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Reconstruction of the Congenital Disorders of the Female Genitalia: Discontinuitas Uterus Cervik

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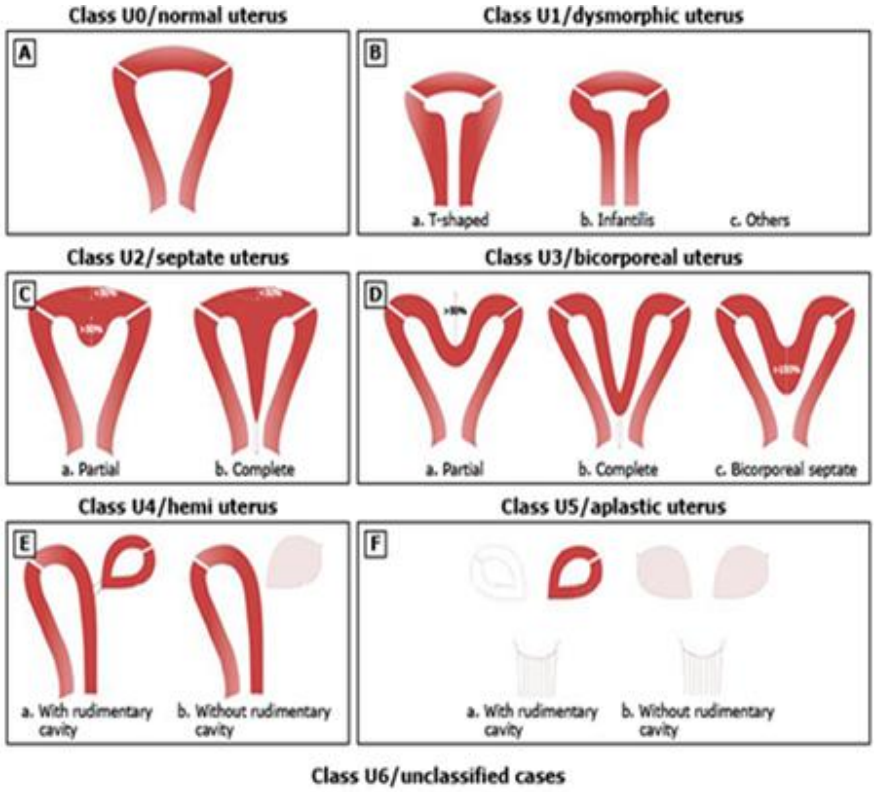
Case Report: A new type of anomaly of mullerian channel development: discontinuitas uterus cervix

Abstract: ***Congenital abnormalities of the uterus and cervix are abnormalities of female genitalia that are usually caused by an anomaly in the development of the Mullerian ducts. We report an operative management for congenital abnormalities of the uterus that is disconnected between the cervix and the uterine body. This disorder does not conform to the consensus classification approved for congenital uterine anomalies by ESHRE / ESGE in 2013 and there have been no previous reports of similar cases. The patient in this case was a 14 year old woman with the chief complaint of having never had a first menstrual period but was suffering from severe monthly lower abdominal pain. On investigation shows that the uterine body appears separate from the cervix and there is a cyst that is suspected to be an endometrioma. Uteroplasty is performed to create anastomosis and canalization between the uterus and cervix using a nelathon catheter that is placed for 3 months. The patient had menstruation about 1 month after surgery and there was no pain. Operative management in this case significantly improved the patient's quality of life.***

Keywords: Congenital Anomaly Uterus; Ductus Mullerian; Uteroplasty

1. Introduction

Congenital uterine anomaly is a female genital disorder caused by an anomaly in the development of the Mullerian duct. The prevalence of congenital uterine abnormalities is about 4 - 7%[1]. A classification system has been proposed for female genital tract anomalies, for effective diagnosis and treatment. However, in this case, uterine and cervical disconnection was found that had not been included in the conventional classification published by ESHRE / ESHE in 2013[2]. To date, there have been no previous reports of uterine anomalies with the cervix separated from the uterine body. In this case report, we present management for anastomosis as well as canalization between the uterus and cervix[1]. The case was published with the written consent of the patient and his family.

