



Surgical Outcomes in Patients with Disorders of Sex Development in Mofid Children's Hospital, 2001-2014

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Abstract

Background: Disorders of Sex Development is a childhood and infantile anomaly affects not only the somatic growth; but also leading to stress and anxiety among parents who are seeking optimal treatments. Accordingly in this study the surgical outcomes in patients with Disorders of Sex Development in Mofid Children's Hospital from 2001 to 2014 were determined.

Materials and Methods: In this retrospective study children with Disorders of Sex Development in Mofid Children's Hospital from 2001 to 2014 who met the eligibility criteria were included. Data were gathered by existing medical documents and were recorded in prepared checklist. The surgical outcomes were assessed with interview and clinical examination after announcement by hospital. The success and complication rate including external genitalia acceptable feature, vaginal stenosis in prepuberty period, intercourse problems during fertility period and gender dysphoria were determined by a multi-disciplinary team including Paediatric Surgeons, Paediatric endocrinologists and Psychologists.

Results: In the current study we have evaluated seventy two patients; 55 (76.38%) affected by Congenital Adrenal Hyperplasia, thirteen (18.05%) by Testicular Feminization, 2 (2.7%) by Ovotesticular Disorder and two case (2.7%) by Mixed Gonadal Dysgenesis. Most common type of applied surgery was Clithroplasty, Genitoplasty and Pull through Vaginoplasty (47 cases). Fifty nine patients (81.9%) had no surgical complications. All patients had good conditions at discharge and no mortality was registered. Three cases of Testicular Feminization (4.2%) Who Underwent pull through colovaginoplasty were married.

Conclusion: According to our findings, surgical outcomes in cases of Disorders of Sex Development are relatively good and satisfactory. However long-term follow-up study is required to determine the final outcomes especially for marital and sexual issues.

Keywords: Clitoroplasty; MixGonadal; Ovotestis; Vaginoplasty

Introduction

Disorders of Sex Development are a childhood and infantile problem affecting not only the somatic growth; but also leading to stress and anxiety among parents who are seeking optimal treatments [1-3]. Although there are different therapeutic options; there is no definite treatment other than surgery [4-7]. One of the important issues for this matter is external genital feature [8-10].

Prompt treatment and alleviation of anatomical problems would result in better outcomes with decreased psychological effects, emotional trauma and mental preparation for children [9-11]. Another importance of optimal therapeutic choice in children with DSD is the corresponding effects on social, sexual, mental, and cultural characteristics before the initiation of sexual function [12-14]. The continuous follow-up in these patients before puberty revealed disordered sexual and marital function and probable fertility problems [11-14]. Recognition and application of the best surgical approaches would lead to better treatment outcomes. Accordingly in this study the surgical outcomes in patients with Disorders of Sex Development in Mofid Children's Hospital from 2001 to 2014 were determined.

Materials and Methods

In this retrospective study children with Disordered Sexual Development in Mofid Children's

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Table 1: Karyotype distribution in patients.

Anomaly	karyotype	Final assignment
CAH	53 XX	Female
	1 deletion 14 and 21	Female
	1 XO-XX	Female
Testicular Feminization	13 XY	Female
Ovotesticular	1 XY	Female
	1 XY	Male
Mixed gonadal Dysgenesis (MGD)	2 XO-XY	Female



Figure 1: Intra operative feature (Clitoral reduction, Genitoplasty) in CAH patient.

Hospital from 2001 to 2014 who met the eligibility criteria were included. Surgical reports, endocrinology assessments, post operative outpatients visits, possible complications and their managements were gathered by existing medical documents and recorded in prepared checklists. All patients were referred by endocrinologists for surgical assessment or correction. The latest outcomes were assessed with interview and clinical examination after announcement by hospital. The success and complication rate including external genitalia acceptable feature, vaginal stenosis in prepuberty period, intercourse problems during fertility period and gender dysphoria were determined by a multi-disciplinary team including Paediatric Surgeons, Paediatric endocrinologists and Psychologists. Diagnosis of



Figure 2: Intra operative feature (Clitoral reduction, Genitoplasty) in CAH patient.

