Anatomical Assessment and Planning

The key to functional outcomes in cloaca management

Richard J. Wood & Geri Hewitt
Center for Colorectal and Pelvic Reconstruction
Nationwide Children’s Hospital
The Ohio State University College of Medicine
• No Disclosures
Complex anatomy problem: Stages in management

- New born period
- Reconstructive planning
- Reconstructive surgery
- Urological functional outcomes
- Colorectal functional outcomes
- Gynecological functional outcomes
Newborn period

• Make diagnosis
• Variations: Classic cloaca, Posterior cloaca, Covered Cloacal extrophy
• Significant co-morbidity (associated anomalies)
  1. Cardiac
  2. TEF
  3. LT clefts
  4. Significant renal abnormalities
Variations
Newborn period

- Need for colostomy: most cases (variations)
- Hydrocolpos: presence or absence
- Functional effects: Obstruction to urinary drainage
  1. Bladder
  2. Ureters
  3. Kidneys
- Drainage options: transabdominal vs transperineal
Newborn female infant with no visible anus

How many perineal orifices?

1. Cloaca
   - Renal & Pelvic USS (Day 1)
   - Hydrocolpos +/- Hydronephrosis
   - NO Hydrocolpos
   - Vaginostomy + Colostomy
     - 3D Cloacaogram + Cystoscopy

2. Vaginal atresia
   - Rectovaginal fistula
     - Rectum NOT reachable through posterior sagittal incision

3. Perineal fistula
   - Vestibular fistula
     - No fistula
     - Cross table radiograph
     - Rectum reachable through posterior sagittal incision

Primary PSARP
Imaging
Vaginostomy creation with septum division
Reconstructive planning

• Timing of reconstruction
• Complexity matters: vaginal replacement
• Data required for planning
• Quality of bladder neck and urethra
• Vaginal and mullerian development (vaginal septum, presence and numbers of cervices)
• Height of rectum, need for MIS
• **Length of the common channel**
Common Channel Length

• Important
• Not an isolated measurement
• Contextualized by other factors.
Cysto-vaginoscopy and EUA

- Multi-disciplinary team
- Urology, Gynecology, Colorectal, Radiology in the OR
- Visualize structures
- Place catheters
- Some spatial understanding
3D cloacagram

- Spatial understanding
- Reduce learning curve
- Combined with C-V EUA reviewed by combined team
- Leads to consolidated pre-operative plan
- More effective family counseling
3-D Cloacogram
Imaging
Modeling
Modeling
Reconstructive Surgery

- Overall strategy
- Vaginal replacement required
- What tissue, colon, rectum or ileum
- Transperineal only, or transabdominal also
- Open vs MIS
- Urological drainage options: SP tube, vesicostomy, circle stent, ureterostomy
Vaginal Replacement (colon)
Vaginal Replacement (Small Bowel)
Vaginal Replacement (rectum)
Longitudinal transection
Vaginal Replacement (rectum) Transverse transection
Reconstructive Surgery

• Spatial perineal planning
• Urethra placement
• Location of anterior border of anal sphincter complex
• Length of perineal body
• Space left for introitus
• Maximize functional outcomes.
Urological functional outcomes

• Ability to drain and store urine
• Renal failure: 30-50 % lifetime risk.
• Nephrons, nephrons, nephrons
• Nephrotoxic drugs
• Renal dosing
• Ensure bladder emptying
• Aggressively treat UTI’s
Urological functional outcomes

- Timing of functional urological reconstruction
- Continent vs incontinent options
- Compliance
- Quality of life vs Nephrons
- Anatomical location of Mitrofanoff and Malone
- Implications on fertility and delivery implications
Colorectal functional outcomes

- Sacral and spinal development
- Complexity of malformation
- Perineal muscle development (subjective)
- Continence index (work in progress)
- Quality of repair
- Optimize potential
Colorectal functional outcomes

• Bowel Management
  1. Laxatives and water soluble fiber
  2. Enemas (stimulant)
     Ante-grade
     Retrograde

• Combined surgery with Urology and Gynecology
Combined reconstructive surgery

- Bladder status
- Ureters and reflux
- Bladder neck function
- Colon status
- Need for ante-grade enema options
- Assessment of Mullerian structures
References:

Gynecologic Considerations
Counseling families and patients with ARM

(this is what they ask me!)

