

**Mayer - Rokitansky- Küster – Hauser
syndrome : Some successful vaginal
creation cases using soft mold
method.**



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1. Introduction

- **Mayer- Rokitansky- Küster- Hauser (MRKH) syndrome was first described in 1829**
- **MRKH is the congenital absence of the vagina, uterus and cervix.**
- **Rarely seen in women.**
- **Usually diagnosis in adult patients with amenorrhea or intercourse inability.**

2. Pathophysiology

- The syndrome present with the Müllerian duct agenesis at the 5th week of pregnancy.
- The uterus, cervix, and 2/3 upper of the vagina are merged and failure to develop together with Müllerian duct → the uterus and the vagina are absent.
- Ovarian function is preserved because the ovaries originated from the ectoderm layer.

3. Symptom

- Amenorrhea but breasts, public hair and external genitalia (labia majora, labia minora, vestibule...) are normal
- Infertility
- Intercourse inability or pain
- 46, XX karyotype
- Normal FSH, LH, testosterone level.
- ultrasound: uterus absent, normal ovaries.

4. Treatment

Vaginal
creation

- wifehood

Infertility

- Mother-
hood

Vagina creation

- ❖ Many procedure are employed in the world
- ✓ Abbe (1898 – vaginoplasty - skin graft)
- ✓ McIndoe và Banister (1930 – vaginoplasty - skin graft)
- ✓ Wee và Joseph (1989 – pudendal-thigh flaps - Singapore)
- ✓ Lansac (vagina creation, hard mold)...
- ❖ Most of the procedures are complicated, expensive and inappropriate to use in VietNam.

Infertility treatment

Before

- **Adoption**

Now

- **Gestational
surrogacy**

National Hospital of Obstetrics and Gynecology

- ❖ 2002: Lansac procedure was first applied in our hospital.**
- ❖ This vagina creation procedure gave the patient the opportunity to become a real wife and a mother by surrogacy.**

Lansac modified procedure



Diagnostic laparoscopy

Vagina creation

Vaginal soft mold

Step 1: Diagnostic laparoscopy

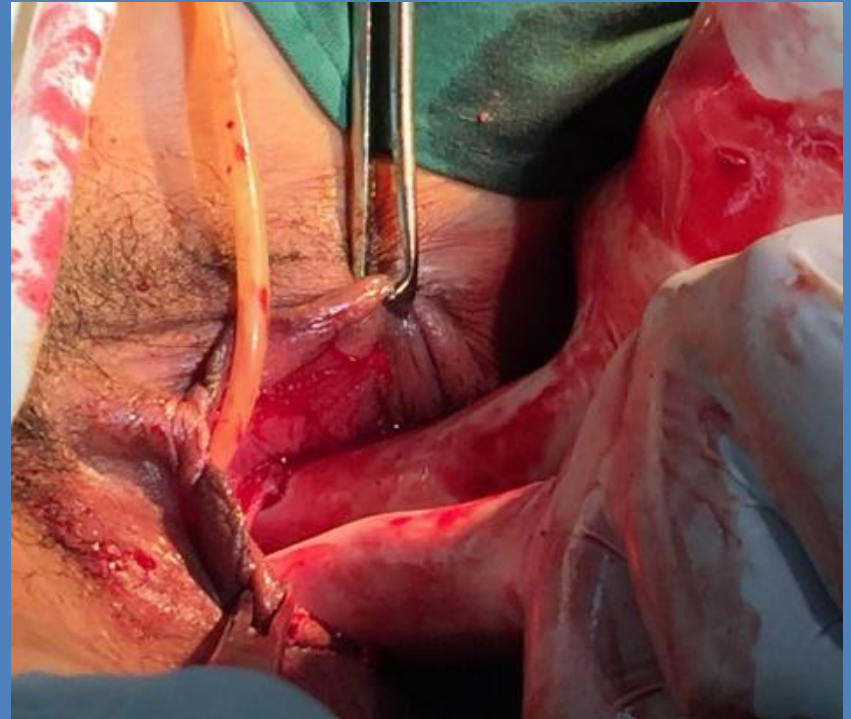
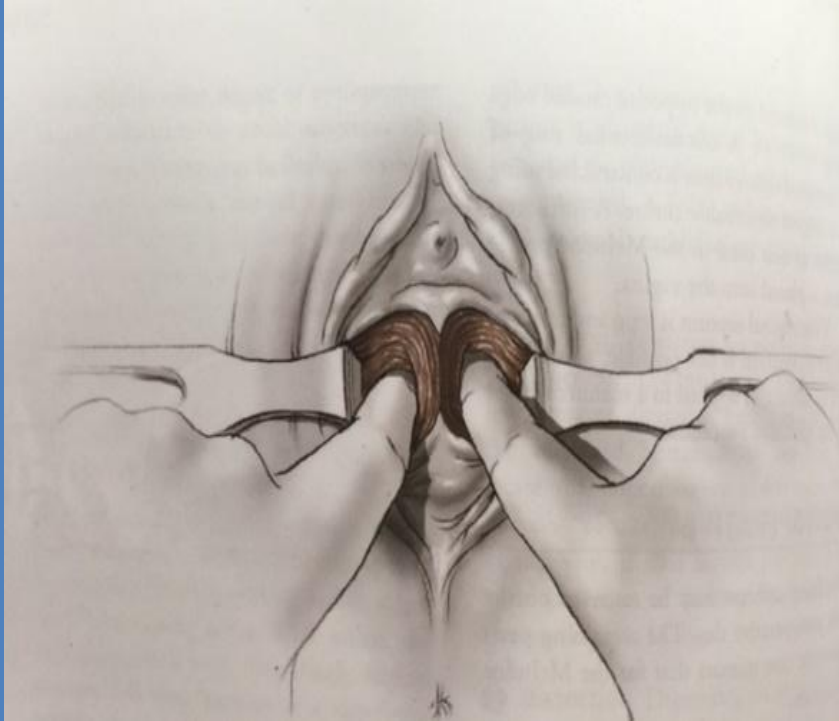


