Robotic Excision of the Vagina in a 46 XX DSD Male Patient. First Pediatric Report

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The Disorders of Sex Differentiation (DSD) represent a wide range of congenital anomalies of the genitalia. Surgical treatment of these cases may become a challenge. We present a case of a 16-year-old boy with 46 XX DSD, SRY negative, presented with persistent dribbling incontinence, recurrent UTI, and perineal pain. Past medical history included right orchiectomy, laparoscopic excision of uterus, fallopian tubes, and left streak gonad at another institution at the age of 2 years. The native vagina was left in place. VCUG confirmed the presence of the residual vagina (8 cm in maximum length), connected with the bulbar urethra. Robotic-assisted laparoscopic excision of the vagina was performed with satisfying short and long-term results.

Keywords: robotic surgery; DSD; UTI; pelvic surgery; minimally invasive surgery

INTRODUCTION

Robotic removal of the vagina in a pediatric male patient has not yet been reported to date. We report a case of a 16-year-old boy affected by 46 XX DSD with a residual vagina removed using the robotic approach.

CASE REPORT

A16-year-old boy with 46 XX DSD, SRY negative, presented with persistent dribbling incontinence, recurrent febrile UTI, and perineal pain. He was raised as a male by his parents. His past medical history included: right inguinal orchiectomy for ovotestes, laparoscopic excision of the uterus, fallopian tubes and intra-abdominal left streak gonad at another institution at the age of 2 years (the native vagina was left in place), two-stage repair of scrotal

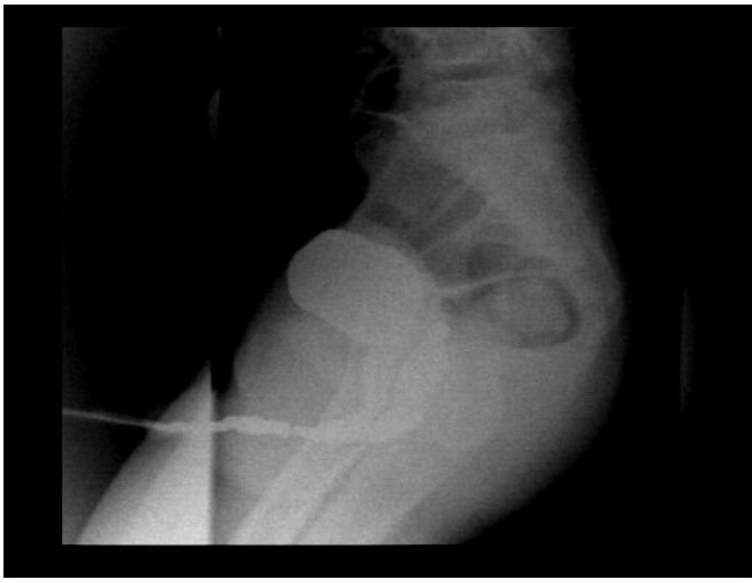

Figure 1. Preoperative VCUG showing the vagina (8 cm), filling up during micturition.

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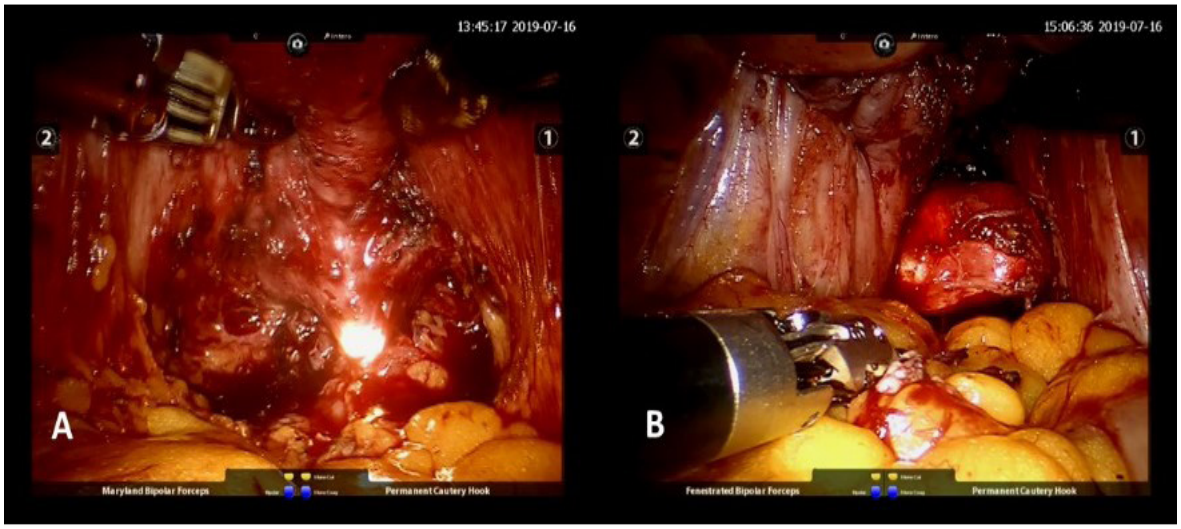

