Hypospadias: What the Primary Care Provider Should Know

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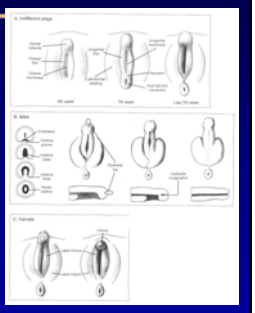
What Is Hypospadias?

- Penile developmental anomaly
  - Hooded prepuce
  - Ventral penile curvature (chordee)
  - Ventral urethral meatus

Embryology of the External Genitalia

- Genital tubercle, Urogenital folds
- Dihydrotestosterone induces elongation, coalescence urogenital folds
- Distalmost urethra formed by invagination from tip of glans
- Foreskin fuses ventrally



Prevalence of Hypospadias

- Metropolitan Atlanta Congenital Defects Program--Severe Hypospadias
  - 0.11 per 1000 live births in 1968
  - 0.27 to 0.55 per 1000 live births in 1990 to 1993
- Birth Defects Monitoring Program--All hypospadias:
  - 2.02 per 1000 live births in 1970
  - 3.97 per 1000 live births in 1993

Paulozzi LJ: Is hypospadias an "environmental" birth defect. (Special issue: The impact of the environment and endocrine disruptors on pediatric urology.) Dial Pediatr Urol 2000;23:8.

Why Is Prevalence Increasing?

- Increased reporting mild forms
  - Ratio of mild:severe is not increasing
- Impact of environmental "endocrine disruptors"
  - Phytoestrogens, DES, DDT, bisphenols
- Better surgical treatment of hypospadias allowing genes to be transmitted
- Better fertility treatment
  - Progesterone effect
  - Treatment of subfertile hypospadiacs

Myers JP: Endocrine disruption: Emerging science vitally important for pediatric urologists. (Special issue: The impact of the environment and endocrine disruptors on pediatric urology.) Dial Pediatr Urol 2000;23:8.

Etiology of Hypospadias

- Genetic: 20-25% have family history
  - Monozygotic twins may be discordant
- Hormonal
  - Hypogonadism
  - 5-alpha reductase deficiency
  - Androgen insensitivity
- Environmental
  - Progesterone/IVF variably associated with increased risk severe hypospadias

Associated Syndromes and Anomalies

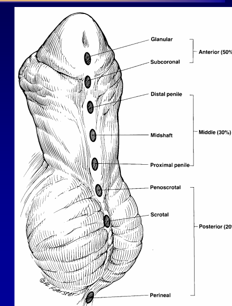
TABLE 65-1 -- Syndromes in Which Hypospadias is a Feature *

