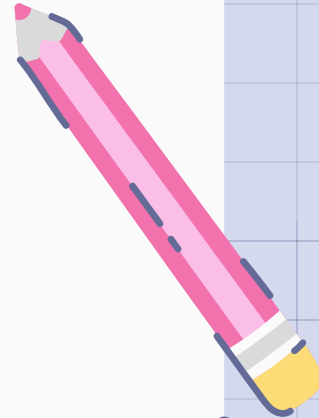




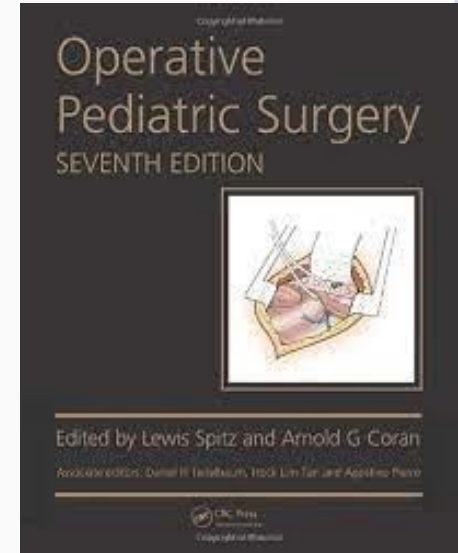
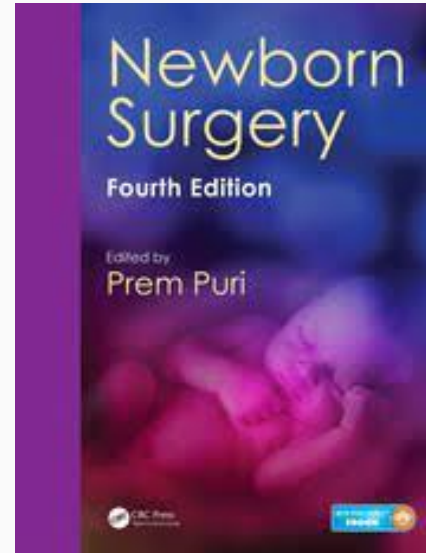
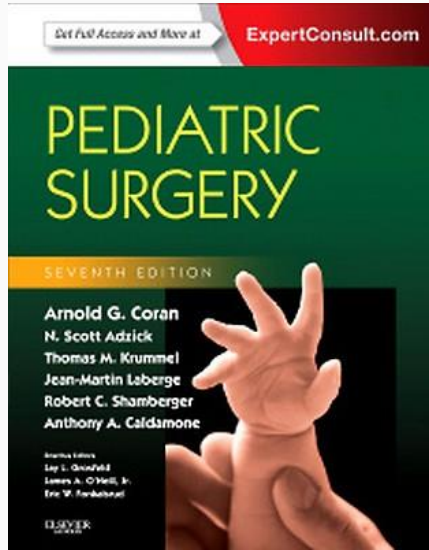
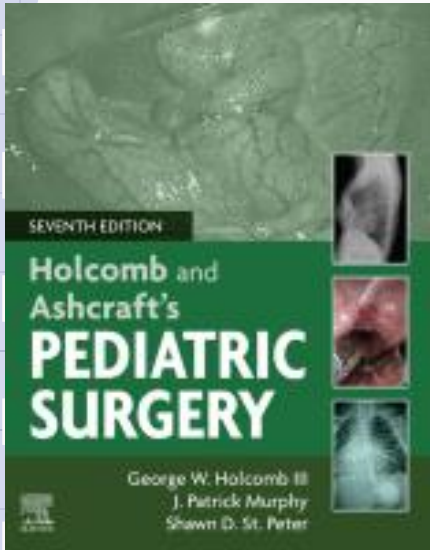
# Bladder and Cloacal

## Exstrophy

R3 Kanyarat Olarachin



# References



# Table of contents

01 Bladder exstrophy

02 Cloacal exstrophy



## Epispadias-Exstrophy Complex (EEC)

- Epispadias
- Classic Bladder Exstrophy
- Cloacal Exstrophy
- Exstrophy Variants

