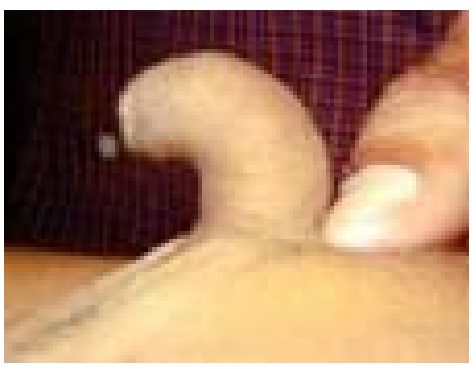
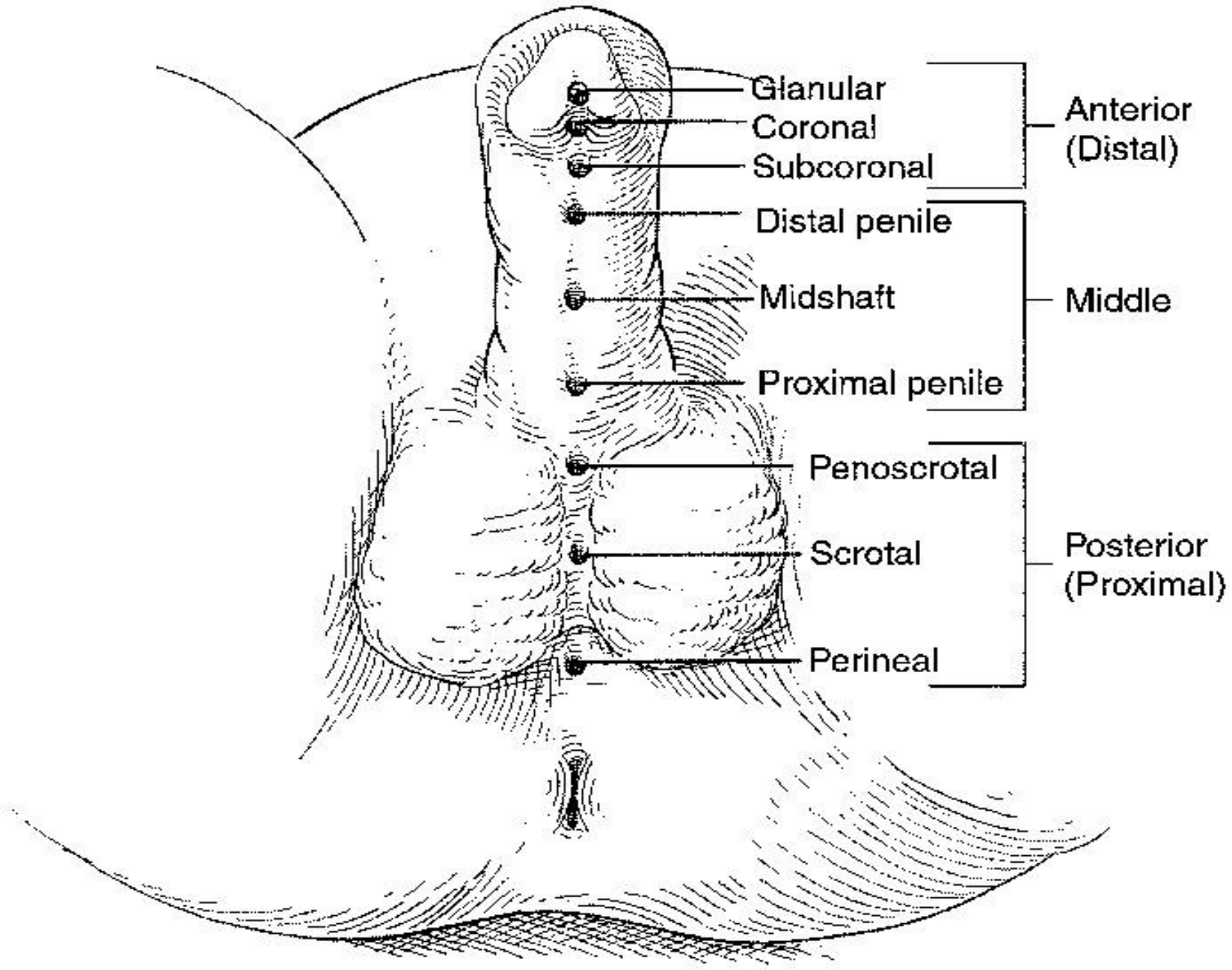
Hypospadias, Epispadias and Posterior urethral valves

Department of Urology

Hypospadias

- Any condition in which the meatus occurs on the undersurface of the penis
- Usually 3 features
  - **ventral meatus**
  - **ventral curvature (chordee)**
  - **Dorsal "hood“; deficient foreskin ventrally**





