrolytes to detect cases of the salt-losing type. High plasma levels of 17-hydroxyprogestrone were indicative of 21-hydroxylase deficiency, whereas raised plasma 11-deoxicortisol levels were indicative of 11β-hydroxylase

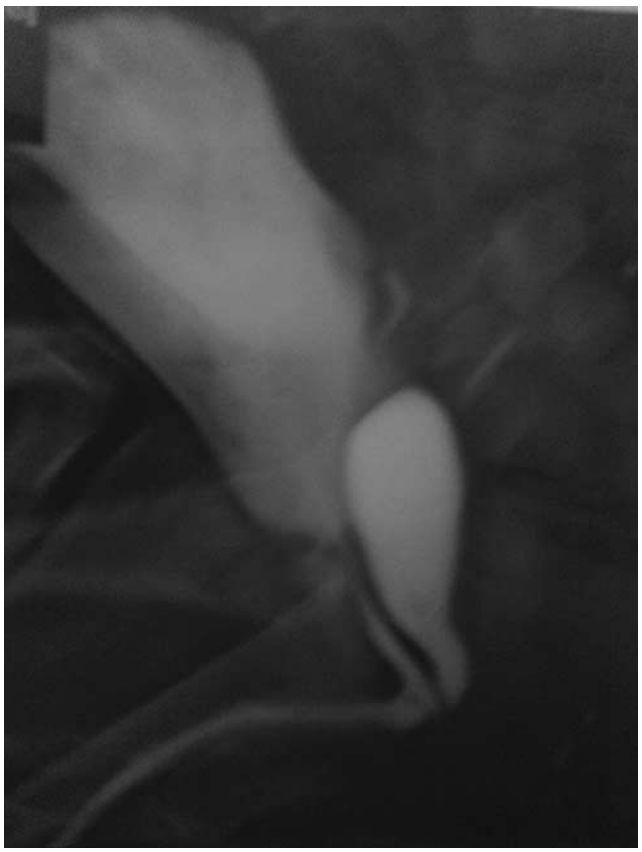
1687-4137 © 2012 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000418527.14707.bc

deficiency. The genotypic sex was confirmed by chromosomal analysis. An abdominopelvic ultrasound was performed to evaluate internal sex organs. All patients had undergone a retrograde flush genitogram. The site of confluence was determined and the lengths of the common channel and the proximal urethra were estimated (Figs 1 and 2). Genitourinary panendoscopy was performed only in three patients in whom evaluation of anatomy by imaging was difficult.

Informed consent was obtained from the parents and the approval of the research Ethics Committee of Mansoura University was obtained. Appropriate steroid replacement was prescribed by pediatric endocrinologists, with doubling of the dose on the morning of the operation. The child was placed in an exaggerated dorsal lithotomy position for the perineal operative approach. A posterior inverted U-shaped skin flap was prepared. The apex of the flap touched the posterior rim of the urogenital sinus (UGS) opening and the incision was extended circumscribing the sinus.

