

2017년 제 18차 대한산부인과내분비학회 학술대회 및 연수강좌
종례위주 환자접근: 미성년



초경이 시작되지 않은 15세 여성

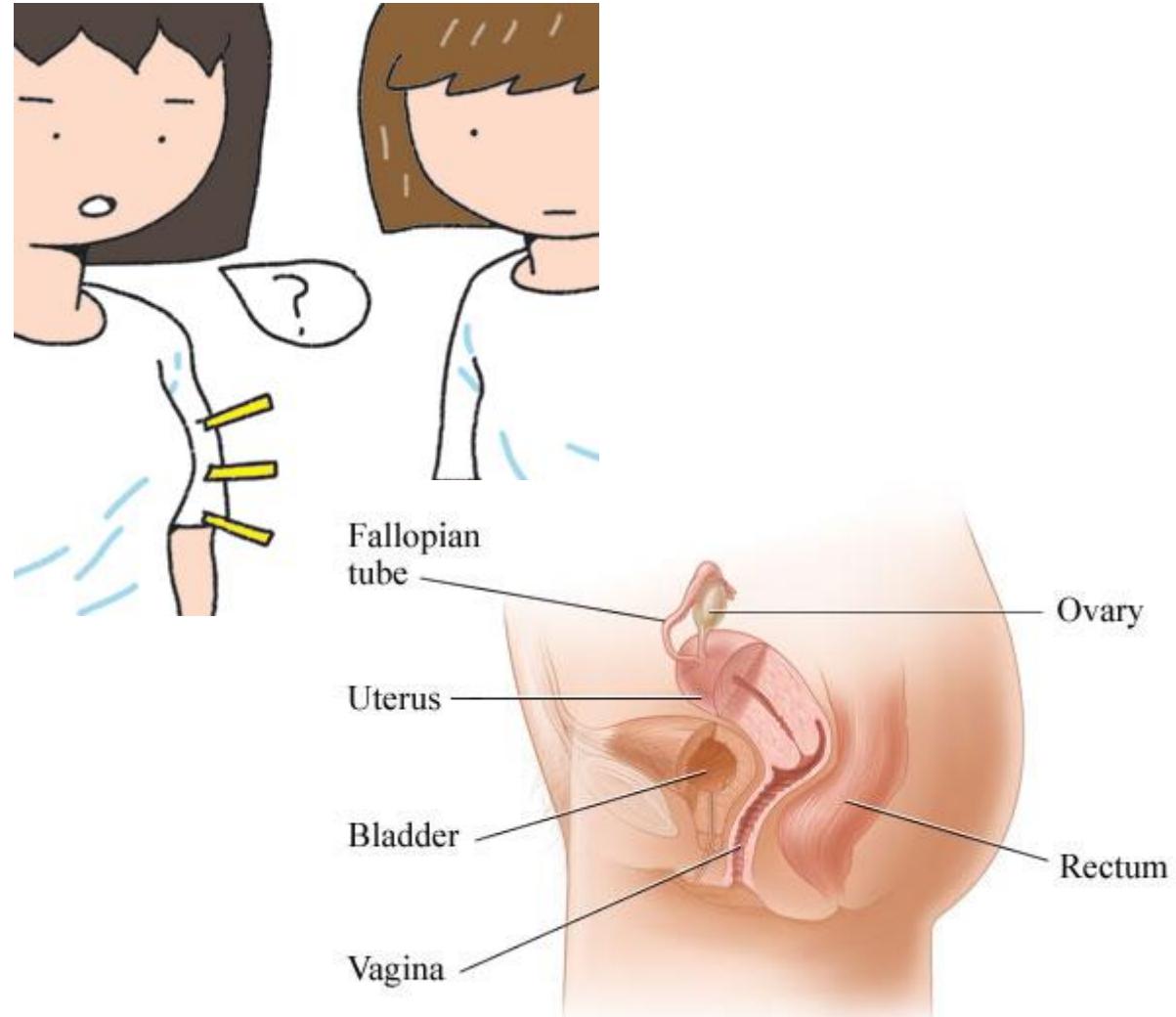
아주대학교 의과대학 산부인과 김미란

DEFINITION



- Primary amenorrhea
- Absence of menses by 13 years of age when there is no visible secondary sexual characteristic development
- By 15 years of age in the presence of normal secondary sexual characteristics

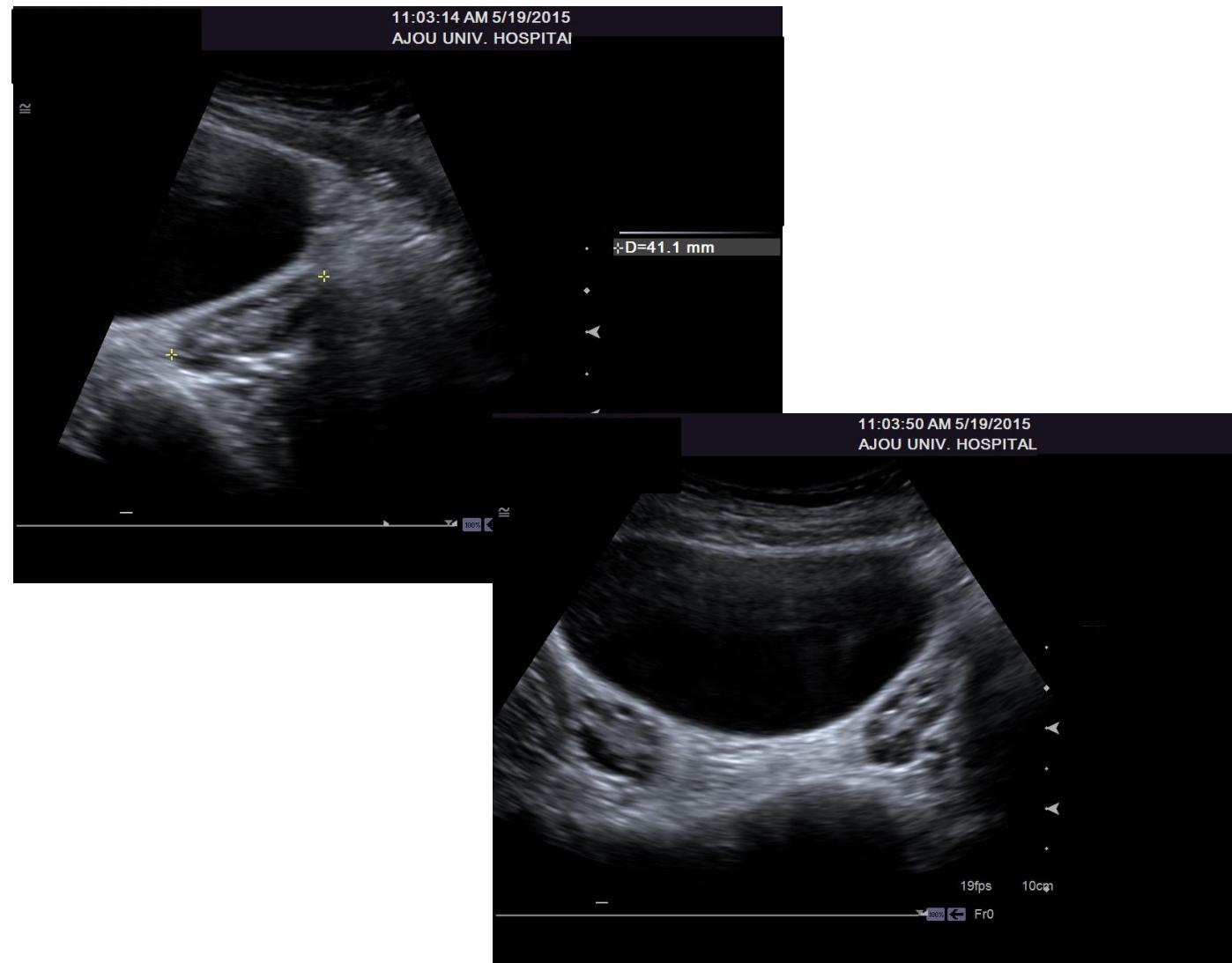
- Amenorrhea without Secondary Sexual Characteristics
- Amenorrhea with Secondary Sexual Characteristics and abnormalities of pelvic anatomy
- Amenorrhea with Secondary Sexual Characteristics and normal pelvic anatomy



