Pediatric & Adolescent Gynecology –
A How To Approach (Didactic)

PROGRAM CHAIR
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Professional Education Information

Target Audience
Educational activities are developed to meet the needs of surgical gynecologists in practice and in training, as well as, other allied healthcare professionals in the field of gynecology.

Accreditation
AAGL is accredited by the Accreditation Council for Continuing Medical Education to provide continuing medical education for physicians.

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Course Description

This course is designed to allow clinicians to establish an “adolescent-friendly environment” in their office setting. Strategies for practice development focused on minimally invasive surgical expertise will be provided. A “how to” Approach is the underlying theme for all lectures in the postgraduate course. Gynecologic surgeons are increasingly being called upon to manage Müllerian anomalies; pre-operative as well as intra-operative expertise will be emphasized. As surgeons we are asked with increasing frequency to assist is fertility preservation when a young patient is faced with a diagnosis of cancer or other chronic debilitating disease. Various surgical approaches that clinicians with advanced minimally invasive expertise should be able to acquire will be presented in a readily applicable manner. Current concepts with regard to management of adnexal masses, torsion, and endometriosis in the young adult will allow surgeons to garner the latest advances of gynecologic surgery in this age group.

Course Objectives

At the conclusion of this course, the participant will be able to: 1) Use the learning process to provide counseling and expertise to facilitate development of an adolescent and young adult gynecologic surgical practice focused on minimally invasive surgical techniques; 2) evaluate and manage Müllerian anomalies with surgical as well as non-surgical approaches will be stressed; and 3) discuss the challenges of managing disorders of sexual development, quantified and streamlined to facilitate counseling and surgical correction.

Course Outline

1:30 Welcome, Introductions and Course Overview  
J.S. Sanfilippo

1:35 Developing a Pediatric and Adolescent Gynecology Minimally Invasive Practice – A How to Approach  
J.S. Sanfilippo

2:00 Minimally Invasive Surgery in the Pediatric and Adolescent Patient:  
Vaginoscopy, Hysteroscopy, Laparoscopy and Robotics  
R.K. Zurawin

2:25 Obstructive Müllerian Anomalies and Hematocolpos – What You Can Do  
H. Appelbaum

2:50 Disorders of Sexual Development  
R.K. Zurawin

3:15 Questions & Answers  
All Faculty

3:25 Break
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<tr>
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<td>Minimally Invasive Surgical Management of Adnexal Masses and Torsion</td>
<td>H. Appelbaum</td>
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<td>4:05</td>
<td>Endometriosis in Adolescents – A Whole Different Ball Game</td>
<td>J.S. Sanfilippo</td>
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<tr>
<td>4:30</td>
<td>Minimally Invasive Surgical Management with Vaginal Agenesis</td>
<td>H. Appelbaum</td>
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<tr>
<td>5:20</td>
<td>Questions &amp; Answers</td>
<td>All Faculty</td>
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The following members of AAGL have been involved in the educational planning of this workshop and have no conflict of interest to disclose (in alphabetical order by last name).
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Frank D. Loffer, Executive Vice President/Medical Director, AAGL*
Linda Michels, Executive Director, AAGL*
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FACULTY DISCLOSURE
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).
Joseph S. Sanfilippo*
Heather Appelbaum*
Robert K. Zurawin
Consultant: Ethicon Endo-Surgery, Ethicon Women's Health & Urology, Conceptus Incorporated, CONMED Corporation, UpToDate
Mark R. Hoffman*
Asterisk (*) denotes no financial relationships to disclose.
41st AAGL GLOBAL CONGRESS
PEDIATRIC & ADOLESCENT GYNECOLOGY
A How To Approach
"Developing a Pediatric & Adolescent Gyn Minimally Invasive Practice-How to Approach"

Joseph S. Sanfilippo, MD, MBA
University of Pittsburgh
Magee-Womens Hospital

Disclosure

I have no financial relationships to disclose.

Objectives

• At the conclusion of this lecture participant will:
  • Review unique aspects of pediatric adolescent Gyn exam
  • Discuss the evaluation & management of common PAG problems
  • Establish a pediatric adolescent focused clinical setting
Fig. 1. Child's position for gynaecological examination.

Fig. 2. Vaginal examination: separation of labia major by pulling the inferior part downward and laterally.

Fig. 3. Vaginal examination: separation of labia major by pulling them anteriorly.
PEDIATRIC PATIENT
- Patient Involved in History
- Frog-legged Position
- Knee-chest Position
- "Show and Tell"
- Low power Magnification
- "Good job"

PELVIC EXAM
- "Do You Use Tampons?"

ADOLESCENT EXAM-PARADIGM SHIFT
- TOOL KIT-ACOG
- First Exam
- "Gynecologic Encounter" 13-15 Y/A
  - Collaborative with Primary Care Provider
  - Support with OB GYN
  - No Pelvic Exam
  - Followed By Annual Visits
- Pap Smear-21 years of age
  - Exception: Sexual Abuse Immunocompromised
  - Not Sexually Active

Stewart F et al JAMA 2001;286:671 ACS, NIH, ACOG 2002
SCREENING FOR SEXUALLY TRANSMITTED DISEASES

- Sexually Active Teens Should be Screened
- “Urine screening Should be Considered When Teens are Reluctant to Have a Pelvic Exam”
  - Urine Ligase Chain Reaction - Less Expense than Cervical Cultures
- “Vaginal Swab” Screening

Health Care for Adolescents ACOG 2002

WHAT DO I NEED TO KNOW ABOUT EXAMINING A PEDIATRIC ADOLESCENT PATIENT?
**TEEN HEALTH GAP**
*Time Magazine*

- Adolescents Have Unique Medical Issues
- A Growing Specialty is Aimed at Addressing Them
- Quote from a Teen (19 y/a)
  - “Because many doctors shy from discussing drugs & alcohol with teens, some kids struggle for years before finding help. (Teen is a recovering addict and grateful for early intervention.)

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**RISK FACTORS—TEENAGE PREGNANCY**

- Poor Educational Performance
- Poverty
- Single-Parent Family
- Family History Teen Pregnancy
- “Pregnancy Viewed as “+” Social Value
- Risk Taking Behavior—Drugs Alcohol Abuse

**PROTECTIVE FACTORS**

- Scholastic Achievement
- Higher Socioeconomic Class
- Intact Family
- Attendance at Religious Services

Elfenbein D, et al Pediatr Clin NA 2003;781

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**Issues & Answers NASPAG 2011**

- Teen Births: 400,000 Annually
  - 9 Times Greater Than Other Developed Countries
  - Greater Maternal Morbidity—Teens
  - “A Battle We Can Win” per Centers for Disease Control (CDC)

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**WHAT CAN WE LEARN FROM TEENS?**

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**Committee Opinion**

Number 357, December 2006 *(Replaces No. 292, November 2003)*

Primary and Preventive Care: Periodic Assessments

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**AN ESSENTIAL GUIDE FOR PARENTS**

*U.S. News*

**MYSTERIES OF THE TEEN YEARS**
LABIAL AGGLUTINATION

- Common-Age 2-3 years
- Chronic Vulvovaginitis
- Urine irritation
- Perineal Hygiene
- Estrogen Cream
- Zinc oxide
- Reoccurrence
**LICHEN SCLEROSIS**
- Pruritis-Burning
- 25% Associated Autoimmune Disease
  - Thyroid, Pernicious Anemia, HLA Class II Antigen
- Vesicles or Bullae
- 1%-2% Hydrocortisone Cream
- 0.05% Clobetasol
- Long-term Recurrence
- 3-5% Risk-Squamous cell Carcinoma
  - 75% Adjacent Lichen Sclerosis

Saravanamuthu J et al Gyn Oncol 2003;89:251

**PSORIASIS**
- Incidence 1-3% Population
- Treatment
  - Domeboro’s Solution
  - Low dose Glucocorticoids
  - Clobetasol 0.05%-Short course
  - Systemic steroids-Refractory Cases
  - Methotrexate
### MOLLUSCUM CONTAGIOSUM

- **Etiology:** Molluscum-poxvirus
- **Inguinal Region & Gluteal Cleft**
- Many Spontaneously Resolve 6-12 mo.
- Observe Unless Not Resolving
  - Spread with Shaving
  - Tretinoin
  - Cidofovir (Topical anti-viral)
  - Imiquimod
  - Curettage

### HERPES SIMPLEX VIRUS

- In Pediatric Population 6% Genital
  - Painful Vesicular Lesions-Ulcerate-Inguinal Adenopathy-Systemic Signs & Symptoms
  - HSV-1 and 2 Usually Gingivitis
  - Always Look for Multiple Sites
  - DDx: Varicella-Herpes Zoster-Impetigo
  - Tx: Acyclovir-Competitive Inhibitor of Viral DNA polymerase-Inhibits DNA synthesis

PINWORMS

- *Enterobius vermicularis*
- Fecal bacteria Carry Pinworms-Vulvitis
- Flashlight Exam Evening vs. “Scotch Tape Test”
- Rx: Mebedazole 100mg repeat in 2 weeks
- Family Members Require Treatment
  - Except if PREGNANT
- Ultrasound
- Little if any Role for Mammography
- Monitoring 1-3 cycles
- FNA
- Malignancy 0.2% of Carcinoma-Breast < 25 y/a
  - Incidence 0.1/1,000,000 per year
- BSE Instruction

Simmons P in Pediatric & Adolescent Gynecology ed. Sanfilippo 2001 Saunders
ARE YOU UP TO DATE ON PCOS?

In the Adolescent?

WHAT DO YOU NEED TO MAKE THE DIAGNOSIS?

- REVISED DIAGNOSTIC CRITERIA-PCOS 2003 CRITERIA
  - Oligo-anovulation
  - Clinical or Biochemical Signs of Hyperandrogenism
  - Polycystic Ovaries
  - And Exclusion of Other Etiologies (CAH, Cushing's Syndrome)

Rotterdam ESHRE/ASRM PCOS Consensus Workshop Group
Fertil Steril 2004'81:19

PCOS-ULTRASOUND

- By Definition:
  - 12 or more Follicles @ 2 mm in diameter
  - Ovary increased
Minimally Invasive Surgery in Children and Adolescents

Robert K. Zurawin, MD
Associate Professor
Director Minimally Invasive Gynecologic Surgery
Baylor College of Medicine
Houston, Texas

History of Pediatric/Adolescent Gynecologic Surgery

- General Surgeons
- Gynecologists
- Pediatric Surgeons and Urologists
- Pediatric and Adolescent Gynecologists

Adoption of Minimally Invasive Surgery

CREOG Objectives

- Pediatric and Adolescent Gynecology
  - “Understand” the medical and surgical treatment of pediatric gynecologic disorders
  - “Describe” appropriate medical and surgical treatments for patients with developmental anomalies
  - “ Treat” adolescent gynecologic disorders medically or surgically

Disclosure

- Consultant: Ethicon Endo-Surgery, Ethicon Women’s Health & Urology, Conceptus Incorporated, CONMED Corporation, UpToDate

CREOG Objectives

<table>
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<tr>
<th>Procedure</th>
<th>Understand</th>
<th>Understand and Perform</th>
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<td>X</td>
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<tr>
<td>Laparoscopy, diagnostic and/or operative</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Lysis of adhesions laparoscopic</td>
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</table>


Point to Remember

In terms of surgery, especially endoscopic surgery:

Children are not "little adults"

Fundamentals

- Optimal surgical outcome depends on the surgeon’s knowledge of
  - Anatomy
    - Intimate, "autonomic" familiarity of pathologic conditions and relevant anatomic structures
  - Technology
    - TOTAL understanding of the surgical instruments
    - Electromechanical principles
    - Troubleshooting ANY malfunction
  - Technique
    - Tissue handling
    - Visual and proprioceptive coordination

Peritoneal Entry

- The sine qua non of laparoscopy
- If you can’t safely enter the peritoneum, you can’t do ANYTHING
- If you can safely enter the peritoneum, you can do EVERYTHING

Extent of the Problem

- ~ 4 million laparoscopies per year in the U.S.
- 0.5 - 3 percent of laparoscopic procedures have complications related to peritoneal entry
- Number of complications = ~ 60,000 per year

Challenges to Peritoneal Access
- Childhood Obesity
- Previous abdominal surgery
  - Previous pediatric surgery
  - Previous laparoscopy!!
  Adhesions to the umbilical undersurface occur in 21.2% of adult patients who have undergone a prior laparoscopy through an umbilical incision; 10.8% in children


Decision Tree
- Umbilicus or Alternative Site?
  - Elevate abdominal wall?
    - Hand elevation or towel clips?
  - Veress needle or Direct Trocar Entry?
  - Bladed or Bladeless Trocar?
  - Optical trocar or non-optical trocar?

