Effects of feminizing surgery for ambiguous genitalia – a novel scale for evaluation of cosmetic and anatomical results

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Key words: ambiguous genitalia; congenital adrenal hyperplasia; hermaphroditism; feminizing surgery; clitoroplasty; genitoplasty; vaginoplasty; assessment scale;

Abstract

INTRODUCTION: Ambiguous genitalia always present diagnostic and therapeutic problem. Long-term results of feminizing operations are unsatisfactory in some cases and reports from follow-up after feminizing genitoplasty are rare in the literature. Systematic studies are needed to evaluate ultimate function of all girls undergoing feminizing surgery. In our opinion, the lack of worldwide-accepted scale for the assessment of long-term effects of feminizing genitoplasty, enabling the possibility of comparing outcomes between institutions and countries, may contribute to this deficit of reliable data. AIM: The aim of the study was the evaluation of the outcomes of surgical management of masculinization using a novel scale developed by the authors. MATERIAL AND METHODS: We examined 43 patients aged 3–24 years (mean age 15.4 years) operated due to ambiguous genitalia. Most of the patients were females with congenital adrenal hyperplasia (CAH) diagnosed in 38 of subjects (88.4%). The patients were operated at the age from 10 months – 15 years (mean age 4.5 years). Five patients had to be reoperated. The effects of surgical management in ambiguous genitalia were assessed using our own scale. We evaluated five anatomical and cosmetic parameters (general appearance, size of pudendal labia, symmetry of pudendal labia symmetry, size and position of clitoris, size of introitus and position of urethra opening), each of them was scored 0–2 points. RESULTS: In 36 of examined patients the result of the surgery was considered good, in 4 patients – satisfactory and in 3 – poor. The most common complication of feminizing genitoplasty in our patients was stenosed vagina (in 10 patients, 23.8%). Location of urinary coil in anterior wall of vagina was found in 6 patients (13.9%) and in 1 patient it was invisible (2.3%). Post-operative clitoral enlargement was found in 3 cases (6.9%). CONCLUSION: Taking into consideration controversial data about the results of feminizing genitoplasty, a standarized, well-defined and commonly accepted scale enabling comparison between methods and institutions is necessary. In our opinion scoring scale makes the evaluation more precise and the results are more comparable.
INTRODUCTION

Intersex refers to conditions in which the phenotype is neither male nor female or the chromosomal or gonadal sex of the individual is at variance with the phenotypic sex (Sax, 2002). Over 80% of intersex disorders is diagnosed with congenital adrenal hyperplasia (Frimberger & Gearhart, 2005). Ambiguous genitalia always present diagnostic and therapeutic problem. Most cases of genital ambiguity are classified as Prader I to III, and are characterized by the presence of a vaginal opening situated distally to the external urethral sphincter and very close to the perineal skin (Prader, 1954; Fortunoff et al., 1964; Spence & Allen, 1964). In complete masculinization, a complete fusion of labia minora and a hypertrophied clitoris with an urethral opening at the tip of the glans clitoridis, appearing as “normal” male external genitalia, except for palpable testes are observed (Prader V) (Prader, 1954). Surgical management in case of variable degree of masculinization should be directed to removing the corpora and preserving the glans with its innervation to create a clitoris with normal sensation, creating a normal-appearing introitus by fashioning labia minora from phallic skin and foreskin and vaginoplasty to provide an adequate opening for the vagina onto perineum (Snyder et al., 1983).

Operation of clitoral shaft resection with preservation of the glans on its neurovascular bundles seems logical, however the data on its later functional/sexual functions are controversial (Crough, 2008). Techniques for vaginal reconstructions have progressed considerably after Hendren and Crawford (1969) defined the concepts of “high” and “low” vagina and established the importance of preserving the external urinary sphincter to prevent postoperative urinary incontinence. When vagina is “low”, e.g. it reaches the urogenital sinus distal to the external urethral sphincter, that is observed in cases of Prader I–III masculinization, it can be simply exteriorised by single flap vaginoplasty described by Fortunoff et al. in 1964. In patients where the urethra communicates with the urogenital sinus proximal to the external sphincter other techniques as “pull-through” vaginoplasty by Hendren and Crawford or Passerini-Glazel are used (Hendren and Crawford, 1969; Passerini-Glazel, 1989). Several modifications of abovementioned techniques have been reported (Farkas et al., 2001; Miranda et al, 2004; Savanelli et al, 2008).