Case

- 2016년 15세 일차성 무월경으로 본원 부인과 방문
- 유방발달 8세 시작, Tanner stage breast III, pubic hair I
- 성조숙증 진단, 10세부터 1년 GnRH agonist 주사치료
- 156cm/60kg (BMI 24.65)
- Waist/HIP: 88/100 (0.88)
- Tanner stage breast IV, pubic hair IV

항목	결과
LH (mIU/ml)	7.1
FSH (mIU/ml)	4.8
E2 (pg/ml)	87
AMH (ng/mL)	10.71
Insulin (uIU/ml)	18.0 (11-11.6)
Glucose (mg/dL)	103
T.Cholesterol (mg/dL)	231 (118-200)
ALT (GPT, U/L)	70 (5-33)
AST (GOT, U/L)	36 (5-32)



- Primary amenorrhea
- More likely hypothalamic amenorrhea related with multifollicular ovary, rather than PCOS
- Weight control and Observation

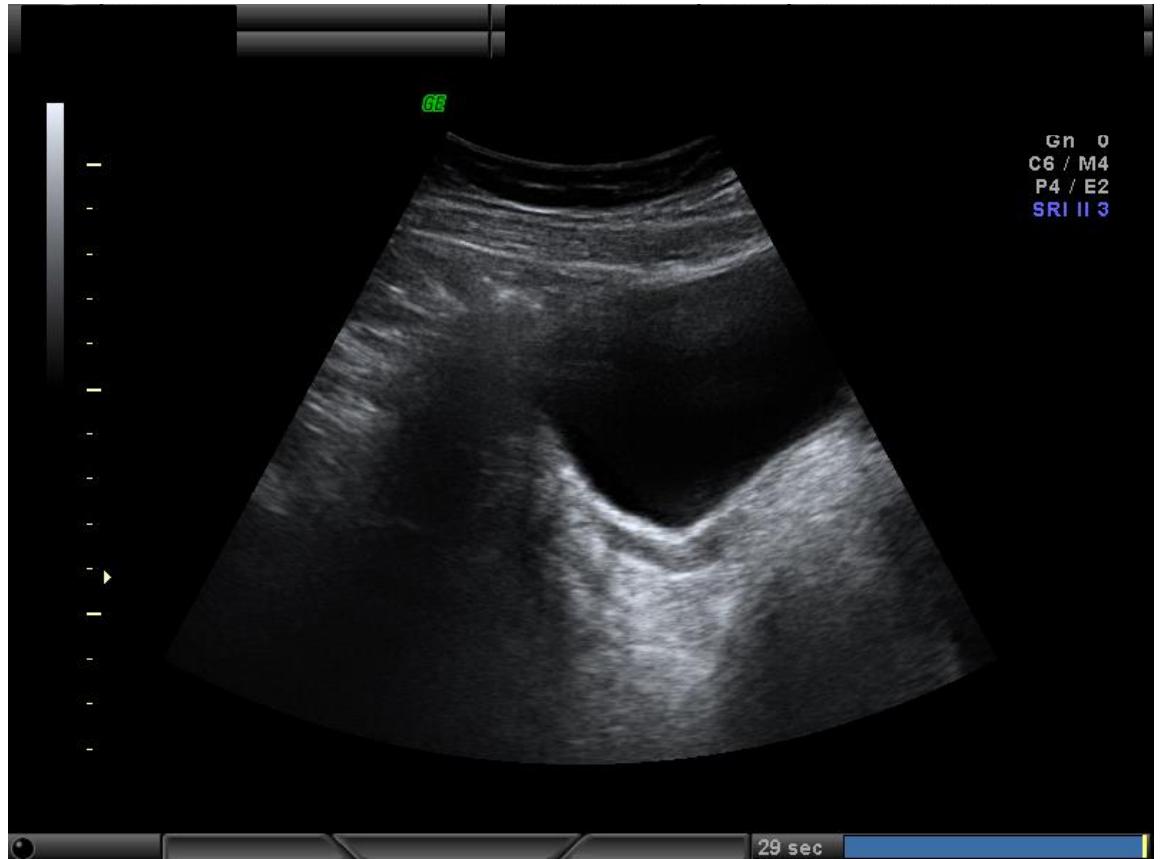
Case – Hypothalamic Hypogonadism

- 2011년 2월 15세 일차성무월경으로 내원
- Tanner stage Breast I, Pubic hair II
- GnRH stimulation test (2010년, 소아청소년과)

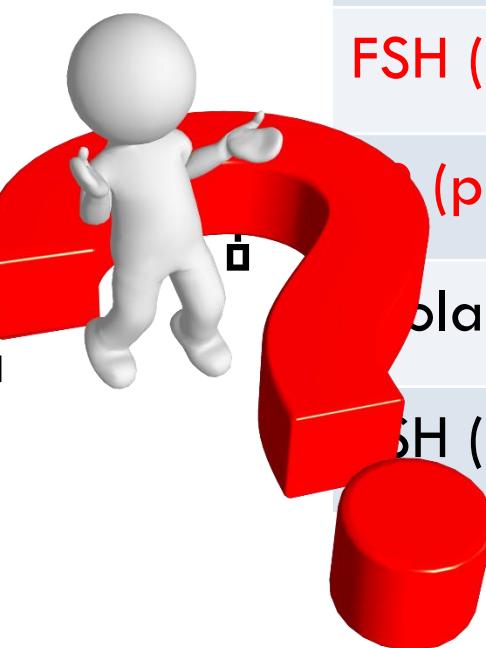
	0	30 min	45 min	60 min	90 min
LH (mIU/ml)	1.9	1.7	1.8	1.8	2.3
FSH (mIU/ml)	0.8	1.1	1.8	1.9	2.4
E2 (pg/ml)	10		13		11

- Hypothalamic Hypogonadism
- 부인과 검사 얻함

- No anosmia
- Female pelvic U/S, MRI
 - Most likely uterine hypoplasia
 - 3.4x1cm sized small uterus
 - 2.4cm, 2cm ovaries
- Brain Sella MRI
 - Normal



- 2012년 1월, 16세부터 여성화로봇 투여 시작
- 2012년 3월 초경 시작
- Progesterone 추가 투여
- 이후 자가 중단
- 2012년 8월(17세) 재내원, 이전에 투여한 항생제 복용
- 2013년 12월 타병원 진료 완료



항목	2012년 8월
LH (mIU/ml)	0.2
FSH (mIU/ml)	0.2
(pg/ml)	5이하
progesterone (ng/mL)	7.1
SH (μ IU/ml)	2.43

Case - Chromosome abnormality

- 2013년 10월 15세 일차성무임경
- 일차병원에서 자궁과 난소가 보이지 않는다고 듣고 어머니만 내원
- 160cm/65kg (BMI 25.4)
- Pelvic MRI
 - Rudimentary uterus and streak gonad.
 - More likely gonadal dysgenesis. DDx. Adrogen insensitivity syndrome

- Vagina 4cm °이상 advanced
- Tanner stage Breast III, Pubic hair II
- Chromosome study
 - ▣ 45,X[15]/46,X,add(X)(q26)[5]
 - ▣ 20 metaphase cells revealed a mosaicism of a monosomy for X in 15cells and additional material of unknown origin replaces the segment Xq26qter in 5cells.

