

# Anorectal anomalies: Genitourinary problems

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# Spectrum of anomalies



# Why worry about Urology

- Prevent renal damage 6%
- Reduce UTI's 22%
- Improve continence 10-35%
- Help with sexual dysfunction

# Role of Urology

- Pre reconstruction
  - Evaluation of whole genitourinary tract
  - Pre reconstruction UTI's
- During reconstruction
  - Understanding anatomy
  - Combining GU reconstruction
- Post reconstruction
  - Protecting renal function
  - Continence
  - Genital problems

# Urological Abnormalities

- 53 patients (83%) had an abnormality of the urinary tract
  - renal structural abnormalities
  - abnormal renal position
  - ectopic ureters
  - VUR
  - Bladder abnormality

# Renal anomalies N= 50

- Renal dysplasia 8
- Solitary kidney 4
- MCDK 4
- Hydronephrosis 4
- Duplex 3
- Fused kidney 2

# UTI's Pre reconstruction

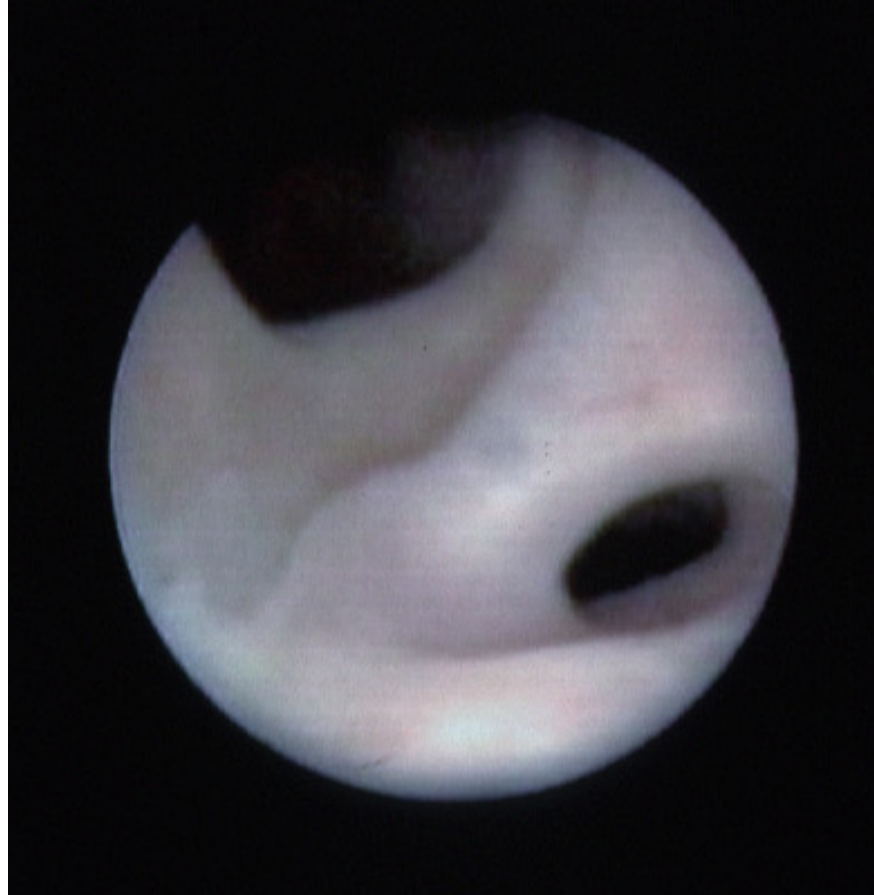
- Consider
  - Feces in the distal colonic loop
  - Not emptying bladder
  - VUR
- Options
  - Ensure no overflow from the proximal stoma
  - Washout distal loop
  - Maximize bladder drainage