• Many have been told ovaries absent on imaging—reassurance required about no evidence of associated ovarian absence/failure
• Concerns around timing and tempo of pubertal changes
• Inquiries about gynecologic anatomy
• Worry about romantic partnership and sexuality
• Questions about fertility potential and childbearing risks as well as mode of delivery—pre-conceptual counseling
• Seek information about types, number, and timing of surgical interventions to maximize reproductive health
Goals in taking care of patients with ARM

• “Our surgical practice for the future in this area should concentrate on preempting the gynecological problems and developing surgical techniques that will minimize the need for additional or multiple procedures later in life and improve functional outcomes.”

• That feels like a tall order!
• The health care community caring for these kids and the families have many similar concerns

Preempting gynecologic problems/multiple surgical interventions

- Hydrocolpos
- Mullerian tract obstruction
- Vaginal septum
- Inadequate perineal body
- Introital/vaginal stenosis
Hydrocolpos

- 30% of patients will have hydrocolpos at birth—associated with longer channel cloaca and UV duplication
- Transperineal or transabdominal drainage indicated
- May lead to infection, scarring, and reduced fertility

Outflow tract obstruction

- Risk of obstruction at time of menarche varies in series from 11-41%
- Obstruction reported as a mixed bag: vaginal (atresia and/or stenosis), cervical, mullerian
- Some patients reported in obstruction rates have yet to undergo vaginal reconstruction
- Obstruction: pain as well as endometriosis and adhesive disease

Mullerian obstruction: the balance

- Management of mullerian anatomy: balancing reproductive potential and risk of obstruction
- What defines a mullerian structure with reproductive potential? Endometrium only? Cervix? Connected to vagina?
- NASPAG committee opinion: removal of obstructed uterine horn—removal of communicating small rudimentary horn to prevent ectopic pregnancy
- Removing mullerian structures is permanent: there’s no redo.
- 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 anatomy

- Literature around mullerian anatomy in patients with cloacal malformations is variable
- At least partly due to inconsistent use of language/nomenclature
- Numerous types of surgeons offering descriptions of mullerian anatomy at very young ages
- We are sometimes limited in determining mullerian anatomy at the time of assessment prior to and/or during primary repair
- Tools: preoperative imaging (usn, mri, cloacogram), vaginoscopy, primary repair, subsequent procedures, post pubertal imaging, symptoms: documented failure prior to puberty
Mullerian anatomy: what do we know?

- From case series, up to 40% will have duplication of mullerian system with two vaginas and uteri
- Other mullerian anomalies are commonly reported: approximately 25% will have amenorrhea from uterine hypoplasia; obstruction rates reported from 11-41%--includes vaginal, cervical, and mullerian obstructions
- Normal menstruation reported from 32-60% of adolescents
- Risk of mullerian anomaly: Couchman series (52%) and our series significantly higher (90%).

- Pradham, S et al. Intraoperative and radiologic assessment of mullerian and ovarian anatomy in patients with cloacal anomalies—in what proportion of patients are we sure? NASPAG abstract 4/17
Mullerian anatomy: preoperative imaging

- In the infant undergoing primary repair, pelvic ultrasound or MRI is often not predictive of intraoperative anatomy.
- In our experience (28 cloacal patients seen first two years of NCH CCPR): USN and/or MRI correlated only 38% of the time with intraoperative findings—(22/28 patients having undergone l/s or laparotomy and 28/28 had EUA/vaginoscopy).
- UK series: 10 patients with USN and/or MRI (age 6-17) suggested hypoplastic uteri who went on to menstruate (age 13-17). 1 with ARM; 1 had UG sinus.

- Pradham, S et al. Intraoperative and radiologic assessment of mullerian and ovarian anatomy in patients with cloacal anomalies—in what proportion of patients are we sure? NASPAG abstract 4/17
- Michala, L et al. The clandestine uterus: or how the uterus escapes detection prior to puberty. BJOG 2010; 117:212-215.
Mullerian anatomy: vaginoscopy

- No cervix
- One cervix
- Two cervices: most predictive of accurate assessment of mullerian structures
Mullerian anatomy: intraoperative intraabdominal assessment

- Identifying anatomy can be challenging in an infant
- Antegrade and retrograde assessment of patency
- How good are we?

