# The Exstrophy Complex

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# Introduction

- **Rare** anomalies (?1:10,000 combined)
- Spectrum of defect in **infraumblical ventral wall**
- **may also involve** urinary, genital, musculoskeletal, GI and neurologic
- Reconstruction requires **multiple surgeries**
- remains one of the **greatest challenges** in pediatric surgery
- Only one paper from Ethiopia…

## *Belachew Dejene, **Bladder Exstrophy management in Tikur Anbessa University Hospital Addis Ababa, Ethiopia**, 2013, Ethiop Med J.*

- **Study period** = 5 years (2007-2012)
- **Sample size** = 46 patients
- **M:F** = 1.7:1
- **Age** = late presentation (> 4years) in 50%
- **Surgery** = staged repair > diversion, ?? No osteotomy
  - **67% (31) staged repair** (all patients < 4 years, plus some older children)
  - **33% (15) diversion** (Mainz II pouch ureterosigmoidostomy)
- **Outcome** = too early to assess (59% (27) of staged still waiting for BNR)
  - Ureterosigmoidostomy = 1 developed fecal incontinence, 1 developed recurrent UTI
  - Staged repair = 1 converted to uretrosigmoidostomy after BNR b/c failed repair

# Outline

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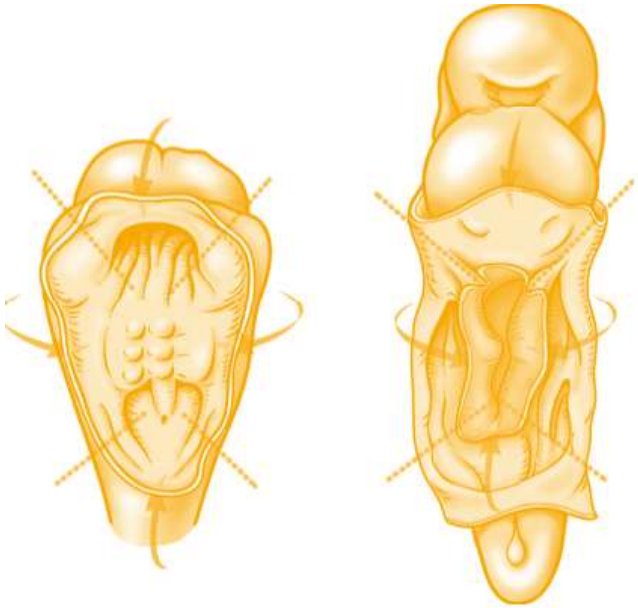
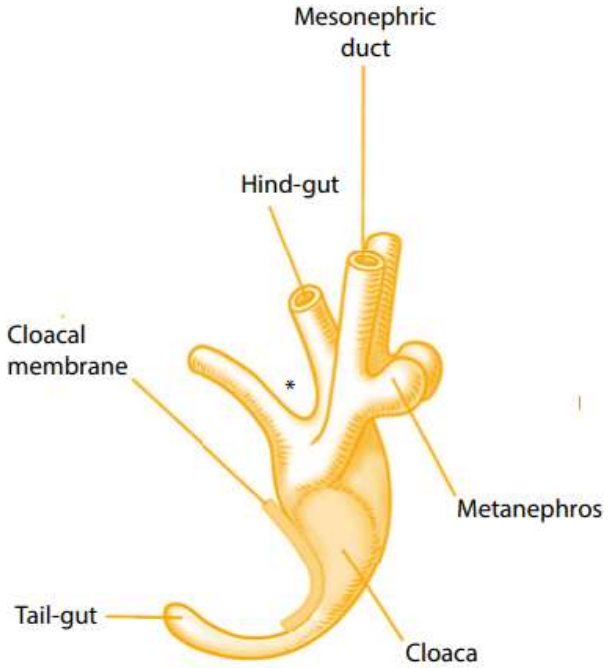
# 1- Embryologic basis

# Normal development

- **4th wk –** lateral body wall (mesoderm) folds**,** centering on future umblical ring. cloacal membrane (infraumblical abd, wall) is bilaminar (ectoderm & endoderm)
- 5th wk **–** cloaca divided by urorectal septum
- **6th wk** – secondary mesoderm (from primitive streak behind cloaca) migrate between ectoderm and endoderm layers of cloacal membrane
- 7th wk - urorectal septum reaches cloacal membrane leading to perforation
- **8th wk** – bladder arises from ventral expansion of urogenital sinus.
- **7th wk -12th wk** – secondary mesoderm enforced by invading somatic mesoderm
- By 12th wk - genital tubercles fuse and urethral folds close over urethral plate

# Normal development






- **Ventral view** of human embryo showing the direction of the four infoldings. And **lateral view** showing division of cloaca

# Theories

- Not an arrest in development (No similar stage in normal embryology)
- Premature rupture of cloacal membrane b/c not enforced with ingrowth of mesoderm
  - ?Limited migration = physical obstruction (cloacal overdevelopment, caudal positioning of genital tubercles or body stalk) , cellular dysfunction
  - ?loss of mesoderm = hypoxemic infarction, Genetic switch leading to senescence
- **timing of rupture determine the severity
  - $5^{th}$ wk = cloacal exstrophy
  - $7^{th}$ wk = classic bladder exstrophy
  - 10 -11 wk = epispadias

# Theories

| Year | Authors | Morphological derangement |
|---|---|---|
| 1952 | Patten and Barry | Caudal positioning of primordia of the genital tubercle relative to the cloacal membrane blocks mesodermal migration |
| 1962 | Marshall | Persistence of cloacal membrane creates wedge effect, blocking mesodermal ingrowth |
| 1964 | Muecke | Abnormally large cloacal membrane blocks medial mesodermal ingrowth |
| 1985 | Thomalla et al. | Ventral hernia of the lower abdominal wall induces exstrophy |
| 1988 | Mildenberger et al. | Caudal insertion of the body stalk obstructs medial mesodermal ingrowth |
| 1977 | Beaudoin et al. | Incomplete rotation of the pelvic ring primordia |
| 2005 | Manner and Kluth | Aneurysmal dorsal aortic swellings distends the pelvis and infraumbilical abdominal wall |
| 2005 | Stephens and Hutson | Embryonic tail compresses the second sacral mesodermal stream that segments the cloacal–allantoic connection and the phallus |
| 2008 | Kulkarmi and Choudari | Cephalad origin of the genital tubercle acts as a wedge to obstruct medial mesodermal ingrowth |
| 2011 | Stec | Cellular or mechanical factors disturb mesodermal ingrowth, yielding an abnormally formed cloacal membrane that prematurely ruptures |
| 2015 | Satish Kumar et al. | Persistent pubic diastasis causes levator maldevelopment as a hammock that pushes the bladder outward and exstrophic, while the lengthening extruded midgut loop returns to the abdomen through the umbilical ring, raising intra-abdominal pressure |

# Animal Models

- Chick model (1964)
  - Inducing early **disruption of cloacal membrane** resulted in cloacal exstrophy
  - Limitation b/c not spontaneous/transgenic (it's mechanically induced)
- Chick model (2005)
  - pericardial injection of suramin led to **transient aortic dilatation** resulting in CE
  - Limitation b/c chicks normally have cloaca (can't create bladder exstrophy)
- Murine model (2013)
  - **P63 knockout mice** resulted in complete spectrum of bladder exstropy
  - Lead to hypotheses regarding pathways involved (dec P63 leading to EEC)

# Etiology

## Likely Multifactorial

- **Genetic**
  - siblings have 1% chance of recurrence
  - children have 1.4% (400x general population)
- **Environmental**
  - Preconception maternal smoking significantly more common in EEC

# 2- Isolated Epispadias

# 2.1 Epidemiology

- Incidence : 1 in 300,000
- M:F = 3-5:1
  - 1 in 100,000 males
  - 1 in 400,00 females

# 2.2 Anatomic features

- Male - urethral meatus located dorsaly
  - Penopubic
  - Penile
  - glanular
- Female
  - least severe degree, the urethral orifice simply appears patulous.
  - intermediate, the urethra is dorsally split along most of the urethra.
  - most severe the urethral cleft involves the entire length of the urethra and sphincteric mechanism and the patient is rendered incontinent

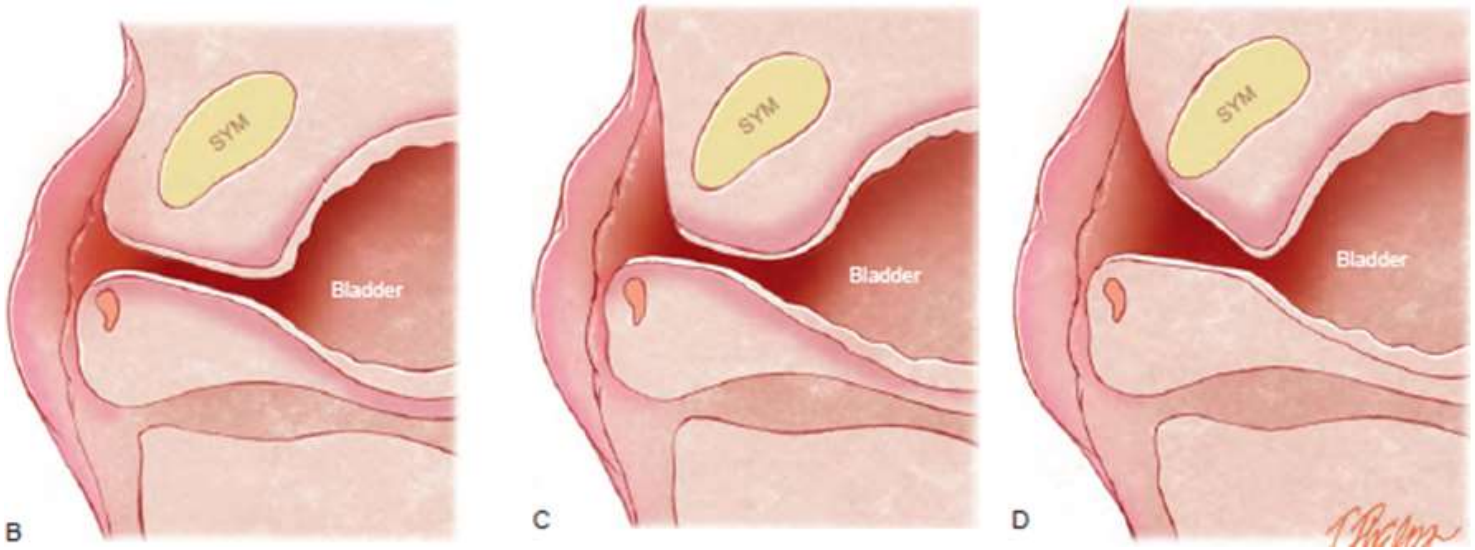
# 2.2 Anatomic features

- **Isolated Male epispadias**: **urethra** is open and exposed. Distal to its ectopic opening. lateral splaying of the **penis** creating a spade-like appearance. phallus is shorter, broader, has dorsal chordee. There might be mild **pubic diasthesis**. **vesicoureteral reflux** may be present (30-40%). **Bladder neck** is involved (short, wide open) but may be spared in boys with distal epispadias. **70% of have complete epispadias with incontinence*.

# 2.2 Anatomic features

- **Female epispadias** : **clitoris** is bifid, the **perineal body** is broadened, and the **vagina** is anterior. **urethral meatus** is not vertically slit-like but is often horizontal, striped, and gaping. **Bladder neck** is wide and incompetent. **Because change in external appearance may be minimal, some children are identified only because of persistent incontinence*

# 2.2 Anatomic features




- **Vesicular epispadias** - urethra mainly normal but relation to clitoris altered.
- **Subsymphyseal epispadias -** defect in anterior wall of urethra for about one half its length. D
- **Retrosymphyseal epispadias** - defect in anterior wall of entire urethra. Sphincter is usually involved

# 2.3 - Management

# Goals of Repair

- achievement of urinary **continence**
- preservation of the **upper urinary tracts**
- reconstruction of **cosmetically** acceptable external genitalia
- establishment of a straight penis of adequate length that is **functional** for sexual intercourse.

# Components of Repair

- correction of dorsal **chordee**
- glanular and **urethral** reconstruction
- closure of penile **Skin**
- **bladder neck** reconstruction can be performed at same time If bladder capacity is adequate
- ****submucosal injection of collagen** in the bladder neck area can have a role in improving stress incontinence when the patient with complete epispadias

# Options of Repair

## Male

- **modified Cantwell-Ransley repair**
  - Partial disassembly of penis, reverse meatal advancement and glanuloplasty
  - ? Short penile length and residual dorsal chordee
- **Complete penile disassembly**
  - urethral plate and each corporeal body along with its hemiglans dissected completely free from each other
  - May result in hypospadic meatus that require a second stage urethroplasty

## Female

- Due to the comparatively shorter urethra, repair is generally done along with YDL - BNR, monsplasty, and clitoroplasty.

# Preop considerations

## Lengthening

- testosterone IM (2 mg/kg) or topical can be administered at 5 and 2 weeks prior to increase penile skin.
- Decision of penile lengthening can then be made
  - Penile lengthening can be achieved by division of all remnants of suspensory ligament and release of crura from inferior pubic rami
  - Further urethral lengthening can be made with grafts.

# Post op considerations

## Male

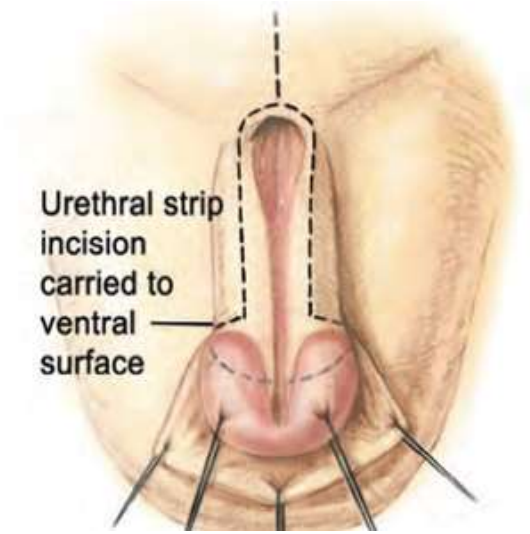
- Urethral catheter – for 2 weeks
- dressing - several days

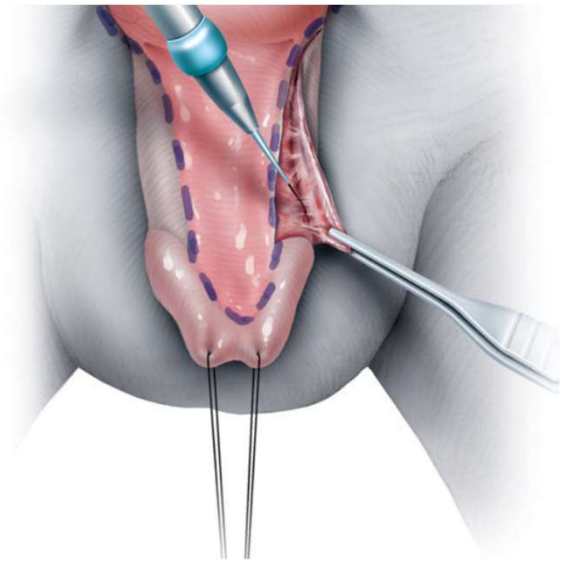
## Female

- 10-Fr catheter - for 5 to 7 days.
- If simultaneous BNR, a Foley catheter is not left in the urethra

# 2.4- Male Epispadias repair Technique

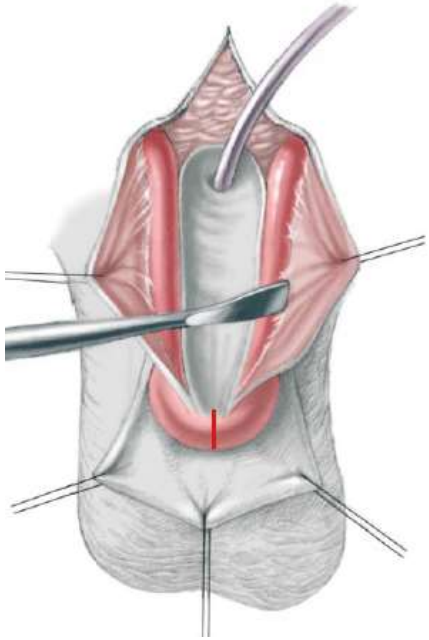
# Incision

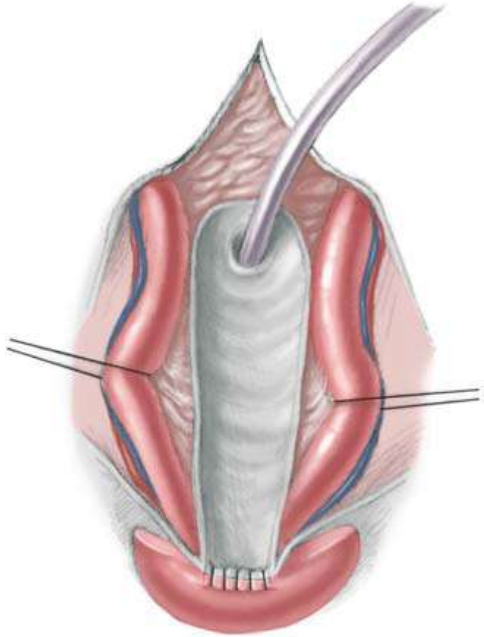


- A U-shaped incision is made along the margins of urethral plate, parallel to corona to deglove the skin. Dissect lateral to plate. The deep dorsal vessels are in Buck's fascia and protected, the superficial vessels should be carefully preserved with the skin.

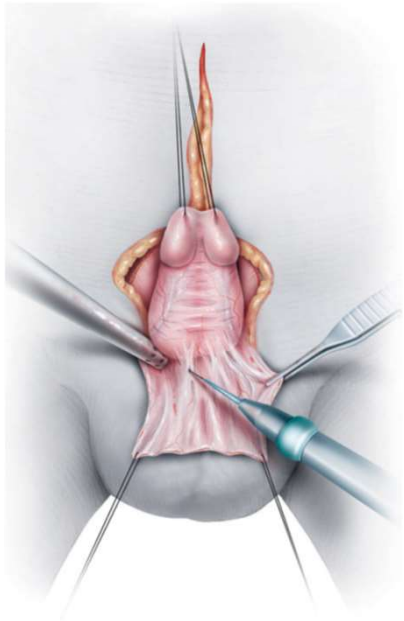
# Reverse MAGPI

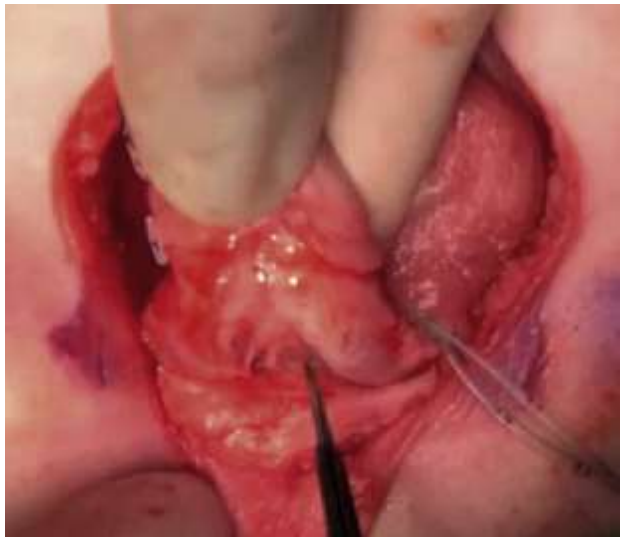




- Modified Cantwell-ransley technique: A reverse meatal advancement and glanduloplasty or IPGAM procedure at the distal urethral plate allows advancement of the urethral meatus onto the glans. Following this, skin incisions are made on the lateral edges of the urethral plate and around the epispadic meatus.

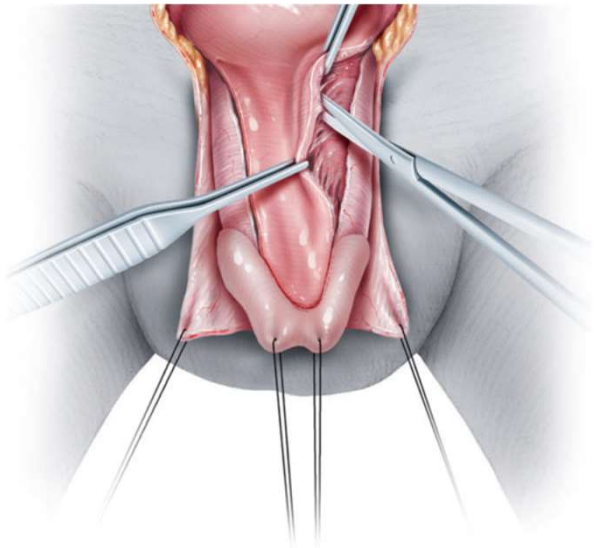
# Dissection of urethral plate

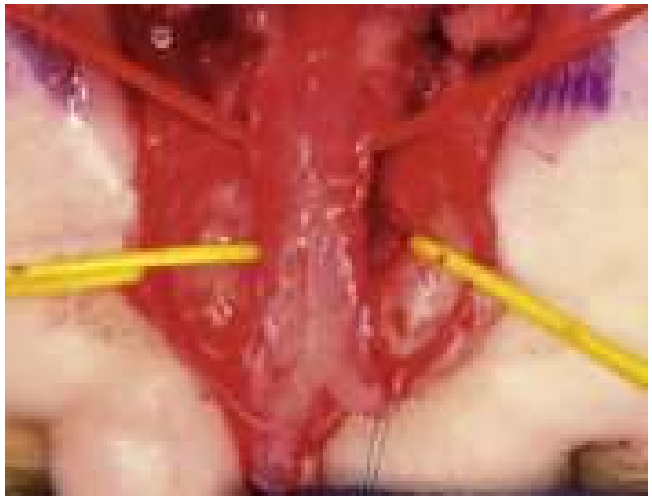




- **Ventral dissection**: spongiosum is dissected from the corporal bodies up to the level of the glans distally and to the prostatic urethra proximally.

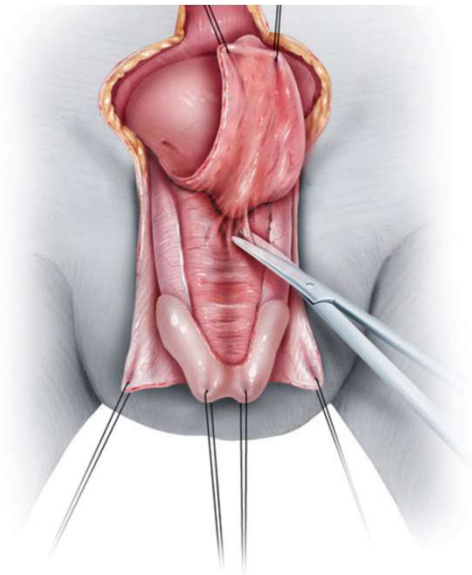
# Dissection of urethral plate

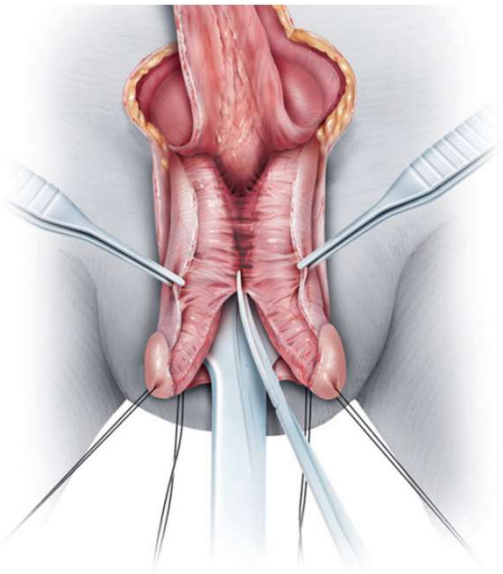


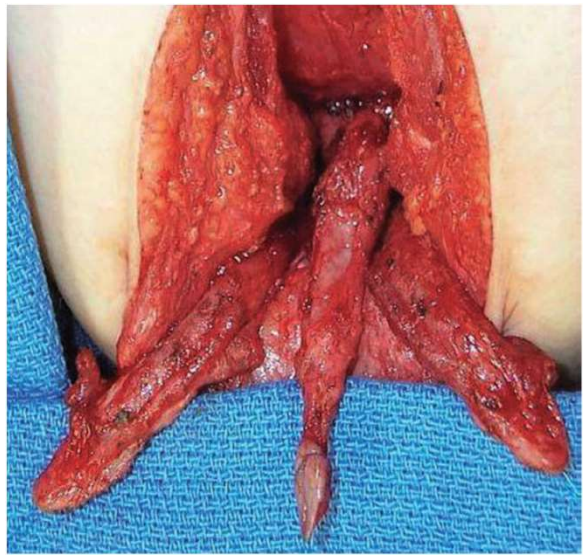



- **Dorsal dissection**: begin at midpoint of lateral edge, separating urethral plate off the corpora cavernosa. Urethral width of 15mm is maintained all the way to bladder neck. In absence of dorsal “bowing” urethra is kept in continuity with the glans.

