FEMINIZING GENITOPLASTY IN CLASSIC CONGENITAL ADRENAL HYPERPLASIA- EXPERIENCE AT KHYBER TEACHING HOSPITAL, PESHAWAR

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ABSTRACT

Objective: The aim of the study is to determine the outcomes of single stage feminizing Genitoplasty in patients with Congenital Adrenal Hyperplasia (CAH) in terms of surgical complications, parents' satisfaction and their determinants.

Material & Methods: In this retrospective chart review, 32 patients who underwent single stage Genitoplasty at Khyber teaching hospital between January 2015 to January 2019 were included. Patients with common channel longer than 2.5 cm and those operated elsewhere were excluded. Parents were interviewed retrospectively and data was collected. Ethical approval was granted by IREB of the institute. Outcome was called ''successful'' in patients with no complication at follow up. Parents and surgeon's satisfaction with cosmesis was recorded from evaluated pre-and postoperative photographs and separately labelled as "satisfied" or "not satisfied". Data was analyzed using SPSS version 22.

Results: Mean age of the patients at surgery was 9.71 ± 1.72 months (6-18 months). Twenty-one (65.25%) patients were in age group 6 to 12 months while 11 (34.37%) were in age group 13 to 18 months. The mean operative time was 180.22+22.41 minutes. No flap ischemia (0.0%), vesicovagnial/veginorectal (0.0%) or surgical site infection (0.0%) was observed in any patient. Urinary incontinence was seen in 1 (3.12%) patient, while urinary tract infections were observed in 3 (9.37%) patients. Two patients (6.25%) had vaginal stenosis at follow up calibration. Parents were satisfied with cosmetic outcome in 30 (93.8%) cases and surgeon in 29 (90.62%) patients. Success rate was found higher (i.e. 90.47% and 100.0%) in younger age groups and lower CAH grades ($P<0.05$), respectively.

Conclusion: The early follow up of females with CAH receiving single stage feminizing Genitoplasty have shown promising results in terms of low complication rates and high satisfaction rates of parents.

Key words: Feminizing Genitoplasty; Congenital Adrenal Hyperplasia; Atypical Genitalia.

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INTRODUCTION

Classical Congenital Adrenal Hyperplasia (CAH) is an autosomal recessive condition resulting from defective cortisol synthesis pathway[1]. The term "classic'' is specific for the commonest variant of CAH in female patients where there is deficiency of 21-hydroxylase enzyme and serum level of 17-hydroxyprogesterone is elevated. The reported incidence of CAH is 1 per 10,000 to 1 per 15,000 live births[2,3]. A spectrum of ambiguity is observed in external genitalia of genetically females bearing 46XX chromosome in classical CAH[4]. In increasing order of ambiguity, clitoromegaly, fused urogenital conduit with single external opening, labial fusion, fused vagina & urethra that opens into common urogenital sinus and impalpable gonads are seen[5]. The gender and genital ambiguity is a psychosocial trauma to both parents and children[6].

As the children retain the potential for normal sexual activity and fertility in classical CAH, the treatment is directed to attain anatomical and psychological female gender and for this, children undergo feminizing Genitoplasty after detailed diagnostic assessment and counselling[7,8]. This treatment includes clitoroplasty, vaginoplasty, relocation of vaginal orifice to normal perineal position and parting of vagina and urethra[9,1].

All these corrective surgeries for treatment of genital ambiguity are technically demanding, with variable outcome and were therefore not always done in one stage[11,12]. Required surgical skills, education of child and family and precise timing of surgery are not studied yet. This study presents the surgical outcomes experienced by

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our department in terms of parent's satisfaction, complications and their determinants in patients with CAH who underwent single stage Genitoplasty.

