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Secondary vaginoplasty for disorders for sexual differentiation: Is there a right time? Challenges with compliance and follow-up at a multidisciplinary center

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Abstract *Purpose:* We present the experience of a multidisciplinary center for disorders of sexual differentiation (DSD) in treating females requiring vaginoplasty. Specifically, we evaluate outcomes and compliance with follow-up protocols in patients undergoing secondary vaginoplasties.
Materials/methods: We retrospectively reviewed consecutive DSD patients who underwent feminizing genitoplasties in 2006—2010. A subset of patients were instructed in vaginal self-dilation at time of secondary vaginoplasty. Through follow-up visits and administered questionnaires we assessed outcomes, compliance and overall satisfaction.
Results: Twenty-two feminizing genitoplasties were completed during the study interval. There were no postoperative complications; average blood loss was 74 ml and mean length of stay was 3 days. Ten females underwent secondary vaginoplasty. The response rate to questionnaires was 5 of 9. Age of initiation for self-dilation ranged from 8 to 24 years. Initial compliance two months postoperatively was good, but diminished 12—24 months after surgery. Responses to the quality-of-life questionnaire were diverse, reflecting a range of patient ages and varied experiences.

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Conclusion: A multidisciplinary, comprehensive approach is necessary to care for patients with DSD due to psychosexual, medical and reconstructive concerns. A secondary vaginoplasty to facilitate menarche and psychosocial concerns is feasible with minimal complications, though compliance and toleration with regard to post-surgical dilation regimens is variable.

Introduction

Children with congenital abnormalities such as ambiguous genitalia from congenital adrenal hyperplasia (CAH) and other disorders of sexual differentiation (DSD) present a complex set of medical and surgical challenges. The incidence of such anomalies varies per report from 1 in 4000 to 1 in 14,000, as definitions of ambiguous genitalia differ [1,2]. The most common of these diagnoses, CAH, occurs in approximately every 1 in 16,000 births [2]. Other diagnoses may include androgen insensitivity syndrome, Mayer–Rokitansky, and mixed gonadal dysgenesis. Despite the infrequency of such anomalies, the pediatric urologist must be aware of the variety of presentations and the appropriate treatment for each. Some of these children require early evaluation and stabilization after birth due to steroid deficiencies. Then, depending on severity, complex urogenital reconstruction may be considered.

All children diagnosed with a DSD, either based on physical exam or newborn screening results, are treated through a multidisciplinary center for DSD at our institution, which includes endocrinology, genetics, psychology and urology. After initial evaluation and stabilization, these children are often evaluated by exam under anesthesia, including cystoscopy, vaginoscopy and possibly genitography as clinically indicated. Surgical correction, if agreed upon by all members of the DSD team and the parents, is generally performed after 6 months of age [3].

The primary goal of vaginoplasty during feminizing genital reconstruction is to construct an adequately sized and appropriately situated vagina with normal-appearing female external genitalia [4]. Historically, these procedures were performed as two separate, staged procedures. Since the 1970s, surgical management of CAH has achieved these goals in a one-staged procedure during infancy [1]. Dictums such as never removing the clitoris and careful preservation of the neurovascular bundles for sensation may improve surgical outcomes. Surgical techniques have evolved, and it is hoped that better outcomes will ameliorate psychosexual concerns, though long-term complications such as vaginal stenosis may occur. There has also been no consensus on the ideal time for secondary intervention when post-surgical vaginal stenosis manifests.

Herein, we present our experience in the surgical management of a consecutive cohort of DSD females undergoing reconstruction, and seek to understand the outcomes. We specifically investigated the compliance of those females performing vaginal dilations utilizing our teaching protocol after delayed or secondary vaginoplasty.

Patients and methods

The institutional review board approved this study. The DSD database of our center was retrospectively interrogated for all female patients undergoing either primary or secondary intervention between January 2006 and December 2010. An interdisciplinary team using standardized guidelines for medical management examined all patients, and recommended reconstructive surgery as needed after extensive consultation with the family in association with a specialist in human sexuality, social work, genetics and urology.

Each patient selected to undergo surgical reconstruction underwent either genitography or endoscopy under anesthesia prior to reconstruction. The surgical technique for repair, length of hospital stay, blood loss, complications and surgical outcomes were reviewed.

Procedures performed included partial mobilization of urogenital sinus with endoscopic evaluation prior to defining anatomy – low versus high urethrovaginal confluence; clitoroplasty with preservation of neurovascular bundles in select cases; vaginoplasty; urethroplasty and perineoplasty as indicated. These procedures utilized a combination of redundant urogenital sinus tissue or non-hair-bearing posterior skin to complete a flap vaginoplasty.

Patients undergoing delayed or secondary vaginoplasty were provided additional information and counseling prior to surgery by the multidisciplinary team. These patients received preoperative surgical teaching and training for vaginal self-dilation, with the goal of preventing recurrent vaginal stenosis, by a nurse practitioner who met with the families in the pre and postoperative context to establish continuity in care.

A non-validated questionnaire measuring Postoperative Dilation Compliance (PDC), created to evaluate compliance with the dilation regimen, and the Glasgow Children's Benefit Inventory (GCBI), a validated quality of life survey for children and adolescents, were administered by mail to a subset of patients who were taught vaginal self-dilation. In the PDC questionnaire, they were asked to review their experiences with self-dilation regimens, noting compliance with dilation at the time of surgery and at current time, as well as overall effects on sense of well-being after the surgical intervention (Addendum A). The GCBI questionnaire (Addendum B) consists of 24 questions with a 5-point Likert answering scale. The total score is transformed to a scale ranging from −100 (maximum harm) to +100 (maximum benefit) with 0 meaning no change at all [5]. The results were scored by calculating the sum of all the responses (Qu. 1–24) on a scale of 1–5 from negative to positive response. The sum of the responses was then divided by 24 to obtain the average response score, and then 3 was subtracted from the average response, and finally multiplied by 50 for the score between −100 and +100. The surgical team did not contact the families directly at any point during the assessment.