# Separating plate, glans and corpora

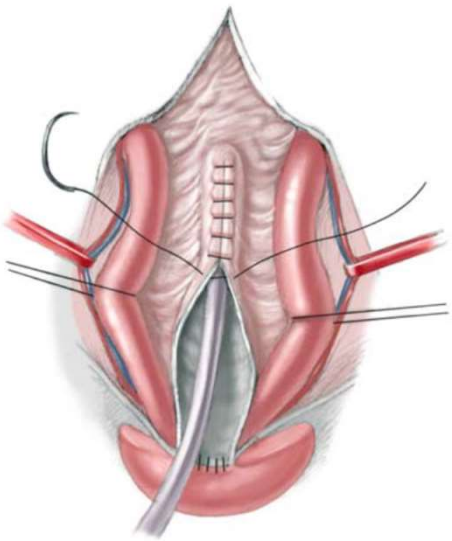


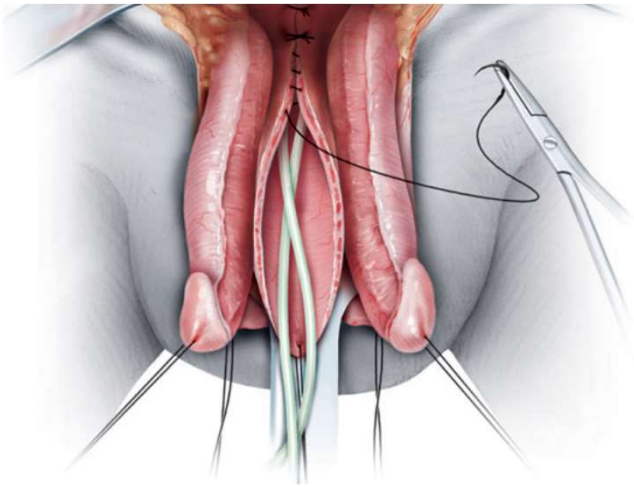


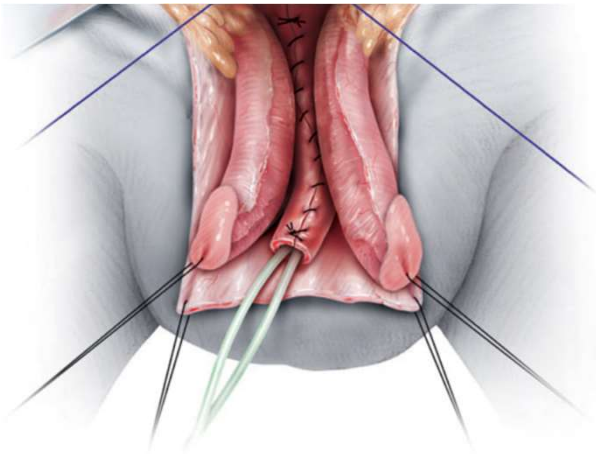


- Complete penile dissasemly technique: If the child has severe chordee and a short urethra that inhibits attempts at penile lengthening, division of urethral plate may be necessary. distal incision to free urethra from glans. corporal bodies are separated from each other which allows them to rotate medially.

# Tubularization

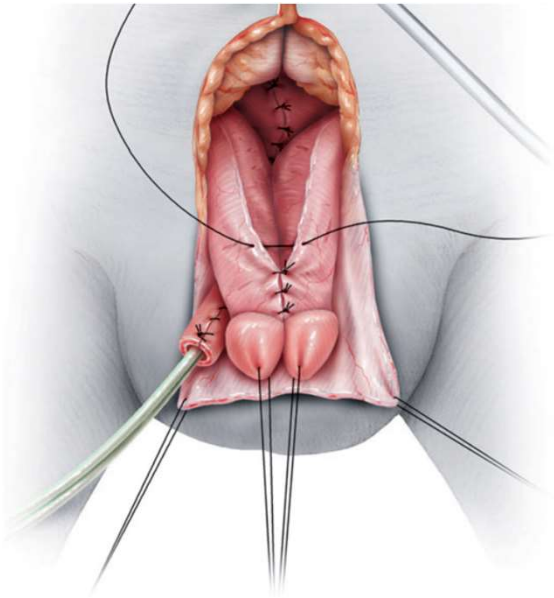


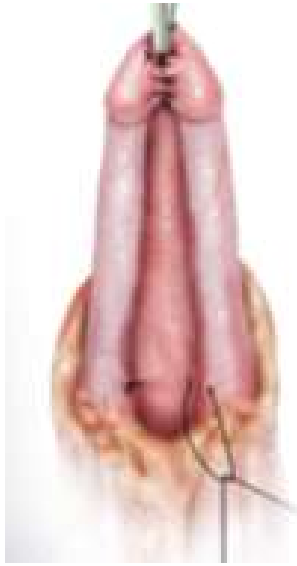




- The urethral plate is tubularized over a 6 or 8 French stent, using 7-0 vicryl interrupted subepithelial stitches as the first layer and a running 7-0 PDS for the second.

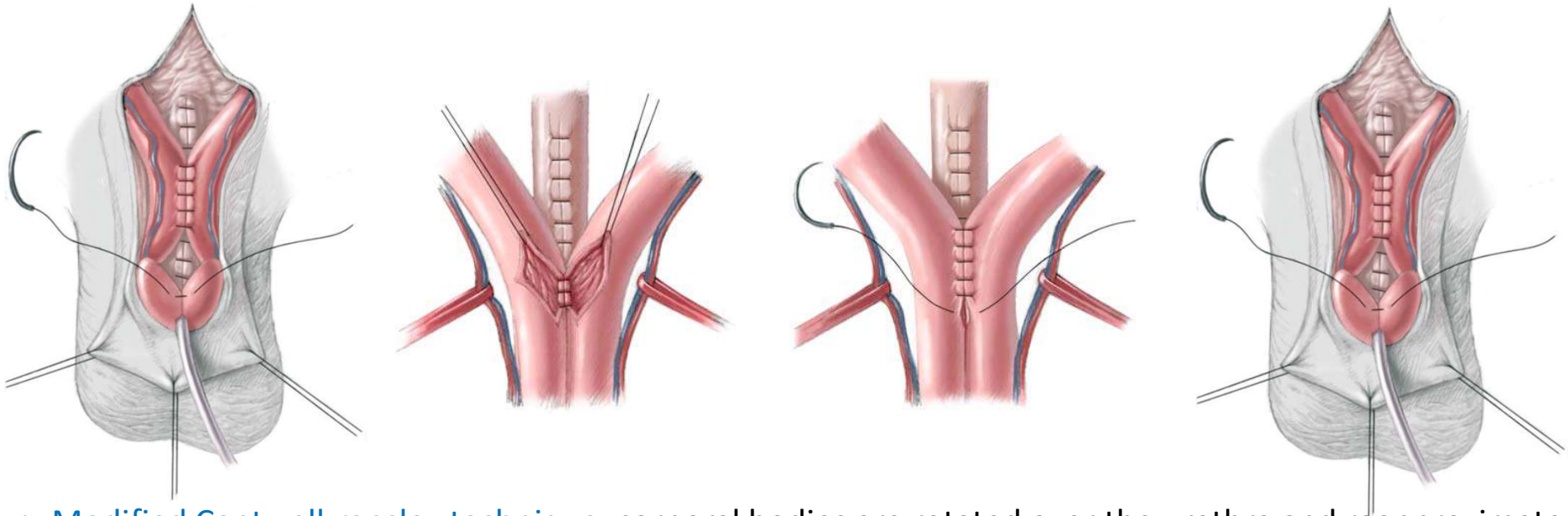
# Corpora approximation & glansplasty





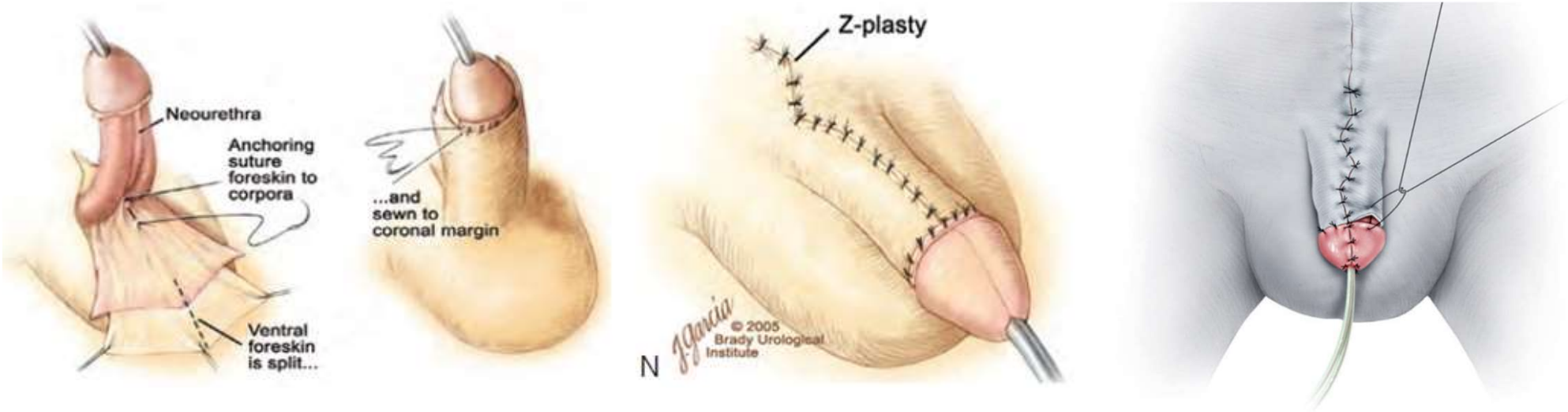
- Complete penile dissasemly : corpora are rotated dorsally and sutured with 5-0 PDS matress. hemiglans with interrupted subepithelial 6-0 vicryl. **often lengthens penis (corpora) so urethra will not reach the correct position. Urethra can be sutured as a cutaneous urethrostomy, and a graft from the prepuce or lip mucosa is placed distally for second stage*

# Corpora approximation and glansplassty



- Modified Cantwell-ransley technique: corporal bodies are rotated over the urethra and reapproximated using a 5-0 absorbable suture in an interrupted fashion. Cavernocavernosotomies (at maximal angulation) may be performed prior to reapproximating to help correct persistent chordee. Midline suturing results in extended corporal length, rolls them medially and neourethra ventrally. glans wings are then closed over the urethra using interrupted 5-0

# Penile shaft skin cover



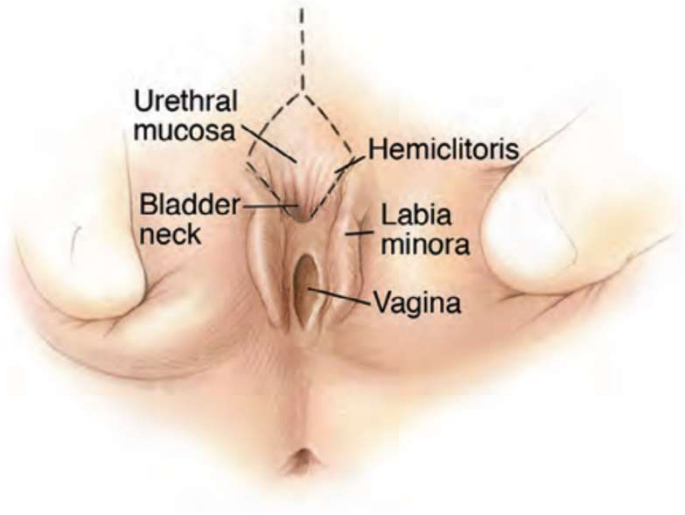

- Reverse Byars flap - Penile shaft skin can be trimmed and tailored to cover the penis using interrupted 5-0 or 6-0 absorbable sutures. Z-plasties at the level of the pubis may decrease the chance of a dorsal retractile scar at the base of the penis. ***Tacking sutures*** *may be placed to prevent skin riding over corporal bodies (3–0, 4–0 PDS at the base of the penis dorsally and ventrally to prevent the skin from migrating down the shaft during the healing phase).*

# Penile shaft skin cover

- Pippi salle The shaft skin can be released, maintaining its ventral vascular pedicle, by a circumferential incision at the penopubic–penoscrotal junction. This creates ventral penile flap that can be rotated 90 degree around the dorsum and then reapproximated. Closure of the skin at the dorsal 12 o’clock position is avoided.

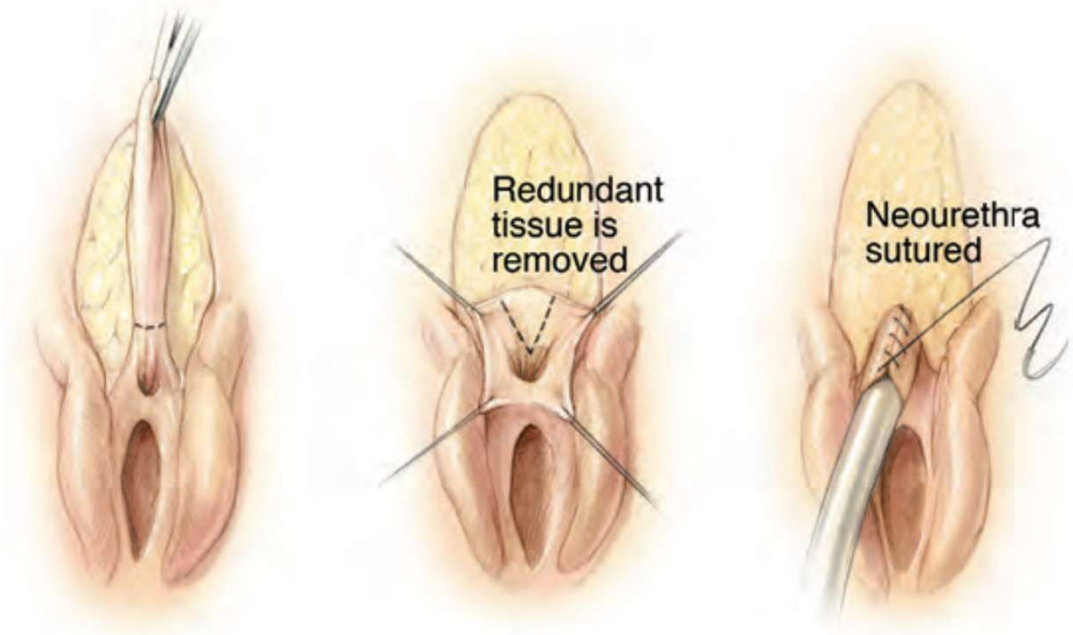
# 2.5- Female epispadias repair Technique

# Incision



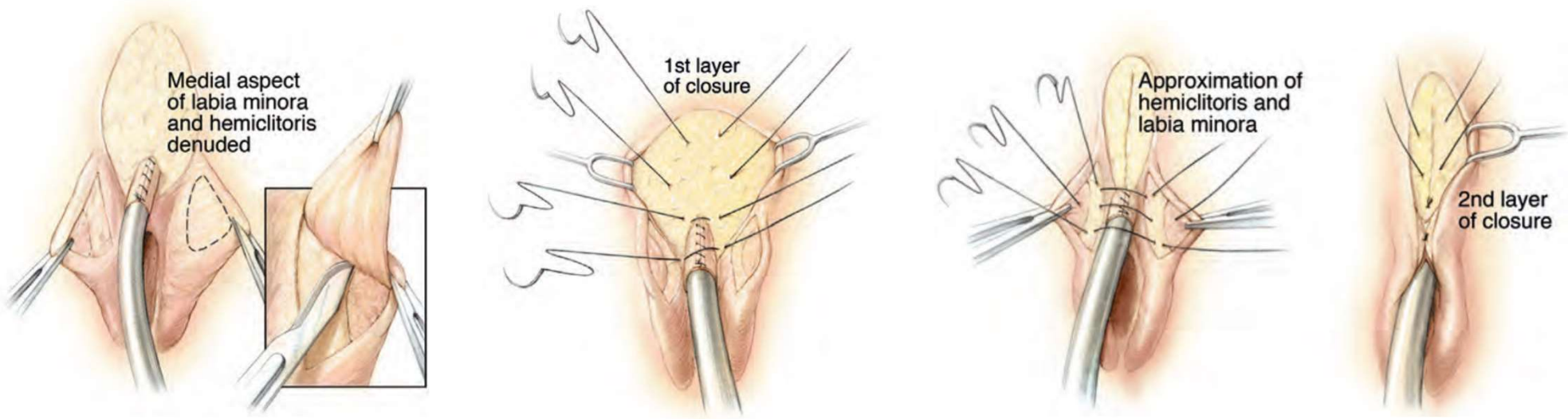

- The urethral incision is begun at the cephalad extent of the vertical incision at the base of the mons and is brought inferiorly through the full thickness of the urethral wall at the 9 o’clock and 3 o’clock positions

# Urethroplasty




- Sutures can be placed in the urethra to permit downward traction so that the roof of the urethra is excised to a level near the bladder neck. Often, one finds the dissection proceeding under the symphysis. An inverting closure of the urethra is then performed over a 10-Fr Foley catheter. Suturing is begun near the bladder neck and progresses distally until closure of the neourethra is accomplished

# Approximating labia and mons




- denuding the medial half of the bifid clitoris and the labia minora. Initial layer of mons closure. Approximation of the labia minora using interrupted sutures of 6-0 polyglycolic acid over the urethral reconstruction. corpora may be partially detached from the anterior ramus of the pubis to aid in closure. Second layer of mons closure is further aided by mobilizing subcutaneous tissue laterally and bringing it medially to fill any prior depression that remains

# Skin closure


- The subcutaneous layer is closed with 4-0 polyglactin suture in an interrupted fashion  The skin is closed with interrupted sutures of 6-0 polypropylene

# 3- Classic Bladder exstrophy

# 3.1 Epidemiology

- Incidence : 1 in 10,000 - 50,000 (most common of the spectrum)
- Sex (M:F)
  - Previous studies =  3-6:1
  - Recent studies = 1:1

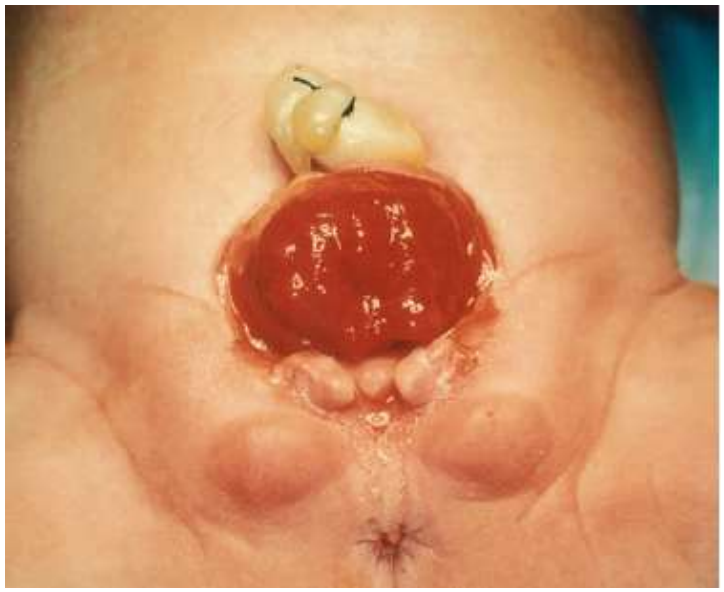
# 3.2 Anatomic features

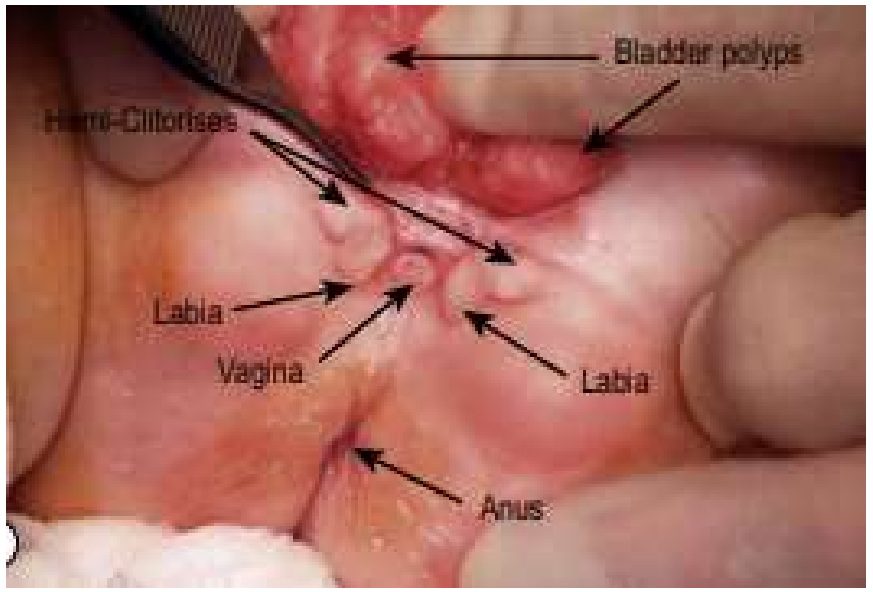
- **triangular defect** in the lower abdominal wall (from the umbilicus to intrasymphyseal band/ attenuated urogenital diaphragm)
- **everted bladder plate** (inside out with the inner surface exposed)
- **umbilical cord lower** than normal, (in relation to ilac crest) usually immediately above the bladder
- **anteriorly displaced anus**
- **Widened and shortened pubic diathesis** and rami. External rotated (outward rotation of legs in some)
- **Umbilical hernias** common
- **inguinal hernias** are common (upto 85%)
- **Minimal uretric intramural tunnel** and ureters insert inferior (reflux after closure)
- Other anomalies are rare, **usually normal upper urinary tract**

# 3.2 Anatomic features

- **Male with bladder exstrophy**: everted urethral **plate**. (top surface of penis ), broad and shortened penis (short and widely separated **corpora**), **Glans** flattened, bivalve, Ventrally hodded **prepuce** , Dorsal **chordee**, **cavernosal nerves** are located on the lateral aspects of the corpora. **scrotum is broad**, with its rugations oriented at a ~45° angle**,** increased distance between base of the penis and scrotum covered by smooth skin,

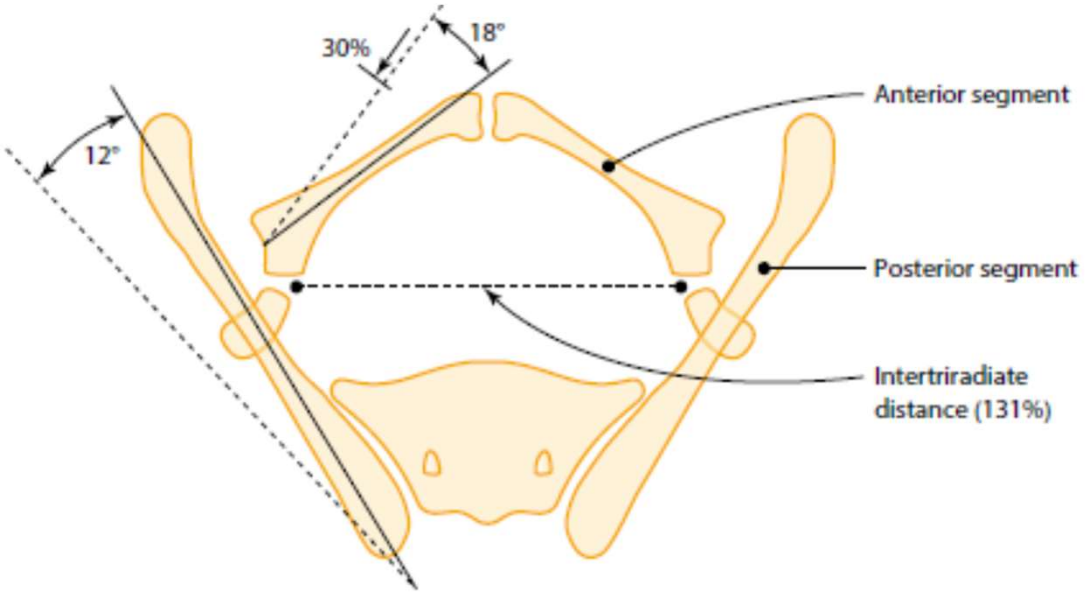
# 3.2 Anatomic features





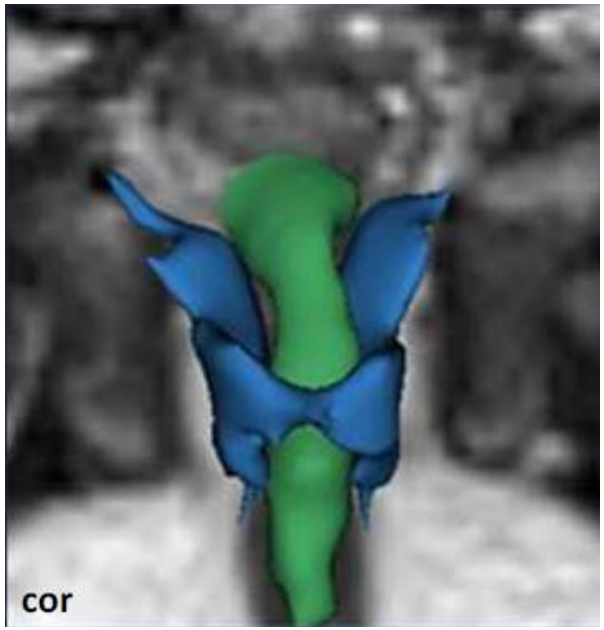

- **Female with bladder exstrophy**: **mons pubis is flattened** and displaced laterally, **divergence of labia, Bifid clitoris** usually located in anterior vaginal wall, **urethra is shortened** and exposed between halves of the clitoris, **Vaginal interoitus** anteriorly displaced and stenotic, **Vaginal canal** Short and normal caliber

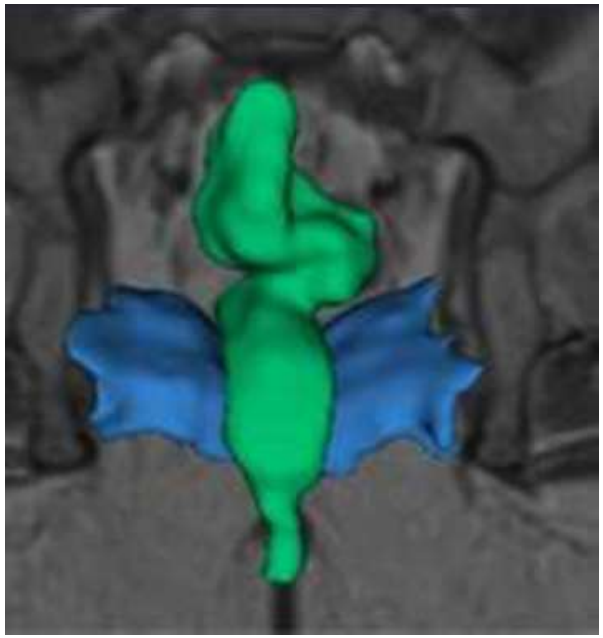
# 3.2 Anatomic features




- **Pelvic bone CT in extrophy vs control:** Pubic bone diastasis resulting from outward rotation of the innominate bones along both sacroiliac joints. Short and rotated pubic rami, Retroverted acetabulae.