# Pre op Investigations

- Renal ultrasound scan
- Renal scan
- VCUG
- Spinal imaging



# Cystoscopy



# Combining GU reconstruction

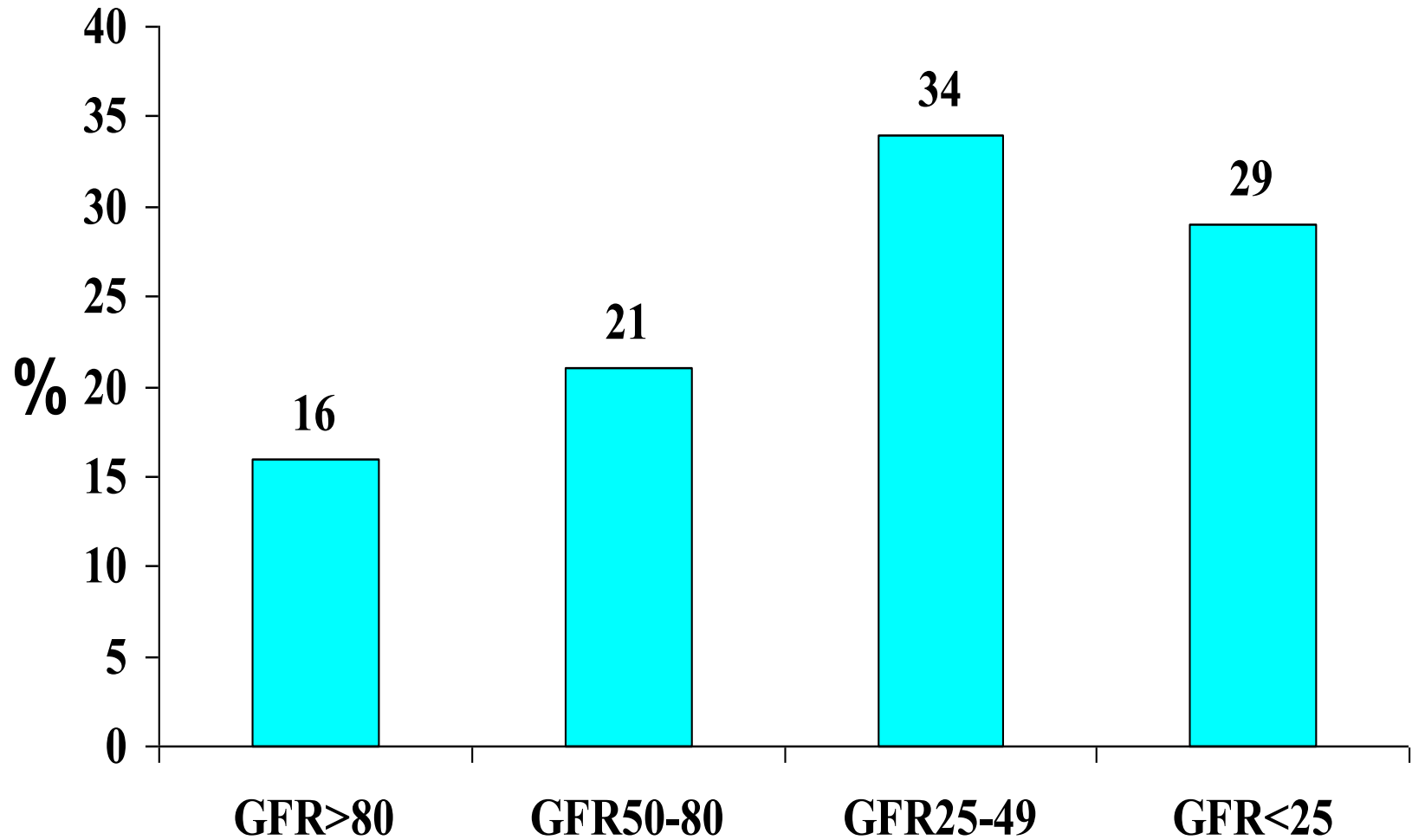
- Undescended Testicle
- Hypospadias
- Vaginal Septum



# Post reconstruction

- Protection of renal function
- Managing continence
- Genital issues
  - Testicular pain
  - Menstrual problems
  - Sexual dysfunction

# Last GFR measurements



# Protecting renal function

- Minimizing UTI's
  - Bladder emptying
  - Constipation
  - Antibiotics
  - Reflux
- Managing hydronephrosis
- Improving bladder mechanics

