AGL 2022

51st GLOBAL CONGRESS ON MIGS

December 1–4, 2022 | Gaylord Rockies Resort and Convention Center | Aurora, Colorado

SYLLABUS

PAGS-618: Optimizing Surgical Management in Today's Pediatric Environment

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The following members of AAGL have been involved in the educational planning and/or review of this course (listed in alphabetical order by last name).

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The following have agreed to provide verbal disclosure of their relationships prior to their presentations. They have also agreed to support their presentations and clinical recommendations with the "best available evidence" from medical literature (in alphabetical order by last name).

Krista Childress, MD*
Kate McKracken, MD*
Nichole A. Tyson, MD
HRA Pharmaceuticals- Consultant
Lissa Yu, MD*

PAGS-618: Optimizing Surgical Management in Today's Pediatric Environment

Co-Chairs: *Nichole A. Tyson, MD, Krista Childress, MD*

Faculty: *Kate McKracken, MD, Lissa Yu, MD*

Course Description

This course provides a comprehensive review on surgical management of common diagnoses that present in the pediatric and adolescent gynecology patient population. The course will address: The diagnosis and management of endometriosis in the adolescent and young adult; The medical and surgical management in the patient who presents with obstructive Mullerian and vaginal anomalies; Optimizing the evaluation and surgical management of young patients with adnexal masses; and an overview on the up-to-date gynecologic care for adolescents and adults with history of complex anorectal and urogenital surgeries.

Learning Objectives

At the conclusion of this course, the participants will be able to: 1) Complete an appropriate surgical evaluation and management for adolescent patients with endometriosis; 2) Employ medical and surgical approaches in the care of the patient with an obstructive Mullerian/Vaginal anomaly; 3) Apply surgical strategies to address the large adnexal mass with a focus on ovarian sparing minimally invasive surgery; and 4) Provide up to date gynecologic care for adolescents and adults with history of complex anorectal and urogenital surgeries.

Course Outline

2:30 pm	Welcome, Introduction and Course Overview	N.A. Tyson/K. Childress
2:35 pm	Endometriosis in Teen and Young Adult	L. Yu
3:05 pm	Management of Adnexal Masses in the Adolescent and Young Adult	N.A. Tyson
3:35 pm	Managing the Patient with Obstructive Mullerian Anomalies	K. Childress
4:05 pm	An Overview of Gynecologic Care for Adolescents and Young Adults with Anorectal Malformations	K. McKracken
4:35 pm	Questions & Answers	All Faculty
5:00 pm	Adjourn	

Endometriosis in Adolescents and Young Adults

Lissa Yu, MD
Assistant Professor, Pediatric and Adolescent Gynecology
University of Washington
Seattle Children's Hospital

Disclosure

- I have no financial relationships to disclose

Objectives

- Describe differences and limitations in the gynecologic care of adolescents compared with adult counterparts
- Complete an appropriate evaluation of a young person with dysmenorrhea
- Identify surgical findings of adolescent endometriosis and offer appropriate treatment

Clinical case

Patient K, age 13

- She has had heavy regular painful periods since menarche 9 months ago.
- She presents with her mom who has endometriosis and reports multiple other family members have painful periods. All her friends in 7th grade have painful periods too, but hers seem to be worse.
- She has tried Ibuprofen and Tylenol as well as heat packs and watching TikTok.

Do you…?

- Start combined hormonal contraceptive pill
- Start progesterone only pill
- Place a progestin IUD
- Book for diagnostic laparoscopy
- Start GnRH agonist or antagonist
- Discuss TLH/BSO

Endometriosis: not just an adult disease

- 9% of autopsied female fetuses (4/52) w endometriosis (Signorile 2012)
- Endometriosis reported prior to and just after menarche (Shim 2019,Marsh 2005, Yamamoto 1997, Goldstein 1979)
- Survey of 4000 adult women with endometriosis: (Ballweg 2003)
  - 2/3 reported their first pelvic symptoms prior 20 years of age
  - 1 in 5 women had pain before 15 years
- Recent systematic review including 19 studies (1243 pts): 648 of 1011(64%) adolescents who underwent laparoscopy had endometriosis (Hirsch 2020)
  - Up to 75% in youth with refractory dysmenorrhea (Janssen 2013, Vercellini 1989)

Gynecologic Evaluation

- Delayed Presentation
  - May present to pediatricians, mental health, and GI teams first
  - Average of 3 doctors prior to GYN assessment
  - Estimates range from 22 months to >9 years to diagnosis (Dun 2015, Ballweg 2004)
  - Care may be in a pediatric setting
- Affected first-degree relative→7-10-fold increased risk (Malinak 1980)
- Consider additional medical history
  - Quantify bleeding, assess for bleeding disorder
  - Other possible associated medical history
    - Auto-immune disease, hypermobility, pain syndromes (Shigesi 2019)
    - Contraindications to medication use

Differences in adolescent medicine

- History
  - Use of menstrual and symptom diaries
  - Family members as collateral source
  - Need for confidential interview
- Physical examination
  - Patients may be unaccustomed to physical exam of abdomen
  - Limited pelvic examination
- Imaging
  - Transvaginal ultrasound may be unacceptable
  - Pelvic ultrasound to rule out obstructive anomalies, other masses
  - MRI may be beneficial (Mertre 2020)

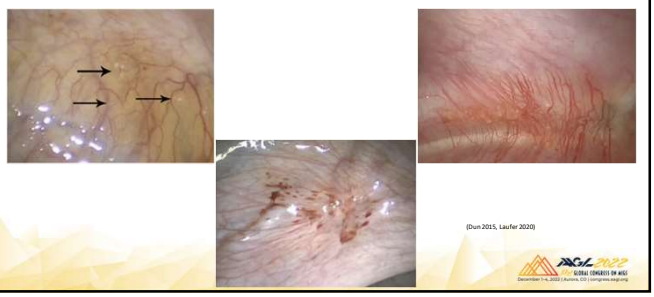
Laparoscopic findings in adolescents

- As with adult women, laparoscopy remains standard of care for definitive diagnosis of endometriosis
- Increased diagnosis of advanced staged endometriosis (Dowlut-McElroy 2017)
- Most commonly, early-stage endometriosis
- Findings may differ slightly from adults: (Dun 2015)
  - Peritoneal defects/windows
  - White, clear, or fibrotic lesions
  - Hemosiderin/pigmented or hemorrhagic lesions
  - Ovarian lesions may be seen but endometriomas less likely

"Earlier stage" does not mean less pain

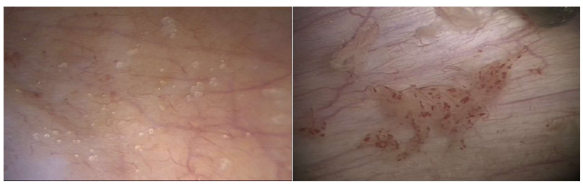
- Surgical staging not correlated with extent of pain symptoms
- Clear and red lesions seen in adolescents are more metabolically active
- Associated with greater prostaglandin production than the "powder burn" lesions seen in adult women (Harel 2012)

Subtle findings in this population

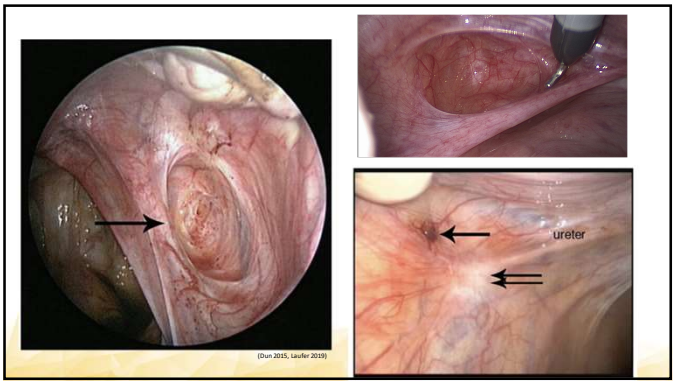


(Dun 2015, Laufer 2020)

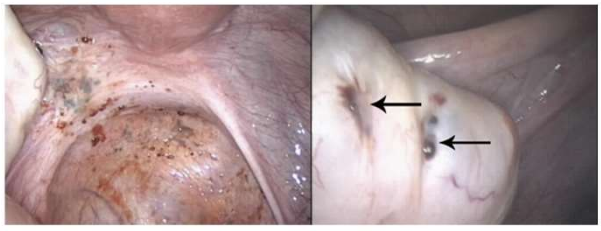
Liquid distention to clarify subtle findings



(Shim 2020, Laufer 1997)



(Dun 2015, Laufer 2019)


(Dun 2015)

Surgical goals: diagnosis and treatment

(ACOG CO 760, Dun 2015, Bafort 2020)

- Reduce bulk of disease and alleviate symptoms
- Minimize risks of surgery
- Maintain reproductive capacity
- Lack of data regarding extensive peritonectomy

Excision or Ablation?