Consensus Guidelines
- Middlesbrough Consensus - International Collaborative Group met in 1999
  - Basic guidelines are still followed today
- Council of the Society of Obstetricians and Gynaecologists of Canada

1 Gynaecological Endoscopy. Volume 8, Issue 6 (p 403-406)

Umbilicus
- Thinnest point on abdominal wall
- Overlies vital bowel and vascular structures
- Frequent site of umbilical hernias with hernia sacs and/or bowel contents
- Adhesions from prior surgery

Safe Veress Needle Entry
- Comparison of elevation of the abdominal wall
  - Hand elevation
  - Towel clips placed 2 cm on either side of umbilicus
  - Towel clips placed at the edge of the umbilicus

<table>
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<th>Technique</th>
<th>Introumbilical</th>
<th>2 cm Lateral</th>
<th>Hand Elevation</th>
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<tr>
<td>Mean distance (cm)</td>
<td>6.8</td>
<td>5.14</td>
<td>3.5</td>
</tr>
<tr>
<td>P-value</td>
<td>P&lt;0.01</td>
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Alternative Sites of Insufflation

- Transuterine
- Cul-de-sac
- Left upper quadrant

Primary Port Placement

Location of deep and superficial vessels of the anterior abdominal wall. Blue circles indicate recommended locations for trocar placement.

Anterior Abdominal Wall

Palmer’s Point


Basic Trocars

Optical Entry

Open entry

Critical principles:
- Force of entry into abdominal wall
  - 4-6 kg in reusable trocars
  - 2-3 kg in shielded bladed retractable trocars and bladeless trocars
- Insufflate to desired Pressure NOT Volume
  - Increase to 20 – 25 mm Hg until all ports are placed, then reduce to 15 mm Hg

You’ve picked the site – now what?

Optical Trocar Entry

Secondary Port Placement

- ALWAYS place secondary ports under direct visual guidance
- Use least amount of force, smallest diameter, and least traumatic puncture
- Avoid critical structures in anterior abdominal wall

Combined View

References:
**Operative Procedures**

- Open laparotomy
- Minimally invasive procedures
  - Laparoscopy
  - Hysteroscopy
  - Vaginoscopy

**Gynecological Operations**

- Congenital Abnormalities
- Foreign body
- Trauma
- Ovarian cysts
- Pelvic Pain/Endometriosis
- Malignancies
- Pelvic Inflammatory Disease
- Ectopic pregnancies

**Gynecological Operations**

- Congenital Abnormalities
- Septae
- Duplications and defects of fusion
- Dysgenetic ovaries

**Bermuda Triangle**

- Inferior Epigastric Vessels
- Medial Umbilical Ligament
- Round Ligament

Slide courtesy of Andrew I. Brill, MD
Septate Uterus

- Historical repair
  - Strassman procedure
  - Tompkins metroplasty

- Hysteroscopic management
  - Blind division with scissors
  - Lasers
  - Monopolar cautery in hypotonic solution
  - Bipolar cautery in normal saline

Duplications

- Didelphys
- Defects of fusion
  - lateral
  - vertical
- Rudimentary horns
  - communicating
  - noncommunicating

Uterus Didelphys
Didelphys with Obstructed Hemivagina

OHVIRA

OHVIRA

Didelphys and Cervical Agenesis

OHVIRA
Principles of Resection

- Preoperative radiologic evaluation
- Laparoscopy/hysteroscopy/vaginoscopy
- Adequate dissection to isolate blood supply
- Midline plane
- Proper instrumentation to insure minimal collateral tissue injury
- Port placement and number

Meyer-Rokitansky (MRKH)

- Vaginal agenesis
- Variable development of internal genitalia
- Problems if viable endometrium – obstruction
- MRI insufficient for diagnosis – need laparoscopy
Dysgenetic Ovaries

- Turner’s syndrome/mosaic
- Any Y-chromosome

Principles of Excision

- Streak ovaries can be very attenuated
- Endo-loop usually not practical – streak ovaries are not pedunculated
- Proximity to pelvic sidewall and ureter requires careful avoidance of collateral injury during dissection
- Attention to hemostasis
Gynecological Operations

- Congenital Abnormalities
- Foreign body
- Trauma
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- Malignancies
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- Pelvic Inflammatory Disease
- Ectopic pregnancies

Ovarian Cysts
- Functional
- Hemorrhagic Corpus Luteum
- Non-functional
  - Benign
  - Malignant

Functional Cyst

Torsion
**Dermoids**

- Tendency to leak, especially if thin, attenuated cyst wall
- Copious irrigation
- Watch for bilaterality
- Negligible risk of complications if spill occurs

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**The Problem**

- The vast majority of adnexal masses are benign
- The vast majority of adnexal masses treated by gynecologists result in preservation of the ovary
- The vast majority of ovarian masses treated by pediatric surgeons end up with salpingo-oophorectomy

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**Gynecological Operations**

- Congenital Abnormalities
- Foreign body
- Trauma
- Ovarian cysts
- Pelvic Pain/Endometriosis
- Malignancies
- Pelvic Inflammatory Disease
- Ectopic pregnancies
Principles of Adhesiolysis

- Careful attention to anatomy, especially ureters and great vessels
- Minimum thermal energy
- Consider adhesion prevention barrier

Laparoscopic Appearance

- Implants seen in adolescents are not typical of what is seen in adults
- Adolescents have clear vesicles, white implants, small hemorrhagic or petechial spots of the pelvic peritoneum
- Endometriosis found microscopically on biopsy of normal appearing peritoneum in 6% of patients (Nisolle FertilSteril 1990;53:984)
Cul de sac

Bullous lesion

Uterosacral ligament
Peritoneal window

Visible Endometriosis
Peritoneal Surface
Peritoneal
Gynecological Operations

- Congenital Abnormalities
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Sexually Transmitted Disease
- Persistent vaginal discharge
- Absence of foreign body
- Inconsistent history of sexual abuse
- Often negative cultures in ER or referring physician’s office
- Look for trauma to hymen, fourchette, but may be absent

Gynecological Operations
- Congenital Abnormalities
- Foreign body
- Trauma
- Ovarian cysts
- Pelvic Pain/Endometriosis
- Malignancies
- Pelvic Inflammatory Disease
- Ectopic pregnancies
Remember

Children are not “little adults”
They require special techniques and instrumentation

Laparoscopic Equipment
- Never need more than 5 mm scope
- 3 mm and 5 mm ports
- Special insufflation requirements in children less than 6 years old
- Consider equipment for heating and humidifying insufflated environment
- Adhesion prevention after ALL non-infected procedures

Emergency Situations
- Ectopic pregnancy
- Pelvic inflammatory disease
- Uncontrolled menorrhagia
- Undiagnosed vaginal bleeding
  - Sexual abuse
  - Foreign body

Objective
Maintain Reproduction Function

Exception
- Associated anomalies or medical conditions that prohibit fertility
  - Congenital heart disease
  - Profound retardation
  - End-stage renal disease
  - Acute life-threatening medical conditions

Emergency Endometrial Ablation
- Failure of hormonal therapy
- Unstable condition preventing emergency hysterectomy
- Failure of balloon compression
The Use of Thermal Balloon Ablation

- 3 pediatric/adolescent patients
- Medical conditions:
  - Sepsis
  - Uncontrolled bleeding in a Jehovah’s Witness
  - Diffuse pulmonary arterial stenosis


Summary

- Minimally invasive surgical techniques are within the grasp of all pediatric and adolescent gynecologists
- It is not enough to have the proper instrumentation available. You must be comfortable with the use of all equipment
- Competent surgical team
- Adequate visualization
- KNOW YOUR ANATOMY AND EMBRYOLOGY
OBSTRUCTIVE MÜLLERIAN ANOMALIES

Heather Appelbaum, MD
Chief, Division of Pediatric and Adolescent Gynecology
Director, Disorders of Sex Development Program
The Steven and Alexandra Cohen Children’s Medical Center
Associate Professor, Obstetrics and Gynecology
Hofstra Northshore LIJ School of Medicine

Objective

1. Define normal and abnormal embryologic development of the reproductive structures
2. Identify the level of obstruction for different Müllerian anomalies
3. Apply appropriate diagnostic and therapeutic strategies for treating hematocolpos

Developmental embryology of the reproductive tract

- Gonads
- Paramesonephros
- Mesonephros
- Metanephros
- Urogenital sinus
- Sinovaginal bulb

Genital Tract Development

- Initially, male and female embryos have both mesonephric and paramesonephric ducts.
- The paired mesonephric ducts connect the mesonephros kidneys to the cloaca.

Müllerian Duct Development

- 6 weeks – 1st identifiable when they elongate caudally and cross the metanephric ducts medially to meet in the midline.
- 7 weeks – The urorectal septum develops and separates the rectum from the urogenital sinus.
- 12 weeks – The caudal portion of the ducts fuse to form the urovesical canal which inserts into the urogenital sinus.

Disclosure

I have no financial relationships to disclose.
**Müllerian Duct Development**
- The 2 müllerian ducts are initially composed of solid tissue and lie side by side.
- Internal canalization of each duct produces 2 channels divided by a septum that is resorbed in a cephalad direction by 20 weeks.
- The cranial, unfused portions develop into the fimbria and fallopian tubes.
- The caudal, fused portions form the uterus and upper vagina.

