Surgery for Female Ambiguous Genitalia

Neonatal management of female intersex
by clitorovaginoplasty

CHAPTER 5

Tom P.V.M. de Jong, Thomas M.L. Boemers

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Abstract

Historically, in female pseudohermaphrodites a staged procedure with early clitoral reduction and delayed vaginoplasty was often the treatment of choice. In recent years several authors have described 1-stage genitoplasty that is performed in the first year of life. The 1-stage procedure for clitoroplasty and vaginal exteriorization is preferable for an optimal functional and cosmetic result. Because of emotional aspects of the family in intersex cases, neonatal reconstruction offers advantages to the child and parents. Furthermore, the perinatal genital hyperstimulation by maternal and placental estrogens that persists into the first 3 to 4 weeks of life produces vaginal enlargement by mucous secretion and vaginal wall hypertrophy, facilitating identification of the cleavage planes and vaginal pull-through. These arguments have led us to treat these patients by neonatal 1 stage clitorovaginoplasty. Cosmetic and functional results in 5 consecutive cases have been excellent without serious complications.

Introduction

Many techniques for clitorovaginoplasty have been reported, mostly accompanied by advice concerning the timing of the operation. Historically, in moderate to high ending vaginas many advocated early clitoroplasty to ameliorate the cosmetic appearance of the external genitalia, followed by vaginoplasty at a later age. Recently, others proposed a 1-stage procedure with relatively early operation in the first year of life. An argument for a 1-stage procedure is that early clitoroplasty can compromise the cosmetic result of later vaginoplasty. Furthermore, some patients have concomitant urinary tract problems based on the urogenital sinus that make early intervention preferable, especially in cases of hydrometrocolpos. In 5 consecutive cases that presented immediately after birth we opted for primary reconstruction in the first weeks of life.
Material and methods

Five virilized female neonates (4 with the adrenogenital syndrome and 1 true hermaphrodite) were treated by clitoroplasty and vaginal exteriorization. Patient age at operation was 1 to 3 weeks. All patients were referred with ambiguous genitalia immediately after birth. In 3 cases the diagnosis of a probable female gender and the adrenogenital syndrome was made at referral by ultrasound and retrograde genitography, and in 1 the diagnosis of the adrenogenital syndrome was confirmed by laparoscopy and endoscopy of the urogenital sinus with retrograde vaginogram. The diagnosis adrenogenital syndrome was confirmed by biochemical data in all cases. A normal 46XX karyotype was noted in these patients. Patient 5 had XO/XY mosaicism on karyotype, and an ovotestis and a streak ovary were removed by laparotomy while she was under anesthesia for genital reconstruction.

All patients underwent surgery using a perineal approach (fig.1). Clitoroplasty was started with circumcision and degloving of the clitoris, followed by the mid shaft lateral incision of Buck’s fascia to create a cleavage plane with the tunica albuginea, thus permitting the dissection of Buck’s fascia (containing the neurovascular bundles) and the glans clitoris off the corpora cavernosa (fig. 2). Subsequently resection of the corpora up to the caudal end of the symphysis past the bifurcation. The spongiosum tissue of the remaining crura of the corpora was destroyed by dilatation with a 2 mm. metal sound to avoid later painful erections. The stumps of the corpora were closed by a running 5-zero polyglycolic acid suture. When necessary, the glans was reduced by wedge excision and/or partial de-epithelization and was sutured to the stumps of the corpora, providing a clitoris in the physiological position at the caudal end of the symphysis. Before vaginal exteriorization a Fogarty catheter was inserted endoscopically into the vagina and a balloon catheter was placed into the bladder. After opening of the perineum with a midline incision that continued into an inverted U-flap above the anus (fig. 1, A), the urogenital sinus was followed proximally until the Fogarty catheter balloon could be palpated (fig. 1, B). The vaginal tip was opened and the anterior vaginal wall freed from the urethra. The opening of the vagina into the urethra/urogenital sinus was closed with 6-zero polyglycolic acid suture,
Figure 1: A. skin incisions are made according to the dotted lines. B. urogenital sinus and vagina are incised according to dotted lines after inserting a small caliber balloon catheter into the vagina endoscopically. C. the situation after closure of the vaginal ending in the urethra/urogenital sinus. D. end-result: the anterior and lateral vagina is sutured to the minor labia consisting of phallic skin. The dorsal vagina is sutured to the inverted U-flap.

