Feminizing Reconstructive Surgery for Ambiguous Genitalia: The Leipzig Experience

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Purpose: We report the results of feminizing reconstructive surgery for ambiguous genitalia with regard to the degree of virilization according to Prader and the long-term outcome with special emphasis on sexual intercourse.

Materials and Methods: Patients 16 to 46 years old with CAH (41) and MPH (17) were followed continuously by an interdisciplinary team that provided standardized hormone substitution and reconstructive surgery. More virilized genitalia were treated using a 2-stage procedure.

Results: Of patients with CAH no surgery and clitoris reduction were done in 4 and 2 (Prader I and II), no surgery, a 1-stage and a 2-stage procedure were done in 2, 3 and 4 (Prader III), a 2-stage procedure, a 1-stage procedure and surgery for fistula were done in 16, 4 and 2, respectively, while 1 refused surgery (Prader IV), and a 2-stage procedure was done in 5, including pull-through vaginoplasty in 2 (Prader V). Of patients with MPH no surgery was done in 6 (Prader I and II), a 1-stage procedure and a 2-stage procedure were done in 3 and 1 (Prader III), respectively, and a 2-stage procedure was done in 6, while 1 refused surgery (Prader IV). Revision vaginoplasties were necessary in 7 patients (12.1%) because of scar stenosis in those who underwent 1-stage as well as 2-stage procedures. None of our patients required reconstructive surgery to create a neovagina. Of the 58 patients 32 achieved sexual intercourse and in 17 the postoperative result would allow sexual intercourse, while in 3 the possibility of sexual intercourse is uncertain. For 3 patients sexual intercourse would not be possible since feminizing reconstructive surgery was refused. One patient could not be followed.

Conclusions: Two-stage surgery leads to a favorable outcome in patients with CAH and MPH, and virilized genitalia (Prader IV and V).

Key Words: abnormalities, adrenal hyperplasia, congenital, pseudohermaphroditism, genitalia, sexual development

The primary goals of feminizing genital reconstruction are to create a normal-looking, sensate clitoris, provide an adequately sized and appropriately situated vagina, and create normal-appearing female external genitalia. Complications include vaginal stenosis, meatal stenosis, vaginal-urethral fistula, female hypospadias, urinary tract injuries, recurrent clitorimegaly and others. In the long term the ultimate goals of feminizing genital reconstruction are to provide good cosmetic results, sexual satisfactory and if possible fertility.

Unfortunately there is little information about long-term followup and possible complications after surgical repair. There is ongoing controversy concerning treatment for more virilized ambiguous genitalia. Different types of feminizing reconstructive procedures and various timing of surgery between 1940 and 1980 were described by Schober.[1] Results were not always satisfactory, especially in more virilized ambiguous genitalia. Various percents of vaginal stenosis requiring additional surgery have been reported by Jones et al (30% in 84 patients),[2] Nihoul-Fekete et al (30% in 48),[3] Bailez et al (78% in 28)[4] and Kitahara et al (12% in 41).[5] Comparison of these data are difficult since most reports do not provide sufficient information about primary genital status. Nevertheless, the number of further vaginal corrections seems to be high. In general more severe virilized genitalia have been corrected with 2-stage procedures.

In the mid 1980s there was a shift by many surgeons in the technique of treating these children, which has remained valid until the present,[6] that is that all patients with intersex should and could be corrected entirely within the first 2 to 6 months of life with a 1-stage procedure. Many other investigators have suggested modified new approaches to improve exposure during surgery for high confluence anomalies, such as the early perineal prone approach, total urogenital sinus mobilization, posterior sagittal transanorectal approach and anterior sagittal transanorectal approach.

We have not used these new techniques at our center for ill children at University of Leipzig. We have exclusively performed a 2-stage procedure for more virilized genitalia (Prader IV and V[7]). We report our results of feminizing reconstructive surgery for ambiguous genitalia with regard to the degree of virilization according to Prader and the long-term outcome with special emphasis on SI.

PATIENTS AND METHODS

The study included 16 to 46-year-old female patients with CAH (41) and MPH (17). Patients were treated by an interdisciplinary team using standardized guidelines for corticoid substitution and reconstructive surgery. The study com-

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prised continuous long-term followup investigations. Patients visited a specialized pediatrician 2 or 3 times yearly. Data were collected from patient records and surgical notes, including the degree of virilization, method of surgery, postoperative course and evaluation at puberty.

We analyzed sexual development and sexual life by a semistructured interview after written consent was provided by the patients. The advantages of this method are the complete evaluation of relevant data as well as the tendency of the answers and further information. Thus, the heterosexual and homosexual behavior, and the orgasmic experiences of every patient could be judged in detail. Furthermore, the study was done using personal communications during outpatient visits. In patients who had not achieved sexual intercourse a gynecological investigation was performed to evaluate whether sexual intercourse would be possible.