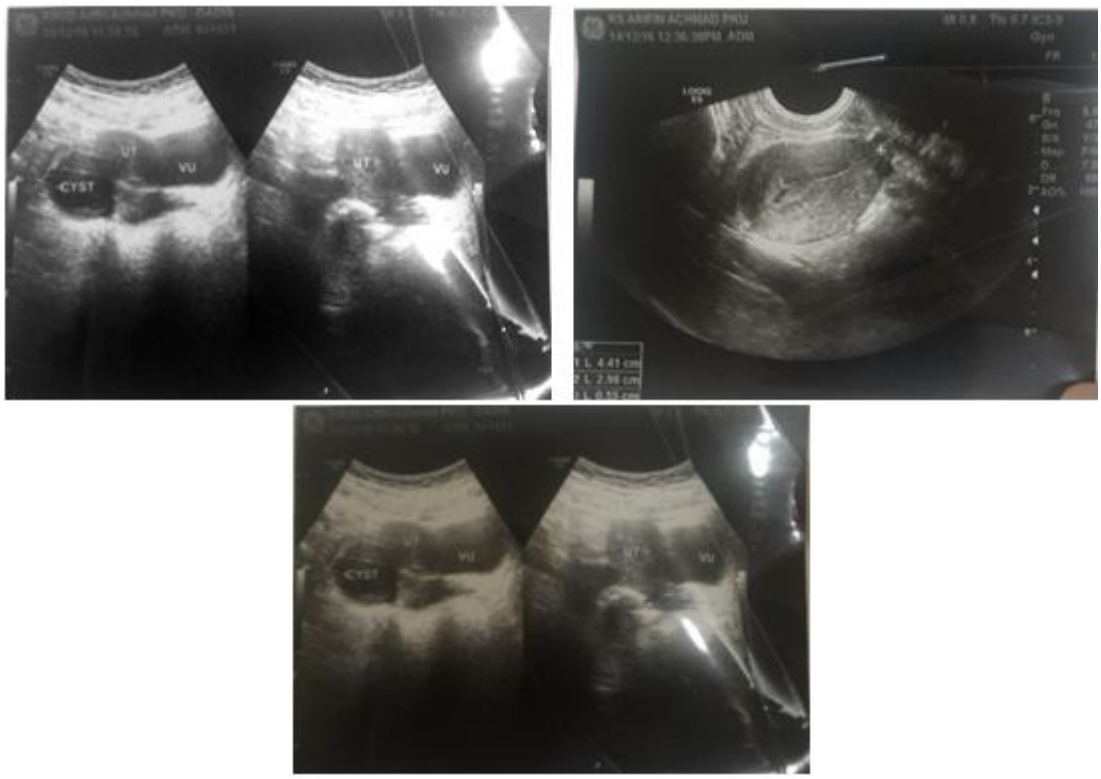


Picture 1: ESHRE / ESGE Classification Scheme for Mullerian Tract Development Anomalies

2. Case Illustration

A woman complained that she had not had a period even though she was 14 years old. Growth of breasts, fat in buttocks and thighs, growth of fine hairs on the folds of the arms and pubis according to age. The patient also felt lower abdominal pain which occurred periodically every month from the age of 2 months, but there was no menstrual bleeding at all. Lower abdominal pain is only felt for 1 - 2 days, then the pain decreases slowly until it goes away in about 4 - 5 days. The patient did not complain of a lump in the lower abdomen, complaints of sudden weight loss, decreased appetite, or urinary disorders and defecation.