Syndrome (s.)	Relative or Approximate % of Cases with Hypospadias	Other Notable Findings of the Genitalia
Aniridia–Wilms tumor association	Frequent	Cryptorchidism
Deletion 4p s.	Frequent	Cryptorchidism
Deletion 11q s.	50%	Cryptorchidism
Deletion 13q s.	Frequent	Cryptorchidism
Fetal trimethadione s. (Tridione s.)	Frequent	Ambiguous genitalia
Fraser s. (cryptophthalmos s.)	Frequent	Cryptorchidism
Fryns s.	86%	Cryptorchidism, bifid scrotum
Opitz s. (hypertelorism-hypospadias s., Opitz-Frias s., Opitz oculo-genito-laryngeal s.)	Frequent	Cryptorchidism, bifid scrotum
Rapp-Hodgkin ectodermal dysplasia s. (hypohidrotic ectodermal dysplasia, autosomal dominant type)	Frequent	—
Rieger s.	Frequent	—
Schinzel-Giedion s.	100%	Micropenis, hypoplastic scrotum
Smith-Lemli-Opitz s.	70%	Micropenis, hypoplastic/bifid scrotum
Triploidy s. and diploid/triploid mixoploidy s.	Frequent	Micropenis, cryptorchidism
X-linked α-thalassemia/mental retardation (ATR-X) s.	Frequent	Micropenis, hypoplastic scrotum
Brachmann-de Lange s. (Cornelia de Lange s., de Lange s.)	33%	Cryptorchidism (73%)
Beckwith-Wiedemann s. (exomphalos-macroglossia-gigantism s.)	Occasional	Cryptorchidism
Craniofrontonasal dysplasia	Occasional	—
Dubowitz s.	Occasional	—
Duplication 4p s. (trisomy for the short arm of chromosome 4, trisomy 4p s.)	Occasional	Micropenis, cryptorchidism
Duplication 9p s. (trisomy 9p s.)	Occasional	Micropenis, cryptorchidism
Escobar s.	Occasional	Cryptorchidism
Duplication 10q s.	Occasional	Cryptorchidism, streak gonads
Fanconi pancytopenia s.	20%	Micropenis, cryptorchidism
Fetal hydantoin s. (fetal dilantin s.)	Occasional	Cryptorchidism, micropenis, ambiguous genitalia
Fetal rubella effects (fetal rubella s.)	Occasional	Cryptorchidism
Fetal valproate s.	Occasional	—
FG s.	Occasional	Cryptorchidism (36%)
Hay-Wells s. of ectodermal dysplasia (ankyloblepharon-ectodermal dysplasia-clefting s., AEC s.)	Occasional	Micropenis
Hydrolethalus s.	Occasional	—
Johanson-Blizzard s.	Occasional	Micropenis, cryptorchidism
Killian/Teschler-Nicola s. (Pallister mosaic s., tetrasomy 12p)	Occasional	—
Lenz-Majewski hyperostosis s.	Occasional	Cryptorchidism
Peters'-Plus s.	Occasional	Cryptorchidism
Levy-Hollister s. (lacrimo-auriculo-dento-digital s., LADD s.)	Occasional [hm1m](coronal)	—
Marden-Walker s.	Occasional	Micropenis, cryptorchidism
Multiple lentigines s. (leopard s.)	Occasional	Cryptorchidism, hypogonadism
Oto-palato-digital s., type II	Occasional	Cryptorchidism
Restrictive dermopathy	Occasional	—
Roberts-SC phocomelia (pseudothalidomide s., hypomelia-hypotrichosis-facial hemangioma s.)	Occasional	Cryptorchidism
Russell-Silver s. (Silver s.)	Occasional	Seminoma
Shprintzen s. (velo-cardio-facial s.)	Occasional	Cryptorchidism
Simpson-Golabi-Behmel s.	Occasional	Cryptorchidism
Townes-Brocks s.	Occasional	—
Trisomy 13 s. (D1 trisomy s.)	<50%	Cryptorchidism (>50%)
Trisomy 18 s.	<10%	Cryptorchidism (≥50%), bifid scrotum (<10%)
XXY s., Klinefelter s.	Occasional	Micropenis, cryptorchidism
XXXY s. and XXXXY s.	Occasional	Micropenis (80%), cryptorchidism (28%)
XYY s.	Occasional	Micropenis, cryptorchidism
Zellweger s. (cerebro-hepato-renal s.)	Occasional	Cryptorchidism

* Adapted from Jones KL: Smith's Recognizable Patterns of Human Malformation, 5th ed. Philadelphia, WB Saunders, 1997, p 835, with permission.