항목	결과
LH (mIU/ml)	12.2
FSH (mIU/ml)	31.0
E2 (pg/ml)	14
Prolactin (ng/mL)	5.2
TSH (μ IU/ml)	4.52
Progesterone (ng/ml)	0.6
Testosterone (ng/ml)	0.36

유전학 클리닉 진료시작

- 터너증후군의 아형, Mosaic Turner SD
- Y 염색체 존재유무 확인 위해 SRY 검사 : negative
- 2013년 11월 여성호르몬 치료 시작
- 초경 후 호르몬치료 유지 중

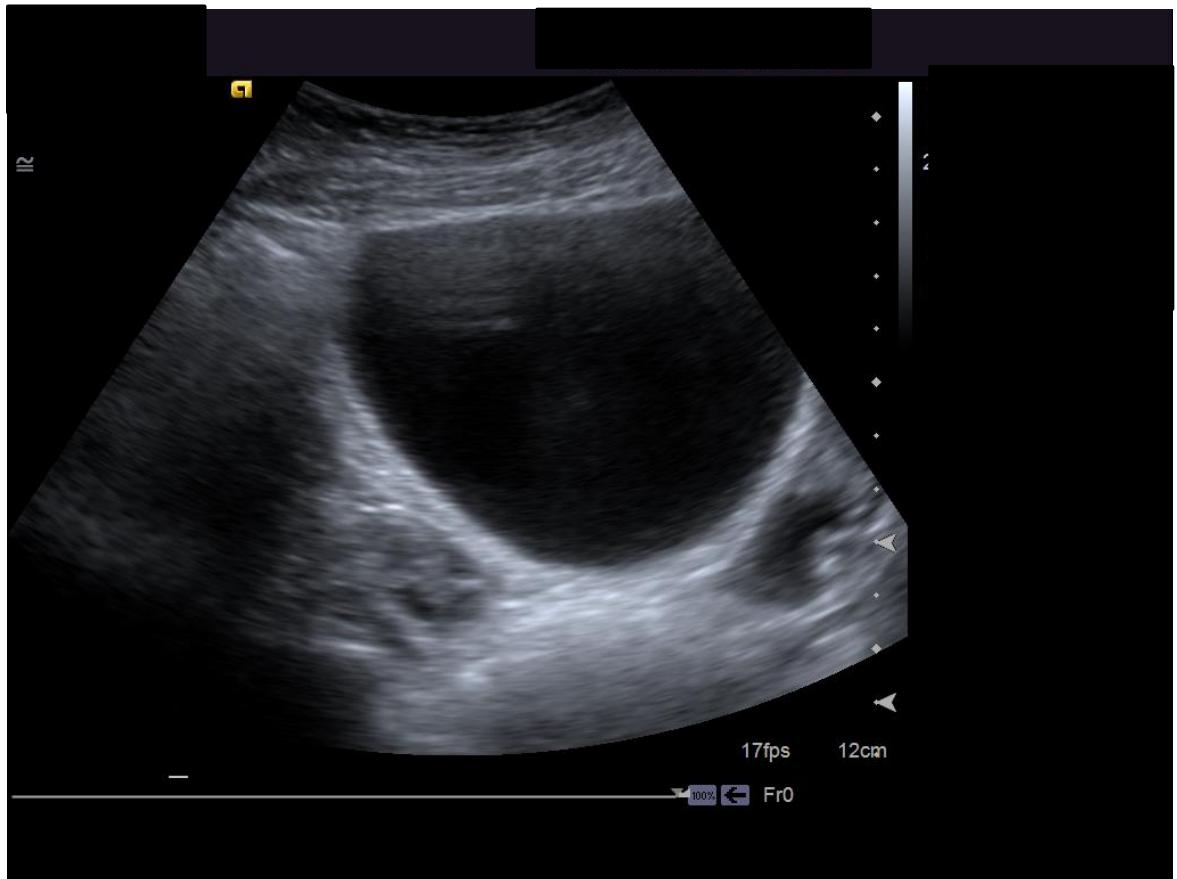
항목	2016년 12월
LH (mIU/ml)	1.5
FSH (mIU/ml)	6.9
E2 (pg/ml)	212
TSH (μ IU/ml)	0.31

CASE - Anomaly

- 2014년 3월 일차성무월경으로 내원
- 157cm/51.7kg (BMI 21)
- Vagina blind pouch
- Chromosome study
 - 46,XX

항목	결과
LH (mIU/ml)	8.44
FSH (mIU/ml)	5.2
E2 (pg/ml)	7
Prolactin (ng/mL)	5.7
TSH (μ IU/ml)	3.22
AMH (ng/mL)	4.59

- Pelvic US
 - R/O agenesis or hypoplasia of uterus
- 20% ↑ Vaginoplasty



Vaginoplasty – Modified Laparoscopic Davydov Operation

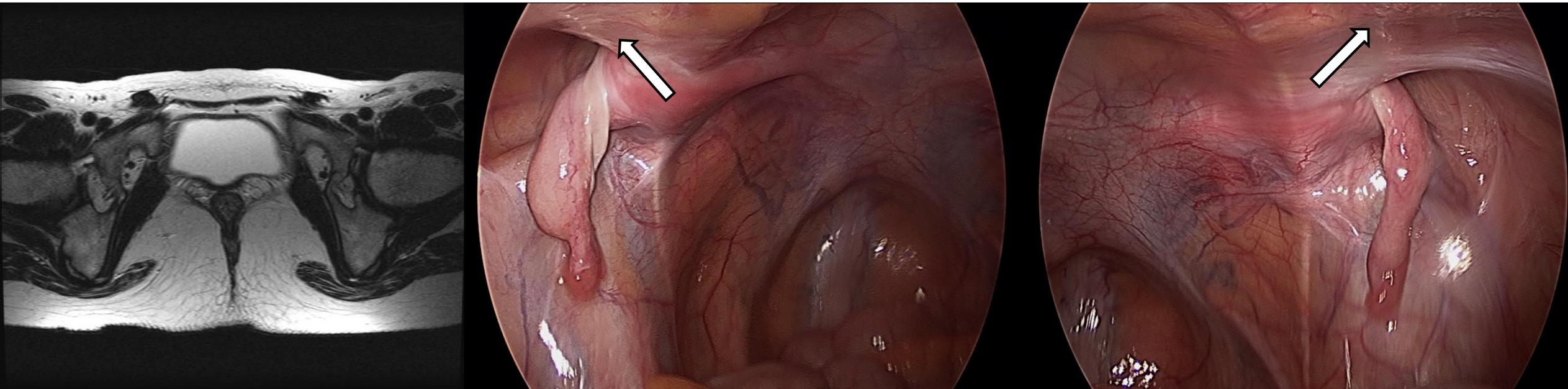


Figure. Magnetic resonance imaging and laparoscopic finding with bilateral inguinal ovaries (arrows)