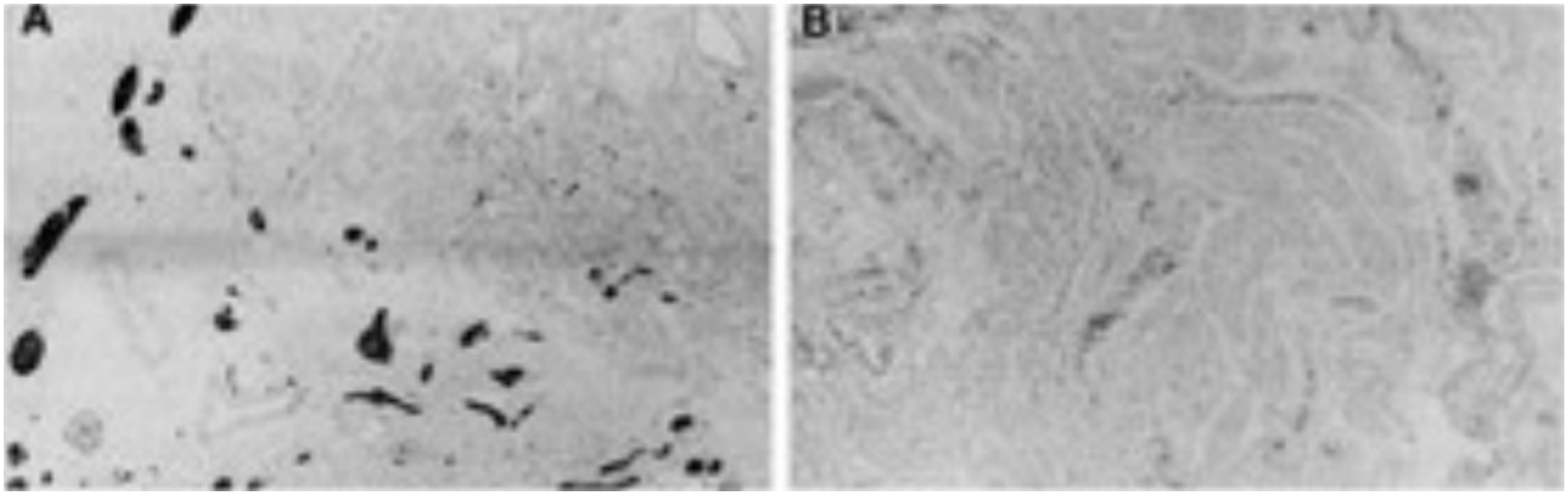
# 3.2 Anatomic features



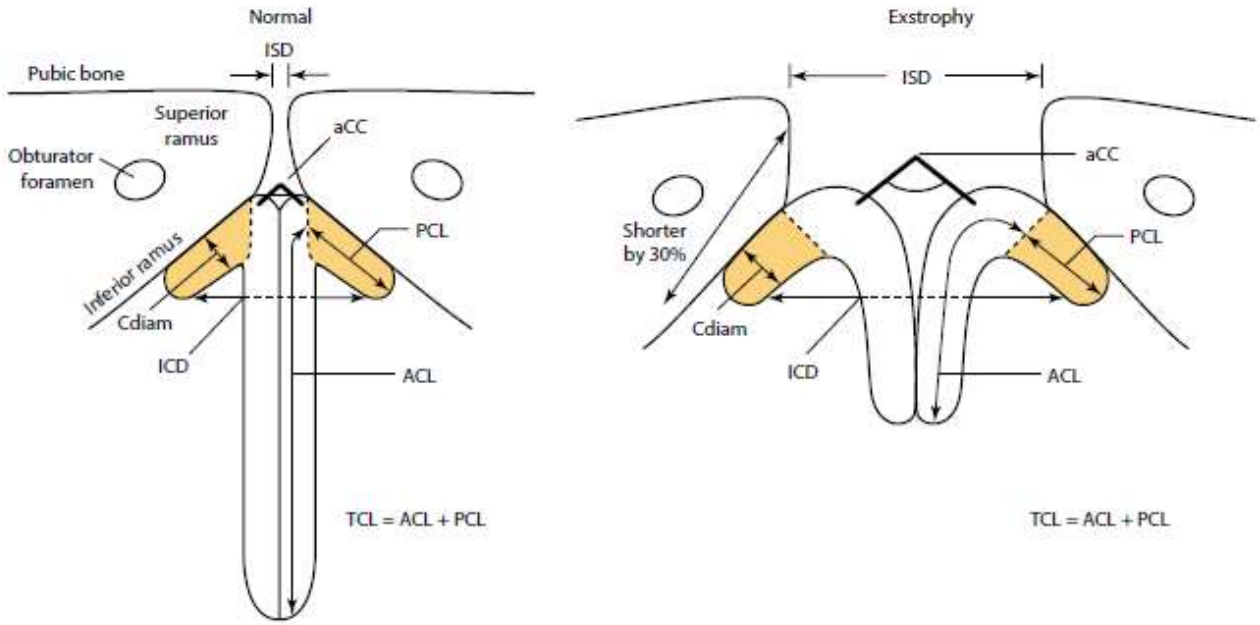
- **Pelvic musculature MRI in extrophy vs control**: More on posterior side of rectum. loss of dome configuration (over all flat and elongated). Rectal prolapse can occur

# 3.2 Anatomic features




- **Histology of Detrusor in BE vs Control**: **fewer myelinated nerve fibers**, increased collagen(3x), overexpression of inflammatory genes and underexpression of developmental genes (detrusor immaturity)

# 3.2 Anatomic features




- **MRI of Penis in BE vs Control:** Reveals the penis is shorter than normal because of a short anterior corporal segment. It has been anecdotally noted that the length of the penis and the size of the exstrophic bladder plate are inversely related.

# 3.3. Management

# Prenatal diagnosis and management

- **Ultrasound** = Absence of urinary bladder in fetal pelvis
  - Protrusion of lower abdominal wall
  - Anteriorly displaced scrotum with small phallus
  - low-set umbilical cord with normal kidneys
  - Abnormal iliac crest widening
- **MRI** = better visualization
- **Educate** about condition and prepare for anticipated care
- **Spontaneous vaginal delivery**

"Approach your child's treatments and path to health in small steps. Try not to feel totally overwhelmed by the diagnosis. It will be a lifelong issue. However, you can overcome the worries as you support your child. And remember: Attitude is everything."

*Parent of a 14-year-old boy with BE*

"Normal (regular) life is still much more prominent than the occasional challenge. The kids will have the same interests and experiences as any other child. Also, we learn how remarkably resilient we are. The kids and the parents will be able to handle the challenges ahead."

*Parent of a 9-year-old girl with BE*

"It made me stronger. Our marriage benefited from it. It brought my wife and I together to 'team up' to support our daughters."

*Parent of a 9-year-old girl with BE*

"They will do everything that other kids do. Bladder exstrophy is just part of who they are."

*Parent of an 8-year-old boy with BE*

# Natural history

*Non-lethal at birth; natural history has been well described in untreated adults.*

- Psychosocial problems (odor & hygiene)
- Skin breakdown (Incontinence)
- UTI
- Urinary caliculi
- Renal failure
- Sexual dysfunction
- Malignancy (chronically exposed bladder plate)
    - ectopic bowel mucosa, polypoid lesions (cystitis cystica/ glandularis) >> squamous metaplasia>> adenoca/ squamous ca

# Goal of treatment

- Reconstruct bladder & urethra and create competent bladder neck
  - renal function
  - Bladder function & Continence
- Reconstruct genitalia
  - Acceptable appearance
  - Sexual function
- Psychological support
  - Ensure child has typical childhood

# Options

- 1. Urinary Diversion
  - Incontinent Diversions – ileal conduit, ileocecal conduit, colon conduit
  - Internal Diversion (Rectal Sphincter-Based Continence) – Sigma, Ghoneim, Gersuny, Heitz–Boyer–Hovelacque, rectal bladder with colostomy, ureterosigmoidostomy, ileocecal ureterosigmoidostomy
  - External Diversion (Urinary Reservoir with catheterizable channel) – Indiana (cecal), Mainz, Penn (ileocecal), kock pouches
  - ** orthotopic neobladder
- 2. Anatomic reconstruction
  - Multistage: MSRE (modern staged repair )
  - Single stage: CPRE (complete primary repair)
  - **Kelly procedure: RSTM (Radical soft tissue mobilization)

# 3.3.1. Urinary Diversion

- creating a functional connection between the urinary and intestinal tracts (diverting from bladder)
- ? bladder and urethra are tubularized and function later in life as a conduit for semen
- achieves goal of dryness better than reconstruction
- Avoids complications such as urinary retention & kidney damage, dependence on CIC
- not commonly performed in developed countries.
  - For patients who have failed multiple attempts at functional reconstruction.
  - Some advocate primary urinary diversion for patients with bladder plates deemed too small to close. However, it is impossible to *accurately predict* which bladder plates will increase significantly in size after primary closure.

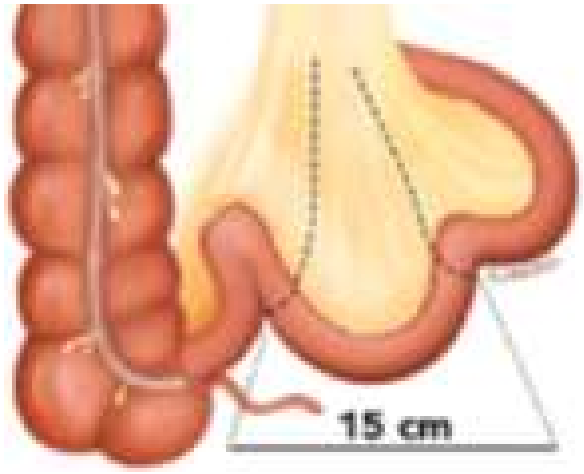
# 3.3.1. Urinary Diversion

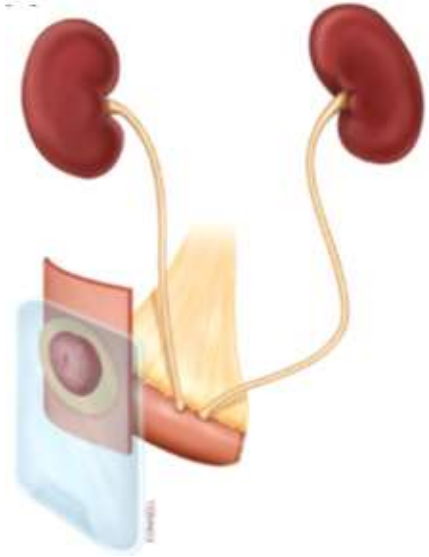
## A) Incontinent urinary diversions

- needed appliance
- had a high rate of bowel stenosis/obstruction
- High rate of infection infection

# 3.3.1. Urinary Diversion

## A) Incontinent Diversion



- **Ileal conduit**: distal ileum taken used to create a connection between the ureters and the anterior abdominal wall via a cutaneous stoma

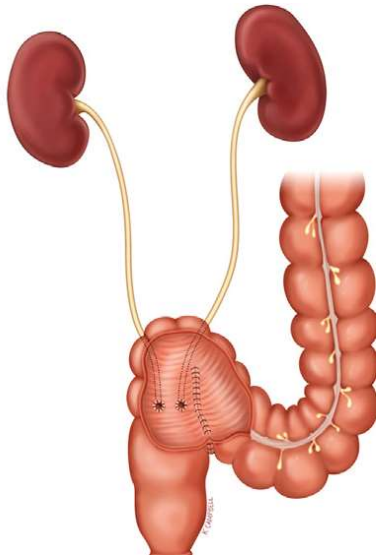
# 3.3.1. Urinary Diversion

## B) Internal diversion: Ureterosigmioidostomy

- one of the oldest and technically easiest continent diversion
  - Use may be limited by anal sphincter problems in BE (weak pelvic floor, anoplasty for anterior anus)
  - high pressure of contractile sigmoid **refluxes infected** urine or feces
  - hyperchloremic metabolic **acidosis**
    - *frequent emptying to reduce contact with rectal mucosa, bicarbonate
  - bladder **calculi**
  - 250 to 300 x risk (occur in ~5% in 25 years) of **adenocarcinoma** at ureteric anastomosis site
    - * modification of reservoir to prevent mixture of urine and feces might help
- **Mainz II pouch** described by Fisch and Hohenfellner in 1991, is a modification of ureterosigmoidostomy with pouch formation**.** It provided significant improvement to USO.
  - Better protection of upper tract (Reduces reservoir pressures ) and improves nighttime continence

# 3.3.1. Urinary Diversion

## B) Internal diversion



- **Mainz II (modified ureterosigmoidostomy)** = creation of a low pressure pouch for a remodeled and detubularized sigmoid colon. The ureters are implanted via a submucosal, antirefluxing tunnel.

# 3.3.1. *Considerations in Mainz II USO

- Age - fecal continence should be achieved
- anal sphincter - mechanism should be intact intact
  - Digitally by anal stimulation
  - having the child hold warm water for a couple of hours (300ml for 2-3 hr day time or 4-6 hr night)
- IVU/CT - to r/o ureteric dilation (contraindication to USO)
- Bowel preparation
- Antibiotic – prophylactic + continued post op until uretric stents are removed

# 3.3.1. *Considerations in Mainz II USO

- Drainage: if < 50ml/24 hr
- rectal tube: when bowel movement start (5th d)
- Uretric stent: release skin fixation on 9th d
- IVP / ultrasound before discharge - detect bilateral hydronephrosis; if present, place percutaneous nephrostomies at once before anastomotic leakage occurs
- ultrasound – at 6 weeks, then regularly detect ureteric stenosis (usually within first 5 years)
- Water soluble contrast enema in case of recurrent infection to detect reflux/obst .
- Blood gas analysis: base excess below -2.5 mmol/L is indication to start alkali substitution and recheck every 2 weeks
- low-chloride diet + Na/K as bicarbonate /citrate – to prevent significant hyperchloremic acidosis in children with renal impairment. Keep the serum bicarbonate level nearly normal. If decompensated might also need frequent evacuation (rectal tube) and fluids
- Occult blood every 3mo & annual colonscopy – starting from 5th year post op to screen for malignancy
  - If detected early, curative surgery including conversion into a continent reservoir is possible

# 3.3.1. Urinary Diversion

## C) External diversion: Augmentation-Mitrofanoff

- Bladder Augmentation usually reserved after closure and BNR
  - If there is **inadequate capacity** (high bladder pressure and VUR) to protect the kidneys
  - to help otherwise irreversible forms of **incontinence**

# 3.3.1. Urinary Diversion

## C) External diversion: Augmentation-Mitrofanoff

- 1911, Coffey described the flap valve technique of ureteral reimplantation in ureterosigmoidostomy. This principle was utilized by Mitrofanoff in 1980 to create a continent, catheterizable appendicovesicostomy
  - narrow, supple **conduit** brought to the skin
  - **antirefluxing connection** between the conduit and the reservoir (flap valve)
  - large, low-pressure **urinary reservoir**;
  - an **antirefluxing mechanism** between the upper urinary tract and the reservoir;
  - **CIC** to allow effective, regular, low-pressure emptying of the reservoir.

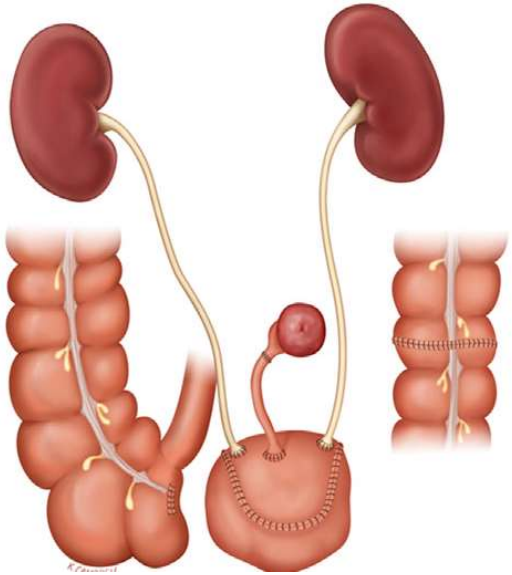
# 3.3.1. Urinary Diversion

## C) External diversion: Augmentation-Mitrofanoff

- Complications include
  - stomal stenosis, ischemia, prolapse, mucus blockage, lekage
  - urinary infections
  - bladder calculi
  - persistence of a high-pressure neoreservoir
  - metabolic and acid–base disturbances
  - Adenocarcinoma
  - difficulty in catheterization leading to false passage

# 3.3.1 Urinary Diversion

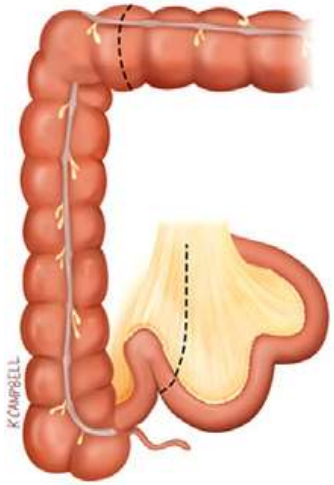
External diversion

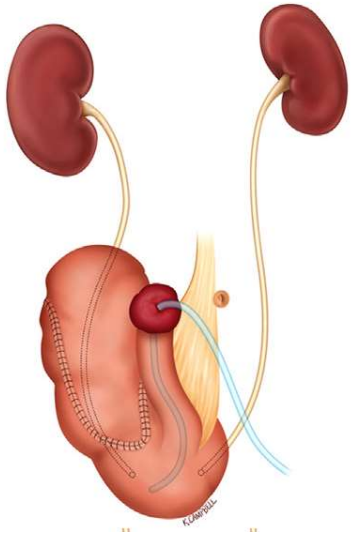


- **Augmentation with Mitrofanoff:** colocystoplasty with appendicovesicostomy

# 3.3.1. Urinary Diversion

External diversion





- **Indiana Pouch** = ureter anastomosed to right colonic reservoir with ileocecal valve. terminal ileal segment used as efferent limb and connected to abdominal wall.

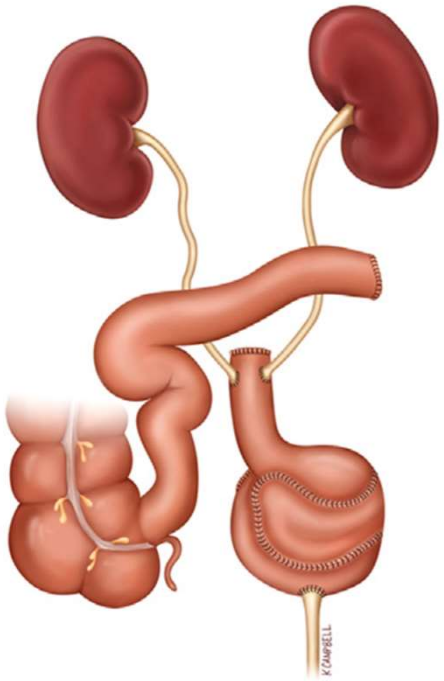
# 3.3.1. Urinary Diversion

## Bladder substitution (Orthotopic neobladder)

- If bladder is **inadequate even to attempt augmentation (**excessively fibrotic or too small**)**
- Alternate to excision of plate with diversion.
- plate may also be left used later for the posterior urethra in an Arap-type procedure

# 3.3.1. Urinary Diversion

## Bladder substitution



- **Studer pouch:** orthotopic neobladder constructed from ileum. Proximal end of ileal segment left intact to be used as afferent limb

# 3.3.1 Considerations in Augmentation-mitrofanoff

- Renal assessment
- Educate the patient and family about CIC.
- Prepare the bowel and administer broad spectrum antibiotics.
- Foley catheter for 2 weeks. (No other drainage needed in mitrofanoff without augmentation)
- Cystostomy tube – requires regular irrigation. remove once routine CIC established.
- Mitrofanoff catheter - 3 weeks
- CIC trial – 3wk after cystogram if no leak detected, clamp cystostomy and start CIC with 8 or 10 fr (may inc to 12-14 fr overtime )
- Ultrasonography - to gauge the effectiveness of catheter drainage.
- Bladder neck closure is necessary in almost all cases.
  - could be left open if there is total dryness between the urethral catheterizations and the impossibility of self-catheterization owing to an orthopedic situation.
  - Alternatively, a urethral sling procedure may be done later if incontinence

# 3.3.1 Considerations in Augmentation-mitrofanoff

- Suprapubic tube irrigated BID with 30-60 ml NS

# 3.3.2. Anatomic Reconstruction

## MSRE

- developed in 1970s due to discouraging results of Single stage repair (**upper tract damage** and the low rate of **urinary continence**)
- Initially meant to be 2nd stage BNR at 2-3 yrs and epispadia repair later.
  - This paradigm was altered when bladder capacity was noted to increase with added resistance of early epispadias repair.
- The current MSRE defined by the Johns Hopkins group (Gearheart)
- Combined MSRE stage 1& 2 can be performed if significantly delayed primary closure or initial closure failure.

# 3.3.2. Anatomic Reconstruction

MSRE Male

- 1st stage **(2 mo)** close bladder, abdominal wall & partial epispadias repair (post. urethra)
  - 5-7 hr procedure (with osteotomy)
- 2nd stage **(6mo -1yr)** Completion of epispadias repair (modified Cantwell–Ransley )
  - When urethral groove is of adequate length
- 3rd stage **(usually 4- 5 yr, --10yr)** BNR (+ almost always need reimplantation)
  - Requires bladder capacity and motivation to participate in voiding program
  - If bladder too small, bladder neck transection, augmentation and continent diversion
- MSRE Female
  - closes bladder, entire urethra, and abdominal wall with later BNR

# 3.3.2. Anatomic Reconstruction

## CPRE

- Later **advances** and advent of **CIC** led to resurgence of single stage (Mitchell).
- Combines first 2 stages of MSRE and Allows bladder cycling to begin earlier, which could optimize bladder development.
- Dec cost and morbidity of repeat operations. But may require further surgeries.
  - Most for BNR for incontinence (60-80%)
  - almost all will require reimplantation.
  - Some for resulting hypospadias
  - Some for Failed primary closure

# 3.3.2. Anatomic Reconstruction

## CPRE Male

- bladder closure, anatomic bladder neck narrowing, urethral elongation, and epispadias repair in a single operation.
- The epispadias repair is done by complete penile disassembly
  - Otherwise attachments pull the urethral plate anteriorly, preventing posterior placement of the proximal urethra and bladder neck in pelvis
- At times, bilateral ureteral reimplantation may be performed at CPRE

## CPRE Female

- Similar to female MSRE but more aggressive mobilization of the vagina and urethral plate posteriorly into the pelvic diaphragm in order to gain bladder outlet resistance

# 3.3.2. Anatomic Reconstruction

RSTM (radical soft tissue mobilization)- The Kelly operation

- CPRE/MSRE without osteotomies + detaching corpora from ischiopubic rami insertion) detachment of the levator ani from the obturator internis fascia. These voluntary and involuntary sphincter muscles are then wrapped around the tubularized
- Reportedly achieves significant penile lengthening
- Not widely used b/c can lead to damage to pudendal NVB

# 3.3.2. Considerations in primary closure

- **Reduce trauma to bladder plate:** umbilical cord **ligated** with silk suture or soft tie (“umbilical tape”) rather than clamp. **Covering** with soft, transparent material to protect from diaper. hydrated gel dressing (vigilon®) or plastic wrap (Saran Wrap®) replace dressing daily and bladder **irrigated** with normal saline with each diaper change. **barrier creams** (zinc oxide) help prevent skin inflammation.