**Function of the Müllarian tract**
1. Menstrual egress
2. Sexual intercourse
3. Fertility
4. Pregnancy
5. Delivery

**Non Obstructive Müllerian Anomalies**
- Septated, cribiform, or microperforate hymen
- Persistent urogenital sinus
- Longitudinal vaginal septum/duplicated vagina
- Uterine didelphys
- Bicornuate uterus
- Uterine septum
- Unicornuate uterus
- Müllerian agenesis

**OBSTRUCTED MÜLLERIAN ANOMALIES**

**Obstructed Müllerian Anomalies**
- Rudimentary horns
- OHVIRA
- Segmental vaginal agenesis
- Transverse vaginal septum
- Cervical atresia or cervical agenesis

**Obstruction to menstrual egress**
- Pelvic pain
- Hematocolpos/Hematometria/Hematosalpinx
- Pyometria/pyosalpinx
- Endometriosis
Clinical Presentation

• Pelvic pain
• Dysmenorrhea
• Abnormal bleeding
• Pelvic mass
• Pregnancy complications

Adolescent with pelvic pain and pelvic mass

Think Obstructed Müllerian Anomaly

Imperforate hymen

- Hydrocolpos/mucocele at birth
- Hematocolpos at puberty
- Untreated imperforate hymen can cause retrograde menstrual flow resulting in endometriosis
- Hymenectomy should be delayed until puberty

Hymenectomy

- Characterizes the number and nature of the Müllerian structures
- Identifies cervical and vaginal anatomy
- Degree of uterine fusion in duplicated systems is delineated
- Associated urinary malformation are identified

Preoperative assessment of obstructed Müllerian anomaly by T2 weighted MRI
**Rudimentary uterine horns**

- The prevalence of a rudimentary uterine horn is 1/100,000
- 48% of rudimentary horns do not have a cavity
- Most cavitary rudimentary horns do not have functional endometrium (Fedele, 1990)

**Non-communicating rudimentary horns**

- 7-10% functional
- MRI or pelvic ultrasound identifies endometrial stipe or hematometria
- Risk of ectopic pregnancy
- Laparoscopic removal of obstructed non-communicating functional horn
- Nonfunctional uterine horns do not require surgical intervention

**Reproductive outcomes with a functional non-communicating rudimentary uterine horn**

- Transperitoneal migration of a sperm or fertilized ovum can result in ectopic pregnancy
- Uterine rupture between 10-20 weeks of gestation (Tufail, 2007)
- Functional cavitated rudimentary horns have a higher ectopic and lower live birth rate than those with a rudimentary horn and no cavity (Heinonen, 1997)

**Complications associated with functional rudimentary uterine horns**

- Retrograde menstruation
- Hematosalpinx
- Pelvic abcess
- Endometriosis
- Hematometria
- Ectopic pregnancy

**Case reports of ectopic pregnancies in rudimentary uterine horns**

- Ectopic pregnancy in a non-communicating tube of a rudimentary horn (Pokoly, 1989)
- Ruptured tubal pregnancy on the same side as the rudimentary horn (Handa, 1999)
- Tubal or rudimentary horn pregnancies were found only in women with unicorunate uterus and rudimentary horn with a cavity (Heinonen, 1997)
Laparoscopic resection of rudimentary horns resulted in successful pregnancies in infertile patients with unicornuate uteri with non-communicating rudimentary horns (Giatras, 1997).

Removal of the rudimentary horn may enhance reproductive performance of the unicornuate uterus (Fedele, 1987).

Removal of the rudimentary horn may enhance reproductive performance of the unicornuate uterus (Fedele, 1987).

Uterine didelphys
Obstructed hemivagina
Ipsilateral renal agenesis

Uterine didelphys
Obstructed hemivagina
Ipsilateral renal agenesis

Uterine didelphys
Obstructing vaginal septum
Unilateral cyclic or constant pelvic pain with normal menses
MRI or pelvic ultrasound identifies hematometrocolpos and normal uterus

Uterine didelphys
Obstructed hemivuterus resulting in hematometrocolpos
Distal hemivaginal agenesis/non-communicating longitudinal vaginal septum/unilateral imperforate vagina

Preoperative schematic

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA)

OHVIRA

What should you do?

Do
- Suppress menses
- Provide analgesia
- Decompress the bladder for urinary retention
- Refer to specialist for surgical intervention

Don’t
- Perforate the dilated structure
- Attempt drainage
- Operate before the anatomy is clearly defined
Transvaginal excision of vaginal septum
- Semilunar convex incision made in most distal aspect left anterolateral wall of the hemivagina
- Septum excised with needle point cautery
- Sequential reapproximation of vaginal mucosa with 3.0 vicryl to the level of 0.5cm from the cervices
- Diagnostic laparoscopy not necessary

Segmental vaginal agenesis
- Greater than 1 cm distance between the upper upper and lower vaginal tract
- Transvaginal surgical repair
  - Segmental graft
  - Vaginal stenting
  - Vaginal pull-through
  - Serial dilation combined with other techniques

Vaginal atresia/segmental vaginal agenesis
- Vertical fusion/obstructive defect
- Urogenital sinus fails to develop the lower vagina
- Mullerian structures remain normal
- Presents with primary amenorrhea and cyclic or constant pelvic pain
- Ultrasound shows hematometra or hematocolpos
- MRI can delineate the thickness of the segment and confirm the presence of a cervix

Surgical technique
- Hematocolpos acts as tissue expander
- Preop mechanical dilation can decrease the thickness of the segment
- Crescentic incision at the vaginal dimple
- Probes, dilators, transrectal ultrasound guide dissection
- Once vaginal mucosa identified, vaginal pull through to the introitus or interpose graft

Transverse vaginal septum
- Transvaginal resection with reapproximation of upper and lower vaginal mucosa
- Preop mechanical dilation to thin the septum
- Z plasty may minimize post operative stenosis

Surgical Technique
- Preoperative imaging is essential to rule out complex defects
- Thin septae should be resected followed by a primary end-to-end anastomosis of the upper and lower vaginas
- Thick septae should be resected with a pull through and circumferential Z-plasty reconstruction technique or with a graft
- Distension of upper vagina with menstrual blood or preoperative dilation of the lower vagina may decrease the thickness of the septum

Cervical agenesis

Middle Vagina 40%  
Upper Vagina 46%  
Lower Vagina 14%  

43
**Uterovaginal procedures for cervical atresia**

- Utero-vaginal Canalization
  - Transvaginal or Transabdominal Approach
  - Create Ostium From Uterus to Vagina
  - Stent with/without Grafting
  - Endocervical Gland Presence—Better Prognosis
  - Restenosis Rate High: 40-60%
  - Risk Infection—Peritonitis-Sepsis-Death
  - Fertility Success Remains Low

  Fujimoto V et al AJOG 1997;177(6):1419

**Fertility with cervical atresia**

- Overall-low fertility
  - High incidence of endometriosis-adhesive disease
- Pregnancy case reports
  - IVF-Transmyometrial Embryo Transfer
  - GIFT
- Gestational Carrier
  - Spont. Preg following Uterovaginal Anastomosis-Graft Reconstruction

**Management of congenital absence of cervix**

- 8 patients with cervical atresia
- Cervical-vaginal fistula created
- No Pregnancies
- Hysterectomy is the treatment of choice


**MANAGEMENT OF CONGENITAL ABSENCE OF CERVIX**

- Overall Incidence Uterine Anomalies 0.5% Deliveries
- 8-Cervical Atresia-Pelvic Pain
- Cervical-Vaginal Fistula Created
- No Pregnancies
- Hysterectomy Treatment of Choice


**UTERO-VAGINAL PROCEDURES**

- Utero-vaginal Canalization
  - Transvaginal or Transabdominal Approach
  - Create Ostium From Uterus to Vagina
  - Stent with/without Grafting
  - Endocervical Gland Presence—Better Prognosis
  - Restenosis Rate High: 40-60%
  - Risk Infection—Peritonitis-Sepsis-Death
  - Fertility Success Remains Low

  Fujimoto V et al AJOG 1997;177(6):1419

**FERTILITY CONSIDERATIONS**

- Overall-Low Fertility
  - Endometriosis-Adhesive Disease
- Case Reports Pregnancy
  - IVF-Transmyometrial Embryo Transfer
  - GIFT
- Gestational Carrier
  - Spont. Preg following Uterovaginal Anastomosis-Graft Reconstruction
Endometriosis

- Retrograde menstrual flow with obstructive anomalies
- Early restoration of outflow tract can limit endometriosis
- Cecal metaplasia may play an additional role

Complex Müllerian Anomalies

key points

- Hemi-hysterectomy is recommended for a cavitated functional rudimentary horn
- No conclusive evidence exists to warrant excision of a non-cavitated rudimentary horn
- The level of the obstruction should dictate the operative approach

Obstructed Müllerian Anomalies

key points

- Obstructive congenital anomalies of the vagina should be approached transvaginally by septal resection or vaginoplasty
- Hysterectomy is the treatment of choice for cervical agenesis
- Hemi-hysterectomy is recommended for a cavitated functional rudimentary horn
- No conclusive evidence exists to warrant excision of a non-cavitated rudimentary horn

Complex Müllerian Anomalies

key points

- Appropriate imaging is essential to assess the level of obstruction
- The level of the obstruction should dictate the operative approach
- Laparoscopic hysterectomy is recommended for a cavitated functional rudimentary horn, and may be indicated for cervical agenesis
- No conclusive evidence exists to warrant excision of a non-cavitated rudimentary horn
**Disorders of Sexual Development**

Robert K. Zurawin, MD  
Associate Professor  
Director, Minimally Invasive Gynecologic Surgery  
Department of Obstetrics and Gynecology  
Baylor College of Medicine  
Houston, Texas

**Vulva**
- Classical Latin – *vulva* was “the womb” - Celsus, *De medicina*, IV, 1.2
- Sanskrit – *ulva*

**Vagina**
- Latin – “the sheath of a sword” or “scabbard” - Julius Caesar, *Gallic Wars* V, 44.8
- Roman farming – “leaf sheath of an ear of wheat”

**Disclosure**
- Consultant: Ethicon Endo-Surgery, Ethicon Women’s Health & Urology, Conceptus Incorporated, CONMED Corporation, UpToDate
Developmental Abnormalities of External Genitalia
- Clitoris – hypertrophy
- Vulva
  - Congenital labial fusion
  - Acquired labial agglutination
  - Hypertrophy of labia minora and majora
  - Prolapse of urethral mucosa
  - Hemangioma

Genetic-Chromosomal Abnormalities
- True hermaphroditism
- Female pseudohermaphroditism
- Male pseudohermaphroditism
- Androgen insensitivity (testicular feminization)
- Mixed gonadal dysgenesis
- Chromosomal abnormalities with vulvovaginal anomalies

Developmental Abnormalities of Internal Genitalia
- Vagina
  - Imperforate hymen/microperforate hymen
  - Transverse vaginal septum
  - Duplication
  - Agenesis
  - Wolffian duct remnants
    - Gartner’s duct cyst
    - Herlyn-Werner syndrome

External Genitalia
- Ambiguous Genitalia
  - Female pseudohermaphrodites
  - Androgen abnormality/insensitivity
- Labial Hypertrophy
- Female Genital Mutilation
  - Type I Clitoridectomy
  - Type II Clitoridectomy and labial excision
  - Type III Modified infibulation
  - Type IV Total infibulation

Clitoral hypertrophy

Clitoral hypertrophy
Labia

- Two of them are “labia majora”
  - One of them is:
    - Labium majus

- Two of them are “labia minora”
  - One of them is:
    - Labium minus

Congenital Labial Fusion

Repair of Labial Fusion

Repair of Labial Fusion

Repair of Labial Fusion

Labial Agglutination
Type I — Partial or total removal of the clitoris and/or the prepuce (clitoridectomy). When it is important to distinguish between the major variations of Type I mutilation, the following subdivisions are proposed: Type Ia, removal of the clitoral hood or prepuce only; Type Ib, removal of the clitoris with the prepuce.

Type II — Partial or total removal of the clitoris and the labia minora, with or without excision of the labia majora (excision). When it is important to distinguish between the major variations that have been documented, the following subdivisions are proposed: Type IIa, removal of the labia minora only; Type IIb, partial or total removal of the clitoris, the labia minora and the labia majora.

Note also that, in French, the term ‘excision’ is often used as a general term covering all types of female genital mutilation.

