

Assessment of Urogenital Sinus Mobilization and Feminizing Genitoplasty in Children with Congenital Adrenal hyperplasia

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Original
Article

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ABSTRACT

Background: The objective of surgical intervention in females with urogenital sinus (UGS) and congenital adrenal hyperplasia (CAH) is to maintain bladder function, restore normal appearance and function of the external genitalia, and enable normal sexual and reproductive development throughout adulthood. We aimed to assess female patients who underwent UGS mobilization as part of feminizing genitoplasty treatment.

Patients and Methods: This study utilized a retrospective descriptive database of 20 patients who underwent feminizing genitoplasty and UGS mobilization in a single session for patients with CAH.

Results: The mean age for the 20 participants included in the study was 27.3±10.55 months. Of these patients, six (30%) were classified as Prader III, whereas 14 (70%) were Prader IV. The length of the UGS was determined to be less than 2.0 cm in two (10%), 2.0 cm in five (25%), 2.5 cm in eight (40%), and less than or equal to 3.0 cm in five (25%) cases. The vaginal orifice was retracted with abnormal introitus in two (10%) cases and was seated in a normal site in the posterior portion of the vestibule in 18 (90%) patients. The labia minora had a satisfactory cosmetic appearance and looked relatively normal in all the studied patients. The labia majora had a satisfactory cosmetic appearance in 18 (90%) and retracted in two (10%) cases. Residual urine occurred in three (15%) patients. There was a complete absence of urinary dribbling in all patients. In our study, a urogenital fistula was observed in one patient, accounting for 5% of the cases. However, no cases of recto-vaginal fistula were identified.

Conclusion: Feminizing genitoplasty combined with UGS mobilization is an effective method for patients with CAH in early childhood. Furthermore, good cosmetic genitalia appearance and urinary continence can be achieved in the majority of patients.

Key Words: Children; congenital adrenal hyperplasia; feminizing genitoplasty; urogenital sinus mobilization.

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INTRODUCTION

Congenital adrenal hyperplasia (CAH) is the etiology of the adrenogenital syndrome. CAH is the primary cause of ambiguous genitalia, accounting for 1 in 15 000 live births^[1]. The underlying etiology of CAH is an autosomal recessive condition where the enzyme responsible for the steroid pathway fails, leading to a hormone deficiency despite normal Müllerian development. The cause of this insufficiency is typically a deficit in 21-hydroxylase, which accounts for 90% of cases. Nevertheless, the clinical presentations differ, which accounts for the variety of clinical presentations^[2].

The surgical repair of the external genitalia of girls with CAH and urogenital sinus (UGS) is a challenging process that requires maintaining bladder function and allowing for normal adult sexual and reproductive lives. The main parameters that determine reconstruction are the degree of virilization and the level of vaginal insertion^[3].

Feminizing genitoplasty is a well-established procedure used to achieve a desired esthetic and functional outcome by correcting the external genitalia and separating the genitalia from the urinary tract^[4]. There is an ongoing debate regarding the optimal timing for corrective surgery, whether it should be performed during early childhood or delayed until adolescence. This debate arises from

the dissatisfaction reported by certain adult patients who underwent genital reconstruction in early childhood, expressing dissatisfaction with the appearance of their new genitalia as well as their sexual experiences^[5].

The concept of feminizing childhood genital surgery has been a participant of debate for more than a decade. This is because there is concern that clitoral surgery may negatively impact an adult's sexual function. In addition, young girls may not necessarily require a functioning vagina^[6]. Currently, there is more focus on one-stage reconstruction^[7]. Pena concept of complete UGS mobilization (TUM) in CAH patients was applied by Rink *et al.*^[8]. Therefore, they experienced fewer problems and good cosmetic benefits. Despite achieving a favorable cosmetic outcome, the aggressive dissection of the bladder neck posed a significant risk of urine incontinence. This led to the eventual development of the notion of partial UGS mobilization (PUM) by Rink *et al.*^[9].

Therefore, our objective was to evaluate female patients undergoing feminizing genitoplasty in situations of CAH by managing the UGS mobilization, with a specific focus on vaginal caliber, introitus position, urine continence, genital appearance, and the evaluation of early surgical sequelae.