# 3.3.2. Considerations in primary closure

- Imaging: **AP pelvic x-ray / KUB** to asses diasthesis and **Ultrasound** to asses kidneys

# 3.3.2. Considerations in primary closure

## Timing of primary closure

- Early (<72 hours)
  - anatomic closure without the use of osteotomies
  - Earlier bladder cycling (improved capacity)
  - reduces the time of bladder exposure (dec risk of precancerous changes)
  - Not applicable if the bladder template too small (<3 cm), covered with polyps, or appears inelastic
- Delayed (4-6 mo, --9 mo)
  - allows transfer to high volume center
  - more stable infant (safer anesthesia)
  - anatomy is safer to repair (especially if baby was premature)
  - more time for parental bonding
  - ?time to stimulate penis with testosterone
  - However requires adequate protection of exposed tissue and has increased cost
  - increases likelihood of eventual need for bladder augmentation due to inadequate bladder capacity

# 3.3.2. Pre-operative Considerations

## Timing of primary closure

- **Indications for delaying repair**
  - inadequate bladder
  - preterm
  - Surgeon preference and experience
  - availability and access to medical resources

- **acceptable capacity -** bulging on crying , indenting on touching by a sterile gloved finger (a small bladder template, if **distensible/contractile**, can enlarge after closure)

- **If bladder remains inadequate (after delay) -** nonrefluxing colon conduit along with abdominal wall closure and epispadias repair. Can be converted to a continent diversion at later age.

# 3.3.2. Timing of primary closure

- **<u>Assessment of bladder plate</u>:** Requires EUA b/c good portion of previously unappreciated bladder can be discovered behind the fascia. But may also find unsuitable bladder, small & undistendable, rigid, stretched between the edges of the small triangular fascial defect

# 3.3.2. Timing of primary closure

- **unsuitable for early primary closure** bladder plate Fibrotic with hamartomatous polyp. attempt at closure will only result in upper tract deterioration . delay of closure may give more time for bladder plate growth. Adequate capacity is estimated ≥ 5 mL at birth, with elasticity (evert with valsalva).

# 3.3.2. Considerations in primary closure

## Materials

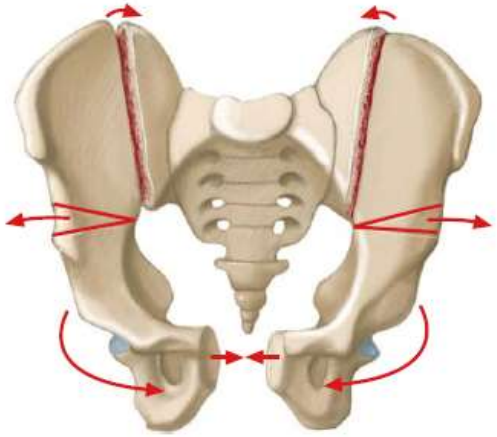
- Tubes
  - 3- and 5-F paediatric feeding tubes (ureteric stents);
  - 10 F Malecot drain (suprapubic tube)
- Sutures
  - 2–0 silk ties (umbilical vessels);
  - 3–0 and 5–0 PDS sutures (bladder and urethral closure);
  - 3–0 nylon, 4–0 chromic, 3–0 polyglactin (secure drains);
  - No. 2 nylon sutures (intrapubic stitch);
- Paediatric Van Buren sounds.
- orthopaedic instruments (if osteotomy required)
  - Interfragmentary pins, external fixator
  - Supplies for traction

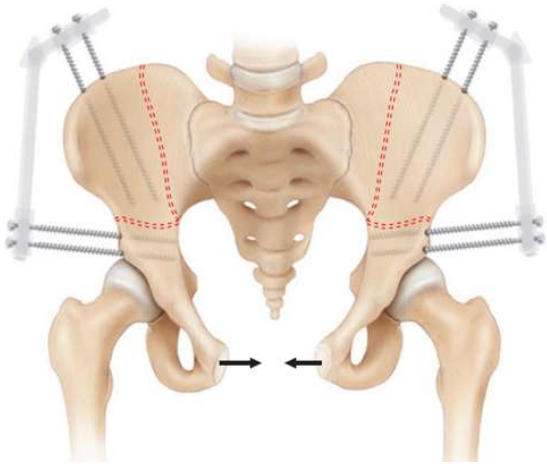
# 3.3.2. Considerations in primary closure

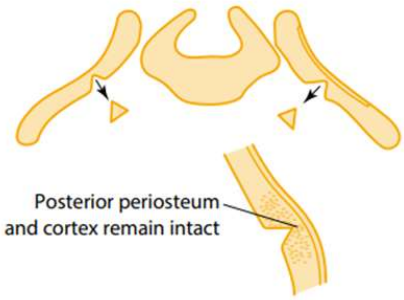
## Iliac osteotomy

- The bone is cut to change its alignment. This decreases tension placed on pelvic bone at closure.
- Pelvic closure helps achieve continence and prevent dehiscence. Also optimizes placement of bladder in pelvis & approx. of corpora/clitoris.  Doesn't impact orthopedic sequele (gait abnormalities)
- Usually performed at time of bladder closure. Indicated for
  - children outside the immediate newborn period (non-malleable pelvis),
  - those with very wide diastasis (>4 cm)
  - reoperations (failed closures).
- It must be decided preop whether a formal osteotomy will be required
  - ensure that there is appropriate orthopedic support
  - parents need to be prepared to support a prolonged admission while the child is supine and in traction after surgery.

# 3.3.2. Iliac osteotomy





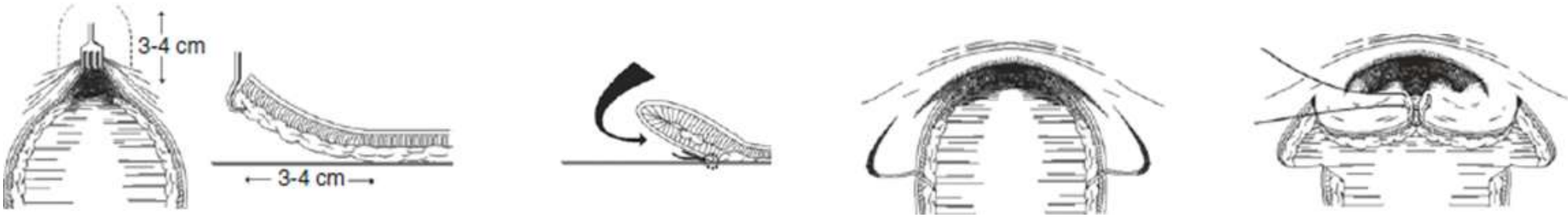



- **Bilateral combined anterior vertical iliac and transverse innominate osteotomies** done before bladder closure and inter-fragmentary pins placed. External fixator placed after completion of soft tissue closure. *Advocated by the Johns Hopkins group b/c their experience of superior result than anterior diagonal iliac osteotomy alone (used by Seattle group)*

# 3.3.2. Considerations in primary closure

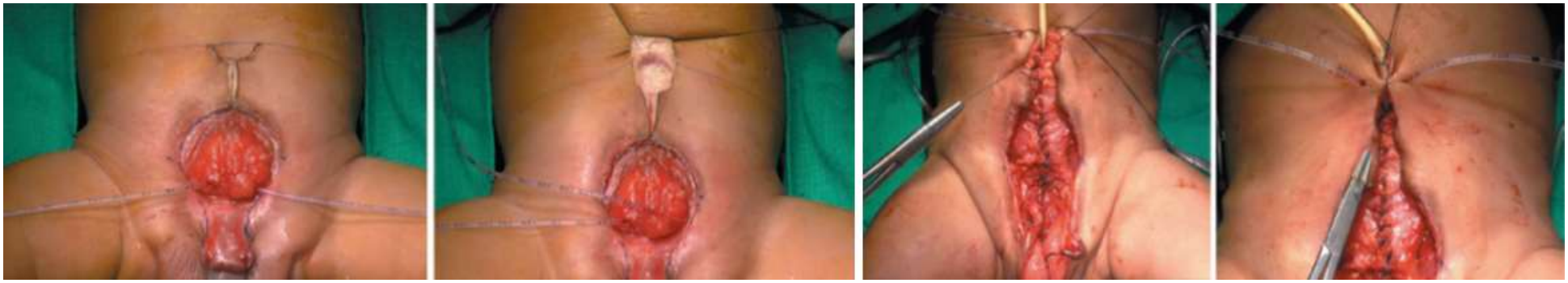
- Umblicoplasty
  - Research has shown its an important aesthetic landmark for most
  - Performed with the initial closure, separately or later with other procedures
- Inguinal exploration and hernia repair
  - Inguinal hernia common b/c enlarged inguinal rings, compromised fascial support and lack of obliquity of the inguinal canal.
  - Reinforcement of the transversalis and internal oblique fascia during hernia repair decreases the incidence of later direct inguinal hernias
- Uretric reimplantation
  - high incidence of VUR after exstrophy closure

# 3.3.2. Umblicoplasty




- **“Manchester umbilicoplasty.”**inverted umbilical reconstruction: At the apex of midline wound, the skin and superficial fascia are elevated off the anterior rectus sheath for 3–5 cm above the apex of the skin incision. The most superior apex of the wound is sutured to the linea alba at as high point as possible, thereby inverting the skin and fixing it to the sheath. Two 1.0 × 0.5 cm skin flaps based superiorly are cut from the margin of the inverted skin. The flaps are rotated medial and sutured to the linea alba to form the base of the new umbilicus.

# 3.3.2 .Umblicoplasty



- **trapezoidal flap**: trapezoid-shaped incision keeping width and length to 3:1 and base of the flap corresponding to the transtubercular line. Raising flap off the linea alba in the plane deep to the Scarpa's fascia. Linea alba approximation around catheter. Approximation of the base of trapezoid flap. Tubularization of the flap.

# 3.3.2. Anesthetic Considerations

- Avoid abdominal distention (compartment syndrome)
  - NGT decompression
  - Avoid NO2
  - Careful fluid management
  - Epidural achives less narcotics and inhaled anesthetics

# 3.3.2. Post op considerations

- Antibiotics to prevent infection, continue PO b/c all will have VUR
- Immobilization to decrease stress on closure (with or without osteotomy)
  - ≈4 weeks for infants and 6–8 weeks for older children
- pain control is essential in maintaining pelvic immobilization
  - Tunneled epidural (3 wk) continuous infusion of 1 mg/mL lidocaine at 0.8 mg/kg/h
  - i.v. or oral opioids
  - enteral acetaminophen or diazepam
- Maximal urinary drainage to divert urine away from the bladder / urethra as it heals
  - ureteral stents - 2-4 wks after closure, as swelling caused by the pressure of closure of a small bladder can obstruct the ureters and give rise to obstruction and transient hypotension
  - suprapubic tube – 4 wk. Before removing the suprapubic catheter the bladder outlet is calibrated with a catheter or sound, to ensure free drainage. Residual urine is estimated by clamping suprapubic tube
- Oxybutnin - until all catheters are removed
- 1-4 wks hospital stay may be needed for healing

# 3.3.2. Immobilization

- **Modified Buck's traction (2-4 wk): used with external fixators (osteotomy)** align the legs and hips in a straight line with the child lying flat on the bed. A posterior lightweight splint can be used in newborns when the child is out of traction to maintain hip adduction. fixators and pins can be surgically removed when good callous formation is seen on pelvic radiography, which usually occurs at 6 to 8 weeks.

# 3.3.2. Immobilization

- **Modified Bryant's traction (4wk):** gently supporting the legs straight upward toward the ceiling, away from the bed, with the legs bent at the hips (90 degree flexion). **may be used if there is no osteotomy.** In hospital for 4-6 wks , may traumatize skin

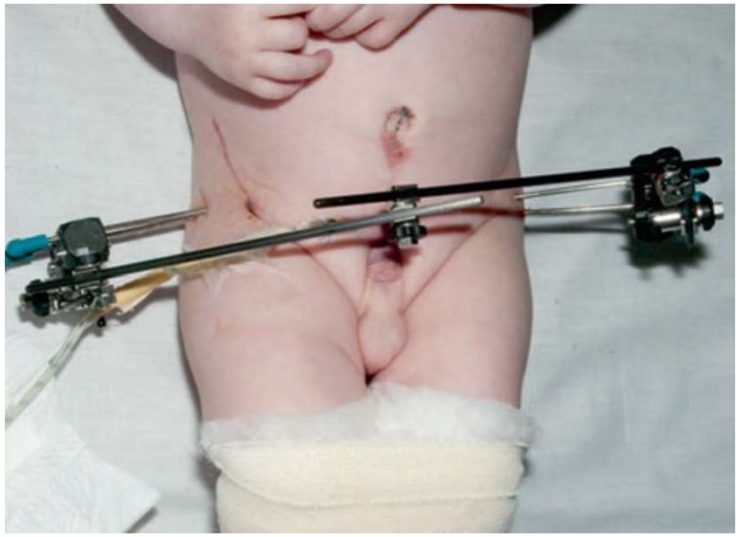
# 3.3.2. Immobilization

- **Spica cast (3-6wk)** following CPRE to reduce tension on the closure by preventing hip abduction. Allows more mobility and earlier discharge from hospital. Preferred technique of immobilization in many centers.

# 3.3.2. Immobilization

- **A hinged spica cast**, which is easier to remove, large anterior section open to allow visualization of the abdomen and posteriorly to access epidural site and to enable diaper changes.

# 3.3.2. Immobilization



- **“mummy wrapping” the child’s legs**, have been called into question after a retrospective study found them to cause higher rates of skin breakdown and have lower success rates

# 3.3.2. Considerations in BNR

- **Cystoscopy and Gravity Cystogram under GA** – b/n 1-3 year to asses reflux and bladder capacity (>85 cc), and stricture
- **Motivation to participate in Training** (voiding pogram) – starting 6 mo
- **Preoperative urodynamic evaluation** should be considered because it allows the detection of detrusor hyperactivity or atony, as well as the assessment of functional bladder capacity and leak point pressures
- Urethral catheter - 7-10 days. However, Some surgeons recommend avoidance because of concerns that it may adversely affect later urinary continence.
- Ureteral stents 10-14 d
- Suprapubic tube – 3 wks
- voiding trials 3rd wk with measurement of PVR to assess for retention before removing the suprapubic tube.
  - If no urine is passed, cystoscopy and urethral stenting may be required for a short period of time.
  - A bladder readjustment period (awareness, capacity) may span several months before day and subsequently night continence is achieve
- **Ultrasound** – before discharge, after 3mo, every 6mo -1yr to document upper tract status

# 3.3.2. Considerations in BNR

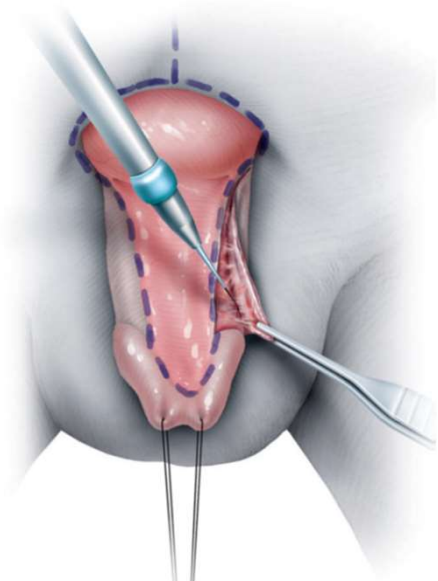
- **<u>Biofeedback evaluation/training</u>**

# 3.4 – Primary closure Technique

# Assessment of Anatomy

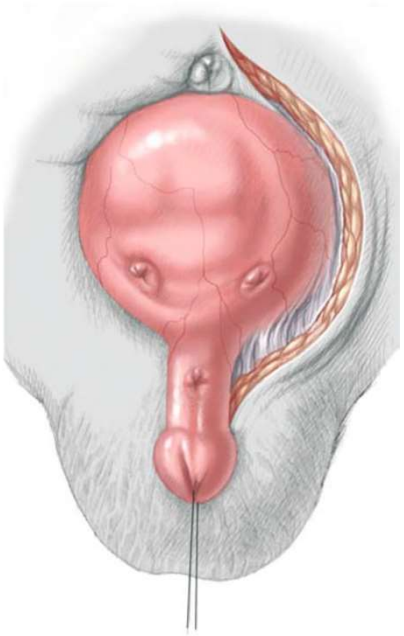
- **Penis reflected ventrally** to expose structures. Bladder and urethral **Plate measured**. **stay suture** is placed transversely in each hemiglans. **Ureter intubated** and secured with stich. Incision site, veromontanum and neoumbilicus **marked**

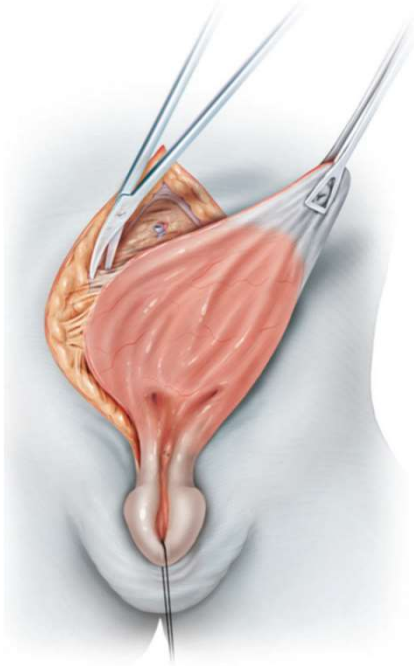
# Initial incision



- Traction sutures placed transversely on each hemiglans. incision begins along the perimeter of the bladder neck by generating **posterior urethra** (incising 2 cm wide mucosa distal to triagone to distal to veromontanum). The **urethral plate** is incised (1.5 cm) *but only upto midshaft of penis in MSRE*. Incision is carried cranially to dissect the **bladder plate** with an **inverted "V" incision** incorporating the umbilical stump.

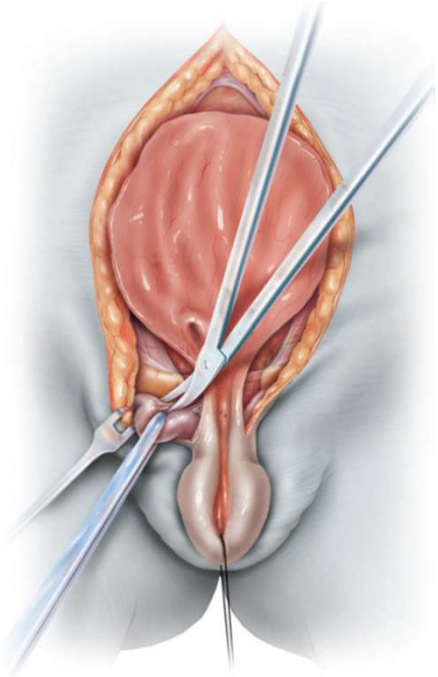
# bladder plate dissection





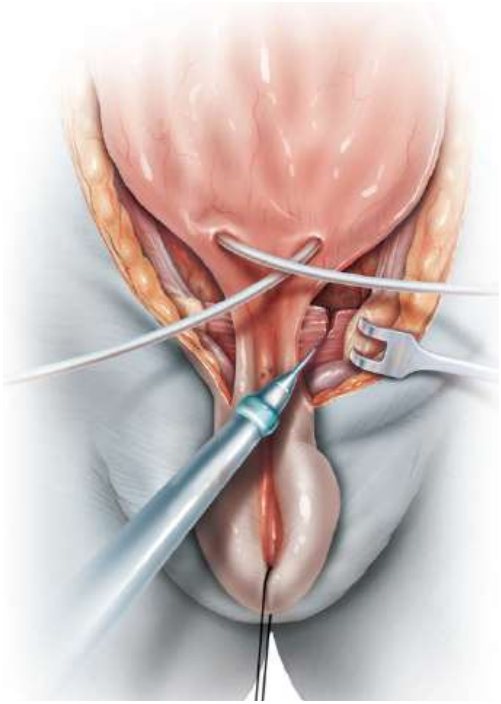
- **Traction** on umbilical stump. umbilical vessels ligated with 3-0 vircyl. **Initial plane between skin & rectus fascia** ("fat is your friend") helps identify plane b/n bladder edge and medial/posterior parts of rectus. inserting index finger inside the bladder & inverting it facilitates dissection. Entire dissection is extraperitoneal. Peritoneum is dissected off external surface of bladder dome to allow free placement in pelvis. *Corporal dissection carried out after bladder plate dissection in CPRE*

# Division of suspensory ligaments



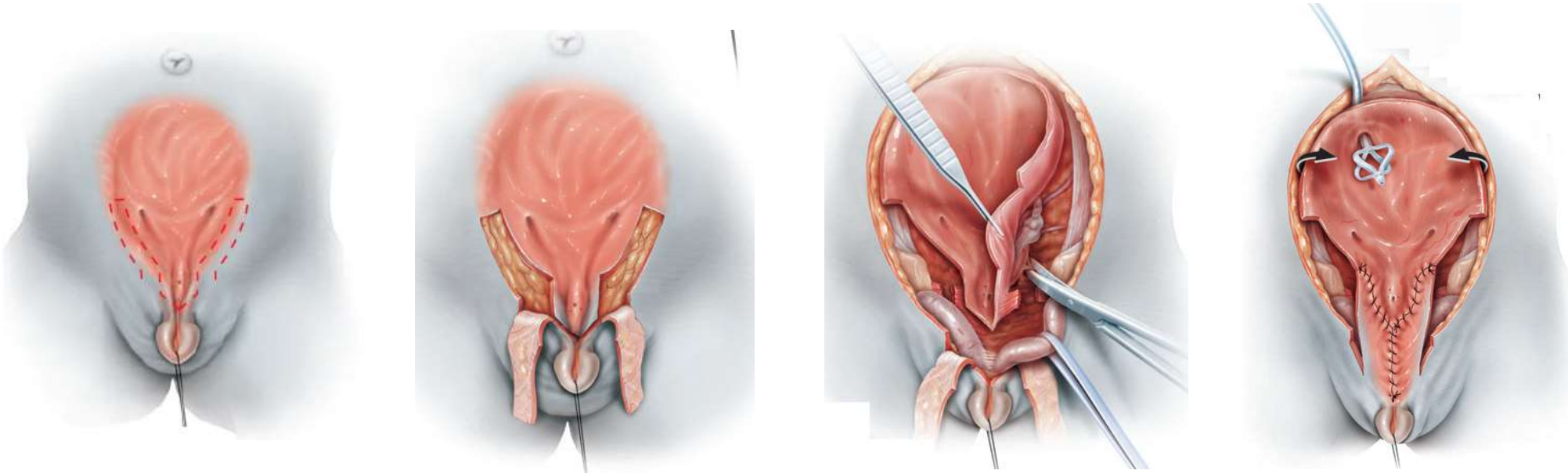
- Continue caudal dissection. Traction on the glans will reveal insertion of corporal body on pubis. Freeing attachments to the **suspensory ligaments** on the anterior part of the inferior pubic rami can increase penile length.

# Division of intersymhyseal bands



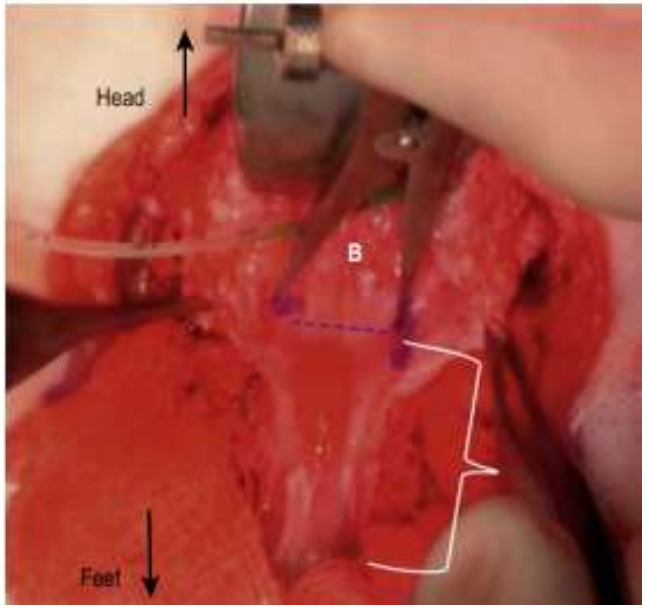
- retract bone laterally to accentuate the **intersymphyseal band attachment** that tether bladder neck. The visualized urogenital diaphragm fibres are taken down sharply with electrocautery to the levator hiatus in their entirety. This will allow appropriate placement of bladder deep in pelvis.