Type III — Narrowing of the vaginal orifice with creation of a covering seal by cutting and appositioning the labia minora and/or the labia majora, with or without excision of the clitoris (infibulation). Type IIIa, removal and apposition of the labia minora; Type IIIb, removal and apposition of the labia majora.

Type IV — All other harmful procedures to the female genitalia for non-medical purposes, for example: piercing, piercing, incising, scraping and cauterization.
Masses

- Ectopic ureter
- Prolapsed ureterocele
- Bladder extrophy
- Vaginal cyst
- Obstructed hemivagina
- Cloacal and urogenital sinus abnormalities

Introital Abnormalities

Urethral Prolapse

Urethral Prolapse Repair
Urethral Prolapse Repair

Anomalies Associated with Vulvovaginal Abnormalities
- Urinary tract
  - Unilateral renal agenesis with obstructed hemivagina
  - Ectopic or duplicated ureter
  - Exstrophy of the bladder
- Intestinal tract
  - Ectopic or imperforate anus
  - Rectovaginal fistula

Developmental Abnormalities of Internal Genitalia
- Vagina
  - Imperforate hymen/microperforate hymen
  - Transverse vaginal septum
  - Duplication
  - Agenesis
  - Wolffian duct remnants
    - Garnder’s duct cyst
    - Herlyn-Werner syndrome
Hymen

- Greek – ἑμήν – “the thin skin or membrane covering the brain and heart” - Aristotle
- hymenoptera ~ “membrane wing” wasps
- Hymen was the god of marriages
- The Greek wedding song was a “hymenaios”

Septate Hymen

Imperforate Hymen

Septate Hymen

Microperforate Hymen

Imperforate Hymen
Disorders of:
- Agenesis
- Hypoplasia
- Vertical fusion (canalization abnormalities resulting from abnormal contact with the urogenital sinus)
- Lateral fusion (duplications)
- Resorption (septa)

Anomalies of the Uterus and Vagina

![Imperforate Hymen](image)

![Anomalies of the Uterus and Vagina](image)

![Transverse Vaginal Septum](image)
**Disorders of Mesonephric Remnants**
- Hydatid of Morgagni cyst
- Cysts of the broad ligament
- Gartner’s canal (duct)

**Wolffian remnants**

**Vaginal Agenesis**

**Creation of Neo-Vagina**
- Frank dilator therapy
- McIndoe
  - “Traditional” skin graft procedure
- Vecchietti
  - Traction using flexible dilator placed at introitus
- Davydov
  - Combined vaginal and laparoscopic approach
**Septae**
- Duplications and defects of fusion
- Dysgenetic ovaries

**Septate Uterus**
- Historical repair
  - Strassman procedure
  - Tompkins metroplasty
- Hysteroscopic management
  - Blind division with scissors
  - Lasers
  - Monopolar cautery in hypotonic solution
  - Bipolar cautery in normal saline

**Duplications**
- Didelphys
- Defects of fusion
  - lateral
  - vertical
- Rudimentary horns
  - communicating
  - noncommunicating
Preoperative radiologic evaluation
Laparoscopy/hysteroscopy/vaginoscopy
Adequate dissection to isolate blood supply
Midline plane
Proper instrumentation to insure minimal collateral tissue injury
Port placement and number

Principles of Resection

Thank you very much!
Minimally Invasive Surgery for Adnexal Masses and Adnexal Torsion in Children: a conservative approach

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Chief, Division of Pediatric and Adolescent Gynecology
Department of Obstetrics and Gynecology
Long Island Jewish Medical Center
Steven and Alexandra Cohen Children’s Medical Center

Disclosure

I have no financial relationships to disclose.

Objectives

- Review the preoperative assessment of adnexal masses in children and adolescents
- Assess when ovarian preservation is indicated in children with adnexal masses
- Review minimally invasive surgical techniques for approaching large adnexal masses in children
- Diagnose, treat and prevent adnexal torsion in children and adolescents.

Adnexal masses

<table>
<thead>
<tr>
<th>Physiologic</th>
<th>Benign Neoplasm</th>
<th>Malignant Neoplasm</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>Follicular cyst</td>
<td>Mature teratoma</td>
<td>Immature teratoma</td>
<td>Ectopic pregnancy</td>
</tr>
<tr>
<td>Corpus Luteal cyst</td>
<td>endometrioma</td>
<td>Dysgerminoma</td>
<td>Obstructive Mullerian anomaly</td>
</tr>
<tr>
<td>Ruptured ovarian cyst</td>
<td>Serous cystadenoma</td>
<td>Granulosa cell tumor</td>
<td>Paraovarian cyst</td>
</tr>
<tr>
<td></td>
<td>Mucinous cystadenoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lymphoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Torsion</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tubo-ovarian abscess</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hematosalpinx</td>
<td></td>
</tr>
</tbody>
</table>

Incidence of ovarian masses by cell type

<table>
<thead>
<tr>
<th>Non neoplastic</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>ovarian torsion</td>
<td>26.5%</td>
<td>46.5%</td>
<td></td>
</tr>
<tr>
<td>corpus luteal cyst</td>
<td>14.2%</td>
<td>14.2%</td>
<td></td>
</tr>
<tr>
<td>paraovarian cyst</td>
<td>15.4%</td>
<td>15.4%</td>
<td></td>
</tr>
<tr>
<td>hematopyosalpinx</td>
<td>1.9%</td>
<td>1.9%</td>
<td></td>
</tr>
<tr>
<td>ovolitis</td>
<td>0.9%</td>
<td>0.9%</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Benign neoplastic</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>mature teratoma (dermoid)</td>
<td>44.3%</td>
<td>36.8%</td>
<td></td>
</tr>
<tr>
<td>cystadenoma</td>
<td>31.8%</td>
<td>29.8%</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Malignant neoplastic</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>dysgerminoma</td>
<td>0.8%</td>
<td>0.8%</td>
<td></td>
</tr>
<tr>
<td>immature teratoma</td>
<td>1.9%</td>
<td>1.9%</td>
<td></td>
</tr>
<tr>
<td>yolk sac tumor</td>
<td>0.9%</td>
<td>0.9%</td>
<td></td>
</tr>
<tr>
<td>granulosa cell tumor</td>
<td>1.9%</td>
<td>1.9%</td>
<td></td>
</tr>
<tr>
<td>serous-Leydig cell tumor</td>
<td>0.9%</td>
<td>0.9%</td>
<td></td>
</tr>
</tbody>
</table>

Prenatal/neonatal ovarian cysts

- 1:2,500 live female births
- Follicular cysts develop in response to maternal hormones
- Differential diagnosis includes congenital urogenital anomalies, mesenteric or omental cysts, volvulus, intestinal duplication or urachal cysts
- 90% spontaneous resolution by three months
Complications associated with prenatal/neonatal cysts
- Hemorrhage
- Rupture
- Torsion and necrosis
- Incarceration in inguinal hernia
- Respiratory distress
- Labor dystocia

Management of prenatal/neonatal ovarian cysts
- Serial ultrasounds at birth and q4-6weeks
- Spontaneous regression occurs in most cases
- Surgical intervention for persistent cysts, symptomatic cysts, cysts increasing in size, or cysts >5cm with ovarian preservation

Ovarian cysts in infants/prepubertal girls
- Rare phenomenon
- Hormonally active cysts can cause precocious puberty
- 26% asymptomatic abdominal masses are malignant
- <1% cysts with abdominal pain are malignant

Management of ovarian cysts in prepubertal girls
- Serial ultrasounds
- Hormonal evaluation
- 90% resolve spontaneously
- Surgical intervention for persistent, large, or symptomatic cysts

Conservative management for benign ovarian cysts in adolescents
- Serial ultrasounds
- Analgesics
- Serial Hct
- Hormonal therapy

Management of postpubertal/adolescent ovarian cysts
- Follicular cysts are <3cm simple cysts and will invariably self resolve
- Functional cysts <6-10cm should be managed conservatively unless symptomatic
- Laparoscopic cystectomy for persistent, large, or symptomatic cysts
- Hormonal therapy will suppress further cyst formation
Complications from ovarian cysts

- Torsion
- Rupture
- Hemorrhage
- Urinary tract obstruction
- Aorto-caval shunting
- Incarceration of inguinal hernia
- Respiratory distress

Case #1

L.H. is a 10y2m old with intermittent right lower quadrant pain of variable intensity over the last 15 hours.

Prior to coming to the ED, her pain was severe and she was nauseated.

On arrival to the ED her pain had improved and she felt much better.

She was afebrile, with normal vital signs.

Tanner staging B1PH1

Abdomen was soft with tenderness in the right lower quadrant. There was no rebound or guarding. There were no palpable masses.

Pelvic examination was not performed.

Transabdominal pelvic ultrasound

- Right ovary 6.4 X 4.8 X 4.0 cm with a 4.2cm simple cyst
- Left ovary 1.4 X 1.7 X 1.3cm
- Prepubertal uterus

What is her diagnosis?

Ovarian Torsion

Twisting of the ovary on its ligamentous supports results in impedance of blood supply.
Factors predisposing to adnexal torsion in children

- Ovarian masses
- Persistent neonatal ovarian cyst
- Polycystic ovaries
- Müllerian anomalies

Ovarian and fallopian tubal torsion

- Elongated utero-ovarian ligament in prepubertal girls
- May be associated with strenuous exercise or sudden increase in abdominal pressure
- Neonates present with abdominal mass, feeding intolerance, vomiting, abdominal distension and irritability

Incidence and Trends of Pediatric Ovarian Torsion Hospitalizations in the US, 2000-2006

- Incidence of ovarian torsion in age group 1-20 yo is 4.9/100,000
- 58% of cases of ovarian torsion in children are associated with benign masses
- Less than 0.5% of ovarian torsion cases were associated with malignant neoplasm
- There were no cases of venous thromboembolism

Symptoms associated with ovarian torsion

- Stabbing pain (70%)
- Nausea and vomiting (70%)
- Sudden and sharp pain in the lower abdomen (59%)
- Pain radiating to back, flank, or groin (51%)
- Peritoneal signs (3%)
- Fever (<2%)

The role of ultrasound in ovarian torsion

- Peripheral follicles with stromal edema
- Heterogenously enlarged ovaries
- Free fluid in the cul-de-sac
- A ratio of torsed adnexal volume to the normal adnexal volume greater than 20 is predictive of a mass inside the ovary
- Color flow Doppler can be appreciated in a torsed ovary

Loss of normal ovarian parenchyma
Enlarged ovary crosses the midline

Whirl pool sign

The role of inflammatory markers in ovarian torsion

Surgical management of ovarian torsion

- Diagnostic and therapeutic laparoscopy
- Exploratory laparotomy
- Ovarian preservation
- Detorsion of ovary
- Cystectomy
- Detorsion with second procedure cystectomy
- Cyst aspiration
- Ovarian bivalving
- Oophoropexy
- Oophorectomy

Assessing ovarian viability

- Color flow Doppler is not a reliable measure of ovarian viability
- Leukocytosis, fever, and signs of peritonitis may indicate irreversible damage to the ovary
- Macroscopic appearance is not a good indicator of viability

What should you do with the purple, black, and ugly ovary?