PATIENTS AND METHODS:

In this study, we conducted a retrospective analysis of the medical records of 20 patients who underwent feminizing genitoplasty using UGS mobilization for a genital condition associated with CAH between January 2019 and January 2023 in the Pediatric Surgery Unit at Minia University Hospital and Qena University Hospital (prior to the surgical procedures, the parents of the patients provided their informed written consent, as well as written consent to participate in the study, which involved an agreement to the data analysis recorded in patients' files. The Institutional Review Board (IRB), Faculty of Medicine, Minia University, has reviewed and approved the study procedures in accordance with the criteria set by the ethical committee (Approval NO: 1041/01/2023).

At the diagnostic stage, karyotyping was performed on all patients to verify their genotype sex, which was confirmed to be 46xx. In addition, abdominal ultrasound was performed for all cases to confirm the presence of female internal sex organs. Laparoscopy was performed in five cases when abdominal ultrasound was inconclusive in determining the presence or absence of female internal reproductive organs.

Furthermore, a retrograde genitogram was performed to evaluate the anatomy of the common UGS. The Prader classification was utilized to indicate the degree of virilization. A pediatric endocrinologist supervised the administration of medical care and monitoring. In

accordance with the endocrinologist/anesthetist protocol for CAH patients, all children with the condition got a "stress dose" steroid replacement during surgery.

Surgical procedure

Operative notes retrieved from patients' files

Prior to the surgical reconstruction, a cystoscopy was performed using the same procedure to assess the length of the common channel and the position of the urethrovaginal confluence. Two catheters were placed, one in the bladder and the other in the vagina, using endoscopic guidance. The length of the common channel was measured using endoscopy.

All patients had a single-stage surgical procedure; however, the specific type of intervention varied according to the level of vaginal confluence. Rebuilding the vaginal opening and severing the vagina from the urethra were typically included in the procedure. Every intervention was performed using a perineal approach while the patient was positioned in the dorsal lithotomy position.

Urogenital sinus mobilization

After following the UGS by a circumscribing incision, a 2 cm mucosal strip was defined and kept beneath the clitoris. The incision was then extended to the tip of an inverted perineal "U" incision by moving it longitudinally (toward the anus).

The posterior aspect of the UGS, in front of the rectum, was the site of the subsequent circumferential dissection. The dissection terminated before reaching the levator ani muscles and proceeded in an anterior direction till reaching the lower border of the pubic bone while protecting the pubourethral ligaments (PUL). An approximately 2–3.5 cm dissection was necessary to reach the urethrovaginal confluence. We continued dissection until reaching the posterior end of the catheter.

Vaginoplasty

Vaginoplasty involves the posterior splitting of the common channel until the urethrovaginal confluence is visible. This process continues until a vagina with a well-vascularized and normal-looking wall is achieved.

The posterior wall of the vagina was then separated from the rectum, releasing tension and enabling the vaginal opening to reach the perineum. Finally, it was sutured to a Fortunoff flap to provide a broad vaginal introitus. When the UGS was low (<2.5 cm), we generated a mucosal-lined vestibule by mobilizing the common channel until the urethrovaginal confluence reached the perineum and incised its distal half ventrally.

When there was a high UGS (>2.5 cm), the vagina was severed at the urethral junction. A minimal amount of dissection was performed to allow for the closure of the UGS, which then became a part of the urethra. The technique created a rim of vaginal tissue that could be anastomosed to the mucosal flap made using the distal portion of the UGS. The Passerini-Glazel^[10] flap was created by opening the UGS at 12 o'clock and inverting its mucosa until sufficient length was obtained to create the distal anterior vaginal wall. In order to avoid vaginal stenosis, the posterior wall of the vagina was extensively incised in the midline. Subsequently, the inverted U perineal flap (Fortunoff flap) was sutured to the apex of the posterior vaginal wall incision^[11]. By expanding the introitus, this method may have prevented vaginal stenosis and retraction.

Reduction clitoroplasty

In all cases, clitoral excision was performed using the method outlined by Kogan *et al.*^[12], which preserved the neurovascular bundle and moved the glans clitoris anatomically by securing it to the corporeal bodies' stump. Fifty percent of the patients had glandular reduction, which involved cutting a wedge out of the glans' core region.