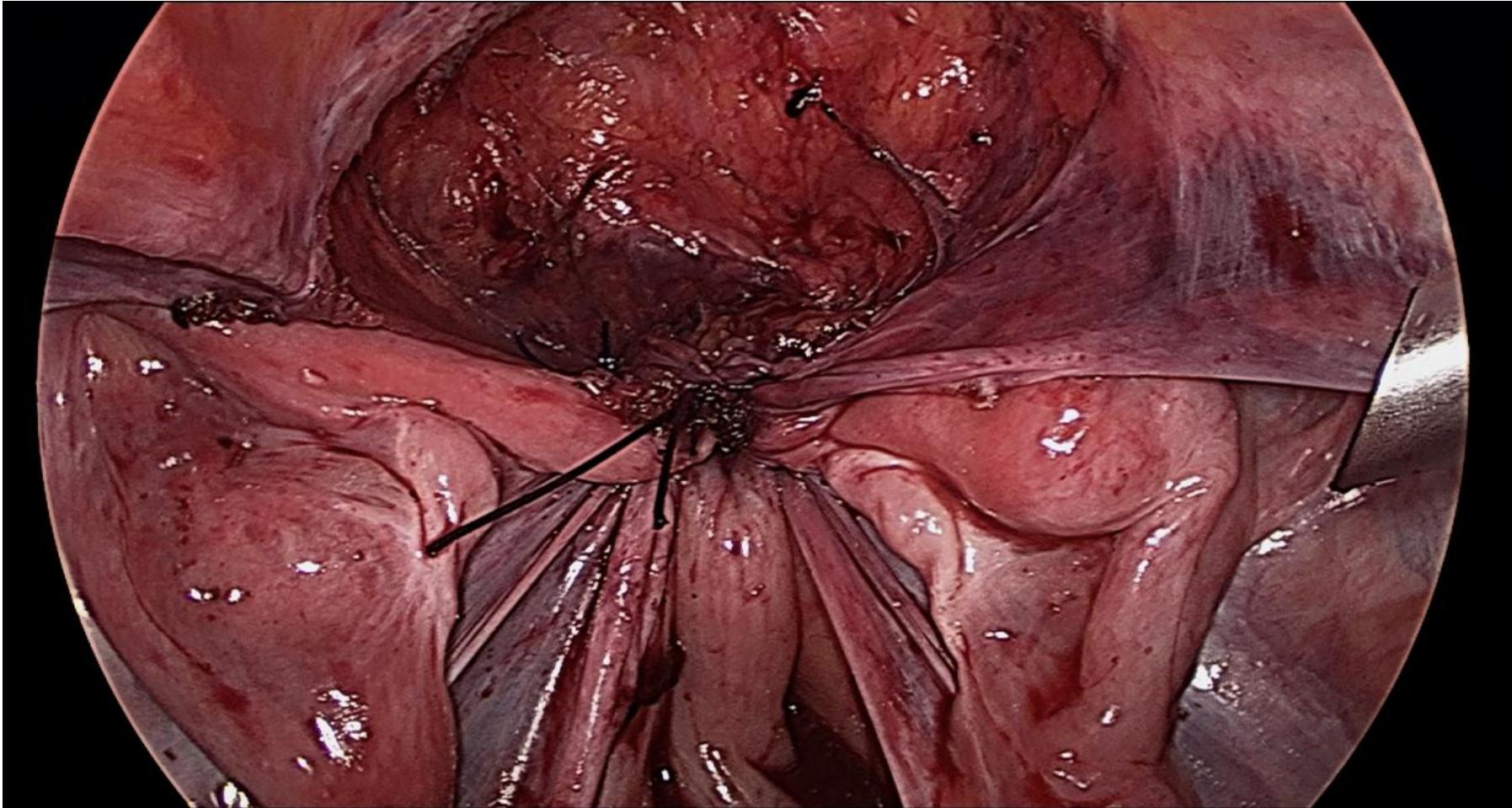


Figure. After inguinal ovaries reposition, vaginal vault was made through middle purse-string suture to prevent vaginal prolapse and maintain of vaginal sufficient length.