**Detorsion of the Ovary**

<table>
<thead>
<tr>
<th>Study</th>
<th>% recovery of ovarian function</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oelsner et al, (1993)</td>
<td>91% (85/92)</td>
</tr>
<tr>
<td>Mage et al., (1989)</td>
<td>94% (16/17)</td>
</tr>
<tr>
<td>Shalev et al. (1995)</td>
<td>94% (48/52)</td>
</tr>
<tr>
<td>Rody et al, (2002)</td>
<td>100% (4/1)</td>
</tr>
<tr>
<td>Aziz et al, (2004)</td>
<td>100% (14/14)</td>
</tr>
<tr>
<td>Celik et al, (2005)</td>
<td>92% (13/14)</td>
</tr>
<tr>
<td>Roussel et al.</td>
<td>100% (15/15)</td>
</tr>
<tr>
<td>Levy et al.</td>
<td>100% (3/3)</td>
</tr>
<tr>
<td>Parsky et al</td>
<td>88% (7/8)</td>
</tr>
<tr>
<td>Cohen et al (1996)</td>
<td>100% (7/7)</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td><strong>93% (211/227)</strong></td>
</tr>
</tbody>
</table>

**Case #1: Post operative ultrasound**

- 2.7 X 1.4 X 0.9 cm prepubertal uterus
- Left ovary is 2.6 X 1.5 X 1.4cm
- **Right ovary is 2.4 X 1.8 X 1.0cm**

**How long is too long to wait to detorse the ovary?**

- The adnexa of rats were twisted for 36 hours or until they became bluish-black in appearance
- All ovaries that were torsed under 24 hours showed no immediate or delayed evidence of necrosis on histologic evaluation
- Ovaries torsed for 36 hours showed immediate and delayed adnexal necrosis


**Ovarian torsion**

- Surgical emergency to preserve ovarian functioning
- Must have high clinical index of suspicion in patients with acute and/or intermittent, variable abdominal pain and nausea/vomiting

**Oophoropexy**

- Utero-ovarian ligament at the ovarian insertion is attached to the ipsilateral uterosacral ligament using permanent suture
- Plication of the utero-ovarian ligament
- Ovary can be sutured to the pelvic sidewall

**How do we prevent recurrent ovarian torsion?**
Oophoropexy

**Pros**
- May prevent recurrent torsion of a detorsed ovary
- May prevent torsion with polycystic ovaries
- Elongated utero-ovarian ligaments likely pose a higher risk of recurrence
- May be prophylactically useful for the contralateral ovary following unilateral oophorectomy

**Cons**
- Theoretical risk of impaired blood supply
- Theoretical risk of peritubal adhesions
- Insufficient data on future fertility functioning

Ovarian torsion key points

- Early intervention for ovarian torsion results in preservation of ovarian function, despite the gross appearance of the ovary
- Ultrasonographic appearance of the ovary is a useful tool for managing ovarian torsion, but color flow Doppler is not reliable
- Oophoropexy of the detorsed adnexa or the contralateral ovary may be appropriate

Complex ovarian masses

Case #2

- 8y3mo with incidental finding of non-tender abdominal mass on physical examination by pediatrician
- Ultrasound showed 11cm complex cystic ovarian mass
- MRI showed 10.1 X 8.4 x7.1 cm lobulated cystic mass with septations with a solid tubular component arising from the left ovary, a normal prepubertal right ovary measuring 1.9 x1.3 x 0.7cm and normal prepubertal uterus measuring 2.7 X 1.5 X 1.0cm, a small amount of free fluid in the peritoneal cavity
- Tumor markers (AFP, HCG, inhibin, LDH) normal

Factors affecting surgical approach: laparotomy vs. laparoscopy?

- Risk of malignancy
- Size and feasibility
- Spillage and adhesion prevention
- Recurrence
Factors affecting ovarian preservation vs. oophorectomy

- Risk of malignancy
- Ratio of volume of neoplasm to volume of normal ovarian tissue
- Bilaterality
- Risk of ipsilateral or contralateral recurrence

Clinical findings suggesting benign vs. malignant mass

**Benign**
- Unilateral
- Cystic
- Mobile
- Smooth
- No ascites
- Slow growing
- Age <35yo

**Malignant**
- Bilateral
- Solid
- Fixed
- Irregular
- Ascites
- Rapid growth
- Age >35yo
- Associated endocrinopathy

Ultrasonographic characteristics of the ovary

**Benign pattern**
- Simple cyst without internal echoes
- Simple cyst with scattered echoes
- Central dense round echoes
- Thin or thick multiple linear echoes

**Malignant pattern**
- Cystic echoes with papillary or indented mural parts
- Irregular thick septations and solid parts
- Heterogeneous component with irregular cystic part
- Completely solid with homogeneous component

Characteristics of benign tumors of the ovary

- Predominantly cystic
- <8cm tumor size
- Normal tumor markers

Risk of malignancy

- Masses > 8cm
- Elevated tumor markers
- Predominantly solid
- Precocious puberty
- Palpable mass

Decision tree for ovarian enlargement

- Ovarian enlargement
  - Ovarian mass <75mm
    - Predominantly cystic
    - Normal tumor markers
  - Ovarian mass >75mm
    - Predominantly solid
    - Elevated tumor markers

Predicting malignancy

- Age
- Masses > 8cm
- Elevated tumor markers
- Predominantly solid
- Precocious puberty
- Palpable mass

Recommendations from Children's Oncology Group

- Tumor markers should be evaluated preoperatively
- Surgical intervention should evaluate the extent of disease, maximize complete tumor resection, spare uninvolved reproductive organs
- Incomplete surgical staging is upgraded and chemotherapy is advised

Serum marker | Associated tumor
---|---
AFP | endodermal sinus, embryonal carcinoma, mixed germ cell
LDH | choriocarcinoma, embryonal carcinoma, mixed germ cell
HCG | dysgerminoma, mixed germ cell
CA125 | epithelial tumors
CEA | serous cystadenocarcinoma, mucinous cystadenocarcinoma
Inhibin | granulosa-theca cell tumor

Which complex ovarian masses require staging?

- Neoplastic ovarian masses in children have 10-20% risk of malignancy
- Tumor markers are positive in only 54% of children with an ovarian malignancy
- Ovarian masses greater than 8 cm
- Prepubertal age range

Staging requirements

- Pelvic washings
- Visual inspection of contralateral ovary, pelvic viscera, omentum, and peritoneal surface
- Palpation of lymph nodes
- Removal of intact specimen with clean margins

Risk of rupture according to surgical procedure and surgical approach

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Risk of Rupture</th>
</tr>
</thead>
<tbody>
<tr>
<td>Laparoscopy</td>
<td>92.8% P&lt;0.001</td>
</tr>
<tr>
<td>Laparotomy</td>
<td>36.8% P&lt;0.001</td>
</tr>
<tr>
<td>Cystectomy</td>
<td>92% P&lt;0.001</td>
</tr>
<tr>
<td>Oophorectomy</td>
<td>14% P&lt;0.001</td>
</tr>
</tbody>
</table>


Tumor spillage

- Spillage upstages a malignant tumor necessitating chemotherapy
- Adhesions
- Chemical peritonitis 0-0.2%
Outcomes for malignant tumors treated with ovarian salvation

- 2.6% recurrence for immature teratoma with ovarian preservation
- No recurrence 4.7y follow up after chemotherapy without oophorectomy for immature teratoma n=8
- Recurrence with oophorectomy plus chemotherapy
- 22% recurrence for borderline ovarian tumors treated with cystectomy only n=22


What is the result of ovarian preservation?

- 3-4% recurrence rate for mature teratomas and 2.6% for immature teratomas
- 0-18% recurrence rate for low malignant potential tumors
- 9.6%-14% recurrence in stage IA, grade 1-2
- 90-100% survival rate with chemotherapy +/- oophorectomy

Is there a safe compromise between laparoscopy and laparotomy?

Mini laparotomy with unilateral oophorectomy

Complex ovarian masses key points

- Preoperative risk assessment for malignancy will help determine surgical approach
- Children have a higher risk of ovarian malignancy than reproductive age women
- Negative tumor markers do not rule out malignancy
Endometriosis in Adolescents
A Whole Different Ball Game

Joseph S. Sanfilippo, MD, MBA
University of Pittsburgh
School of Medicine

Disclosure

I have no financial relationships to disclose.

Objectives

Upon Completion of this Lecture the Participant Will Understand:

- Endometriosis is a Premenarchal Disease
- Family History is Associated with Incidence of 30% vs. 7.6% in Adults
- Importance of Multisystem Evaluation with Chronic Pelvic Pain

ENDOMETRIOSIS FIRST REPORTED

- von Roikitansky 1860
- Sampson Variable Appearance Endometriotic Implants 1920
- Path Should be Obtained Endometrioma > 3cm.
- Peritoneal Lesions:
  - Papular or Vesicular with Serous or Hemorrhagic Content

THEORIES OF CAUSATION

- Retrograde Menstruation: “Sampson”
  - Coelomic Metaplasia
  - Lymphatic Metastasis
  - Vascular Metastasis
  - Iatrogenic Dissemination
- Cell-mediated or Immunologic Defects
  Gleisher N et al OG 1987:70:115
PATHOPHYSIOLOGY

- Defective Immune Surveillance
- "A Local Pelvic-Inflammatory Process"
- Peritoneal Macrophages—Their Secretory Products, Cytokines, Neovascularization
  - Cytokines: TNF alpha, Interleukins, Chemokines

PREMENARCHEAL

- Two Premenarcheal Girls 12 y/a & 13 y/a
- Retrospective Review 67 Adolescents
- Emory University 1992-1994
- Average Duration of Symptoms: 2.4 years
  - Laparoscopy or Laparotomy for Pelvic Pain
  - Majority Stage I (ASRM Class.)
    - "Superficial Red Lesions"
  - Stage III (6.1%) and Stage IV (2%) with NO Outflow Tract Obstruction
    - Age 12: Stage I No Mullerian Anomaly
    - Age 13: Cervical Dysgenesis Hematometra

Reese K et al J Pediatr Adolesc Gynecol 1996;9(3):125

Endometriosis in Premenarcheal Girls Without Uterine Anomalies

- Five premenarcheal girls with chronic (>6 mos) pelvic pain
- Negative gastrointestinal evaluation
- All had laparoscopic biopsy proven endometriosis and ablative treatment
- All had marked improvement in pain
- Two had repeat laparoscopy 6 and 8 years later for pathologically confirmed endometriosis


Incidence of Endometriosis in Adolescents

- 25–38% of adolescents with chronic pelvic pain
  - J Reprod Med 1989;34:827
  - Clin Exp Obstet Gynecol 1999;26:76
- 50-70% of adolescents with pelvic pain not controlled with OCPs and NSAIDs
**Age of First Pelvic Symptoms**


**Symptoms of Endometriosis in Adolescents (n=49)**
- Cyclic pain (67%)
- Noncyclic pain (39%)
- Dysmenorrhea (100%)
- Gastrointestinal symptoms (67%)
- Abdominal pain (58%)
- Referred pain (31%)


**What Are You Likely to Find in Adolescents?**
- Stage I or II
- "Atypical Red Lesions"

**Diagnosis: Standard Technique and Systematic Investigation**
- Panoramic view
- Vesicouterine peritoneum
- Anterior and posterior uterus
- Cul-de-sac and sigmoid
- Left ovarian fossa, ovary and tube
- Right ovarian fossa, ovary and tube
- Appendix and upper abdomen
### Appearance of Endometriotic Implants in Adolescents