Labial reconstruction

The labia minora was reconstructed using the dorsal phallic skin. In order to achieve a natural-looking reconstruction of the vestibule, V-Y incisions were performed following the scrotal folds, and Labia Majora flaps were created. The data collected during the follow-up were analyzed. Based on modified Creighton *et al.*^[13] criteria, genital symmetry, clitoral size, vaginal location, and genital skin quality were used to determine cosmetic results. Based on these criteria, the genital appearance was considered satisfactory if two or fewer matched the standards for esthetics and if all the requirements were fulfilled. Following a perineal examination, the vaginal diameter was measured by gently calibrating with the largest appropriate Hegar dilators. Urinary continence was assessed using a voiding diary and ultrasonography to measure the post-void residual in individuals who had received toilet training. In addition, a voiding cystourethrogram was performed 3 months later to evaluate the bladder's shape, length, and urethral diameter (Figs 1–7).

Parents satisfaction was assessed using a five-point Likert scale (1 being extremely unsatisfied and 5 being extremely satisfied)^[14]. The assessments were conducted by anesthesiologists who were not responsible for patient intraoperative management or block administration.

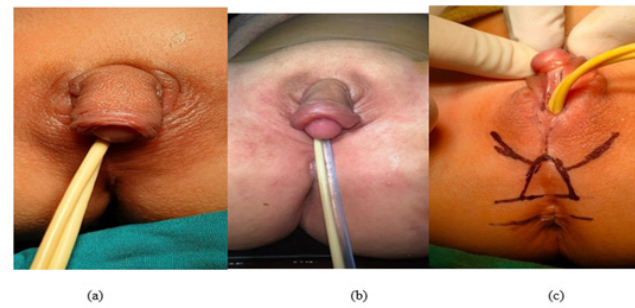


Fig. 1: (a, b) Preoperative photos after cystoscopic guided insertion of two catheters, one in the bladder and the other in the vagina, and (c) perineal skin flap (Fortunoff flap).

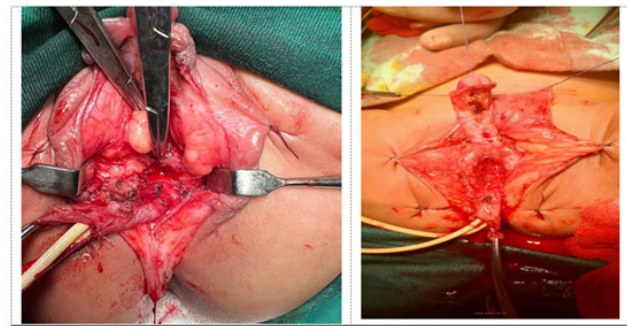


Fig. 2: Urogenital sinus mobilization.

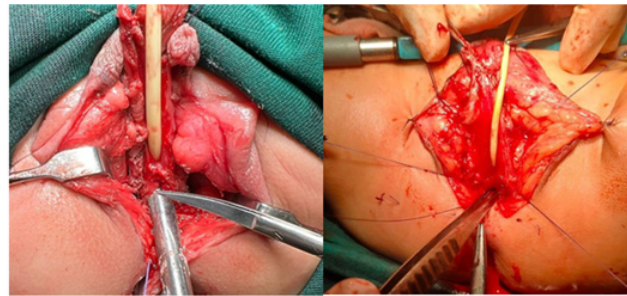


Fig. 3: Division of the posterior wall of the UGS, delivery of the vagina, and fixation with the perineal skin flap. UGS, urogenital sinus.

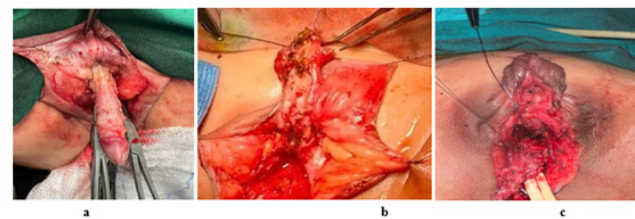


Fig. 4: (a, b) Excision of the corpus cavernosum on either side with preservation of the neurovascular bundle, (c) plication of the clitoris.