# Continence rate -Intermediate term

Fistula site	Voiding Dry	Wet	CIC
Cloaca (14)	4 (29%)	1 (7%)	9 (64%)
Bladder neck (5)	0	1 (20%)	4 (80%)
No fistula (3)	1 (33%)	2 (67%)	0
Low (28)	25 (89%)	3 (11%)	0

# Adult rate of Urinary Incontinence

	<b>Total n = 74</b>	<b>Low ARM n = 23</b>	<b>High ARM n = 25</b>	<b>Cloaca n=15</b>	<b>Unclassifiable ARM n=11</b>
<b>None</b>	<b>44 (59%)</b>	<b>14 (61%)</b>	<b>18 (72%)</b>	<b>3 (20%)</b>	<b>9 (82%)</b>
<b>Monthly</b>	<b>1 (1%)</b>	<b>0</b>	<b>0</b>	<b>1 (7%)</b>	<b>0</b>
<b>Weekly</b>	<b>16 (22%)</b>	<b>8 (35%)</b>	<b>3 (12%)</b>	<b>4 (27%)</b>	<b>1 (9%)</b>
<b>Daily</b>	<b>9 (12%)</b>	<b>1 (4%)</b>	<b>3 (12%)</b>	<b>4 (27%)</b>	<b>1 (9%)</b>
<b>All of the time</b>	<b>4 (5%)</b>	<b>0</b>	<b>1 (4%)</b>	<b>3 (20%)</b>	<b>0</b>

# Long term outcome – bladder emptying

	<b>Total n = 74</b>	<b>Low ARM n = 23</b>	<b>High ARM n = 25</b>	<b>Cloaca n=15</b>	<b>Unclassifiable ARM n=11</b>
<b>Normal</b>	<b>57 (77%)</b>	<b>23 (100%)</b>	<b>20 (80%)</b>	<b>5 (33%)</b>	<b>9 (82%)</b>
<b>Self-catheter</b>	<b>8 (11%)</b>	<b>0</b>	<b>3 (12%)</b>	<b>3 (20%)</b>	<b>2 (18%)</b>
<b>Mitroff</b>	<b>7 (9%)</b>	<b>0</b>	<b>0</b>	<b>7 (47%)</b>	<b>0</b>
<b>Ileal con</b>	<b>1 (1%)</b>	<b>0</b>	<b>1 (4%)</b>	<b>0</b>	<b>0</b>
<b>Express bladder</b>	<b>1 (1%)</b>	<b>0</b>	<b>1 (4%)</b>	<b>0</b>	<b>0</b>



# Functional Anomalies in ARM patients

## Incontinence of Urine

- Wiener '74 31%
- Hassink'83 22%
- Rintala '94 (Adult FU) 33%
- Boemers '95 24%
- Pena '95 10%

# Risk factors for incontinence

- Spinal anomalies
- Severity of anomaly
- Bladder Neck
- Surgery

# Urinary incontinence

	Incidence
• Normal sacrum	11%
• Abnormal sacrum	56%
• Overall	24%

# Functional anomalies

## Effect of PSARP

- 32 patients
- Temporary anomaly 3
- Permanent anomaly 3

# Male genital anomalies

- Up to 52% have an anomaly
- Undescended testicles 10 -40%
- Bifid scrotum
- Penile anomalies 14 -25%
  - Hypospadias
  - Chordee
  - Epispadias
  - Penile duplication

# Testicular pain

- Testicular torsion
- Epididymitis



# Long term outcome

- 30 males
- Fertility
  - 11 men had partners that conceived
  - 4 required IVF

# Female genital anomalies

- Internal genitalia
  - Up to 30 -45%
  - 60% of cloacal anomalies will have septation anomalies
- External genitalia
  - Hamartomas
  - Hemangiomas



# Long term outcome

- 44 women
- 40 menstruate normally
  - 2 hysterectomy
- Fertility
  - 7 women reported 16 pregnancies
  - 1 is undergoing IVF
  - 36 have not tried to conceive