<table>
<thead>
<tr>
<th>Lesion type</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Red</td>
<td>82</td>
</tr>
<tr>
<td>Pigmented (Black/Blue)</td>
<td>76</td>
</tr>
<tr>
<td>Vesicular (Clear)</td>
<td>41</td>
</tr>
<tr>
<td>White</td>
<td>6</td>
</tr>
<tr>
<td>Superficial</td>
<td>98</td>
</tr>
<tr>
<td>Peritoneal pocket</td>
<td>18</td>
</tr>
<tr>
<td>Deep</td>
<td>12</td>
</tr>
</tbody>
</table>


### Location of Superficial Endometriosis in Adolescents (N=36)

<table>
<thead>
<tr>
<th>Location</th>
<th>Superficial</th>
<th>Deep (&gt;3mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Broad ligament</td>
<td>26 (73%)</td>
<td>-</td>
</tr>
<tr>
<td>Cul-de-sac</td>
<td>25 (69%)</td>
<td>11 (31%)</td>
</tr>
<tr>
<td>Ovary</td>
<td>20 (56%)</td>
<td>7 (19%)</td>
</tr>
<tr>
<td>Uterosacral ligament</td>
<td>12 (33%)</td>
<td>21 (58%)</td>
</tr>
<tr>
<td>Rectum</td>
<td>11 (31%)</td>
<td>7 (19%)</td>
</tr>
<tr>
<td>Peritoneal pocket</td>
<td>6 (17%)</td>
<td>-</td>
</tr>
<tr>
<td>Bladder</td>
<td>-</td>
<td>4 (11%)</td>
</tr>
</tbody>
</table>


### Check out Peritoneal Pockets

Adolescents:
Red Lesions "Flamelike" "Polyps" or "Vesicles"

Batt R J Pediatr Adolesc Gynecol 2003;16:337

### Endometriosis & Mayer-Roikatansky-Kuster-Hauser Syn.

- 20 y/o known Dx MRKH Syn.
- Increasing Pelvic Pain
- Operative Laparoscopy-Endometriosis as Red Polypoid Lesions
- (Stage I)
- Pelvic Kidney
- Implication: Coelomic Metaplasia Theory for Etiology-Endometriosis

Mok-Lin E, Laufer M et al JPAG 2009

### Dilute Vasopressin & Endometrioma's

Less Coagulation Required
Preservation of Ovarian Follicular Activity
Management Options

- Surgical
  - Conservative
  - Correction of Müllerian anomaly
- Medical
  - NSAIDs
  - Oral contraceptives
  - Progestins
  - GnRH agonists
- Combination Surgical/Medical
- Alternative therapies for pain

Laparoscopic Treatment of Endometriosis in Teenagers

- 31 patients (13-20 years old) underwent laparoscopy for chronic pelvic pain unresponsive to NSAIDs and/or OCPs
- Endometriosis found in 11 (36%)
  - Stage I/II, N=5
  - Stage III, N=6
- 3 with Stage I/II and 5 with Stage III were "pain free or greatly improved" following surgery
- 1 with Stage I and 1 with Stage III reported "partial improvement"


Question:
Can visualization of the endometriotic lesion accurately determine the depth of infiltration?

Tip of the Iceberg

Overview Depth of Infiltration

25% - 48% lesions are ≥5mm deep


Question:
How deep are most endometriotic implants?
Limitations of Fulguration

Monopolar destroys tissue at its tips

Bipolar destroys tissue between forceps

Bipolar Instruments

LigaSure Sealing System
- Continuous bipolar waveform

PK (PlasmaKinetic) Sealing System
- Pulsed bipolar waveform
- Allows cooling off pd between bursts of energy, reduces drying of tissue at contact point, and results in less sticking

Thermal Spread of Bipolar Electrosurgery

<table>
<thead>
<tr>
<th>Vessel size</th>
<th>LigaSure™</th>
<th>PlasmaKinetic™</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>2–3 mm</td>
<td>1.2 ± 0.5</td>
<td>1.5 ± 0.2</td>
<td>.27</td>
</tr>
<tr>
<td>4–5 mm</td>
<td>2.4 ± 0.5</td>
<td>2.4 ± 0.4</td>
<td>.79</td>
</tr>
<tr>
<td>6–7 mm</td>
<td>2.5 ± 1.3</td>
<td>3.2 ± 0.5</td>
<td>.32</td>
</tr>
</tbody>
</table>

Thermal spread beyond the bipolar tips is <5mm.


Excision of Endometriosis


-39 pts w/ any stage of Endometriosis

- Randomized to:
  - Immediate group vs. Delayed Group
    - L/S#1 excision vs. L/S#2 (6mo) staging vs. excision
  - 6 & 12 month post-op evaluation

GnRH Agonists

For: Teens > 16 y/a
Add:
- Calcium
- Vit. D.
Add Back: Norethindrone

GnRH Agonist and Bone Mineral Density

- Indicated in > 16 y/a
- Monitor at 6-8mo. Then Every 2 yrs.
- All on Calcium & Vit D & Norethindrone Acetate 5mg/d (Add Back)
- Skeletal Defects: Spine NOT Hip (n=50)
- Bone Health Ctr by Gyn Program @ Children’s Hosp. Boston 1995-2005

Divasta A et al J Pediatr Adolesc Gynecol 2007;20:293s
### Effect of GnRH on Bone Density

<table>
<thead>
<tr>
<th>Author (Date)</th>
<th>Drug</th>
<th>Lumbar spine BMD ∆(%)</th>
<th>6 mos RX</th>
<th>Post RX</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fukushima (1995)</td>
<td>Buserelin</td>
<td>-10.8%</td>
<td>-8.1% (6 mos)</td>
<td></td>
</tr>
<tr>
<td>Revilla (1995)</td>
<td>Triptorelin</td>
<td>-2.9%</td>
<td>-1.0% (6 mos)</td>
<td></td>
</tr>
<tr>
<td>Dawood (1995)</td>
<td>Leuprolide</td>
<td>-14.0%</td>
<td>-3.3% (1 yr)</td>
<td></td>
</tr>
<tr>
<td>Paoletti (1996)</td>
<td>Goserelin</td>
<td>-4.0%</td>
<td>-6.0% (6 mos)</td>
<td></td>
</tr>
<tr>
<td>Taga (1996)</td>
<td>Nafarelin</td>
<td>-3.3%</td>
<td>-2.2% (6 mos)</td>
<td></td>
</tr>
</tbody>
</table>

### Effect of GnRH With Add-Back on Bone Density

<table>
<thead>
<tr>
<th>Author (Date)</th>
<th>Drug</th>
<th>Lumbar spine BMD ∆(%)</th>
<th>6 mos RX</th>
<th>Post RX (6 mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Edmond (1994)</td>
<td>Goserelin</td>
<td>-3.7%</td>
<td>-2.1%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>25 ug E2/5 mg MPA</td>
<td>-2.3%</td>
<td>-1.6%</td>
<td></td>
</tr>
<tr>
<td>Howell (1995)</td>
<td>Goserelin</td>
<td>-4.1%</td>
<td>-1.9%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>25 ug E2/5 mg MPA</td>
<td>-2.3%</td>
<td>-1.6%</td>
<td></td>
</tr>
<tr>
<td>Moghissi (1996)</td>
<td>Goserelin</td>
<td>-4.1%</td>
<td>-1.6%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>0.3 mg CEE/5 mg MPA</td>
<td>-2.0%</td>
<td>-1.6%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>0.625 mg CEE/5 mg MPA</td>
<td>-1.5%</td>
<td>-1.6%</td>
<td></td>
</tr>
</tbody>
</table>

### WHAT WORKS ?

- NSAIDs
  - Selective Drugs
- OCPs-Continuous ? Depending on Etiology
- Neuropathic Analgesics
  - Tricyclic Antidepressants
  - SSRIs
- Muscle Relaxants or Spasmolytics
- GnRHagonists

### OTHER THERAPIES

- SPRMs (Selective Progesterone Receptor Modulators)
- Aromatase Inhibitors
- Intra-uterine Contraceptive Systems

### MULLERIAN ANOMALIES & OUTFLOW TRACT OBSTRUCTION

- Incidence: 0.1-3.8% Adolescents-Pelvic Pain
- Presentation
- Incomplete Obstruction
- Imperforate Hymen Diagnosis & Management
- Transverse Vaginal Septum
Are You Interested in the Genetics?

If there is a Family History of Endometriosis

- Adolescent with Chronic Pelvic Pain Refractory to Medical Therapy
  - 30% Incidence vs 7.6% in Adults
- Genetics:
  - Two Loci: Chrom. 10q26 & 20p13
- Significance in Future Diagnosis or Predisposition to Endometriosis Screening

---

ENDOMETRIOSIS IN ADOLESCENTS KEY POINTS

- Failure to Respond to NSAIDs OCPs
- Empiric Treatment: GnRHag
- If Pain Subsides=Diagnosis Endometriosis
- GnRHag ≥ 16 y/a
- Add back: Norethindrone 5 mg/d
- 2% Femoral Neck bone Loss
- 5% Trabecular Bone Loss
- Majority of Bone Mass Growth Achieved by 20 y/a

---

PAIN EVALUATION
CHRONIC PELVIC PAIN

- Definition: > 6 months
- Prevalence: 38/1000 Females Aged 15-73
- PQRST Approach
  - Provocative & Palliative
  - Quality of Pain
  - Radiation & Relief
  - Timing

MULTISYSTEM APPROACH

- GI
- GU
- GYN
- MUSCULOSKELETAL
- PSYCHOLOGICAL/PSYCHIATRIC

Ectopic Endometrial Glands/Stroma

LABORATORY ASSESSMENT

- CBC
- Pregnancy Test
- Sed Rate
- Cervical Cultures
- Plain Film Abdomen (Stool)
- Pelvic Ultrasound
- Role of Laparoscopy

Alternative Therapies for Pain

- Correct Diagnosis
  - Role of Laparoscopy - Systems Approach First
- Cognitive and behavioral
  - Guided Imagery
  - Progressive muscle relaxation
  - Biofeedback
  - Hypnosis
- Physical Therapy
- Trigger-Point Injections or Peripheral Nerve Blocks
  - Sacral Nerve Stimulation?
- Acupuncture (Pediatrics 2000;104:941)
- Internet
- Support Team
Endometriosis: Difficult to Dx

- Irregular Capillary Lesions
- Black Powder-Burned Lesions
- White Scarred Lesions
- Hemorrhagic Lesions
- Vesicular Lesions

Clinical Case in Pediatric & Adolescent Gynecology

- An Educational Program from ACOG and NASPAG
- Include 32 clinical cases in Pediatric and Adolescent Gynecology

Available for purchase

- Online at http://sales.acog.org or http://www.naspag.org/store.cfm
- By phone at 800-762-ACOG
Minimally Invasive Surgical Approach to Vaginal Agenesis

Heather Appelbaum, MD, FACOG
Associate Professor, Obstetrics and Gynecology
Hofstra NSLIJ School of Medicine
Chief, Division of Pediatric and Adolescent Gynecology
Steven and Alexandra Cohen Children’s Medical Center of New York

Disclosure

• I have no financial relationships to disclose.