01

# Bladder Exstrophy



Head



Bladder



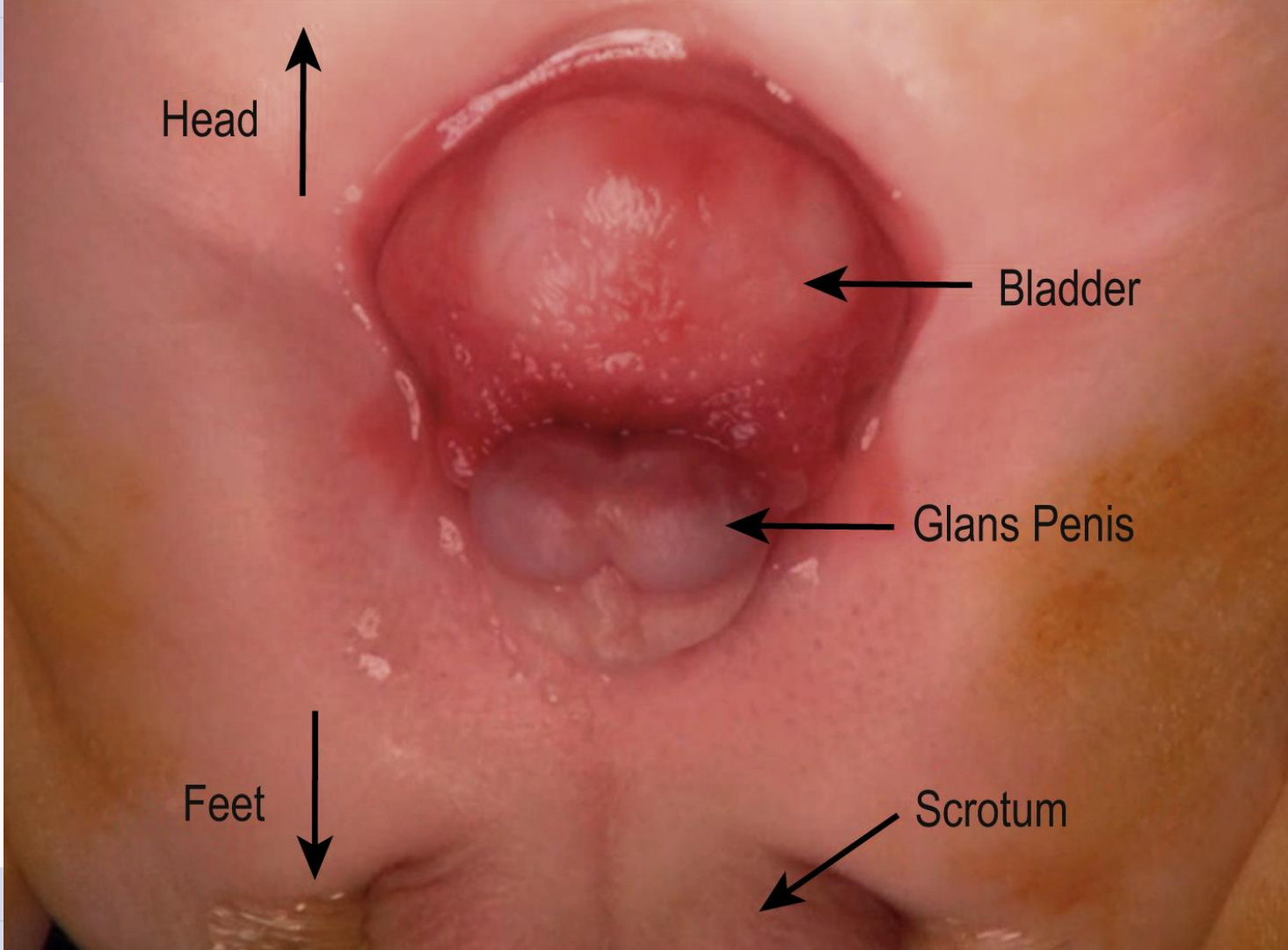
Glans Penis



Feet



Scrotum



# Bladder exstrophy

- Incidence 1: 50,000 live birth
- Male : Female 1:1 – 6:1
- Rarely associated with other organ system malformation.

## Features

- Anteriorly located anus.
- Female genital anatomy
  - More vertically oriented vaginal opening following repair
  - Wider and shorter vagina than normal.
- Male genital anatomy
  - Foreshortened the anterior component of the penis

# Statistics

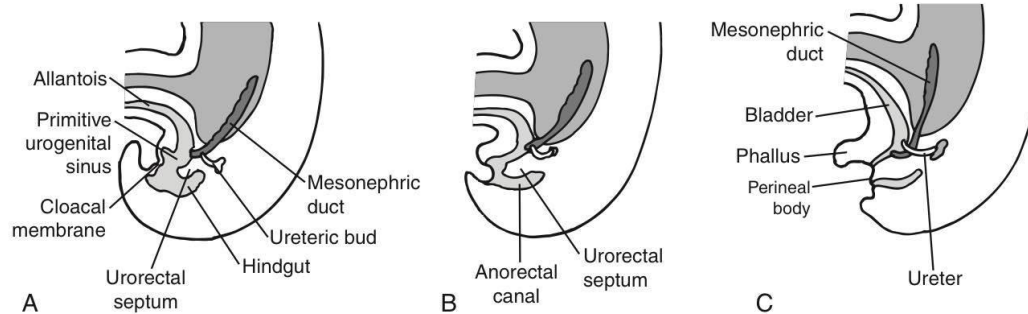
Data from small familial series

- Siblings of children with BE have an incidence of BE that ranges from 0.3–2.3%, much higher than normal
- Children with parent with an exstrophy have incidence of BE 1.4%, or 400-fold higher than the general population.
- No clear pattern of inheritance
- No link between exstrophy and parental age, maternal reproductive history, or periconceptional maternal exposure to alcohol, drugs, chemical noxae, radiation, or infections but **maternal exposure to smoking** has significantly more common in patients with EEC



# Pathogenesis

- Prescientific era : trauma to unborn child
- Theory : Mesenchymal cells in intermediate layer of mesodermal membrane fold do not migrate properly during 4<sup>th</sup> week, resulting in rupture of the cloacal membrane
  - Ischemic injury
  - Genetic switch



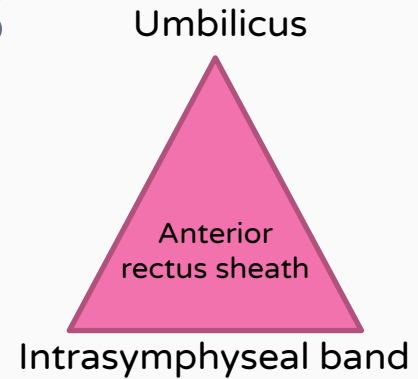
**FIGURE 120-1** Division of the cloaca in the urogenital sinus and rectum. **A**, At the end of the fifth week. **B**, At 7 weeks. **C**, At 8 weeks. (Modified from Sadler TW: Langman's Medical Embryology, 8th ed. Philadelphia, Lippincott Williams and Wilkins, 2000, p 316.)

