

Vaginal agenesis: A case report*

BY REYALU T. TAN, MD; SIGRID A. BARINAGA, MD, FPOGS; AND MARIE JANICE S. ALCANTARA, MD, FPOGS

Department of Obstetrics and Gynecology, Southern Philippine Medical Center

ABSTRACT

Congenital anomalies of the vagina are rare congenital anomalies. Women born with this anomaly present with collection of blood in the uterine cavity or hematometra and pelvic pain. Presented is a case of a 12-year old girl with hypogastric pain and primary amenorrhea complicated by vaginal agenesis. She was managed conservatively by creating a neovagina with the use of bipudendal flap or Modified Singapore flap. Management can be non-surgical or surgical but the management of congenital vaginal agenesis remains controversial. The decision to do a conservative surgical procedure or a hysterectomy depends on the clinical profile of the patient, the expertise of the surgeons, the extent of the anomaly, and its association to other congenital anomalies.

Keywords: Vaginal Agenesis, Hematometra, Primary Amenorrhea, Modified Singapore flap

INTRODUCTION

Developmental anomalies in mullerian ducts and urogenital sinus represent some of the most interesting disorders in Obstetrics and Gynecology. Normal development of the female reproductive system leads to differentiation of the reproductive structures. Vaginal agenesis is the congenital absence of vagina where there is failure of formation of the sinovaginal bulb which leads to outflow tract obstruction and infertility. It occurs in 1 out of 4,000–10,000 females. Complete vaginal agenesis is discovered in 75% of patients with Mayer-Rokitansky-Kuster-Hauser syndrome. Approximately 25% of patients have a short vaginal pouch.¹ At present, no local studies cite the mean incidence of these anomalies in the Philippines. The Journal of the Reproductive Endocrinology and Infertility has published 2 cases with combination of cervical and vaginal agenesis.^{2,3} Patients with vaginal agenesis usually manifest with symptoms of obstructed menstrual flow leading to collection of blood in the endometrial cavity, fallopian tubes and peritoneal cavity.

CASE REPORT

This is a case of RV, a 12 year-old, nulligravid, who came in due to abdominal pain and no menarche. The patient had no known illnesses. She underwent Exploratory Laparotomy, Left Fimbrioplasty, Hysterotomy with Evacuation of Hematometra, Perineal Exploration last November 2014 for Hematocolpometra. The family history is unremarkable with no similar menstrual disorder or

congenital anomaly. The patient is an Elementary student, non-smoker, non-alcoholic beverage drinker, 2nd child of a G5P5 mother.

Two months prior to admission, the patient had sudden onset of severe abdominal pain. Admitted at a local hospital and managed as a case of Ovarian New Growth with complication. At laparotomy, the patient was noted with hemoperitoneum (100 milliliter) with the left fallopian tube enlarged to 5 x 9 centimeter with a clubbed end with dripping blood. Fimbrioplasty was done. The uterus was asymmetrically enlarged to 12 week size. Hysterotomy was done at the vesicouterine fold with a 1 centimeter incision where hematometra (300 milliliter) was suctioned. The right fallopian tube and both ovaries were grossly normal. On perineal exploration, a stenotic vaginal introitus was advanced and explored. No vagina and only periurethral and perirectal membranes were noted. The Postoperative diagnosis was Hemoperitoneum secondary to retrograde menstruation; Hematometra and hematosalpinx secondary to congenital absence of the vagina and cervix. The patient was discharged improved.

Recurrence of the hypogastric pain a month prior to admission was noted. The patient was brought to tertiary government hospital and was examined awake, ambulatory with normal vital signs. The patient had pink palpebral conjunctivae, anicteric sclerae, symmetric chest expansion and clear breath sounds. The breasts were Tanner stage 2, with axillary hair. (Figure 1) The abdomen was soft, flabby, and non-tender, with infraumbilical midline scar. The vulva was grossly normal, with pubic hair, clitoris, urethral opening, and vaginal dimple. The vagina ends in a pouch, 2-3 centimeter from the hymenal ring (Figure 2). Digital rectal examination revealed good sphincteric tone, intact vault, with cystic non tender mass noted 3-4 centimeter from the anal opening.