- Minimal difference in outcomes between excision or ablation in early endometriosis (Wright 2005, Healey 2014)
- Can we extrapolate from studies performed in adults?
  - Different study metrics (e.g. dyspareunia)
  - Do not all start on medications
  - Type of peritoneal ablation varied
  - Long term differences unknown (multiple decades and surgeries)

Combination treatment

- Surgery alone may be insufficient
  - Recurrent disease and/or adhesions despite peritoneal excision (Laufer 2019)
- Benefit of extensive excisional surgery for severe disease unknown in adolescent population with minimal disease
- Combine with medications for ongoing suppression (Doyle 2009, ACOG CO 760)


(Laufer 2019)

Hormonal treatments?

Similarities to adults:

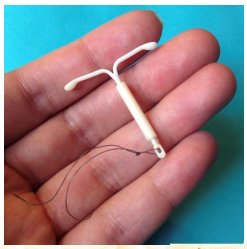
- Hormonal pills, injection, patch, and vaginal ring may provide ongoing suppression and contraception (ACOG CO 760)
- GnRH agonists and antagonists (DiVasta 2015, Taylor 2017)
- Methyltestosterone and danazol may be helpful in patients who seek masculinization effects (Shim 2020)

Differences to consider:

- Bone health/accrual (DiVasta 2015, Taylor 2017)
- Side effect tolerability
- Patient compliance
- Insurance coverage
- Need for continuous contraception

Placement of Progestin IUD

- Concurrent placement with laparoscopy minimizes need for potentially difficult office procedure (Yoost 2013, Abou-Setta 2013)
  - Treatment for dysmenorrhea
  - Ongoing prevention of pregnancy
- Important to confidentially assess contraception desires
- Can be used with additional endometriosis treatment



(Univ Michigan)

Additional treatments

Similarities to adults:

- NSAIDs, multimodal meds
- Support groups
- Online communities
- Pelvic floor physical therapy
- Management of comorbid disease (GI, psych, heme)
- TENS units
- Acupuncture
- Mindfulness

Differences to consider:

- Use of social media
- May not tolerate endovaginal PT techniques or have transportation for additional visits
- Increased psychological burden during COVID era, moreso in teens and in girls in particular (Liu 2022)

Conclusions

- Endometriosis should be considered in young people with refractory dysmenorrhea
- Laparoscopy remains the standard of care for diagnosis and treatment (excision or ablation) with ongoing medical treatment
- A close relationship between MIGS and PAG providers allows for appropriate initial treatment and ongoing longitudinal care

Acknowledgments

- AAGL PAG SIG
- NASPAG

NASPAG

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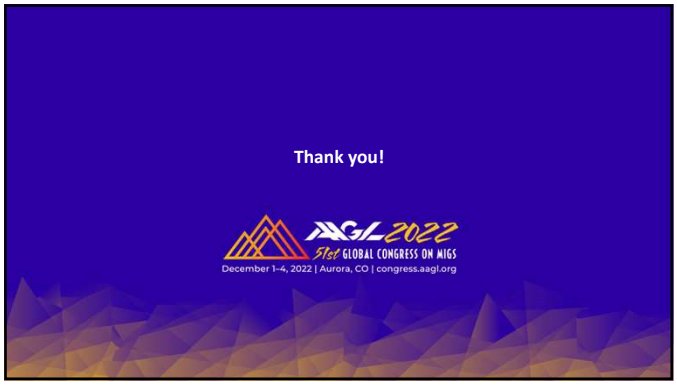
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Management of Adnexal Masses in the Adolescent & Young Adult

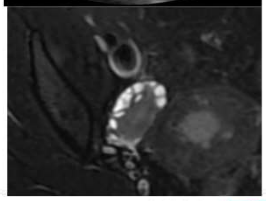
Nichole Tyson, MD
Clinical Associate Professor
Pediatric and Adolescent Gynecology
AAGL Post Graduate Course, 2022

Disclosures

- HRA Pharmaceuticals- Consultant to apply for FDA approval for adolescent use of the over the counter birth control pill

Objectives

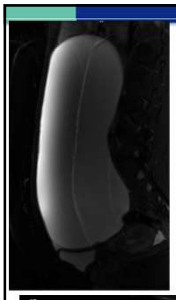
- Describe surgical strategies to address the large adnexal mass with a focus on ovarian sparing minimally invasive surgery

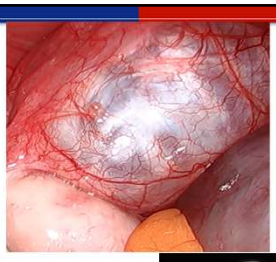


- PCOS

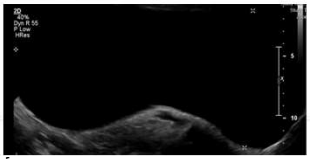
Ponnatapura, J., Dyer, R.B. & Ou, J.J. The ovarian "string-of-pearls" sign. *Abdom Radiol* **44**, 2019

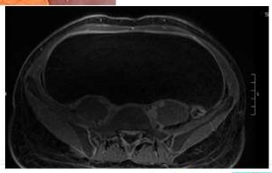
- Hemorrhagic cyst





Paratubal cyst





- Endometrioma

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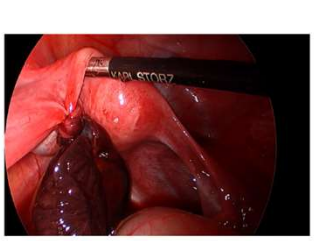

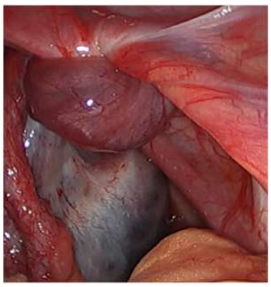

- Torsion

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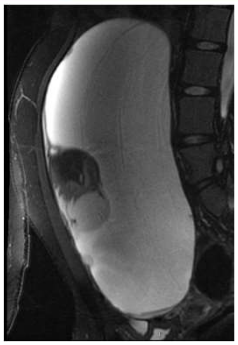

- Dermoid

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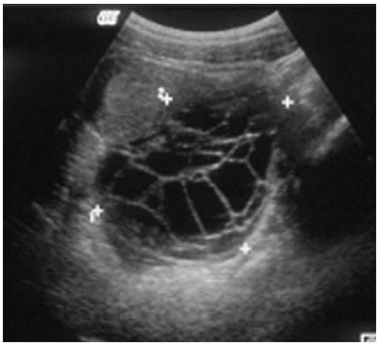

- Granulosa cell

9

Nathalie A. Fleming, et al Juvenile Granulosa and Theca Cell Tumor of the Ovary as a Rare Cause of Precocious Puberty: Case Report and Review of Literature, JPAG. 2010.

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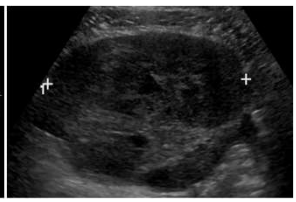

- Sertoli Leydig

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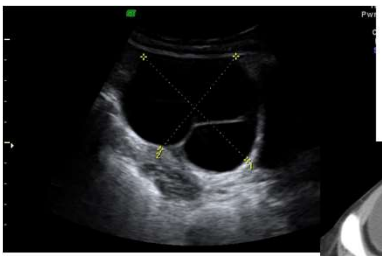

- Cystadenoma

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Case courtesy of Dr Praveen Jha, Radiopaedia.org, rID: 16979

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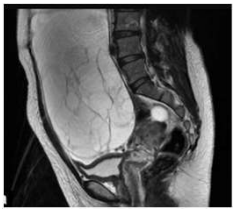

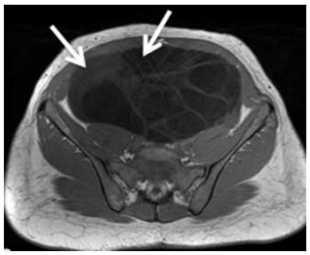

- Mucinous tumors

P.-E. Laurent, J. et al
Mucin-producing tumors of the ovary: MR imaging appearance,
Diagnostic and Interventional Imaging, 2015.

