International Journal of Chemical and Biochemical Sciences
(ISSN 2226-9614)

Journal Home page: www.iscientific.org/Journal.html

Outcomes of feminizing genitoplasty operations in virilized females with congenital adrenal hyperplasia in relation to the previous assessment

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Abstract

Congenital adrenal hyperplasia (CAH) refers to a group of autosomal recessive disorders resulting from deficiency of one of the five enzymes required for synthesis of cortisol in the adrenal gland. CAH is the most common cause of genital ambiguity. Hormonal level and Level of confluence, proved to be the most important factor in technical difficulty of the corrective surgery. In this study we aimed to describe the preoperative surgical anatomy regarding depth of vaginal calibration and how assessing that anatomy provided guidance to the choice of surgical approach and surgical decision making. This is a prospective observational study conducted at Cairo University children hospitals, from 2019-2022 after approving of the ethical committee. The study was a collaboration of the endocrinology unit with the pediatric surgery department to include all CAH Females who presented with virilization and in need of surgical reconstruction were included in this study. Parents satisfaction was excellent 40% satisfaction by surgical outcomes correlated to hormonal control. The authors noted that women with CAH were less satisfied with their genitalia whether operated or not. We concluded that there was no significant difference between total and partial urogenital mobilization procedures regarding postoperative urinary continence in accordance with the literature with favorable good anatomical outcomes and surgical satisfaction to the parents.

Keywords: Feminizing, Genitoplasty, Virilized, CAH.

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1. Introduction

Congenital adrenal hyperplasia (CAH) refers to a group of autosomal recessive disorders resulting from deficiency of one of the five enzymes required for synthesis of cortisol in the adrenal gland. CAH is the most common cause of genital ambiguity [1]. Hormonal level and Level of confluence, proved to be the most important factor in technical difficulty of the corrective surgery [2]. Vaginal reconstruction techniques generally take one of four types: 1- The “cut-back” vaginoplasty, which is rarely used nowadays 2-The “flap” vaginoplasty (Fortunoff) is applicable to a low (distal) vaginal confluence, 3-The pull-through vaginoplasty may be used for any level of confluence but is generally reserved for a very high confluence, 4 - UGS mobilization, either partial or total, 5- Complete vaginal replacement can be achieved by many techniques, but this is used for a rudimentary or absent vagina [3]. In this study we aim to describe the preoperative surgical anatomy regarding depth of vaginal calibration and how assessing that anatomy provided guidance to the choice of surgical approach and surgical decision making.

2. Patients and methods

This is a prospective observational study conducted at Cairo University children hospitals, from 2019-2022 after approving of the ethical committee. The study was a collaboration of the endocrinology unit with the pediatric surgery department to include all CAH Females who

presented with virilization and in need of surgical reconstruction were included in this study. The included patients were above age of 5 years and were Controlled by glucocorticoid and mineralocorticoid replacement therapy, to be clinically and hormonally fit for surgical interventions. Patients with Debatable sex of rearing; mixed gonadal dysgenesis, ovotesticular disorder of sexual development and partial androgen insensitivity syndrome were excluded. We also excluded patients with inadequate compliance with corticosteroid, uncontrolled hormonal assay, thereby, rendering surgery or interventions clinically hazardous or unsuitable. The assessment included pre and intraoperative assessment to correlate level of virilization with type of surgery. Diagnosis of CAH was made preoperatively by 21 hydroxylase deficiency and followed up mainly by `7 hydroxyprogesterone and Hormonal assay including cortisol, ACTH, 17-OH progesterone, DHEA, androstenedione, 11-deoxicortisone, testosterone if needed. The Preoperative assessment included Full history and preoperative data regarding prader score, genitography especially confluence depth, Age, Clinical examination included the external genitalia in presence of the guardian, Karyotyping, Genitography assessing the confluence depth. Dose of hydrocortisone for replacement therapy is adjusted in mg/m2/day every 3-6 months guided by growth parameters, serum electrolytes, degree of virilization and hormonal profile. The performed surgical techniques correlated to preoperative assessment. Cut-back, PUM, TUM and Passerine-Glazel.