Transrectal ultrasound revealed Hematocolpometra

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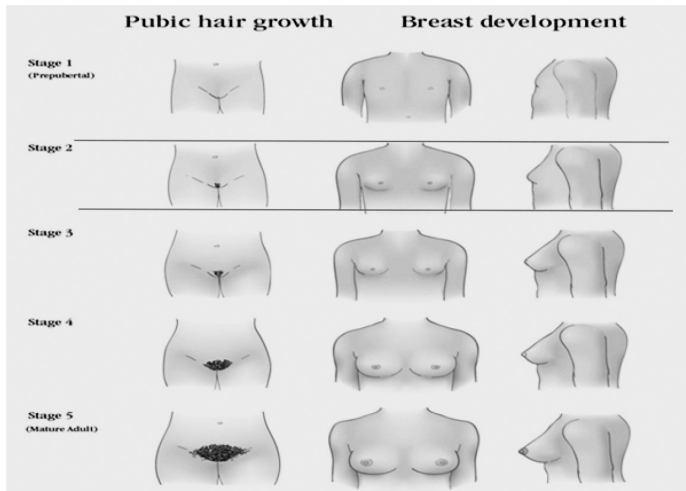


Figure 1. Tanner staging



Figure 2. Perineum (Anterior view)

(12 milliliter) due to an outlet obstruction consider Complete Transverse Vaginal Septum mid/distal 3rd of the vaginal canal, thin endometrium, sonographically normal looking uterine corpus, small ovaries with hemato/hydrosalpinx (1.2 milliliter), left. (Figure 3) Ultrasound of KUB was done to identify if there were any accompanying urologic structure abnormalities which revealed cystitis with sonographically normal kidneys. Abdominal Magnetic Resonance Imaging demonstrated Hematometra (3.7

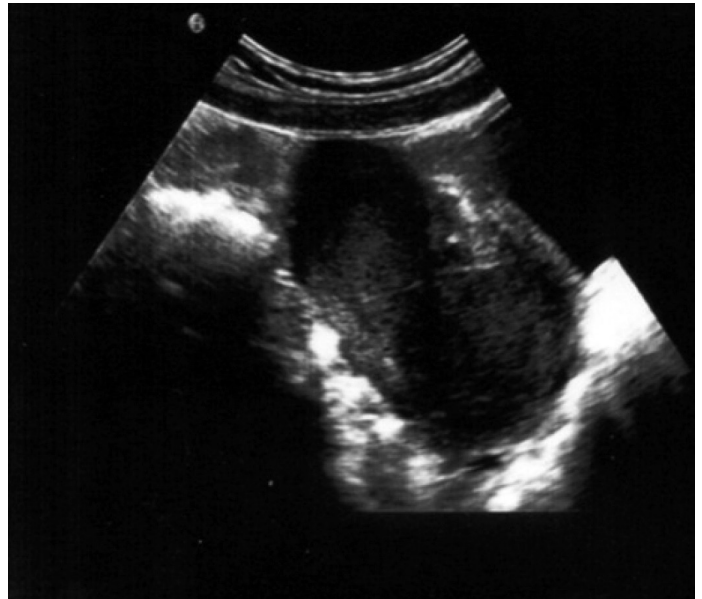


Figure 3. Right ovarian cyst. (A) The right ovarian cyst measured 1.5 x 2.0 x 1.5 cm irregular, yellow tan, soft mass. (B) Cut section showing minute cartilage-like spicules (black arrows).

centimeter maximum antero-posterior diameter of the endometrial collection) secondary to vaginal canal agenesis. There was so-called cervix noted about 4 centimeters from the hymenal ring. (Figure 4)

A conference involving the Obstetrics and Gynecology, Infectious Diseases, Reproductive Endocrinology and Infertility, Urogynecology, Plastic and Reconstructive Surgery, Pediatrics, Psychiatry and Radiology, was made to arrive at the most appropriate management on the patient. A surgical approach was decided to address the cyclical pain by providing an outflow tract to the menstrual blood and sexual satisfaction to the patient in the later part of her life, and if successful, this may also provide a chance to conceive a child. But the limitations include, in an early age, the patient will be exposed to vaginal dilatation to maintain the patency of the neovagina, a second time surgery may have lesser success outcome due to a possible fibrosis formation to the 1st explored vaginal tissue. The extent of the success depends on how well the surgery was done, the motivation and maturity of the patient as to how well she understood the treatment plan and her role in the success of the surgery by doing frequent vaginal dilatation and follow-up until the desired opening and vaginal length is achieved to provide an outflow tract. The creation of a neovagina with bipudendal flap to provide an outflow tract of the cyclic menstruation was the plan of the service. The patient and her family were counseled and referred to the Department of Pediatrics for medical clearance and Department of Psychiatry for counseling to help in the patient's preparation physically, psychologically and emotionally. The patient was then cleared and scheduled for the said operation.