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- TOA

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Ovarian Masses in Children and Adolescents - An Analysis of 521 Clinical Cases

Pathology	Number of Patients	Percent total
Non-neoplastic	92	18
Ovarian torsion	8	1.5
Corpus luteum cyst	9	1.7
Follicular cyst	27	5
Simple ovarian cyst	16	3
Endometrioma	32	6

MZhang, et al.Ovarian Masses in Children and Adolescents - An Analysis of 521 Clinical Cases, JPAG, 2014

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Adnexal masses- Neoplastic & Benign

Pathology	Number of Patients	Percent total
Mature teratoma	270	52
Cystadenoma	86	16
Fibrothecoma	4	0.8
Struma ovarii	3	0.6
Sclerosing stromal tumor	3	0.6
Ovarian theca cell tumor	1	0.2
Fibroma	1	0.2
Ovary sex cord tumor	1	0.2
Borderline cystadenoma	13	2.5

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Adnexal masses- Neoplastic & Malignant

Pathology	Number of Patients	Percent total
Immature teratoma	19	3.6
Invasive mucinous cystadenoma	8	1.5
Yolk sac tumor	5	1
Dysgerminoma	4	0.8
Mixed germ cell tumor	4	0.8
Sertoli Leydig cell tumor	4	0.8
Gonadoblastoma	2	0.4
Juvenile granulosa cell tumor	1	0.2

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History

- Obtain history from parent/s and patient
- Gestational history
  - Maternal hypothyroidism
  - Gestational diabetes
  - Ovarian cyst on antenatal ultrasound
- Prepubertal pertinent history
  - Thelarche
  - Growth spurt
  - Vaginal discharge
  - Uterine bleeding

May indicate physiologic ovarian cyst or E2 secreting granulosa cell tumor

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History

- Symptoms of virilization
  - Hirsuitism
  - Acne
  - Deepening of voice
  - Clitoromegaly

May be indicative of testosterone excess from Sertoli Leydig tumor

History in adolescent

- Menstrual history
  - Thelarche, menarche, LMP
  - Dysmenorrhea-quality, severity & duration of pain
  - Irregular cycles

Help assess for physiologic cysts and endometriosis

- Severe pain

Think ruptured hemorrhagic corpus lutuem and if + nausea/vomiting think--torsion

Adolescent confidential visit

- Sexually active?
  - Is this a pregnancy related adnexal mass
  - Ectopic pregnancy
  - TOA?

Exam considerations unique to PAG

- Breast budding before age 7-8 may reflect elevated E 2 levels from central stimulation (LH/FSH) or from ovarian tumor or physiologic cyst
- Growth charts can help assess growth spurt indicating puberty
- Vaginal exams are often particularly painful.
- Rectal examinations in prepubertal and young teens are often much less painful and should be considered
- Single digit bimanual exam

When to get tumor markers?

- Solid component > 2 cm
- Thick septations
- Ascites
- Precocious puberty or virilization

TUMOR MARKERS

- AFP
- BHCG
- LDH
- Inhibin A & B

- CA 125- can be elevated for a variety of benign reasons in post pubertal patients (endometriosis, torsion, even menses)
- CEA in young adults

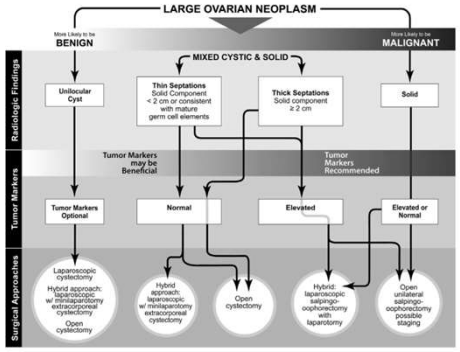
Neoplasm	Elevated tumor markers
Germ cell tumors-dysgerminoma, choriocarcinoma, embryonal, yolk sac tumor, immature teratoma, mixed germ cell	AFP HCG LDH
Granulosa cell	Estradiol Inhibin
Thecoma	Estradiol Testosterone
Sertoli Leydig	Testosterone Inhibin

Imaging

- U/S is gold standard/first line
- MRI has been explored as diagnostic option for unusual or rare situations where the U/S diagnosis is not clear
- Expensive
- Preop planning

Expectant Management

- Imaging suggests physiologic
- Post pubertal and < 8 cm
- Prepubertal and < 5 cm
- No solid component beyond septations/debris from hemorrhage
- Negative tumor markers
- No torsion concerns
- Family compliance w/ follow up plan



A.-M.E. Amies Oelschlager et al. / J Pediatr Adolesc Gynecol 29 (2016) 88e94

Large cyst and CA risk

- Childhood incidence of ovarian cancer is 2.6/100,000
- 424 patients with 11% malignancy
- 521 patients with 9% malignancy
- Malignancy was associated with mean mass size of 17 cm, benign mass size mean was 8 cm
- No malignancy was seen when size was < 6 cm

Oltmann S, Garcia N, Barber R, et al: Can we preoperatively risk stratify ovarian masses for malignancy? J Pediatr Surg 2010; 45:130
Zhang et al JPAG 2014

Frozen section

- Sensitivity is less than 70%
- 23% of tumors declared borderline at frozen section were found to be malignant on final pathology
- Decision to proceed with cystectomy vs. oophorectomy should be based on imaging/tumor markers rather than frozen section

Puls L, Heidtman E, Hunter JE, et al: The accuracy of frozen section by tumor weight for ovarian epithelial neoplasms. Gynecol Oncol 1997; 67:16

Oophorectomy vs. Cystectomy

- The only absolute indication for the decision to perform an oophorectomy of a large ovarian cyst is known malignancy.
- Many oophorectomies performed in the pediatric population, which in retrospect may have been unnecessary
- Unilateral oophorectomy has an odds ratio of early menopause (<40) of 4.3 and a decrease in AMH-more to learn
- PRESERVE the Ovary unless suspect malignancy

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There's no ovary left…

- Some erroneously believe that no normal functional ovary remaining when an ovary is stretched by tumor.
- During an ovarian cystectomy, the cyst can be dissected from the ovarian cortex
- Histological examination of the ovarian cortex maximally stretched by the tumor demonstrates normal ovarian tissue

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Maneschi, Lorenzo et al, Ovarian cortex surrounding benign neoplasms: A histologic study. American Journal of Obstetrics and Gynecology, 1993

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Cyst rupture

Chemical peritonitis

Historical concern

Chemical peritonitis is rare

Overall outcomes are not different

Fertility

Historical concern

One study found that those with intraoperative rupture had a significantly higher rate of spontaneous pregnancy (100%) versus 68.9% without intraoperative rupture (P = .01).

G. Zanetta, *et al.* **Laparoscopic excision of ovarian dermoid cysts with controlled intraoperative spillage. Safety and effectiveness** J Reprod Med 1999

M. Pansky, *et al.* **Inadvertent rupture of benign cystic teratoma does not impair future fertility** Am J Obstet Gynecol, 2010

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Surgical Approach
Benefits of Laparoscopy

- less pain
- shorter hospitalization
- reduced recovery time
- lower incidence of infection
- less bleeding
- more satisfaction with scars
- lower rates of adhesive disease which may impact future fertility.
- more cost effective than laparotomy despite increased equipment cost and increased operating time

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Surgical approach

- Great video of laparoscopic cystectomy

https://www.jmig.org/article/S1553-4650(19)30222-5/fulltext

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Minilaparotomy for LARGE cysts

- Alexis wound retractor
- Purse string suture around wound
- Oschner trocar (Gallbladder suction)
- Dermabond to cyst
- Sterile ultrasound bag/ decompress cyst
- Drain/decompress to exteriorize

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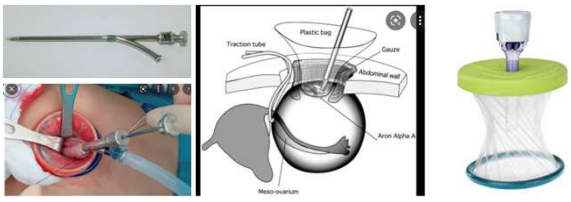
Tools and Tricks



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CYST RUPTURE- CA

- Tumor rupture, upstages the cancer
- Some studies demonstrate serious cystadenocarcinomas w/ intraop rupture in adults have not had increased mortality or morbidity with intraoperative cyst rupture compared to patients where there was positive cytology upon entry into the abdomen
- Limited applicability to pediatric and adolescent females where epithelial malignancies are rare
- Small studies and anecdotal experience—rupture upstaged and extensive peritoneal disease
- Granulosa cell tumors present with early stage disease, unilateral salpingo-oophorectomy should be performed

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Tumor Recurrence

ADNEXAL MASS	RECURRENCE RATE
Mature Teratoma	7.6% at 2 years Contralateral recurrence 14% at 3 years Bilateral post op surveillance
Benign Mucinous Cystadenoma	3/11 w/ cystectomy (3 had intraop rupture) 0/28 with adnexectomy Post op surveillance
Immature Teratoma	Not enough data Diagnosis may not be clear or finalized until final path, so may need second surgery (USO)
Borderline Epithelial Tumor	USO- 15% recurrence Cystectomy 36% recurrence Even with bilateral adnexectomy-5.7% recurrence Cystectomy w/ post op surveillance

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QUESTIONS?