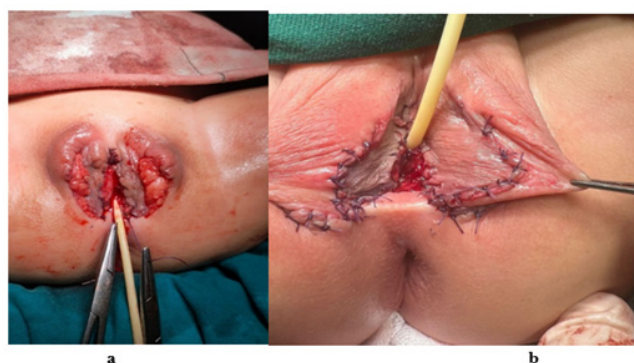


Fig. 5: Labioplasty (a) after division of the posterior wall of the UGS to do vaginoplasty, the phallus skin was used to do the labia minora, (b) V-Y plasty of the outer skin (previous scrotum) to do the labia majora. UGS, urogenital sinus.



Fig. 6: Feminizing genitoplasty at the end of the operation.



Fig. 7: A case 1 year after the operation.

Statistical analysis

Data statistical analysis was performed using IBM Inc. (Armonk, New York, USA). Quantitative variables were expressed using the mean and SD. Qualitative factors were expressed as percentages and frequencies.

RESULTS:

Table 1 displays the baseline characteristics of the studied patients, encompassing a total of 20 patients. Patients' mean age was 27.3 ± 10.55 months, with a mean follow-up duration of 18.65 ± 8.86 months.

Concerning the Prader classification, six (30%) patients were class III, and 14 (70%) were class IV. Regarding the length of UGS, of 20 children evaluated using cystoscopy, the urethrovaginal confluence from the perineal skin surface was determined to be less than 2.0 cm in two (10%), 2.0 cm in five (25%), 2.5 cm in eight (40%), and more than or equal to 3.0 cm in five (25%).

In this study, 16 (80%) patients underwent partial urogenital mobilization, whereas four (20%) underwent total urogenital mobilization. With respect to the postoperative complications, only residual urine occurred in three (15%) patients, 20 ml in two cases, and 50 ml in one case, as evidenced by abdominal and pelvic ultrasound. None of the cases experienced urine dribbling. A urogenital fistula was observed in one patient, accounting for 5% of the cases. This fistula closed on its own after a urethral catheter was inserted for 10 days. However, no cases of recto-vaginal fistula were observed in any of the patients, as confirmed by postoperative genitogram. No patient has experienced a urinary tract infection (Table 2).

Regarding the cosmetic results of the studied patients, the vaginal orifice was retracted with abnormal introitus in two (10%) cases and was located superficially in the posterior part of the vestibule with normal-looking vaginal introitus in 18 (90%) patients. The vaginal calibration had regular dilatation in 18 (90%) patients and was stenosed in two (10%) patients. Cases with vaginal stenosis required dilatation under general anesthesia, followed by a continuous dilatation regimen done by the mother. Clitoral head size was accepted and seated in the upper portion of the vestibule in 19 (95%) patients, and it was large in the follow-up period in one (5%) patients. The clitoris was covered by skin in three (15%) patients and was in good condition in 17 (85%) patients. The labia minora was cosmetically good in all the studied patients. The labia majora was cosmetically good in 18 (90%) patients, was retracted and surgically pulled in one (5%) patient, and was retracted and healed by dressing in one (5%) patient. The genitalia appearance was good in 15 (75%) patients and satisfactory in five (25%) patients, as shown in (Table 3).

Parents were extremely satisfied in 14 (70%) cases, with good satisfaction in five (25%) cases, and were unsatisfied in one (5%) case due to the retraction of the labia majora, which required correction and pulling under anesthesia (Table 4).

Table 1: Baseline characteristics of the studied patients

	Total (N=20)
Age (months)	27.3±10.55
Prader classification	
III	6 (30)
IV	14 (70)
Length of UGS (cm)	
<2 cm	2 (10)
2 cm	5 (25)
2.5 cm	8 (40)
≥3 cm	5 (25)

Data presented as mean±SD or frequency (%).
UGS, urogenital sinus.