Figure 2 A-B. Separation of the vagina from the surrounding structures using the light of the cystoscope as guidance.

hypospadias when he was 3-year-old, bilateral mastectomy at the age of 13 years. Because of the persistence of recurrent febrile UTI and perineal pain, a voiding cystourethrogram (VCUG) was performed, confirming the presence of a residual vagina (8 cm maximum length) (**Figure 1**). The decision to perform a vaginectomy was taken. The patient was placed in a combined lithotomy/supine position to perform simultaneously the robotic surgery and vaginoscopy. The preliminary cystoscopy showed a normal urethra, without stenosis, with the vagina opening at the level of the bulbar urethra. The 12-mm optical port was inserted via the umbilicus, and two other 8-mm robotic working ports were placed on the para-rectal lines and at the level of the transverse umbilical line. The bladder was suspended to the anterior abdominal. The light of the cystoscope, into the vagina, was used as guidance to separate the vagina from the surrounding tissues (**Figure 2A-B**). Dissection was performed between the vagina and the surrounding tissues. Suspension of the vagina, good magnification ensured by the robotic approach, and the Trendelenburg position allowed us to avoid injuries to the sacral plexus. The vagina was transected as close as possible to the urethra leaving in place a little sidewall of the vagina near the urethra. This opening was closed with an absorbable stitch, avoiding urethral strictures (**Video 1**). The specimen was then removed from the optical port. The pathologic examination confirmed the removal of the vagina. The operative time was 240 minutes.

The post-operative period was uneventful. The patient was discharged 7 days after the procedure The Foley catheter was removed on the 20th post-operative day. A post-operative VCUG showed neither significant remnants at the level of the bulbar urethra or leakage with normal bladder emptying (Figure 3). After a two-year follow-up, the patient shows a complete resolution of the symptoms.

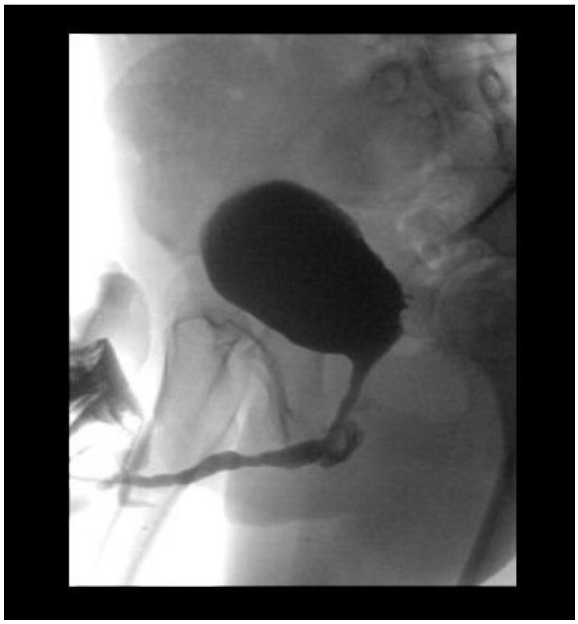

Figure 3. Post-operative VCUG showing the removal of the vagina with a little stump close to the urethra.

DISCUSSION

Surgical removal of a residual vagina may be a real challenge in the pediatric population due to the proximity to the pelvic nerves, rectum, ureters, bladder, rectal sphincters, and because of the narrow surgical space[1,2,3]. There is a gaining consensus about robotic surgery feasibility and efficacy in the pediatric population due to an increasing number of appreciable surgical results reported in the literature in a wide range of congenital anomalies[1]. In the case of pelvic anomalies, open surgery often needs a combined approach with two incisions at the level of the abdomen and perineum respectively. The laparoscopic approach, despite shorter hospitalization and good control of post-operative pain, rarely allows safe access to the perineal space [4,5]. The minimally invasive approach, based on robotic surgery, has been recommended in several reports[4,6,7] because of the clearer view of the deep pelvic structures. Robotic-assisted surgery has been successfully used to obtain a complete removal of a large utricle in a pediatric patient who had previously undergone a laparoscopic procedure[8]. In the present case, the previ-

ous laparoscopic approach did not allow the complete removal of the vagina. For this reason, we opted for robotic-assisted laparoscopy in the redo procedure to combine the advantages of laparoscopy with the improved three-dimensional (3D) visualization associated with high instruments dexterity[9]. This technique has been previously used in transgender adults[10]. To our knowledge, no cases of robot-assisted excision of the residual vagina in a pediatric DSD male patient has been described in the literature to date.

In conclusion, because of the excellent magnification, the high dexterity ensured by robotic instruments, the low risk of iatrogenic lesions, the short hospital stay and the low postoperative pain, robotic-assisted surgery should be considered also in pediatric patients.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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