Classification of Hypospadias

- Incomplete foreskin
- Stretched penile length (?micropenis)
- Presence, degree of chordee (curvature)
- Location of urethral meatus
- Presence of testes



Megameatus Intact Prepuce

- Variant
- Intact prepuce--often discovered after circumcision
- No chordee

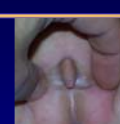
Evaluation of the Child With Hypospadias

- Mild hypospadias
  - No specific evaluation
- Severe hypospadias
  - Consider underlying endocrine abnormality
- Renal ultrasound not necessary
- One or both testes nonpalpable:
  - Consider intersex state

Hypospadias vs. Intersex

- Bilateral descended gonads
  - Intersex unlikely
  - Consider androgen insensitivity, 5-alpha reductase deficiency
- Unilateral nonpalpable gonad
  - Must consider mixed gonadal dysgenesis
- Bilateral nonpalpable gonads
  - MUST consider 46XX with CAH

Hypospadias vs.. Intersex

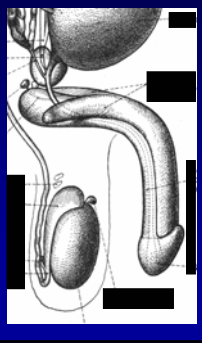

Goals of Surgical Therapy

- Straighten penis (chordee correction)
- Relocate urethral meatus to tip of glans
- Cosmetic penile skin coverage
- Repair scrotal anatomy



Surgical Anatomy

- Corpora Cavernosa--erectile bodies
- Corpora Spongiosum--urethral body
- Neurovascular bundle runs dorsally under buck's fascia, which surrounds all corpora



Now let's get this straight…

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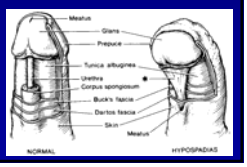
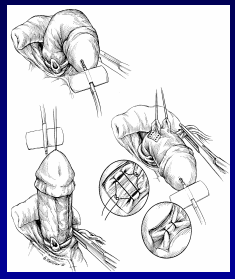
Chordee Correction

- Most chordee is corrected by degloving penis at the level of Buck's fascia
  - Dorsal neurovascular bundle is under Buck's
- Often thick chordee tissue under ventral penile skin
- When chordee is intrinsic to corpora, 2 options
  - Dorsal tucks
  - Ventral grafting

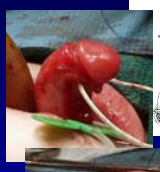
Dorsal Plication of the Corpora

- To preserve urethral plate
- Several techniques
- Avoid neurovascular bundles by dissecting them, with Buck's fascia, off of dorsal corpora
- Test by artificial erection




Ventral Grafting of the Corpora

- Requires division of urethral plate
- Dermal graft or tunica vaginalis--now SIS
- Artificial erection to prove effect
- Allow some redundancy for graft shrinkage

Urethral Reconstruction

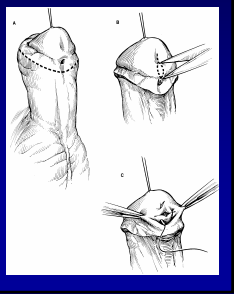
- Can be divided into two general techniques:
  - Urethral plate intact
  - Urethral plate divided
- Within those groups there are:
  - Grafts
  - Flaps
  - Tubularization techniques
  - Urethral mobilization

Intact Urethral Plate Repairs

- Generally for milder hypospadias
- Can be extended to proximal shaft, occasionally scrotal
- Have advantage in most of avoiding circumferential anastomosis
  - Decreased risk of stricture
  - Technically more straightforward

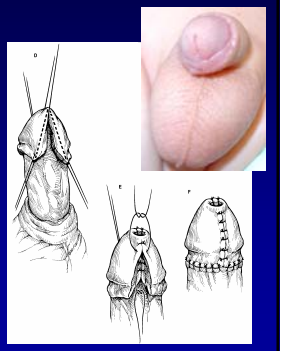
Distal Hypospadias Repair-- The MAGPI

- **M**eatal **a**dvancement and **g**lans**p**lasti
- Only applicable to glanular hypospadias
- "Sleight of hand" technique
- Step 1: advance dorsal urethra



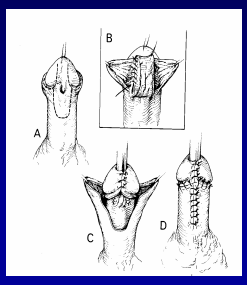
MAGPI--continued

- Second step: glansplasty to "advance" ventral urethra
- Most common complication is meatal regression