Age at which feminizing genitoplasty should be performed remains still controversial. Although a Consensus Statement (LWPES/ESPE CAH Working Group, 2002) recommends surgical reconstruction in early childhood (first 2–6 months), there are also some data, that early vaginal surgery bears the risk of fibrosis and scarring, so that reconstruction of the vagina should probably be deferred until puberty (Alizei et al, 1999).

Long-term results of these operations are unsatisfactory in some cases and reports from follow-up after feminizing genitoplasty are rare in the literature. Systematic studies are needed to evaluate ultimate function of all girls undergoing feminizing surgery. In our opinion, the lack of worldwide-accepted scale for the assessment of long-term effects of feminizing genitoplasty, enabling the possibility of comparing outcomes between institutions and countries, may contribute to this deficit of reliable data. Creighton et al. (2001) proposed a well-described, standardized approach that assesses the clitoris, labia minora and majora and introitus. Apart from obtaining genital appearance compatible with gender and good adult sexual and reproductive function, the third important goal of the surgery is unobstructed urinary emptying without incontinence or infections (LWPES/ESPE CAH Working Group, 2002). Thus, in our opinion, an evaluation of the position of external urethra opening may be important and useful and we developed our own scale for the assessment of cosmetic and anatomical effects of feminizing genitoplasty.

This study was undertaken to evaluate the outcomes of surgical management of masculinization in patients operated in our institution using a novel scale developed by the authors.

MATERIAL AND METHODS

We examined 43 patients aged 3–24 years (mean age 15.4 years) operated due to ambiguous genitalia at the Department of Urology, Silesian University of Medicine in Katowice, Poland from Jan 1989 to Nov 2002 (Table 1). The most frequent cause of ambiguous genitalia in examined group was CAH, diagnosed in 38 of subjects (88.4%), there were also 2 other cases of pseudohermaphroditism (4.7%) or mixed gonadal dysgenesis (3 patients, 6.9%). The karyotype was 46, XX in 38 patients and 46, XY in 5 patients. The diagnostic procedures before the surgery in all patients included: ultrasonography of pelvis, genitoscopy and genitography. In 3 patients abdominal CT was performed, 4 patients underwent urography, in 5 patients dysgenetic gonads were removed before genitoplasty by laparotomy (n=3) or laparoscopy (n=2).

The masculinization degree before the operation was assessed according to Prader scale. The majority of patients were considered as Prader III (74.5%) (Table 2).

The patients were operated at the age from 10 months – 15 years (mean age 4.5 years) (Table 3). Five patients had to be reoperated. The first surgery was performed at the age 2–3 years (mean 3 years), the reoperation was done after 3–5 years (mean 4.2 years) after the initial surgery. All patients were operated in the technique that is applied in Urology Dept., Silesian Medical University in Zabrze, Poland (Paradysz et al., 1995).

**List of abbreviations:**
- CAH – congenital adrenal hyperplasia
- CT – computed tomography
- UTI – urinary tract infection
In patients recognized as Prader II a "cut-back" vaginoplasty and reconstructive operation of clitoris with the excision of cavitory bodies with preservation of dorsal vascular and neural bundle and glans of clitoris was performed. Surgery in Prader III consisted of reconstruction of vestibule of the vagina using gluteal and perineal lobe in shape of "U" letter turned upside down and clitoroplasty. In Prader IV additionally pudendal labia were reconstructed. Patients with Prader V underwent vaginoplasty by "pull-through" method, preservative clitoroplasty and reconstruction of pudendal labia.

The effects of surgical management in ambiguous genitalia were assessed using our own scale developed for this purpose (Table 4). We evaluated five anatomical and cosmetic parameters, each of them was scored 0–2 points. Overall outcome was considered as good, when the total score was 9–12, satisfactory, when 5–8 and poor, when scored 0–3 points. All subjects were evaluated by the blinded urologist (not the operator), who was not involved in the care of the patients.

RESULTS

Results of our assessment are presented in Table 5. In 36 of examined patients the result of the surgery was scored as good. This group included 5 reoperated patients.

In 4 patients the effect were considered satisfactory. In this group narrowed introitus was found in 2 patients, and in the other 2 the examination revealed protruding clitoris. In 3 of these 4 patients external ostium of urinary coil was located in anterior wall of vagina and in 1 it was invisible.