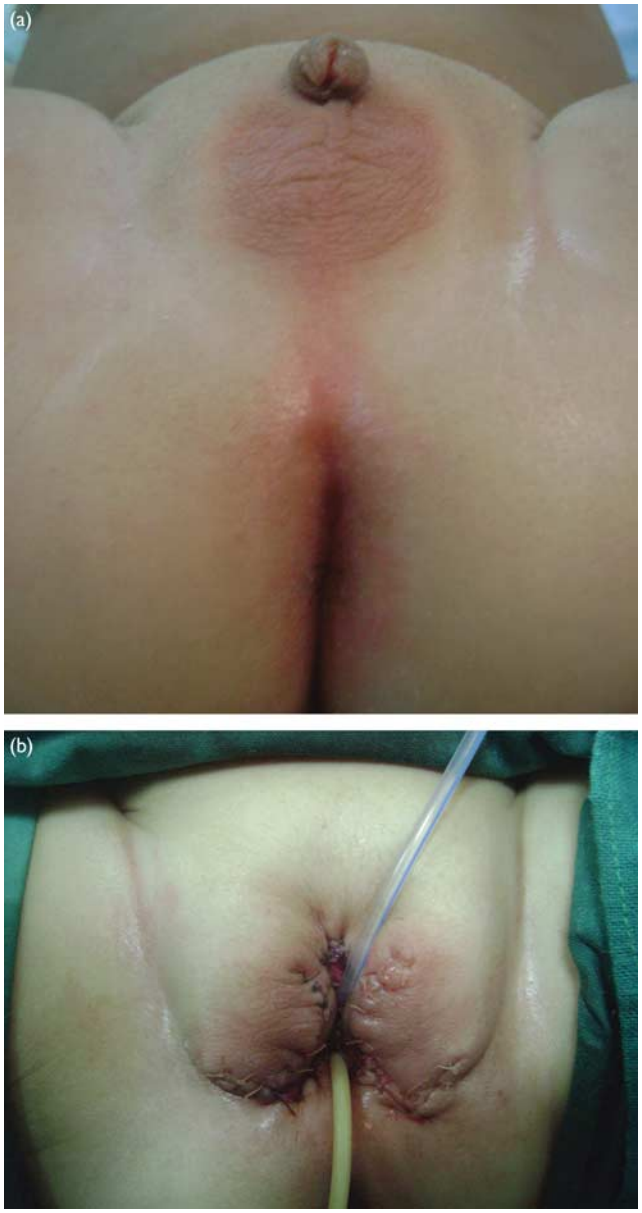
Clitoral reduction was performed in nine patients using the Kogan technique [11] of subtunical excision of the two corpora leaving the glans with the intact neurovascular bundle. Clitoral recession was performed in the remaining five patients with less severe clitoral enlargement. The common UGS was managed by TUM as described by Peña [7] using the modified perineal approach recommended by Ludwikowski *et al.* [8]. However, division of the pubourethral ligament was needed in only three patients and partial urogenital mobilization as described by Rink *et al.* [9] was sufficient for the other 11 patients. The tissues of the UGS were used according to the modifications recommended by Rink *et al.* [12]. In three patients the UGS was divided dorsally and the tissue of the UGS was reflected ventrally to reconstruct the anterior vaginal wall. In the other 11 patients, the UGS was divided ventrally to be used as a mucosa lined vestibule. The tissues of the common UGS were not discarded in any case. After suturing the flaps to the vagina, the skin of the labia majora was advanced inferiorly until it reached the base of the inverted U-flap. The split clitoral skin flaps were used to construct the labia minora.

Fig. 1



Retrograde flush genitogram.

Fig. 2



(a) Preoperative image of a female patient with congenital adrenal hyperplasia, (b) feminine external genitalia after feminizing genitoplasty.

A Foley's catheter was left in the urethra and removed after 5 days. Oral feeding was initiated 2–3 h post-operatively. An appropriate analgesic is administered for pain control in the immediate postoperative period. Broad spectrum antibiotics were administered for the whole period of catheterization.

The follow-up period ranged from 5 to 38 months. The postoperative cosmetic and anatomic results were evaluated according to criteria described by Creighton *et al.* [13]. These cosmetic assessment criteria included the following: genital proportions and symmetry, pubic hair distribution, clitoral hood size and shape, glans clitoris shape and prominence, clitoral body size and prominence, labial positions and proportions, vaginal introital position and appearance, and genital skin quality. Three categories for cosmesis were defined: good (no abnormal features), satisfactory (up to two minor abnormalities), and poor (three or more abnormal features). Follow-up also included clinical assessment of the site and size of the urethral and vaginal openings and clinical evaluation of urinary continence, which was defined as complete dryness during intervals between spontaneous voiding with an average interval of 3 h.

Results

The age of patients ranged from 6 months to 8 years (96 months) with a mean age of 33 months; the largest group consisted of patients of the youngest age group (< 1 year), which included five patients (35.7%; Table 1). Eight patients (57.1%) scored III and five patients (35.7%) scored IV. The most severe virilization with a Prader score of V was reported in one patient (7.2%). Only one patient was diagnosed as having CAH because of 3-β-hydroxysteroid dehydrogenase deficiency, whereas all other patients were diagnosed as having CAH because of 21-hydroxylase deficiency (92.8%). The latter group included five patients of the simple virilizing type (38.5%) and eight patient of salt-wasting type (61.5%).