# Genetics

- Multifactorial
- ISL-1 on chromosome 5q11.1
- Whole exome sequencing and genome wide association studies are in progress

# Anatomic considerations

- Musculoskeletal defects : widening pubic symphysis
- Abdominal wall defects : triangular fascial defect, IIH
- Anorectal defects : anterior anus
- Male genital defects : shorter and wider anterior corporal length
- Female genital defects : bifid clitoris, shorter vagina but normal caliber
- Urinary defects :
  - Bladder : ectopic bowel mucosa or isolated bowel loop may be present, hamatomatous polyp may be present
  - Upper tract : normal
  - Ureteral openings : misplaced



# Diagnosis

## Prenatal ultrasound

- Absence urinary bladder in fetal pelvis
- Semisolid mass protruding from abdominal wall
- A lower abdominal protrusion
- An anteriorly displaced scrotum with a small phallus in male fetuses
- Normal kidney with a low-set of umbilical cord
- An abnormal iliac crest widening (about  $110^\circ$ , normally  $90^\circ$ )

Perform fetal MRI if any abnormality is seen to characterize the variants and distinguish from another abdominal wall defect

# Prenatal management

- Prenatal counseling
- Transfer patient to pediatric center with pediatric urologist

# Principle of reconstruction

- Close the bladder and urethra
- Reconstruct the genitalia
- Function needed : continence, voiding, and sex.
  - Continence : competent bladder neck
  - Voiding : bladder neck that can relax to allow a sustained detrusor contraction, resulting in normal voiding with complete emptying

## Pros :

- Minimizes the risk of upper urinary tract deterioration
- Prevents UTI and VUR
- Decreases the risk of urinary calculi

# Natural History and Early Attempts at Treatment

- Untreated still survive but ...
  - Skin breakdown secondary to total urinary incontinence
  - Tumor development within the chronically exposed bladder plate
  - Significant psychosocial morbidity
- Very early efforts in BE management : partial reconstruction of the abdominal wall to allow the application of a urinary receptacle to collect urine.
- Early attempts at closing the bladder were fraught with complications  
-> urinary diversion through the ureterosigmoidostomies (USO) (poor results)
- Current management was developed after knowledge of urinary tract and bladder physiology (the effect of BE repair on urine storage and emptying, and the concept of clean intermittent catheterization)
- Still in imperfect state.

# Operative approaches

- Urinary diversion
- Anatomic reconstruction
  - Single stage repair
  - Multistage repair

Surgeon preference and experience

Patient anatomy

History of previous operations

Available of tertiary care

Access of medical care and resources



# Urinary diversion

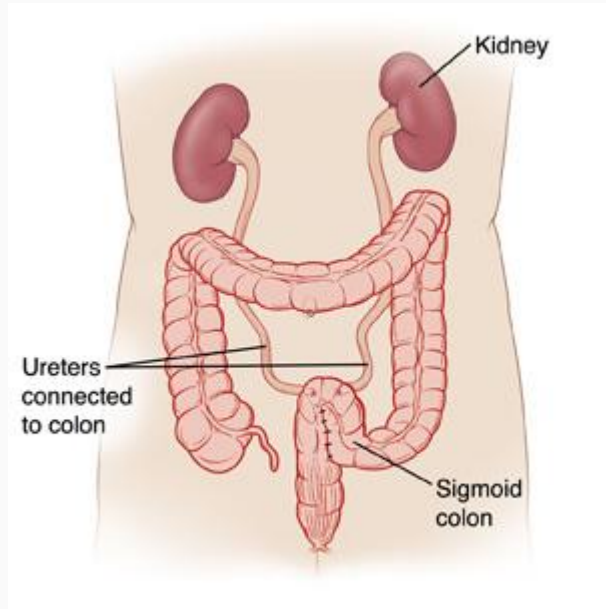
- Primary diversion is not commonly performed
- 
- Continent urinary diversion
  - More consistent degree of dryness
  - Fewer required operations and fewer early complications compared with anatomic reconstruction
  - Require catheterization
- Incontinence urinary diversion
  - Avoids the complications associated with continent reconstruction such as a failed continence mechanism resulting in persistent incontinence, urinary retention, stomal complications, and dependence on CIC to empty the bladder.