Embryology

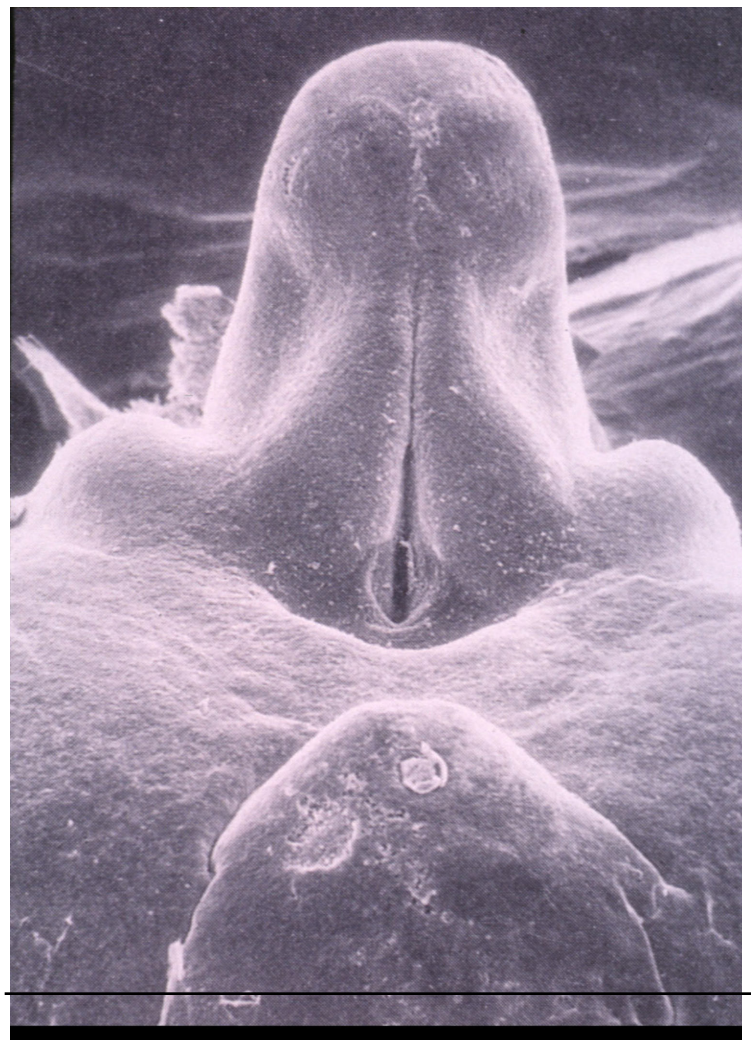
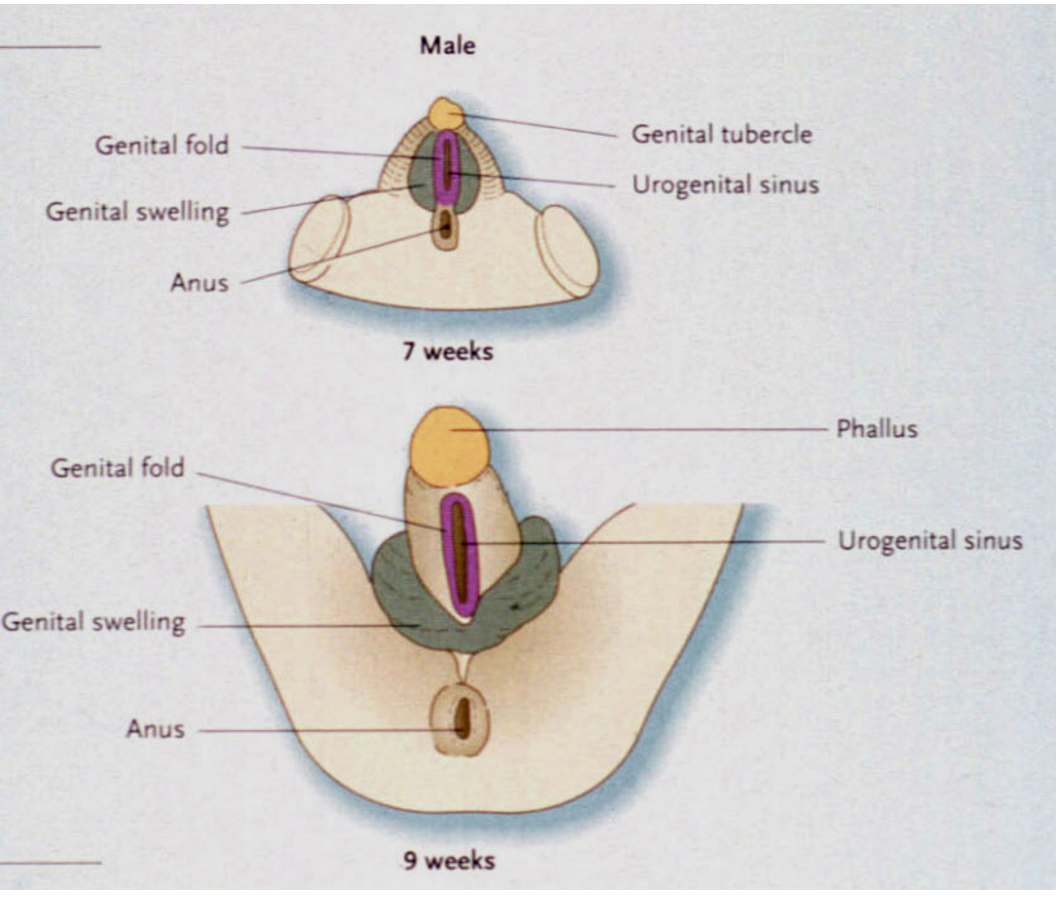


Figure 15-33

- Genital tubercle fuses in midline
- Mesodermal folds create the urethral and genital folds
- coalesce in midline as phallus elongates
- Distal glans channel tunnels to proximal urethra as solid core then undergoes canalization

Embryology



Prepuce forms as ridge of skin from corona

Hypospadias

- Failure of ventral aspect to form
- Dorsal hood

Chordee

- Differential growth between normally developed dorsal tissue and underdeveloped ventral corporal tissue
- Fibrous tissue distal to hypospadiac meatus

Incidence

- 1:300 live male births
- 6000 boys each year in the US
- Some genetic component
  - 8% of patients have father with hypospadias
  - 14% of patients have male siblings with hypospadias
  - If child with hypospadias, risk to next child
    - 12% risk with negative family history
    - 19% if cousin or uncle with hypospadias
    - 26% if father or sibling
- More common in Caucasians (Jews and Italians)
- Higher incidence in monozygotic twins (8.5x)

Associated Anomalies

- Undescended testes 9% and inguinal hernia 9%
- Upper tract anomalies rare (1-3%)
- *Utriculus masculinus*
  - 10 to 15% in perineal or penoscrotal hypospadias
  - Incomplete mullerian duct regression

Associated Anomalies

- Rule out intersex, especially with cryptorchidism
  - Adrenogenital syndrome
  - Mixed gonadal dysgenesis
  - Incomplete pseudohermaphroditism
  - True hermaphrotidism

Associated Anomalies

- **hypospadias** and **cryptorchidism**
  - high index of suspicion for an intersex state
- Walsh reported the incidence of intersexuality in children with cryptorchidism, hypospadias, and otherwise nonambiguous genitalia to be 27%
  - **nonpalpable testis were at least threefold more likely to have an intersex condition than those with a palpable undescended testis (50% versus 15%**)

