From Diagnosis to Motherhood: Management of Primary Amenorrhea with Vaginal and Cervical Agenesis Resulting in Successful Pregnancy - A 6-Year Follow-up Case Study

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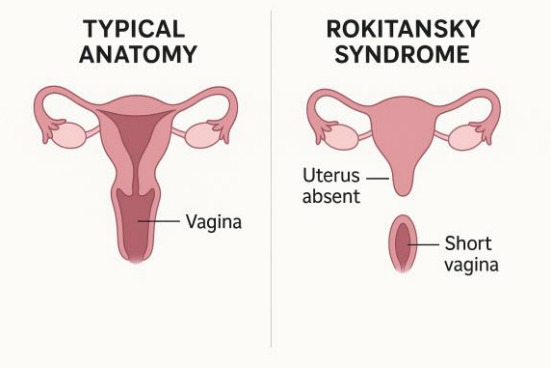
Background

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a genetic condition causing the absence or underdevelopment of the uterus and absence of the vagina, affecting approximately 1 in every 4,500 to 5,000 female newborns. This defect impacts fertility and reproductive health and can be divided into two types: type 1 (complete uterine loss with normal ovaries) and type 2 (skeletal and renal defects). Women with MRKH struggle to conceive or carry a pregnancy, impacting their experience of womanhood and motherhood. Here, we replicate a rare instance of an MRKH woman who successfully underwent neovaginal and neocervical construction and gave birth to a healthy baby.

Case

Our case was a 14-year-old female patient presented with primary amenorrhea in 2018 and underwent abdominal ultrasonography. She had a uterus measuring 38 x 58 mm with an endometrial thickness of 4 mm without cervix and vagina. In 2019, she returned with persistent amenorrhea and abdominal pain, and her uterus measured 96x44 mm with 3 mm endometrial thickness, hematometra, left hydrosalpinx, and fluid collection in the pelvis and posterior cul-de-sac.



she underwent combined vaginal and abdominal surgery for neovagina and neo-cervical creation due to primary amenorrhea and hematometra. a second surgery was performed in 2021 to address recurrent symptoms with indwelling catheter in the neocervix. in April 2023, a laparoscopic removal of the cervicouterine catheter was performed, she had a 6-7 cm neovagina. She married at 16 and we used to do cervical dilatation every 3 months. So she conceived 1 year after marriage, shortly after her final cervical dilation procedure. She had overt diabetes, and hospitalized at 34 weeks of pregnancy and a cesarean section was performed at 36 weeks (end of 2024) because of severe oligohydramnios, resulting in the delivery of a healthy infant.

Concluding message

Our case's unique, spontaneous conception process demonstrates how advances in reproductive medicine, such as uterine transplantation or genital tract construction, are enhancing the quality of life for women with MRKH and allowing them to achieve their motherhood dreams. Also, it is suggested to consider hysterectomy as the last option for women with cervical Agenesis.