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Managing the Patient with Obstructive Mullerian Anomalies

Krista J. Childress, MD
Associate Professor, Pediatric & Adolescent Gynecology
University of Utah/Primary Children's Hospital

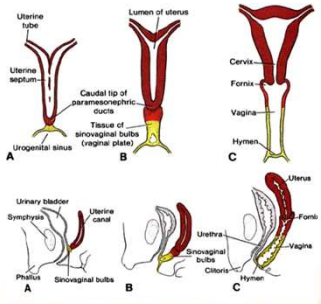
AGL 2022 Star Global Congress on MIGS
December 1-4, 2022 | Aurora, CO | congress.aagl.org

Disclosure

I have no financial relationships to disclose"

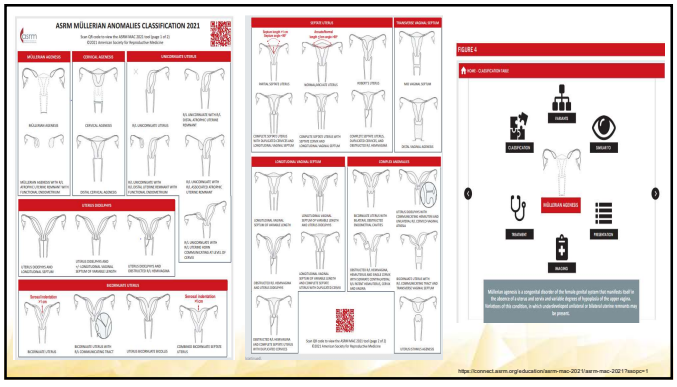
Objectives

- Review embryology
- Understand ASRM classification of Mullerian Anomalies
- Comprehend how to evaluate and manage common obstructive Mullerian anomalies
- Identify considerations for surgical management in the pediatric and adolescent patient population.



Paramesonephric (Mullerian) ducts
- Uterus, fallopian tubes, upper 1/3 vagina
- Fusion and canalization

Urogenital sinus (UG sinus)
- Lower 2/3 vagina and vestibule



Associated Anomalies

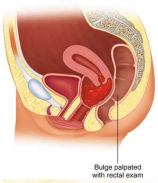
- 20-30% have renal anomalies
  - Up to 40% with unicornuate uterus
- Skeletal/spinal 16.4%
- Gastrointestinal 12%
- Heart defects 5.0%
- Abnormal neurologic status 1.3%

Kapcsik K, et al. Obstructive Müllerian Anomalies in Menstruating Adolescent Girls: A Report of 22 Cases. J Pediatr Adolesc Gynecol. 2018

Common Obstructive Mullerian Anomalies

- Imperforate hymen
- Lower vaginal atresia
- Transverse vaginal septum
- Obstructive hemivagina and ipsilateral renal anomaly (OHVIRA)
- Non-communicating uterine remnants
- Cervical or cervicovaginal agenesis

- **Presentation**
  - Usually found after puberty
  - Primary amenorrhea and pain
  - Progressive dysmenorrhea
  - Abdominal mass
  - Urinary retention
  - Constipation
- **Physical examination**
  - Vaginal introitus present?
  - Length of the vagina?
  - Cervix?
  - Rectal exam: distance of hematocolpos from perineum
- **Imaging**
  - US, MRI, 3D US



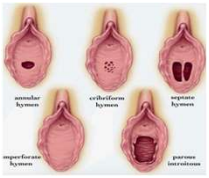
Hymen Abnormalities

Origin: Urogenital sinus

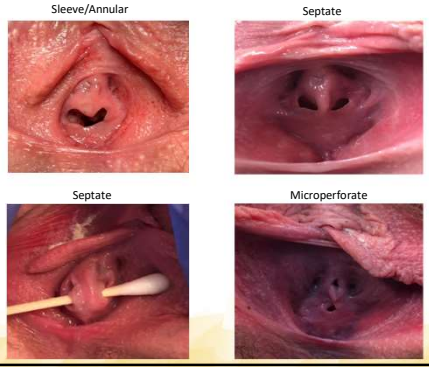
Variants

Symptoms
- Difficulty with tampon insertion
- Abdominal pain

Imperforate hymen
- 1/2000 girls



Dietrich et al. Obstructive Mullerian Anomalies. IPAG 2014.



Case 1

- 12 year old seen in the ED twice for pelvic pain. Diagnosed with UTI and constipation (negative cultures).
- 3rd presentation to the ED she was having difficulty voiding
- Transabdominal pelvic US ➔ prompted a genital exam…

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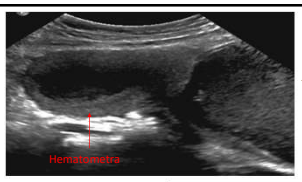
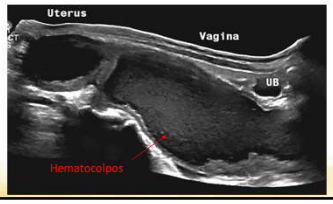


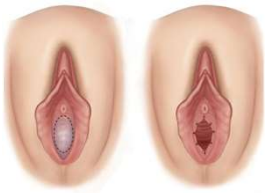
Uterus + patent cervix



Uterus + large hematocolpos

Classic exam findings:
- Bulging blue hymen
- Protrudes with valsalva
- No true hymen tissue noted

→ **Imperforate Hymen**



- Very thin tissue → +/- sutures
- Occasionally imperforate hymen ruptures
- Rarely requires dilation post-operatively

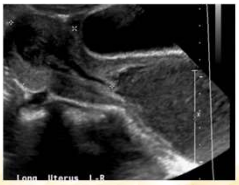
Imperforate Hymen Resection

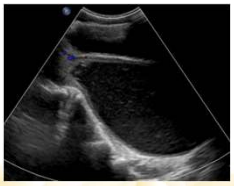
Hymenectomy (Imperforate Hymen) - YouTube

Children's Healthcare of Atlanta

Case 2

13 year old girl presented to the ED with 1 week of worsening abdominal pain. Thelarche at the age of 11. Pelvic US was done prior to a genital exam.





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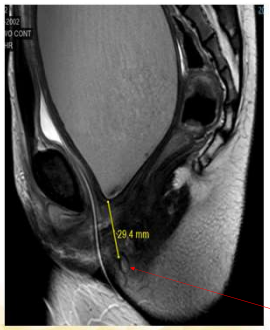
vs

- External exam not classically imperforate hymen
- Rectal exam: 2cm from perineum to hematocolpos
- Valsalva: minimal protrusion
- Further imaging?



- Measure length of atresia
  - Vitamin E capsule
  - US gel
- Clinical examination more reliable than imaging
- ?? 3 cm cut-off for graft ??
  - Exam with 1-2 cm distance
  - Distal progression with valsalva
- Management: OR for vaginal pull-through

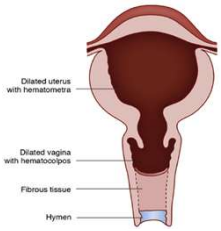
Vitamin E capsule

Lower Vaginal Atresia/Agenesis

- **Origin**: UG sinus
- Complete or partial
- Upper vagina present
- Usually no perineal bulge

- **Diagnosis:**
  - US
  - MRI

- **Treatment**:
  - < 3 cm → Vaginal pull-through
    - Native vagina*
  - > 3 cm → risk stenosis → consider graft



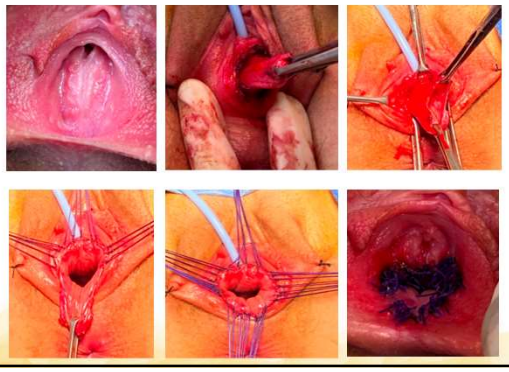
Vaginal Pull-Through



Surgical technique
- Mobilize upper vagina to the introitus
- Two fingerbreadths = adequate introitus

Concerns
- Introital stenosis
- Vaginal dilation

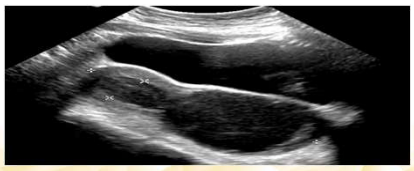
Top pictures courtesy of Holly Hartgen, MD




Cooper Surgical Milex Vaginal-Hymenal Silicone Dilators - Milex Vaginal-Hymenal Silicone Dilator, Size XL - MX20IXL

Case 3

13 year old girl presented to the ED with 1 week of worsening abdominal pain. Thelarche at the age of 11. Pelvic US was done prior to a genital exam.