Small rudimentary uterine bulbs are presented with normal fallopian tubes and two normal ovaries.

Step 2: Vagina creation

- ❖ **Transverse vaginal incision , 2-2,5 cm**
- ❖ **Use blunt-tipped scissors to dissect the connective tissue between the urethra and bladder anterior and the rectum posterior, under laparoscopy guidance. The dissection goes to the peritoneum.**
- ❖ **The canals are formed and spread gently by using the scissors. Index fingers are then insinuated into the forming tunnels, and pressure is exerted laterally to extend the canals.**

Step 2: Vagina creation



Step 3: Vaginal soft mold

- ❖ **Initially, rigid dilator (wood mold) was applied but during postoperative care, the patients suffered from pain and the mold was easily loose.**
- Low success rate.**

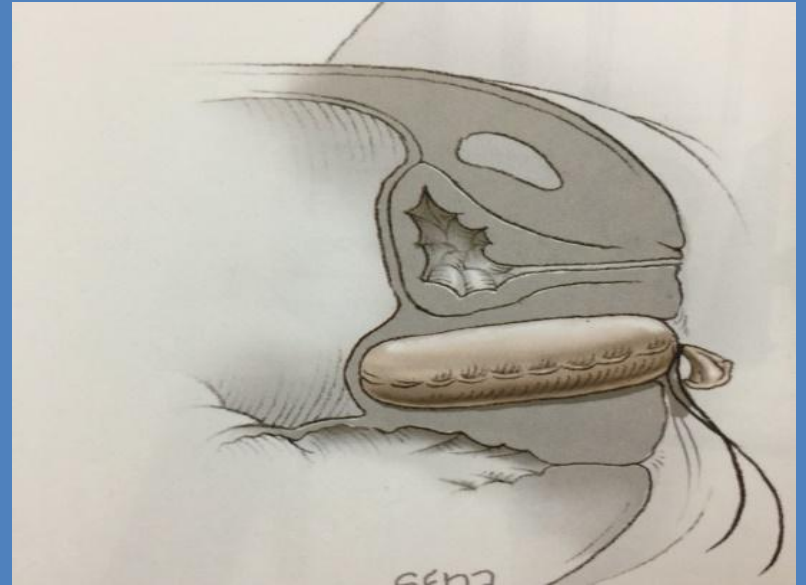
Step 3: Vaginal soft mold

- ❖ **Improvement:**
 - ❖ A mold was created by using a cylindrical medical gauze wrapped by a condom.
 - ❖ A mold inserted and held in the neovagina by stitching two labia majora
- ❖ **Advantages:**
 - ❖ Hemostasis
 - ❖ Adherence reduction
 - ❖ Cheap.

