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Vaginoplasty and creating labia minora in children with disorders of sex development

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Abstract

Purpose To report current results of vaginoplasty using the mucosa of the prepuce, and creating labia minora by penile skin in children with disorders of sex development (DSD).

Methods In 10 years, we have performed 22 vaginoplasties using the described technique of vaginoplasty, 21 patients with 46, XX DSD and 1 ovotesticular DSD. The assessment of the results of this technique of vaginoplasty was undertaken in several stages: (a) The evaluation of the cosmetic result. (b) The research for a urinary incontinence and urethrovaginal fistulas. (c) The research for a vaginal stenosis by the introduction of a lubricated feeding tube into the vaginal cavity.
The labia minora was evaluated by three criteria: its skin should be thin and very supple, it should have a free edge which partially or totally covers the clitoris, urethral meatus, and vaginal orifice; and it is preferable that its color be darker than the rest of the skin.

Results The cosmetic outcome was considered by parents and the surgeon as very satisfactory in 11 patients (50% of cases), satisfactory in 4 patients (18.2%), and unsatisfactory in 7 patients (31.8%). The postoperative complications were five cases of proximal stenosis (22.7%), one distal stenosis (stenosis of introitus) and two necrosis of the preputial flap. No urethrovaginal fistula and urinary incontinence were reported.

Conclusions In infant and young child, when it is difficult to make use of complete urogenital mobilization, the mucosa of the prepuce can be an alternative to create a neovagina, its histological constitution is identical to a vaginal wall, and it does not prevent to have a good labia minora.

Keywords Disorders of sex development · Ambiguous genitalia · Feminizing genitoplasty · Vaginoplasty · Labia minora · Labioplasty

Introduction

We talk about ambiguous genitalia or disorders of sex development (DSD) when the appearance of the external genitalia of a new-born, does not resemble that of a girl or a boy, but rather a form between the two. And when a person has well-developed external genitalia, but the phenotype does not correspond to the genetic sex. In such cases, this is usually discovered later, at puberty [1].

In patients with 46, XX DSD and some other types of DSD, the female gender is proposed, and a surgical correction should be performed as early as possible to permit the development of a good gender identity in the patients. This surgical correction consists of reducing the size of the phallus (clitoroplasty), creating labia minora, and vaginoplasty.

The vaginoplasty and creating labia minora in patients with DSD remain a challenge for pediatric urologist. Several procedures have been described for vaginal reconstruction one among which is the use of preputial flap [2]. In this paper, we report current results of vaginoplasty using the mucosa of the prepuce, and creating labia minora by penile skin.

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Materials and methods

During 10 years (from March 2008 to March 2018), we performed 22 vaginoplasties using the described procedure, 21 patients with 46, XX DSD (2 patients had 11-hydroxylase deficiency and 19 had 21-hydroxylase), and 1 ovotesticular DSD. Two types of medical imaging were performed in all patients:

- Ultrasound in search of female genital structures (uterus, ovaries, fallopian tubes), and to evaluate the adrenal gland size.
- Genitography allowed us to show the upper vaginal cavity, and located the level where it joined the urethra (Fig. 1).

The Prader classification showed that 8 patients had grade III and 14 had grade IV–V. However, 4 patients with 46, XX DSD had a simple virilizing form and 17 had an additional salt loss.

The patients included in the study came from all regions of our country. They were hospitalized 2 to 3 days before the surgical repair to provide psychological support of parents and children. Then, they stay 7 to 15 days at the hospital after surgery. These patients were treated according to the following criteria: operated for the first time, in one stage, by a single surgeon. The age of the patients at surgery ranged from 4 to 47 months. The delay of the surgical repair encountered in some cases was due to the rarity of centers that treat this type of pathology. The median follow-up was 5 years (range 4 months to 10 years).