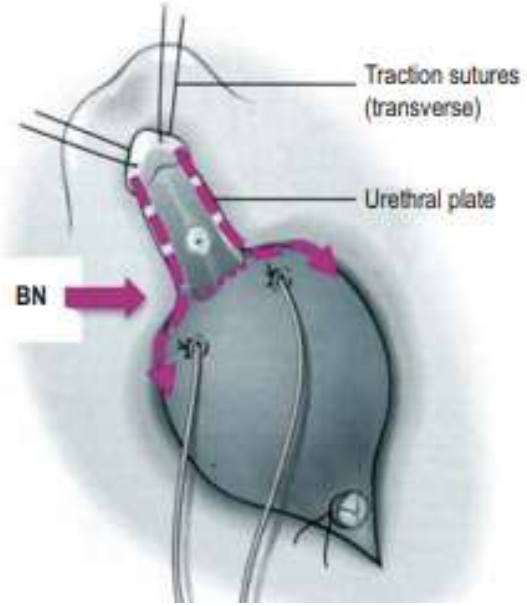
# **modified Duckett paraexstrophy skin-flap technique



- **** short urethra and severe dorsal chordee**: **At this point decision may be made to transect urethral groove**. Requires lengthening by paraexstrophy skin.(further division of suspensory ligament and urogenital bands, dissection behind prostate and lower bladder, suturing flap to prostatic urethra 5-0 PDS) should be used with great caution. The authors have noted a 40% complication rate associated with their use and significant comorbidity

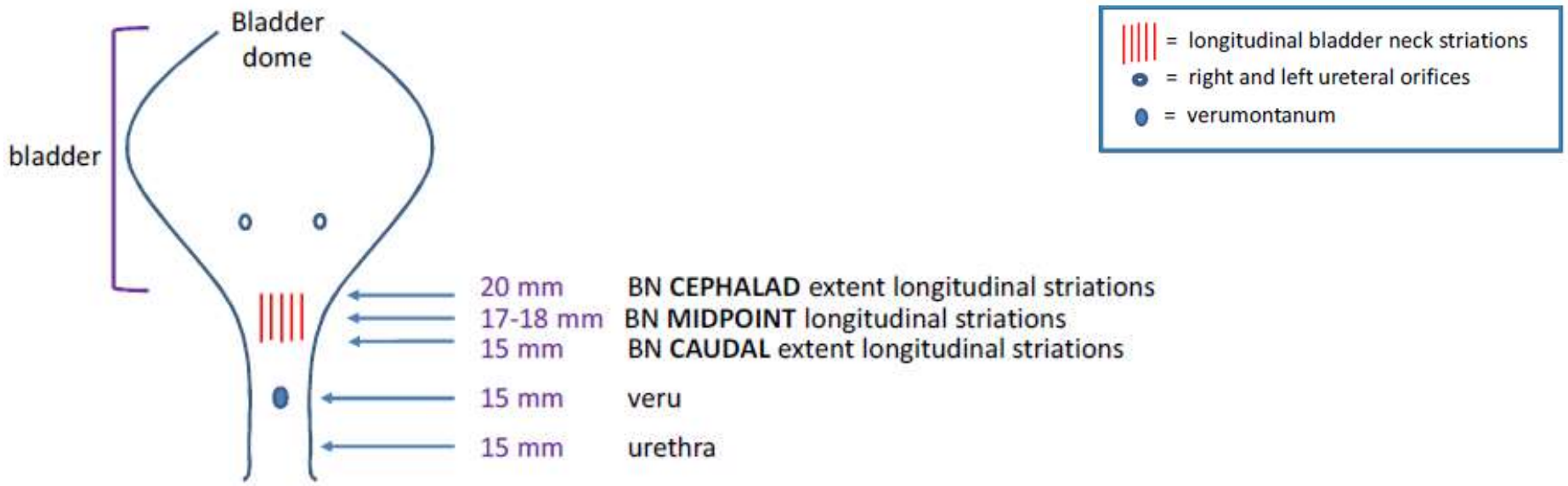
# Elongation of urethra/bladder neck development



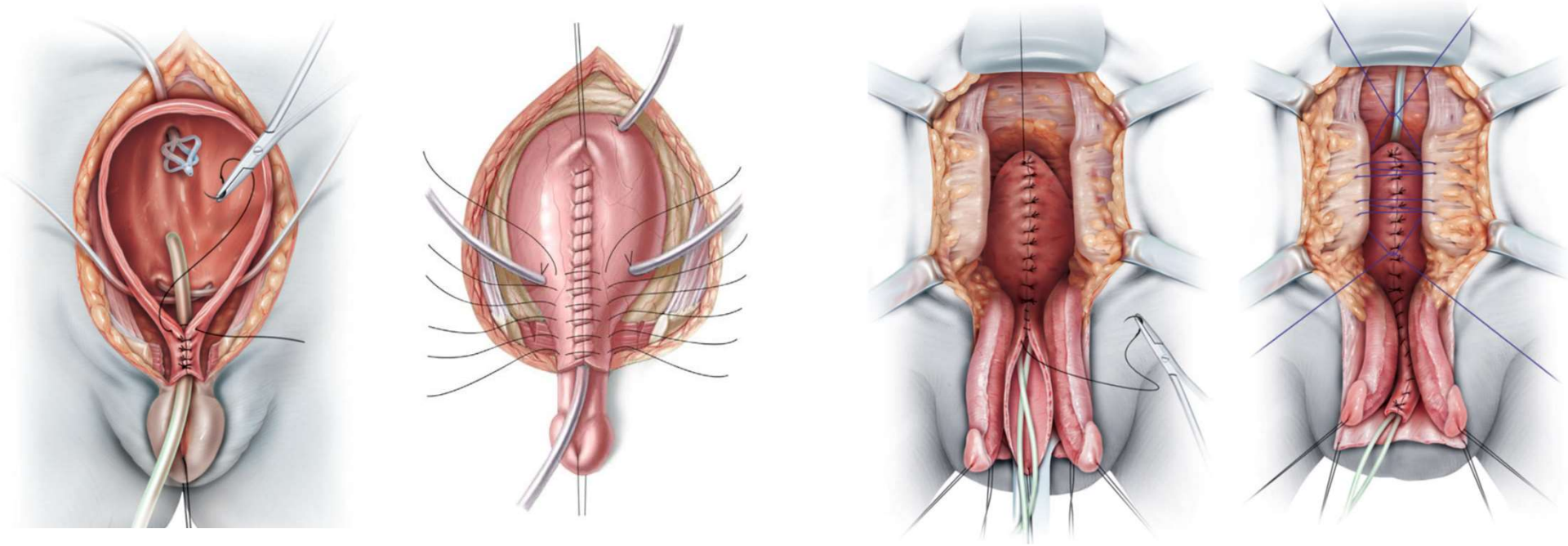

- CPRE: dissection on either side of the bladder neck, but limited to what is necessary to provide a tension-free anastomosis. bladder neck is identified b/n verumontanum and ureteral orifices by ridges or longitudinal folds. Urethral plate being dissected toward bladder neck (dotted line) at a more acute angle. bladder neck width is typically marked out to be 2–3 mm wider than the urethral plate

# Elongation of urethra/bladder neck development



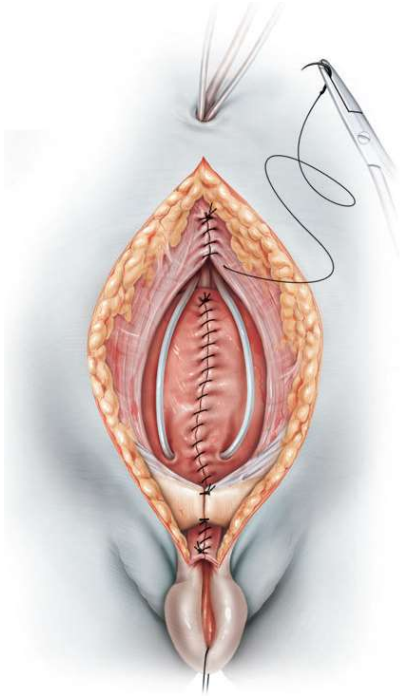

- CPRE: Considerations in bladder neck construction = approximate width for a 2 month old

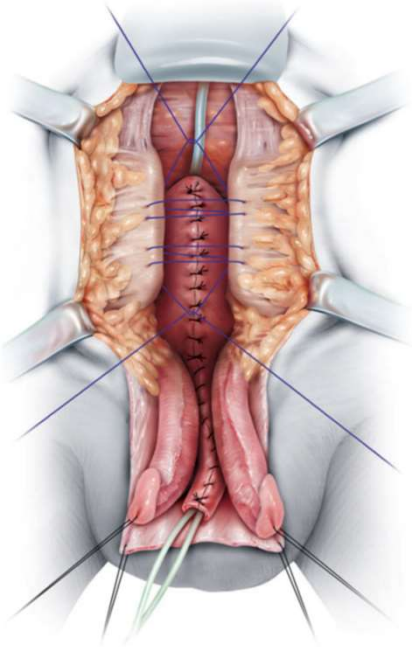
# Bladder closure



- 10-12 fr sound placed to calibrate (enough resistance for adaptation but not to cause upper tract changes). **Bladder** closed with PDS 4-0 interrupted. **Umblical stump** should be excised and **suprapubic tube** passed through neoumblicus. *Urethroplasty also done in CPRE starting from meatus, inverting the mucosa using PDS/Maxon, 6-0 & 5-0. neourethra can be matured to the surrounding lower abdominal and penile skin with interrupted 5–0 in MSRE*.

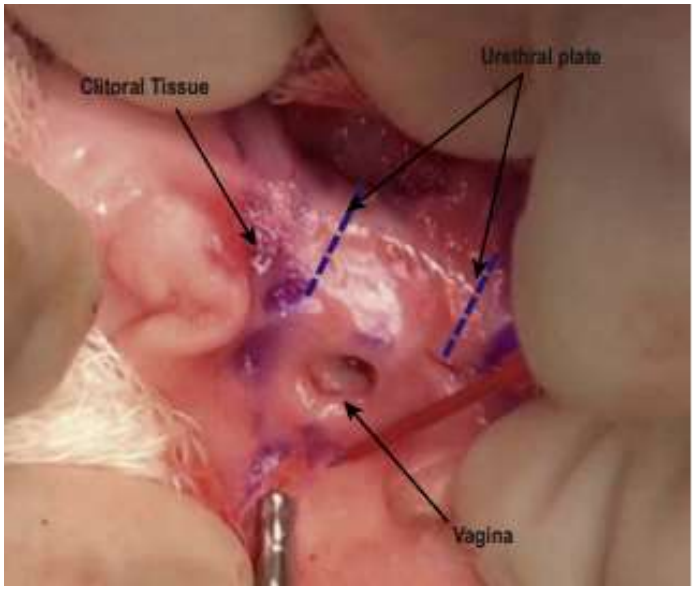
# Pubic bone and abdominal wall closure

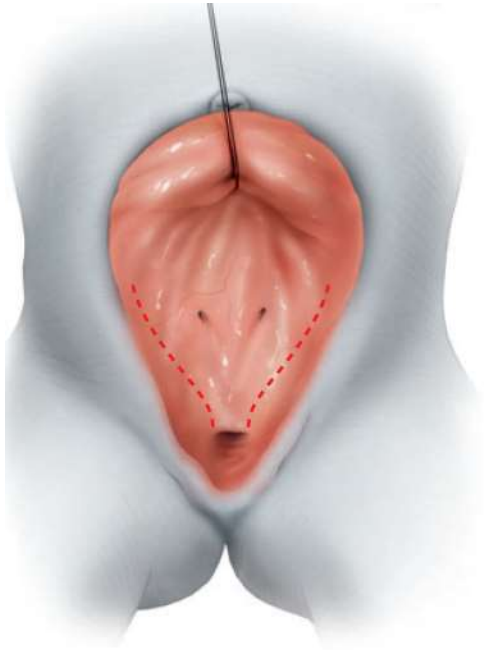




- Manual approximation with pressure over the greater trochanters. Subcutaneous tissue elevated off anterior surface and **Pubis symphesis** closed with 1 or 0 PDS interrupted or horizontal matress. *In CPRE coropora approximation, gansplasty & penile skin closure continue. Before glansplasty, Edge of each hemiglans is trimmed to check for ischemia. If compromised, pubis sutures are released and replaced with less tension.*

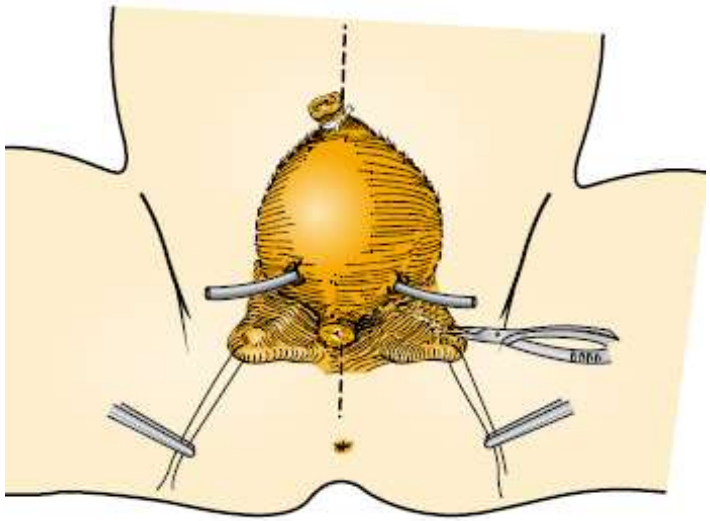
# Modification in girls

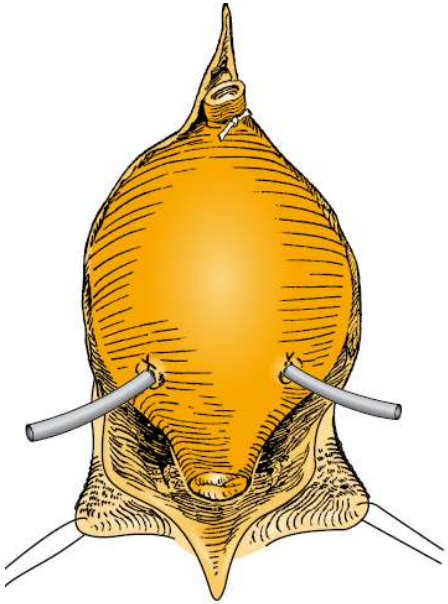


- **Initial incision**: Urethral plate **outlined b/n corporas (2cm wide mucosa)**of the clitoris and incision extends up to the vagina. Incision surrounds vagina and advanced caudally toward the anus along the perineum as a **Y–V advancement**. Labia also advanced as needed .

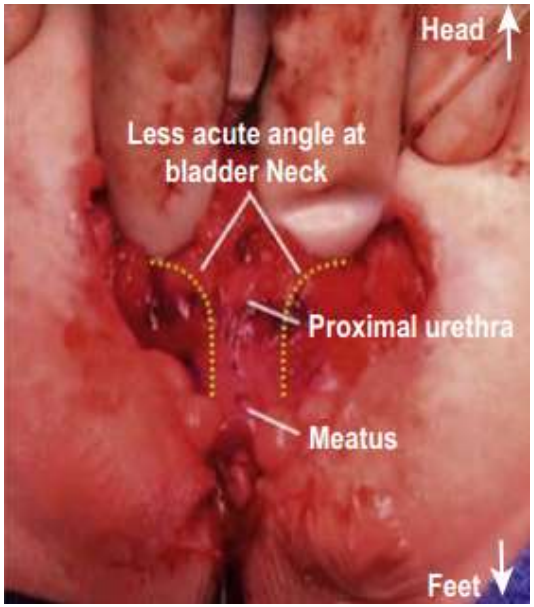
# Modification in girls

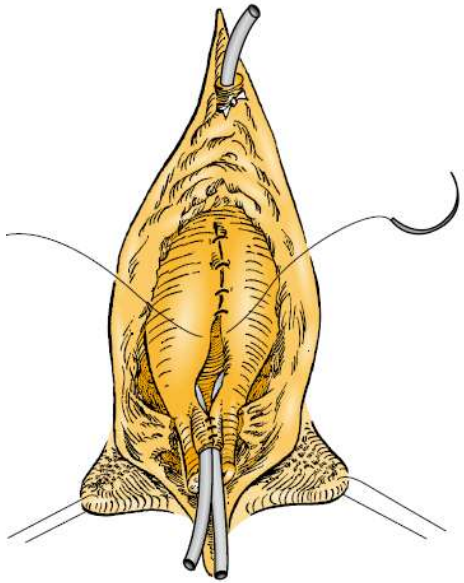




- **Dissection**: Plane of dissection medial to clitoral halves anteriorly and lateral to vagina posteriorly. Following adequate dissection, the vagina and urethraare moved posteriorly in the perineum with a Y-V plasty. The medial aspect of each hemiclitoris is deepithelialized to permit approximation of the two glans clitori and reconstruction of the mons.

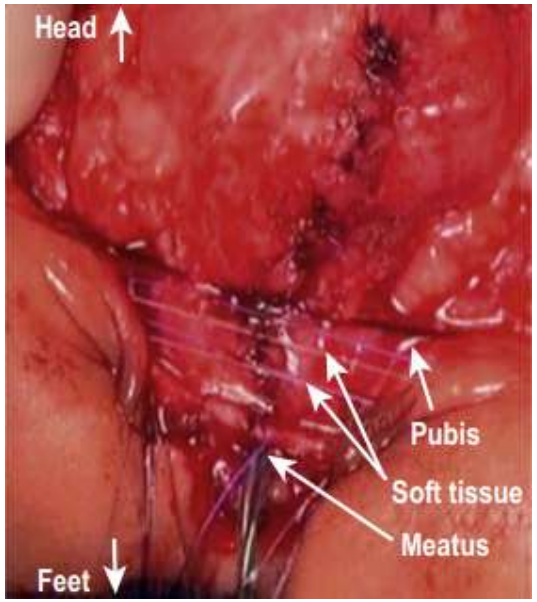
# Modification in girls

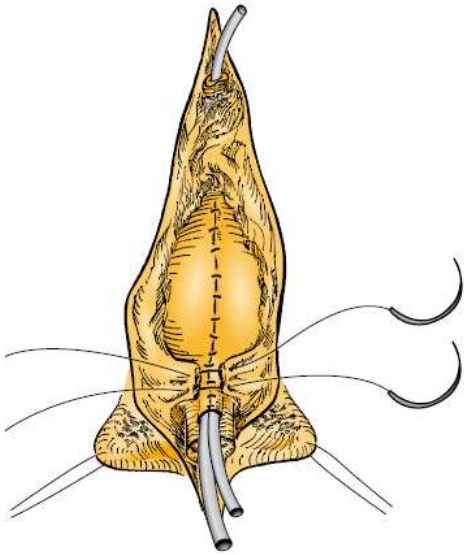


- **Bladder closure and urethroplasty**: Bladder neck gradually tapered into elongated proximal urethra (dotted lines). Urethroplasty with interrupted PDS 6-0, 5-0 stiches. Bladder with 4-0 interrupted,

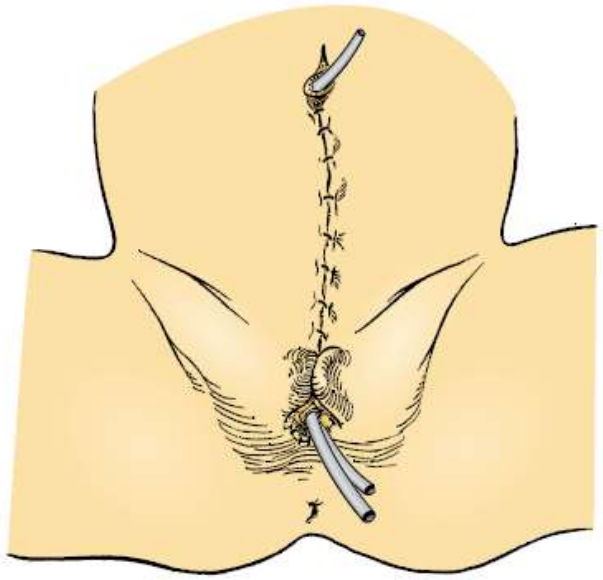
# Modification in girls



- **Pelvic closure**: A “four corner” suture is placed in the **distal urethral** tissue (but not tied) for maturing urethral meatus. **Anterior perineal soft tissue** sutures then placed, followed by sutures on **pubic bone**. Finally  tied in reverse order of placement

# Modification in girls:



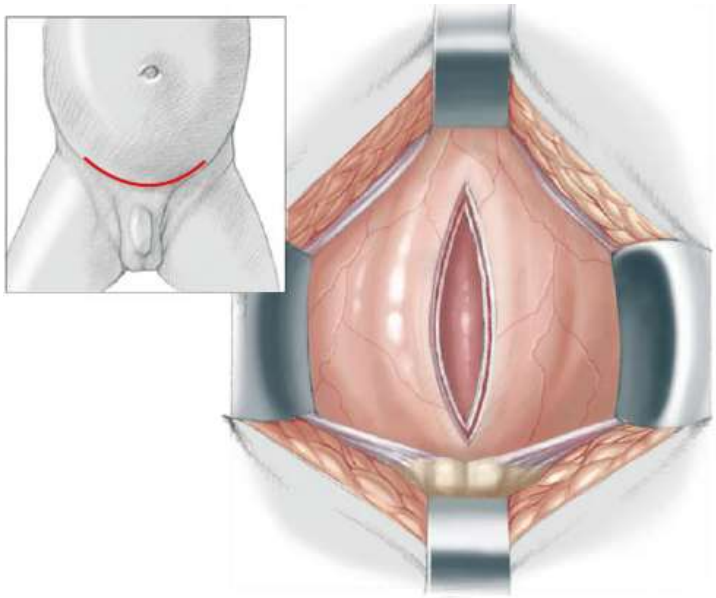
- Skin closure: The neourethra is matured with 5.0 Vicryl sutures and reapproximates the already denuded bifid clitoris medially so that they fuse together after suturing with 7.0 Maxxon suture. The labia majora is advanced posteriorly to the perineum at this time to improve cosmesis of the external genitalia.

# Complete CPRE

- Completed CPRE in a male and female. If ureteral stents are brought out the neoumbilical site, then a Silastic catheter can be used to stent the neourethra. *The urethra is not stented in MSRE stage 1, to avoid necrosis with accumulation of secretions in the neourethra.*

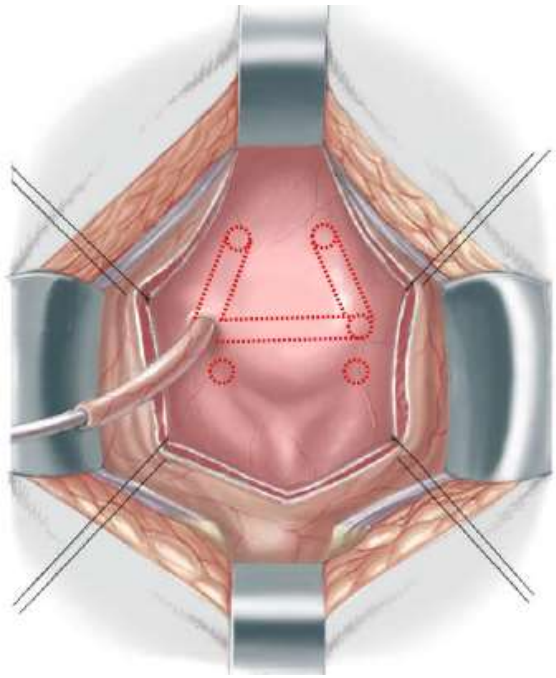
# 3.5 Modified Young–Dees–Leadbetter BNR technique (post MSRE-2)

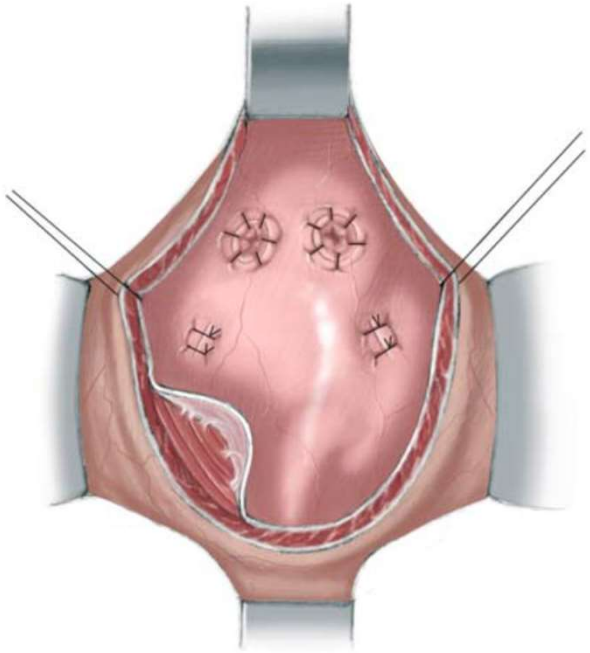
# Initial incision



- Bladder neck is extensively dissected and a vertical cystotomy is made.