# Surgical options for Urinary Diversion

External Diversion (Continent Urinary Reservoir)	Internal Diversion (Rectal Sphincter-Based Continence)	Incontinent Diversions
Indiana pouch (cecal reservoir with ileal catheterizable channel)	Sigma pouch Ghoneim reservoir	Ileal conduit Colon conduit
Mainz pouch	Gersuny	Ileocecal conduit
Penn pouch (ileocecal reservoir with appendiceal catheterizable channel)	Heitz–Boyer–Hovelacque Rectal bladder with proximal colostomy	
Kock pouch	Ureterosigmoidostomy	
	Ileocecal ureterosigmoidostomy	

# Ureterosigmoidostomy

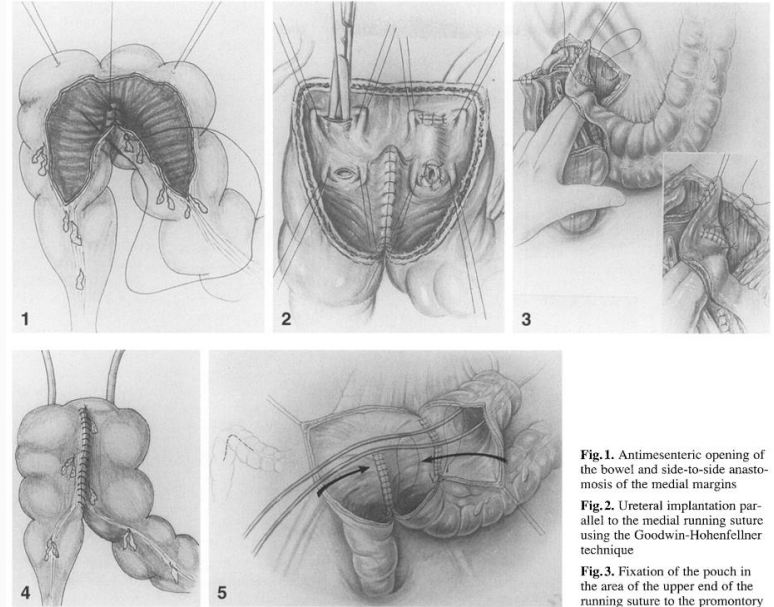
- The earliest continence urinary diversion
- Caution : Contraction from sigmoid colon will reflux the infected urine or feces back into the urinary system resulting in severe infections.
- Long-term complications
  - Hyperchloremic metabolic acidosis
  - Chronic pyelonephritis
  - Bladder calculi
  - Increased risk of adenocarcinoma developing at the anastomosis of the ureter(s) to the colon 250-300X



<https://api.kramesstaywell.com/Content/6066ca30-310a-4170-b001-a4ab013d61fd/ucr-images-v1/Images/front-view-of-torso-showing-kidneys-connected-to-sigmoid-colon-by-ureters-268415>

# Sigma pouch

- Mainz II pouch
- Detubularized sigmoid pouch
- Reduces reservoir and ureteral pressures, and improves night-time continence



**Fig. 1.** Antimesenteric opening of the bowel and side-to-side anastomosis of the medial margins

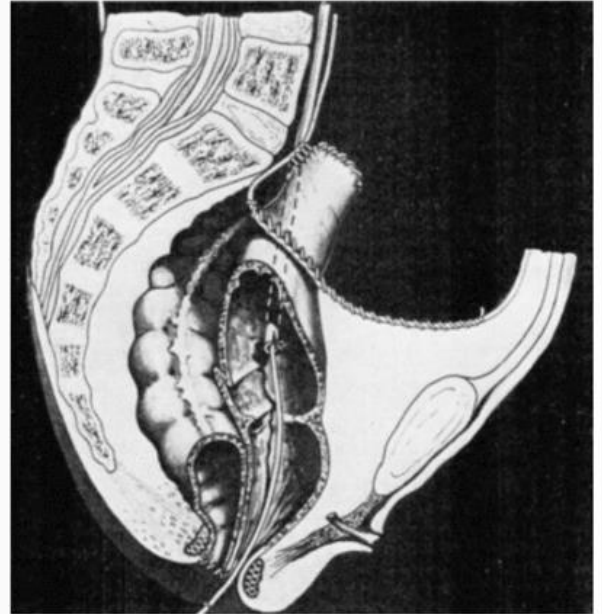
**Fig. 2.** Ureteral implantation parallel to the medial running suture using the Goodwin-Hohenfellner technique

**Fig. 3.** Fixation of the pouch in the area of the upper end of the running suture to the promontory

Fisch M, Wammack R, Hohenfellner R. The sigma rectum pouch (Mainz pouch II). *World J Urol.* 1996;14(2):68-72. doi: 10.1007/BF00182560. PMID: 8731120.

# Heitz–Boyer–Hovelacque procedure

- Principle : isolation of a rectal segment for ureteral implantation followed by posterior sagittal pull-through of the sigmoid colon through the anal sphincter
- Used both for primary treatment and salvage repairs
- Continence rate 52-97%



TACCIUOLI, M., LAURENTI, C. and RACHELI, T. (1977), Sixteen Years' Experience with the Heitz Boyer-Hovelacque Procedure for Exstrophy of the Bladder. *British Journal of Urology*, 49: 385-390.