Figure 4. Magnetic Resonance Imaging

Intraoperatively, cystourethroscopic findings revealed normal urethral meatus and mucosa. The ureteral orifice was appreciated. There was a 2x2 centimeter bump-like extravesical mass noted inferior to the trigonal area pushing the trigonal spaces. (Figure 5) Perineal exploration was initiated by forming a “U” incision along the vaginal dimple. (Figure 6) Blunt dissection with concomitant direct rectal examination was done to create a plane between the bladder and the rectum. The vaginal length was 5 centimeter and can accommodate 1 index finger. The cervix was identified and noted to be rudimentary and effaced. It was punctured and hematometra (100 milliliter) was evacuated. The cervix was tagged using vicryl suture in 12, 2, 6, 8, and 10 o’clock positions. The Plastic and Reconstructive Surgery service proceeded with the creation of neovagina. Bipudendal flap/Modified Singapore flap, was harvested along the thigh crease measuring 7x3 centimeter, the posterior skin margin was at the level of the introitus. (Figure 7.1) The flap was raised



Figure 5. Cystourethroscopy



Figure 6. Perineal Exploration

along with the deep fascia of the thigh and epimysium of the adductor thigh muscles. (Figure 7.2) The flap was tunneled under the labia (Figure 7.3) and sutured at the midline to form the vaginal pouch. (Figure 7.4) The flap tip was anchored to the previously tagged cervix. The mucosal edge was sutured to labia minora. (Figure 7.5) Closure of the donor site was done using vicryl 3-0 suture, and a vaginal tube was inserted to maintain the opening. (Figure 7.6) The patient has an uneventful post-operative recovery. The removal of the vaginal tube was done after 1 week with intact sutures on the cervix and the neovagina

was noted with good flap take. The neovaginal opening admits 2 fingers with ease and the length was 4-5 centimeter. Transrectal ultrasound was done 10 days post-operative which revealed thin endometrium, normal-sized anteverted uterus and ovaries with several follicles, consider bilateral hydrosalpinges, minimal pelvic ascites with adhesions. The external cervical os was dilated with fluid within. There was a minimal vaginal fluid (6.2 milliliter). (Figure 8) The patient was discharged improved and daily vaginal dilatation using a modified dilator made of plaster of Paris patterned to a Hegar's dilator, with condom sheath. (Figure 9)