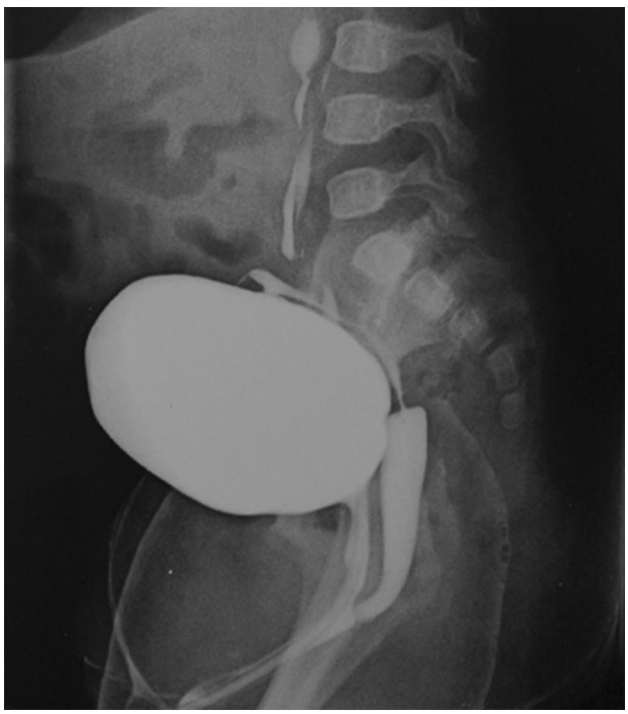

Fig. 1 Genitography, which shows a uterus, tubes, and a vaginal cavity and especially. It allowed us to locate the level where this vaginal cavity join the urethra

To avoid a possible decompensation of the adrenal insufficiency during the surgical act, the daily dose of hydrocortisone was multiplied by three times during 4 days (1 day before surgery and 3 days later). Then, the dose was gradually reduced.

The vaginoplasty by tubularized preputial flap in children with DSD was reported in 2013 (2). The patient was placed in the lithotomy position for the best exposure of the perineum. A Foley catheter (Caliber of 6 Fr) was placed into the vagina to facilitate its identification during dissection. However, in five cases, the vaginal opening was closed and the introduction of a catheter was impossible. A traction suture was placed through the glans, and an incision in the perineal area described an inverted U above the anus and extended distally to the meatus of urogenital sinus by another incision (Y-shape incision). After the circumferential subcoronal incision, the skin and dartos fascia were dissected from the shaft. The clitoroplasty was made in all cases according to the technique described some years ago [3]. This technique consists of a complete mobilization of the glans with its neurovascular bundle to achieve the necessary symmetric reduction of the length and diameter of the corpora cavernosa, as well as the volume of the glans.

The dissection and separation of the vagina from the posterior urethra wall was the most challenging part of the operation, especially in cases with high confluence. The urethra was sutured with a double layer of continuous and interrupted 4–0 polyglactin sutures. In all patients, a Foley catheter was maintained into the bladder for 7 to 14 days.

The dissection of the preputial flap with its pedicle was done after measuring the distance between the native vagina and perineum. The mucosa of the prepuce provides a flap of 12–16 mm wide and extensible. If the mucosa is very narrow, the dissection can be expanded to the outer face of the prepuce. This preputial flap was converted into a cylinder around the vaginal stent (caliber of 12 or 18). This cylinder was brought posteriorly, and its free end was sutured to the native vagina with interrupted 4–0 polyglactin sutures, and its outer end to the perineal skin. Postoperatively, the vaginal stent was left in the vagina for 5–7 days and antibiotic prophylaxis was maintained for 7 days.

Before the age of 2 years, in cases with low and intermediate vaginal confluence, the distance between the native vagina and the perineal skin is < 2–2.5 cm, and the preputial flap was sufficient. However, in high vaginal confluence, this technique was associated with partial urogenital mobilization.

Clitoroplasty and creating labia minora were realized at the same operative time with vaginoplasty, a surgical operation which can last 5 h. Usually, creating labia minora is done by the penile skin and the prepuce. However, the extensive dissection of the foreskin with its pedicle used in vaginoplasty gave a very thin penile skin. This skin often poorly vascularized was divided into two parts. Then, the distal part of each flap was folded in two and sutured to have a free edge. The clitoroplasty was achieved in all patients by complete mobilization of the glans with its neurovascular pedicle to realize a significant and symmetric reduction of the length and diameter of the corpora cavernosa, as well as the volume of the glans [3].

The evaluation of the results of this surgical technique was made in several stages:

- The search for urinary incontinence and urethrovaginal fistulas.
- Perineal examination to evaluate the aesthetic result of feminizing genitoplasty.
- Introduction of a lubricated feeding tube (caliber of 6, 8, and 10 Fr) into vaginal cavity to evaluate its permeability (Fig. 2).

Postoperatively, the quality of labia minora was appreciated by three criteria: its flexibility (it should be thin and very supple), its color (it is preferable to be darker than the rest of the skin), and its free edge which partially or totally covers the clitoris, urethral meatus, and the vaginal orifice.

Results

Bleeding was the main intraoperative complication. However, this bleeding was never been very significant, endangering the patient's life.