3. Results and Discussion

This study prospectively reviewed CAH patients who underwent complex, primary vaginoplasty at a single center by a multiple surgeon. This study assigned to anatomical and functional outcomes regardless the cosmetics except for parents' satisfaction and third person observer. The patients' age in this study ranged from 6 to 14 -year- old this was in concordance with Stites et al. who investigated 45 patients aged between 6- and 18-year-old (Table 1). The selection of this certain age group represents an important factor to have a proper assessment of function [4]. In this study we included different surgical procedures for urogenital reconstruction. We have five cases who had TUM, 15 had PUM and two cases who had Passerine-Glazel procedure. Moreover, we included another 4 cases who had cut back only; thus, they didn't meet the typical inclusion criteria. Braga et al. [5] included in his series different feminizing genitoplasty procedures in his series in which he assessed function, infection, and patient satisfaction [5]. All patients included in this group did not experience complications neither in the form of hormonal control compliance nor a calibration during evaluation (Table 2). On the other hand, this did not go in concordance with literature as Stites et al. reported that 20% of patients who underwent TUM had serious incontinence issues [4]. All cases regarding cut back showed no urological symptoms, the same PUM and passerini vaginoplasty all cases underwent noninvasive uroflowmetry. All of them were within normal range of bladder volume and normal range of Q max and time to urinate in this study, in the study by Wisniewski and colleagues, the physicians perceived the genital appearance as better than did the patients [6]. Callens and associates noted that feminizing surgery did not seem to improve or hamper psychosexual outcome, especially in those patients with severe virilization [7]. There has been some concern that lower urinary tract symptoms are more likely in those patients with CAH but in the study by Fagerholm and colleagues, symptoms did not seem to differ in female DSD patients versus controls. Occasional lower urinary tract symptoms were common in both patients and controls [8]. The results of TUM and PUM are very early. These procedures are technically easier, and the cosmetic results are superior, but whether the functional results are better is unknown (Table 3). The potential for stress incontinence or denervation of the sphincteric mechanisms is also unknown. Most authors to date have not found continence to be altered by TUM [9]. Until these results are available, these procedures should be used with caution (Table 4). Recently there has been an effort to understand sexual function and mental health following feminizing genitoplasty [10-13].

Table 1: Percents of pre-operative confluence depth according to pre-operative hormonal control

		Hormonal control Pre	
		Satisfactory	Unsatisfactory
Low	Count	3	1
	% within Confluence depth	75.0%	25.0%
	% within Hormonal control	15.8%	14.3%
Intermediate	Count	11	3
	% within Confluence depth	78.6%	21.4%
	% within Hormonal control	57.9%	42.9%
High	Count	5	3
	% within Confluence depth	62.5%	37.5%
	% within Hormonal control	26.3%	42.9%
	Count	19	7
	% within Confluence depth	73.1%	26.9%
	% within Hormonal control	100.0%	100.0%

Table 2: Showing preoperative genitography and Prader classifications correlated with operations were done.

	Genitography			Operation	
			prader		
	confluence depth	depth in mm			
				Vaginoplasty	**Labioplasty**
1	Intermediate	19	III	PUM	F,Kogan
2	Intermediate	20	II	PUM	F,kogan
3	Intermediate	18	III	TUM	F,kogan
4	High	32	V	TUM	diamond F,Kogan ,lateral vaginal wall
5	Intermediate	19	III	PUM	F,VC
6	High	33	IV	Passerini	F,VC
7	Intermediate	18	III	PUM	F,VC
8	Intermediate	10	III	Passerini	F,kogan
9	Intermediate	15	III	PUM	F,Kogan
10	Intermediate	14	II	PUM	F,Kogan
11	Intermediate	15	III	PUM	F,VC
12	High	35	III	PUM	F,Kogan,UGS lat vaginal wall
13	low	7	II	Cutback	Kogan
14	High	32	III	PUM	VC
15	High	35	IV	PUM	F,Kogan
16	Intermedite	18	III	PUM	F,Kogan
17	low	8	II	Cutback	Kogan
18	low	7	III	PUM	F,VC
19	Intermediate	17	II	Cutback	VC
20	Intermedite	18	IV	PUM ,inner surface of prepuce	Kogan
21	Intermediate	17	III	PUM ,inner surface of prepuce	Kogan
22	High	30	III	PUM ,inner surface of prepuce	Kogan
23	Intermediate	15	IV	TUM ,inner surface of prepuce	Kogan
24	High	30	IV	TUM ,inner surface of prepuce	Kogan
25	High	30	IV	TUM ,inner surface of prepuce	Kogan
26	low	15	II	Cutback	VC