Further Evaluation

- Only with severe hypospadias and sexual ambiguity
  - Includes testicular abnormalities
  - Up to 25% of these patients have enlarged utricles or other female structures
- The incidence of abnormalities with other forms of hypospadias approximates that of the general population
  - Therefore no further evaluation is indicated

Treatment

- Meatoplasty and glanuloplasty
  - Multiple techniques
- Orthoplasty
  - Utilize artificial erection
  - Release urethra from fibrous tissue
  - Plicate dorsal tunica albuginea
  - Ventral graft if needed

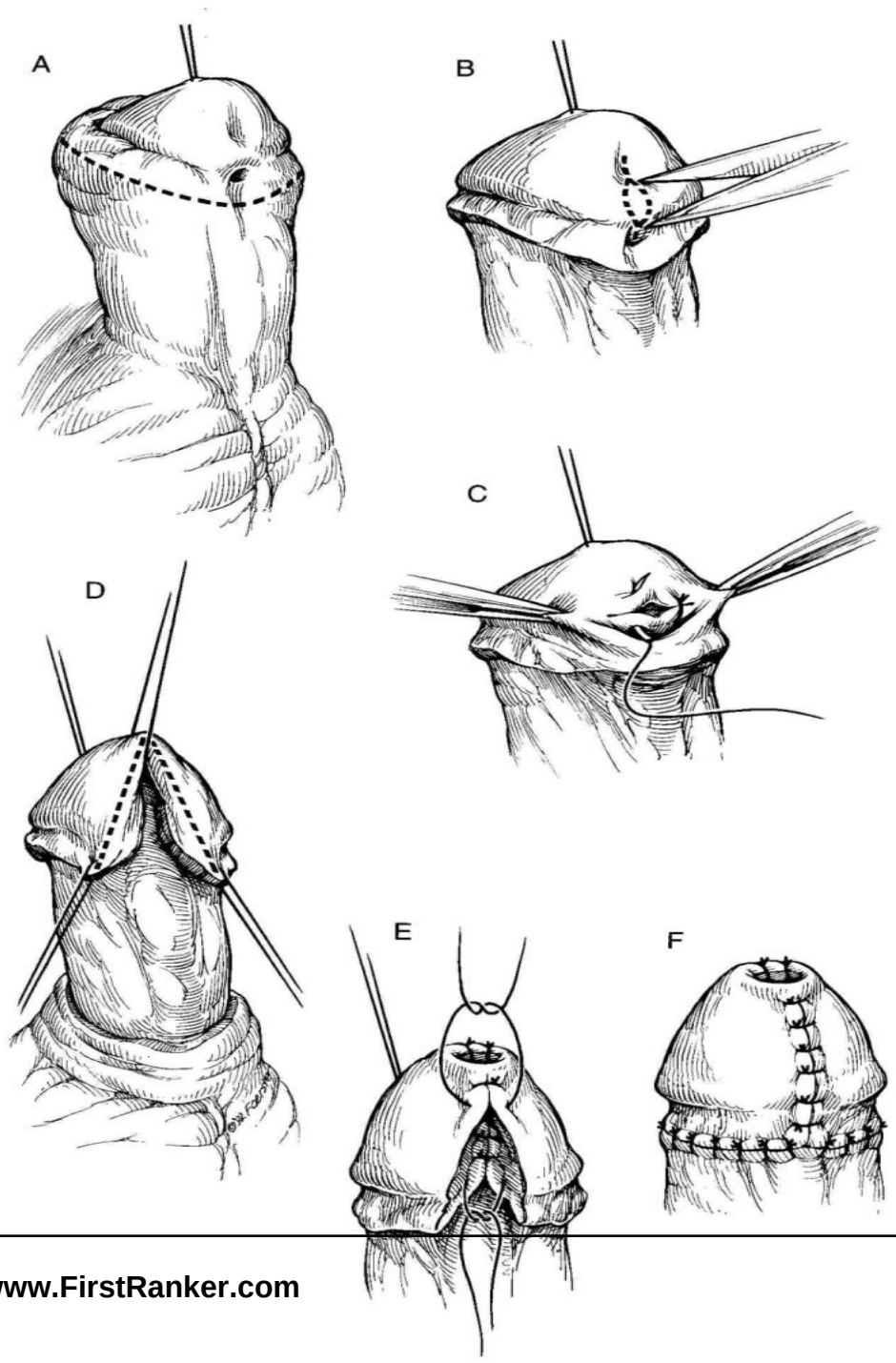
Treatment

- Urethroplasty
  - Onlay vascularized flap
  - Tubularized flap
  - Free graft
- Skin cover
  - Mobilized dorsal prepuce and penile skin
  - Double faced island flap
- Scrotoplasty

