

Case report: Successful Pregnancy In A Patient With Isolated Subtotal Vaginal Agenesis Accompanied With A Cervical Vestibular Fistula Which Has Been Treated Surgically

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Abstract:

Obstructive reproductive tract anomalies usually diagnosed in first year after menses should have occurred, so because the menstrual blood cannot get out, it accumulates resulting in hematocolpos or hematometrocolpos which in turn causes pelvic cyclic pain raised in severe by time associates primary amenorrhea.

We report a case of isolated subtotal vaginal agenesis -which is one of the obstructive anomalies- but without facing any complication before getting married except some pain during menses because there was a fistula making it possible to pass out menstrual blood. It is important to detect the exact malformation in order to conduct a successful following surgery. we mention its surgical treatment in which we use peritoneum as a flap to line neovagina and the following up of the patient who started to use frank's dilators after surgery for some time to improve vaginal length and prevent stenosis.

Key Words: Isolated Subtotal Vaginal Agenesis, Cervical Vestibular Fistula

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تقرير حالة: حمل ناجح عند مريضة عدم تصنع مهبل شبه تام معزول مترافق مع ناسور عنقي دهليزي تم تدبيره جراحياً

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الملخص:

تشخص تشوهات الطريق التناسلي الانسدادية غالباً في السنة الأولى بعد أن يبدأ الطمث، وبسبب عدم امكانية خروج دم الطمث فإنه يتجمع في المهبل أو الرحم والذي بدوره يسبب ألم حوضي دوري يزداد بشدته بمرور الزمن مترافقاً مع انقطاع طمث بدني. لقد قمنا بتسجيل حالة عدم تصنع مهبل شبه تام معزولة -والذي هو واحد من التشوهات الانسدادية- لكن دون وجود أي شكوى قبل الزواج سوى بعض الألم أثناء الدورة بسبب وجود ناسور جعل من الممكن لدم الطمث الخروج خلاله. من المهم تحديد التشوه بدقة لاجراء تداخل جراحي لاحق بشكل ناجح. قمنا بذكر تدبيره الجراحي والذي قمنا فيه باستخدام البيروتوان كشريحة لتبطين المهبل الجديد ومتابعة المريضة التي بدأت باستخدام موسعات فرانك بعد الجراحة لبعض الوقت لتحسين طول المهبل ومنع التضيق اللاحق.

الكلمات المفتاحية: مهبل شبه تام معزول، ناسور عنقي دهليزي.

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Introduction:

Cellular differentiation, migration, fusion, and canalization are a series of events that occur in the course of the development of the female reproductive pathway.

Therefore, a defect in any of the previous stages leads to congenital anomalies, which in turn may affect menstrual cycles, sexual activity, infertility or childbirth later. Mostly after puberty, but it may be detected during childhood (1. **Stallion, A., 2000, Vol. 9, No. 3, pp. 128-134**). It is important to ascertain here that the development of gonads is independent of the development of the reproductive pathway, so those women who have ovaries and normal levels of female hormones. However, this does not negate the existence of urinary abnormalities and anomalies, since the female genital tract closely associated with the development of the kidneys, ureters and bladder.

Therefore, the accompanying urinary malformations in these patients must be excluded by appropriate radiographical procedures (1 **Stallion, A., 2000, Vol. 9, No. 3, pp. 128-134**, 4. **Laufe, M.R., 2019, Stand: Oktober 2021**). To date, there is no universally accepted classification system for abnormalities of the reproductive tract, which is a problem nowadays (2). Vaginal agenesis is one of these abnormalities that result from obstruction of the uterovaginal path and occurs when part of the vagina fails to form, where it is replaced by fibrous tissue.

Vaginal atresia or vaginal aplasia may be an isolated developmental defect (rare) or as part of a complex abnormality (more common). The management is either non-surgical using vaginal dilators or surgical with various interventions (1. **Stallion, A., 2000, Vol. 9, No. 3, pp. 128-134**, 3. **Linder, B.J., 2021, 453-455**). In our case, the vaginal agenesis included the most of the vagina with a fistula that allows the passage of menstrual blood. This case was managed surgically by laparotomy and using the peritoneum as a slide to line the new vagina and continue with the use of dilators after surgery.