Table 2: Postoperative complications of the studied patients

	Total (N=20)
Dribbling	0
Residual urine	3 (15)
Urogenital fistula	1 (5)
Recto-vaginal fistula	0

Data presented as frequency (%).

Table 3: Cosmetic results of the studied patients

	Total (N=20)
Vaginal orifice	
Retracted	2 (10)
Seated at the vestibule	18 (90)
Vaginal calibration	
Average (hegar 10)	18 (90)
Stenosed	2 (10)
Clitoral head size	
Accepted size	19 (95)
Large in the follow-up period	1 (5)
Clitoris size	
Small sized	3 (15)
With average size	17 (85)
Labia minora	
Cosmetically good	20 (100)
Labia majora	
Cosmetically good	18 (90)
Retracted and surgically pulled	1 (5)
Retracted and healed by dressing	1 (5)
Genitalia appearance	
Good	15 (75)
Satisfactory	5 (25)

Data presented as frequency (%).

Table 4: Parent satisfaction of the studied patients

	Total (N=20)
Extremely satisfied	14 (70)
Satisfied	5 (25)
Unsatisfied	1 (5)

Data presented as frequency (%).

DISCUSSION

The CAH procedure often includes clitoroplasty, labioplasty, urethroplasty, and vaginoplasty^[15]. Vaginal-urethral separation is a complex surgical procedure that requires advanced training to execute^[16]. Common complications following surgery include the recurrence of urethrovaginal fistula, vaginal or urethral stenosis, and urinary incontinence.

Total UGS mobilization was first documented to repair cloacas to aid reconstruction and lower the risk of complications such as urethrovaginal fistula^[17]. Subsequently, Rink *et al.*^[8] used a perineal-prone approach to apply this procedure to children with CAH.

This modification facilitated the restoration of the urethra and vagina, resulting in reduced surgical duration. In addition, it enabled the mobilization of the urethrovaginal confluence en bloc by moving the vaginal and urethral orifices to the desired locations. The whole mobilization procedure has raised concerns about postoperative urine incontinence due to the disruption of the PUL despite the reduced risk of urinary sphincter injury. Based on the currently available limited long-term follow-up data, these theoretical issues have not been determined to have any clinical significance. For instance, Kryger and González^[18] found that with complete urogenital mobilization, nine children with CAH maintained their urine continence. Unfortunately, the potential for long-term complications has not been thoroughly investigated. Considering the agreement between Oshiba *et al.*^[19], Farkas *et al.*^[20], and Passerini-Glazel^[10] that achieving optimal reconstruction without affecting the sphincteric mechanism is technically possible, regardless of the extent of dissection, we decided to preserve the PUL if it does not compromise surgical outcomes.

In a trial including 55 patients, Bailez *et al.*^[21] found that TUM provides good outcomes with a mix of several approaches. Ludwikowski *et al.*^[22] illustrated that TUM has been modified for DSD and is appropriate in cases when the UGS is more than or equal to 3 cm. It also minimizes the risk of urethral innervation injury and shortens surgical procedures because there is no need for a urethral and vaginal

separation. With a UGS less than or equal to 2–3 cm, it can be inferred that a non-intensive dissection and U-flap vaginoplasty would be adequate.

In contrast, Palmer *et al.*^[23] found no correlation between a TUM or PUM and patients' increased incontinence. According to Stites *et al.*^[24], after a TUM, patients experience severe urine incontinence, which is why they would instead use a PUM. None of the patients exhibited any symptoms of incontinence. The implementation of UGS mobilization is clearly a positive advancement in feminizing genitoplasty despite particular concerns related to continence.

Consistent with our findings, Farkas *et al.*^[20] and Passerini-Glazel^[10] demonstrated that protecting the sphincteric mechanism during the dissection is simple. Therefore, if the surgical outcomes are not compromised, we opt to preserve the PULs. In addition, our findings are compatible with those of Palmer *et al.*^[23], who found no differences in urine continence between PUM and TUM in UGS and cloacal abnormalities in their study on urinary incontinence.