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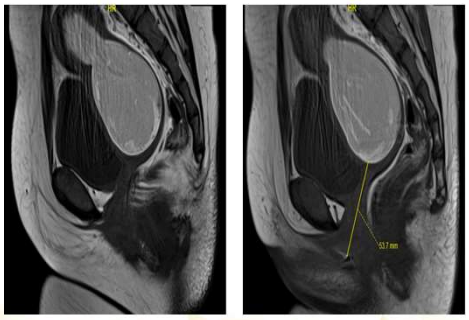
- No bulge with Valsalva
- No obvious hymen tissue
- Patulous urethra
- Rectal exam

Hematocolpos

↓

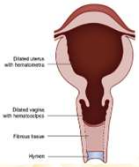
5-6cm from introitus



MRI: 5.4cm lower vaginal atresia

Counseling

- **Considerations**
  - Will need graft (> 3cm)
  - Dilation
    - Shrink the atretic tissue distance
    - Post-operatively
  - Concern for re-stenosis with early surgery
- **Options**
  - Vaginoplasty now
  - *Menstrual suppression until mature
  - +/- drainage



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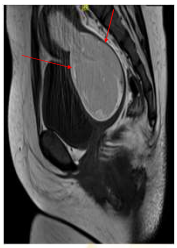
Failure of Menstrual Suppression

Symptomatic relief

↓

Drain hematometrocolpos

- **Laparoscopy**: vagina/uterus
- **Interventional radiology**
  - No good literature
  - Transabdominal
  - Case series presented at NASPAG 2021



Gennie et al. Laparoscopic drainage of hematocolpos. 2010

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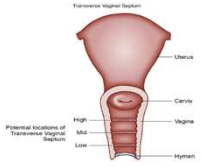
Vaginal Septum

Transverse vaginal septum

- Failed canalization
- Incomplete: allows menstruation
- Upper vagina > lower vagina



Longitudinal vaginal septum

- Failed fusion → 2 canals
- Associated with Mullerian anomalies (Didelphys)

Management: surgical resection

Dietrich et al.Non-Obstructive and Obstructive Mullerian Anomalies. JPAG 2014.

Longitudinal Septum

- Fibrous tissue
- How?
  - Ligasure
  - Clamp/cut/tie
- Re-approximate vaginal mucosa with interrupted sutures (3-vicryl or 4-0 vicryl)
- +/- vaginal stent

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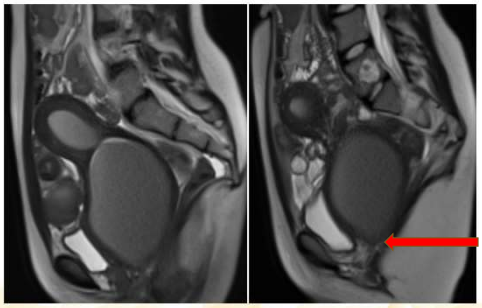
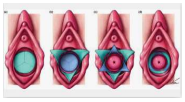
Longitudinal Vaginal Septum Resection

https://www.youtube.com/watch?v=ifiBFR__ISI

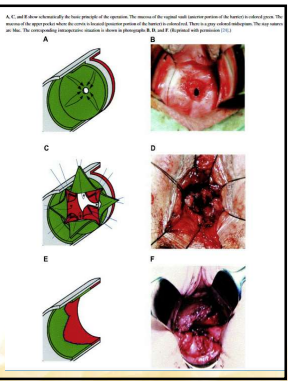


US gel into vagina to see thickness of septum

Counseling



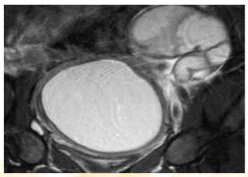
- **Considerations**
  - Septum mid vagina→ very difficult surgery
  - Z-plasty necessary?
  - Must do vaginal dilation
  - Too young to dilate
  - Concern for stenosis with early surgery
- **Options**
  - Vaginoplasty now + stent use
  - IR drainage (unable to access dilated vagina)
  - Allow further menses to push septum distal
    - Easier resection
    - Further examinations→ patient more comfortable
  - Menstrual suppression → allow to mature



Skinner B, Quint E. Obstructive reproductive tract anomalies: a review of surgical management. JMIG. 2017. Vo 24, No. 6, 901-908

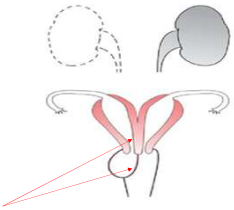
14 yo presents to the ED with abdominal pain. Menarchal x 6 months. Known history of left renal agenesis. Not followed by urology. Never seen a gynecologist. Regular periods.

Imaging shows…



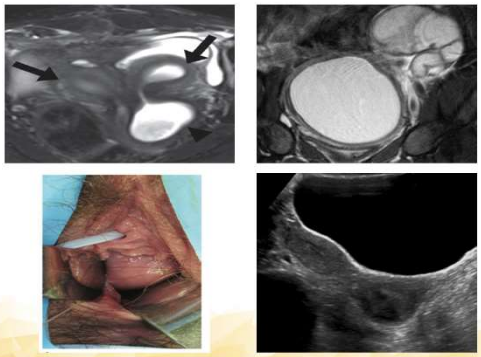
Obstructed Hemivagina Ipsilateral Renal Anomaly (OHVIRA)

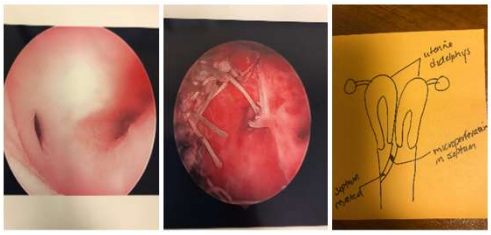
- **Renal Anomaly**
  - Renal agenesis, pelvic kidney, ectopic ureter
- **Diagnosis**
  - Menstrual history
  - Abdominal mass and/or vaginal bulge
  - Solitary cervix
  - US/MRI → what's the exact anatomy?
- **Septum/cervix:** +/- microperforation
- **Treatment**:
  - Transvaginal resection of septum



Uterine duplication
Obstructed hemivagina
Ipsilateral renal anomaly

Smith NA, Lauder MR. Fertil Steril. 2007; 87(4):918–22

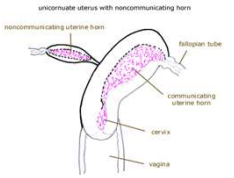


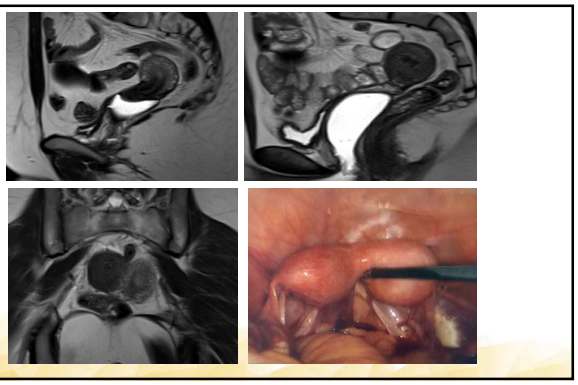


Surgical Correction
- Vaginoscopy → +/- microperforation
- Remove as much vaginal septum as possible without injuring the other cervix

Non-communicating Uterine Remnants

- Functional endometrium: distend with menstrual products→ pain
- Association with vaginal agenesis
- Diagnosis:
  - MRI
  - Hysteroscopy: 1-2 tubal ostia
  - Laparoscopy
- Management:
  - Removal of functional complete or partially obstructed uterine horns



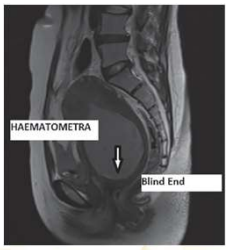


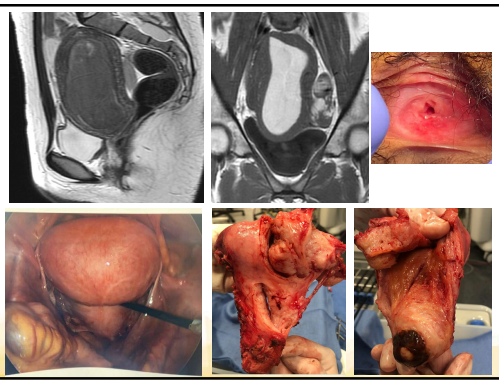
Non-Communicating Remnant Horn Resection

https://www.youtube.com/watch?v=BnWeAtRpcR0
Min 1:58

Cervical Agenesis and Cervicovaginal Agenesis

- Normally developed uterus
- Absent cervix
- Vaginal agenesis (50%)
- Mullerian anomalies (33%)
- **Diagnosis**: US and MRI
  - Hematometra
  - Hematosalpinx
  - MRI: not great at evaluating cervical tissues
- **Treatment**:
  - Menstrual suppression
  - Uterine remnant resection
  - +/- neocervix creation or attach vagina to uterus