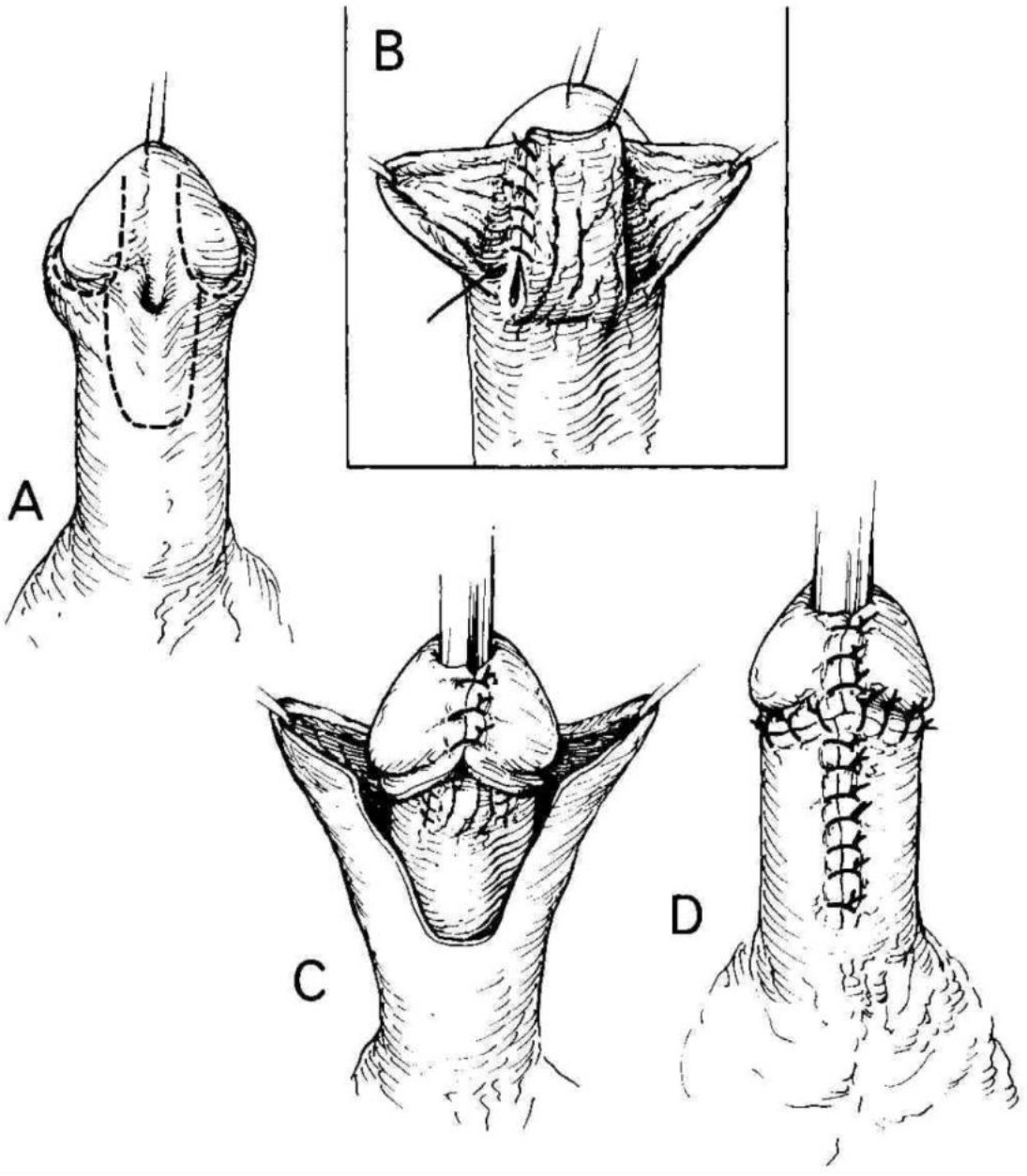
Hypospadias Repair

- Over 150 operations have be described
- Distal hypospadias
  - Tubulization of the incised urethral plate (Snodgrass)
  - Meatal advancement (MAGPI)
  - Meatal-based flaps (Mathieu)
- Proximal hypospadias
  - Onlay grafts
  - Vascularized inner preputial transfer flaps (Duckett)
  - Free grafts (skin, buccal mucosa)