Figure 7.1. Flap Marking



Figure 7.2. Flap Harvesting



Figure 7.3. Flap Tunneling



Figure 7.4. Vaginal Pouch Formation



Figure 7.5. Flap Insetting



Figure 7.6. Vaginal Tube Insertion

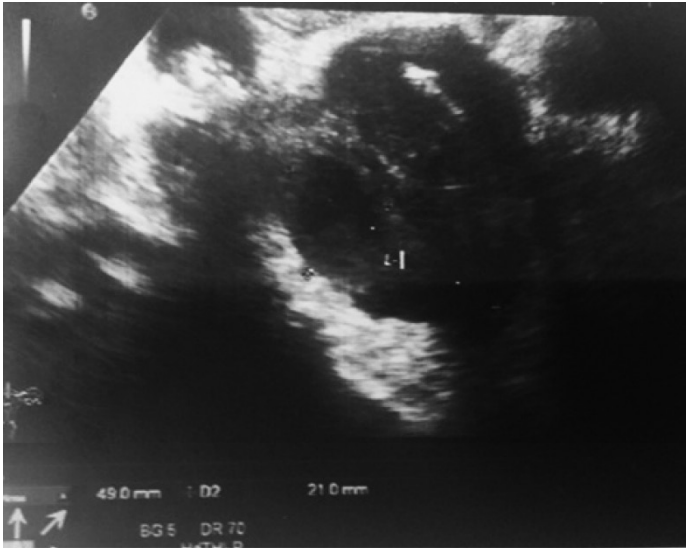


Figure 8. Post-Operative Transrectal Ultrasound

After 2 months from the reconstruction, the patient was seen without episode of menstrual flow, no recurrence of hypogastric pain and a flap in good condition but a stricture was noted in the neovagina. A repeat Transrectal ultrasound (Figure 10) was done which revealed a normal cervix with hemato/hydrocolpos (2.43 milliliter), thin endometrium with hemato/hydrometra (1.44 milliliter). A small anteverted uterus and unilocular cystic right ovary with benign features were seen. The left ovary was small. There was no free fluid. The patient was readmitted and vaginal dilatation under spinal anesthesia was done. The vaginal canal was advanced and a stricture was noted at the proximal 3rd of the vagina. (Figure 11) The neovaginal length was measured 9 centimeter and admits 1 finger. The patient was discharged and advised to continue vaginal dilatation with the use of the modified vaginal dilator. After five months from the reconstruction, the patient was with normal menstrual flow and noted irregularly occurring up to the present.



Figure 9. Modified Vaginal Dilator

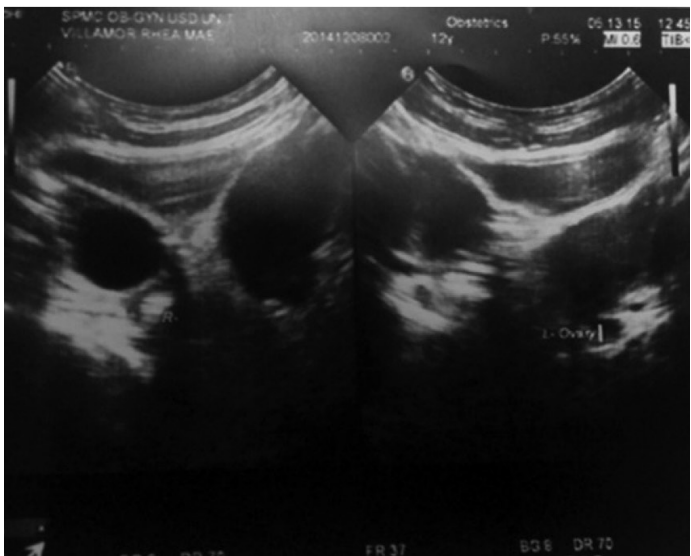


Figure 10. Transrectal Ultrasound (2 months after the reconstruction)



Figure 11. Neovagina(after 2 months)

CASE DISCUSSION

This is a case of a 12-year old nulligravid who presented a chief complaint of hypogastric pain and primary amenorrhea. She has normal development of the secondary sexual characteristics. Perineal examination revealed no vaginal opening but a dimple that ends in a pouch. Sonography showed hematocolpometra due to an outflow tract obstruction with a consideration of a complete transverse vaginal septum.

The differential diagnoses include transverse vaginal septum, and imperforate hymen. The patient with a low transverse vaginal septum will have a normal hymen with more proximal obstruction of the vaginal canal. The patient with an imperforate hymen will not have the typical fringe of hymenal tissue. In addition to presenting with primary amenorrhea, the 2 conditions occur with symptoms of cyclic abdominal or pelvic pain and a pelvic mass due to the obstructed outflow tract and associated hematocolpos.⁴ *The index patient belonged to tanner*

stage 2 both breasts and pubis and an ultrasound result confirming the presence of uterus with hematometra, fallopian tubes with hydro/hematosalpinx and ovaries. Primary amenorrhea secondary to an outflow tract obstruction such as transverse vaginal septum or vaginal agenesis was highly considered. The different causes of the outflow tract obstruction were discussed with the patient and her parents. Treatment options such a creation of neovagina, constant vaginal dilatation and their complications were discussed before the operation.