# Complications of Rectal Sphincter-Based Continence

- Urine contact with rectal mucosa
  - Metabolic electrolyte imbalances
  - Treatment
    - Oral bicarbonate replacement
    - Frequent bladder emptying
  - Malignancy risk at ureterointestinal anastomosis
    - Prevent admixture of feces and urine may decrease the incidence of cancer

# History of Anatomic Reconstruction

- The first attempt was unsuccessful
- 1881 – Pubic symphysis approximation
- Early 20<sup>th</sup> century - Convert to urinary diversion
- Late 20<sup>th</sup> century - single stage reconstruction (continence rate 10-30%, complication : renal damage 90% due to BOO)
- 1970 - Staged reconstruction -> **Modern staged repair of exstrophy (MSRE)**
- Knowledge of CIC – **Complete primary repair of exstrophy (CPRE)**

# MSRE vs CPRE

MSRE	CPRE
need later bladder neck reconstruction (BNR)	More aggressive mobilization of the vagina and urethral plate posteriorly into the pelvic diaphragm in order to gain bladder outlet resistance that allows for normal voiding with continence
Staged procedure (closes and repositions of the bladder and posterior urethra, epispadias repair, bladder neck reconstruction)	Single stage procedure



# Operative considerations

- Immediate newborn period, prior to 72 hours of life.
  - Allows for anatomic closure without the use of osteotomies
  - Decreases bladder exposure that can lead to histologic changes
  - Helps to “mature” the bladder with bladder cycling
- Delayed primary closure
  - Safer
    - The patient’s condition
    - More well-prepared operation (personnel, team)
  - Allows bonding (need to separate for traction after operation 4-6 wk)
  - Allow times to stimulate the penis with testosterone
  - Caution
    - Need good protection of the bladder plate
    - Increased cost of care 50%

# Preoperative Care

## After delivery

- Ligate umbilical cord with suture, **not clamp**
- Hydrated gel dressing or plastic wrap to protect the exposed bladder

## Investigation

- USG KUB
- Film pelvis AP (to evaluate pelvic diastasis)
- USG spine if sacral dimpling or other signs of spina bifida is observed

## Preoperative antibiotics

- Pre-op ATB is not required , but perioperative and post-operative ATB are used to decrease risk of infection following reconstruction

# Perioperative and Postoperative Adjuncts to Successful Bladder Exstrophy Closure

Intraoperative	Postoperative
Avoid abdominal distention <ul style="list-style-type: none"><li>• NG tube for continuous decompression</li><li>• Avoid nitrous oxide</li></ul>	Tunneled epidural for pain management
Epidural to avoid narcotics	Maximal urinary drainage
Careful fluid management	Immobilization with traction or spica cast
Antibiotics	

# Adjuncts Aspect of the Repair

## Indirect inguinal hernia

- Commonly associated
- From one study, 9 patients need hernia repair from 25 patients that hernia repair were not performed during BE closure
- Inguinal exploration and repair of an inguinal hernia at the time of exstrophy closure

# Adjuncts Aspect of the Repair

## Vesicourethral Reflux

- High incidence (up to 75%)
- Perform urethral reimplantation at the time of initial closure
  - Providing a more gradual course through to the neohiatus, which provides more distance from the bladder neck to the ureteral orifices

# Adjuncts Aspect of the Repair

## Osteotomies

- Pelvic bone of BE patient : external rotation of the posterior pelvis, shortening and external rotation of the pubic rami, and a wide pubic diastasis
- Advantages
  - Optimizing pubic symphysis apposition, decrease tension of the fascial repair
  - Optimizing placement of the bladder, bladder neck, and urethra in the pelvis
  - Improving the re-approximation of the corporal and clitoral bodies
  - decreasing the chance for later uterine prolapse
  - Improves rates of continence in children closed outside of the immediate newborn period

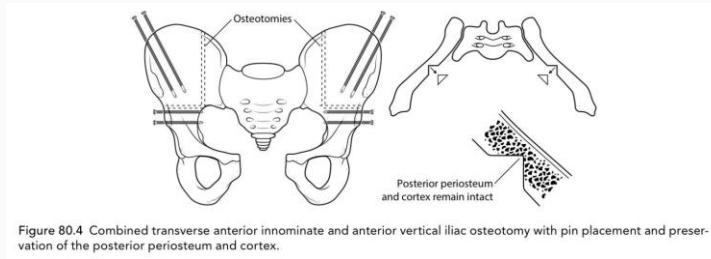
# Adjuncts Aspect of the Repair

When to perform osteotomies

- At the time of BE closure
- Any BE child age > 72 hr (< 72 also valuable)
- NB with exceptionally wide diastasis (> 4 cm)
- Reoperation

# Osteotomies

- Posterior approach : Prone position (Perform after bladder closure)
- Anterior approach : Supine position (Avoid repositioning the patient)
  - Anterior diagonal technique
  - **Anterior diagonal and vertical iliac osteotomies**
    - Excellent initial and long-term results compared with anterior iliac osteotomies alone in some series.



- Diagonal mid-iliac osteotomy (less common)
  - Same incision as the exstrophy closure,
- Division of the superior pubic ramus (less common)
  - Newborn period.