Postoperatively, there were necrosis of the preputial flap in two patients which were manifested immediately after the surgical act by a cyanosis of the flap and disappearance of labia minora after a few days. This redoubtable complication was due to excessive stretching of the preputial flap pedicle.

Introduction of a lubricated feeding tube into vaginal cavity revealed the presence of proximal stenosis in 5 cases (22.7%) and 1 distal stenosis (stenosis of vaginal introitus).

No urinary incontinence and urethrovaginal fistulas were noticed in all patients.

The cosmetic result was considered by parents and the surgeon as very satisfactory in 11 patients (50% of cases), satisfactory in 4 patients (18.2%), and unsatisfactory in 7 patients (31.8%):

- The 11 patients judged very satisfied had labia minora well developed with a free edge, a small apparent part of glans (length < 5 mm), an area "A" (Fig. 3) covered by a red mucosa and orifices (vaginal and urethral) in place.
- The seven patients with unsatisfactory aesthetic result, had in five patients a perineal skin in zone "A," a voluminous glans in 4, and absence of labia minora with voluminous glans in 2.

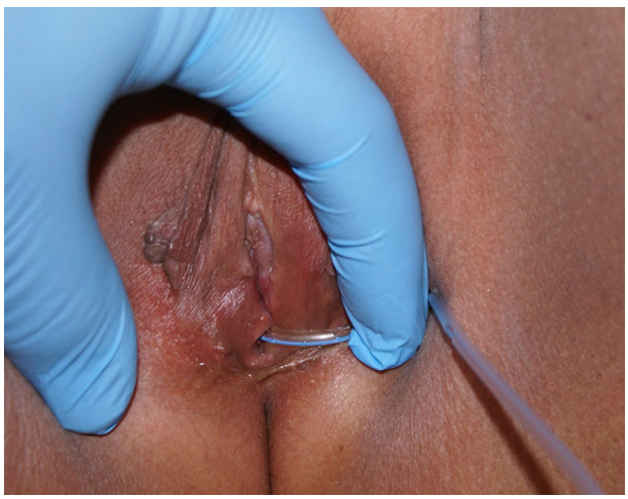

Fig. 2 Introduction of a lubricated feeding tube (caliber of 6, 8, and 10) into vaginal cavity looking for proximal stenosis

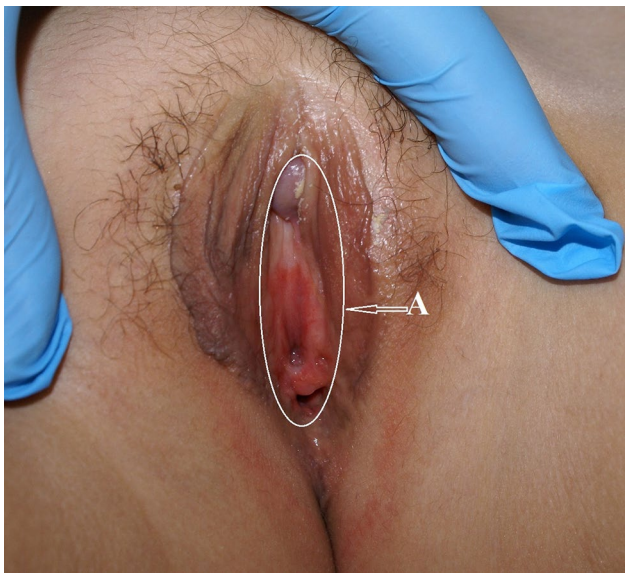

Fig. 3 The area "A" located between two labia minora should be covered by the wall of the common urogenital canal, never by a perineal skin

– All four patients with satisfactory result had the absence of labia minora and apparent part of glans < 10 mm.

Discussion

Patients with DSD which require vaginoplasty could be divided into two groups:

– Patients with vaginal agenesis: the complete androgen insensitivity syndrome and some cases of 46, XY DSD as Leydig cell hypoplasia type 1 (a rare malformation characterized by female external genitalia with complete regression of Müllerian duct: absence of fallopian tubes, vagina, and uterus). These forms are often discovered later, in older children or at puberty, by clinical signs as such inguinal hernia; virilization or delayed of primary amenorrhea in a girl; and breast development or apparition of cyclical haematuria in a boy. Thus, the timing of vaginoplasty does not arise. The first recommended treatment of vaginal agenesis is the vaginal dilatation (Frank's technique modified by Ingram), and the surgery should only be proposed if the dilatation fail or if it is impossible to do because of perineal scarring from previous surgical procedures [4]. Also, vaginal dilatations are often necessary after surgical repair. The surgical techniques that can be used are McIndoe technique, Vecchietti, Williams and vaginal replacement (by an intestinal tube, peritoneum or Davydov procedure, amniotic membrane, buccal mucosa …).
– Patients with persistent Müllerian duct: 46, XX DSD, ovotestis DSD, mixed gonadal dysgenesis, and 46XY gonadal dysgenesis, are often discovered at birth and are characterized by the presence of internal genital structures (uterus, fallopian tubes, and upper vagina) with external genitalia which does not resemble neither to a girl or a boy, but rather a form between the two. The surgical correction of these forms of DSD should be performed as early as possible to allow good development of the gender identity. This repair includes clitoroplasty, creating labia minora, and vaginoplasty. The aim of the vaginoplasty is to separate the openings of the vagina and urethra, and to connect the vaginal cul-de-sac to perineal skin.