Meatal Based Flaps

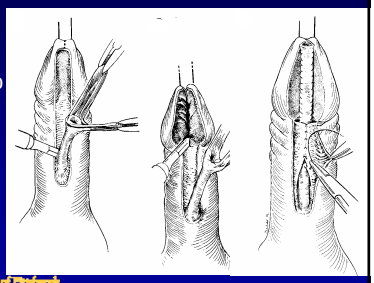
- "Flip-flap", Matthieu, Ombredanne, etc.
- Flap is not separated from SQ tissue
- Midshaft and beyond
- Simple technique applicable to many situations



Mathieu P: Traitement en un temps de l'**hypospadias** balanique ou juxtabalanique. J Chir 1932;39:481-486.

Urethral Mobilization

- Advance urethra to normal position
- Often applicable to distal reoperative procedures
- Presumably decreased risk fistula



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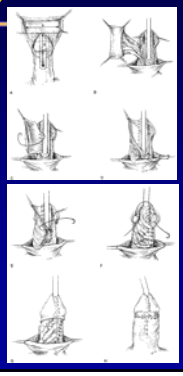
Dorsal Island Onlay

- Dorsal inner prepuce harvested on pedicle
- Used to create ventral aspect of urethra
- Previously, the most common repair of mid- to distal hypospadias



Elder JS, Duckett JW, Snyder HM: Onlay island flap in the repair of mid and distal penile **hypospadias** without chordee. J Urol 1987;138:376-379.

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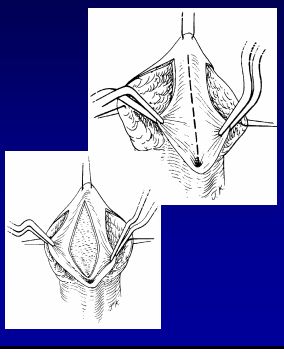
The "Snodgrass" Urethroplasty

- "Unhinging" incision midline urethral plate
- Primary tubularization
- Generally used for distal hypospadias
- Obviates need for flap or graft



Snodgrass W: Tubularized, incised plate urethroplasty for distal **hypospadias**. J Urol 1994;151:464-465.

The Snodgrass Urethroplasty

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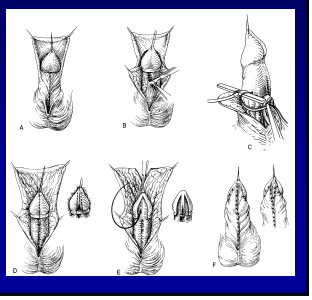
Dividing the Urethral Plate

- Short urethra prevents penile straightening
- Generally in severe forms hypospadias
- Can be divided into two techniques
  - Two stage repairs
  - One stage repairs

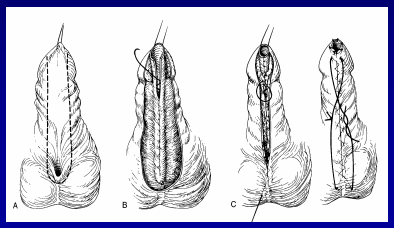
First Stage Hypospadias Repair--Chordee Correction

- Penis is degloved
- Urethral plate is split
- Chordee is corrected
- Prepuce is transposed to ventral penis for future urethroplasty (6 months later)



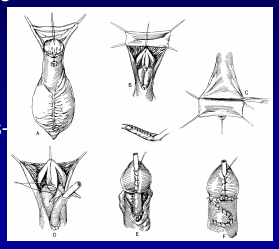
Second Stage Hypospadias Repair

- Modified Thiersch-Duplay tube



Horton-Devine Free Prepuce Graft

- First major one-stage repair
- Principles of full-thickness skin grafting
- Prepuce is hairless-ideal for urethral reconstruction