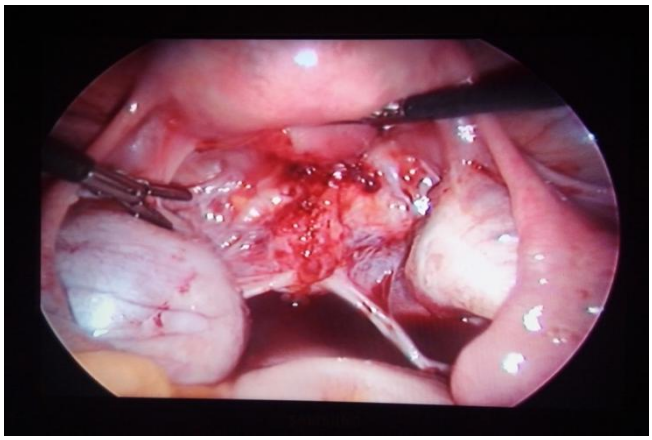
On the physical examination by Rectal Toucher by consultant urogynecologist, there was a vaginal impression within normal limits, there was no hematocolpos, cervical palpable, and there was a cystic mass in the left adnexa suspected of an endometriosis cyst. The patient was then consulted to the endocrinology department and performed a trans rectal ultrasound examination. The uterus was normal in shape and size, positive endometrial line. The cervix is not connected to the uterus and there is a mass in the left adnexal measuring 3.5 x 3.0 cm and free fluid in the Douglas cavity.



Picture 2: Ultrasound Examination Results

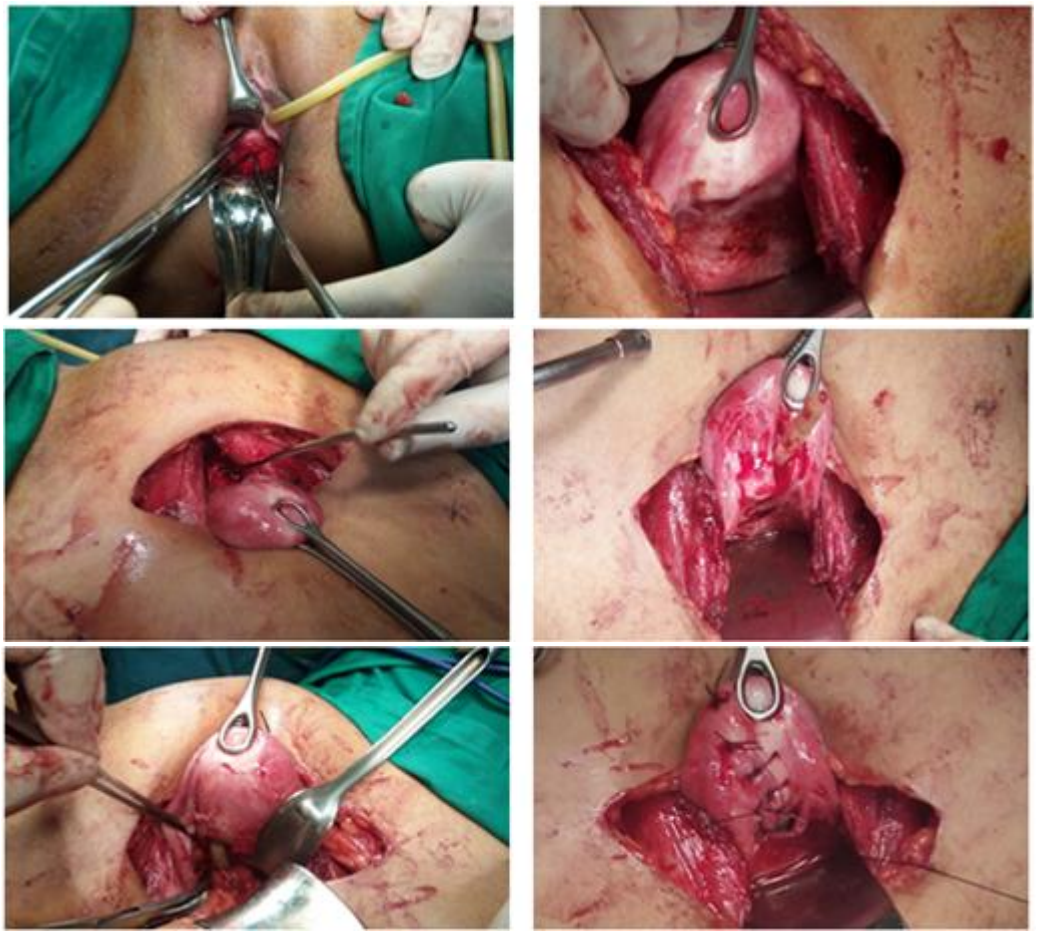
The CT scan results also showed a normal shape and size of the uterus with a cystic mass of 3.98 x 3.89 x 3.53 cm in the left adnexal. In other intra - abdominal organs, the impression is within normal limits. The patient was later diagnosed with Amenorrhea Primer et causa congenital uterine abnormality (uterine - cervical disconnection) with endometriotic cysts. The patient is then prepared for laparoscopy in preparation for laparotomy and repair which will require inspecular action. The family received the explanation and agreed to take the action planned by the team from Sanglah Hospital –Bali.