Pelvic ultrasound was successful in detecting internal female sex organs in 10 patients (71.4%). A retrograde flush genitogram delineated both the urethra and vagina in all patients. However, the radiological lengths of the common channel measured by the genitogram were compared with the operative lengths and they were found to be the same in nine patients (64.3% sensitivity).

The mean operative time was 201 min (range: 175–235 min). The lengths of common channels ranged from 1.5 to 3.5 cm, with a mean length of 2.2 cm. Half of the patients belong to the middle group having common channels of intermediate length (2–3 cm). Only two patients (14.3%) had a long channel (> 3 cm), whereas five patients (35.7%) had a short channel (< 2 cm). Blood transfusion of 20 ml/kg was required in three patients only.

Table 1 Age of patients at the time of urogenital mobilization

Age group	*N* (%)
<1 year	5 (35.7%)
1–3 years	4 (28.6%)
3–5 years	2 (14.3%)
>5 years	3 (21.4%)

Table 2 Relationship between length of common channel and degree of virilization assessed using the Prader score

	Operative length of common channel (cm)			
	<2	2–3	>3	Total
Prader score				
III				
Count	3	5	0	8
% Within the Prader score	37.5	62.5	0	100.0
IV				
Count	2	2	1	5
% Within the Prader score	40.0	40.0	20.0	100.0
V				
Count	0	0	1	1
% Within the Prader score	0	0	100.0	100.0
Total				
Count	5	7	2	14
% Within the Prader score	35.7	50	14.3	100.0

The relationship between the length of the common channel and the degree of virilization assessed using the Prader score was studied (Table 2). No patient with a Prader score of III had a long common channel, whereas the only patient who scored V had a long common channel. Nevertheless, 62.5% of patients with score III had intermediate length versus only 40% of patients who scored IV. Accordingly, it was found that the common channel are not necessarily longer in highly virilized patients ($P = 0.106$).

Cosmetic and anatomic outcome of external genitalia after TUM was evaluated according to criteria described by Creighton *et al.* [13]. No abnormalities were reported in the clitoral position or in the position of vaginal introitus. Abnormalities in the labia minora were most commonly reported (three patients). Overall evaluation revealed good cosmetic outcome in 10 patients (71.4%; Table 3). Postoperative complications were reported in four patients (28.6%). However, all were mild complications managed by simple maneuvers (Table 4). Wound infection occurred in one patient and was resolved using systemic and topical antibiotics. Ischemia of labia minora flaps occurred in one patient and was managed using topical applications that promote wound healing. Flaps left to heal resulting in poor labia minora. Late vaginal stenosis occurred in two patients after 6–9 months and was treated effectively by just regular vaginal dilatation. Nine patients were at an appropriate age for evaluation of urinary continence. All were able to keep dry intervals between spontaneous voiding and were considered clinically continent. No patient developed postoperative urinary symptoms that required further investigations.

Discussion

CAH is a continuum of disorders that affect patients throughout their lives [14]. In this study, 92.8% of cases of CAH were found to be due to 21-hydroxylase deficiency and 7.2% of cases were due to 11-β-hydroxylase deficiency. This high incidence of 21-hydroxylase deficiency

Table 3 Cosmetic and anatomic outcome according to criteria described by Creighton and colleagues

Cosmetic outcome	*N* (%)
Clitoris size	
Normal	12 (85.7%)
Large	1 (7.15%)
Small	1 (7.15%)
Absent	0 (0%)
Clitoris position	
Normal	14 (100%)
Low	0 (0%)
Absent	0 (0%)
Vaginal introitus	
Normal	12 (85.7%)
Small (stenosis)	2 (14.3%)
Interoital position	
Normal	14 (100%)
Not surrounded by labia	0 (0%)
Labia majora	
Normal	12 (85.7%)
Small	0 (0%)
Scrotalized	2 (14.3%)
Labia minora	
Normal	11 (78.6%)
Small	2 (14.3%)
Poor	1 (7.1%)
Absent	0 (0%)
Overall	
Good	10 (71.4%)
Satisfactory	2 (14.3%)
Poor	2 (14.3%)

Table 4 Postoperative complications

Complications	*N* (%)
Wound infection	1 (7.15%)
Flaps ischemia	1 (7.15%)
Urethral complications	0 (0%)
Vaginal stenosis	2 (14.3%)
Urinary incontinence	0 (0%)
Total complications	4 (28.6%)

is similar to the 95% incidence reported in the other studies [15,16].