Figure 2: A. longitudinal section of phallus and symphysis. After degloving of the phallus Buck’s fascia with the neurovascular bundles is removed from the corporal bodies. B. the corporal bodies are resected up till the bifurcation at the caudal end of the symphysis, the corporal stumps are closed after destroying the remaining erectile tissue with a metal sound. C. after reduction of the glans this is sutured to the remaining stump of the corporal bodies.
leaving a redundant length of urethra (figure 1. C). The urogenital sinus was partially opened until a urethra remained that was rather long for patient age. The open strip of the urogenital sinus epithelium was sutured cranially to the clitoris and caudally to the anterior vaginal wall. The phallic skin is remodeled to provide a preputial hood proximally as well as labia minora, which are distally sutured to the lateral vaginal wall. The inverted U-flap was then sutured to the dorsal vaginal wall. The labial folds are reduced and transposed posteriorly to create normal appearing labia majora.

Results

Convalescence was uneventful in all cases. With proper endocrinologic care no electrolyte or fluid complications developed in the post-operative period. Cosmetic appearance is considered excellent in all cases. Follow-up in the 5 cases is 28, 26, 12, 11 and 9 month, respectively. One patient underwent meatomy of the urethral meatus at 6 and 12 months after reconstruction with an uneventful followup of 16 months after the last meatomy. In the other patients, a 10F catheter passes the urethral meatus and a 14F catheter passes the vaginal orifice without resistance and any need for dilatation.

Discussion

Vaginal exteriorization in neonates seems to be relatively easy compared to reconstruction at a later age. A transtrigonal approach was not necessary in our patients although in 2 the vaginal ending into the urethra was considered to be high (Prader IV to V). Others who described this approach in these procedures indicate that reconstruction can be done early\textsuperscript{1,3} but should not be done in high ending vaginas.\textsuperscript{2} However, to our knowledge the results of neonatal procedures have not been previously published. The reason for easy vaginal exteriorization is
the neonatal hypertrophy of the external and internal genitalia at birth, based on maternal and placental hyperstimulation with estrogens. Indeed, all of our patients had a distended vagina filled with whitish mucous (fig. 3). This effect can probably be used to advantage during the first 2 weeks of life. There is no sign whatsoever for less perfect results in neonatal reconstructions (fig. 4). The psychological advantage to the parents and children is enormous, while there are no signs of greater risk for these patients with current pediatric anesthesiological procedures. In contrast, in the same period we operated on 14 similar patients aged 4 years or older, and the patients and parents had severe psychological problems. Of these patients, 10 experienced considerable urinary tract morbidity that has partly been based on the untreated urogenital sinus. Four patients had recurrent urinary tract infections, 2 complained of dysuria and urge

Figure 3: retrograde genitography in a neonate with urogenital sinus showing a dilated vagina with a mucous plug.
incontinence, and 2 had dribbling incontinence after voiding. These complaints subsided after reconstruction. Furthermore, vesicoureteral reflux probably unrelated to the urogenital sinus was present in 1 patient and infravesical obstruction resulting in a giant bladder diverticulum was present in 1. Both anomalies were treated during reconstruction without morbidity. The percentage of urinary tract complications in this older group is high compared to the 35% described by Lobe et al. In several older patients with adrenogenital syndrome and a high ending vagina proximal to the external sphincter who underwent surgery by other methods there has been stress incontinence due to an extremely short urethra after reconstruction. A review of these cases indicates the origin of incontinence: to create a normal vaginal introitus the short urethra was mobilized up to the level of the bladder neck to pull the orifice in level with the introitus. This procedure seems to have resulted in some damage of the sphincteric mechanism and lack of adequate functional urethral length after primary reconstruction. Our procedure as described provides a urethra with normal or redundant length for age without the need of mobilizing the proximal urethra. We hope that this adequately prevents incontinence based on a short functional urethra, even in high ending vaginas. In our first 2 patients normal daytime continence developed.

Figure 4: A. pre-operative photograph of a patient with adrenogenital syndrome and high ending vagina. B. postoperative view of the same patient. C. the same patient 8 months after operation.
However, followup is too short to make definitive statements on this aspect. In conclusion, neonatal 1-stage reconstruction seems to be the most feasible approach to treat these patients. It will take many more years to prove definitely that the benefits of an early operation are maintained throughout puberty.

Postscript: The follow-up of the girls described in this article nowadays ranges between 7 and 10 years. They experienced not a single urinary tract complication over these years. Many other patients have been operated using this scheme with similar results. This paper has been the first publication worldwide that propagates neonatal reconstruction of female intersex patients. A survey on the treatment of female intersex patients was done by interactive panel discussion at the combined meeting of the European and the American Pediatric Urological meeting in Tours, France in June 2000. Twenty percent of the pediatric urologists and pediatric surgeons have adopted our policy and treat the children immediately after birth. Of six papers on this subject, published in leading journals since 1996, this article has been cited in 5.
Literature