On January 16, 2017, a laparoscopic procedure was performed. The uterus appeared normal in shape and size, both tubes were normal, the left ovarian cyst was the size of a chicken egg, the impression was a brown cyst. The right ovary appears normal and menstrual blood appears to fill the cavity of Douglas. On exploration of the posterior part, there is non - communicating between the uterus and the cervix with the peritoneal tissue as a connector. In the caudal part, the position of the sacrouterine ligament is shifted more inferiorly and is relatively smaller in size. It was decided to do a laparoscopic cystectomy, a brown discharge from the impression of an endometriosis cyst. The cyst capsule is evacuated and the abdominal cavity washed. The procedure is followed by a laparotomy to perform anastomosis and cervical uterine canalization.



Picture 3: View of the Structure of the Internal Genitalia through Laparoscopy

A pfannenstiel incision was performed, the abdomen was opened layer by layer, after the peritoneum was opened, the uterus, two tubes, and both ovaries were seen within normal limits. On palpation, the uterus is palpable apart from the cervix, it was decided to identify the cervix through the vagina. Inspeculo was performed and the cervix was identified and then clamped with a tenaculum. The cervix was normal in size, but there was no cervical canal. With sondase, identification of the cervix is carried out by observation from the abdomen. It appears that the cervix is not united with the uterus. Formed a cervical canal with a sonde until it penetrates intothe peritoneum, inserting a 14 F foley catheter up to the intraperitoneum, the procedure is continued with perabdominam.



Picture 4: Uterine and Cervical Anastomosis Reconstruction Stages

The catheter from the cervix was identified, it was decided to do a hysterotomy by penetrating the base of the uterus. Canalization is performed with a foley catheter inserted through the artificial canal to the uterine cavity, then suture fixation is performed on the fundus and catheter tip that is inside the uterine cavity with Silk no.2.0. Followed by hysteroplasty of the uterus with interrupted PGA thread no.1. The cervix and uterus were joined with interrupted sutures with PGA no.1. Make sure the uterus and cervix are well connected, there is no bleeding, the catheter balloon is filled with 3 - 4 cc of fluid. Ensure the catheter is properly fixed, the action is complete. Operation time is about 4 hours. It is planned to maintain the anastomotic catheter for 3 months and then the fixation node will be removed per laparoscopy while evaluating the uterine and cervical connection.

Postoperatively the patient was treated for 3 days given antibiotics then went home with a good postoperative wound and was planned for control every 2 weeks with education for sitz baths with antiseptic solutions during outpatient care. In a control, on February 2, 2017, the patient experienced menstruation on the 6th day, it appeared that menstrual blood came out of the vagina. Menstruation lasts for 7 days and is not accompanied by pain. There are no signs of infection and the catheter is still well fixed. It is planned to remove catheter fixation per laparoscopy 3 months postoperatively.

3. Discussion

Congenital abnormalities of the female genital tract are caused by disorders of the embryological development of the Mullerian or Paramesonephric ducts. The consensus for this classification of disorders has been published by the European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynecological Endoscopy (ESGE) [2, 3]. This case involved separating the cervix from the uterine body, and did not fit the conventional category of uterine anomalies. A literature search revealed no similar case reports. Therefore, this case represents a new type of female genital anomaly without developmental defects in the Mullerian duct.