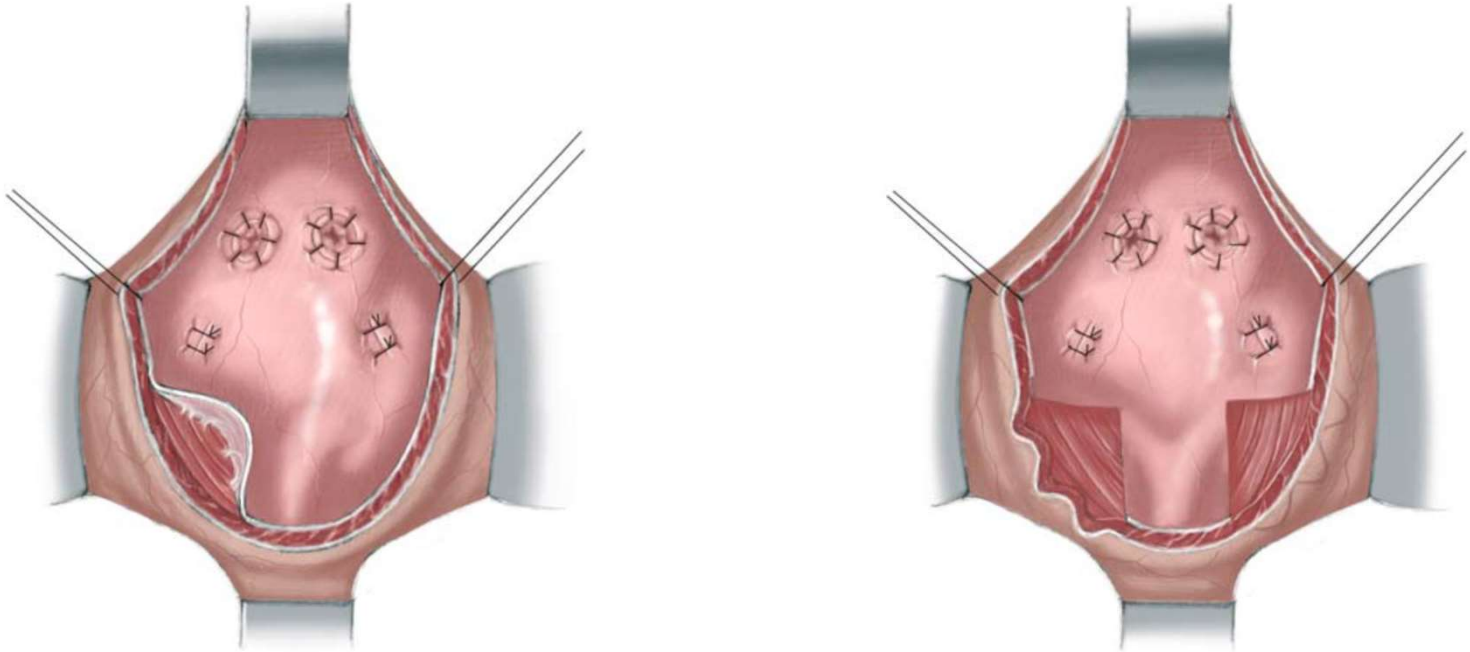
# Ureteral reimplantation





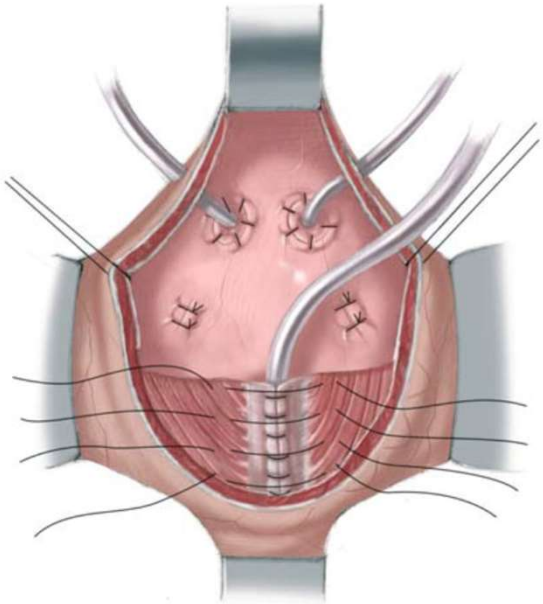
- transtrigonal or cephalotrigonal bilateral ureteral reimplantation

# Mucosal strips



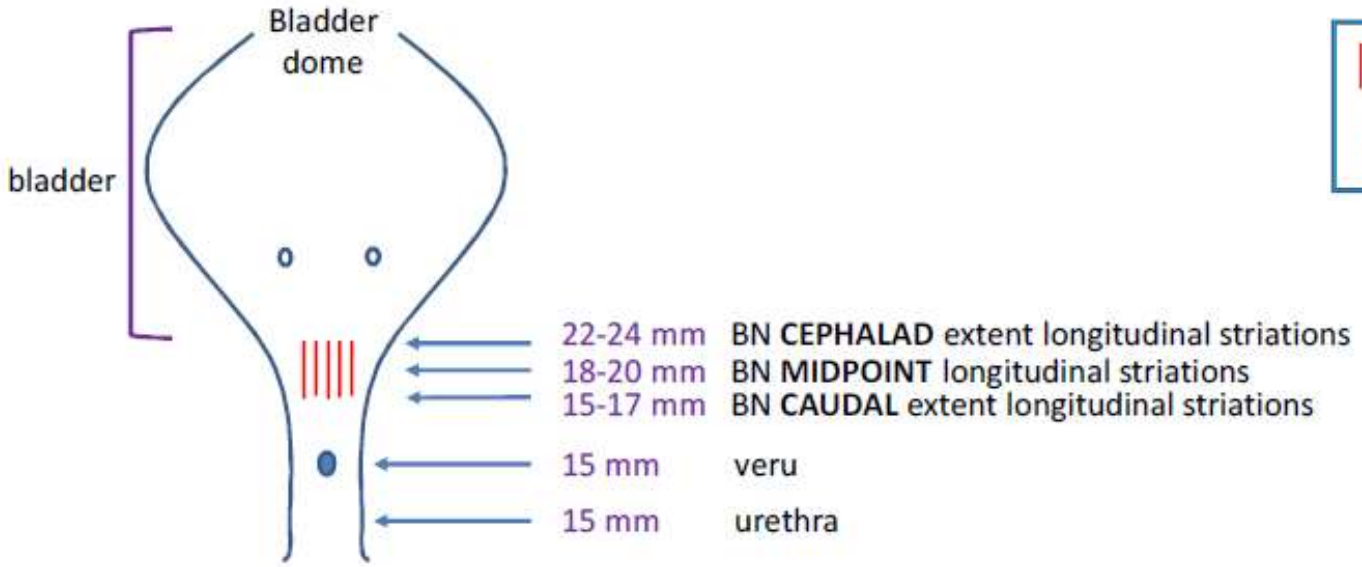
- strip of bladder mucosa approximately 1.5–1.8 cm wide and 3–4 cm long is generated and the lateral **bladder triangles** are **demucosalized**. Use of an epinephrine-soaked sponge during this dissection aids in hemostasis and visualization.

# Tubularization



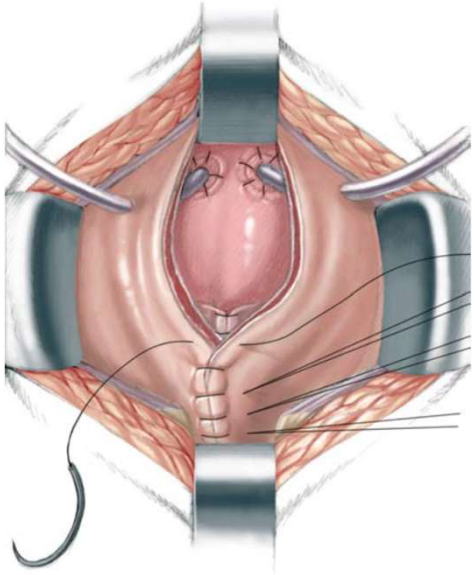
- The neourethra is tubularized over an 8 French stent using interrupted or running vicryl (4-0 or 5-0). The first of these sutures is a 3-0 PDS, demarcating the bladder neck

# Tubularization




- Consideration in BNR: approximate width for a 5-10 yr old

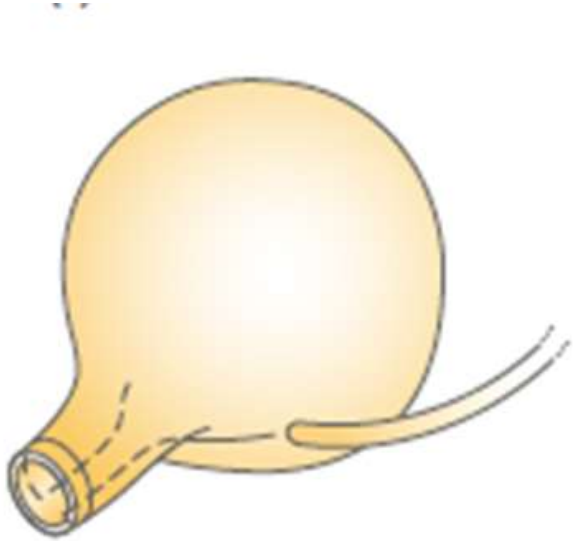
# Reinforcing neobladder neck



- two triangular regions of the demucosalized detrusor muscle are then closed over the mucosal tube in a two-layer “vest over pants” double-breasted technique using 3-0 vicryl. Sutures in the third layer are not cut since they are used in the Marshall– Marchetti–Krantz bladder neck suspension. These sutures are tied over rectus fascia

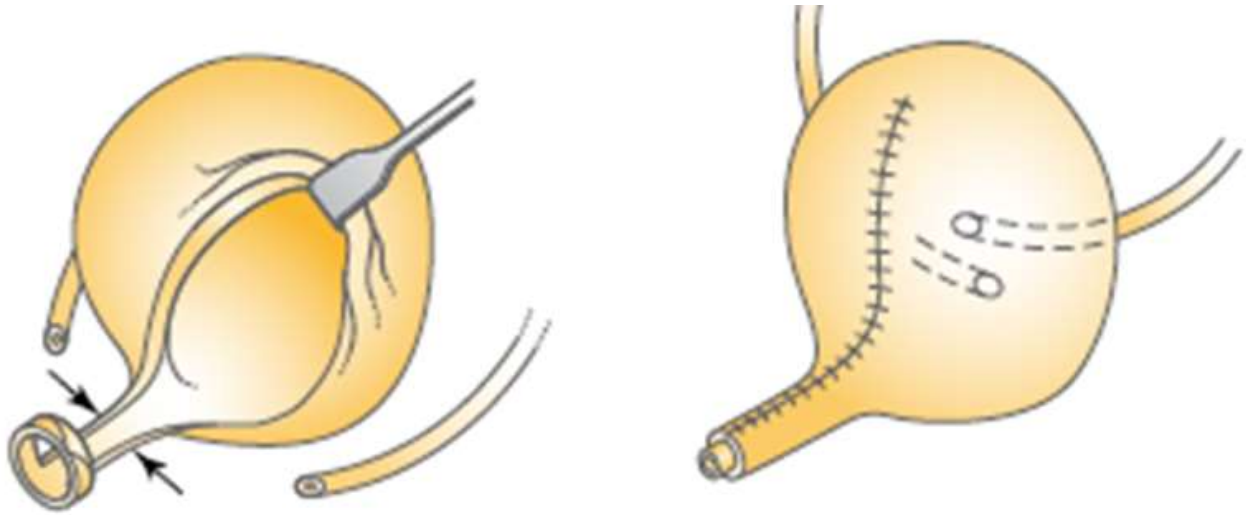
# 3.6 Mitchell BNR technique (post CPRE)

# Incision



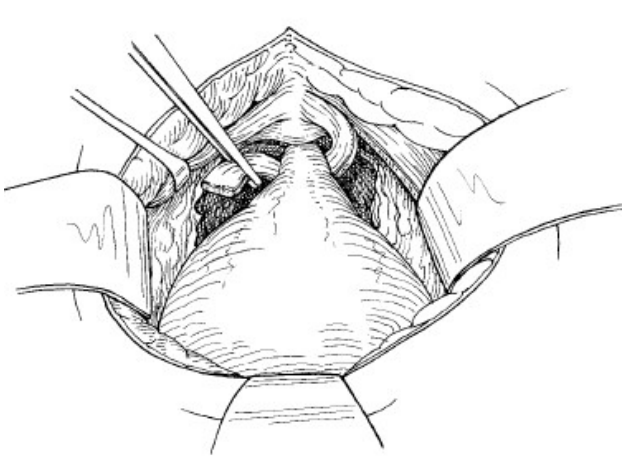
- anterior urethra is incised *transversely and the incision is extended cephalad*. The incision is made full thickness

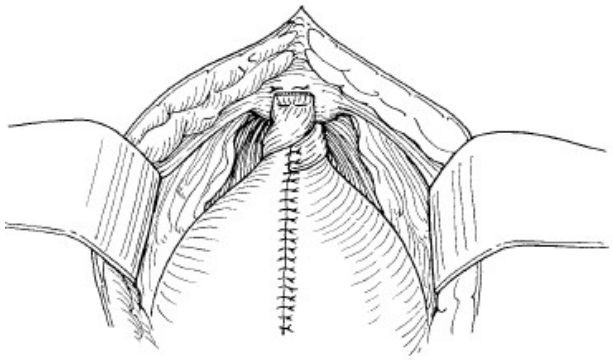
# Tubularization

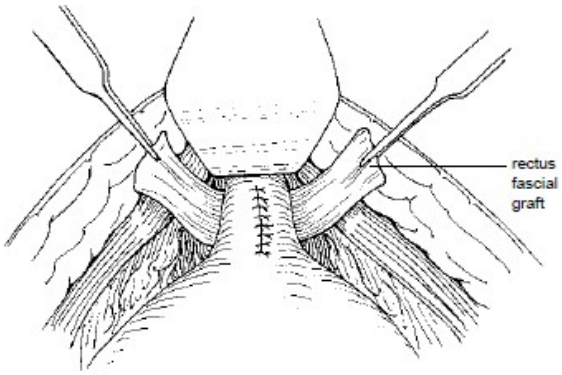


- After cross-trigonal ureteral reimplantation, the urethral strip is tubularized in two layers using a Vicryl or Monocryl suture (4-0 or 5-0) over an 8–10 French urethral catheter. Bladder may be closed incontnuity

# Reinforcing neobladder neck



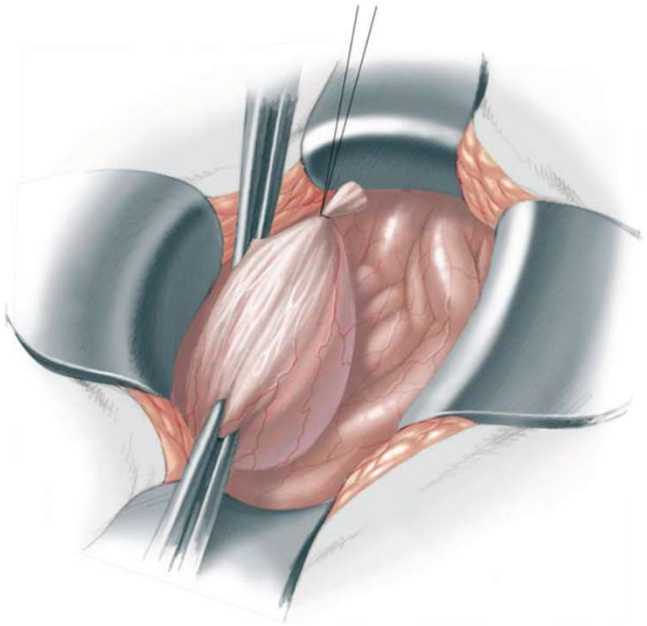


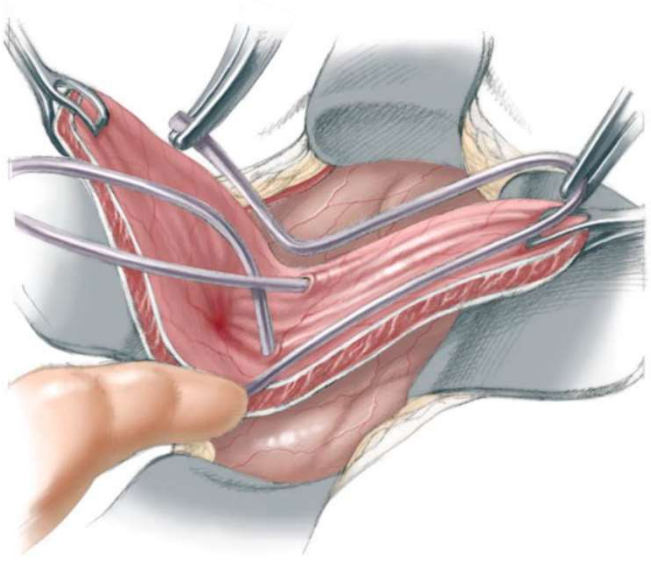



- Following the closure, dissection around the new bladder neck may be performed if a combined bladder neck wrap or sling will be placed simultaneously (**Rectus Myofascial Wrap, Bladder Wall Pedicle or Rectus Fascial Wrap**)

# 3.7 Augmentation cystoplasty (Ileocystoplasty), appendicovesicostomy (mitrofanoff principle) and bladder neck closure techniques

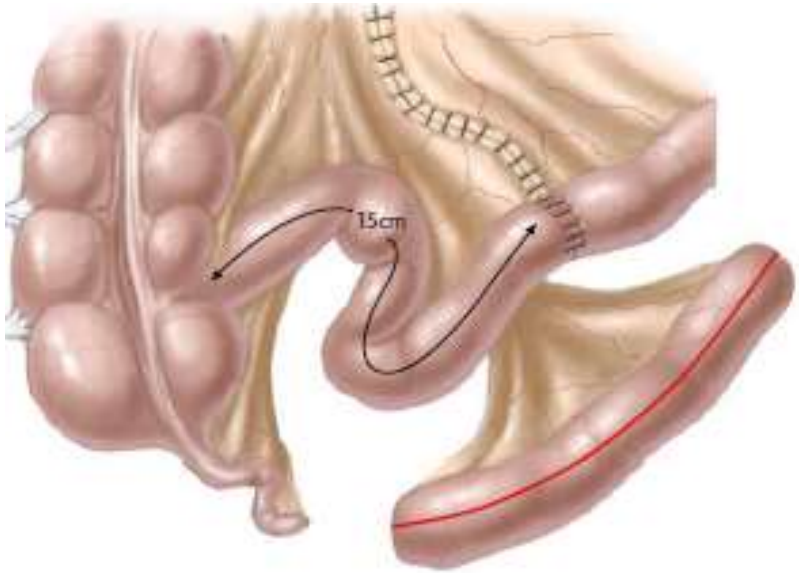
# Augmentation: Bladder exposure and incision





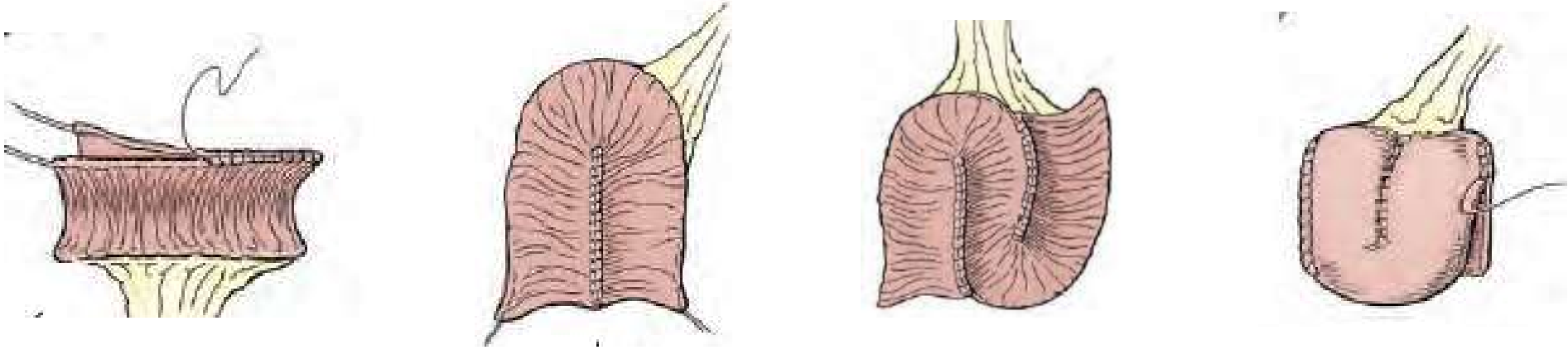
- Lower midline or pfannestine incision used to enter retropubic space. Urachus ligated and held with traction suture. Safe incision is achieved by starting on the bladder dome and opening the bladder using diathermy point down one side at a time. circumference of the bisected bladder is then measured to definethe required length of the bowel segment

# Augmentation: Ileal resection



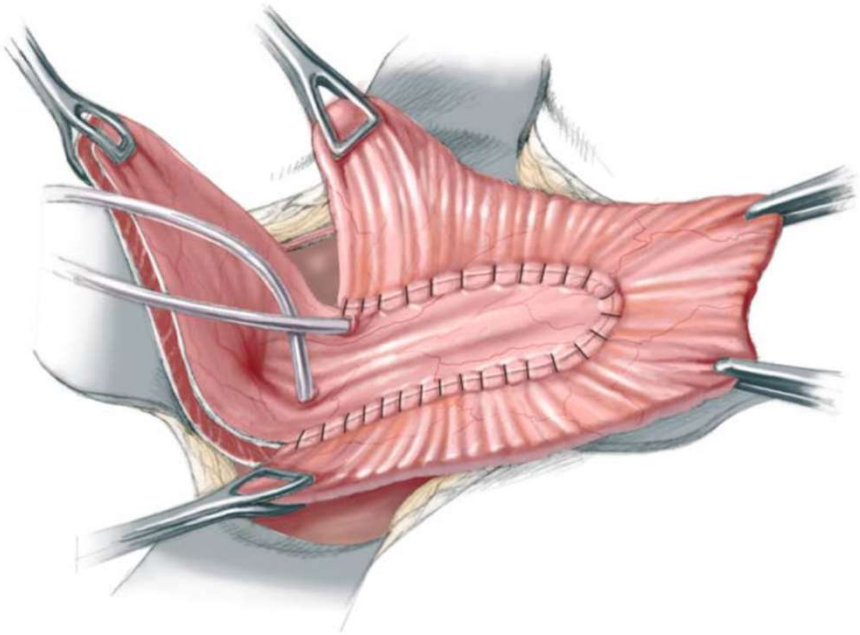

- 20- to 40-cm segment of ileum at least 15 cm from the ileocecal valve is removed and opened on its antimesenteric border

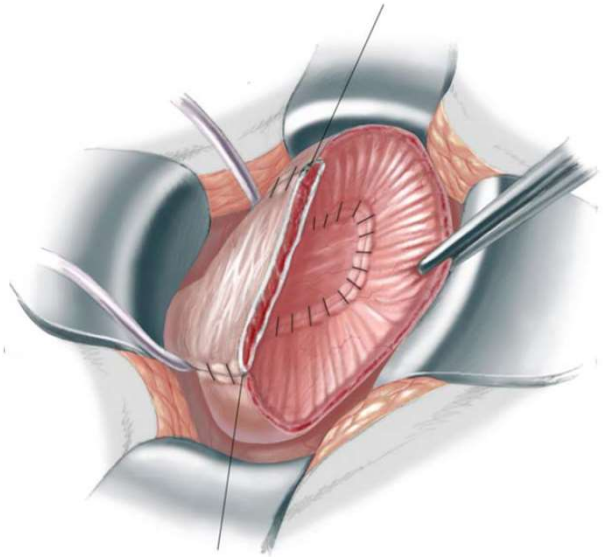
# Augmentation: Reconfiguration



- The opened ileal segment should be reconfigured. This can be done in a **U, S, W** configuration. It can be further folded as a **cup patch**.

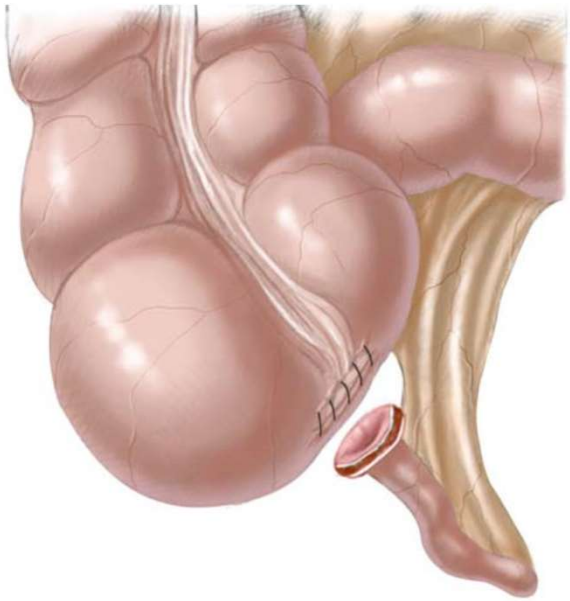
# Augmentation: Bladder Anastomosis





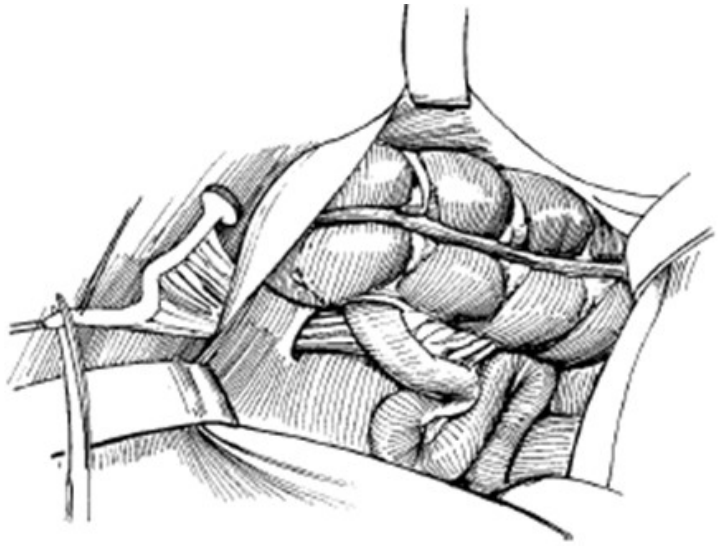
- The reconfigured ileal segment is anastomosed widely to the native bladder.