In 3 patients an overall result of surgical management was poor. In 2 subjects it was complicated by slit-like introitus and in 1 patient vaginal stenosis and protruding clitoris were stated. All 3 patients ranged to this group had urinary coil located in anterior wall of vagina. The most common complication of feminizing genitoplasty in our patients was stenosed vagina (in 10 patients, 23.8%). Location of urinary coil in anterior wall of vagina was found in 6 patients (13.9%) and in 1 patient it was invisible (2.3%). Post-operational clitoral enlargement was found in 3 cases (6.9%).

DISCUSSION

Many technical improvements have been made for vaginoplasty in the last few years. However, there is still significant consumer dissatisfaction, with intersex peer support groups around the world expressing condemnation of infant genital surgery performed for cosmetic reasons (Creighton, 2005).

In children with ambiguous genitalia assigned female vaginoplasty is an integral part of feminizing genitoplasty and is commonly performed during the first year of life. However, the longitudinal results of this procedure are controversial and reports on anatomical and cosmetic outcomes after genital surgery for children with ambiguous genitalia are mixed.

Krege et al. (2000) examined 27 patients with CAH with special emphasis on vaginal functional outcome and sexual activity after vaginoplasty. Mean age at the time of first operation was slightly lower than in our patients and was 3.6 (1–9) years. Vaginoplasty was performed in 24 patients as described by Fortunoff et al. (Fortunoff, 1964) and one patient required "pull-through" technique. During the long-term follow-up time (they evaluated patients aged 14–33 years), more than one third (36%) of observed patients had secondary vaginal stenosis. Six patients of them had Prader III and three Prader IV grade.

On the other hand, in the study by Stikkelpbroek et al. results of vaginoplasty performed at the age 0.1–3.7 years are poor, as 6 of the 7 operated patients needed re-do vaginoplasty in puberty due to vaginal stenosis, and 2 of the 6 patients experienced some urinary incon tinence. It should be noticed, that observed group was very small. They were operated by the same paediatric surgeon. This brings a question about experience of the institution, because the Consensus says that feminizing genitoplasty should be performed at the centre with experience of at least 3–4 cases/yr. (LWPES/ESPE CAH Working Group, 2002).

Other authors (Hoepffner, 2006) report that revision vaginoplasties were necessary in 12% of patients, because of scar stenosis in patients who underwent 1-stage as well as 2-stage procedures. These data are consistent with observations made by Burgu et al. (2007). They identified 63 patients who underwent a total 71 vaginoplasties using posterior skin flap, pull-through and intestinal replacement. The majority of operations were performed before puberty and as primary procedures in patients with various causes of intersex genitalia. Strictures and discharge were the most common problems, and nine patients underwent revision surgery. Also Sircili et al. (2006) obtained good results of the single-stage feminizing genitoplasty. In their study the incidence of vaginal introitus stenosis was about 12%.

Farkas et al (2001) showed good cosmetic and functional results of 1-stage feminizing genitoplasty performed by modified technique of maximal mobilization of argental sinus en bloc including the vagina and urethra via perineal approach. The patients were operated very early (0.9±0.3 years). They report, that girls who reached puberty menstruate normally and there is no evidence of fibrosis of scarring of the perineum. In smaller girls they were able to calibrate the vaginal opening easily with a 20 to 22Fr bougie. However, this follow-up seems to be rather short (4.7±2.6 years), and the authors do not provide more detailed data about the patients (as e.g. Prader stage).

Similar excellent results are obtained by Miranda et al. (2004). They used labioscrotal island flaps to enlarge the vaginal introitus in all procedures. From 11 oper-
lated patients, 7 were high Prader stage (IV or V). This technique is also used by us. The postoperative follow-up ranged from 6 months to 8 years and its course was not complicated by vaginal stenosis in any case. In 3 girls the cosmetic result was not fully satisfied because of corrugated labia majora with excess skin. There was also one instance of a minor dehiscence of the suture of labia majora that resolved spontaneously.

In the study by Braga (2006), adequate caliber of mobilized vagina was achieved in 23 of 24 patients.

These data suggest, that the effects of operations performed using partial or total urogenital sinus mobilization are better than in case of traditional techniques.

