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Cervicovaginal agenesis treated with modified Yang-Monti technique in two steps: Case report and literature review

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ABSTRACT

INTRODUCTION: Müllerian duct anomalies are rare with less than 200 cases published in the literature. Recently, the implementation of the Yang-Monti principle for the creation of the neovagina has been used in 10 previous published cases. Here, we report the first case of cervicovaginal agenesis treated with the modified Yang-Monti technique in two steps in México.

PRESENTATION OF CASE: A 14-year-old female presented to the pediatric consult with a history of primary amenorrhea and a chronic-cyclic pelvic pain. She had normal external genitalia and secondary sexual characteristics, with a small vaginal pouch. Pelvic ultrasonography and magnetic resonance showed the absence of cervix and proximal vagina. The surgical approach was carried out in two steps. In the first, a neovagina was created with the modified Yang-Monti technique. On a second step one year after, the neovaginal dome was anastomosed with the uterus in a diamond shape, using Gore-tex® in the cervicovaginal anastomosis, and a Foley catheter to prevent stenosis. After an 18-month follow-up, no complications have been observed. She has been taking Drospirenone and Ethinyl Estradiol with regular menstrual cycles.

DISCUSSION: The surgical treatment of cervicovaginal agenesis has evolved. The advantages of the modified Yang-Monti technique lie in the possible diminution of the tension on the vascular pedicle, and the gained length of the neovagina. To perform this procedure in two steps, likely diminishes the risk of neovaginal ischemia and leakage of the anastomosis.

CONCLUSION: The two-steps modified Yang-Monti technique represents a safe alternative for the management of cervicovaginal agenesis.

1. Introduction

Müllerian duct anomalies (MDA) are rare [1,2] and may present as any abnormality in the fallopian tube, uterus, cervix and upper two-thirds of the vagina [3]. The American Society of Reproductive Medicine (ASRM) classified these anomalies into seven categories, where the hypoplasia/agenesis of the vagina and cervix corresponds to types 1a and 1b, respectively [4]. The incidence of cervicovaginal agenesis (CVA) is unknown, and there are less than 200 cases published [1], where only 7% of the cases had functional endometrial tissue [5]. There are several techniques for the surgical management of CVA, which range from hysterectomy to different methods of cervicovaginal reconstruction [1,2,6]. Important aspects of the surgical management of CVA includes the accurate diagnosis of both the underlying abnormality and its anatomy, as well as the evaluation of any associated renal or skeletal anomalies, and the proper psychological preparation of the patient [7]. Hendren et al. [8] and Kisku et al. [2] used the sigmoid colon to create a neovagina. Tu et al. [9] and Garcia-Roig et al. [10] described the implementation of the Yang-Monti principle [11,12] to create a neovagina using the colon. Here, we report the first case of CVA managed with the modified Yang-Monti technique in México in two steps. The following accomplishes the SCARE criteria[13]. The legal tutor agreed with the publication of the case and signed the informed consent.

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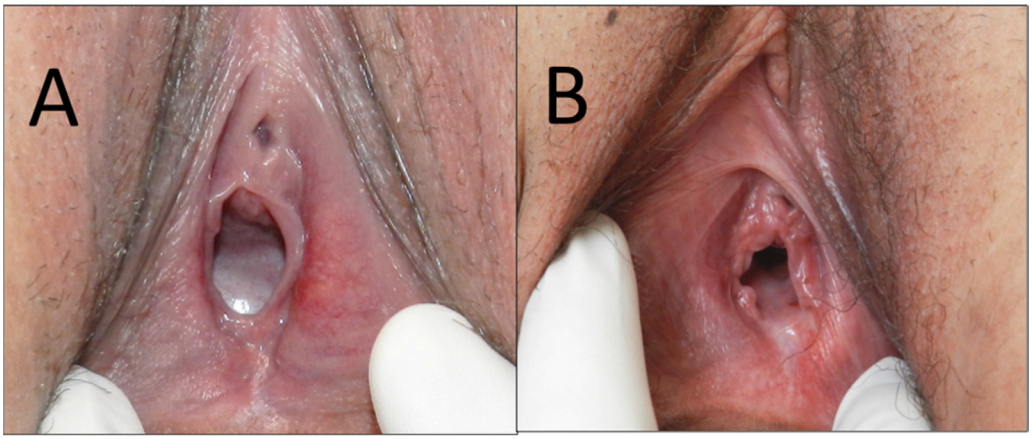

Fig. 1. A preoperative physical examination; B 1-month postoperative examination.