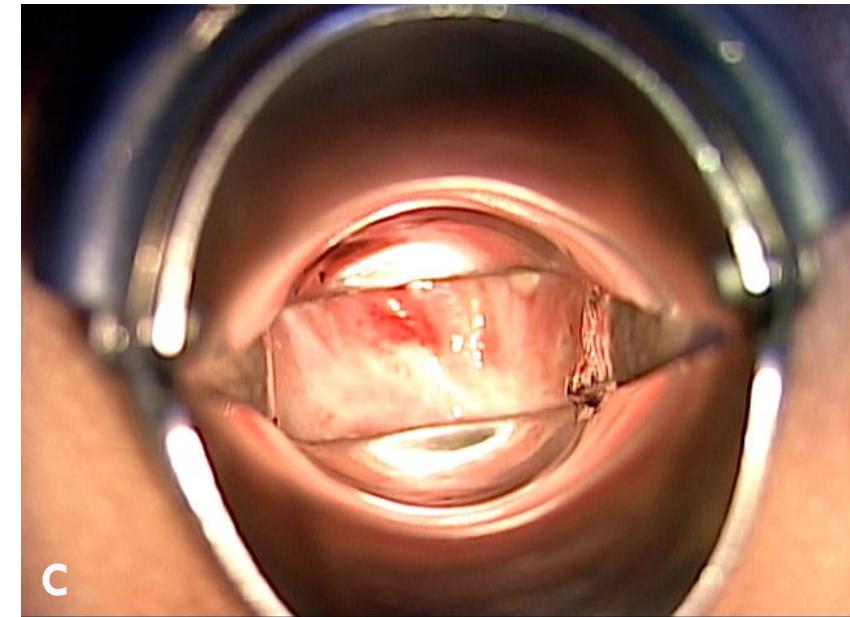
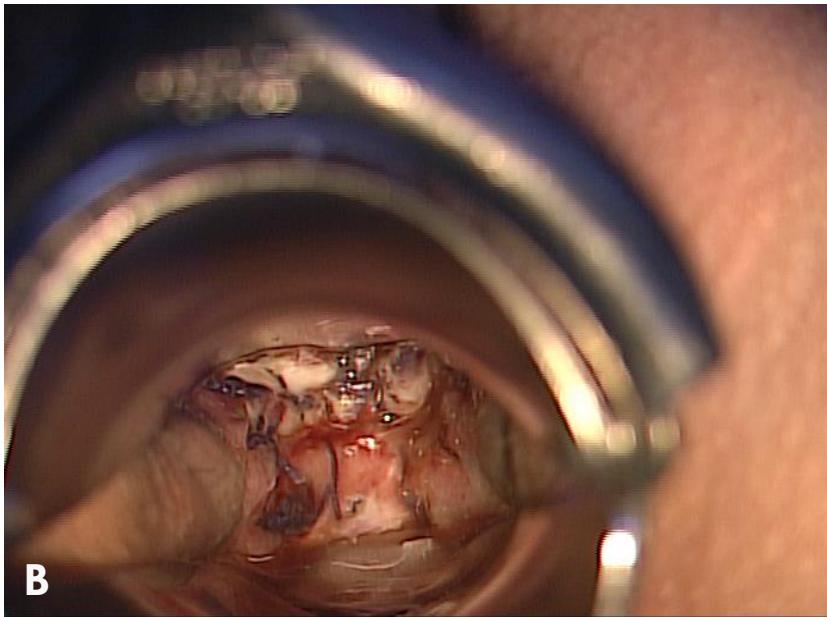
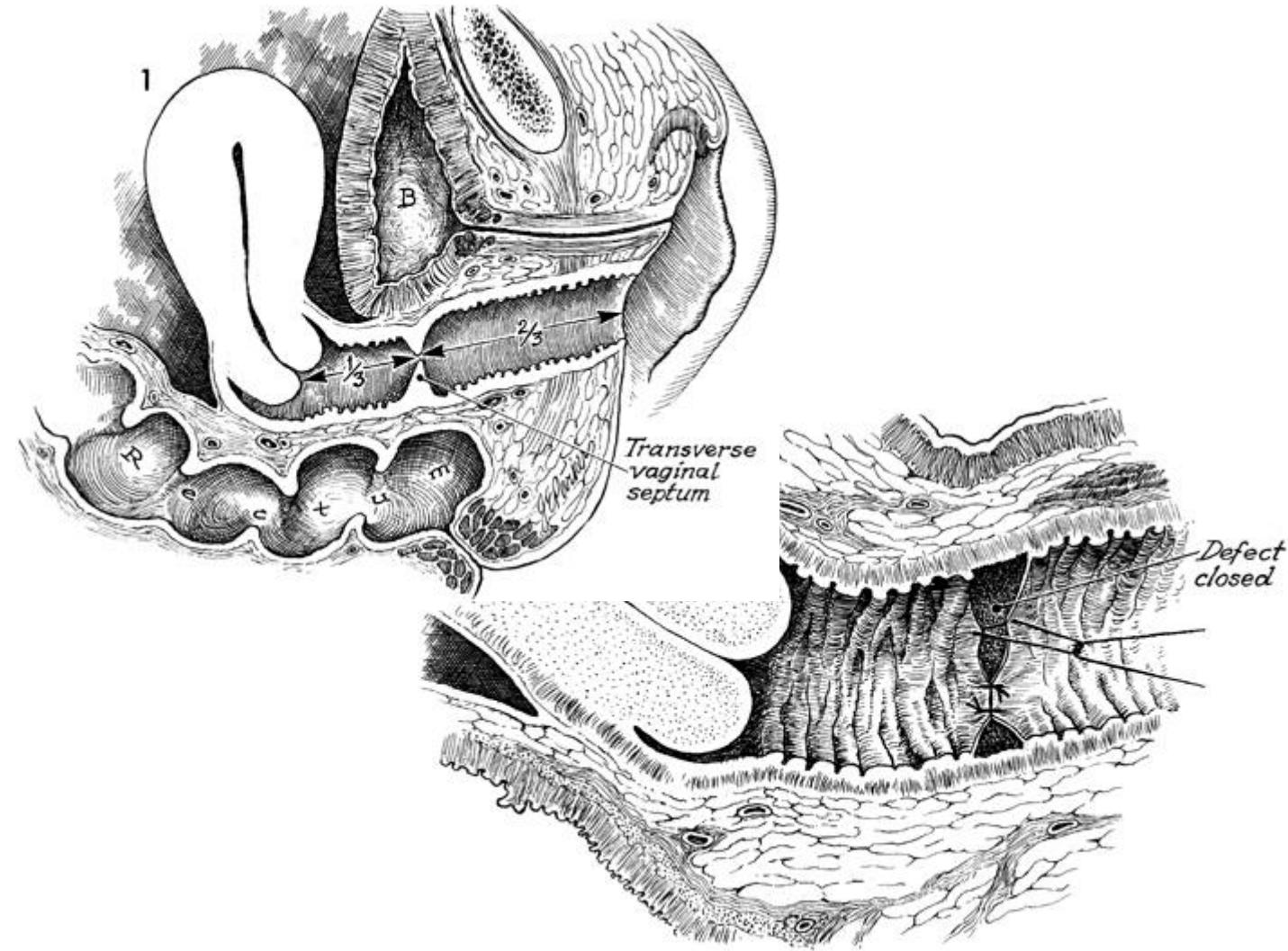
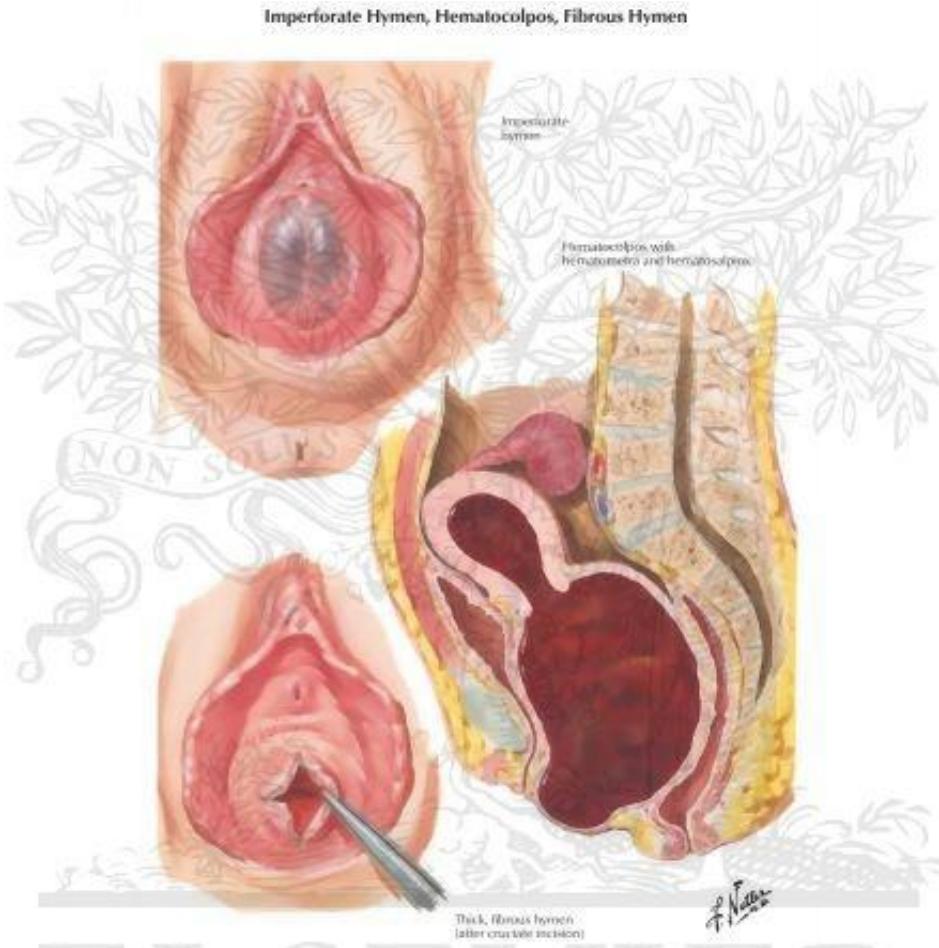


Figure. Post operative appearance of the newly created vaginal opening (A), 15 days after surgery, the edge of pelvic peritoneum (B), and vaginal vault after surgery at 12months (C).