OBJECTIVE

1. Review normal and abnormal embryologic development of the Müllerian structures and the urogenital sinus
2. Describe different minimally invasive surgical approaches to creating a neovagina
3. Compare the advantages and disadvantages of minimally invasive surgical vs. non-surgical approaches to creating a neovagina

Müllerian agenesis

Mayer
Described vaginal dysgenesis (1829)

Rokitansky
Further characterized vaginal agenesis (1938)

Küster
Identified associated renal anomalies (1910)

Hauser
Differentiated vaginal agenesis from androgen insensitivity (1961)

Vaginal agenesis/aplasia

• Occurs in 1 in 4,000 to 5,000 live female births
• Variable Müllerian duct development
• 10% obstructed uterus
• 90% fibromuscular bilateral uterine remnants

Associated defects

• 30-50% have associated renal anomalies
• Associated skeletal and auditory anomalies
• 5% of anorectal malformations have associated vaginal agenesis
Developmental embryology of the reproductive tract

- Gonads
- Paramesonephros
- Mesonephros
- Metanephros
- Urogenital sinus
- Sinovaginal bulb

Müllerian Duct Development

- The 2 Müllerian ducts are initially composed of solid tissue and lie side by side.
- Internal canalization of each duct produces 2 channels divided by a septum that is resorbed in a cephalad direction by 20 weeks.
- The cranial, unfused portions develop into the fimbria and fallopian tubes.
- The caudal, fused portions form the uterus, cervix, and upper vagina.

Female genital tract development

- Müllerian duct progression
- Wolffian duct regression
- Renal development
- Cloacal differentiation

Müllerian Duct Development

- 6 weeks – Müllerian ducts elongate caudally and cross the metanephric ducts medially to meet in the midline
- 7 weeks – The urorectal septum develops and separates the rectum from the urogenital sinus
- 12 weeks – The caudal portion of the ducts fuse to form the uterovaginal canal which inserts into the urogenital sinus.

Vaginal agenesis

- Failure of the sinovaginal bulbs to develop and form the vaginal plate
- May be caused by improper induction of the sinovaginal bulbs from the neighboring uterovaginal primordium.
- Hymenal fringe is usually present along with a small vaginal dimple because they are both derived embryologically from the urogenital sinus.

Mayer-Rokitansky-Küster-Hauser Syndrome

- Congenital absence of the uterus and vagina
- Karyotype 46,XX
- Normal ovaries
- Normal secondary sex characteristics
- Normal external genitalia
- Vaginal dimple proximal to hymen
- Associated renal agenesis or malformation
Androgen insensitivity

- 46, XY
- Female body habitus, breast development and external genitalia
- Short blind end vagina
- No Müllerian structures
- Absent axillary or pubic hair growth

Creation of functional vagina

- Coital incidental dilation
- Serial intermittent mechanical self dilation
- Continuous mechanical dilation
  - Vecchietti procedure
  - Balloon vaginoplasty
- Graft vaginoplasty
  - Split thickness skin graft
  - Buccal mucosa
  - Bowel
  - Peritoneum
- Williams labial flap vulvovaginoplasty

Mechanical intermittent self dilation: Frank or Ingram method

Technique

- Dilators of gradual sizes are used to create a vaginal space
- Dilator is placed by the patient at the vaginal dimple
- Pressure is applied for two hours daily

- Alternatively, sit on a bicycle seat, lean slightly forward with the mold in place in a pushing manner for 20 minutes 3 times a day

Frank or Ingram method

Advantages

- Non surgical approach
- 85-90% success rate

Disadvantages

- Requires months to years
- Poor compliance
- Inadvertent urethral or rectal dilation

Vaginal dilation

Laparoscopic assisted creation of a neovagina

- Modified Vecchietti procedure
- Modified Davydov procedure
- Laparoscopic assisted bowel graft vaginoplasty
- Modified balloon vaginoplasty

Continuous mechanical dilation: Modified laparoscopic Vecchietti procedure

- Laparoscopically placed traction dilator device applies continuous pressure resulting in invagination of the vaginal mucosa
- Dilator remains in situ for 7-10 days on continuous traction
- Post procedural intermittent maintenance dilation with functional vagina after six weeks
The Vecchietti procedure

- 1965 Vecchietti devised a traction device
- 1992 modified Vecchietti device for laparoscopic application
- 2009 modified Vecchietti instrument set FDA approved for use in the US

Anatomical landmarks

OR preparation

Laparoscopic suture placement

Placement of the graduated dilator

- Sutures attached to graduated dilator are threaded through eye at the tip of the dissector and then carried through the rectovesicular space
- Post dilator placement requires cystoscopy and proctoscopy to confirm integrity of bladder and rectum

Placement of contralateral suture
Affixing the device

Post operative care
- Hospitalization for pain management 5-7 days
- Adjust traction device q48hours
- Device removed after 7-10 days
- Maintenance dilation with estrogen cream and rigid dilator twice weekly
- Regular intercourse

Surgical outcomes
- n=71
  - Mean duration of surgery 47.5 minutes
  - Mean hospital stay 8.6 days
  - Mean post operative vaginal length 9.6 cm
  - Mean 3 month and 6 month vaginal length 10.7 cm
  - Epithelialization of vagina after 10.1 months

Surgical complications
- 11% developed UTI
- 2.8% accidental perforation of the bladder
- 1.4% bladder hematoma
- 1.4% urethral necrosis
- 1.4% vaginal synechiae
- 1.4% granulation tissue
- No rectal lesions

Patient satisfaction
- 5 cases over 3 years reported satisfactory intercourse with improvement in self-confidence, self-esteem, general well being
- 8 patients over 9 years median vaginal length was 1.5 cm
  - Satisfactory intercourse (7.8/10)
  - 4/6 had minor pain with sexual activity
  - 7/8 would have the procedure again

Long term outcomes
- 110 patients underwent the laparoscopic modified Vecchietti technique
  - Followed at 1 month, 3 month, 6 month and 12 month postoperative
  - Vaginal length, Schiller’s test, quality of sexual intercourse assessed by Rosen’s Female Sexual Function Index (FSFI)


Fertility and Sterility


**Long term outcomes**

- Anatomic success was obtained in 104/106 (98%) patients.
- Functional success was obtained in 103/104 (99%) with no significant difference in desire, arousal, and satisfaction.
- Vaginoscopy showed 90% iodine-positive vaginal type epithelium.
- Vaginal biopsies showed normal glycogen-rich normal squamous epithelium.

**Advantages to the Vecchietti procedure**

- Minimally invasive.
- Functional vagina created in approximately one week.
- No long term post operative complications.
- Good long term sexual satisfaction.

**Davydov technique**

**Postoperative care**

- Vaginal mold left in situ for six weeks.
- Functional vagina after six weeks must be maintained by intermittent dilation or regular intercourse.

**Surgical outcomes n=18**

- 16/18 sexually active.
- 14/16 sexually satisfied.
- 2/16 dyspareunia.
- 0/18 vaginal stenosis.
- 1/18 rectovaginal fistula.
- Vaginal length 6-9cm.
- 0/18 vault prolapse.

**Laparoscopic bowel colpoplasty**

**Surgical technique**

- Segment of bowel is mobilized on vascular pedicle.
- End-to-end anastomosis of the bowel.
- Trochar site enlarged to 3cm to allow for distal end of graft to be exteriorized.
- Distal end of graft closed in a purse string.
- Dissection of the rectovaginal space under laparoscopic guidance.
- Tension free anastomosis to introitus with interrupted circumferential sutures.
**Laparoscopic intestinal graft vaginoplasty**

**Advantages**
- Most successful redo procedure
- Excellent option for patients with combined anorectal malformations and vaginal agenesis
- Adequate vaginal length
- Natural lubrication
- Early coitus
- Lack of shrinkage
- # minimally invasive

**Disadvantages**
- Intestinal complications
- Not really minimally invasive
- Intestinal stenosis
- Leukorrhea


---

**Outcomes laparoscopic sigmoid colpoplasty**

*n=7*

- Mean operative time=312 (220-450) min
- Mean blood loss=decrease in Hb=3.6g/dl
- Mean hospital stay=7.7 days
- Mean vaginal length=11.5 cm (7-15cm)
- Introital dilation required=2/7
- UTI=1/7
- Vulvar hematoma=1/7


---

**Satisfaction**

- 4/7 sexually active
- 0/4 dyspareunia
- 3/4 completely satisfied
- 7/7 satisfied with surgical outcome


---

**Balloon vaginoplasty**

- Laparoscopic suction irrigator used to apply downward pressure through rectovesicular space to level of vaginal dimple
- Cannula used to guide needle intraperitoneally through the rectovesicular space and then exited out abdominally
- Silk sutures affixed to 18g Foley catheter
- Foley carried through intraperitoneal space to exit at the vaginal fovea
- Inflate balloon with 15cc saline
- Sutures removed and countertraction applied
- Serial inflation of balloon


---

**Surgical outcomes**

*n=6*

- 1/6 urethral injury
- 1/6 rectal injury
- Neovagina 9-12cm
- 5/6 sexual active
- 90% sexual satisfaction


---

**A look into the future of treatment for Müllerian agenesis**

- Controlled studies with quality indicators comparing anatomical and functional outcomes of different approaches to vaginal agenesis are needed
- A look toward future human uterus transplantation (Green Journal, June 2012)
References


Our case... Ms. BH

Ms. BH is an 18yo G0 AAF with PMH significant for Hodgkin’s Lymphoma, HTN, DM who initially presented to the Pedi Gyn clinic in 09/2006 prior to initiation of chemotherapy for discussion of her fertility.

Hodgkin’s lymphoma

- Most children with HL present with painless lymphadenopathy, usually cervical, supraclavicular, axillary, or, less often, inguinal - feel rubbery.
- They also have many nonspecific systemic symptoms including fatigue, anorexia, and weight loss.
  - Fewer than 20 percent of children with HL have the classic fever and night sweats that are seen in adults.
- Twenty years after diagnosis of HL, the cumulative incidence of second malignancies was 7.6%.
  - Breast cancer, thyroid cancer, and soft tissue sarcomas were the most common ones.

Cancer diagnosis

- 4% of all people newly diagnosed with cancer are younger than 35 years old (40,000 per year).
- 1-2% of all people newly diagnosed with cancer are younger than 19 years old (12,000 per year).
- In 2010, it is estimated that 1/250 adults will be childhood cancer survivors.

Disclosure

- Consultant: Ethicon Endo-Surgery, Ethicon Women's Health & Urology, Conceptus Incorporated, CONMED Corporation, UpToDate

Preserving Fertility in Adolescents with Cancer

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Infertility risk

- Risk of infertility is associated with 3 key aspects of cancer therapy: patient gender and age, type of chemotherapy used, whether or not radiation is used (where and what dosage).
- Greatest infertility risk is associated with chemotherapy using alkylating agents: cyclophosphamide, ifosfamide, nitrosoureas, chlorambucil, busulfan, procarbazine. Causes accelerated oocyte apoptosis...
- Radiation risk highest with any type of abdominal pelvic radiation, but also with cranial!

A refresher...

- The number of oocytes that females have at birth is FIXED at approximately 1-2 million.
  - 6 month old: 700,000
  - 7 years old: 300,000
  - 37 years old: 25,000
  - 50 years old: 1,000

Infertility

- Rates of permanent infertility depends greatly on many factors...
- In females, infertility can be related to decreased available primordial follicles or, alterations in blood supply available to the reproductive tract, or disruptions in the "normal" anatomic locations of the reproductive organs, or disruptions in hormone production.
- Resumption of menses is NOT an indicator of fertility, as patients typically believe.
- And retained fertility immediately after treatment does not mean a normal duration of fertility (aka - increased risk of POF)

Possible treatment effects

- Immediate infertility
- Increased risk of miscarriages
- Premature ovarian failure
  - Menopause before age 40
  - Childhood Cancer Survivor Study showed us that 8% had POF overall and those who received radiation to the pelvis or abdomen had a 30% chance of developing POF.
- Increased risk of POF

Bone marrow transplant

- Bone marrow transplant is associated with a >90% risk of POF secondary to pre-therapy whole body radiation.
- Only 9 reports of return of ovarian function in a population of 144 patients studied (all < 25 years old).