Disordered Sexual Development was not obtained during perinatal or neonatal period in all patients. Data analysis was performed by SPSS (version 18.0) software [Statistical Procedures for Social Sciences; Chicago, Illinois, USA]. Mean and standard deviation were reported for numerical data and the frequency and percent were reported for categorical data.

Results

Seventy two patients met the defined eligibility criteria of available comprehensive medical documents and possibility of refer to hospital for surgical, endocrinological and psychological assessment. The mean age at the time of surgery was 4 years and 5 months: minimum age was 2 months and maximum age was 18 years. 53 patients with XX karyotype, 16 Patients with XY karyotype and the remaining 3 patients were found to have mosaic karyotype. As shown in Table 1. In five cases (6.9%) there was positive family history of DSD. Female pseudohermaphroditism, male pseudohermaphroditism, and other types of DSD including Mixed Gonadal Dysgenesis: and Ovotesticular were seen in 55 (76.4%), 13 (18.1%), 2 (2.75%) and 2 (2.75%) patients, respectively. In 54 patients (75 percent) there were no associated anomalies (Table 2). The most common used surgery was clitoroplasty, vaginoplasty plus genitoplasty (Figure 1,2). In high confluence CAH patients we did not performed total mobilization

Table 2: Associated anomalies in patients.

Associated Anomalies	Diagnosis	Frequency	Percent	Cumulative Percent
Inguinal hernia	MGD	1	2.8	2.8
	CAH	1		
Hypospadias	CAH	3	4.2	6.9
Cryptorchidism	TF	1	1.4	8.3
Chromosomal anomalies (der (14;21)(q10;q10) balanced robertsonian translocation)	CAH	1	1.4	9.7
Inguinal hernia+Cryptorchidism	TF	2	2.8	12.5
TTN +VUR	CAH	1	1.4	13.9
VUR+FTT	CAH	1	1.4	15.3
brain atrophy in MRI	CAH	1	1.4	16.7
microcephaly	CAH	1	1.4	18.1
turner characteristics	CAH	1	1.4	19.4
ASD		1	1.4	20.8
Congenital right calcaneous valgus	CAH	1	1.4	22.2
Cardiac Anomaly	TF	1	1.4	23.6
Single kidney(Recurrent UTI)	CAH	1	1.4	25
None		54	75	100

TTN: Transient Tachypnea of Neonate; VUR: Vesicoureteral Reflux; FTT: Failure to Thrive; ASD: Atrial Septal Defect; UTI: Urinary Tract Infection

Table 3: Surgical techniques.

Type of surgery	Frequency	Valid Percent	CAH	Testicular Feminization	Ovotesticular	Mixed gonadal Dysgenesis (MGD)
1.Clitoroplasty+Vaginoplasty+Genitoplasty	47	65.3	46	-	1	1
2.Clitoroplasty+Genitoplasty+ colovaginoplasty	9	12.5	9	-	-	-
3.Hysterosalpingectomy+LeftGonadectomy+RightOrchidopexy	1	1.4	-	-	1	-
4.Genitoplasty+ColoVaginoplasty+Left Orchidectomy	1	1.4	-	-	-	1
5.ColoVaginoplasty+BilateralGonadectomy	13	18.1	-	13	-	-
Total	72	100	55	13	2	2

Table 4: Complication patients.

Complication		Anomaly	Frequency	Percent
Early	Hematoma	CAH	1	1.4
	abdominal evisceration	CAH	1	1.4
Late	Urinary retention	CAH	1	1.4
	Vaginal Complications(Stenosis)	Testicular Feminization and CAH	8	11.1
	Fistula Formation	CAH	2	2.8

of urogenital sinus due to risk of urinary incontinency. In two of High Confluence patients we saved the common channel as urethra and separated vagina at the confluence site and colovaginoplasty for genital tract reconstruction. In three cases of high confluence CAH we did Clitoroplasty and Genitoplasty in first operation followed by pull through colovaginoplasty later as the second operation. Female karyotype to female phenotype was the most type of gender assignment (Table 1). Fifty nine patients (81.9%) had no complications (Table 4). The most common mentioned complication was vaginal stenosis which were managed by serial dilatation plan (Table 5). All patients had good conditions at discharge and no mortality was reported.