In the past few years the appropriate timing of vaginoplasty is under dispute. It has been suggested, that delayed vaginoplasty until puberty may lead to better vaginal healing. This would avoid young children undergoing a painful operation to produce a vagina which is unnecessary in childhood and may well need further revision surgery (Alizei, 1999; Creighton, 2005; Burgu, 2007). It seems obvious, the higher Prader stage, the more difficult operational conditions are (Oświęcimska, 2006) and especially in cases of high vaginal confluence, the surgery should be deferred. Göllü et al. (2007) delayed vaginoplasty in cases of high vagina until 2 to 4 years of age. Typically, vaginal stenosis was the most common complication and was observed in 10 of 18 children who underwent feminization procedures. This supports the hypothesis by Passerini-Glazel (1999), that a single stage procedure is mainly advantageous for the most virilized patients when the vagina is relatively small, and contemporary use of the split urogenital sinus allows the best mucosal lining and reconstruction of the vulvar region. This advantage is lost if vaginoplasty is performed at a later stage than clitoroplasty.

Concomitant clitoroplasty and vaginoplasty gives the opportunity to use the mucosal layer of preputial skin to give extra length to penile flaps on vaginal introitus and labia minora reconstruction (Savanelli et al, 2008). Fourteen patients Prader III–V were operated with this technique, and only one vaginal stenosis was observed in a case with high confluence.

In our study stenosed vaginal introitus was found in 23.8%. The range of age at our patients were operated was very wide (1–15 years). Most of the cases were operated until they were 3 years old, but in some patients the operation was deferred due to different reasons.

Cosmetic results of clitoroplasty are much better than vaginoplasty. In our study post-operational clitoral enlargement was found in 3 cases (6.9%).

Farkas et al. (2001) reported 1 case of total clitoris loss and 2 cases of clitoromegaly for 49 operated patients. Miranda et al. (2004) observed only one case of clitoris positioned within the labia with the thigh in an adducted position for 11 evaluated girls. In the study by Gupta et al. (2006) the cosmetic outcome of clitoroplasty was excellent in 37, satisfactory in 10 and poor in 3 of total 50 evaluated patients. Very similar results were obtained by Göllü et al. (2007). Among 50 patients who underwent clitoroplasty, cosmetic appearance was excellent in 35 (70%) patients, satisfactory in 11 (22%) and poor in 4 (8%) (atrophy clitoris in 2 and clitoromegaly in 2) patients. However, very little is known about the function of clitoris after surgery. Any incision to the clitoral glans, corpora or hood may risk damage to the innervation (Creighton, 2005). In women, who underwent the feminizing genital surgery there is a significant impairment to sensitivity in the clitoris compared to controls. Moreover, linear relationship between the impairment in sensitivity and difficulties in sexual function (Crouch, 2008). In our opinion future research should be directed to establishing standardized methods of functional assessment of clitoris in operated women. It should be also noted, that clitoromegaly appears mainly due to insufficient hormonal treatment (i.e. excessive androgen production). In our patients the main reason for that was poor patient’s compliance.

The first proposition of unified scale for the assessment of results of feminizing surgery was published in 2001 by Creighton. They assigned patients to one of three categories for cosmesis: good (genitalia appear normal), satisfactory (up to two minor abnormalities, unlikely to be judged abnormal by a non-medically trained person) or poor (genitalia look abnormal; three or more abnormal features). The assessment criteria included: genital proportions and symmetry, pubic hair distribution, clitoral hood size and shape, glans clitoral shape and prominence, clitoral body size and prominence, labial positions and proportions, vaginal introital position and appearance, and genital skin quality. Anatomical dimensions were assessed against normal values for clitoral and vaginal size. Overall cosmetic result was good in 18%, satisfactory in 41% and poor in 41%. Since this study has been published, a few modifications of this scale have been proposed (Lean et al, 2005; Gupta et al, 2006; Braga et al. 2006). All of them are rather of descriptive character, none of these scales apart from Stikkkelbroeck et al. (2003) uses score points for the assessment. In our opinion scoring scale makes the evaluation more precise and the results are more comparable. For example, a very good, systematic study by Sircili et al. (2006) would be of greater value if an objective, scoring scale has been used for the assessment of outcomes.

Moreover, in most of papers “genital” part of the assessment is emphasized, whereas anatomic (urethra opening location) and functional (continence, urinary tract infections) aspects of the “urinary” part are neglected.

