



**UNIVERSIDAD NACIONAL AUTÓNOMA
DE MÉXICO**

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DIVISIÓN DE ESTUDIOS DE POSGRADO E INVESTIGACIÓN
SECRETARÍA DE SALUD
INSTITUTO NACIONAL DE PEDIATRÍA

**“LAPAROSCOPIC APPROACH FOR NEOVAGINAL CREATION WITH SIGMOIDS
IN ADOLESCENTS WITH RECTOVESTIBULAR ANORECTAL MALFORMATION
PREVIOUSLY CORRECTED”**

TESIS

PARA OBTENER EL GRADO DE
ESPECIALISTA EN CIRUGÍA PEDIÁTRICA

PRESENTA:

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MÉXICO, D.F.

2014



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Abstract

Objective: To describe the creation of a neovagina with sigmoid using laparoscopic approach in adolescents diagnosed of anorectal malformation (ARM) and Mayer-Rokitansky- Küster-Hauser (MRKH) syndrome that were previously intervened using posterior sagittal anorectoplasty (PSARP) approach in the first months of life without correction of vaginal agenesis.

Materials and methods: Two adolescents with rectovestibular ARM with a history of primary PSARP in the first months of life without the correction of vaginal agenesis were included. The patients had been completed approach for diagnosis MRKH, including enema colon, all of which pointed to MRKH syndrome and on the basis of which a laparoscopic neovaginal creation with the sigmoid were carried out.

Results: The creation of neovagina with sigmoids was factible in the patients with all the cosmetic advantages and recovery that minimal invassive surgery could offer. Presently, complications such as stenosis, prolapsed, urinary and intestinal function deteriorations have not been seen and the patients are enjoying a better psychosocial life.

Conclusions: In patients with ARM + MRKH, a complete pre-operative assessment of colorectal conditions is paramount and foremost to the selection of the technique or intestinal segment to be employed. The advantages of minimal invassive surgery offered excellent cosmetic results that were harvested in psychological well-being of the adolescents. Surgical history of PSARP should not be a limiting

factor for creating a neovagina using minimal invasive approach. Gradually, laparoscopy surgery would replace laparotomy in these procedures.

Keywords: Mayer Rokitansky Kuster Hauser, rectovestibular anorectal malformation, primary posterior sagittal anorectoplasty, laparoscopy, neovagina

Introduction

MRKH consists of partial or total absence of vagina and uterus with functionally normal ovaries and rudimental Fallopian tubes. It is usually seen in women possessing 46XX chromosomes and normal external genitals. It could be linked with other alterations within which the most frequent are renal and skeletal alterations among others. The association with ARM is very rare with a frequency of less than 1%^{1,2} of all the ARMs. The etiological origin of this malformation is due to an interruption in the embryonary development of Mullerian ducts without affectation of the ovaries. For this reason, the diagnosis is frequently late for primary amenorrhea which inevitably leads to a psychological shock in the patient and the family,³ and invariably brings about a deep affectation of her self-esteem and personal safety. The management is multidisciplinary with the construction of neovagina as the paramount objective to uplift and improve the psychosocial well-being of the patient. Since it was first described,⁴⁻⁸ different techniques of neovaginal creation have been put forward⁹⁻¹² as well as different modifications of the same.¹³⁻²¹ However, even when laudable advances in the management of this type of patients have been made coupled with the availability of initial management proposals,²² still there are conditions where these conducts cannot be generalized as it is the case of ARM and MRKH, where apart from few experiences in their management, little effort has been made to explore technical innovations of minimal invasive surgery. This work described the construction of neovagina with sigmoid using minimal invasive surgery in adolescents previously intervened by posterior sagittal anorectoplasty in the first years of life.

Materials and Methods

Two adolescents with a history of ARM and rectovestibular fistula who were surgically intervened for PSARP at 5 and 6 months old were study. In the follow-up of the patients, diagnosis of MRKH was established through karyotype 46XX, CAT, and NMR which revealed the absence of uterus. The renal function and structure were normal. Both patients depicted their concern and desire to receive vaginal reconstruction therapy. The two were suffering from moderate constipation occasionally controlled with laxants²³. The sigmoids were of normal dimension and the bladder emptying was normal also. The assessment of sexual development and mental health was carried out by multidisciplinary team consisting of psychiatrics, psychologists, gynecologists, endocrinologists, and pediatric surgeons.