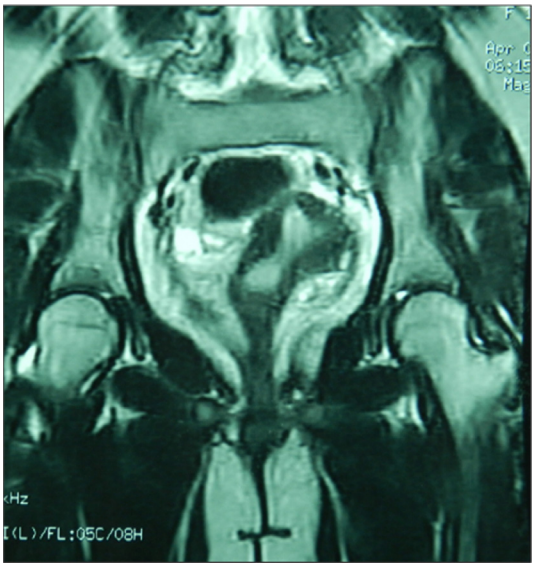

Fig. 2. A Magnetic resonance imaging demonstrating the cervical atresia and the vaginal agenesis.

2. Case presentation

A 14-year-old female presented to the pediatric consult with a history of primary amenorrhea and a chronic-cyclic pelvic pain of two years. During the physical examination, she had age-appropriate secondary sexual characters, normal female external genitalia with a small vaginal pouch of approximately 2 cm (Fig. 1A). The ultrasound demonstrated an anteflexed uterus of $7.6 \times 3.6 \times 4.2$ cm, an endometrium of 5 mm with normal adnexa and absence of cervix and proximal vagina; this was corroborated by a magnetic resonance of the pelvis (Fig. 2). The excretory urogram was normal. She had a 46XX Karyotype. The patient presented to the emergency room with an episode of pelvic pain, and decided to perform a diagnostic laparoscopy and found a hemoperitoneum secondary to blood outlet from the Fallopian tube. The patient was managed with medroxyprogesterone for one-year until the mother of the patient accepted the surgery.

The surgical procedure was performed as follows: Diagnostic laparoscopy in which we detected endometriosis that was managed by ablation. The rectovesical pouch was dissected laparoscopically assisted, then, the laparotomy was done, and the neovagina created using the technique described by Tu et al. [9], and Garcia-Roig et al. [10] (Fig. 3). An 8–10 cm of the sigmoid colon was resected, taking great care that the vascular supply was kept in the middle portion of the isolated segment (Fig. 3A). The sigmoid section was opened on its anterior border 1 cm from the mesenteric edge (Fig. 3B) and retubularized longitudinally in two layers (Fig. 3C–D). Finally, the neovagina was delivered to the vaginal introit (Fig. 1B). We left the vaginal dome at the level of the cervix with the uterus in situ.

The postoperative period was accomplished without complications, the patient was discharged on the fifth postoperative day, with vaginal dilatations with the 22 Fr Hegar dilatator for one month, and medroxyprogesterone for one year. During the follow-up, the patient not presented vaginal stenosis (Fig. 1B) or other complication.

A year later, we did the second surgical procedure to reconnect the uterus with the neovagina:

Through the previous incision the uterus was identified, we did an incision in the lower segment of it, until we found the uterine lumen, then we extended the incision 1 cm longitudinal, washed out the hematometra and did a permeability test of the Fallopian tubes by instillation of methylene blue, observing the methylene blue reached the abdominal cavity through the left Fallopian tube; the right Fallopian tube was ingurgitated. A transverse incision of 1 cm was done in the neovaginal dome, and a diamond shaped anastomosis was carried out with separate stitches with 3-0 polyglactin 910. We left a Gore-Tex® of 2 cm fixed to the uterovaginal anastomosis, and a Foley catheter of 12 Fr through the Gore-Tex® for preventing stenosis of the anastomosis.

The patient had a good evolution and was discharged at day 7-postoperative. We took out the Foley catheter at day 21, and the Gore-Tex® was expelled spontaneously after three months. The patient has been managed with Ethynyl Estradiol and Drospirenone for 18-month with regular menstruation.