# Adjuncts Aspect of the Repair

## Immobilization

- To decrease stress on the closure
- Types of immobilization
  - Modified Bryant's traction
    - Immobilizes the baby in a bed and the hospital for 4–6 weeks
    - Can cause injury to the skin
  - External fixation
    - Pin tract infection
    - Need wound dressing OD
    - Recommend for older children
  - Spica cast
    - Provides mobilization and early discharge
    - Lower cost
    - Hinge spica case

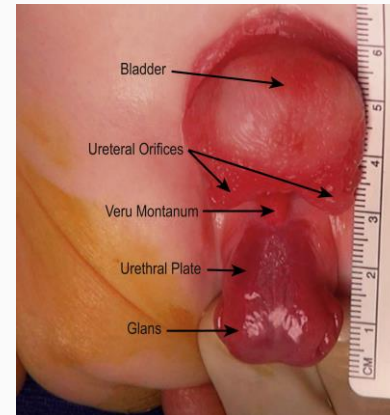
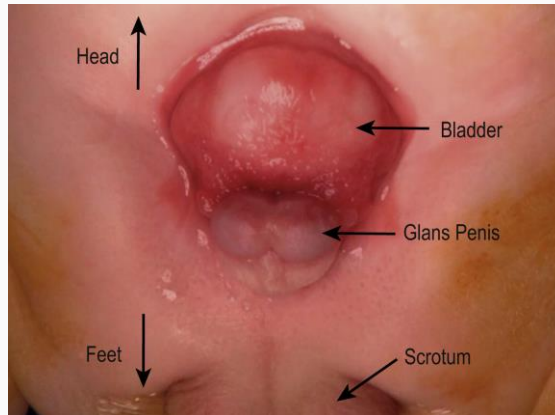


# Complete Primary Repair of Exstrophy (CPRE)

- Combination of bladder closure, anatomic bladder neck narrowing, urethral elongation, and epispadias repair in a single operation in order to provide an environment for bladder cycling.
- At times, anatomy permitting, bilateral ureteral reimplantation is performed at CPRE in order to achieve the goals of urinary continence and preservation of renal function.
- Limit tissue injury

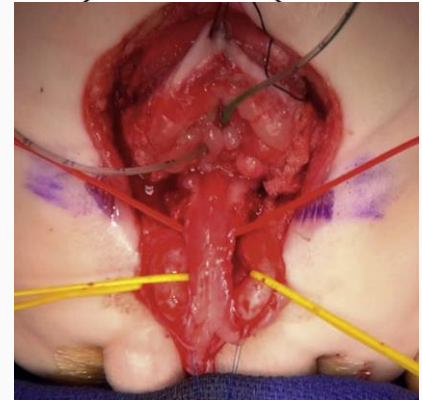
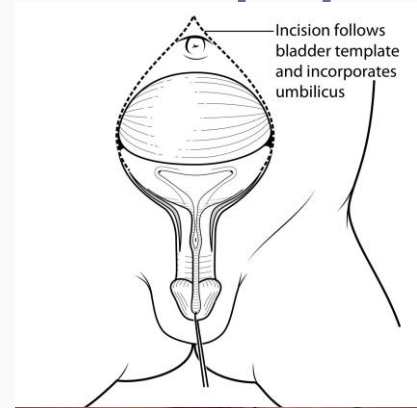
# CPRE Technique in the Boy with Bladder Exstrophy

- Total body preparation
- Bilateral iliac osteotomies are completed prior to sterile prep if a posterior approach is used, or after prepping if an anterior approach is chosen.
- The anatomy should be evaluated at the outset of the procedure
- Tegaderm (3M) is placed over the anus.
- Measure bladder and urethral plate, identify each landmark structures



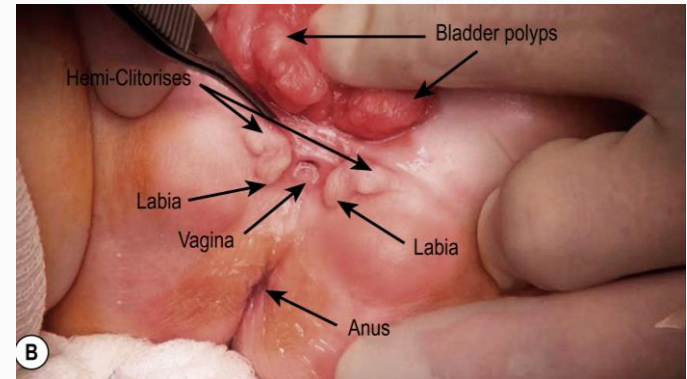
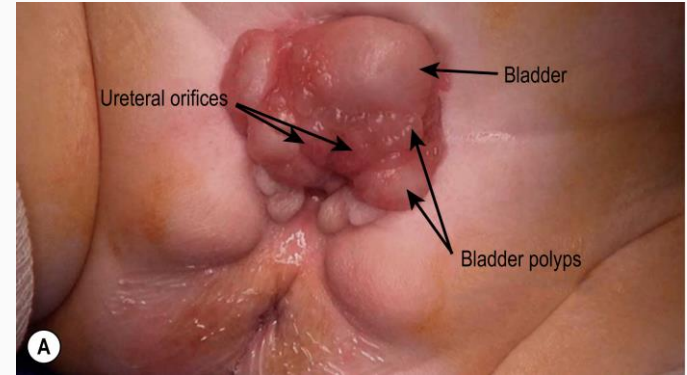
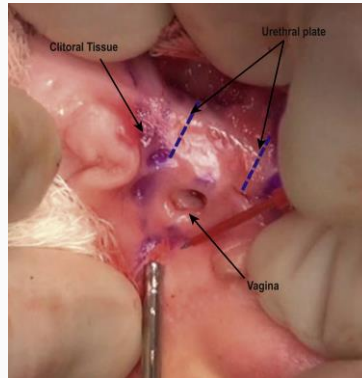
# CPRE Technique in the Boy with Bladder Exstrophy