Discussion

In the current study, the mean age at the time of surgery was 4 years and 5 months. In the study by Heng Zhang et al. [15] the median age of patients at the time of surgery was subdivided into three discrete lives time period including 6.4 years during childhood, 14.2 years during adolescence and 23.2 years during adulthood [15]. While in the study by Gulnur Gollu et al. [16] the mean at the time of surgery was 4.4 ± 2.3 years [16]. Furthermore the mean age of surgical reconstruction was mentioned to be 3.5 ± 1 year in the study by Eroglu Get et al. [17].

Finally the mean age of surgical intervention in the current study was similar to the study conducted by Gulner Gollu. The clear impact of timing in surgical reconstruction has yet to be defined based on psychological surveys and sexual function status during marital period. The most common surgical technique applied in the current study was clitoroplasty, vaginoplasty and genitoplasty among 65.3% of patients, while colovaginoplasty and Bilateral gonadectomy was undertaken in 13 patients who were affected by testicular feminization. The third most common type of surgery was clitoroplasty, genitoplasty and colovaginoplasty in 9 patients who were suffering from high-confluence CAH.

Among the superiorities of our study was surgical technique classification based on the specific type of Disordered Sexual development including CAH, TF, MGD and Ovotesticular disorder. In the study by Heng Zhang et al. [15] surgical procedures were subdivided to two main subgroups of those who were assigned to be

Table 5: Vaginal stenosis as a complication.

Diagnosis	Occurrence of vaginal stenosis (frequency)	$\leq 2 / \geq 3$ frequency of Dilatation
CAH	2	≥ 3
	5	≤ 2
TF	1	≥ 3
MGD	-	-
OVD	-	-

male versus female. Twenty five out of a hundred-sixty three female assigned patients underwent Gonadectomy without Genitoplasty and Genitoplasty was performed in the remaining 138 patients [15]. In the study conducted by Gulnur Gollu et al. [16] male assignment was mentioned in five CAH patients who were reared as male due to delayed diagnosis [16].

In all those five patients hypospadias repair in multiple stages, resection of mullerian remnant structures and testicular prostheses placement were undertaken, while in our study only one case of male assigned CAH underwent urethroplasty, scrotoplasty and multiple hypospadias repair.

In the study conducted by Eroglu G et al. [17] methods of applied surgical techniques were categorized based on four main DSD subtypes including 1 (Table 3). Female pseudohermaphroditism as the most common type of DSD patients participating in the study (CAH), 41 patients in whom clitorovaginoplasty, staging procedures, clitoroplasty, clitoridectomy and total mobilization of urogenital sinus in the manner of reducing frequency were undertaken 2. Male pseudohermaphroditism, in whom similar to our study Gonadectomy and bowel vaginoplasty was undertaken in 10 patients and in 3 others Clitoroplasty was performed. In only one case of Mixed Gonadal Dysgenesis, colovaginoplasty was mentioned [17].

In the current study; we included 2 patients affected by MGD and due to female assignment in; Genitoplasty, ColoVaginoplasty and uni lateral Orchidectomy were undertaken while Clitoroplasty, Vaginoplasty and Genitoplasty in the other one. Regarding considerable aspects of our study was categorizing postoperative complications in two discrete subtypes of early and late complications

including hematoma and evisceration in the former and urinary retention, vaginal stenosis and fistula formation in the later group. The most common complication was vaginal stenosis demanding further dilation. In the study by Heng Zhang et al. [15] the postoperative complications were classified as Genital and Urologic ones. Vaginal stenosis demanding dilation or surgical correction in the former group, penile augmentation, urethral stricture and difficult urination in the later one was mentioned [15].

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