Table 3: Showing type of UGS operations included.

	Frequency	Percent
Cutback	4	15.4
Passerini	2	7.7
PUM	15	57.7
TUM	5	19.2
Total	26	100.0

Table 4: showing type of UGS operations included (further classified)

	Frequency	Percent
Cutback	4	15.4
Passerini	2	7.7
PUM	12	46.2
PUM ,inner surface of prepuce	3	11.5
TUM	2	7.7
TUM ,inner surface of prepuce	3	11.5
Total	26	100.0

Table 5: Showing Percents of types of Surgical Procedure according to pre-operative hormonal control.

		Surgical Procedure			
		Cutback	Passerini	PUM	TUM
Satisfactory	Count	3	0	13	3
	% within Hormonal control	15.8%	0.0%	68.4%	15.8%
	% within VPL	75.0%	0.0%	86.7%	60.0%
Unsatisfactory	Count	1	2	2	2
	% within Hormonal control	14.3%	28.6%	28.6%	28.6%
	% within VPL	25.0%	100.0%	13.3%	40.0%
	Count	4	2	15	5
	% within Hormonal control	15.4%	7.7%	57.7%	19.2%
	% within VPL	100.0%	100.0%	100.0%	100.0%

Table 6: Showing Percents of pre-operative confluence depth according to VPL type.

		Surgical Procedure			
		Cutback	Passerini	PUM	TUM
High	Count	0	1	4	3
	% within Confluence depth	0.0%	12.5%	50.0%	37.5%
	% within VPL	0.0%	50.0%	26.7%	60.0%
Intermediate	Count	1	1	10	2
	% within Confluence depth	7.1%	7.1%	71.4%	14.3%
	% within VPL	25.0%	50.0%	66.7%	40.0%
low	Count	3	0	1	0
	% within Confluence depth	75.0%	0.0%	25.0%	0.0%
	% within VPL	75.0%	0.0%	6.7%	0.0%
	Count	4	2	15	5
	% within Confluence depth	15.4%	7.7%	57.7%	19.2%
	% within VPL	100.0%	100.0%	100.0%	100.0%

Parents satisfaction was excellent 40% satisfaction by surgical outcomes correlated to hormonal control. The authors noted that women with CAH were less satisfied with their genitalia whether operated or not (Table 5). Nordenström and associates noted that clitoral sensitivity was affected in nearly all patients who had undergone clitoroplasty. However, those with partial clitoral resection sparing more nerve function had a more favorable outcome and did not differ from those with no surgery. Initial degree of virilization also affected clitoral sensitivity [14]. Fagerholm and colleagues noted that quality of life and health-related quality of life studies appeared normal in most patients and that mental health was similar to or better than published data from Finland. There were 5 of 24 patients who had some poorer scores because of distressful memories of too-late surgery, the operative treatment itself, or poor sexual function. This same group also noted that intercoital relationships started later in females who underwent genital reconstruction in childhood as compared to the normal population. In addition, the patients preferred surgery to be done early (Table 6) [8].

4. Conclusion and recommendations

We concluded that there was no significant difference between total and partial urogenital mobilization procedures regarding postoperative urinary continence in accordance with the literature with favorable good anatomical outcomes and surgical satisfaction to the parents.

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