Data concerning the frequency lower urinary tract symptoms are few and contradictory. Davies et al. (2005) emphasize increased frequency of urge and stress incontinence after feminizing genital surgery in CAH. These data confirm the previous findings (Stikkelbroeck et al, 2003). On the other hand, Braga et al. (2006) did not observed urinary incontinence or recur-
Table 1. Age at the patients were assessed

<table>
<thead>
<tr>
<th>Age [years]</th>
<th>3–6</th>
<th>7–10</th>
<th>11–14</th>
<th>15–18</th>
<th>Over 19</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>10</td>
<td>15</td>
<td>10</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>%</td>
<td>23.3</td>
<td>34.9</td>
<td>23.3</td>
<td>9.3</td>
<td>9.3</td>
</tr>
<tr>
<td>Total</td>
<td>43</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 2. Age at patients were operated

<table>
<thead>
<tr>
<th>Age [years]</th>
<th>1–3</th>
<th>4–6</th>
<th>7–9</th>
<th>10–12</th>
<th>13–15</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>27</td>
<td>7</td>
<td>4</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>%</td>
<td>62.7</td>
<td>16.2</td>
<td>9.3</td>
<td>4.6</td>
<td>6.9</td>
</tr>
<tr>
<td>Total</td>
<td>43</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 3. The degree of masculinization according to Prader’s scale

<table>
<thead>
<tr>
<th>Prader scale</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>0</td>
<td>7</td>
<td>32</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>%</td>
<td>0</td>
<td>16.3</td>
<td>74.5</td>
<td>4.6</td>
<td>4.6</td>
</tr>
<tr>
<td>Total</td>
<td>43</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 4. Assessment Scale for anatomic and cosmetic results of surgical management in ambiguous genitalia

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>General appearance</td>
<td>abnormal</td>
</tr>
<tr>
<td>Pudendal labia size</td>
<td>markedly hypertrophic or</td>
</tr>
<tr>
<td></td>
<td>hypotrophic</td>
</tr>
<tr>
<td>Pudendal labia symmetry</td>
<td>markedly assymetric</td>
</tr>
<tr>
<td>Size and position of clitoris</td>
<td>markedly enlarged, protruding</td>
</tr>
<tr>
<td>Size of introitus</td>
<td>slit-like</td>
</tr>
<tr>
<td>Position of urethra opening</td>
<td>external ostium of urinary</td>
</tr>
<tr>
<td></td>
<td>coil invisible</td>
</tr>
<tr>
<td></td>
<td>external ostium of urinary</td>
</tr>
<tr>
<td></td>
<td>coil located in anterior</td>
</tr>
<tr>
<td></td>
<td>wall of vagina</td>
</tr>
<tr>
<td></td>
<td>normal</td>
</tr>
</tbody>
</table>

Table 5. Results of the assessment of anatomic and cosmetic results of surgical management in ambiguous genitalia in our group of patients

<table>
<thead>
<tr>
<th>Result</th>
<th>Number of patients</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>good</td>
<td>36</td>
<td>83.7</td>
</tr>
<tr>
<td>satisfactory</td>
<td>4</td>
<td>9.3</td>
</tr>
<tr>
<td>poor</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Total</td>
<td>43</td>
<td>100</td>
</tr>
</tbody>
</table>
rent UTIs, with normal bladder emptying in most of the patients and small post-void residual in 10% of them. In the study by Nabhan et al. (2006), the incidence of UTIs in children with CAH was similar to that in the general population, but the others report recurrent UTIs as complication of feminizing surgery (Krege et al. 2000; Stikkelbroeck et al. 2003). However, the relationship between urogenital sinus presence and recurrent UTIs has not been established, yet. The existence of 2 orifices was included to the general cosmetic assessment by Sirici et al. (2006), they showed the existence of the urogenital sinus in 11 from 34 operated patients. Miranda et al. (2004) were able to visualize the urethral meatus in 90.9% of patients. Braga et al. (2006) obtained separate and easy to identify in 21 of 24 operated girls. Based on these data we suggest including the position of urethra opening to the assessment scale of cosmetic results of feminizing genitoplasty. Clear and easy to apply functional criteria are urgently needed.

We conclude that taking into consideration controversial data about the results of feminizing genitoplasty, a standarized, well-defined and commonly accepted scale enabling comparison between methods and institutions is necessary. In our opinion scoring scale makes the evaluation more precise and the results are more comparable.

REFERENCES