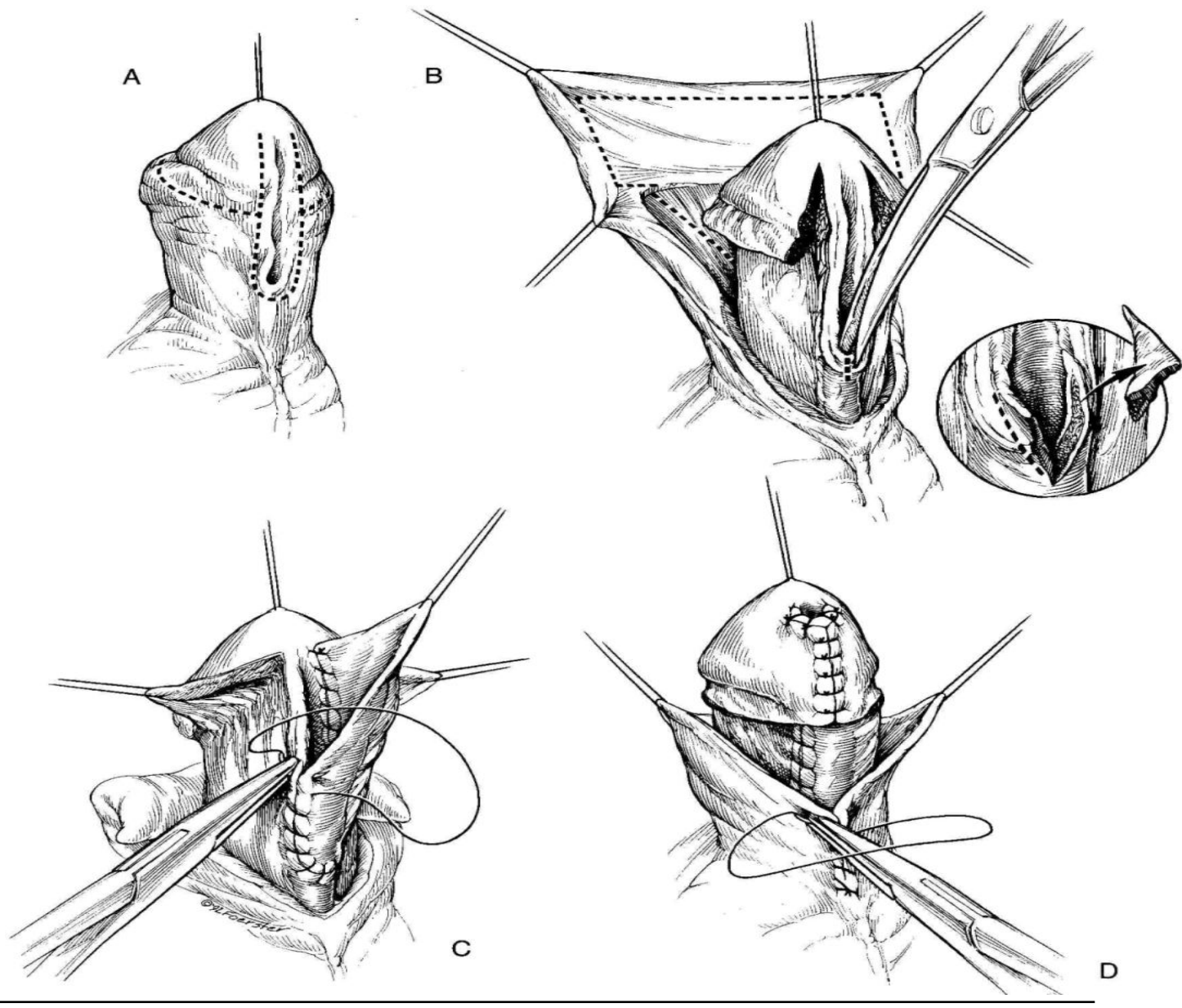
MAGPI



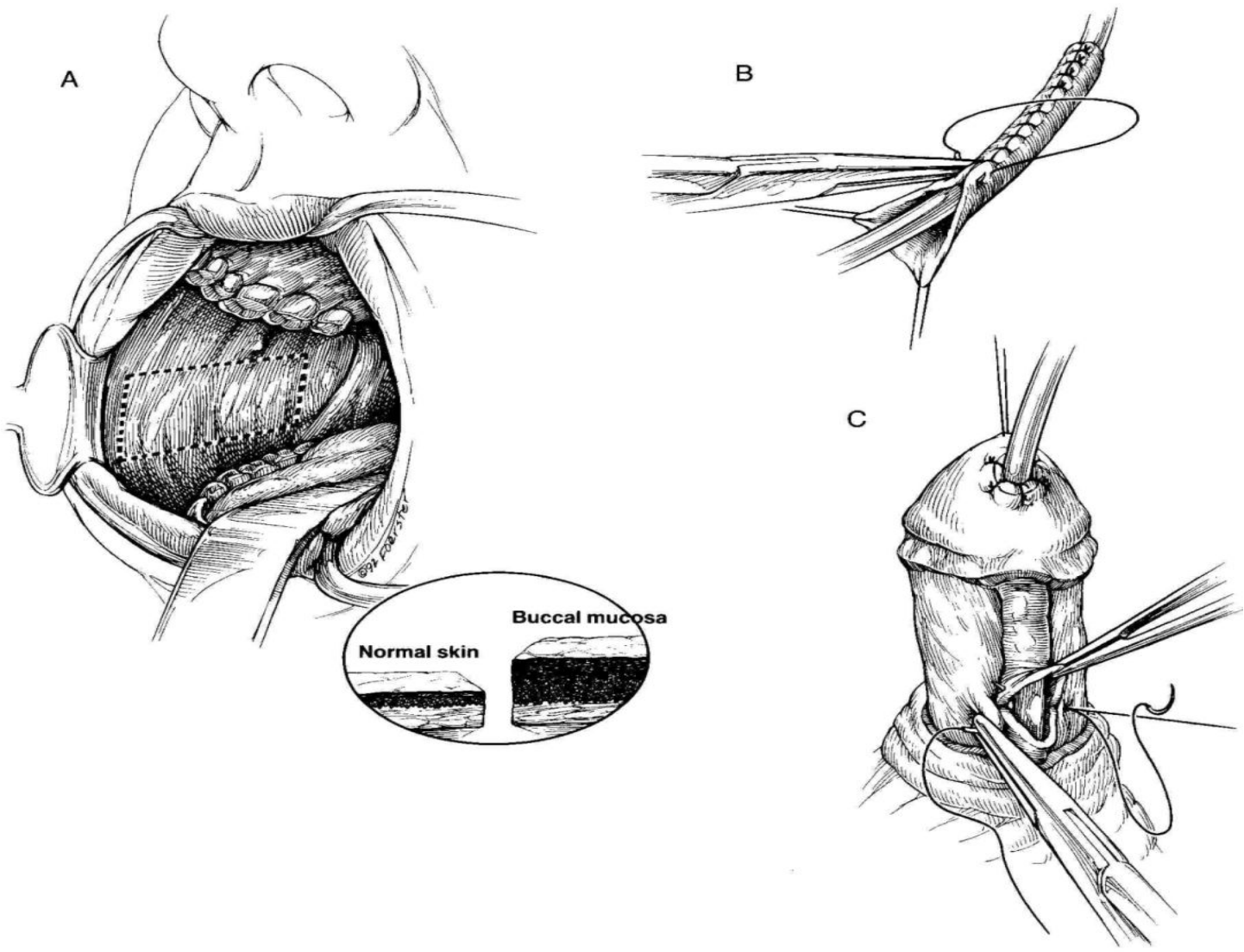
Mathieu



Island Onlay



Buccal Mucosal Graft



Epispadias

Introduction

- An **epispadias** is a rare type of malformation
- of the penis in which the urethra ends in an opening on the upper aspect (the dorsum) of the penis.
- It can also develop in females when the urethra develops too far anteriorly

Etiology

- It is a partial form of a spectrum of failures of abdominal and pelvic fusion in the first months of embryogenesis known as the **exstrophy - epispadias complex.**
- It occurs as a result of defective migration of the genital tubercle primordii to the cloacal membrane, and so malformation of the genital tubercle, at about the 5th week of gestation.

Associated anomalies

- The causes of epispadias are not known. It may occur because the pubic bone does not develop properly.
- Epispadias can occur with bladder exstrophy. In this rare birth defect, the bladder is inside out and sticks through the abdomen wall. Epispadias can also occur with other birth defects.

Presentation

- Most cases involve a small and bifid penis, which requires surgical closure soon after birth, often including a reconstruction of the urethra. Where it is part of a larger Exstrophy, not only the urethra but also the bladder (bladder exstrophy) or the entire perineum (cloacal exstrophy) are open and exposed on birth, requiring closure.

Presentation.....

- Males usually have a short, wide penis with an abnormal curve. The urethra usually opens on the top or side of the penis instead of the tip. However, the urethra may be open along the whole length of the penis.
- Females have an abnormal clitoris and labia. The opening is usually between the clitoris and the labia, but it may be in the belly area. They may have trouble controlling urination (urinary incontinence).