Clinical presentation:

A 25 years nulliparous patient presented to the University Hospital of Obstetrics and

Gynecology in Damascus, she has been married for 4 months, and presented with a complaint of dyspareunia, and lower abdominal pain for two months. She had no complaints before marriage and her menstrual cycle was normal in terms of duration and amount of menstrual blood but she suffered from dysmenorrhea. She had no past medical or family history.

She reported a story of a surgical operation 5 days ago, after which it was faulty diagnosed with a transverse vaginal septum therefore she underwent a failed surgical operation and the patient was informed of the need to visit a specialized tertiary hospital. In our hospital, an Ultrasound of the pelvis and abdomen was performed to evaluate the urinary and reproductive tract which showed that the kidneys were apparently normal in size and echogenicity, the bladder was within normal limits (Figure 1). The uterus was homogeneously echogenic, with regular walls and lining of normal thickness in relation to the menstrual cycle, measuring about 12 mm. The ovaries were in normal size with homogeneous appearance and contained many peripheral and central follicles (figure 2). Laboratory tests were performed and were within normal limits: Hemoglobin: 11.6 mg/dl, platelets: 198,000/ μ l, creatinine: 0.89 mg/dl. Vaginal examination showed a fistula that reaches the cervix and through which the menstrual blood passes to the remaining vagina without any signs of blockage of blood flow. We decided to perform a surgical procedure based on the patient's desire after discussing the available conservative and surgical treatments, the advantages and disadvantages of each, and ascertained on the necessity of adhering on the recommendations following the surgery and their importance for the success of the surgical procedure and that it may take a period of time to reach the best result. The surgery was performed under general anesthesia and sterilization of the abdominal area, vulva and vagina with povidone-iodine was taken. We catheterized the bladder using a Foley catheter, then laparotomy was performed through Pfannenstiel incision to reach the abdominal cavity, where the uterus and appendages appeared of normal

size and shape (Figure 3), and through a vaginal approach, a second surgeon made a transverse incision with a length of 1.5 cm under the orifice of the urethra by about 2 cm, then a kocher clamp was inserted through the incision towards the pelvis, taking into account that no damage was caused to the bladder, while the other surgeon dissected the peritoneum that covering the uterus and bladder, then an incision was made in the place of the vault that occurred as a result of the insertion of the kocher through the vagina. A blunt dissection was performed downward to meet the other surgeon who used the kocher until the full technique was performed between the bladder anteriorly and the uterus posteriorly, then peritoneum was pulled through the vesicouterine diverticulum into the new canal and sewed it on the front surface of the vestibule of the vagina (figure 4), then the peritoneum covering the uterus was sewed on the bladder in order to close the peritoneum. Thus, the anterior wall of the vagina was formed by the peritoneum, while the posterior wall of the vagina was formed from the fibrous tissue of the obstructed vagina and the peritoneum covering the rectum. After the reconstruction the cervix appeared to be in an anterior position, but with follow-up we noticed that the cervix returned to its normal position. The patient was trained to put vaginal dilators several times a day, gradually with increasing their sizes for a period of 40 days, and the patient was trained to use the dilator step by step and the way to clean them. Disinfected vaginal creams were used. The patient was discharged home. Follow-up was arranged frequent and the patient was guided to have intercourse after 40 days. Through the follow-up visits, the vagina increased in dimensions, and the cervix returned to its normal central position. The patient was pregnant and pregnancy was confirmed by ultrasound.