3. Discussion

CVA is a complex MDA that usually presents with a functional uterine fundus and Fallopian tubes, normal ovaries, cervical and vaginal atresia with normal external genitalia [2,14], as the patient. The frequency of vaginal atresia is of 1 in 4000 to 1 in 5000 female subjects with abnormal development of the uterine corpus observed in >90% (Mayer-Rokitansky-Kuster-Hauser syndrome) [2,15], but the incidence of CVA is unknown. In 1999, Fujimoto et al. [15] described only 50 patients reported in the literature, 48% associated with a normal vagina, 42% with complete vaginal agenesis, and only 10% with a "shortened blind vaginal pouch" as the patient. In 2011, Roberts et al. [1] estimated that there were less than 200 cases of CVA published in the literature since 1942.

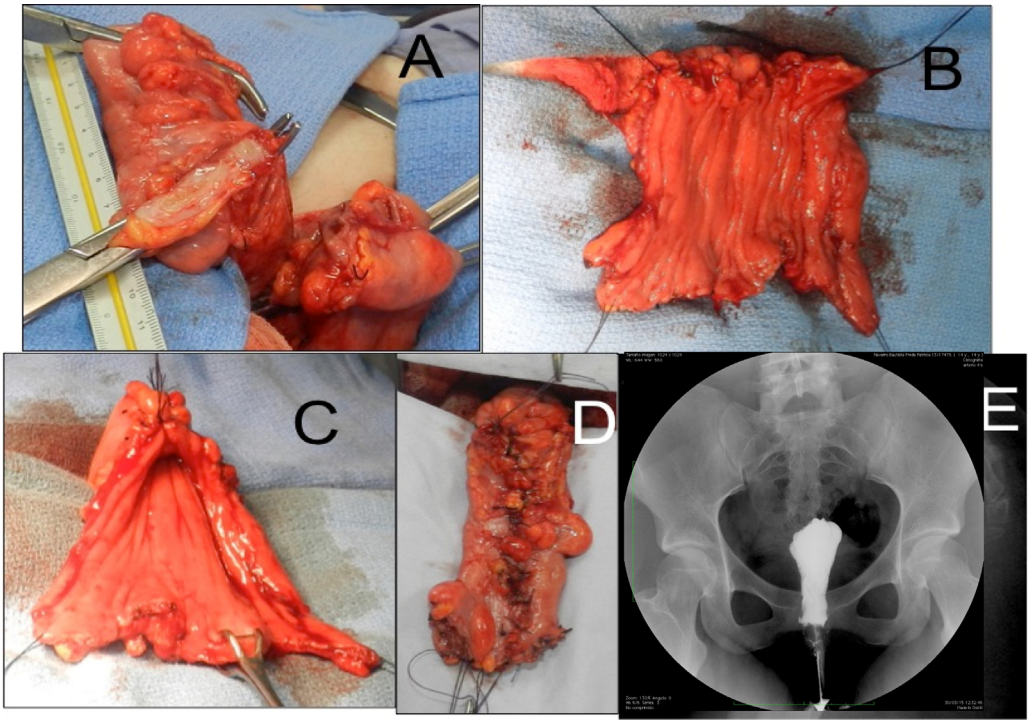

Fig. 3. A Resection of the sigmoid colon; B Longitudinal incision; C-D Retubularization; E 6-month postoperative vaginography.

CVA arises from a defect in the growth of the Müllerian ducts [1–3]. The proper development of the Müllerian ducts depends on the completion of three phases: organogenesis, fusion, and septal resorption. The failure in the organogenesis will develop uterine agenesis or hypoplasia [3]. Along with the development of the Müllerian ducts, the mesonephric ducts appears. The failure of the distal Müllerian structures to develop may give rise to cervical and vaginal agenesis; these anomalies are closely related to a wide range of associated malformations of the genitourinary tract in 29% of the times [3,5].

There is not a consensus on the primary symptom of a patient with CVA, but must of them presents between the age of 12–20 years and have two key features, one is the primary amenorrhea, and the other is a chronic cyclic pain [2,14,15]. On physical examination, normal secondary sex characteristics will be found, with a complete transverse vaginal septum located in the upper vagina in 46%, in the middle vagina in 40%, or in the lower vagina in 14% [16]. The complete obstruction of the menstrual flow will develop an hematometra or an hematosalpinx which may lead to endometriosis [14,16]. On the clinical approach in the study of a patient with MDA, an abdominal ultrasound is recommended because adequately allows characterizing the uterus, adnexa, and kidneys. An excretory urogram is also mandatory because of the association with nephrological and urological anomalies. However, pelvic MRI is considered the best imaging modality for MDA diagnosis [3,16].