Examination

- Abnormal opening from the bladder neck to the area above the normal urethra opening
- Backward flow of urine into the kidney (reflux nephropathy)
- Urinary incontinence
- Urinary tract infections
- Widened pubic bone

Treatment

The main treatment for isolated epispadias is a comprehensive surgical repair of the genito-urinary area usually during the first 7 years of life, including **reconstruction of the urethra, closure of the penile shaft and mobilisation of the corpora**.

modified Cantwell-Ransley approach

- The most popular and successful technique is known as the **modified Cantwell-Ransley** approach.

complete penile disassembly technique

- In recent decades however increasing success has been achieved with the **complete penile disassembly** technique despite its association with greater and more serious risk of damage

Complications

- Recurrence
- Sexual problems
- Incontinence
- UTI
- Infertility
- Psycho-social stress

Posterior Urethral Valves

Posterior Urethral Valves (PUV)

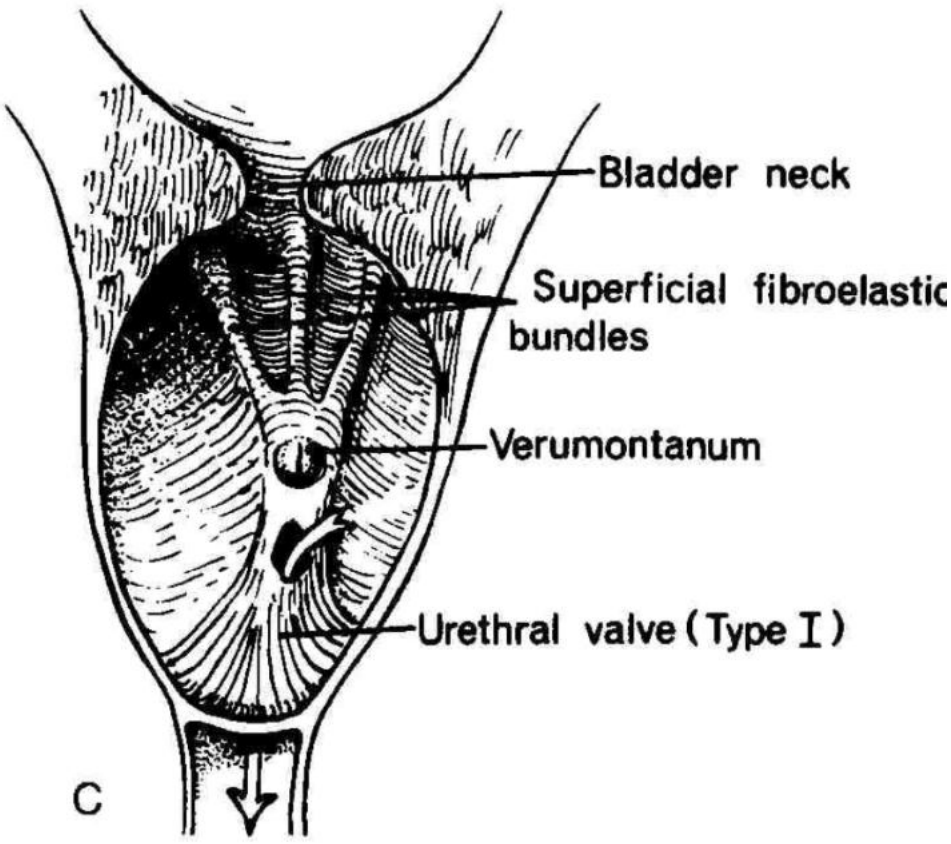
- Congenital Proximal Urethral Obstruction
- Abnormal congenital mucosal folds in the prostatic urethra that look like a thin membrane that impairs bladder drainage

PUV Defined

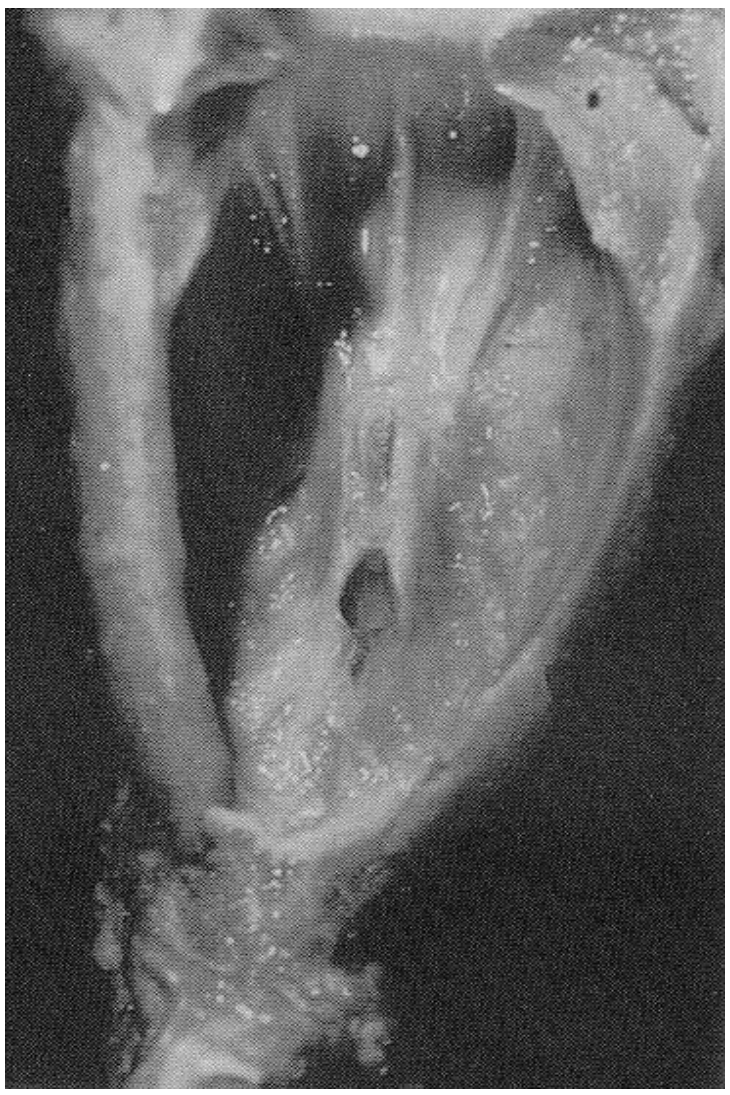
- Type I
  - Obstructing membrane that extends distally from each side of the verumontanum towards the membranous urethra where they fuse anteriorly
- Type II
  - Described as folds extending cephalad from the verumontanum to the bladder neck
- Type III
  - Represent a diaphragm or ring-like membrane with a central aperture just distal to the verumontanum
  - Thought to represent incompelte dissolution of the urogenital membrane