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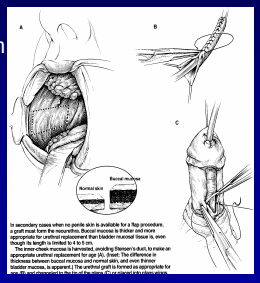
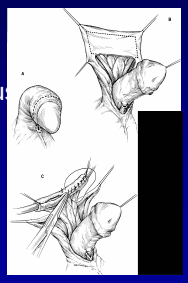
Buccal Mucosa Graft

- Currently non-skin graft of choice
- Can be obtained from cheek or inner lower lip or can extend throughout
- Reported success is high, lower in tubed repairs



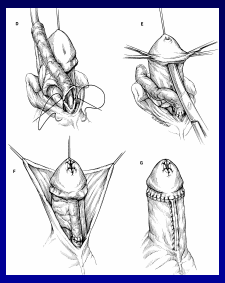
Transverse Island Pedicle Flap

- Has supplanted free grafts in most cases
- Inner prepuce is freed, along with subcutaneou pedicle
- Flap is tubularized to appropriate length



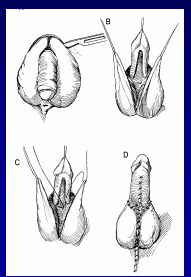
Transverse Island Pedicle Flap 2

- Suture line is placed against corpora
- Pedicle can cause penile torsion if too short
- Tunneling through glans is not preferred--split glans and raise flaps



Correction of Scrotal Transposition

- Bilateral rotation flaps with excision of tethering midline raphe
- Creates circumferential incision at base of penis--may compromise vascularity of island flap urethroplasty



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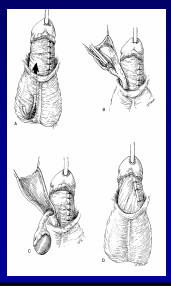
Buttressing Layers

- Extra coverage for repair using:
  - Pedicle to transverse island flap (with or without use of skin)
  - De-epithelialized skin flap
  - Tunica vaginalis--especially in cases requiring scrotoplasty



The SCUFF Procedure

- Variation on the theme of buttressing layers
- For megameatus, distal failed hypospadias

Docimo, S.G.: Subcutaneous Frenular Flap (SCUFF) for Megameatus and Reoperative Hypospadias Repair, Urology, 2001

SCUFF Technique

Radical Mobilization of the Bulbar Urethra

- In severe hypospadias:
  - Can dissection result in “less severe” hypospadias?
  - Can chordee be repaired without dividing the urethral plate?

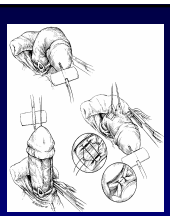
Baker, L.A., Mathews, R., **Docimo, S.G.:** Radical bulbar dissection to correct severe chordee and proximal hypospadias., J. Urol. 2000

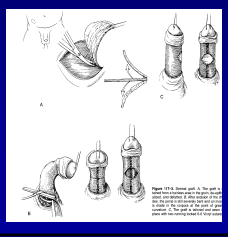
The Problem:

- Proximal hypospadias, scrotal or perineal with severe chordee
- One stage or two?

The Standard Approach

- Deglove penis
- If chordee severe:
  - Dorsal tucks
  - Divide urethral plate
    - Transverse Island Tube
    - Two-stage repair





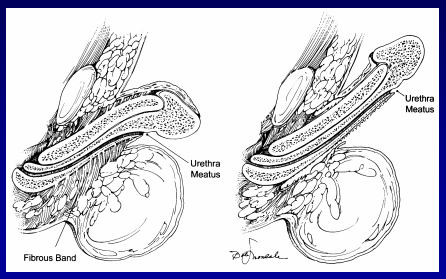
The Challenge

- Can urethral plate be preserved even with severe chordee for Snodgrass or onlay repair?
- Is the anatomy causing chordee also proximal to meatus?