Once the diagnosis of MDA is made, the treatment will depend on the ASRM classification. In the case of a patient with a CVA, which is classified as a type 1b [14], the treatment has evolved. The earlier reports advocate a conservative management or canalization procedures, but 33% of this patients required a hysterectomy after unsuccessful canalization procedures [15]. Other more recent reports demonstrate that although hysterectomy is recommended as an elective procedure [1,14], the use of a colovaginoplasty when the patient has a functional uterus is also a safe procedure [2,5,8–10,14]. In 1994, Hendren and Atala [8] described the use of bowel (sigmoid colon, small intestine, and rectum), for the creation of a neovagina. Kannaiyan et al. [5] and, Kisku et al. [2], published separately series of patients were a neovagina was created with sigmoid colon by rotating 180° degrees, and anastomosed the proximal end of the sigmoid colon to the vagina and, the distal end segment of the sigmoid colon to the cut end of the uterus in an antiperistaltic anastomosis. Kannaiyan et al. [5] reported that its patients were not advised to have any vaginal dilatations, and no indwelling molds or stents were used. They reported stenosis of the neovaginal orifice in 9%, without other complications such as endometritis or pelvic inflammatory disease. In contrast, Kisku et al. [2] documented a leakage of the anastomosed colon in 5%, recurrence of the pain due to the recurrence of chocolate cysts in 10%, mild stenosis of the neovagina in 10%, on daily self-dilatation as a precautionary measure in 15%.

We decided to use a technique for the creation of the neovagina first documented by Tu et al. [9] in 2012, and repeated by Garcia-Roig et al. [10] in 2014, in the believe that this technique diminish the tension on the vascular pedicle, especially in cases with short mesentery, obesity or a masculinized pelvis [10]. Furthermore, the decision to do it in two steps was for diminishing the risk of neovaginal ischemia, anastomotic leakage, and endometritis. This technique allows us to create a larger neovagina without compromising the vascular pedicle, gaining between four to six centimeters in length [9]. Tu et al. [9] reported no complications in the immediate postoperative period nor the follow-up, with a median follow-up of six years. On the contrary, Garcia-Roig et al. [10] documented one patient (16.6%) with mild vaginal stenosis. It is important to highlight that together, the series of Kannaiyan et al. [5] and, Kisku et al. [2] had 31 patients, and the cases of Tu et al. [9] and, Garcia-Roig et al. [10] had only ten patients. Therefore, more patients treated with this technique are necessary for comparing the outcomes in the immediate, mediate and long term. The first case of cervicovaginal agenesis treated with the modified Yang-Monti technique in two steps in México is presented here. With a follow-up of 18-month from the second surgery, the patient denies troublesome mucus production, with painless, regular, and cyclic menstrual periods, without vaginal stenosis or the requirement of self-vaginal dilatations, nor any other complication related

to the surgery. As to our knowledge, in México there is only one cases series of patients treated for vaginal agenesis were different techniques of skin graft were used [17].

4. Conclusion

The MDA represent a rare disease, the correct classification, and adequate treatment are fundamental for a proper quality of life. In the case of cervicovaginal agenesis, the surgical approach is not standardized. The option of the modified Yang-Monti technique for the creation of the neovagina seems to be a good procedure when compared to the standard colovaginoplasty. We learned from this case that performing the surgery in two steps, allowed the tissues to heal. Therefore, the uterovaginal anastomosis was done in a well-irrigated tissue without tension, diminishing the risk of anastomotic leak. Additionally, the abdominal adherences were less, enabling the possibility to perform the second step of the surgery by a laparoscopic approach.

Conflicts of interest

All authors declare no conflict of interest about the publication of this article. No external funding was needed.

Ethical approval

The written consent was signed by the legal tutor.

Consent

No personal information is given nor modified.

Author contribution

Acosta-León J.- study concept, writing the paper, final decision to publish, data collection.

Pantoja-Rojas A.- study concept, data collection.

Corona-Rivera JR.- data collection and analysis.

López-Marure E.- data collection and analysis.

Ploneda-Valencia CF.- study concept, writing the paper, data collection, final decision to publish.

Guarantor

Jorge Acosta-León and César Felipe Ploneda-Valencia.

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