MATERIAL AND METHODS

We enrolled all 32 female patients with CAH and atypical genitalia operated over a duration of 4 years between January 2015 and January 2019 at Khyber teaching hospital, Peshawar. Data was retrieved from files and follow up findings from charts documented at 3 weeks follow up meeting. Pre operatively, at initial encounter, patients were categorized into 5 grades of CAH severity according to Prader Scale; grade 1: clitoral hypertrophy, grade 2: clitoral hypertrophy, urethral and vaginal orifices present, but very near, grade 3: clitoral hypertrophy, single urogenital orifices, posterior fusion of labia majora: grade 4: penile clitoris, perineoscrotal hypospadias, complete fusion of the labia majora: grade 5: complete masculinization (normal looking male genitalia) but no palpable testis. All the patients had received a single stage genitoplasty (clitoroplasty, vaginoplasty or combined procedure). Record of operative procedure and post-operative examination for complications and cosmesis at three weeks were reviewed retrospectively. Parents' satisfaction with cosmesis labelled as ''satisfied'' or ''not satisfied'' evaluated from comparison of pre-operative and post-operative pictures was recorded. Surgeon's satisfaction was determined in a similar way. Complications were assessed at 3 weeks follow up recorded findings on clinical examination. Complications and parents' satisfaction were compared for the severity and age of Genitoplasty to know if they were the determinants of the outcome parameters. Success was labelled to those patients who had no complications of surgery. Data was analyzed using SPSS version 22. For age of patients and duration of study, mean and SD were calculated while for grades of CAH, patient satisfaction and outcomes frequency and percentages were calculated. For comparing of variables chi-square test was applied and P value of equal or less than 0.05 was taken as significant.

RESULTS

In our study, out of 32 patients, 5 (15.62%) patients underwent clitoroplasty, 7 (21.87%) patients underwent vaginoplasty and 20 (62.5%) patients underwent combined procedure (clitoroplasty and vaginoplasty). Parents and treating pediatricians were satisfied with cosmetic outcome in 30 (93.8%) and 29 (9.62%) patients, respectively. The mean operative time was 180.22+22.41 minutes. Surgical site infection, flap necrosis, urethral complications were not observed in any patient (0.0%) after surgery. Postoperatively, vaginal stenosis was found in 2 (6.25%) patients. Vesicovagnial and rectovaginal fistula were not observed in any patient (0.0%). Urinary incontinence was seen in 1 (3.12%) patient, while urinary tract infections were observed in 3 (9.37%) patients. Mean hospital stay was 5.92 ± 3.41 days. None of the patient required re-do surgery and there was no mortality (0.0%). Success rate was 81.25%.

Characteristics of patients with CAH are shown in Table 1. Stratification of success rate with effect modifier i.e. age and CAH grades are shown in table 2 and 3. Par-

Table 1: Characteristics of patients with CAH (n=32).

Parameters		No. of patients (%)
Age (Months)	Mean ± SD	9.71 ± 1.72
	6 -12	21 (65.62%)
	13 - 18	11 (34.37%)
Associated congenital disorders		0 (0.0%)
Past history of Genitoplasty		0 (0.0%)
Grades of CAH	Grade I	3 (9.37%)
	Grade II	6 (18.75%)
	Grade III	13 (40.62%)
	Grade IV	10 (31.25%)
	Grade V	0 (0.0%)

Table 2: Stratification of data (Success Rate) with age (n=32).

Age (Months)	Success rate	
	Yes n(%)	Non(%)
6-12 (n=21)	16 (76.19%)	5 (23.80%)
13-18 (n=11)	10 (90.90%)	1 (3.12%)
Total	26 (81.25%)	6 (18.75%)
p-value*	0.0003**	

Table 3: Stratification of data (Success Rate) with CAH grades (n=32).

Grades of CAH	Success rate	
	Yes n(%)	Non(%)
Grade I (n=3)	2 (66.67%)	1 (33.33%)
Grade II (n=6)	5 (83.33%)	1 (16.67%)
Grade III (n=13)	12 (92.30%)	1 (7.69%)
Grade IV (n=10)	7 (70.0%)	3 (30.0%)
Grade V (n=0)	0 (0.0%)	0 (0.0%)
Total	26 (81.25%)	6 (18.75%)
p-value*	0.0001**	

Table 4: Parents & Surgeon's satisfaction.

Parameters		No. of patients (%)
Parents' satisfaction	Yes	30 (93.8%)
	No	2 (6.25%)
Surgeons' satisfaction	Yes	29 (90.62%)
	No	3 (9.37%)

Table 5: Early Operative outcome of feminizing Genitoplasty.