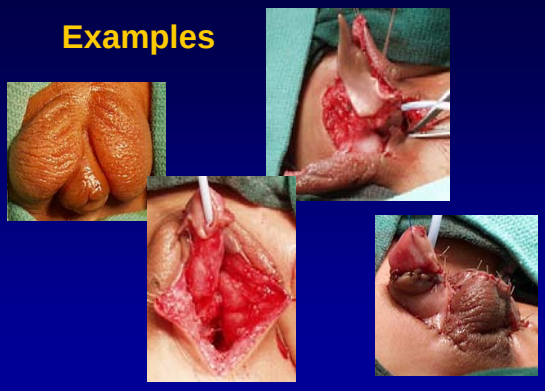
Technique

- Penis is degloved
- Dissection carried proximal to hypospadiac meatus
- Tethering bands to urethra and corpora divided in perineum, releasing proximal ventral shaft
- Can dissect to/beyond perineal body

Principle



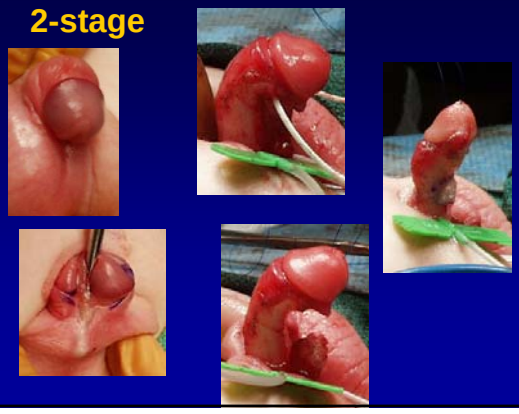
Examples



Examples

Examples

2-stage



Postoperative Care

- Most cases done as outpatient, except free grafts
- May have urethral stent draining into diaper
- Simple tegaderm dressing
- Stent in for 2 to 14 days, depending on repair
- Older children or adults may rarely need suprapubic tube, other form of drainage

Complications

- Bleeding
- Infection
- Persistent chordee
- Breakdown of urethroplasty
  - Complete
  - Urethrocutaneous fistula, 3-25% based on complexity
  - Meatal regression

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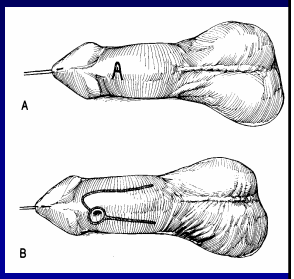
Principles of Fistula Repair

- Wait 6 months for tissues to heal
- Excise fistulous tract
- Close defect in urethra
- Advance or rotate skin flap and subQ tissue to give layered closure/offset suture lines



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Management of Hypospadias Failures

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Options for Failed Hypospadias

- Two-stage free graft (Bracka)
  - Remove urethra
  - Line ventral penis with buccal mucosa graft
  - 6 months later, roll into tube
- Must have full complement of “tricks” to deal with potential reconstructive dilemmas

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Miscellaneous Principles

- Avoid scrotal or proximal shaft skin in urethra--these will be hair bearing
- Testosterone stimulation may be used for microphallus or penis with little skin--rarely needed
- Most repairs can be done at 5-6 months of age, best if done by 18 months

Summary

- Hypospadias consists of three defects:
  - Urethral meatus
  - Chordee
  - Hooded prepuce
- Etiology may be hormonal; usually multifactorial in normal boy
- Chordee correction:
  - Radical mobilization bulbar urethra
  - Dorsal tucks
  - Divide urethral plate and graft corpora

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Summary (cont)

- Urethral repairs
  - Intact plate
    - Tubularization (Snodgrass)
    - Flaps (Matthieu, onlay)
    - Free grafts
  - Divided plate
    - Dorsal island flap tube (Duckett)
    - Two-stage (Retik)

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Summary (cont.)

- Buttressing layers
  - Dorsal flap
  - Tunica vaginalis flap
  - SCUFF flap (megameatus, redo)

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The Goal:

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