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Does common channel length affect surgical choice in female congenital adrenal hyperplasia patients? ☆

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Abstract *Objective:* Partial/total urogenital sinus mobilization (UGSM) is one of the recommended techniques for treatment of female congenital adrenal hyperplasia (CAH). In this study we compared the length of common channel (CC) and type of operation performed in CAH patients.

Patients and methods: We retrospectively analyzed data of patients receiving surgery for female CAH. Patients were separated into three groups: group 1 had partial UGSM, group 2 had total UGSM, and group 3 had total UGSM plus the vaginal anterior wall was made from CC. Age at surgery, length of CC, surgical time, follow-up time, and complications were compared.

Results: There were a total of 29 patients. For groups 1, 2, and 3, the average age at surgery was 47.2 months, 14.4 months, and 21.3 months, respectively, and the average CC length was 1.25 cm, 3.1 cm, 4.3 cm, respectively. The average time of surgery was 165 min, 193.1 min, 282.5 min, respectively. The average follow-up time was 34.7 months, 36.3 months, 28.3 months, respectively. There were two complications (UGS flap necrosis and opening of sutures) in the third group.

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Conclusion: We advise the use of partial UGSM for CC of 0.5–2 cm, total UGSM for CC of 2.5–3.5 cm, and total USM with use of CC as the anterior vaginal wall in CC \geq 4 cm in length. Good cosmetic and functional results are obtained with this approach.

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Introduction

Congenital adrenal hyperplasia (CAH) is an autosomal recessive disorder of cortisol biosynthesis. It leads to phenotypic changes due to excessive endogenous androgen production during fetal development in females. Characteristic findings are an enlarged clitoris, partly fused and rugose labia majora, and a common urogenital sinus in place of a separate urethra and vagina [1].

Examination of the external genitalia reveals the degrees of virilization. Prader stages I–V describe increasing verification from a phenotypic female with mild cliteromegaly (stage I) to a phenotypic male with glandular hypospadias or normal penis appearance (stage V) [2].

Cliteroplasty, vaginoplasty, and labioplasty are the components of feminizing genitoplasty surgery of female CAH patients [3,4]. However, the surgical treatment of female CAH patients is still challenging and at times controversial. After Pena [5] described total urogenital sinus mobilization (UGSM) in patients with cloaca in 1997, partial/total UGSM became one of the recommended techniques in CAH. The urogenital sinus and vaginal confluence are described as being “high” or “low” according to the location when compared to the external sphincter, and vaginal “high insertion” is accepted as an urethral length of 1.5–2 cm [6]. Despite no objective criteria accepted as standard in the literature, these cystoscopic findings are important to determine the type of surgery that will be performed.

The aim of this study was to compare the length of common channel (CC) and the type of UGSM operation that was performed in female CAH patients.

Patients and methods

The clinical and operative files of 29 patients who underwent surgery for female CAH due to 21-hydroxylase deficiency between October 2008 and May 2013 at our institute were retrospectively analyzed.

CC lengths of all patients were measured by cystoscopy prior to major genital surgery. Patients that were diagnosed with CAH during the newborn period had cystoscopy at 6–7 months and total/partial UGSM around 1 year of age, whereas patients with late diagnosis (over 1 year) had cystoscopy and major genital surgery soon after diagnosis.

When retrospectively analyzed, the patients were found to have had three surgical methods performed (PUSM, TUSM, or TUSM plus CC as the vaginal wall) and were therefore divided into three groups for comparison of CC length. The age of patients was recorded and surgery times were analyzed. The short-term results and complications were also recorded.

Surgical techniques

Measuring the common channel

Before cystoscopic examination or UGSM, all patients received medication for stress coverage. In the lithotomy position, cystoscopy is performed. The CC is examined and the location of the vaginal confluence noted. To measure the CC, the cystoscope is advanced into the vagina. A 3F ureter catheter is inserted through the cystoscope. The cystoscope is reintroduced and performed alongside the catheter. The catheter is adjusted so that the first centimeter mark is at the level of the confluence. The length of the CC, the distance from the vaginal confluence to the perineal meatus, is measured as the cystoscope is withdrawn. A Foley catheter is introduced into the bladder. A Fogarty catheter is placed as a guide in the vagina in preparation for UGSM.

Cliteroplasty

The operation starts by placing a traction suture in the glans. A subcoronal circumferential incision is then made 0.5 cm proximal to the glans. The shaft skin is degloved and dorsal foreskin is split longitudinally; the flaps obtained are used for construction of the labia minora later. The dorsal neurovascular bundle and the glans are separated from the corporal body as described by Poppas et al. [7]. The CC is transected from the corporeal bodies by bipolarcauthery. Corporal bodies at 1.5–2 cm distal to the bifurcation are sutured and distal parts of corporeal bodies are resected by cautery. According to the age of the patient and the size of the glans, glans reduction is done either by wedge resection or by de-epithelization. The glans is settled onto the rest of the corporeal bodies by sutures.

Partial UGSM

We begin with a midline and U-shaped perineal flap incision. After cliteroplasty described as above, CC dissection is performed circumferentially to the pubic bone. Anteriorly we reach the pubourethral ligament but do not dissect it. Posteriorly the Fogarty catheter balloon is palpated. The CC is opened at the 6 o'clock position to the vaginal confluence. The posterior vaginal wall is opened and the U-flap skin is sutured. The CC mucosa is tailored to the clitoris by sutures (Fig. 1).

Total UGSM

In this group, for further mobilization when PUSM is not sufficient, the pubourethral ligament is dissected anteriorly

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