Imperforate hymen vs. Transverse vaginal septum

일차성무월경



CASE- Anomaly

- 2014년 9월 아랫배통으로 병원 응급실 방문.
아파서 잠을 못 잔다. 11세
- 유판상 imperforate hymen ↑↑ ↑↑
- 이차성진 breast III, Pubic II
- Pelvic MRI
 - ▣ More likely vaginal atresia or septum, differential diagnosis include imperforate hymen.
 - ▣ Hematocolpos, hematometra, and endometriotic cyst on Lt adnexa.



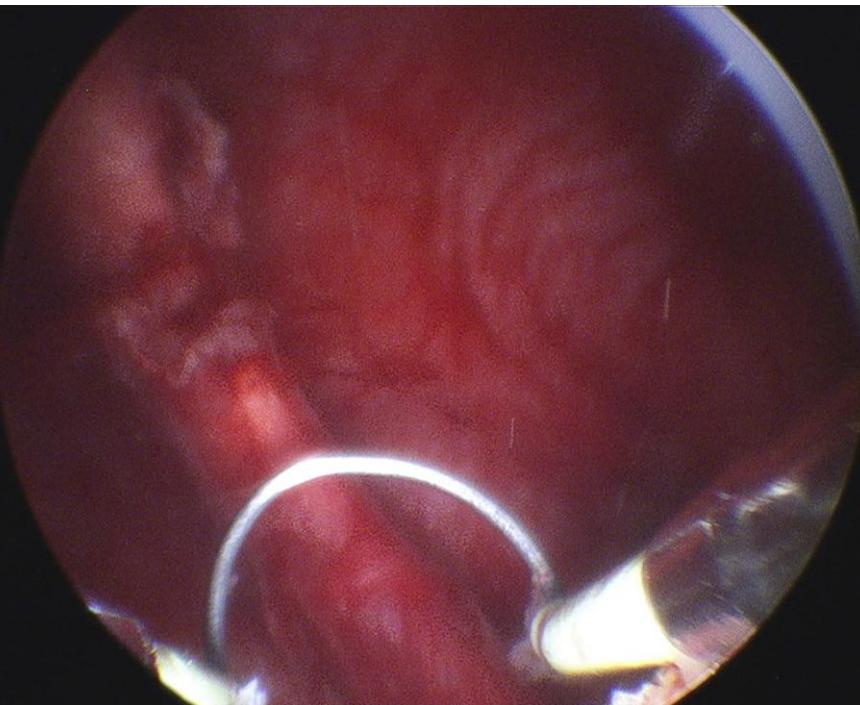
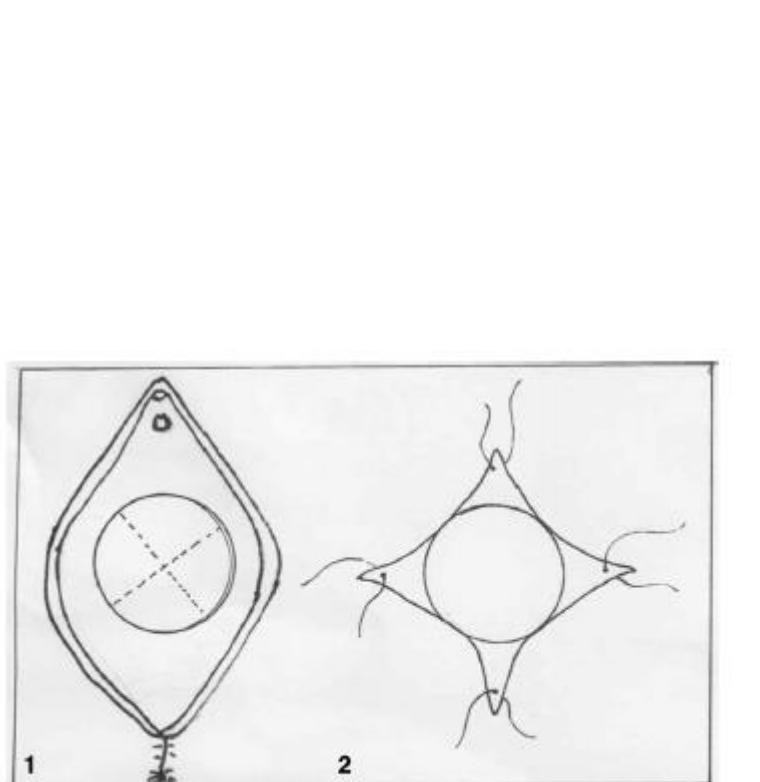
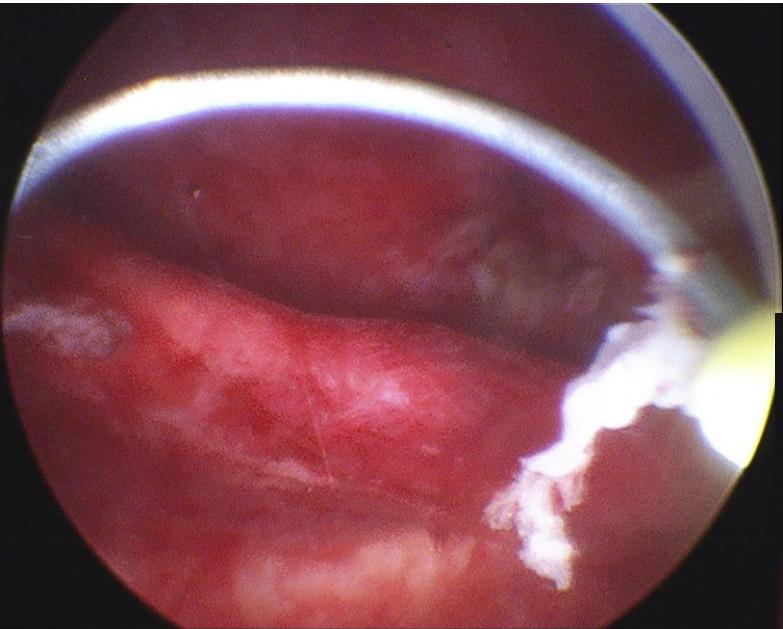
- 2014년 9월
 - ▣ Hymen ring 1.5cm 상방 transvaginal septum 밑부
 - ▣ Spetum 제거
- 2014년 11월 pelvic MRI
 - ▣ Hemorrhagic fluid collection in intrauterine cavity has been improved compared with previous study.
 - ▣ Linear structure below the cervical os is noted. R/O residual transvaginal septum.
 - ▣ R/O possibility of residual transvaginal septum.