The abdominal sonography technique is found to have a sensitivity of 100% in identification of internal female genitalia [17]. In our study, a lower ultrasound sensitivity of 71.4% was reported. This low sensitivity may be related to the experience of the radiologist and the quality of the ultrasound machine. However, the sensitivity obtained by us is very close to the sensitivity of 77.8% reported by Elhalaby [18]. Retrograde genitography was found to have a sensitivity of 64.3% in determining the length of the common channel. This is close to the results reported by other studies (60% by Chertin *et al.* [17], 55.6% by Elhalaby [18] and 72% by Vanderbrink *et al.* [19]). Hence, endoscopy is considered to be a crucial step in genitoplasty, as it identifies the anatomy of the common channel and enables insertion of a Foley or Fogarty catheter into the vagina through the communication to the UGS [20].

In the present study, the policy of early one-stage correction was adopted. Graziano *et al.* [21] agreed that early intervention is associated with better compliance with dilatation, a reduction in the parents' concerns regarding their child, and a lesser psychological impact on the child at an early age. In contrast, Alizai *et al.* [22] support postponing vaginoplasty until puberty claiming the availability of supple and genital skin and avoidance of revision after early surgery. However, we agree with Elhalaby [18] on the ease of handling the vagina in young children.

The length of the urethra proximal to the confluence with the vagina is more important than the length of the common channel, with a high risk of incontinence when the urethra is too short [23,24]. This study found that the relationship between the length of the common channel and the degree of virilization is not statistically significant ($P = 0.106$). However, Escala *et al.* [25] found a direct relationship between the length of common sinus and the Prader score.

The criteria described by Creighton *et al.* [13] were used to evaluate the cosmetic and anatomic outcome of external genitalia. We reported poor cosmetic outcome in two patients (14.3%). Similar results were reported by Elhalaby [18] (11.1%) and Bragga *et al.* [26] (12.5%) using the same Creighton criteria. However, Creighton *et al.* [13] through a more objective assessment found poor cosmetic outcome in 41% of patients. These large differences may be because the variability in assessment of minor abnormalities among different surgeons. Hence, a more objective scoring system is needed for more accurate evaluation.

Vaginal stenosis is the most common complication after vaginoplasty [27]. Before the era of TUM, there was a high incidence of vaginal stenosis of up to 78.5% and most patients required further vaginal reconstructive procedures [28]. A much lower incidence of vaginal stenosis was reported on using TUM, ranging from 0% [23,29] to 9.1% [24]. This is obviously because TUM decreased the incidence of vaginal ischemia. We reported vaginal stenosis in two patients (14.3%) and they needed only vaginal dilatation. Similar to other studies [23,29], we reported 0% incidence of urethrovaginal fistula or hypospadic urethral meatus. Furthermore, postoperative retraction of the urethra did not occur in any patient, and an obvious and easily accessible urethral meatus was obtained in all patients. Overall, on short-term follow-up, no patient needed any revision surgery after TUM and all reported complications that could be managed through minor interventions.

Studies on urinary function after TUM revealed well-preserved continence mechanisms postoperatively [23,30]. However, Rink *et al.* [9] discussed the concerns associated with the very proximal, circumferential dissection above the pubourethral ligament and described the technique of partial urogenital mobilization that limits the chance of injury to the urinary sphincter, clitoris, and their innervation. This latter technique was used in most of the patients in this study, and all evaluated patients were found to be clinically continent.

Conclusion

UGS mobilization is a valuable tool in feminizing surgery in many aspects. It solves technical problems and facilitates dissection, maintains adequate blood supply to the urethra and vagina, minimizes blood loss and

requirements for blood transfusion, and has good cosmetic outcome with low incidence of complications and without affecting mechanisms of urinary continence.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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