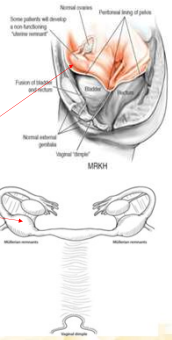




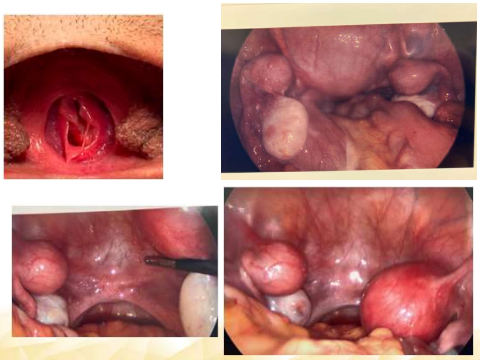
Vaginal Agenesis (MRKH)

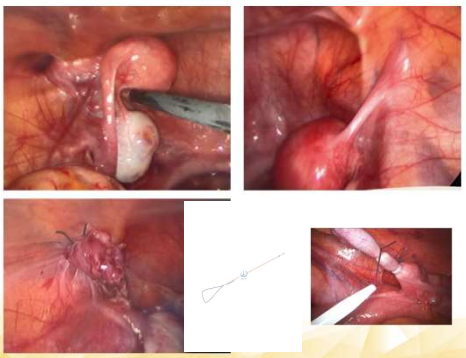
Mayer-Rokitansky-Kuster-Hauser Syndrome

- Agenesis of the uterus, cervix, and vagina
- 1 in ~5,000 female births
- Normal ovarian function
- Normal female phenotype
- Diagnosis: 46 XX, MRI
- **Uterine remnants** (47-84%)
  - 7-10% functional
    - Menstrual suppression
    - Surgical resection



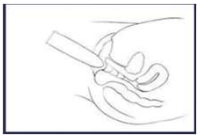
Dietrich et al. Non-Obstructive Mullerian Anomalies. JPAG 2016.





Neovagina Creation

- **Non-surgical**
  - Vaginal dilation
- **Surgical**
  - McIndoe
  - Modified McIndoe
  - Vecchietti
  - Davydov
  - Bowel



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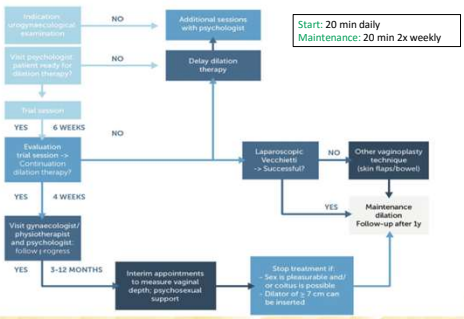
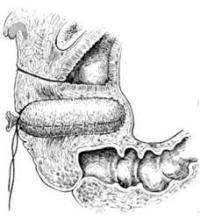
ACOG CO 728. 2018.

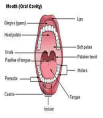
Dilation Protocol



Start: 20 min daily
Maintenance: 20 min 2x weekly

McIndoe + Modified





Classic: split-thickness skin grafts

Modified
- *Buccal
- Amnion
- Peritoneum
- Interceed
- Silicone
- Cellulose

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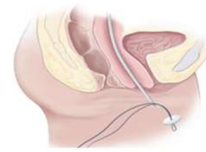
McIndoe AH. Banister JB. J Obstet Gynaecol Br Commonwealth. 1938; 45:490.
Abbe R. Med Record. 1898; 54:836.

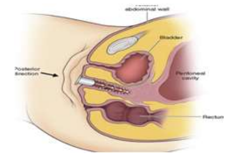
Modified McIndoe

https://www.youtube.com/watch?v=wDN-ubELdE4

Minute 1:40 starts dissection, then can fastforward
Minute 3:40 making the stent (they use placental amnion)
Minute 5:11 making of stent

Vecchietti (Traction)





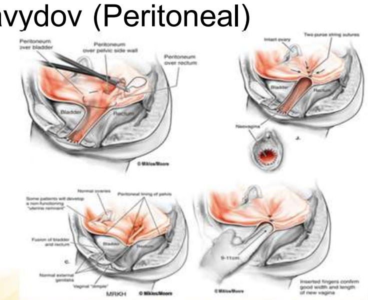
Modified
https://www.youtube.com/watch?v=gEgreEsmRek

NW
https://www.youtube.com/watch?v=LX7RjvYXVqc

Modified Laparoscopic Vecchietti's Procedure (surgeryu.com)

Brucker et a. Neovagina creation in vaginal agenesis. Fertil. Steril. 2008

Davydov (Peritoneal)



Vaginal lengthening
https://www.youtube.com/watch?v=0N7r7b0XNew

Vaginal agenesis
https://www.youtube.com/watch?v=OPQq13IjpDs

Plenary 1 - Laparoscopy - Creation of a Neovagina: A Modified Davydov Approach (surgeryu.com)

Technique	Advantages	Disadvantages
Pre-Operative Dilation method (Frank/Ingram)	• Non-invasive • No hospitalization • Preserves vaginal tissue • Cost–effective, inexpensive • Minimal morbidity and complications	• Motivated and mature patients required (low compliance) • Time-consuming • Discomfort, manual fatigue, and inability to complete • Limited success in younger patients • Poor results with skin dimple • Increased risk of vaginal prolapse
McIndoe procedure (split-thickness skin grafts)	• Vaginal approach avoids laparotomy • No bowel anastomosis • High rate of graft take • Low rate of prolapse	• Post-operative dilation needed • High rate of graft contracture and neovaginal stenosis • Disfiguring scar at the donor site • Potential of graft infection • Lack of lubrication • Risk for squamous cell carcinoma • Complication rate up to 14%
McIndoe (modified) Allogenic tissue	• Reduction of the occurrence of chronic granulation tissue	• Risk of allograft rejection • Risk of transmission of infectious diseases
Intestinal vaginoplasty (sigmoid colon, ileum, jejunum)	• Lubricated segment with reliable blood supply • Grows with the patient • No scarring • No dilation needed • Low rate of stricture • Can be performed with previous extensive surgery	• Requires laparotomy and bowel anastomosis • Prolapse 3–8% • Excessive discharge • Complication rate of 16–26% (post-operative ileus, bowel obstruction, diversion, ulcerative colitis, adenocarcinoma)
Full-thickness skin grafts Myocutaneous/ fasciocutaneous flaps Pudendal thigh flap Labia minora flaps Malaga flap Singapore flap Scapula flap	• Good blood supply, low infection rate • Flap incision incorporated into laparotomy site • Pedicled tissue form better than free-tissue, because no dilation required • No skin graft with pudendal thigh flaps	• High rate of prolapse and flap loss (gracilis) • Skin appendages • Deficient lubrication • Disfiguring scar at the donor site • Wound site infection • Hair regrowth in the neovagina

Callens N, et al. An update on surgical and non-surgical treatment for vaginal hypoplasia. Hum Repro Updat 2014; 20: 775-801. Table 3: Modified for space & relevance

Acknowledgments

- AAGL PAG SIG
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  - Dr. Kate McCracken
  - Dr. Lissa Yu
  - Dr. Strickland
  - Dr. Sanfillippo
  - All members!

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Dietrich J, Millar D, Quint E. Obstructive reproductive tract anomalies. JPAG. Vol 27, Issue 6, p396-402.
Romanski P, Bortoletto P, Pfeifer S. Unilateral obstructed Mullerian anomalies: a series of unusual variants of known anomalies
Callens N, et al. An update on surgical and non-surgical treatment for vaginal hypoplasia. Hum Repro Updat 2014; 20: 775-801. Table 3: Modified for space & relevance
Brucker et a. Neovagina creation in vaginal agenesis. Fertil. Steril. 2008
Nikolaev 1998, Miller 2008, Mansour 2015
Dennie et al. Laparoscopic drainage of hematocolpos. 2010
https://connect.asrm.org/education/asrm-mac-2021/asrm-mac-2021?ssopc=1

You Tube & Surgery U Videos

https://www.youtube.com/watch?v=0N7r7b0XNew
https://www.youtube.com/watch?v=OPQq13IjpDs
Plenary 1 - Laparoscopy - Creation of a Neovagina: A Modified Davydov Approach (surgeryu.com)
Modified
https://www.youtube.com/watch?v=gEgreEsmRek
https://www.youtube.com/watch?v=LX7RjvYXVqc
Modified Laparoscopic Vecchietti's Procedure (surgeryu.com)
https://www.youtube.com/watch?v=wDN-ubELdE4
https://www.youtube.com/watch?v=BnWeAtRpcR
https://www.youtube.com/watch?v=ifiBFR__ISI
Hymenectomy (Imperforate Hymen) - YouTube


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An Overview Of Gynecologic Care For Adolescents And Young Adults With Anorectal Malformations

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Disclosure

- I have no financial relationships to disclose

Objective

- Understand the importance of comprehensive, longitudinal gynecologic care for girls/women with anorectal malformations

Reproductive Health – a longitudinal, lifespan assessment

- Initial evaluation & surgical planning
- Pubertal evaluation
- Sexuality, family planning, obstetrical outcomes



Anorectal Malformations: Why include gynecology?