□ 2014년 11월

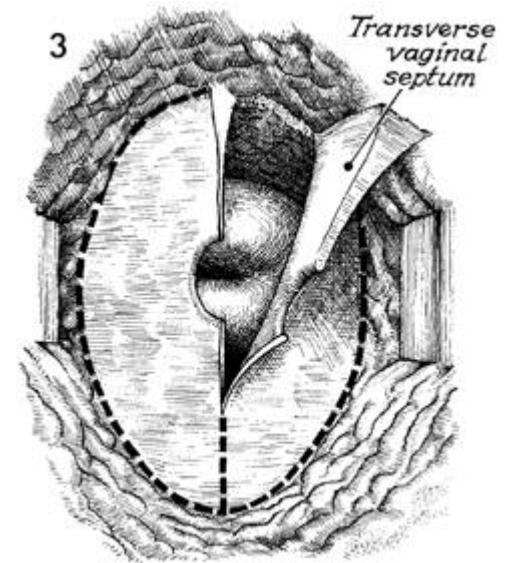
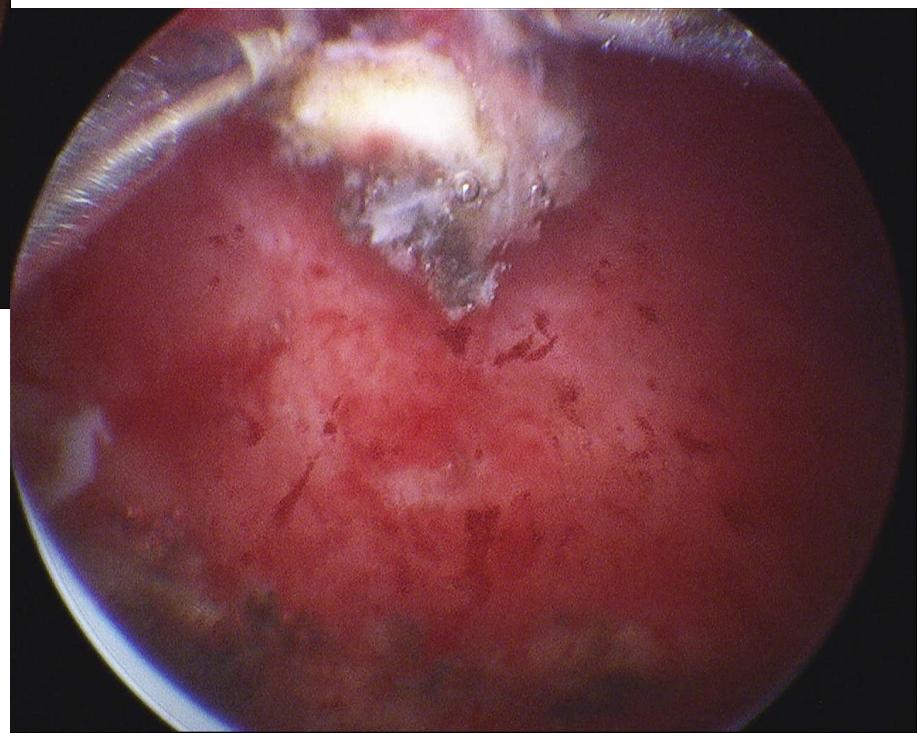


□ 2015년 4월 pelvic U
□ Hematocolpos 6cm

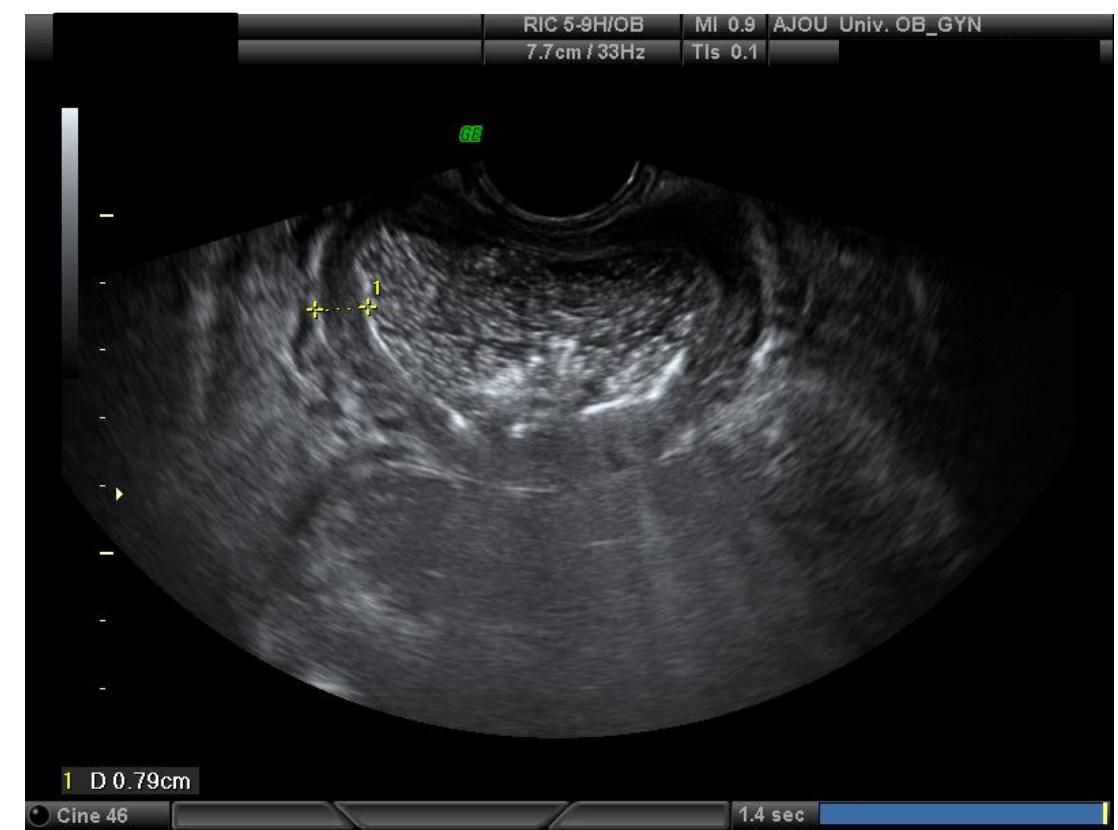




2015년 5월 2차 촬영



- ↑↑ ↑ vaginal mold ↑↑ ↑
- 그러나 ...
- 전위



Amenorrhea with secondary sexual characteristics



- ***Abnormal physical examination***
- ***Hypergonadotropic hypogonadism***
- ***Hypogonadotropic hypogonadism***

