



Feminizing Genitoplasty for Children with Disorders of Sex Development

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Abstract

The purpose of this work is to present two new surgical techniques used for feminizing genitoplasty:

Clitoroplasty: We developed and used this technique since 2004. This technique consists of a complete separation of the glands with its neurovascular bundle; this allows us to achieve an important and symmetrical reduction of the length and diameter of the corpora cavernosa, as well as the volume of the glands.

We have also developed a new technique of vaginoplasty, which is more physiological than previous techniques. In this technique, we use the inner surface or mucosa of the prepuce, a similar tissue to that of a true vaginal wall (non keratinized stratified squamous epithelium).

Both surgical techniques can be used for feminizing genitoplasty of children with 46, XX DSD, or ovotesticular DSD (true hermaphroditism), or mixed gonadal dysgenesis or 46, XY gonadal dysgenesis (dysgenetic male pseudohermaphroditism). These techniques are simple to carry out with good final cosmetic aspects and fewer complications.

Keywords

Disorders of sexual development; Ambiguous Genitalia; Feminizing Genitoplasty; Clitoroplasty; Vaginoplasty; Ovotesticular DSD; Mixed Gonadal Dysgenesis; 46,XY Gonadal Dysgenesis; Dysgenetic Male Pseudohermaphroditism

Introduction

A patient is diagnosed with disorders of sex development (DSD), when his/her phenotype resembles neither that of a boy, nor that of a girl, but it is a form between the two. But this definition is incomplete, since there are disorders of sex development where the external genitalia are well trained, but the phenotype does not match the genetic sex. These are forms late-onset, during the pubertal spurt. In four types of sexual ambiguity: 46, XX DSD (female pseudohermaphroditism), ovotesticular DSD (true hermaphroditism), mixed gonadal dysgenesis and gonadal dysgenesis 46XY (pure gonadal dysgenesis), the female gender is proposed for these children and the feminization of external genitalia or feminizing genitoplasty is performed after the birth. It includes three stages:

- Clitoroplasty: reducing the size of the clitoris or the penis

- Vaginoplasty: Creating a vaginal cavity
- Creating labia minora

In the feminizing genitoplasty, we use two new techniques: Clitoroplasty technique [1] and a technique of vaginoplasty [2].

Clitoroplasty

Surgical correction of an enlarged clitoris should be performed as soon as possible, to allow for a good development of the child's sexual identity. The aim of surgical correction is to create a female phenotype corresponding to genre. In the past, the clitoral hypertrophy was treated by clitorodectomy (excision of the clitoris) [3,4]. However, the gland is an important organ for the erotic sensitivity of the woman, and its ablation is a mutilating and illogical practice. So the concern of surgeons was to develop a technique that permits reducing the size of the phallus while preserving a sensitive gland.

The first Clitoroplasty was described by Lattimer in 1961 [5]. The principle of this technique is to dissect the corpora cavernosa which are thereafter buried in a tunnel created under the skin. However, this technique does not correct the malformation but hides it and it is responsible for pain during erection at puberty [6]. The technique of choice used in a large number of centers is that which has been described in 1974 by Kumar et al [7]. This technique consists of a partial resection of the corpora cavernosa after release of the neurovascular pedicle of the gland, followed by restoration of continuity by suturing the edges by non-resorbable fine wire. But this technique is responsible of a unacceptable cosmetic appearance, a voluminous glands, disproportionate with its environment. And in both techniques, there is no reduction in the diameter of the corpora cavernosa which gives a voluminous appearance to the clitoris during the erection. Several other techniques have been proposed to reduce the apparent volume of the glands [8,9].

Recently, two new techniques have been described (a few months apart) for the surgical correction of a voluminous clitoris. Both techniques have in common the complete separation of the glands with its neurovascular bundle. Our technique [1] and the technique of Pippi Sale et al. [10] who places the corpora cavernosa around the vaginal orifice and the glands is repositioned at the bifurcation of the corpora cavernosa.