- Insert small-caliber feeding tube as a ureteral stent and secured with absorbable suture.
- Assess bladder mucosa and excise polyp if seen
- Create inverted V incision using fine needle electrocautery
- Ligate the obliterated umbilical arteries and vein
- Separate peritoneum from bladder
- Divide intersymphyseal band
- Urethral plate dissection starting from ventral side and then dorsal side
- Continue deep dissection to the level of prostatic urethra and bladder neck
- Bladder neck reconstruction
- Urethroplasty, bladder and bladder neck approximation
- Pubic bone approximation with PDS 1-0 or 0-0
- Place suprapubic cystostomy
- Penile reconstruction
- Abdominal wall closure
- Umblicoplasty



# CPRE Technique in the Girls with Bladder Exstrophy

- Same preparation as male patient
- Create inverted V skin incision
- Create “Y-V” incision in order to advance the vaginal wall more posterior into the perineum
- Urethroplasty
- Place perineal sutures
- Pubic bone approximation with PDS 0-0 or 1-0 \* 2 ties
- Tie perineal sutures
- Abdominal wall closure
- Umbilicoplasty
- Drainage
- Immobilization





## Other Primary Reconstructive Techniques

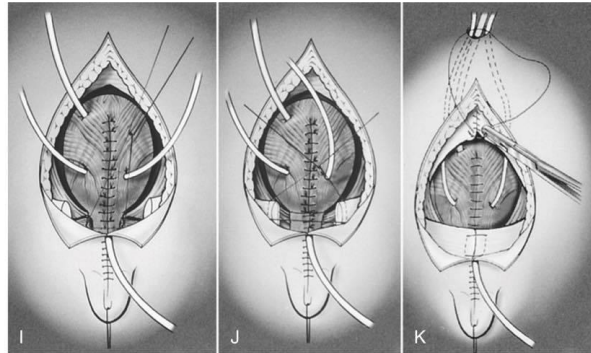
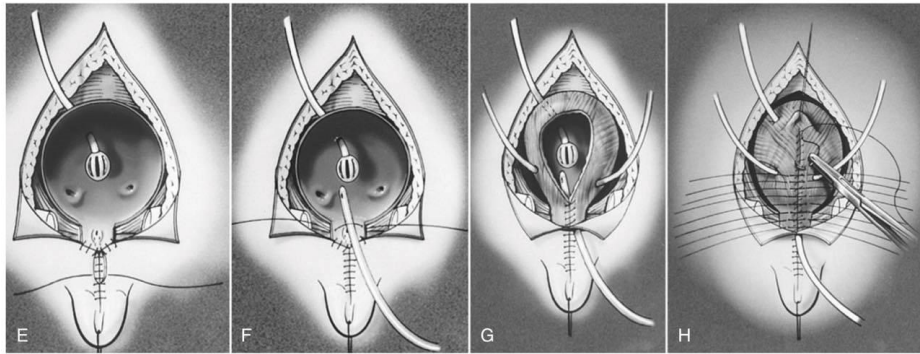
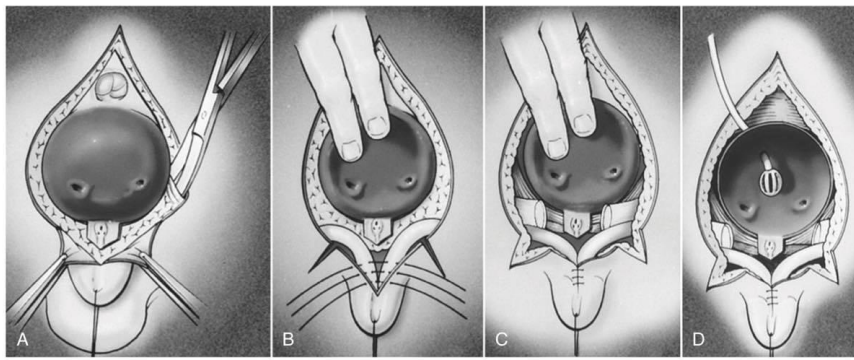
- Rösch and colleagues : bladder closure, epispadias repair, and bilateral ureteral reimplantation without iliac osteotomy
- Baka-Jakubiak : bladder neck, urethral and penile reconstruction
- **Kelly operation** (radical soft tissue mobilization) : detaching the insertion of the corpora cavernosa from the ischiopubic rami, but damage to the pudendal neurovascular supply could be occurred (no osteotomies)

# Modern Staged Repair of Bladder Exstrophy (MSRE)

Stage I: Bladder Closure (Includes repair of the proximal urethra in a male)

- Closure of the bladder, posterior urethra, and the abdominal wall defect results in the creation of complete epispadias (in the boy)
- Perform within the first 2 months of life.
- May not be advisable if there is a small-sized and/or stiff bladder “plate,” or if the infant is premature.





# Modern Staged Repair of Bladder Exstrophy (MSRE)

## Stage II: Epispadias Repair

- Create a **straight and functional penis** with a glanular meatus in the boy, and an **acceptable cosmetic appearance**.
- Perform at approximately **6 months to 1 year of age**
- Exogenous **testosterone** supplementation can be given prior to this surgery in the boy based on surgeon preference and penile size.
- Techniques
  - Complete Penile Disassembly approach
  - Modified Cantwell–Ransley technique.  
(A controversial component of this operation is whether to rotate the corpora inward or outward following pubic bone closure.)