Type I PUV

- Obstructing membrane radiating distally from the posterior edge of the verumontanum to the membranous urethra
- During voiding, the fused anterior portion bulges into the urethra with a narrow posterior opening
- Possibly due to anomalous insertion of the mesonephric ducts into the primitive fetal cloaca

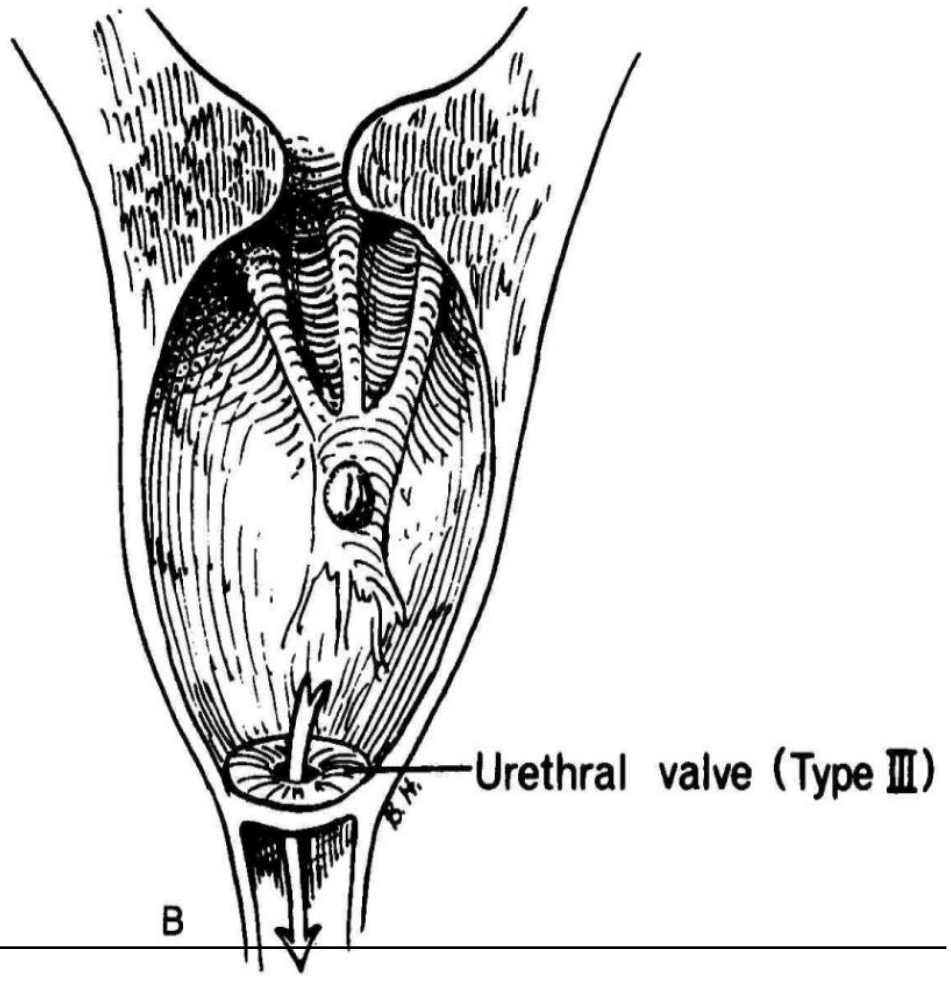


Type I PUV



Type III PUV

- Represent incomplete dissolution of the UG membrane
- Distal to the verumontanum at the membranous urethra
- Ring-like with a central opening, “wind sock valve”



Incidence

- Males only
- 1:5000 – 8000 male births
- Type I > 95%
- Type III - 5%
- Children with Type III PUVs have a worse prognosis as a group
- 50% of patients with PUV will have vesicoureteral reflux
  - 50% unilateral, 50% bilateral

Clinical Presentation

- Varies by degree of obstruction
  - Symptoms vary by age of presentation
- Antenatal
  - Bilateral hydronephrosis
  - Distended and thickened bladder
  - Dilated prostatic urethra
  - Oligohydramnios - accounts for co-presentation of pulmonary hypoplasia.

Clinical Presentation

- Newborn
  - Palpable abdominal mass
    - Distended bladder, hydronephrotic kidney
    - Bladder may feel like a small walnut in the suprapubic area
  - Ascites
    - 40% of time due to obstructive uropathy
  - History of Oligohydramnios
  - Respiratory distress from pulmonary hypoplasia
    - Severity often does not correlate with degree obstruction
    - Primary cause of death in newborns

Clinical Presentation

- Early Infancy
  - Dribbling / poor urinary stream
  - Urosepsis
  - Dehydration
  - Electrolyte abnormalities
  - Uremia
  - Failure to thrive; due to renal insufficiency
- Toddlers
  - Better renal function (less obstruction)
  - Febrile UTI
  - Voiding dysfunction – incontinence
  - Daytime incontinence may be the only symptom in boys with less severe obstruction

Initial Management

- Bladder Drainage
  - A 5 or 8 Fr pediatric feeding tube is ideal
  - A Foley catheter should not be used, due to the tendency of the balloon to occlude the ureteral orifice and cause a bladder spasm.
    - Secondary obstruction
  - Broad spectrum antibiotic coverage
  - Metabolic panel
    - Assess renal function and metabolic abnormalities
    - Acidosis, hyperkalemia common problems