- Recent data reviewing 466 female patients with ARMs revealed:
  - 31% total had associated gynecologic anomaly/anomalies
  - Most common anomalies included:
    - Longitudinal vaginal septum
    - Uterine didelphys
    - Vaginal Atresia
  - Likelihood of gynecologic anomalies increases with more complex ARMs
    - Perineal Fistula: 5%
    - Rectovestibular Fistula: 18%
    - Cloaca: 68%

Anorectal Malformations: Why include gynecology?

- Likelihood of gynecologic differences increases with more complex ARMs
  - Rectoperineal Fistula: 5%
  - Rectovestibular Fistula: 18%
  - Cloaca: 68%



Why do associated gynecological anomalies occur?

- The Müllerian ducts form the internal female reproductive structures:
  - Most of the fallopian tubes, uterus, cervix, and upper vagina.
- During development, the Müllerian ducts undergo **growth and development at the same time as bowel and urinary structures.**
- Müllerian obstructions are more common in patients with renal, vertebral, anorectal, cardiac, tracheoesophageal and limb anomalies
- Think about with kidney anomalies!

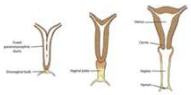
Epidemiology

- Uterovaginal differences have an incidence ranging from 3% in a population without symptoms to 67% in patients with cloacal anomalies
- Among female patients with a kidney anomaly, 29% also have a Müllerian anomaly

O'Flynn O'Brien KL, Bhatia V, Homafar M, Gong YY, Winsten MT, Gerber J, Dietrich JE. The Prevalence of Müllerian Anomalies in Women with a Diagnosed Renal Anomaly. J Pediatr Adolesc Gynecol. 2021 Apr;34(2):154-160.

Embryologic Development

- Müllerian ducts fuse and form the fallopian tubes, uterus, cervix and upper vagina
- Distally, the urogenital sinus separates into the urethra and distal vagina



- Müllerian obstructions are more common in patients with renal, vertebral, anorectal, cardiac, tracheoesophageal and limb anomalies

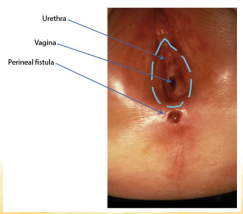
Infancy

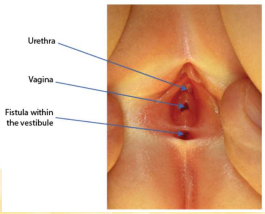
Initial evaluation

- At the time of primary surgery: suspected anatomy (vagina, septum, cervices, mullerian structures)
- Subsequent surgeries: opportunities to confirm/monitor for development of gynecologic structures
- Think about the pelvis and gynecologic structures anytime you are in the abdomen!!

Three openings

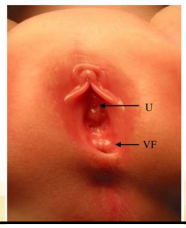
- Rectovestibular fistula
- Perineal fistula
- H-type rectovaginal





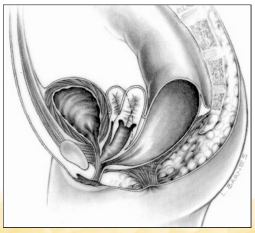
Two openings

- Vaginal atresia
- Rectovaginal fistula



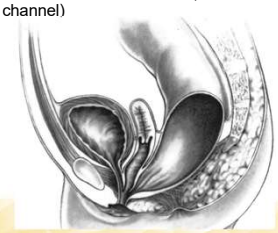
One opening

- Cloacal malformation



One opening

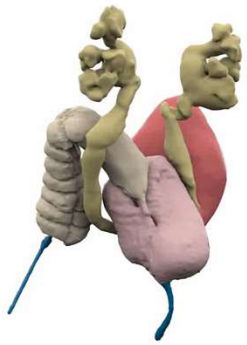
- Cloacal malformation (short common channel vs. long common channel)



More complex ARMs will undergo EUA, cysto-vaginoscopy and cloacagram

- **Common channel length***
- **Urethral length***
- Vaginal length
- ? Vaginal septum
- Number of cervices

*biggest factors in determining type of procedure for primary reconstruction: TUM vs separation



Vaginoscopy: Cloaca with 2 cm common channel

Limitations

- Pre-operative imaging is limited
- The structures are TINY and have not yet fully developed
- Unable to assess functional status – is there an endometrium?

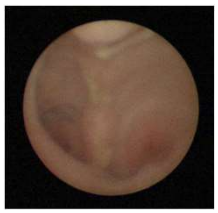
Number of cervices

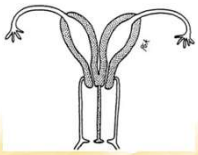
- Important to try to understand anatomy
- Risk of obstruction
- Information about reproductive potential

- **0**: does not guarantee absence of functional mullerian tissue
- **1**: does not promise normal mullerian anatomy
- **2**: duplicated uteri—typically associated with longitudinal vaginal septum

Presence of a longitudinal vaginal septum

- Removal at the time of the primary reconstruction versus at puberty
- Implies duplicated uteri (uterine didelphys)





Vaginal septum - longitudinal

Mullerian structures: the balance

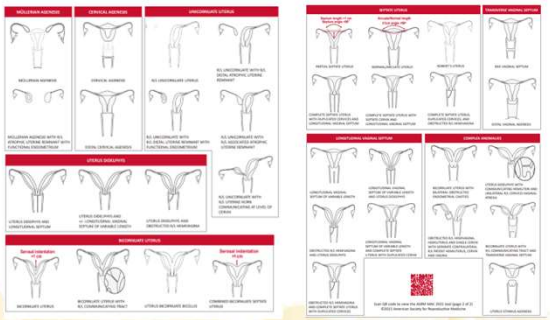
- Management of mullerian anatomy: balancing reproductive potential & risk of obstruction
- What defines a mullerian structure with reproductive potential?
  - Endometrium only?
  - Cervix?
  - Connected to vagina?
- Considerations – removing mullerian structures is permanent.
- Intraoperative decision making: can we identify anatomy at time of primary repair?

Dietrich, JE et al. NASPAG committee opinions: non-obstructive mullerian anomalies, obstructive mullerian anomalies

Mullerian structures: bottom line

- "It should be made clear to parents that the potential for uterine function cannot be assessed in childhood. These data suggest that where there is uncertainty, care should be taken when removing tissue with uterine potential."
- We err on the side of retention of mullerian structures to maximize reproductive potential and accept that we may increase risk of obstruction and need for additional surgery after menarche

Fernando, M. The long-term management and outcomes of cloacal anomalies. *Pediatri Nephr* 2015; 30:759-765.



Puberty

Puberty

- The ovaries should be normal—no associated problems with ovaries
- If healthy and normal gut absorption, puberty should happen at the normal time, in the normal tempo
- Estrogen from the ovaries: patients get taller, develop breasts, get their period
  - Thelarche (10y) then typically a 2-year interval until menarche (12y)
- Estrogen stimulation → see how the reproductive structures develop
  - Watch for menarche
  - Pain or lack of bleeding = red flags for obstructed menstrual outflow

Initial Goals

- **Ensure normal pubertal development**

MENSES = VITAL SIGN

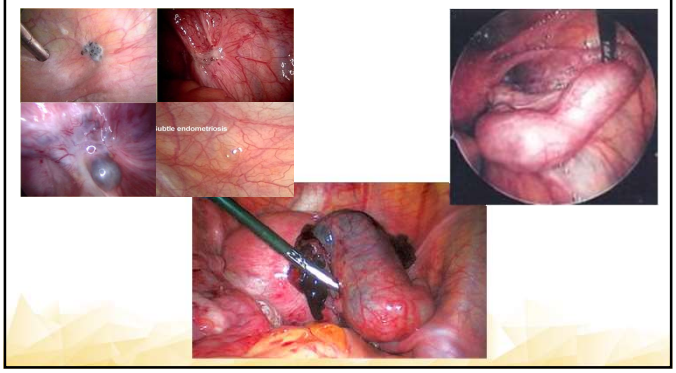
- **Ensure a patent outflow tract for menses**
  - Effective menstruation requires adequate uterine development, active endometrium, & a patent outflow tract
  - Pelvic ultrasound after breast budding
  - Uterine horn, cervical stenosis, vaginal stenosis, introital stenosis