At first glance this condition is similar to a condition called Mayer - Rokitansky - Küster - Hauser syndrome or MRKH syndrome. However, in MRKH syndrome what occurs is agenesis in the Mullerian duct and can be accompanied by agenesis of organ structures that are embryologically close together such as the kidney[4, 5]. It is not suitable in this patient because the uterus, ovarian tubes and kidneys and cervix are well formed. The only abnormality that occurs is a disconnection between the uterus and cervix.

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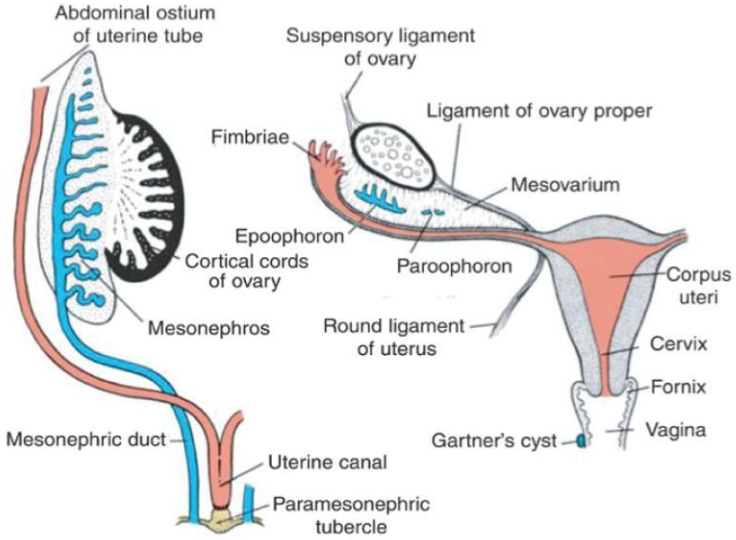

Picture 5: Embryological Development of Female Internal Genitalia

Preoperative evaluation by ultrasound and CT - scan is very important to understand the anatomical position of the uterus and adnexa for planning surgical reconstruction in this case[6, 7]. Penetrating and widening the tip of the cervix to the uterine cavity using a sondase and catheter is also effective for the prevention of cervical stenosis. However, care is required in this procedure as intrauterine insertion of the catheter can cause infection[1], although this was not the case in our case.

Anastomosis technique for the uterine and vaginal corpus is also performed in cases of radical trachelectomy which can be applied to conjugate the cervix with the corpus of the uterus[8]. We successfully anastomosed the separate cervix and uterine corpus using modified hysteroplasty. The tip of the catheter is fixed by suturing it to the uterine fundus using a silk thread to prevent the catheter from being pulled out if it is pulled by the patient. It was planned to untie the knot per laparoscopy 3 months after surgery while evaluating the results of the anastomotic suture that we did previously.

With the aim of facilitating periodic menstruation, as well as reducing monthly abdominal pain. The cause of lower abdominal pain that begins at the age of 12 years can be due to the excretion of menstrual blood because there is no communication from the uterus to the cervix so that the pressure in the uterine cavity becomes high and menstrual blood is excreted into the peritoneal cavity. This can also explain the emergence of intra - abdominal free fluid and the presence of endometriosis cysts in patients.

Before the patient underwent surgery, the patient complained that it was difficult to attend school every month because of lower abdominal pain. However, patients feel helped and can return to their daily activities after surgery because they have less complaints and have experienced their first menstruation. Therefore, reconstructive measures are very significant and greatly improve the patient's quality of life and even reduce anxiety in the patient's parents. However, it is unclear whether pregnancy will occur and what method of delivery is best for this case. Given the abnormal sacrouterine ligament, it is possible to develop uterine prolapse in the future. So even though menstruation can occur, long - term follow - up is needed to assess whether this treatment is really successful or not. Hopefully, this case report will trigger another report that could spark a new classification of mullerian channel development anomalies.

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