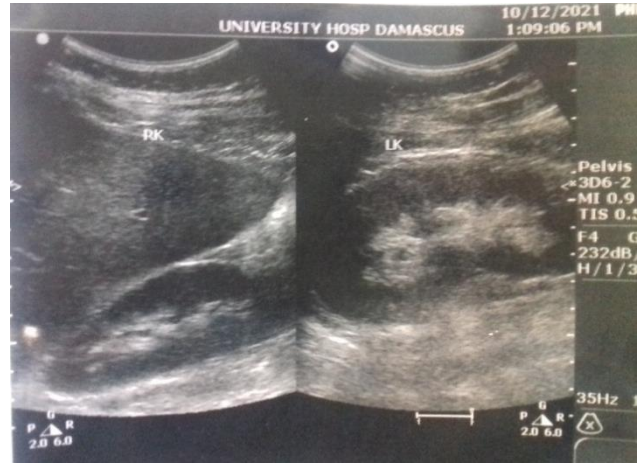


Figure 1



Figure2

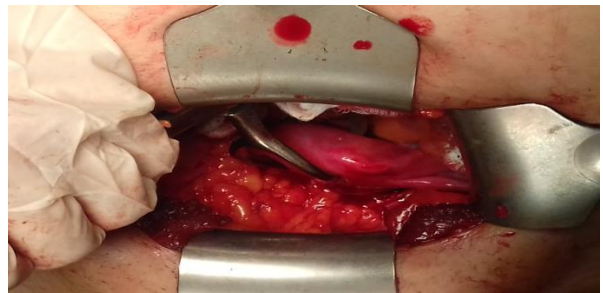


Figure 3

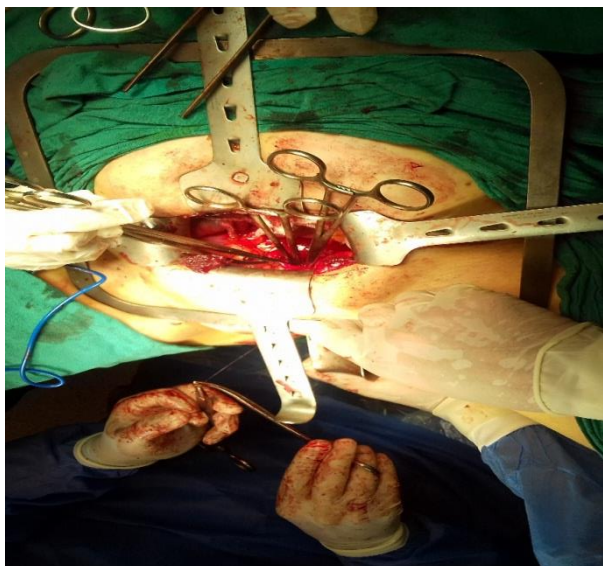


Figure 4

Discussion:

Mullerian's ducts develop in the absence of androgen and anti-mullerian hormone to form the female reproductive tract and external genitalia, and this occurs in conjunction with the regression of the Wolff's ducts. The two Muller's ducts begin to elongate caudally by the sixth gestational week to fuse medially by the eighth week, forming a single tube in the middle called the uterovaginal canal, which will eventually give rise to the uterus, cervix, and the upper third of the vagina. The development of the vagina begins at week 12 starting from the Muller tubercle where the uterovaginal canal meets the urogenital sinus. The vaginal plate is formed from the fusion of the two bulbs of the vaginal sinus, and by week 26 it has completed its growth and becomes a cord of cells. Central cells erupt and desquamate in the vaginal lamina from the caudal direction to the cephalic, resulting in the formation of vaginal lumen.

The anterior part of the urogenital sinus forms the bladder and urethra, while the posterior part forms the vestibule of the vagina and hymen. (1 **Stallion, A., 2000, Vol. 9, No. 3, pp. 128-134**

..5. **Miller, R.J, 2008, 51(2), pp.223-236**) Therefore, it is important to know the embryonic development of the genital tract and its stages, as any defect in any of the previous stages leads to a specific abnormality.

Similarly, changes that occur in the balance between genetic, developmental and hormonal influences can lead to a wide range of abnormalities of the reproductive system, including vaginal agenesis. Genetically, several genes have been identified to have a strong influence on the development of the Muller duct. However, no study has found a single gene mutation responsible for the abnormalities of the Muller duct (7. **Dietrich, J.E., 2014, 27(6), pp.396-402**).

The defect in the development of the urogenital sinus constitutes most of the developmental problems, which explains the frequency of abnormalities of the lower part of the vagina and the hymen, and the association with the occurrence of urinary abnormalities in many cases. Vaginal aplasia is the congenital absence of the vagina or part of it with an incidence of 1/4000-5000 girl with normal karyotype and functional ovaries, while the term vaginal atresia is best used to describe vaginal obstruction secondary to radiographic treatment, previous surgery or trauma, where it is classified into complete, which is often associated with abnormalities of the cervix, or partial including the proximal or distal part of the vagina, and often the distal atresia is better in terms of ease and results of reconstruction and less association with post-operative stenosis.

