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Failure of laparoscopic Vecchietti procedure in a woman with androgen insensitivity syndrome

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Summary

The authors describe a case with androgen insensitivity syndrome (AIS) who underwent the laparoscopic Vecchietti procedure for creation of a neovagina. Postoperatively, the patient achieved anatomic success, with a vaginal length of about eight cm, and she was advised to use vaginal dilators after discharge. The patient reported improved sexual function, but presented about six months later for shortening of her vagina and difficult vaginal intercourse. Physical examination revealed an obliterated vaginal canal about two cm long. Further examination revealed lack of vaginal epithelization. The patient was instructed to continue using vaginal dilators in combination with estrogen cream; however, the patient did not achieve a vaginal length > two cm. The authors believe that the laparoscopic Vecchietti procedure may not be appropriate for women with AIS due to lack of epithelization.

Key words: Androgen insensitivity syndrome; Vecchietti procedure; Epithelization.

Introduction

Surgical vaginal creation may be necessary in cases where the vagina is short or absent. Such is the case in patients with Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome and complete or partial androgen insensitivity syndrome (AIS). While the use of non-surgical methods such as dilators have been shown to be effective in patients with a short or absent vagina, [1] these may not be suitable for some women. Women who have had several vaginal surgeries may not benefit from dilation owing to the incapacity for the vaginal tissue to conveniently stretch by dilators alone. Moreover, the use of vaginal dilators carries deep psychological and emotional implications that make it intrusive, with many women reporting that dilators remind them of their abnormality [2]. For these women, surgical reconstruction of a neovagina may be beneficial. Current methods for vaginal reconstruction have replaced the traditional surgical procedures where the neovaginal space was lined with a split-thickness skin graft (McIndoe-Reed procedure) [3] or with a section of the intestine [4]. Among the recent methods, the Vecchietti and Davydov techniques have been used and both procedures are now performed laparoscopically. Previous reports have shown low complication rates associated with these procedures [5, 6]. In addition, some studies have reported the Vecchietti procedure to be successful in patients with AIS [7, 8]. The authors present the case of a patient with AIS who had vaginal agenesis in whom the laparoscopic Vecchietti procedure failed.

Case Report

The patient is an 18-year-old, divorced woman who was referred to King Abdulaziz University Hospital for management of AIS. At the age of 14 years, she complained of amenorrhea, but she was reassured. The patient presented later at the age of 18 years with complaints of painful and difficult intercourse. Physical examination revealed that secondary sexual characteristics were well developed. Pelvic examination showed absent vagina. Investigations showed normal hormonal profile, 46, XY karyotype, and normal intravenous pyelography. Pelvic ultrasonography and magnetic resonance imaging (MRI) confirmed the absence of the uterus. A diagnosis of AIS was confirmed and laparoscopic creation of a neovagina using Vecchietti's method was offered. She underwent uneventful laparascopic Vecchietti procedure as described before [6]. Post-surgery, the patient was noted to have a vaginal length of about eight cm. She was instructed to use vaginal dilators after discharge. Initially, she reported improved sexual function but presented about six months later with a complaint of shortening of her vagina and difficult vaginal intercourse. Upon examination, the vaginal canal was obliterated and measured about two cm. Further examination revealed lack of vaginal epithelization. The patient was instructed to continue using vaginal dilators in combination with estrogen cream; however, during subsequent visits, the patient did not achieve a vaginal length more than two cm.

Discussion

AIS is characterized by evidence of feminization of the external genital organs at birth, abnormal secondary sexual development in puberty, and infertility in individuals with a 46,XY karyotype [9]. Patients with AIS demonstrate a range of defects in androgen action. Three distinct forms

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of AIS have been identified based on phenotype [9]: 1) complete androgen insensitivity syndrome (CAIS) where the patient has typical female external genital organs; 2) partial androgen insensitivity syndrome (PAIS) where the patient has predominantly female, predominantly male, or ambiguous external genital organs; 3) mild androgen insensitivity syndrome (MAIS) where the patient has typical male external genitalia.

The characteristics of the present patient were consistent with CAIS. MRKH syndrome can also mimic CAIS. Similar to patients with CAIS, those with MRKH demonstrate primary amenorrhea, underdeveloped vagina, and normal breast development; however, the 46, XX karyotype is critical in differentiating both syndromes, [10] which was the case in the present patient.

While vaginal dilation is reportedly the treatment of choice in patients with short vaginal length, surgery is recommended when dilatation fails. However, the timing and nature of the intervention is critical and surgeons need to work closely with the patient and other physicians involved in the care of the patient [9]. In the present case, vaginal dilatation was not explored initially, as the patient was married and had already engaged in sexual activity. In addition, the authors believed that delaying surgery may have had a negative impact on the patient's social and mental well-being and, consequently, jeopardized the functional success of the operation. Besides, the patient must be motivated and be aware of the procedure as well as of the need for a post-operative a postoperative phase.

The laparoscopic approach for creating a neovagina with the Vecchietti method has been reported to be simple, safe, and effective [5, 6]. Anatomic success, with vaginal lengths > six cm and good functional outcomes have been described in one case series of 86 patients with vaginal agenesis [11]. In their report, the authors found that 84 of the patients achieved a vaginal length > six cm within eight days after laparoscopic Vecchietti; after six months, the length of the neo-vagina was 6.5 cm in four patients and seven cm in all remaining patients. In addition, 82 of the patients in their series showed evidence of vaginal epithelization within six months after surgery. While in the present case, the patient initially achieved a vaginal length of eight cm and reported improved sexual function, the vaginal canal was obliterated and measured about two cm six months after the procedure. Further examination revealed lack of vaginal epithelization, and continued use of vaginal dilators in combination with estrogen cream did not result in anatomic success.

Finally, while there are isolated reports [7, 8] of successful vaginoplasty with the Vecchietti laproscopic procedure in patients with AIS, the present case prompts the authors to believe that further clinical evaluation is required to explore this treatment option in AIS. In addition, the decision for surgery should be individualized to the patient and the patient's desire for vaginoplasty.

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