Surgical technique

The surgery was carried out under general anesthesia and peridural block. The patients were placed in Trendelenburg position with the urethra splinted using Foley catheter followed by decompression of the bladder. A total of four trocars, two of 12 mm and two of 5 mm were used. One of the 12 mm trocars was placed at the level of umbilicus and the other on the right flank while one of the 5mm was placed on the left flank and the remainder on the suprapubic. A 5 mm, 30 grade lent and 12 mmHg pressure pneumoperitoneum were used. An evaluation of the vascularity, mobility, and sigmoid longitude was carried out with a selection of a segment of approximately 10-12 cm at peritoneal deflection level that would be used in the construction of a neovagina. In the mesenterium of this segment, a

proximal and distal window was created using decompressed Foley catheter EnSeal Trio® (Ethicon, Cincinnati, OH, USA). A proximal and distal shot was placed on each of the windows with a sharp stapler of 60 mm (echelon 60 endopath stapler, Ethicon Endo-surgery Johnson & Johnson, Cincinnati, OH, USA) thereby separating the intestinal grafted segment. Later, colon proximal cape was extracted through the umbilical scar for the placement of intraluminal transrectal enlargement head of 33mm (PROXIMATE® ILS Curved Intraluminal Stapler, Ethicon Endo-Surgery, Cincinnati, OH, USA) which was fixed with a knuckle of Polyglactin 910, 2-0 Vicryl® Ethicon, Cincinnati, OH. This was newly embedded in the abdominal cavity in order to join the proximal colon with the rectus by introducing intraluminal stapler through the later, followed by exploration and dissection of vaginal introitus in roma form through the perineal under laparoscopic vision avoiding any damage to the urethra and the rectus and creating a space where the neovagina should be lodged. The distal end of the graft was placed in the vaginal introitus and anastomosed with 3 zero vicryl using simple points (figure 1). A point of fixation in the anterior face of the interposed sigmoids was placed via laparoscope. The lavage of the cavity was carried out with the extraction of pneumoperitoneum and trocars without leaving any drainage.



Figure 1. Immediate post-operative

Case Report

Patient 1

An adolescent of 17 years old with diagnosis of ARM plus rectovestibular fistula and constipation that was occasionally treated with laxants, who evacuated 1-2 times a day with occasional blood stains but normal bladder emptying. She had a boyfriend but the absence of vagina was a product of anxiety and anguish to her, reason why she sought for vaginal reconstruction. The colon for enema showed a sigmoid with normal dimension.

Apparently a healthy with a weight of 37 kg and a height of 1.37 m (constitutional underweight), breast 5, and genital Tanner in line with undersized vulva (for decreased dorsoventral axis) without clitorimegaly. The vestibule was short and the urethral meatus was central. The hymen was replaced by thick and estrogenous mucus. The anus was located 1.5 cm from the vulva vestibule without mucosal prolapsed and with well formed perianal fossa.

During the surgery, the ovaries were found in the internal inguinal orifice and were of normal size and shape. However, the fimbrias were rudimental and joined by fibrous cord that corresponds to Mullerian remnants which were resected.

After the procedure, recovery of intestinal function was on the 2nd day and oral nutrition was begun in the 3rd day. She was discharged on the 5th day following the retirement of urinary catheter. She underwent a period of intestinal adaptation with evacuations of up to four times per day without important accidents of blood stains and only required transitory dietetic modifications. In 2½ years of follow-up, she

has been presenting two evacuations of normal consistence in a day with normal anal sensitivity and preserved bladder function. Presently, she is neither on special diet nor does she use laxants. Vaginal cleansing every 72 hrs was prescribed a month after the surgery and this is continued till date. No report of mucosal discharge and therefore the use of sanitary towels are not required. The genital appearance is normal without visible evidence of cicatrix with humid estrogenous mucous membrane. Abdominal cicatrices were absent as well as intestinal occlusion events. The calibration of the neovagina is done with Hegar 22 dilator however; she has not started sexual life.

Patient 2

A girl of 18 years old with additional diagnosis of interauricular and interventricular communications without hemodynamic repercussion; metabolic syndrome; cervicothoracic scoliosis secondary to C6 vertebral hypoplasia and C5 hemivertebrate; phalangeal and right hand metacarpal agenesis as well as right supernumerary ribs. The colon by enema showed a sigmoid with normal dimension. Metabolic control was with metformin and diet. At 14 years old, she underwent depressive episode associated with vaginal agenesis, coupled with moderate constipation occasionally managed with laxants, and stains probably menorrhoeal every 15 days. Presently, she is a high school student with good academic performance but no sentimental partner.

Morphometrically, the body weight was 53.5 kg with a height of 1.35 m (BWI=29.3) and breast 5. Evaluation of the urogenital organs showed a genital Tanner, hypertrophic major lips, central urethral meatus, short estrogenous vestibule and normal clitoris. The anus was located 1.5 cm from the vulvar vestibule without mucous prolapsed and with well-formed perianal fossa.

The ovaries were evaluated during the surgery, finding them to be normal with rudimentary fimbrias and fibrous cord that was resected during this process. Double cecal appendices and Meckel diverticulum were surgical findings showing no data of inflammation and were left without resection.