Description of our technique: The innervations and vascularization of the penis are provided by two neurovascular pedicles which pass longitudinally under the Buck's fascia, at the 1-and 11-o'clock positions. In the glands, the nerves and the vessels form a network around of the distal part of corpora cavernosa. Gearhart et al. [11] demonstrated that electromyography genital response in the glands after stimulation of the neurovascular bundle remains intact after releasing the pedicle and reducing the length of the corpora cavernosa.

A traction suture with wire (4/0) is placed through the glands. After the circumferential sub coronal incision, the entire phallus skin envelope is retracted. The penile base is surrounded by a rubber band, which serves as a tourniquet. Next, the dorsal side of the corporeal bodies is exposed. Two parallel longitudinal incisions of Buck's fascia are made at the 2- and 10-o'clock positions until the tunica albuginea

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Received: April 22, 2015 Accepted: April 24, 2015 Published: April 30, 2015

is seen. To keep in contact with this area, fine scissors are placed behind Buck's fascia to lift it up with the dorsal neurovascular bundle. The dissection needs to remain directly on top of the tunica albuginea to prevent neuronal and vascular injury. This dissection must be extended proximally and distally.

When the entire pedicle is released, the glands can be separated easily and safely of the corpora cavernosa, and remains attached only by its pedicle. The complete release of the glands and its neurovascular bundle greatly facilitates the surgical act (Figure 1). It allows us to realize an important and symmetrical reduction of the length and diameter of the corpora cavernosa by resection after labeling of internal and distal part of these bodies. Then we reunite what remains of two bodies by two running suture.

The released gland in the form of a cap is often voluminous and requires a reduction of its size: a longitudinal incision made in the midline of the anterior wall, allows us to make a harmonious reduction by excision of the edges. Subsequently, the glands is reconstructed and fixed to the end of the corpora cavernosa by using a fine wire (6/0).

Since 2003, we have used this technique in 28 patients; we deplore a case of necrosis of the glands. Since, we give great importance to the position of the pedicle. We also had 4 atrophy of the glands, but unintentionally, the atrophy of glands enters in the goal of treatment.

We think that with this new surgical technique, we can obtain excellent cosmetic appearance of the external genitalia.

Vaginoplasty

Vaginoplasty remains a challenge for the pediatric surgeon and several techniques have been described to create a neo-vagina. Some of these techniques (use of an intestinal loop, free thin skin or total skin, with or without tissue expansion, amnion and oral mucosa) are generally indicated for vaginal agenesis or atresia. It is not the case of certain forms of ambiguous genitalia: 46, XX DSD (female pseudohermaphroditism), ovotesticular DSD (true hermaphroditism), mixed gonadal dysgenesis and gonadal dysgenesis 46XY (pure gonadal dysgenesis), where the regression Muller channels do not occur and the internal female structures are present (uterus, fallopian tubes and upper vagina that opens into the urethra).

So, the objective of any vaginoplasty is to separate the vagina of the urethra with creating a new vagina which opens in the normal position in the perineum. This neo-vagina should allow the patient to have in adulthood a normal sex life, a normal menstrual cycle, and be able to give birth by natural way without problems.

Traditionally, the choice of the technique of the vaginoplasty for congenital adrenal hyperplasia dependent on the location of the urogenital confluence: YV plasty described by Fortunoff [12] was used in the lower forms, and in higher forms, the Pull-through described Hendren and Crawford [13] using shreds of perineal skin, usually in two stages. However, these techniques are often complicated by stenosis [14] and periodic dilations are needed. The majority of patients who have undergone these techniques are not satisfied with their body image, and suffer from anxiety during intercourse with orgasm problems [15]. The repeated vaginal dilations for stenosis in childhood are causing scarring and fibrosis, which can compromise a possible surgical reoperation and sexual life of these patients in adulthood. To avoid these problems, many authors prefer to delay vaginoplasty until puberty [15,16].

Another technique has had great success, the technique described by Paressini-Glazer [17]. But it seems that this technique gives vaginal stenosis in a third of cases [18] with a high rate of urethral fistulas

In 1999, Ludwikowski et al. [19] propose the use of the mobilization of the urogenital sinus in children with adrenal hyperplasia, described by Peña two years rather [20] in the treatment of malformations of the cloaca. Currently, this method (total or partial mobilization of the urogenital sinus) is the preferred technique of the majority of centers. But, the gain with a total mobilization of the urogenital sinus, cannot exceed 1.5 cm, and therefore it cannot be used alone in case of high form. In addition, the lowering of the urogenital sinus toward the perineum carried with it the neck of the bladder and the external urethral sphincter. This may constitute a long-term risk on urinary continence of these patients in adulthood.