Operative outcomes	No. of patients (%)
Mean duration of surgery (Mean ± SD)	180.22+22.41 minutes
Flaps ischemia	0 (0.0%)
Urinary dribbling/retention	1 (3.12%)
Urethral complications	0 (0.0%)
Vaginal stenosis	2 (6.25%)
Surgical site infection	0 (0.0%)
Urinary tract infections	3 (9.37%)
Vesicovagnial fistula	0 (0.0%)
Rectovaginal fistula	0 (0.0%)
Re-do surgery	0 (0.0%)
Successful outcome	26 (81.25%)

ents and surgeon satisfaction are shown in table 4.

DISCUSSION

This study conducted on the single stage Genitoplasty included 32 patients with CAH. In a study by Almasri J [13] et al, the mean age of patients was 2.7 6 4.7 years. Another study by Shoeir HM [14] et al, the observed mean age was 33 months in a range of 6 months to 8 years. majority of patients i.e. 35.7% in younger age group (< 1 year) in this study. In our study, we did not encounter any other congenital anatomic disorder in any patient and none of the patients had undergone surgery for genital reconstruction elsewhere. In our study, majority of patients i.e. 40.62% had grade III CAH, followed by grade IV (31.25%), grade II (18.75%) and grade I (9.37%). Shoeir HM [14] et al, in his study showed that majority of patients i.e. 57.1% were grade III. There was no patient (0.0%) with grade V in our study, however in a study by Shoeir HM [14] et al, 7.2% patients were with grade V.

The mean operative time was 180.22+22.41 minutes in ours study. In another study by Shoeir HM [14] et al, the mean operative time was 201 min (range: 175–235min). Surgical site infection, flap necrosis, urethral complications were not observed in any of our patient after surgery. However, in the study by Shoeir HM [14] et al, wound infection and flap ischemia were observed in 7.15 % and 7.15% patients, respectively. Postoperatively, vaginal stenosis was found in 2 (6.25%) patients, in our study. In a study by Almasri J [13] et al, the vaginal stenosis was significantly high i.e. 27% of patients developed vaginal stenosis. In another study by Shoeir HM [14] et al, vaginal stenosis was reported in 14.3% patients. Vesicovagnial and rectovaginal fistula were not observed in any patient in our study. Urinary incontinence was observed in 1 (3.12%) patient, while urinary tract infections were found in 3 (9.37%) patients in our study in contrast to the study by Shoeir HM. Mean hospital stay was 5.92 ± 3.41 days, in our study. No patient required re-do reconstruction and there was no mortality in our study. Success rate was 81.25% in our study.

In our study, parents and surgeon were satisfied with cosmetic outcome in 30 (93.8%) and 29 (9.62%) patients, respectively In a study by Almasri J [13] et al, the patients and surgeon satisfaction were 79.4% and 71.8%, respectively.

In our study, we stratified the success rate with age and concluded that success rate was higher i.e. 90.47% in younger age group (6-12 months) as compared to older age group (13-18 months) i.e. 63.63% . The results were statistically significant (p=0.0003). We also stratified the success rate with grades of CAH and observed that the success rate is more i.e. 100.0% in lower grade (grade I & II) and less i.e. 92.30% and 50.0% in higher grades (grade III & IV), respectively. The results were again statistically significant (p=0.0001).

It was a single center study based on experience of single surgeon with limited number of patients, which is one of limitations of the study. Further large scale studies are needed to triangulate these results.

CONCLUSION

The early follow up of females with CAH receiving single stage feminizing Genitoplasty have shown promising results in terms of low complication rates and high satisfaction rates by parents and pediatricians. Urinary tract infection is more frequent complication. Success rate was found to be higher in younger age groups and lower CAH grades in this study.

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AUTHOR'S CONTRIBUTION

Following authors have made substantial contributions to the manuscript as under

Rehman IU: Conception and design.

Ali S: Acquisition of data and critical review.

Uzair M: Analysis and interpretation of data.

Abdullah F: Acquisition of data and critical review.

Rehman FU: Final approval of version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.