Following the procedure, intestinal function was normalized on 2nd day and oral nutrition was begun on 3rd day and the patient was discharged on 5th day after retiring the urinary catheter. The period of intestinal adaptation took place in the first month evacuating 4 times a day without accidents and only required dietetic control. In 2½ years of follow-up, she has been presenting two evacuations of normal consistence in a day with normal anal sensitivity and preserved bladder function. She is neither on special diet nor does she use laxants. Till date, there is no report of mucosal discharge and therefore the use of sanitary towel is not required save vaginal cleaning every 72 hrs which was prescribed a month following the surgery and continued until now. Presently, she is neither on special diet nor does she use laxants. The genital appearance is normal without visible evidence of cicatrix with humid estrogenous mucous membrane. Abdominal cicatrices as well as intestinal occlusion events were absent. Her state of mood and that of her family are favorable and until now, has not presented depressive

signs. The calibration of the neovagina is carried out with Hegar 22 dilator however; she has not started sexual life.

Results

The dissection of a space from the vaginal introitus to the peritoneal cavity for the descent of the graft was not impeded by the history of PSARP. The urinary catheter maintained the perineum dry and there was no report of early surgical complications and in the follow-up. Intestinal movement and the first evacuation were established in the first 48 hours in both cases. Pain control did not require the use of opioids.

The two patients presented a one-month period of intestinal adaptation. The rectal and bladder functions following the surgery were maintained without changes. Mucosal discharge is being resolved by vaginal cleaning every 72 hours without serious social problem.

The genitals remained without perceptible cicatrices with normal appearance, estrogenous and humid mucus (Figure 2). The neovagina is maintained dilated with Hegar 20 which is prescribed two to three times per month and the longitude of the segment is 10 – 12 cm. Both patients have not started sexual life. Self-perception of both is a “complete woman”. The patient with sentimental partner refers feeling surer in her relation, while the other patient has not presented depressive crisis after the operation. Parents of both patients are satisfied with the result of the surgery until now.



Figure 2. Late post-operative

Discussion

Mullerian dysgenesis occurs in approximately 1:5000 live newborn females. The absence of vagina provokes important psychological, physiological and social problems to a woman and her families.³ Based on the progress made on this ailment, there are initial proposals of non-surgical treatment.²² The association of MRKH and ARM is very rare,^{1,2} and vaginal creation has been described at the time of ARM correction. There are few evidences of posterior sagittal anorectoplasty in adolescents without the correction of vaginal agenesis.^{24,25}

The history of ARM surgical correction makes us to imagine that the formation of neovagina is something difficult for secondary fibrosis due to previous surgical manipulation. However, there was no difficulty in creating the space to lodge the neovagina. This was in accordance with the report of Miller who in similar conditions created a neovagina using the technique of Vecchietti.²⁴ We consider that surgical history could not be a limiting factor.

The psychological impact of MRHK in patients and families is widely known,^{3,26} an aspect which could be worsened in our patients for its association with other malformations and for the age. Minimal invasive procedure is considered in the treatment, for the fact that the esthetic advantages are important in this age group. Moreover, the advantages of lubrication, permeability, and longitude of the segment have been described by other authors when sigmoid segments are used and this can be observed in the patients.

Prior to the procedure, the intestinal and bladder functions were considered with the finding of urinary and fecal continence that required occasional use of laxants for evacuation, conditions that did not contraindicate the use of sigmoid segments and a point which has only been analyzed by few publications.²⁷ However, we deemed it fundamental in order to avoid increasing the risk of fecal incontinence.

Both patients underwent through a period of adaptation of approximately one month during which the number of evacuations increased and only one of the patients had an accident of soiling. Modification of diet was just transitory and there was a remission of all the changes.

The complications referred to in intestinal segment were no longer observed in both patients²⁸. Mucous discharge, tackled with vaginal cleaning every 72 hours during bath, became reduced along the time in both patients.

The psychosocial aspects of the patients have not been evaluated, however; self-perception, esthetic outlook, and social environment are vital aspects giving an important support to this function as was described after neovagina creation.²⁹⁻³²

Conclusions

There are few reported experiences in the management of adolescents with uncorrected ARM and MRKH. The only limitation of this work is the number of patients (two), however, the result obtained is in accordance with what were published in other series.

We consider that in patients with ARM + MRKH, a complete pre-operative evaluation of colorectal conditions is indispensable before choosing the technique or intestinal segment. Surgical history of the septum or vaginal introitus in the first reparation of PSARP should not be a limiting factor for carrying out a technique which otherwise would be thought impossible taking into account that many years have passed following the first intervention. What is needed is only a careful dissection to avoid a possible lesion of the urethra or rectus. It is relatively easy to create the space for the neovagina.

Laparoscopic creation of a neovagina offers all the advantages of minimal invasive surgery with excellent cosmetic results. This can be translated to psychological well-being of the adolescent, a situation which is quite understandable for the age and very important given the emotional feelings of youths, and consequently the esthetic impact as seen by her future sexual partner. We believe that laparoscopic surgery would gradually replace laparotomy with creation of surgeons and gynecologists with experience in gynecological and digestive surgery.

Disclosure Statement

No competing financial interests exist.

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