It was found that four patients, which account for 20% of the total, received entire urogenital mobilization, whereas the remaining 16 patients, accounting for 80% of the total, experienced partial mobilization. In relation to postoperative complications, a total of three (15%) patients had residual urine, two cases with 20 ml, one case with 50 ml, and one (5%) patient with urogenital fistula. In our study, there were no cases of urinary incontinence, dribbling, or recto-vaginal fistulas, and no patients had urinary tract infections. By combining UGS mobilization with feminizing genitoplasty in a single procedure, it was possible to achieve good exposure. It enabled the reconstruction of the anterior vaginal wall by division of the mobilized UGS, making a wide mucosal-lined vestibule. These advantages were evident in patients' cosmetic outcomes. Out of the total number of patients, two (10%) patients had their vaginal opening retracted with abnormal vaginal introitus, whereas 18 (90%) had it seated at its typical location. In 18 (90%) individuals, the vaginal calibration exhibited regular dilatation, while in two (10%) patients, it was stenotic and needed dilatation under general anesthesia. In 19 (95%) of the patients, the clitoral head size was accepted and concealed in the vestibule, whereas in one (5%) of the patients, it was noticeable and enlarged in size during the follow-up period. In three (15%) of the patients, the clitoris was small in size, while in 17 (85%) of the patients, it was in good condition. The labia minora in every participant under study had good cosmetic results. Of all the patients, 90% had good cosmetic results with their labia majora, while 5% underwent surgical correction, and the remaining 5% had their labia retracted and treated

with a dressing until it healed. Out of the total of 20 patients, the genitalia appearance was deemed good in 15 (75%) individuals, whereas it was considered satisfactory in five (25%) individuals. In 14 (70%) cases, parents were extremely satisfied; in five (25%) cases, parents were satisfied; and in one (5%) cases, parents were unsatisfied. These results are consistent with Oshiba *et al.*^[19], who assessed the early results of PUM in feminizing genitoplasty in terms of vaginal caliber, urine continence, and genital appearance in females with CAH. It was found that 24% of the 19 patients had satisfactory cosmetic improvements, which amounts to six patients. The remaining 76% did not have satisfactory cosmetic changes. Out of the 24 participants, 96% had an adequate caliber mobilized vagina. Seven potty-trained kids had their bladder function evaluated. Only one patient had daytime incontinence, but after a few months, frequent voiding aided in the patient's recovery.

Our study included 20 children with a mean age of 27.3 ± 10.55 months. Early reconstruction is suitable for our society and accurately represents the psychological state of young girls and their parents. Furthermore, it was discovered that the technique used in the early stages contributed to mobilizing the UGS and performing genitoplasty in a single operation, resulting in satisfactory outcomes. Using the PUM technique at an early age was concomitant with Braga *et al.*^[25]. They operated on 24 children, with a mean age of 28 months. They observed both a satisfactory cosmetic outcome and separate vaginal and urethral orifices with good vaginal caliber. They also found no urinary tract infection or urinary incontinence in those toilette-trained girls.

Recent studies have brought attention to the potential long-term effects of early feminizing genitoplasty by disclosing infrequent cases of sex dysphoria and poor cosmetic outcomes in women who underwent surgery as infants^[26,27]. Certain authors have proposed that surgery be postponed until the patient demonstrates an interest in having their genitalia corrected.

According to Peter E. Clayton *et al.*^[28], delaying correction in DSD individuals may have negative consequences. The primary objective of Oshiba *et al.*'s^[19] study in early sex assignment is to lessen parental concern so young girls can develop greater self-worth and a stronger sense of femininity. Research revealed that most girls with CAH favored making an early repair decision at an early age^[29].

Our study's retrospective design has some limitations, such as the length of the UGS before surgery, the small number of infants who operated in more than one stage, the small sample size, limited tools to assess urinary continence, and the abbreviated

follow-up period. Larger cohort studies are required to evaluate the interaction of several factors, including age, virilization level, surgical expertise, and a specialized surgeon's commitment to feminizing genitoplasty.

CONCLUSION

Feminizing genitoplasty combined with UGS mobilization is an effective method for patients with CAH in early childhood. Good cosmetic genitalia appearance and urinary continence can be achieved in the majority of patients.

CONFLICT OF INTEREST

There are no conflicts of interest.

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