

Spontaneous pregnancy after surgical management of Mayer Rokitansky Kauser Hauser syndrome.

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INTRODUCTION

Vaginal agenesis is a rare congenital anomaly associated with primary amenorrhoea and cyclical pelvic pain. The incidence is 1 in 4500 female births. The most common cause leading to this condition is Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome. It is a disorder that affects the female reproductive system.

In this condition the external genitalia development is normal and often the uterus is absent. In 9% of the cases, vaginal agenesis can occur in the presence of the uterus (3)

The failure in the development of the terminal portion of the paramesonephric ducts causes vaginal agenesis (4)

OBJECTIVE

To report a spontaneous pregnancy after surgical management of Mayer Rokitansky Kauser Hauser syndrome.

CASE REPORT

A 28 year old female married for 7 months presented in the outpatient clinic with history of primary amenorrhoea and severe cyclical dysmenorrhoea. She had well developed secondary sexual characteristics but there was blind ending vagina.

CASE REPORT

Ultrasound scan and MRI showed cervical atresia and non- developed upper 1/3rd of the vagina. Uterus and ovaries appeared normal.

A decision to perform vaginocervicoplasty was made after detailed discussion with the patient.

Intraoperatively on vaginal examination a blind ended vagina and a small fibrotic cervical nodule was felt. Pfannenstiel incision was given and Uterus mobilised. A tract was created from the cervix to the vagina with the help of a probe. Uterus was pushed down and cervix stitched with the vaginal wall by interrupted stitches to ensure continuity of the tract.

A Nelton tube was passed from the cervix to vagina and secured in the vagina by means of a stitch. The tube was to be retained for 4-6 weeks.

Patient was commenced on Norethisterone tablets for 4 weeks.

At 6 weeks follow up appointment the Nelton tube was removed in theatre.

The cervix was open and a good vaginal length was felt on examination.

Patient presented with spontaneous conception at 20 months post surgery.

CONCLUSION

In this particular case timely surgical intervention lead to a favourable outcome.

Very limited cases of a successful pregnancy outcome are reported in literature therefore more research is needed.

REFERENCES

- [1] J. E. Dietrich, D. M. Millar, et E. H. Quint, « Obstructive reproductive tract anomalies », J. Pediatr. Adolesc. Gynecol., vol. 27, n° 6, p. 396–402, déc. 2014, doi:<https://doi.org/10.1016/j.jpog.2014.09.001>.
- [2] D. Wilson, B. Bordoni, « Embryology, Mullerian Ducts (Paramesonephric Ducts) », in *StatPearls*, Treasure Island (FL): StatPearls Publishing, 2024. Consulté le: 24 février 2024. [En ligne]. Disponible sur: <http://www.ncbi.nlm.nih.gov/books/NBK557727/>.
- [3] C. Sohrabi, G. Mathew, N. Maria, A. Kerwan, T. Franchi, et R. A. Agha, « The SCARE 2023 guideline: updating consensus surgical Case Report (SCARE) guidelines », Int. J. Surg. Lond. Engl., vol. 109, n° 5, p. 1136–1140, avr. 2023