Traditionally, the choice of surgical procedure was based on the location of the urogenital confluence. When the vaginal confluence was low, a simple Y-V vaginoplasty described by Fortunoff et al. [5] was performed. And, when it was high, the pull-through operation of Hendren and Crawford [6] was achieved, usually in two stages. However, these two techniques widely used in the past are often complicated by stenosis of the vagina [7–9] and periodic dilatations are required. Those dilations can last for years and are often complicated by scarring and fibrosis, which can subsequently affect the sexual life of the woman and make a surgical reoperation difficult. To avoid these problems, many authors prefer delaying vaginoplasty until puberty [9]. The preferred technique for a large number of centers is the total urogenital mobilization described initially by Peña [10] for cloacal anomalies repair. In 1999, Ludwikowski et al. [11] applied this concept to repair the ambiguous genitalia associated with congenital adrenal hyperplasia. It is an excellent procedure which gives a very good appearance of external genitalia. A surgical technique which we have used in the cases with low confluence. However, in infant and young child, this procedure provides only a gain of 1 to 1.5 cm, at the cost of a reduction of the length of the urethra which can be responsible for urinary incontinence at adulthood. Another procedure which deserves mention is the technique described by Passerini-Glazel [12]. We have used this surgical technique in some patients with low or intermediate confluence when the prepuce was small.

A high rate of poor cosmetic results and difficult sexual intercourses in women underwent feminizing genitoplasty for ambiguous genitalia during their childhood were reported [13]. Thus, it was clear that the clitoroplasty, creating labia minora and vaginoplasty in childhood need to be improved.

The use of foreskin provides us a lot of advantages. It is a tissue identical to a true vaginal wall, a stratified squamous epithelium non-keratinized. Thus, the wall of the neovagina contains no hair, no pilosebaceous glands, and no adipose tissue under the skin (a tissue which can increase the risk of fibrosis). In addition, the foreskin is very rich in nerve endings with the presence of Taylor's bands, very erogenous area.

The cosmetic result depends on three important criteria: the volume of the glans, the presence or not of the labia minora and its appearance, and the nature of the tissue used in covering the area located between two labia minora. This area, which we call area A (Fig. 3), should be covered by the wall of the common urogenital canal, never by perineal skin. We think that the presence of a labia majora does not have the same importance in the cosmetic result than the labia minora.

Proximal stenosis was the most frequent complication encountered in patients. We believe that this complication does not need immediate dilatations. It will be done automatically at the age of sexual activity. Proximal stenosis and necrosis of preputial flap can constitute a disadvantage to the use of this surgical technique, if the causes of proximal stenosis remain unknown. To avoid necrosis and loss of the preputial flap, and the disappearance of the labia minora, the dissection of the flap should be meticulous and it should to avoid pulling too much on the preputial flap pedicle. The rate

of distal vaginal stenosis was very low, developed in only one case, it was loose, and easily treated by a single dilation.

The majority of authors prefer to realize the vaginoplasty during adolescence. However, we think that with this procedure, the vaginoplasty can be accomplished during early infancy, at the same time with clitoroplasty and creating labia minora, preferably between 6 and 20 months.

Conclusions

The total urogenital mobilization remains the best procedure for the treatment of the persistent urogenital sinus. However, the mucosa of the prepuce which constitutes a good tissue to create a vagina could be an alternative to create a neovagina. Particularly, in infant and young child.

Compliance with ethical standards

Conflict of interest All authors declare that they have no conflict of interest.

Ethical approval This study conforms to the World Medical Association Declaration of Helsinki (June 1964) and subsequent amendments and the investigations were carried out to a high ethical standard. However, in our country, there is not a formal and documented ethical approval from an appropriately constituted Research Ethics Committee, which should be obtained for all studies involving human.

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