Agenesis in our case included the most of the vagina except a small normal portion in the distal part of vagina about 2 cm length. which had a small canal or fistula with a diameter of 3 mm that connects the cervix to the vestibule of the vagina (figure 5), which explains the absence of symptoms before marriage, the cervix appeared healthy during surgery. (1. **Stallion, A., 2000, Vol. 9, No. 3, pp. 128-134**, 8. **Mei, L., 2021, 61(4), pp.127-13**) There are several classifications of female reproductive tract malformations, including the AFS classification, which is based on the anatomy of the reproductive system, the American Society of Reproductive Medicine (ASRM), the VCUAM classification, and other classifications based on fetal origin or associated urinary anomalies.

Despite this, there are still cases that the doctor must decide to categorize them with a group of abnormalities, as they lie between them, and there are other cases in which the diagnosis before surgery

was not precise, so accurate classification is important in choosing the appropriate surgical technique and avoiding the complications of such difficult surgeries, which explains the need to classify them into clear, easy way to handle. (9 *Acién, P, 2011., 17(5), pp.693-705.*) The diagnosis is often made at the time of puberty, which confirms the importance of conception of such anomalies and determin them at the time of puberty, the menstrual blood is trapped and does not come out to collect in the vagina or uterus, leading to periodic pain with the absence of menstrual blood, which later develops into pain crises that increase in intensity with time, and we may be able to palpitate a mass abdominally. Ultrasound is used in the diagnosis, which is the first and least expensive method, but magnetic resonance imaging is considered the golden method of diagnostical imaging. (1. *Stallion, A., 2000, Vol. 9, No. 3, pp. 128-134.*, 7. *Dietrich, J.E., 2014, 27(6), pp.396-402*) The collection of blood in the vagina helps to limit the use of slides, as it naturally takes the place of mechanical dilators, so it is preferable to postpone the surgery so that the blood pool grows, but before it reaches and accumulates in the uterus.

However, in our case, the blood pool didn't accumulate because of the presenting fistula, which was draining menstrual blood and preventing its collection. (5. *Miller, R.J, 2008, 51(2), pp.223-236*)

In our case, there was a fistula that connects the cervix to the remaining part of the vagina, which contributed to the occurrence of normal but painful menstrual cycles, however when the patient got married, she suffered from dyspareunia so she went to a hospital and the case was misdiagnosed as a transverse vaginal septum, where surgery was performed to resect the septum without success. It is important to search for the associated urinary malformations, due to the high incidence of renal malformations 40% and vertebral malformations 10-20% and to ensure that there is no cervical dysplasia, and non-dystrophy may be associated with other syndromes associated with polydactyly, congenital heart malformations, Retinal dystrophy and others. (1. *Stallion, A., 2000, Vol. 9, No. 3, pp. 128-134.*, 7. *Dietrich, J.E., 2014, 27(6), pp.396-402*)

The ideal treatment method is still unknown, and management is conservative using noninvasive or surgical, either conventional or endoscopic.

Treatment with dilators are used to increase the length of the vagina in patients with vaginal aplasia or in case of vaginal stenosis, whether after surgery or radiotherapy. Frank's method involves the use of gradually expanded dilators, and later Dr. Ingram used dilators attached to a bicycle seat. The use of dilators is less expensive and less complicated compared to surgical methods, and has a shorter hospital stay with no surgical scar, but this method may take longer to obtain the result This procedure is personal and is linked to the patient's interaction and complete desire and willingness to perform it despite the length of time. The length of the new vagina is directly proportional to the duration of use of the dilators. There are several surgical methods, the aim of which is to manufacture a vagina of appropriate length and breadth in a suitable anatomical axis to achieve comfort and sexual pleasure, and these methods are: Duptryen and sheares: making a cut between the bladder and intestine and inserting a connection and leaving it to allow appearance, vecchietti and balloon: a continuous mechanical expansion procedure surgically and the modified procedure Theoretically, the Mcldoe procedure, where manufacturing is done using a skin slice, and one of the disadvantages of this procedure is the defect where the skin slice was taken, Davydov's procedure, where the peritoneum is used as a slice for manufacturing, and we have adopted this method, and the labia or the oral mucosa can be used as a slide. The part of the intestine most used historically and is done using the ileum or the jejunum. The largest length of the vagina was obtained after fabrication using this method. (1. *Stallion, A., 2000, Vol. 9, No. 3, pp. 128-13.*, 3. *Linder, B.J., 2021, 453-455.*, 5. *Miller, R.J, 2008, 51(2), pp.223-236.* 6 *McQuillan, S.K, 2014, 25(3), pp.299-311*