PAIN = RED FLAG

- **Evaluate the introitus** – ensure adequate diameter for tampon use
- **Evaluate the vagina** – for stenosis, residual vaginal septum, graft prolapse

Uterine Horns / Remnants

- Management of mullerian anatomy: balancing reproductive potential & risk of obstruction
- Communicating vs non-communicating
- Functional vs non-functional
- Resect or not
- Risk of retrograde menstruation – endometriosis, scarring, hydrosalpinx, ectopic pregnancy



Ongoing Goals

- **Evaluate & discuss contraceptive needs** – not all methods may be appropriate
- **Evaluate the introitus** – ensure adequate diameter for vaginal intercourse if desired
- **Evaluate the vagina** – for stenosis, graft prolapse
- **Evaluate the perineal body**
- **Routine gynecologic care**
  - HPV vaccine
  - PAP testing
  - Sexually transmitted infection prevention/screening

Goals:
-Menstrual outflow
-Ability to engage in comfortable, satisfying sexual activity
-Ability to become pregnant & maintain pregnancy

Indications for procedures during puberty, adulthood

- **Obstructed menstrual outflow**
- **Introital stenosis**
  - Difficulty with menstrual hygiene & intercourse
- **Lack of perineal body**
  - Obstetrical indications
  - Sexual function – "orgasmic platform"
- **Rectovaginal fistula**
  - Fecal incontinence
- **Residual vaginal septum**
  - Difficulty with menstrual hygiene & intercourse

Sexuality, family planning obstetrical outcomes

Sexuality

WHO IS YOUR TEAM?
- Patient
- Nurses
- Counselor
- Pelvic floor physical therapist
- Gynecologist

- Lots of encouraging news – it does happen
  - On average ARM patients initiate sexual activity at an older age
  - Patients give positive reports of sexual functioning
- Gynecologic assessment prior to penetrative vaginal intercourse to evaluate vaginal opening, length, caliber
  - Some patients may need help with vaginal dilation, pelvic floor physical therapy, surgery
- Empower & educate regarding congenital and post surgical anatomy
  - Body acceptance
  - Sexuality is a normal part of human development / life
  - Sexual function is multidimensional
  - Sexuality is impacted by chronic illness – physical, psychological

Considerations in ARM patients

- **Anatomy**
  - Native vagina versus neovagina
  - Introitus – caliber, location
  - Presence or absence of mullerian (uterine) structures (perceived pregnancy risk)
- **Confidence**
  - Continence – urinary, stool
  - Stomas
  - Bowel regimen – ability to be spontaneous / autonomy
- **Pain**
  - Caliber of introitus
  - Scar tissue
  - Vaginismus
- **Past experiences**
- **Contraception**
  - General health considerations (congenital heart, renal disease)
  - Mullerian anatomy
  - WHO/CDC guidance of best/safest birth control methods

Obstetric Considerations

- **Discuss reproductive potential (pregnancy is possible in ARM pa**
  - Dependent upon anatomy
  - Alternative means for creating a family
  - Provide contraception if patient does not desire pregnancy
- **Encourage pre-conception counseling with a high risk obstetrician**
  - Obtain copies of operative reports
  - Evaluation of concurrent medical conditions (i.e. renal insufficiency)
- **Discuss potential for preterm labor / preterm delivery / malpresentation**
  - Dependent upon mullerian anatomy
  - Knowledge of the patient's gynecologic anatomy is crucial for counseling – use every available opportunity to document the upper & lower tract anatomy
- **Discuss recommended mode of delivery**
  - Vaginal versus cesarean section
  - Urologic and colorectal considerations

Obstetrical concerns

- Many reported births to women with ARM
- No maternal mortalities
- Most common complication: recurrent UTIs
- All mode of delivery reported (vaginal and cesarean section)
- On average, patients deliver early (36 weeks) due to preterm labor
- Consider other VACTERL associated conditions
  - renal (infection/preeclampsia)
  - spine (anesthesia)
  - cardiac (fetal risk/maternal risk with pregnancy)
  - uterine anatomy (preterm delivery/malpresentation)

Mode of Delivery

CESAREAN SECTION	VAGINAL DELIVERY
Repaired cloacal anomaly Vaginal replacement	Native vagina Women not relying on sphincters & sensation to remain clean/dry

- Shared medical decision making & individualization of care
- Risk / benefit discussion – continence matters
- Planned surgical approach – OB, colorectal, urology
- Where are the patient's stomas?

Gynecologic concerns in patients with cloacal anomaly. Breech L. Sem Pediat Surg 25 (2016) 90-95.

Additional GYN Considerations

- Spinal: IUD insertion for menstrual management if EUA
- Sexual health questions
- Period management
- HPV vaccine initiative
- Non-ARM GYN diagnoses: ovarian cysts, dysmenorrhea, labial adhesions
- Constipation, pelvic pain, pelvic floor dysfunction, urinary dysfunction

Take Home Points

- Longitudinal / lifespan approach is necessary
- Comprehensive reproductive health care is essential for ARM patients
- Linking the patient to adult providers who have familiarity with complex anomalies may be difficult – but is crucial for best outcomes
- Pregnancy should be managed in a multidisciplinary model
- There is a paucity of data regarding sexual function and obstetrical outcomes – this is an opportunity for collaborative research

Questions? Thank you!

kate.mccracken@nationwidechildrens.org

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March 23-25, 2023

CULTURAL AND LINGUISTIC COMPETENCY & IMPLICIT BIAS

The California Medical Association (CMA) announced new standards for Cultural Linguistic Competency and Implicit Bias in CME. The goal of the standards is to support the role of accredited CME in advancing diversity, health equity, and inclusion in healthcare. These standards are relevant to ACCME-accredited, CMA-accredited, and jointly accredited providers located in California. AAGL is ACCME-accredited and headquartered in California.

CMA developed the standards in response to California legislation (Business and Professions (B&P) Code Section 2190.1), which directs CMA to draft a set of standards for the inclusion of cultural and linguistic competency (CLC) and implicit bias (IB) in accredited CME.

The standards are intended to support CME providers in meeting the expectations of the legislation. CME provider organizations physically located in California and accredited by CMA CME or ACCME, as well as jointly accredited providers whose target audience includes physicians, are expected to meet these expectations beginning January 1, 2022. AAGL has been proactively adopting processes that meet and often exceed the required expectations of the legislation.

CMA CME offers a variety of resources and tools to help providers meet the standards and successfully incorporate CLC & IB into their CME activities, including FAQ, definitions, a planning worksheet, and best practices. These resources are available on the CLC and IB standards page on the CMA website.

Important Definitions:

Cultural and Linguistic Competency (CLC) – The ability and readiness of health care providers and organizations to humbly and respectfully demonstrate, effectively communicate, and tailor delivery of care to patients with diverse values, beliefs, identities and behaviors, in order to meet social, cultural and linguistic needs as they relate to patient health.

Implicit Bias (IB) – The attitudes, stereotypes and feelings, either positive or negative, that affect our understanding, actions and decisions without conscious knowledge or control. Implicit bias is a universal phenomenon. When negative, implicit bias often contributes to unequal treatment and disparities in diagnosis, treatment decisions, levels of care and health care outcomes of people based on race, ethnicity, gender identity, sexual orientation, age, disability and other characteristics.

Diversity – Having many different forms, types or ideas; showing variety. Demographic diversity can mean a group composed of people of different genders, races/ethnicities, cultures, religions, physical abilities, sexual orientations or preferences, ages, etc.

Direct links to AB1195 (CLC), AB241 (IB), and the B&P Code 2190.1:

Bill Text – AB-1195 Continuing education: cultural and linguistic competency.
Bill Text – AB-241 Implicit bias: continuing education: requirements.
Business and Professions (B&P) Code Section 2190.1

CLC & IB Online Resources:

Diversity-Wheel-as-used-at-Johns-Hopkins-University-12.png (850×839) (researchgate.net)
Cultural Competence In Health and Human Services | NPIN (cdc.gov)
Cultural Competency – The Office of Minority Health (hhs.gov)
Implicit Bias, Microaggressions, and Stereotypes Resources | NEA
Unconscious Bias Resources | diversity.ucsf.edu
Act, Communicating, Implicit Bias (racialequitytools.org)
https://kirwaninstitute.osu.edu/implicit-bias-training
https://www.uptodate.com/contents/racial-and-ethnic-disparities-in-obstetric-and-gynecologic-care-and-role-of-implicitbiases
https://www.contemporaryobgyn.net/view/overcoming-racism-and-unconscious-bias-in-ob-gyn
https://pubmed.ncbi.nlm.nih.gov/34016820/