Vaginal agenesis is the congenital absence of vagina, there is failure of formation of the sinovaginal bulb from the fusion of the caudal end of the mullerian duct system and the urogenital sinus. Patients with vaginal agenesis typically present with primary amenorrhea in adolescence with normal growth and development. The female genital tract developed from 3 origins. The ovaries developed from the germ cells that migrate from the yolk sac into the mesenchyme of the peritoneal cavity and developed into an ova and supporting cells. The lower 3rd of the vagina developed from the ascending sinovaginal bulbs that came from the fusion of the mullerian ducts and urogenital sinus. The uterus, fallopian tubes, and upper 2/3 of the vagina developed from the fusion of the two mullerian ducts.⁵ *The failure of the fusion of the caudal tip of the mullerian ducts and the tissue of sinovaginal bulbs will result to vaginal agenesis that is presented in our index patient with no noted vaginal opening on physical examination.*

Diagnosis of vaginal agenesis is quite difficult and requires high index of suspicion. Since most patients presenting with a clinical profile of vaginal agenesis are adolescents with no previous sexual contact, examination is limited to a direct rectal examination making it more difficult in differentiating a transverse vaginal septum from a vaginal agenesis. Laparoscopy may be inconclusive and costly. Three-dimensional ultrasonography has reported higher accuracy rate over conventional 2-dimensional scanning. In experts' hands, 3-Dimensional ultrasound has sensitivity of 93 % and specificity of 100%.⁶ MRI is the gold standard in the imaging evaluation with a reported accuracy rate of 100 %.⁷ It has the ability to delineate uterine, cervical, and vaginal anomalies; and not an operator dependent, not limited to bowel loops, which is also the advantage over sonography. *Both 2-Dimensional ultrasonography and MRI were done in the index patient. Both can help make an accurate diagnosis of the anomaly when findings are correlated with a complete history and thorough physical examination of the patient.*

The management of vaginal agenesis is challenging and with limited experience. Hysterectomy with ovarian preservation is still the proposed treatment option. But recent advances in the reconstructive surgery,

laparoscopy, and assisted reproductive technology make room for conservative management in carefully selected group of patients to achieve pregnancy in the future. This includes canalization, uterovaginal anastomosis and vaginal reconstruction. The overall success rates of these techniques are about 60 %. There are 6 reported cases with successful pregnancies after restoration of the uterovaginal route.⁸

Creation of neovagina with vaginal agenesis may be done by several methods. The operations develop the potential space between the bladder and the rectum and replaced this space with a stent utilizing tissue, most commonly a split-thickness skin graft or synthetic materials. *In our index case, neovagina was created with the use of bipudendal flap, also known as Modified Singapore flap.* Bipudendal flap is a versatile fasciocutaneous flap, reliable and with robust blood supply. It is a sensate flap since it retains pudendal nerve and posterior cutaneous nerve of the thigh.⁹ The bipudendal flap constructs the vagina in congenital and acquired conditions and is simple and at the same time safe. The reconstructed vagina has a natural, physiologic angle and it has inconspicuous donor site scarring.¹⁰ Overall clinical results as to the index patient include 100% flap survival, simple technique, operative time of about 2-2.5 hours, there was little blood loss, and early wound healing, no stents needed, there was physiologic angle of inclination, the linear scars are well hidden in the groin crease and a sensate reconstructed vagina was formed. Further investigations and follow up

to the patient is necessary to check for the long term patency, flap healing and the most awaited is to gauge the success with the passage of menstrual flow as to creating a successful outflow tract in a case of vaginal agenesis.

In the index patient, the uterovaginal canalization was attempted due to the presence of the cervix and creation of neovagina would be the best way to do such treatment. Sustained psychosocial support is of utmost importance in patients undergoing conservative management, especially since most patients, like the index patient, belong to the adolescent age group and they may be assisted to fully comprehend the extent of their condition.

SUMMARY

Vaginal agenesis represents one of the challenging disorders in Gynecology. Thorough history and physical examination are needed to arrive at a proper diagnosis. Vaginal agenesis, although a rare cause of outflow tract obstruction, clinicians should consider in adolescent patients presenting pain and amenorrhea. Ultrasonography and magnetic resonance imaging will aid in the correct diagnosis and planning for the appropriate management. In patients with vaginal agenesis, the goals of the management are relief of symptoms of pain and preservation of sexual and reproductive function. Early and accurate pre-operative diagnosis, comprehensive treatment plan, careful selection of patients and holistic approach are the cornerstone of the management. ■

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