.,11. *Wu M, 2020 Nov 19;40(12):NP694-NP702*) Obtaining sexual pleasure is one of the indicators of the success of reconstruction, and there are certain criteria to measure the degree of pleasure after the various measures. (10. *Dabaghi, S. 2019, J 30, 353-362*) Our surgical option must be appropriate and successful in the beginning, based on accurate clinical evaluation and on ultrasound and magnetic resonance imaging if necessary to establish an appropriate diagnosis and classify the deformity and

associated malformations, because failure in the first surgery increases the risk of the next intervention and increases the chance of injury to neighboring organs during the procedure (9. *Acién, P, 2011., 17(5), pp.693-705*), and this What happened in our case, where the initial diagnosis was wrong and lead to a wrong surgical procedure, which made the management more difficult and dangerous.

The expansion following most of the surgeries of its variety is required and necessary to reduce the risk of stenosis, where the patient must be taught how to apply the expanders step by step and the need to empty the bladder before dilation, topical lidocaine can be applied 20 minutes before using the expander to allow it to be absorbed and reduce the pain resulting from the expansion, and the patient must be taught How to wash the expander with soap and water, and the need to apply the expansion to 1-3 times a day for 10-30 minutes each time immediately after the surgical procedure, or you can wait 4-6 weeks. A period when the patient feels ready and willing to do so to increase the expansion, of course, with the support and assistance of the partner, which also has an important role in improving results. Duration of application of expanders and replacement of expanders later. Strategies for the prevention of sexually transmitted diseases and HPV should be discussed. In cases of high risk of stenosis, it is recommended to continue dilating 3 times a week for two years. The patient must be closely followed up and advised to visit the clinic frequently and inform the doctor when bleeding, pain or urinary symptoms occur. In each visit, the improvement of the condition must be monitored, the length of the vagina measured, and the question about the previous symptoms, a topical estrogen cream can be prescribed to improve the elasticity of the vaginal mucosa, reduce bleeding or suggest comfort A day in the event of a small amount of bleeding, and a urine analysis and culture must be performed when urinary symptoms appear, and to ensure that the urethra does not expand or pressure on the bladder during the procedure and expand at the wrong angle, and exercises must also be performed Kegel exercises should also be performed to strengthen the pelvic floor. (12. *Oelschläger, A.M.A. 2019, 32(4), pp.354-358*)

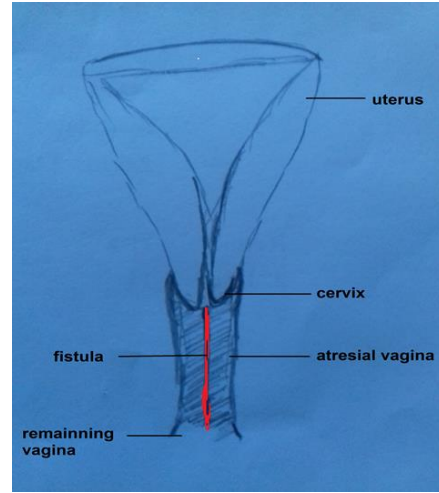


Figure 5

Conclusion:

It is necessary to be familiar with the exact anatomy of the genital area, with a good knowledge to the embryonic origin of each part of the genital tract. This is necessary to classify the malformation and anticipate its severity, extent, or association with other urinary or reproductive malformations in order to decide the appropriate treatment. It is necessary to emphasize on the importance of establishing the correct diagnosis while denying or confirming the presence of accompanying urinary abnormalities to choose the appropriate option that will achieve the best result in order to spare the patient the risks of fruitless surgery that may adversely affect her due to the poor prognosis. The procedures that follow the surgical procedure, such as the use of the expanders and the application of recommendations to prevent side complications, are of importance as they guarantee the preservation of the results of the surgery and even reach better results and prevent the subsequent stenosis, so attention must be taken of it and to the training the patient in detail with periodic follow-up of the results of the expansion. The patient's full conception and awareness of the nature of the treatment and its compliance and application of the recommendations is the basis for the success of the reconstruction process and the preservation and improvement of the obtained result.

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