Step 3: Vaginal soft mold



Step 3: Vaginal soft mold



Postoperative

- ❖ **The new mold are replaced after 2 days following surgery.**
- ❖ **Mold removal after 4 days.**
- ❖ **Postoperative dilatations everyday**
- ❖ **Patients are instructed to wear the dilator after discharged.**

Postoperative

- ❖ **Check – up after 2 and 4 weeks.**
- ❖ **For the 6 weeks following surgery, patient wears the dilator 2-3 times/day**
- ❖ **After the initial month, either wear the dilator or engage in intercourse.**

Clinical cases

Patient characteristics

- ❖ **2014 – 2016, we performed 20 cases using modified LANSAC procedure.**

Patient characteristics

- ❖ **Average age: 25.2 Oldest: 39 Youngest: 19**
- ❖ **Diagnosis time: adolescent amenorrhea**
- ❖ **Presenting complaint: sex intercourse inability (17/20 women are going to be married, 3 married women)**

Patient characteristics

Average operation time: 23,5 minutes.

Average length of stay: 7,2 days.

	N	Vaginal lenght	
Intraoperative	20	10,7	2,2cm
Preoperative	20	10,3	1,8cm
2 weeks following discharged	16	9,7	1,35cm
4 weeks following discharged	12	9,8	1,4cm

One clinical case

Patient History

- ❖ **Name: Hoang Ngọc H** **YB: 1977**
- ❖ **Occupation: worker** **Hometown: Ha Tinh**
- ❖ **3 sister in this family had MRKH syndrome, patient is the oldest.**
- ❖ **Her second and third sister was successful operated with the LANSAC procedure in 2/2014 and 4/2016**

Past medical history

- ❖ Patient has been married for 13 years.
- ❖ 2009: “Pudendal- thigh flaps Singapore” procedure was performed at Tu Du Hospital
- ❖ The case was unsuccessful.

Clinical examination

- ❖ Height: 150cm, Weight: 45kg
- ❖ Normal breast and pubic hair
- ❖ External genitalia:
- ❖ Short vagina: 2.5 cm → intercourse inability.
- ❖ Fundament examination: uterus undefined

Clinical examination

Sub - clinical

- ❖ **Karyotype: 46XX**
- ❖ **Female sex hormones, Thyroid function: normal**
- ❖ **Ultrasound: no uterus, 2 normal ovaries**
- ❖ ***Diagnosis:***
 - ❖ **MRKH syndrome**

Treatment

- ❖ **Old skin flap cut out**
- ❖ **Vagina creation: Modified LANSAC procedure**
- ❖ **Operation time: 30 minutes**
- ❖ **Vagina length: 11.5 cm.**

Treatment



Postoperative

- ❖ **Replace new mold after 3 days.**
- ❖ **Stitch two labia majora to hold the mold inside the vagina.**
- ❖ **Remove the mold after 5 days.**
- ❖ **Day 5,6,7,8: Patient is instructed to wear the dilator.**
- ❖ **Discharged on 22/11/2016.**
Vagina length: 11 cm

Following check up

- ❖ **Instruct patient to use the dilator with betadine ointment 2-3 times/day, 15-30 minutes per time. Soft mold is use for night.**
- ❖ **Check up after 2 weeks and 1 month for vagina length (10.5 cm and 10 cm respectively)**
- ❖ **Result: Patient be able to have sexual intercourse**

VIDEO: Following check up

To: Bn Hoàng Ngọc  Rokitanski

Thu, Dec 22, 7:48 PM

Chị ơi ,hôm nay kỉ niệm 13 năm ngày cưới của bọn em. Lần đầu tiên em đc làm vợ đúng nghĩa chị ạ. Chồng em rất hài lòng chị ạ. Bọn em cảm ơn chị và bác Quyết rất nhiều .

Chúc mừng em nhé!

Dạ vâng chị.

Conclusion

- ❖ **These procedure create an opportunity for the patient with MRKH syndrome to have a normal sexual life and become a mother by surrogacy.**
- ❖ **The modified LANSAC procedure, with low cost, short operation time, uncomplicated instruction for training doctors, is an affordable method to apply in Vietnam.**