Abdominal/Pelvic radiation

- In one case series, 71% of treated pre-pubertal girls failed to enter into puberty, and 26% of the cases that did experience puberty had POF (rad doses all between 2000-3000 cGy)
- Ovarian tissues have an LD_{50} value of 600 cGy
- It has become apparent that ovaries which are located outside of radiation field continue to function much more normally than direct or indirectly radiated ovaries (volume dependent).
**Degree of risk**

**High risk (80%):**
- Hematopoietic stem cell transplantation with cyclophosphamide/total body irradiation or cyclophosphamide/busulfan
- External beam radiation to a field that includes the ovaries
- CMF, CEF, CAF 6 cycles in women age 40 and older (adjuvant breast cancer therapy with combinations of cyclophosphamide, methotrexate, fluorouracil, doxorubicin, epirubicin)

**Intermediate risk:**
- CMF, CEF, CAF 6 cycles in women age 30-39 (adjuvant breast cancer therapy with combinations of cyclophosphamide, methotrexate, fluorouracil, doxorubicin, epirubicin)
- AC 4 in women age 40 and older (adjuvant breast cancer therapy with doxorubicin/cyclophosphamide)

**Lower risk (20%):**
- ABVD (doxorubicin/bleomycin/vinblastin/dacarbazine)
- CHOP 4-6 cycles (cyclophosphamide/doxorubicin/vincristine/prednisone)
- CVP (cyclophosphamide/vincristine/prednisone)
- AML therapy (anthracycline/cytarabine)
- ALL therapy (multi-agent)

**Very low or no risk:**
- Vincristine, Methotrexate, 5-fluorouracil

**Unknown risk:**
- Taxanes, Oxaliplatin, Irinotecan, Monoclonal antibodies (trastuzumab, bevacizumab, cetuximab), Tyrosine kinase inhibitors (erlotinib, imatinib)

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**Risk assessment**

- Assessment of risk for fertility preservation
- Refer to specialists with expertise in fertility preservation

**Fertility preservation options**

- Depends on the age, diagnosis, type of treatment needed, whether or not the patient has a partner...
- Surveys have found that at least 50% of all men and women treated with cancer during their reproductive ages do NOT recall ever having discussed the issue of fertility with their oncologist and many of those that DO report having had such a discussion felt that their concerns were not appropriately addressed.
- Psychologic counseling also should be offered as part of such discussions...

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**Hodgkin’s Lymphoma**

- Typical chemotherapy regimen consists of: chlorambucil, vinblastine, procarbazine, prednisolone.
  - Risk of gonadal failure in men – 86%
  - Risk of gonadal failure in women – 50%
- Alternative regimen (without alkylating agent): adriamycin, bleomycin, vinblastin, dacarbazine

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**Embryo cryopreservation**

- Obviously requires a partner...
- After puberty
- Delay of cancer treatment: 2-6 weeks
- Live birth rates depend on the patient’s age and number of embryos able to be cryopreserved (usually about 10-25% per embryo).
- Pt must be willing to undergo ovarian hyperstimulation with daily hormone injections x 2 weeks and travel to multiple u/s appts to eval follicles prior to collection procedure.
- The most established technique for fertility preservation in women.
- Cost: approx $8,000 per cycle, $350 per year storage fees.
Oocyte cryopreservation (investigational)

- No partner/donor required.
- After puberty
- Same time commitment, same cost.
- Definitely investigational... small case series and case reports; as of 2005, 120 deliveries reported, approximately 1-3% live births per thawed oocyte (3-4 times lower than standard IVF).

Ovarian tissue cryopreservation (investigational)

- Before or after puberty
- Clearly not suitable if cancer is suspected to have metastasized to ovarian tissue.
- Still in early stages... case reports only; as of September 2012, only twenty live births reported.
- Re-implantation can restore hormonal function
- Cost: >12,000


Strategies to preserve fertility in female cancer patients through freezing

- Ovarian stimulation
- Oocyte retrieval
- In vitro fertilization (IVF)
- Egg donation
- Ovarian tissue harvesting
- Ovarian tissue storage
- Ovarian tissue re-implantation
- Whole-embryo freezing

Gonadal shielding during radiation therapy (studied)

- Only selectively possible.
- Expertise IS required to ensure that shielding does not interfere with other nearby areas (unwanted increases in dosing, etc).
- No additional cost.

Ovarian transposition/oophoropexy (studied)

- Same day, outpatient procedure (minimal time commitment compared to others)
- Helps to prevent radiation damage, specifically, and should be done two weeks or less prior to therapy initiation to prevent dislocation.
- May need re-positioning later, or IVF.
- Large cohort studies and case series suggest approximately 50% chance of success due to altered ovarian blood flow and scattered radiation.

Oophoropexy

- The ovaries and their attached vascular supply from the ovarian vessels are brought out of the pelvis and sutured lateral and above the psoas muscle to get them out of the field of radiation.
  - Some authors recommend using permanent suture for this procedure and not dividing any of the attachments.
**Oophoropexy**

- Studied
- Obviously for young cervical cancer cases...
- Limited to early stage cases only
- Requires inpatient admission and usually about 6 weeks of recovery prior to treatment initiation.
- Expertise may be lacking
- Would require cerclage to maintain future pregnancy.

**Ovarian suppression with GnRH analogs or antagonists (investigational)**

- Chemotherapy protective.
- After puberty.
- Given before and during treatment with chemo to stop “ovarian activity” and theoretically protect the adnexa at the cellular level.
- Reduces risk of POF from about 58% to 3%, especially when cytotoxic alkylating agents are used.
- Give agonist for immediate treatment initiation and OK to give antagonist if can delay treatment for approx 4 weeks.
- Cost: approximately $500/mo (monthly injections)

**The ethics question**

- Deciding to pursue or forgo potential fertility sparing procedures/treatments requires a high level of decision making capacity and is very often made by the child’s parents.
- Is parenthood something that these children would even want in the future?
- It is hard to take out the personal bias of parents from this decision... too unfocused and dreaming of grandchildren someday vs too focused on saving the patient now. Parental judgement may not reflect the patient’s own best interest in the future.

**Back to our case...**

The patient denied sexual activity or being in a stable relationship. It was clear, however, that she desired to have children in the future but she was wary of operations and was told that she needed to start her chemotherapy soon...

**What would you do?**
What we did...

We offered her Lupron for suppression during her chemotherapy and she accepted this option. She continued it and did very well – chemo ended 11/2006.

But, then, pelvic lymphadenopathy was noted... oncology plan: local radiation.

What we did...

Long discussion (2 hours) was had with the patient and her mother regarding the patient’s wishes for future fertility – it was decided that she would undergo surgery for oophoropexy in preparation for her need for localized pelvic therapy.

Unresolved issues in human ovarian transplantation

- Patient selection and exclusion criteria
- Optimization of freeze-thaw protocols
- Optimal graft site(s)
- Quality of oocytes matured in a graft
- Efficacy of transplantation for restoration of fertility
- Safety issues
- Ischemia-reperfusion injury
- Prospects for in vitro follicle culture
- Long term adverse effects on offspring


Pregnancy after cancer treatment

- Pregnancy may be complicated with an overall increased risk of organ impairment, especially of the heart, lungs, and uterus (consider testing pre-conceptually).
- There is evidence that pregnancy may increase the risk of worsening cardiac ejection fraction in women treated with doxorubicin for childhood cancer, and uterine or total-body irradiation appears to increase the risk of miscarriage, prematurity and low birth weight.
- Also, if the patient will be using IVF, the increased risk of multiple gestation worsens these above risks/considerations.

The progeny

- Aside from hereditary genetic syndromes, however, there is scant evidence that a history of cancer, cancer therapy, or fertility interventions increases the risk of problems in the progeny.
- Birth defects of progeny of cancer survivors carry the same overall risks of birth defects as the general population (2-3%).

Additional parenting options

- IVF with donor eggs or embryos
  - Expensive (10,000 per cycle)
- Surrogacy
  - Expensive (20-100,000)
- Adoption
  - Expensive (5-35,000)
  - Personal h/o cancer CAN be prohibitive with waiting period, etc
ASRM Ethics Committee Statement

1. Physicians should inform cancer patients about options for fertility preservation and future reproduction prior to treatment.

2. The only established methods of fertility preservation are sperm cryopreservation in men and embryo cryopreservation in women.

3. Experimental procedures such as oocyte or ovarian tissue cryopreservation should be offered only in a research setting with IRB oversight.

4. Concerns about the welfare of resulting offspring should not be cause for denying cancer patients assistance in reproducing.

5. Parents may act to preserve fertility of cancer patients who are minors if the child assents and the intervention is likely to provide net benefits to the child.

6. Precise instructions should be given about the disposition of stored gametes, embryos, or gonadal tissue in the event of the patient’s death, unavailability, or other contingency.

7. Preimplantation genetic diagnosis to avoid the birth of offspring with a high risk of inherited cancer is ethically acceptable.

Latest Information

- ASRM resources on Fertility Preservation

THE END!

Any questions?
CULTURAL AND LINGUISTIC COMPETENCY

Governor Arnold Schwarzenegger signed into law AB 1195 (eff. 7/1/06) requiring local CME providers, such as the AAGL, to assist in enhancing the cultural and linguistic competency of California's physicians (researchers and doctors without patient contact are exempt). This mandate follows the federal Civil Rights Act of 1964, Executive Order 13166 (2000) and the Dymally-Alatorre Bilingual Services Act (1973), all of which recognize, as confirmed by the US Census Bureau, that substantial numbers of patients possess limited English proficiency (LEP).

California Business & Professions Code §2190.1(c)(3) requires a review and explanation of the laws identified above so as to fulfill AAGL’s obligations pursuant to California law. Additional guidance is provided by the Institute for Medical Quality at http://www.imq.org.

Title VI of the Civil Rights Act of 1964 prohibits recipients of federal financial assistance from discriminating against or otherwise excluding individuals on the basis of race, color, or national origin in any of their activities. In 1974, the US Supreme Court recognized LEP individuals as potential victims of national origin discrimination. In all situations, federal agencies are required to assess the number or proportion of LEP individuals in the eligible service population, the frequency with which they come into contact with the program, the importance of the services, and the resources available to the recipient, including the mix of oral and written language services. Additional details may be found in the Department of Justice Policy Guidance Document: Enforcement of Title VI of the Civil Rights Act of 1964 http://www.usdoj.gov/crt/cor/pubs.htm.

Executive Order 13166, “Improving Access to Services for Persons with Limited English Proficiency”, signed by the President on August 11, 2000 http://www.usdoj.gov/crt/cor/13166.htm was the genesis of the Guidance Document mentioned above. The Executive Order requires all federal agencies, including those which provide federal financial assistance, to examine the services they provide, identify any need for services to LEP individuals, and develop and implement a system to provide those services so LEP persons can have meaningful access.

Dymally-Alatorre Bilingual Services Act (California Government Code §7290 et seq.) requires every California state agency which either provides information to, or has contact with, the public to provide bilingual interpreters as well as translated materials explaining those services whenever the local agency serves LEP members of a group whose numbers exceed 5% of the general population.

If you add staff to assist with LEP patients, confirm their translation skills, not just their language skills. A 2007 Northern California study from Sutter Health confirmed that being bilingual does not guarantee competence as a medical interpreter. http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=2078538.

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