Radiologic Evaluation of the Lower Tract

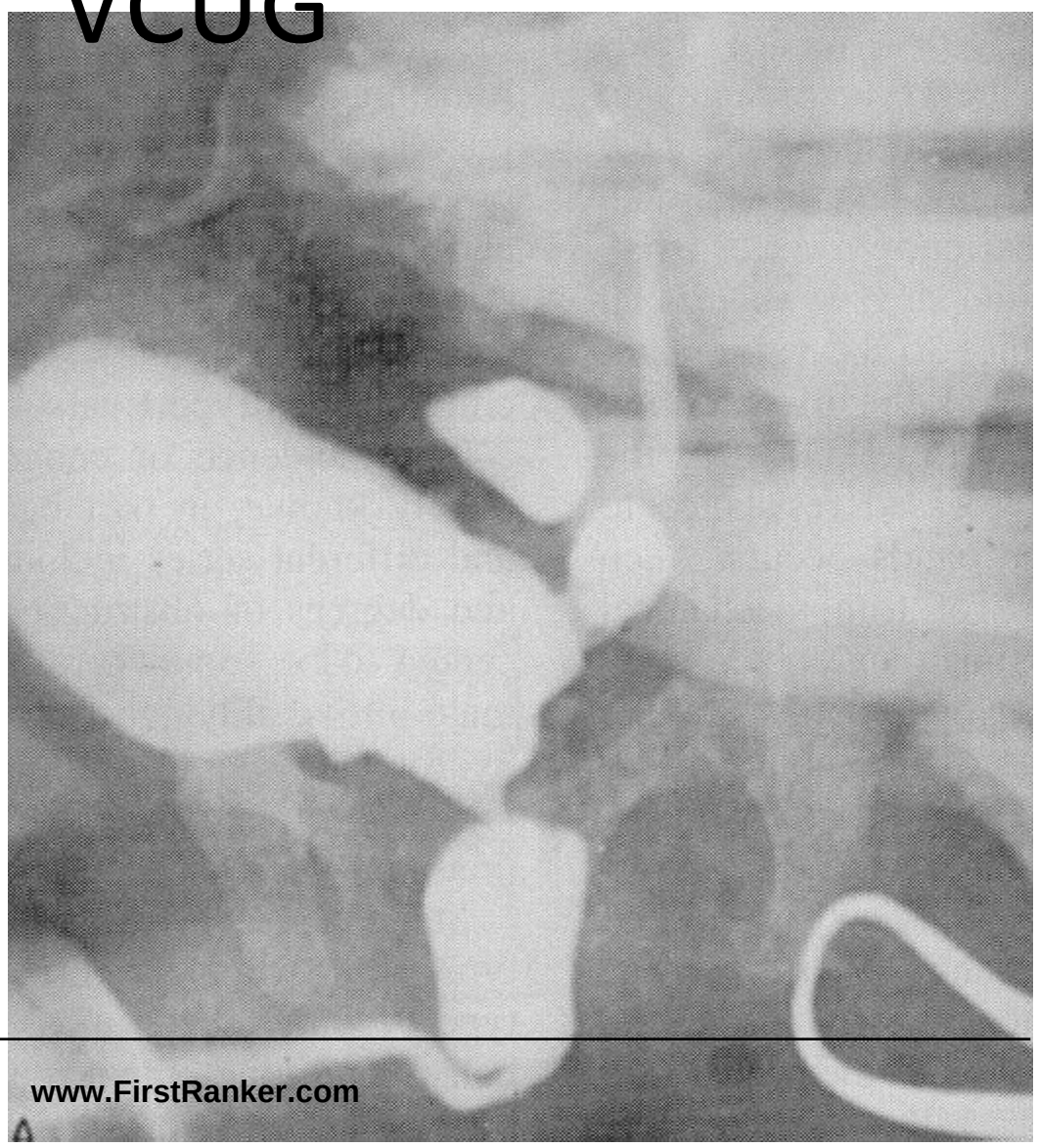
- VCUG
  - Mandatory for all PUV evaluations
  - Showing a dilated prostatic urethra, valve leaflets, detrusor hypertrophy, bladder diverticula, bladder neck hypertrophy, and narrow penile urethra stream, as well as possible incomplete emptying

Radiologic Evaluation of the Lower Tract

- U/S
  - Examining the prostatic urethra for characteristic dilation and thickening of the bladder wall

VCUG

- dilated prostatic urethra
- valve leaflets
- detrusor hypertrophy
- cellules or bladder diverticula
- bladder neck hypertrophy
- narrow penile urethra stream
- possible incomplete emptying



Radiologic Studies- Upper Tract

- Renal Ultrasound
  - Examination for bilateral hydronephrosis and signs of lower tract obstructive process
- Renal Scan
  - Assesses the function of the kidneys

Management

- Transurethral Valve Ablation
  - Incise at 4, 8 & 12 o'clock positions via Pediatric resectoscope
    - Avoid urethral sphincter
    - Catheter drainage for 1-2 days
    - VCUG at 2 months to ensure destruction of valves
    - Regular U/S to evaluate resolution of hydronephrosis

Management

- Transurethral Valve Ablation
  - Alternatively, 8F cystoscope with a Bugbee electrode adjacent
  - Insulated crochet hook (“Whitaker hook”)
    - When urethra too small to accommodate cystoscope/Bugbee

Vesicoureteral Reflux

- Present in 33 - 50%
- Usually Secondary
  - High intravesical pressures
- 33% resolve spontaneously when obstruction treated
- 33% do well on prophylactic antibiotics

Vesical Dysfunction

- 50% have abnormal bladder function
- Presents as incontinence
  - Not due to sphincter dysfunction or damage
- Primary myogenic failure
- Uninhibited contractions
- May lead to progressive renal deterioration

Favorable Prognostic Factors

- Creatinine falling below 1.0 one month after treatment initiated
- Absence of VUR
- Preservation of the corticomedullary junction of the kidneys by renal U/S
- Radiologic evidence of a “pop-off” valve