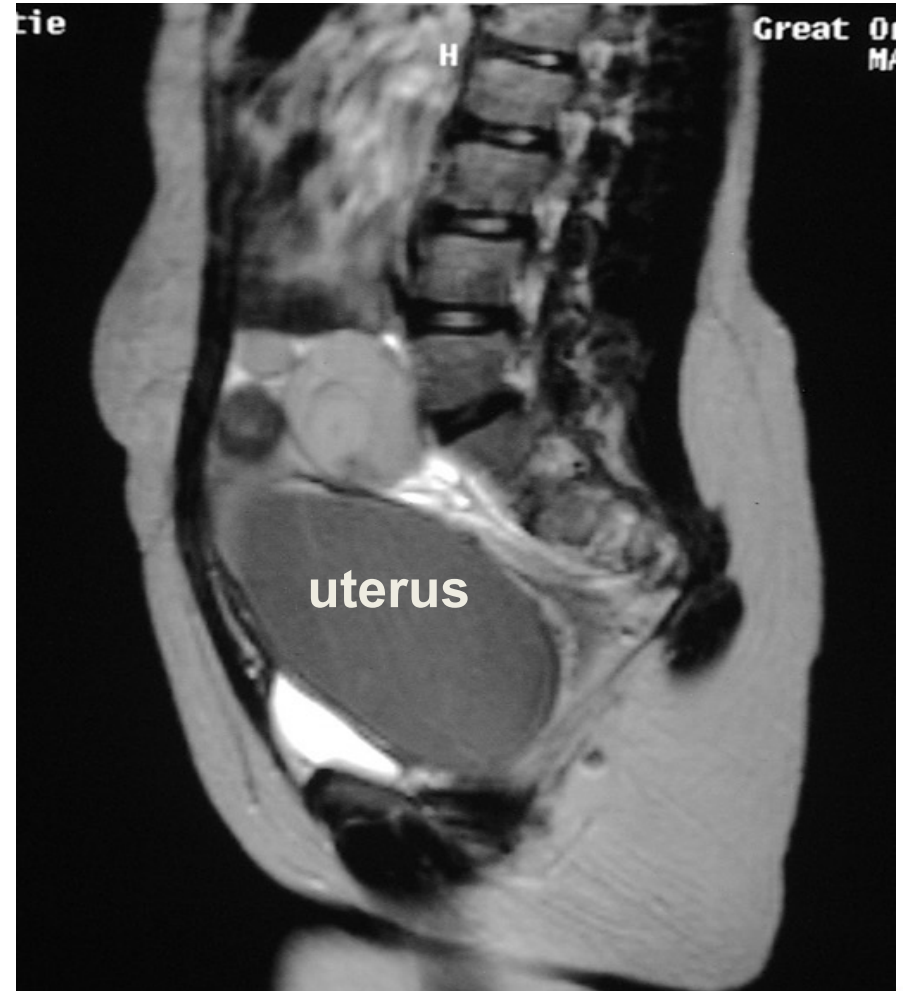
# Cloacal Outcome at Puberty

• Normal Menstruation	13	32%
• Hematometra	15	36%
• Vestigial uterus	8	20%
• Early puberty	3	7%
• Under investigation	2	5%

\* 6/10 diagnosed absent uterus at laparotomy developed uterine function

# Etiology of Obstructed Uterus

Vaginal Stenosis	3
post surgery	
Urogenital sinus	11
(no previous surgery)	
Cervical Stenosis	1



# Adult Outcome - Cloaca

21 women aged 17-32 (mean 24) years

- Sexually active 12 (57%)
- Adequate Vagina 6 (29%)

(6/18 additional adult vaginal surgery)

No pregnancies to date

# Follow up

- At least until potty trained or socially continent
- Renal function
- Bladder function
- Menstruating normally

# Why worry about Urology

- Incontinence
- Chronic Kidney Disease
- Genital anomalies
- Sexual dysfunction

# Finally when to call Urology

- Pre reconstruction
  - Any thing but a simple anomaly
  - Sacral anomalies (tethered cord)
  - Upper tract anomalies
  - GU anomalies
  - UTI's
- Post reconstruction
  - UTI's
  - Incontinence
- When in doubt