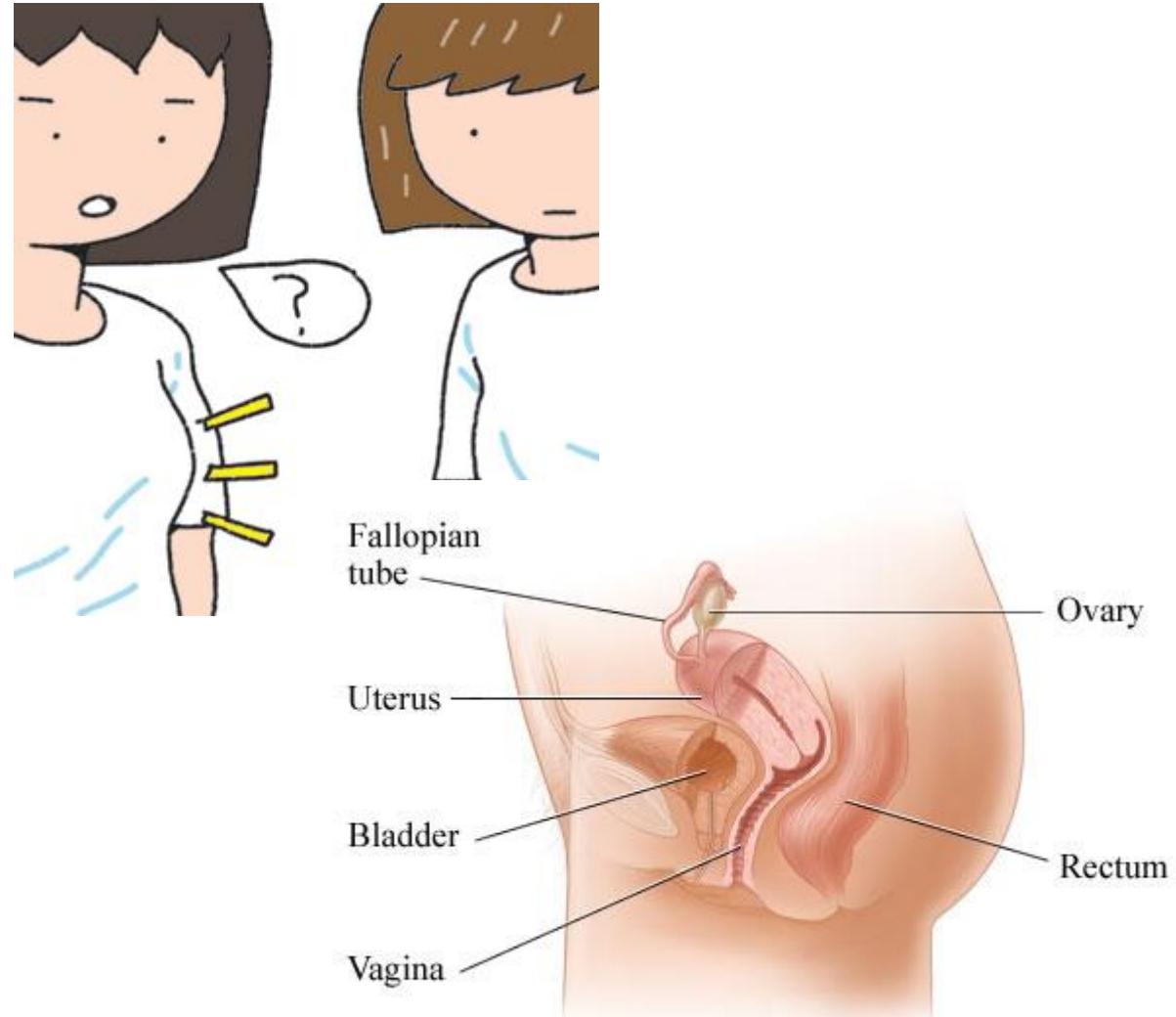
Amenorrhea without secondary sexual characteristics and abnormalities of pelvic anatomy

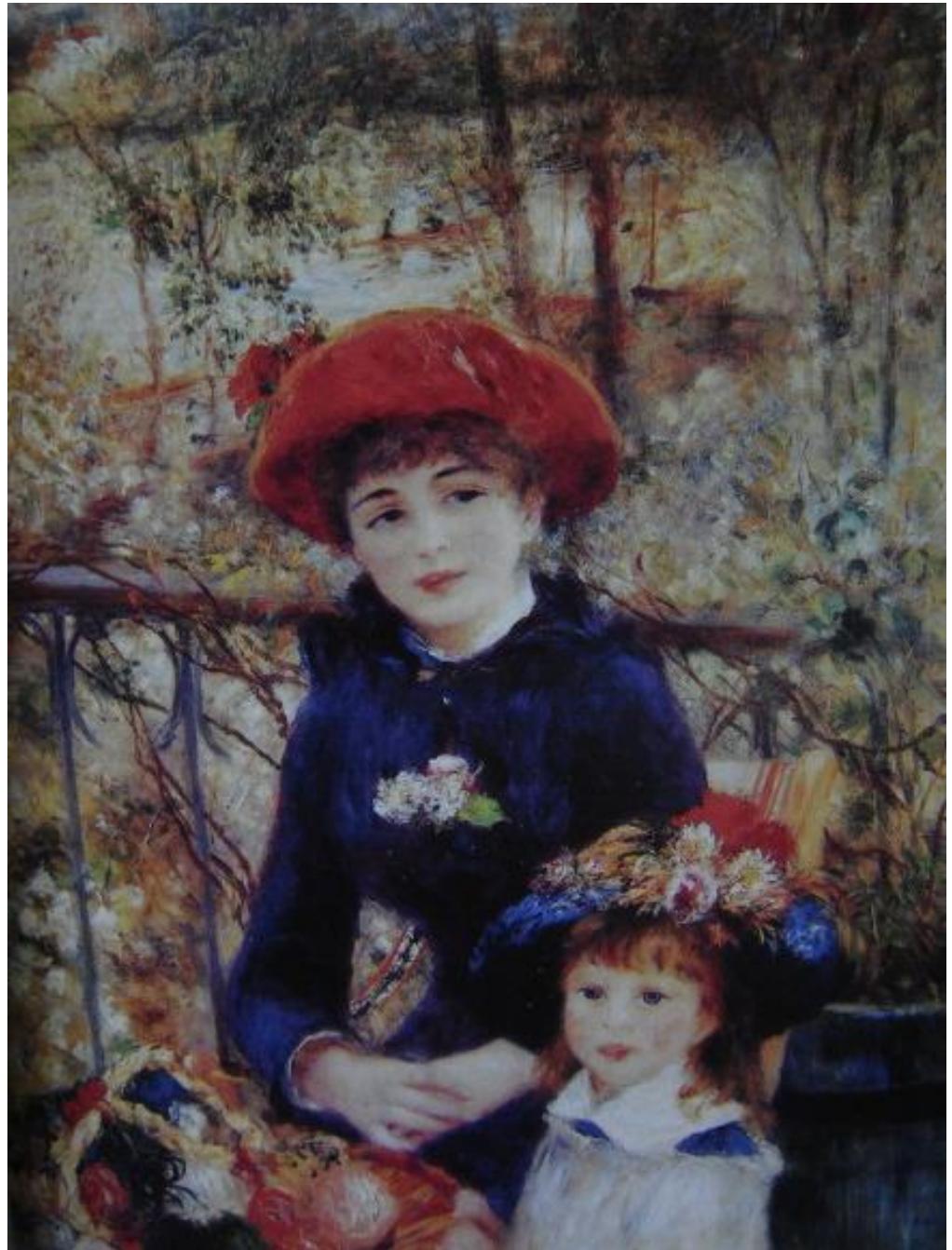
- **Outflow and Mullerian anomalies**
- Androgen Insensitivity
- True Hermaphroditism

Amenorrhea without secondary sexual characteristics and normal pelvic anatomy

- **Polycystic ovarian syndrome**
- Hyperprolactinemia
- **Primary ovarian insufficiency
(Premature ovarian Failure)**
- Pituitary/Hypothalamic Lesions
- Altered Hypothalamic
Gonadotropin-releasing
Hormone Secretion
- Eating disorder
- Weight Loss and Dieting
- Anorexia Nervosa
- Exercise
- Stress
- **Obesity**
- Other Hormonal Factors

- Amenorrhea without Secondary Sexual Characteristics
- Amenorrhea with Secondary Sexual Characteristics and abnormalities of pelvic anatomy
- Amenorrhea with Secondary Sexual Characteristics and normal pelvic anatomy





감사합니다