# Mitrofanoff: Harvesting appendix



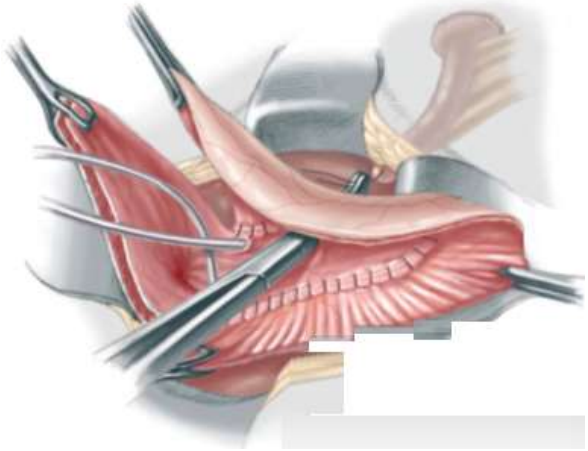
- **appendix is harvested with a cuff of cecum** on a wide pedicle based on the appendiceal artery. Mobilize the cecum for easy placement of the appendix without stretching of the appendiceal vessels. Separate the appendiceal mesentery for a short distance from that of the cecum, preserving all of the blood supply to the appendix. A flap of cecum can be harvested in continuity with the appendix and tubularized to increase the length.  Close defect by 2 layer 3-0 sutures or stapler.

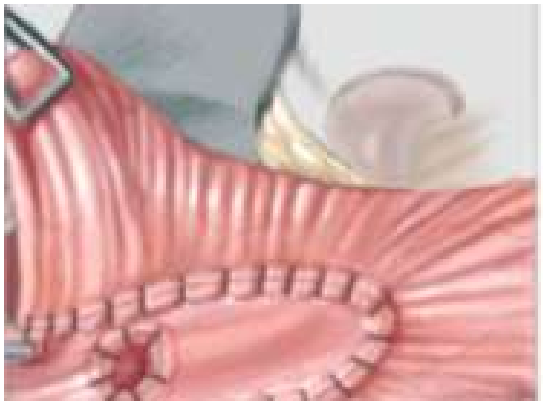
# Mitrofanoff: Extraperitonealize appendix



- **Extraperitonealize the appendix** through a small opening in the peritoneum behind the ileocecal junction. Close the peritoneum with a running suture. Successively trim back the appendiceal tip with Mayo scissors until an adequate lumen is exposed. larger peritoneal dissection of the right lower quadrant is necessary to place the appendix and its mesentery under the peritoneum in the best way.

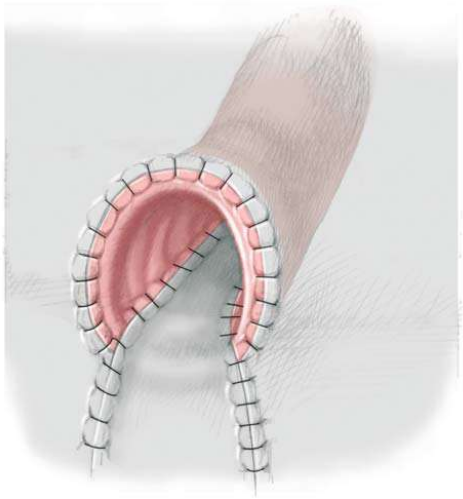
# Mitrofanoff: anastomose to bladder

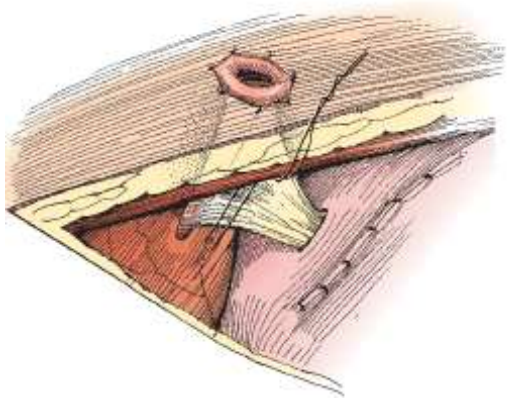




- **distal appendix is tunneled into the bladder** to provide continence after the end is amputated. Tunnel above right uretral orfice as in the Cohen procedure but make it longer (4 to 5 cm). Implant the appendix and its mesentery in the tunnel, stretching it so that it is not kinked The proximal end of the appendix, the **cecal cuff, is brought to the umbilicus or right lower quadrant** as a catheterizable stoma.

# Mitrofanoff: Creating stoma



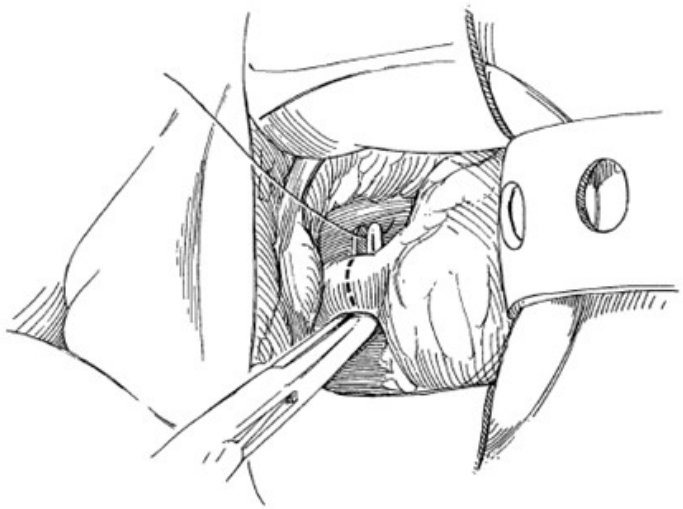


- Pass the appendiceal base through a large opening in the abdominal muscles and a small opening in the skin. Incise a 1- × 1-cm, laterally placed **V-flap** on one side of the umbilicus. Incise the antimesenteric wall of the appendix to correspond to the skin flap. Suture the V-flap into the appendiceal V-incision with interrupted 4-0 polyglycolic sutures. Leave a 10-12 F polyethylene catheter through the appendix. **bladder hitch** to the anterior abdominal wall is useful to avoid kinking of the appendix between the abdominal muscles and bladder and also if the appendix is short.

# Mitrofanoff: alternative to the appendix

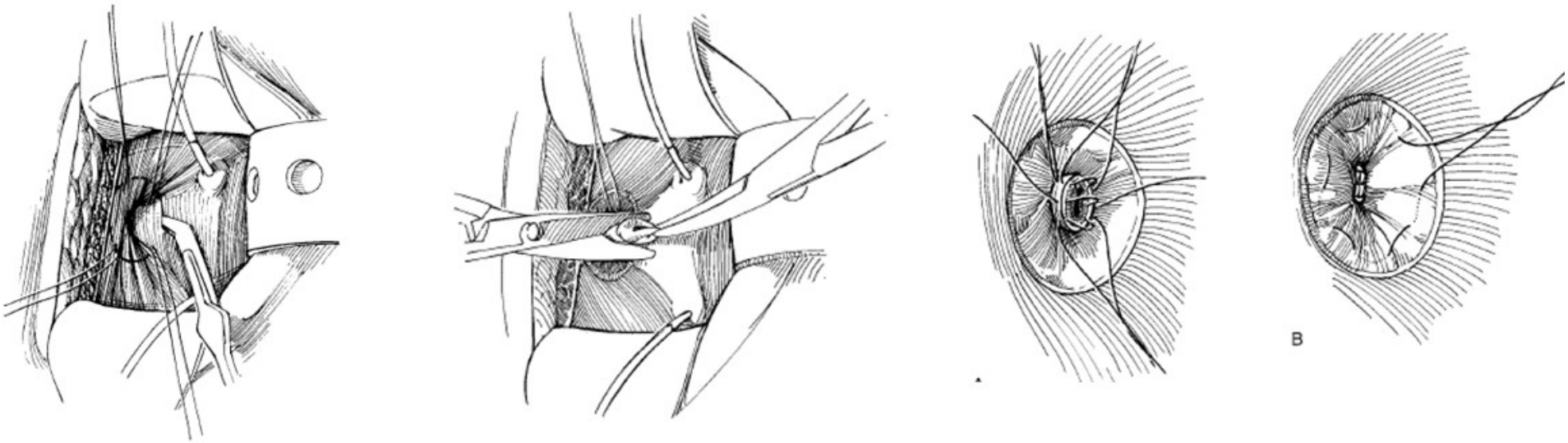

- **Yang-Monti procedure**: transversely tubularized ileum

# Bladder neck closure: extravesical approach



- Enter the retropubic space, and free the urethra by dividing the pubourethral ligaments. Dissect the urethra distally from the prostate or vagina with the aid of a finger in the rectum or vagina, respectively. Elevate the urethra from the vagina or rectum by sharp dissection with a knife to include the adventitia. The longer the segment freed by sharp dissection, the easier the procedure. Divide the urethra and ligate both ends, leaving the proximal suture long. Consider placing omentum  between the distal urethral stump and ligated bladder.

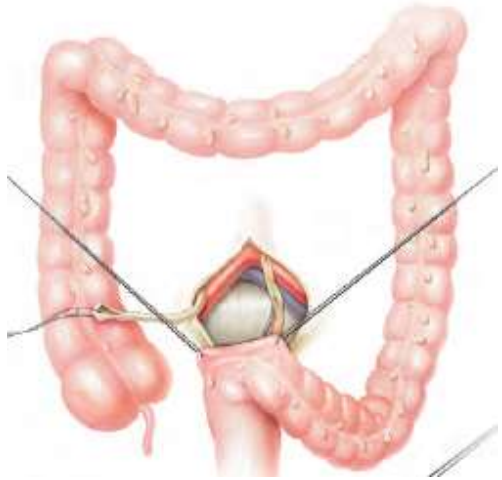
# Bladder neck closure: intravesical approach




- Place traction sutures around vesical neck. Cut through the epithelium circumferentially 1 to 2 cm away from the outlet with hooked knife, and free up the epithelial margins with scissors. Trim the freed epithelium flush with the outlet. Place a 3-0 or 4-0 purse-string suture 1 cm from the urethra and a second circumferential suture 1 cm outside the first to invert urethra. Close the epithelium of the bladder over the repair with interrupted 4-0.

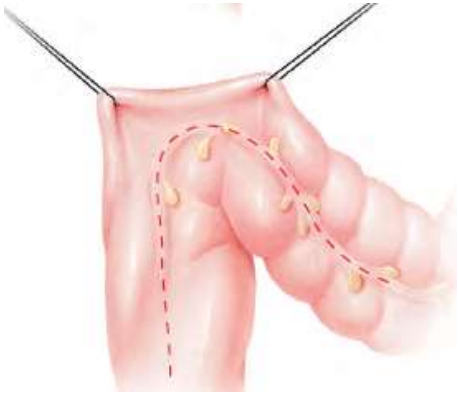
# 3.8 Mainz pouch II Technique

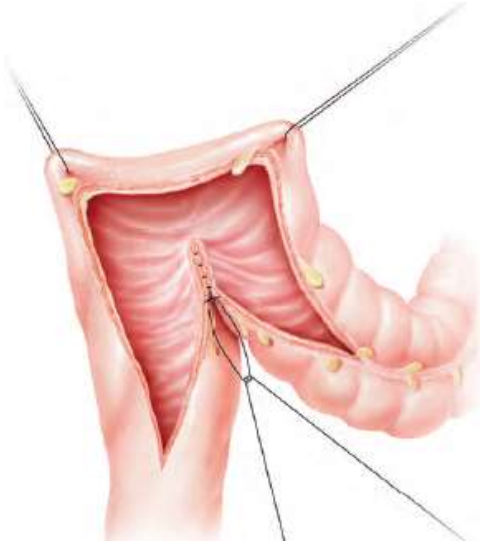
# Incision

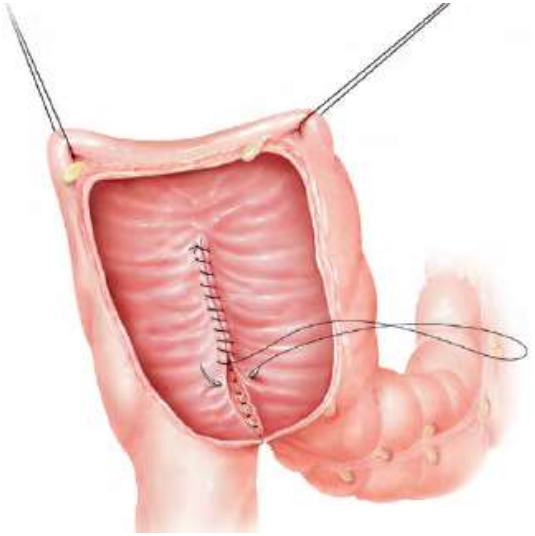


- Supine position. Insert rectal tube. Midline incision. Excise the bladder first, leaving the transected ureters to drain freely. Pack the small bowel in the upper part of the abdominal cavity. Two stay sutures are placed into the rectosigmoid at a position, where it reaches tension free to the promontory,

# Sigma rectum pouch

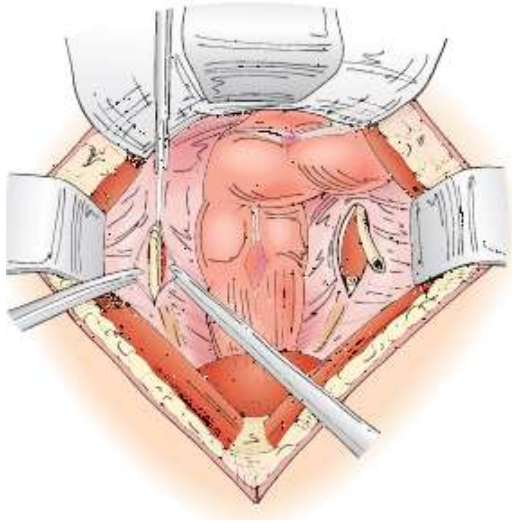


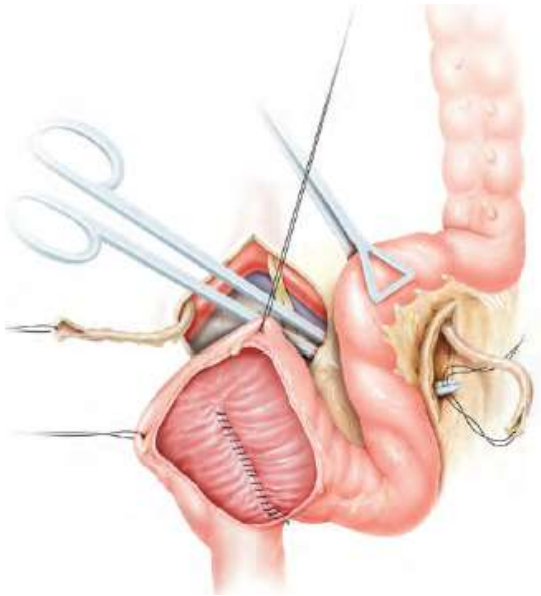




- Sigmoid detubularized by opening along the anterior tenia. Then mechanically cleansed by several wet swabs with gentamycin. Reconfigured to spherical shape by anastomosing posterior wall side to side

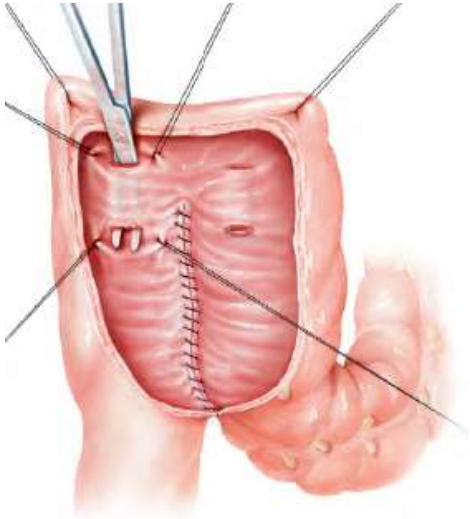
# Mobilization of ureters





- Both ureters are mobilized up into Gerota fascia. preserve the ureteral adventitia with its longitudinal blood supply. Vascular connections between the gonadal vessel and the ureter should be preserved. The left ureter is pulled through the mesentery of the descending colon or sigmoid colon at a location where compression from the inferior mesenteric artery or another major vessel of the mesentery is unlikely into a position in front of the promontory so that a straight course without kinking is achieved.

# Tunneling

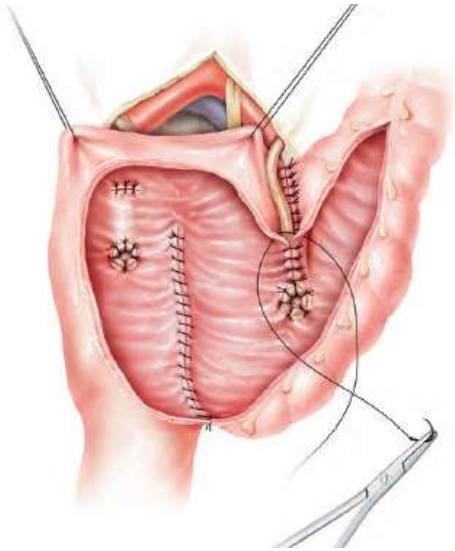


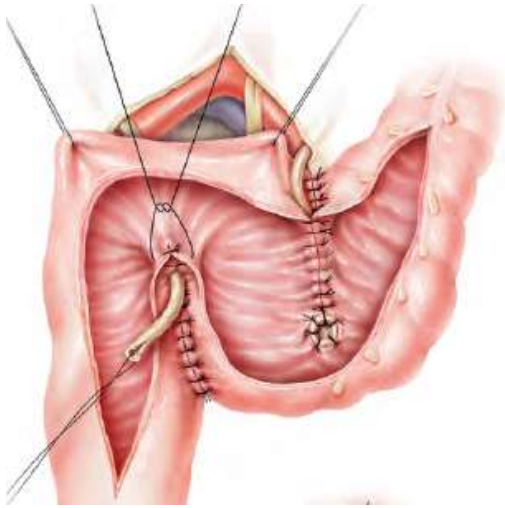
- four stay sutures are placed over a distance of 4 cm. small segment of mucosa is excised. bowel wall may be elevated by the index finger. Previous submucosal injection of a small amount (1–2 mL) of saline eases separation of mucosa from muscularis. Incision of the mucosa over the tip of the curved clamp determines the distal end of the submucosal tunnel

# Implantation

- At the proximal end of the submucosal tunnel, the muscular layer of the posterior wall of the pouch is incised crosswise to allow an unobstructed pull-through of the ureter. Kinking/angulation should also be avoidedl

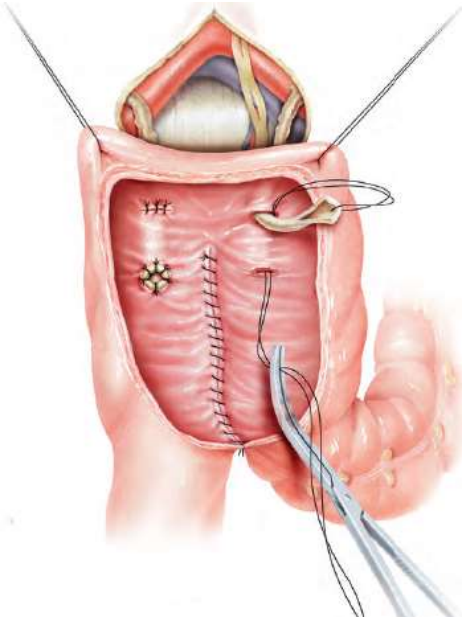
# Implantation for dilated ureter

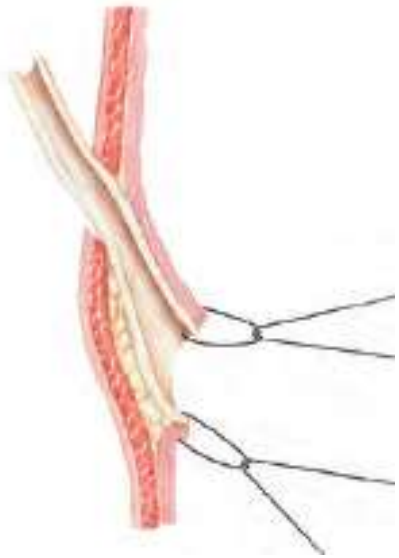




- serosa lined extramural tunnel: **for the left ureter** a third sigmoid loop is required (S shape of the pouch), which is opened antimesenterically. Close to their mesenteries, the seromuscularis of the proximal loop is anastomosed side to side to the seromuscularis of the following loop. anterior aspect of the tunnel is completed by double-layer sutures of the intestinal margins. **The right ureter** is implanted on the first side to side anastomosis.

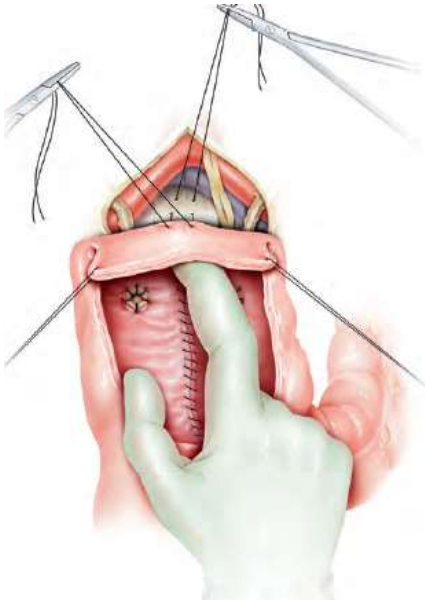
# Anastomosis

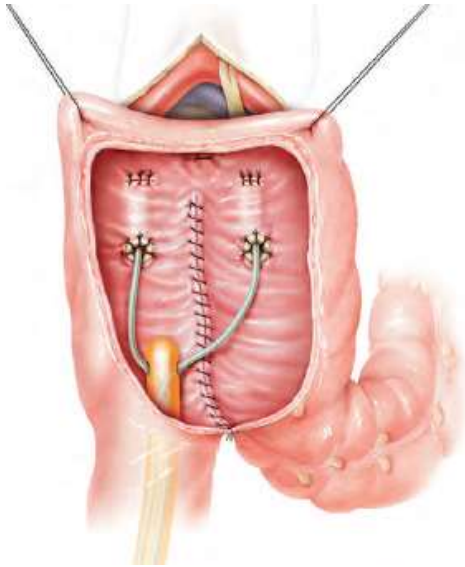




- Ureter spatulated and anastomosed.

# Closure





- back wall of the pouch is **fixed** with one or two nonabsorbable sutures (4-0 Prolene) through the seromuscularis to the periosteum of the promontory on the right side of the mesentery of the sigmoid**. 6 F stent** inserted through side holes in rectal tube. **Rectal tube** secured to the perianal skin by a stitch. Anterior wall of pouch  is closed in two layers (mucosa and seromuscular). mesenteric windows are **closed**, and the pouch is covered with **greater omentum.** Jackson-Pratt **drains** are placed behind the pouch into pelvis if cystectomy was performed.