Feminizing genital reconstruction surgery was performed by 2 pediatric surgical consultants. Total CE was performed until 1975. Thereafter we preferred plastic CR, as performed according to Marberger et al.[8] The procedure involves complete resection of the corpora cavernosa with preservation of the glans and neurovascular bundle, and the creation of labia minora from phallic skin. The goal is to create external genitalia with a normal appearance and with an innervated but nonerectile clitoral phallus. Furthermore, normal bladder function and genital sensitivity must be retained. The VP technique depended on the size of confluence. As described by Fortunoff et al,[9] posterior flap vaginoplasty was used for low confluence and a vaginal pull-through procedure according to Hendren and Crawford[10] was done in cases with high take-off of the vagina from the urogenital sinus.

The timing of surgery and surgical strategy generally depended on the degree of virilization according to Prader. In genital type Prader I and II clitoral reduction was performed only according to family preference. Genital type Prader III was generally corrected with a 1-stage procedure, ie clitoral reduction and vaginoplasty. Genital type Prader IV was corrected with a 2-stage procedure by performing clitoral reduction at infant or preschool age and vaginoplasty during puberty. The timing of the second procedure was fixed as the result of consultation of the pediatrician with the families between patient ages 13 and 17 years. Genital type Prader V was also corrected with clitoral reduction at infant or preschool age. According to the result of genitoscopy before the second operation at the pubertal age vaginoplasty or vaginal pull-through was performed.

RESULTS

Table 1 shows results in patients with CAH with regard to the degree of virilization.

Genital type Prader I and II. Two patients presented with progressive enlargement of the clitoris due to long-term refusal of hormone treatment. These patients underwent clitoral reduction.

Genital type Prader III (table 1). We performed 1 and 2-stage procedures. RVP was necessary in 2 patients. One patient underwent temporary calibration.

TABLE 1. *Patients with CAH*

Pt No.—Age	Surgery (age at surgery)	SI (age at first SI)	Remarks
	Prader I + II (6 pts)		
1—33	CR (14)*	Yes (15)	
2—29	None	Yes (17)	2 Children
3—24	CR (23)*	Yes (18)	Twins
4—20	None	Possible	
5—17	None	Yes (18)	
6—17	None	Possible	
	Prader III (9 pts)		
7—46	CE (8), VP (19)	Yes (19)	1 Child
8—37	CE+VP (5), RVP (7), RVP (19)	Yes (22)	
9—36	CE (2), VP (5)	Yes (18)	1 Child
10—30	CR (3), VP (18)	Yes (18)	1 Child
11—29	None	Not possible	Asexual, refused surgery
12—26	None	Yes (16)	
13—21	CR+VP (6), RVP (15)	Possible	Lesbian
14—20	CR (3), VP (12)	Possible	
15—16	CR+VP (2), calibration (16)	Possible	
	Prader IV (21 pts)		
16—40	CE (6), VP (8)	Yes (16)	
17—34	CE (2), VP (7), RVP (16)	Yes (18)	
18—33	CE (3), VP (17)	Yes (22)	1 Child
19—33	CE (4), VP (17)	Possible	Lesbian
20—32	CR (11)	Not possible	No sexual interest, refused VP
21—32	CE (2), VP (8), RVP (16)	Yes (18)	
22—32	CE (2), VP (13), RVP (17)	Yes (19)	
23—29	CE (2), VP (15)	Yes (20)	2 Children
24—28	CR+VP (7)	Yes (17)	
25—27	CE (5), VP (27)	Possible	No sexual interest
26—24	CR (5), VP (17)	Possible	
27—22	CR (3), VP (14)	Yes (19)	
28—22	No CR, VP (11)	Yes (16)	1 Child
29—22	CR (3), VP (15)	Yes (18)	
30—21	CR (2), VP (13)	Possible	
31—20	CR (2), VP (18)	Yes (18)	
32—20	CR+VP (17)	Yes (19)	
33—19	CR (2), VP (12), stitch fistulas	Possible	
34—18	CR (3), VP (15)	Possible	
35—17	CR+VP (2), RVP (14)	Yes (16)	
36—16	CR (1), VP (15), stitch fistulas	Possible	
	Prader V (5 pts)		
27—32	CE (2), VP (15)	Doubtfully possible	No sexual interest
38—31	CR (6), PVP (11), RVP (20)	Possible	Lesbian
39—26	CE (1), meatal stenosis (1), VP (17)	Possible	No sexual interest
40—26	CR (2), VP (10)	?	No followup
41—19	CR (2), PVP (15)	Doubtfully possible	Sexual interest not yet known

* Refused hormonal replacement therapy in adolescence.

Genital type Prader IV (table 1). These patients generally underwent a 2-stage procedure, although patient 24 underwent a 1-stage procedure. Three patients required a further corrective vaginal procedure after 2-stage feminizing