Mullerian anatomy: intraoperative intraabdominal assessment--misfires

- Warne series: 10 patient intraoperatively identified “early” with uterine hypoplasia; 6/10 went on to have normal menstrual function
- Skerritt literature review (vaginal replacement for vaginal agenesis associated with ARM): 1 patient with uterine hypoplasia diagnosed intraoperatively went on to menstruate. Six patients diagnosed with vaginal-cervical agenesis had no complications after anastomosis of bowel neovagina to ”lower uterine segment”.
- Intraoperative decision making? How good are we? Not perfect.
Mullerian anatomy: the black box

- A proportion of patients with cloacal malformations will have uncertain mullerian anatomy as they enter puberty
- In our series, about 1/3 of our first 28 cloacal patients do not have confirmation of their mullerian structures
- All patients require a high index of suspicion for an associated mullerian anomaly
- We routinely do pelvic ultrasound 6 months after thelarche, ask patients to call us with menarche, ask them to report symptoms of dysmenorrhea, and ask them to repeat pelvic ultrasound after menarche

- Pradham, S et al. Intraoperative and radiologic assessment of mullerian and ovarian anatomy in patients with cloacal anomalies—in what proportion of patients are we sure? NASPAG abstract 4/17
Mullerian anatomy: 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.

Vaginal septum

- Can impact sexual functioning as well as menstrual hygiene
- Seen in patients with cloacal anomalies as well as less complex ARM (5% of patients with recto vestibular fistula)
- Removal at time of primary reconstruction—minimize anesthesia exposure, excellent surgical exposure
- If present after primary repair, can be removed at time of another procedure without evidence of increased morbidity or if as an isolated procedure, consider pubertal timing

Inadequate perineal body

- An adequate perineal body is important not only for fecal continence, but also hygiene, sexual functioning, cosmesis
- Patients may present to gynecologists with complaints involving perineal body after previous surgical repair or undiagnosed ARM
- In our series, 60% (15/28) of patients (aged 6 months to 12 years) referred for evaluation of previously repaired ARM required redo perineoplasties, most commonly due to anteriorly mislocated anus.
- Identified gynecologic abnormalities: introital stenosis (n=4), retained vaginal septum (n=3), remnant of recto vestibular fistula (n=2)

Inadequate perineal body
Inadequate perineal body
After re-do perineoplasty
Introital/vaginal stenosis

• Risk that patients may need additional surgery after their primary repair to address introital and/or vaginal stenosis
• With early reconstruction, typical vaginal stenosis rate reported between 36-41%
• Couchman series: 9/16 (56%) of cloacal patients who underwent vaginoplasty in first year of life required further reconstruction for menstrual egress or sexual activity
• Warne series: 3/30 patients required additional surgery for vaginal stenosis after previous early reconstruction

Functional outcomes: sexuality

- Factors that may impact sexual functioning: anatomy or anatomic repair, poorly developed sacrum, nerve damage, shortened perineum, stenosis, scar tissue/adhesive disease, psychosocial factors, body image, concerns around continence
- CURE-Net: 65% of adult patients with ARM were sexually active
- CURE-Net: patients with ARM, regardless of gender, had sexual debut (both partnered and alone) at later age

Functional outcomes: sexuality

- Davies series: 44 adult women with ARM (all types); the majority were sexually active
- Direct linear relationship between urinary and fecal incontinence and raised sexual anxiety, greater sexual depression, and fear of sexual relationships
- Couchman series: 19 patients (cloaca) (aged 13-35) 8 (42%) were sexually active—one reported difficult penetration—no data on sexual satisfaction

Functional outcomes: fertility

- Little is known about fertility potential in ARM patients
- Limited information available, particularly about attempts to achieve conception or length of time required
- Some case series reporting childbirth rate and obstetrical outcomes
- Females with complex ARM appear to have lower child birth rates compared to both healthy controls and patients with less complex ARM: possible explanations include mullerian anomalies, iatrogenic damage, psychosocial aspects

Functional outcomes: obstetrics

• Know information about isolated mullerian anomalies and obstetrical outcomes may not be completely applicable to cloacal patients
• As cloacal patients as a cohort are aging, more reports of pregnancy outcomes
• Preconceptual care with co-morbidities is important
• Complexity of delivery with previous abdominal or pelvic surgery, Malone or Mitrofanoff in place, renal or cardiac abnormalities
• Most likely cesarean delivery
Summary: Longitudinal assessment and care

• Limited information on long term gynecological outcomes available when counseling families
• Mullerian anatomy may be a “black box” and require transition through puberty for full delineation
• Intraoperative mullerian management: balancing reproductive opportunity and risk of obstruction
• Multiple surgical interventions may be required—consider collaborative procedures and pubertal timing
• Comprehensive pre-conceptual care especially with more complex ARM patients: multiple pelvic surgeries, medical co-morbidities
• Longitudinal, multidisciplinary care for best functional outcomes