# 4- Cloacal exstrophy

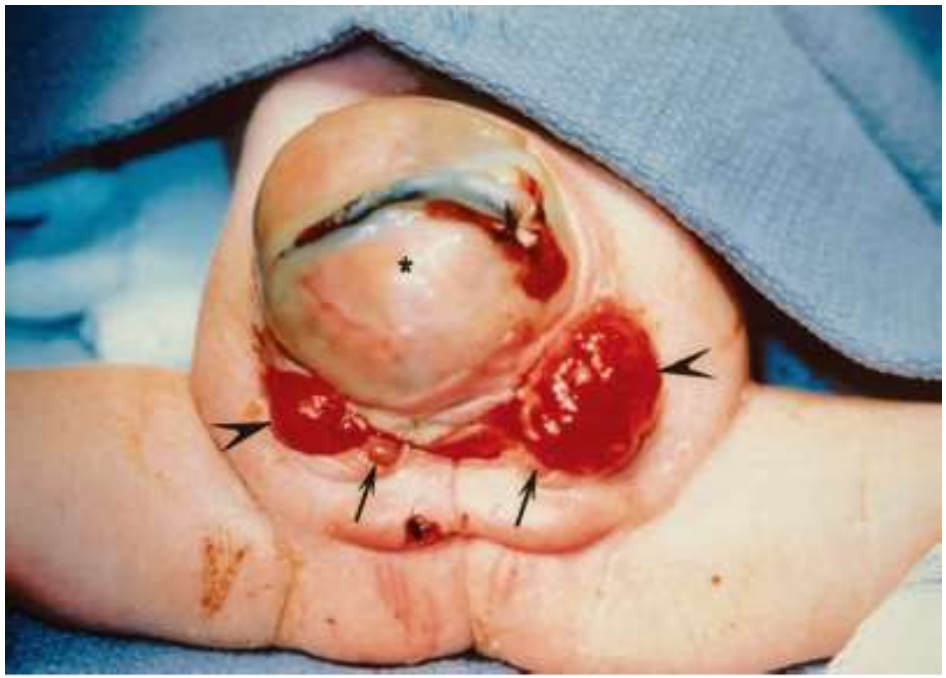
# 4.1 Epidemiology

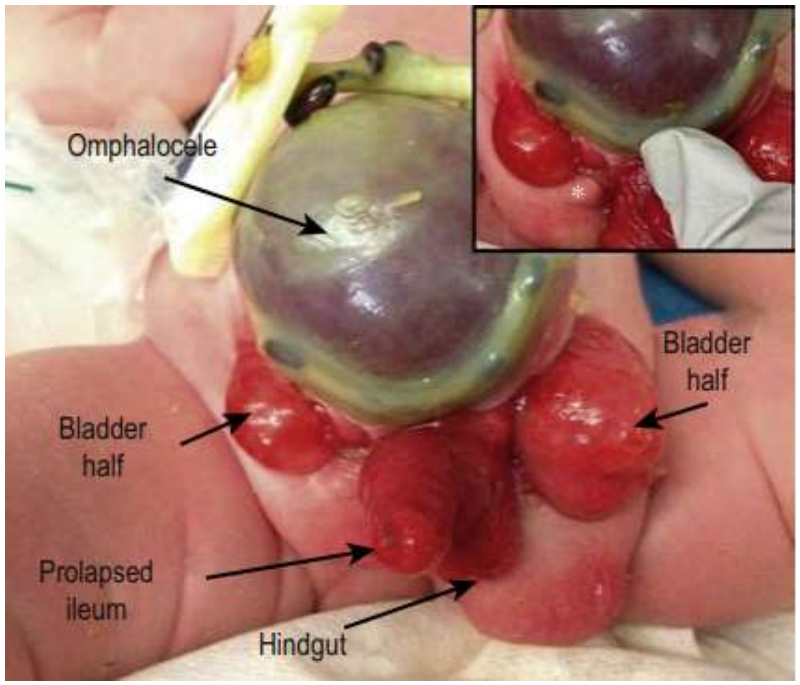
- Incidence : 1 in 300,000
- Higher in female and stillborn
- Higher mortality due to electrolyte, malnutrition and sepsis
- most will require TPN for pre-op optimization

# 4.2 Anatomic features

- **bladder plate** associated with CE is divided in half by the hindgut plate
- similar, but more severe, **pelvic floor** musculature  abnormalities
- the **penis** is often separated into two hemiphalluses due to the wide pubic diastasis
- **Cryptorchidism** is generally the rule.
- **Müllerian fusion anomalies** in 85%, Vaginal agenesis in one-third
- **ileocecal junction** are open plate, **Hindgut** is truncated and terminates at perineum (blind ending microcolon)
- **omphalocele**, imperforate anus, rudimentary hindgut, malrotation, short gut syndrome rudimentary hindgut
- **Upper urinary tract** anomalies 48% (upj, horseshoe, ectopoic kidney)
- **neural tube defects 85-90%**, vertebral anomalies, spinal myelodysplasia, spinal dysraphism, and tethered cord

# 4.2 Anatomic features

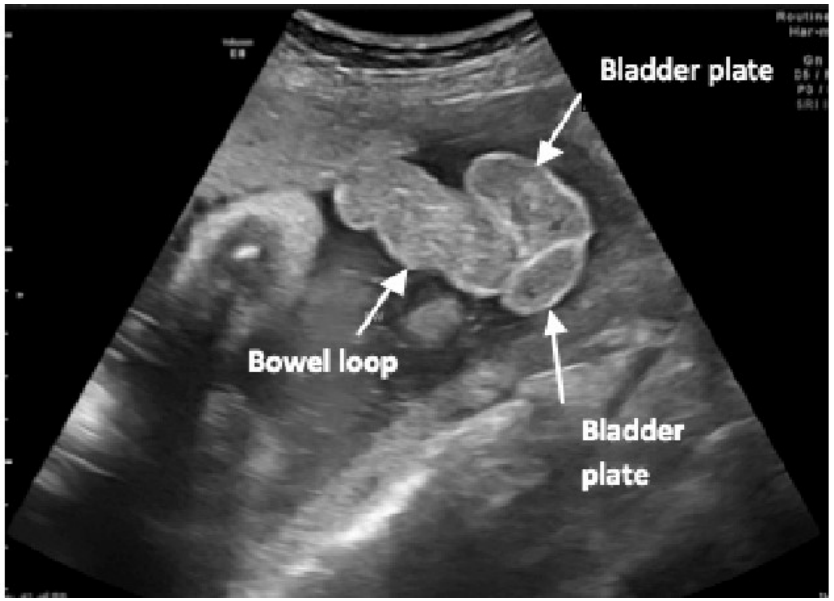





- **Cloacal exstrophy AKA (OEIS complex)** - omphalocele, exstrophy, imperforate anus, spinal defect
- Ileum may intussuscept into hindgut creating appearance of the "**trunk of an elephant's face**"

# 4.3 - Management

# Prenatal care




- **Antenatal ultrasound** might show prolapsed ileum which may look like an “elephant trunk”.
- Labor should be induced to have **predictable delivery** arranged at a specialized center

# Pre-op consideration

## Initial care

- Abdominal film
- Ultrasound (upper urinary tract, internal genital structures and spinal cord)
- Neurosurgery consultation within the first 48–72 hours
- Karyotyping

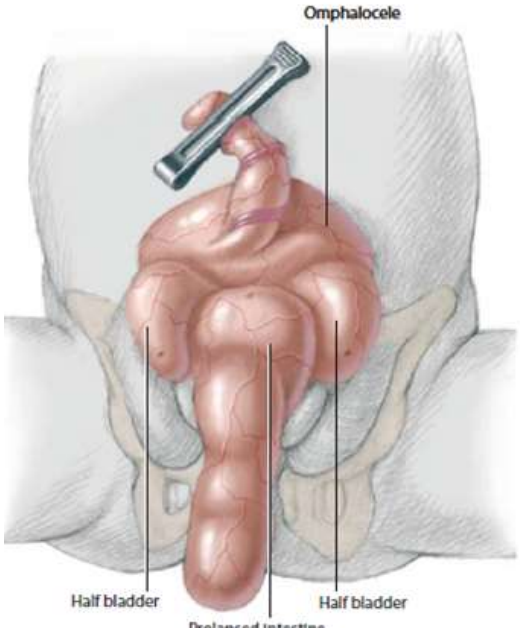
# Pre-op consideration

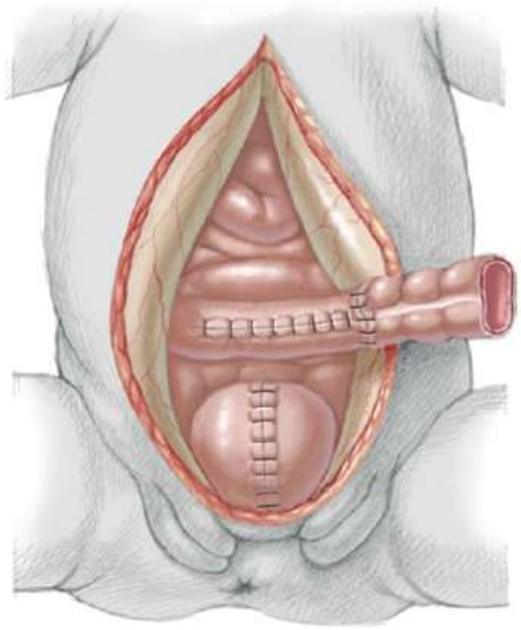
- Short-gut syndrome
  - usually present, even in those with a normal length of bowel.
  - effects of malabsorption and fluid loss especially early in life.
  - hindgut should be placed in continuity with the intestine during initial operation.
  - will also promote growth so that it can be used in later reconstruction urinary/vagina
- Gender assignment
  - Many males have an unreconstructable, diminutive phallus and UDT
  - may elect to have their son undergo a gonadectomy and be raised as a female.
  - Controversial issue given concept of androgen imprinting.

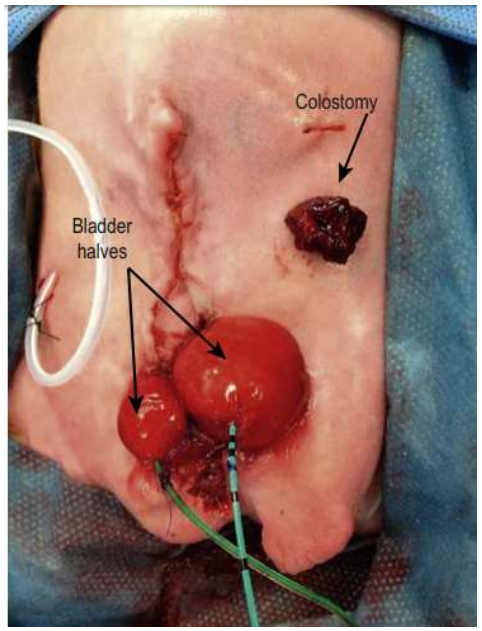
# Surgery

- **Stage 1** – 48-72hr
  - **tubularization** of the cecal plate
  - **Diversion** of rudimentary hindgut proximal to an imperforate anus
    - important to preserve any rudimentary hindgut and to not discard any bowel,
  - **omphalocele repair**.
  - **Primary closure** (Bladder closure + genital revision + osteotomy)
- **Other approach** is delaying primary closure for 2$^{nd}$ stage (6mo)
  - associated anomalies, not tolerating surgery, small bladder, too large diastasis
- **Subsequent stages** follow principles of BE closure
- Majority require **augmentation** and CIC/CUD

# First stage CE repair





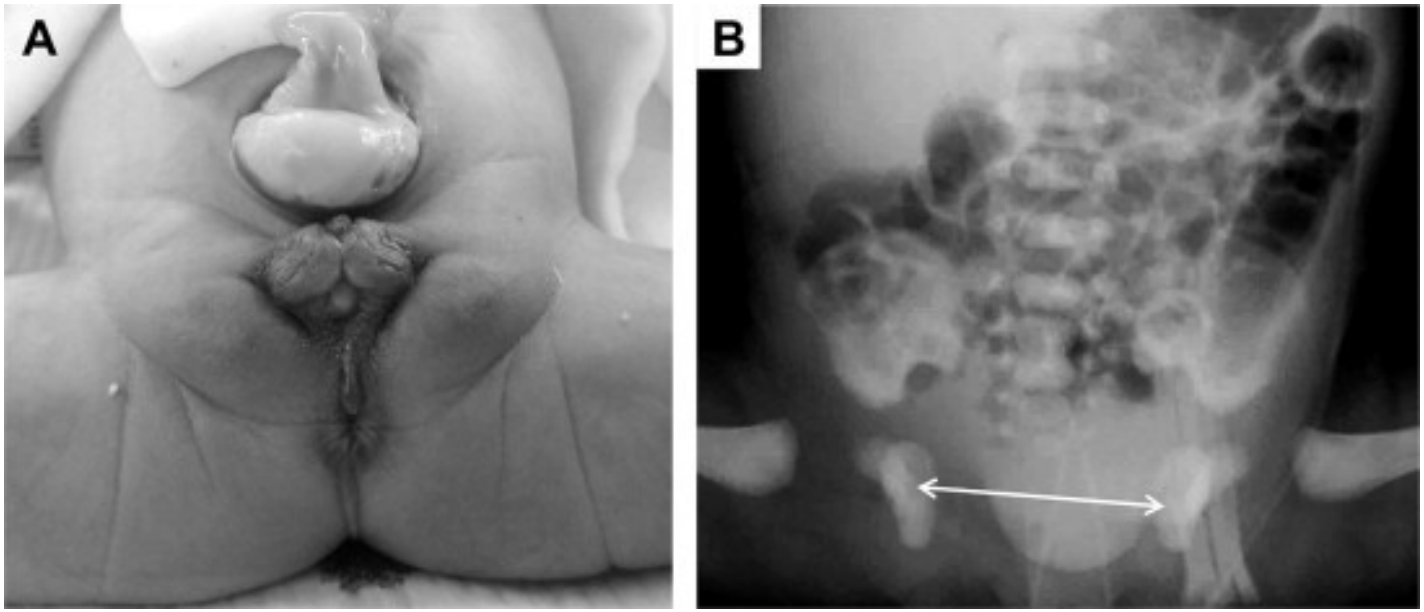

- **First stage**: creation of a colostomy, closure of the omphalocele, and approximation of the bladder plate halves in the midline converts CE to CBE. Alternatively, primary closure of bladder with osteotomy and genital revision can also be done.

# 5- Exstrophy variants

# Variants

- some, but not all, of the typical features of bladder exstrophy or CE
- may have anatomically normal genitalia despite their bladder and colon anomalies
- lower incidence of spinal abnormalities and a higher rate of fecal continence compared with their classic presentations
- Two patient groups had better outcomes than those with classic presentations—superior vesical fissure and skin-covered cloacal exstrophy.
- Because sphincter is intact, other than superior vesical fissure, all should be managed with formal exstrophy closure at birth and followed inthe same manner as their classic presentations.

# Pseudoexstrophy

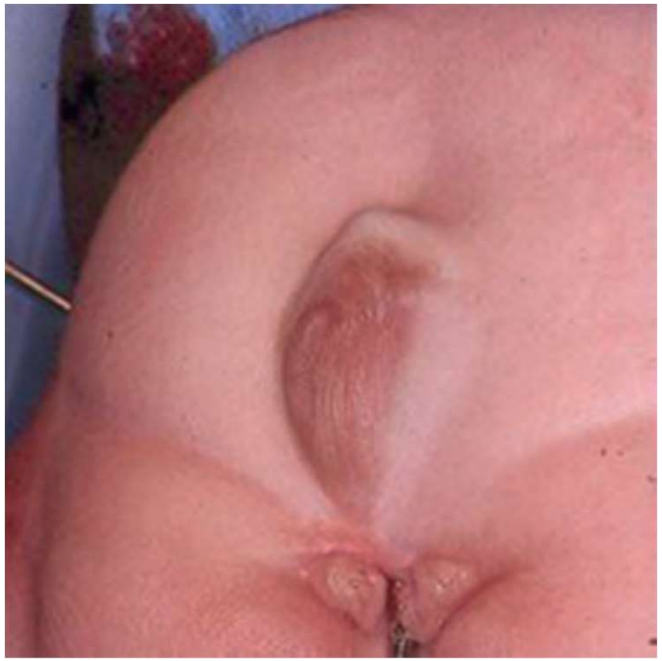



- characteristic musculoskeletal defect of the exstrophy anomaly with no major defect in the urinary tract. low-set umbilicus and divergent rectus muscles that attach to the separated pubic bones. mesodermal migration has been interrupted in its superior aspect only, thus wedging apart the musculoskeletal elements without obstructing the formation of the genital tubercle

# Covered exstrophy


- **Pseudoexstrophy + isolated ectopic bowel segment** on inferior abdominal wall near the genital area, which can be either colon or ileum with no connection with the underlying gastrointestinal tract and only epispadias in the male.

# Covered exstrophy



- Note urethral  sound in the bladder and the subcutaneous position below the skin of the abdominal wall. one could actually see the bladder through a thin membrane of lower abdominal skin. No bowel is sequestered on the abdominal wall, as has been reported in some patients with this variant

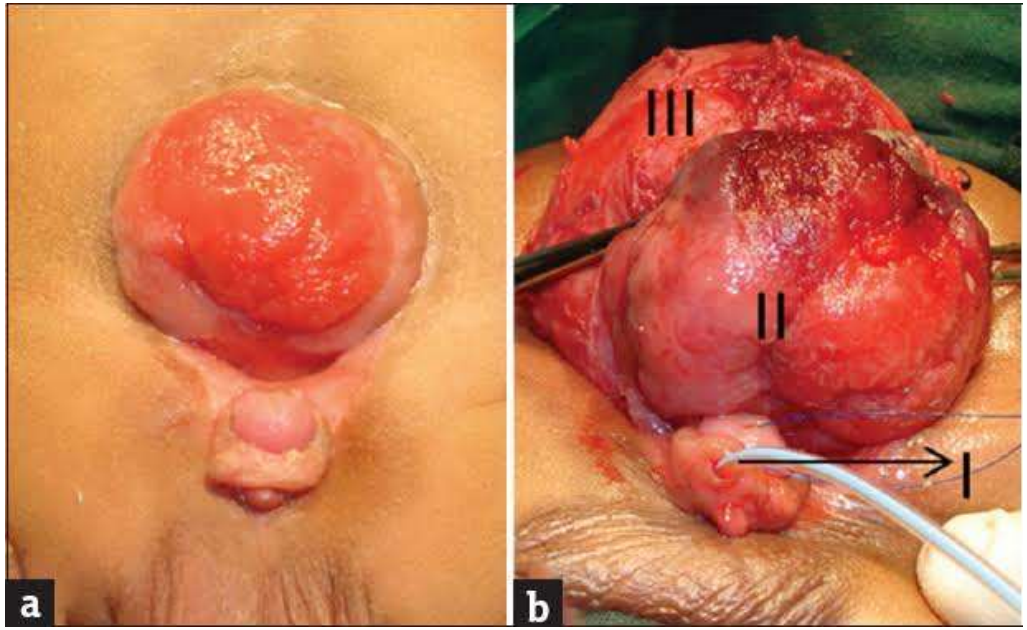
# Superior vesical fissure

- only the upper portion of the bladder is affected (no epispadias). Bladder extrusion is minimal, only over umbilicus, resembles vesicostomy

# 5.4 Duplex exstrophy

- **anteroposterior bladder duplication** with a patch of everted bladder mucosa on the anterior abdominal wall and a second bladder lying in the pelvis.
  - The ureters attach to the closed bladder, rendering the superficial mucosa dry
  - The mainstay of treatment has been resection of the ectopic mucosa and closure of the abdominal wall defect.
- **Side to side duplication** with two separately formed bladder halves in a left-right orientation with a midline septum between the bladders containing muscle.
  - Each bladder has its own ureter and an intact sphincter.

# Duplex exstrophy




- **Duplicate exstrophy in a boy with an intact lower urinary tract.**

# Cloacal exstrophy variants

- exposed hindgut plate and bladder plate, without typical genital involvement.

# 6 – Outcome

# Epispadias

- Continence
  - 75% of male incontinent, but 80% - 90% are continent postoperatively
  - paucity of literature on isolated female epispadias, but 87% to 100% continence after repair
- UCF and stricture
  - Ransley and colleagues (Kajbafzadeh et al, 1995)
    - UCF 4%,
    - urethral stricture rate 5.3%.
  - Mollard and coworkers (1998)
    - UCF <than 10%.
  - Surer and colleagues (2000)
    - UCF 19%
    - urethral stricture < 10%.
  - Baird and colleagues (2005)
    - UCF 13% of primary and 25% of secondary
    - urethral stricture in one patient

# Cloacal exstrophy

- Neurologic complications
  - May be wheelchair bound
- GI complications
  - Short gut syndrome
  - fecal incontinence
- Growth failure
- Psychosexual
  - Short phallus
  - gender conversion

# Bladder Exstrophy

- Given exstrophy is a complex birth defect to reconstruct, no matter which method is chosen, complications will occur, most of which require further surgical interventions.
- **Survival rates** in CE have risen to nearly 100%
- **Boston group** found that bladder capacity was equivalent but CPRE had better bladder stability and bladder compliance than MSRE
- **Johns Hopkins group** report 70% continence with MSRE with minimal complications
- CPRE
  - **blood transfusion** required in 75% male and 29% female newborns.
  - **Hypospadias** after CPRE have ranged from 50% -82%
  - **Bladder neck fistulas** have been observed in as many as 41%

# Potential complications

- **Failed primary closure**
  - Wound dehiscence
  - Bladder prolapse
  - Bladder outlet obstruction
  - Vesicocutaneous fistula
  - recurrent pubic diastasis (failed osteotomy)
- **Osteotomy complications**
  - transient nerve and muscle palsies
  - delayed ileal union
  - pin sites infection
  - Gait abnormalities, Pelvic and hip pain
- **Epispadias repair complications**
  - Persistent choree
  - UCF
  - Wound dehiscence
  - Glans/corporal ischemia (in Mitchel repair)
- **Failed BNR**
  - Retention, UTI
  - Incontinence
  - VUR
- **Phalloplasty/ Inflatable penile prosthesis**
  - partial necrosis, flap loss
  - infection and erosion
- **Intestinal complications**
  - SBO, ileus, Volvulus,
- **Diversion/augmentaion complications**

# UCF

- most common complication (5-40%)
- Site is penopubic angle dorsally in males
- Some say more common in CPRE, some show similar result with MSRE
- Initial conservative management and diversion (suprapubic cystostomy)
- Redo reconstruction (if required) at 6-12 mo.
- Cystoscopy to check distal obstruction & other abnormalities

# UCF

- Ucf at penopubic angle

# Ischemic penile injury

- rare
- leads to atrophy of the glans, corpora cavernosa, or urethra
- Mechanism unknown but may be
  - compression of the pudendal vessels
  - venous congestion
  - unintentional injury
  - abnormal blood supply and collaterals that prevent sufficient blood flow during the procedure
  - congenitally altered vascular supply
  - intraoperative use of epinephrine
  - aggressive dissection and detachment of the corpora and its neurovascular bundle from the ischiopubic ramus and Alcock's canal
  - pelvis compression on approximation of the pubic symphysis (more if no osteotomy), or postop edema.
- can occur even in experienced hands so just be cognizant during repair
- If noted postoperatively, two successful reports of the use of medicinal leeches on the dark glans.

# Ischemic penile injury

- loss of right glans and urethra

# Bladder & abdominal wall dehiscence

- uncommon
- May indicate significant post op infection or technical error
- Complex reoperation may be required depending on location and size

# Bladder & abdominal wall dehiscence

- Major bladder prolapse after CPRE

# Bladder outlet obstruction

- noted after catheter removed or when child develops chronic UTI
- may lead to bladder perforation (especially if augmented)
- May be due to
  - poor bladder contraction
  - urethral stricture
  - complete urethral obliteration (females), or posterior urethral (males)
- Early intervention with CIC and suppressive antibiotics (for VUR)

# UTI & VUR

- UTI Common, risk Increases in bladder augmentation
- VUR in nearly all following bladder closure
  - ureters pass deep into pelvis and enter the detrusor in a direct path with a very short tunnel

# Scars

- **Scar revision** surgeries may be required around adolescence
  - depend on the severity of scarring and the type, size and location of the scar
- **Monsplasty**
  - For significant scarring and/or depression of the tissues in the mons pubis area
  - removes scar tissue present, flattens the area of the mons pubis and joins hair-bearing skin for a more normal appearance.
- **Vaginal reconstruction**
  - Vaginal stenosis can occur with with changes in vaginal tissue.
  - May require vaginoplasty or more compex reconstructions
- **Penile reconstruction**
  - To revise surgical scars that remain from the original surgery and/or to straighten or lengthen the penis.

# Scars

- **Monsplasty** - Removes thinned midline skin and  Builds up interpubic area. if inadequate tissue to excise scar, expanders are placed

# Incontinence

- Multiple procedures are required for to achieve continence and reconstruction of anatomy
- the single most important predictor of long-term bladder growth and continence is successful primary bladder closure
  - Continence after primary closure ranges from 80 to 100%.
  - One failed closure dec to 60%, 2 failure dec to 20%
- After BNR, surgical success is defined as a dry interval of >2–3 hours and spontaneous voiding without catheterization
  - YDL-BNR 30%–80%
  - Only 20–56% of CPRE who remain incontinent achieved continence following additional BNR
- Reported **continence** of 67-87% female patients with good bladder capacity
- initially successful bladder closure and BNR may require further reconstruction in their second decade of life because of poorly compliant, low-capacity bladders that caused urinary incontinence
- Failed BNR might require CIC or Bladder augmentation with bladder neck closure

# Bladder malignancy

- Adenocarcinoma is ~ 400 x more common in CBE
- At risk bladder plate
  - bladders closed later in life
  - those augmented with colon.
- Long-term monitoring with cystoscopic evaluation is recommended

# Renal deterioration

- Due to
  - Recurrent pyelonephritis
  - Pressure on the kidneys during storage/emptying due to outlet resistance
- Renal function should be followed closely after closure

# Sexual function: female

- Females tend to fare better in all aspects of sexual health with most being satisfied with the appearance of their genitalia and all being sexually active
- **pelvic organ prolapse** in 20 - 38%
  - Most common complication
  - future urinary reconstruction should include prophylactic anterior fixation of the uterus
- **Dyspareunia** is reported
- **miscarriage and preterm labor**
  - b/c prolapse / lack of pelvic floor support
  - C/S delivery using paramedian incision is the safe approach.
- hydronephrosis and bacteriuria during preganacy  may necessitate antimicrobial prophylaxis

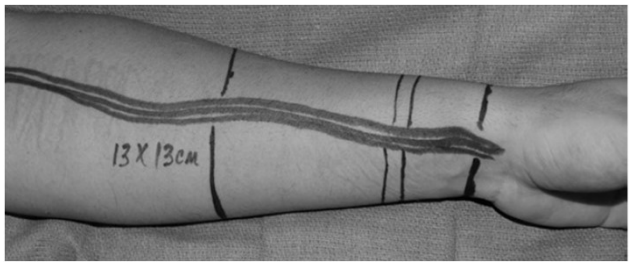
# Sexual function: male

- Despite severe epispadias associated with exstrophy, most patients completing staged reconstruction can expect to enjoy sexual relations
- Epididymitis 19–33%
  - should prompt evaluation for high-pressure voiding due to a urethral stricture
  - may require epididymectomy, vasectomy, or even orchiectomy
- Impaired Fertility
  - Retrograde ejaculation (underlying bladder neck abnormality) may impair fertility
  - Ejaculation possible but seminal emission may be slow and continue hours after orgasm.
  - Sperm quality and quantity is often diminished
  - Chordee and small penile size can create difficulty with intercourse
  - Reconstruction with radial forearm flap might help with size.
  - Some elect to have inflatable penile prosthesis

# Sexual function: male

- Inadequate phallus for intercourse - Neophallus creation

# Sexual function: male




- **Radial forearm flap**

# Psychosocial distress

- anxiety and suicidal ideation is greater than the general population
- related to phallic appearance and urinary incontinence
- regular assessment of social development as early as 12-18mo, including psychiatric evaluation and parental education

# References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