In 1990, Gonzalez and Fernandez [21] have described a technique that uses a flap consisting from skin and prepuce of the clitoris to construct the anterior wall of the vagina, while the back and side walls are formed by a flap of skin taken from perineum. However, the use of the skin of the clitoris, in which two holes is practiced at the midline to pass the glands and urethral meatus, gives a bad cosmetic appearance of the external genitalia and this technique is used very little.

The age of vaginoplasty remains controversial. I think that with this technique, even in severe virilization of the external genitalia, the feminizing genitoplasty with vaginoplasty should be performed in a single stage at an early age of life for the following reasons: The birth of a child with ambiguous genitalia is a tragedy for the parents and prolong the suffering is unjustifiable, the missing part of the vagina is not very important in infants, early correction of the genital malformation is important for a good psychosexual development and before 4 years, the vessels surrounding the corpora cavernosa which come from dorsal pedicle are not yet developed. A long-term follow-up of these patients will be essential to reassess the results of this technique in adulthood.

Description of our technique

We think that the vaginoplasty must be performed simultaneously with the Clitoroplasty at an early age and in one-stage. Therefore, after Clitoroplasty, a Foley catheter F6 was placed in the vagina to facilitate identification of structures during dissection, sometimes the vaginal opening was closed and the introduction of a catheter was impossible. The incision is shown in (Figure 2). The dissection and separation of the vagina from the urogenital sinus is the most challenging part of the operation, especially in cases with a high confluence, which can be made more difficult by limited exposure. The urethra opening is closed with interrupted sutures (6-0 polyglactin) and a urinary stent is maintained in the bladder for 5-7 days.

The dissecting a flap from the mucosa of the prepuce with its neurovascular bundle must be done after measuring the distance between the native vagina and perineum. This flap, Wide normally 14 to 18 mm and extensible, can be extended to the external surface of the prepuce if the mucosa is very narrow. Then it is tubulated on a rectal probe (F18 or 20). The proximal edge of the prepuce of the tube is sutured to the upper vaginal and its distal edge to the perineal skin. In case of low urogenital sinus (less than 2 cm), the use of a single preputial flap is often sufficient. However, when the urogenital sinus is located very high, this technique is associated with a total mobilization of the urogenital sinus. The rectal probe placed in the



Figure 1: Clitoroplasty: The complete release of the glands and the neurovascular bundle allows us to achieve a significant and symmetrical reduction of the length and diameter of the corpora cavernosa, and the volume of the glands.



Figure 2: Vaginoplasty: With this technique the number of vaginal stenosis is low compared to older techniques.

new vagina is cut at 10 mm from the vagina and left 3 to 5 days postoperatively.

At the end of the intervention, the remainder of the clitoral skin is used to create the labia minora according to the method of Bailez et al. [22].

This technique of Vaginoplasty is more physiological than other techniques because of the use of a mucosa similar to that of a true vaginal wall, a non-keratinized stratified squamous epithelium, a wall which does not contain pilosebaceous follicles, which gives it an advantage compared to the perineal skin that contains hair, sebaceous glands and subcutaneous tissue, source of fibrosis. In addition, the foreskin is rich in endings erogenous nerve [23] and contains Taylor's striated bands, an area very erogenous [24].

This simple technique in its realization responds to goal of treatment and gives few complications. It can be used in the feminizing genitoplasty of young children with 46, XX DSD (female pseudohermaphroditism) ovotesticular DSD (true hermaphroditism), mixed gonadal dysgenesis and gonadal dysgenesis 46XY (pure gonadal dysgenesis), alone or combined with total mobilization of the urogenital sinus (in case of very high form).

Conclusion

We think that with both techniques, the surgical correction of DSD has become simpler in its realization, gives few complications and responds to the goal of treatment.

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