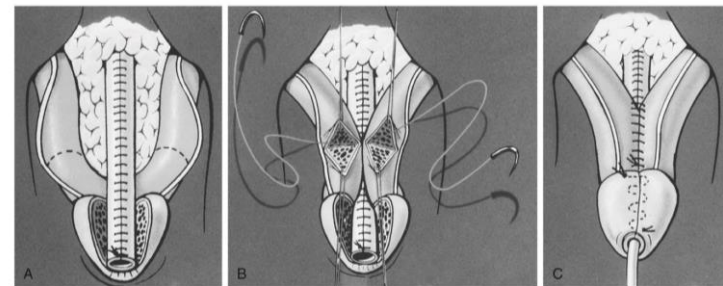
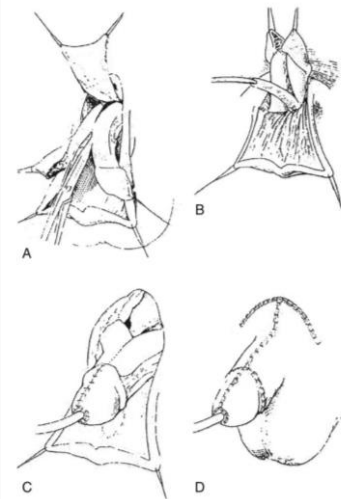
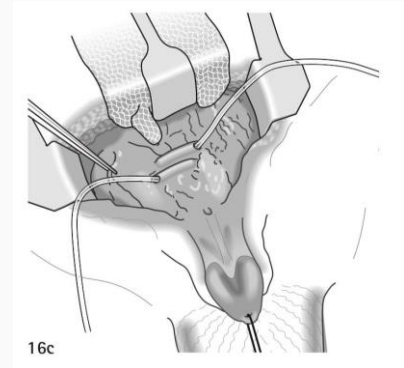


FIGURE 120-12 Steps in the Cantwell-Ransley epispadias repair. **A**, The urethral plate is dissected from the corpora and is tubularized, taking care to preserve the lateral blood supply of the urethra and the neurovascular bundles. **B**, Corporotomies are created at the midphallus, and the urethra is transposed to the ventral surface. **C**, The corpora cavernosa are rotated medially and reapproximated at the corporotomy sites, pulling the corporal bodies inward and providing coverage of the neourethra. This procedure permits further urethral lengthening, approximation of the corpora with preservation of the blood supply, and full coverage of the urethra.

# Modern Staged Repair of Bladder Exstrophy (MSRE)

## Stage III: Bladder Neck Reconstruction and Bilateral Ureteral Reimplantation

- At approximately **4–5 years of age**
- To provide **urinary continence**
- Performed when there is documented **adequate bladder capacity** and motivation of the patient to participate in a **postoperative voiding program**.
  - Techniques
    - Mitchell
    - Young–Dees–Leadbetter (YDL) techniques (continence 30-80%)
- Bilateral ureteral reimplantation
  - To creating space near the caudal extent of the bladder (bladder trigone) in **preparation for the BNR procedure**.
  - Techniques
    - Cross-trigonal transureteral advancement (Cohen procedure)
    - Cephalotrigonal technique



# Outcomes

- Difficult to interpret due to no consensus definition of continence and standard algorithm of treatment
- Most common use definition : Maintenance of dry periods of 3 or more hours during the day without stress incontinence
- Vary rate of continence (37 – 90%)
- A multi-institutional BE consortium (MIBEC) was recently formed to facilitate refinement of the CPRE technique

# Early complication

- Ischemic penile injury (during dissection or venous congestion)
- Urethrocutaneous fistula (penopubic angle dorsally) 5-40%
  - Management :
    - Initial conservative treatment
    - suprapubic cystostomy tube (will promote spontaneous closure)
    - Closure fistula next 6-12 months if the fistula is not closed
- Wound dehiscence, bladder prolapse
  - Post-operative infection, technical error
- Bladder outlet obstruction
  - Poor bladder contraction
  - Stricture
  - Complete urethral obliteration
- UTI
  - VUR

# Long-Term Outcomes

## Male

- Epididymitis 19-33% (evaluate urethral stricture)
- May require epididymectomy, vasectomy, orchiectomy in some rare cases

## Female

- Pelvic organ prolapse 20-30%
- Malignancy risk of the bladder plate
- Chronic renal disease resulting from recurrent pyelonephritis (which cause renal scarring), increase bladder outlet resistance
- Gait anomalies later in life after osteotomy in some patients

# Quality of Life

- Highly variable
- Depend on other chronic medical conditions
- Male : social relationship problem due to phallic appearance and urinary incontinence
- Recommend regular assessment

# Sexual function

## Male

- Parental concern
- Normal erectile function and sensation in most BE males
- Chordee and small penile size maybe problems for sexual intercourse
- Diminished sperm quality and quantity due to partial obstruction after exstrophy repair

## Female

- Intact sexual function (dyspareunia in some patients)
- Unimpaired fertility
- Uterine prolapse is common due to lack of pelvic floor support
- High risk pregnancy
  - Hydronephrosis, bacteriuria (need ATB prophylaxis)
  - Higher risk of spontaneous abortion (22%)
  - Risk of preterm labor

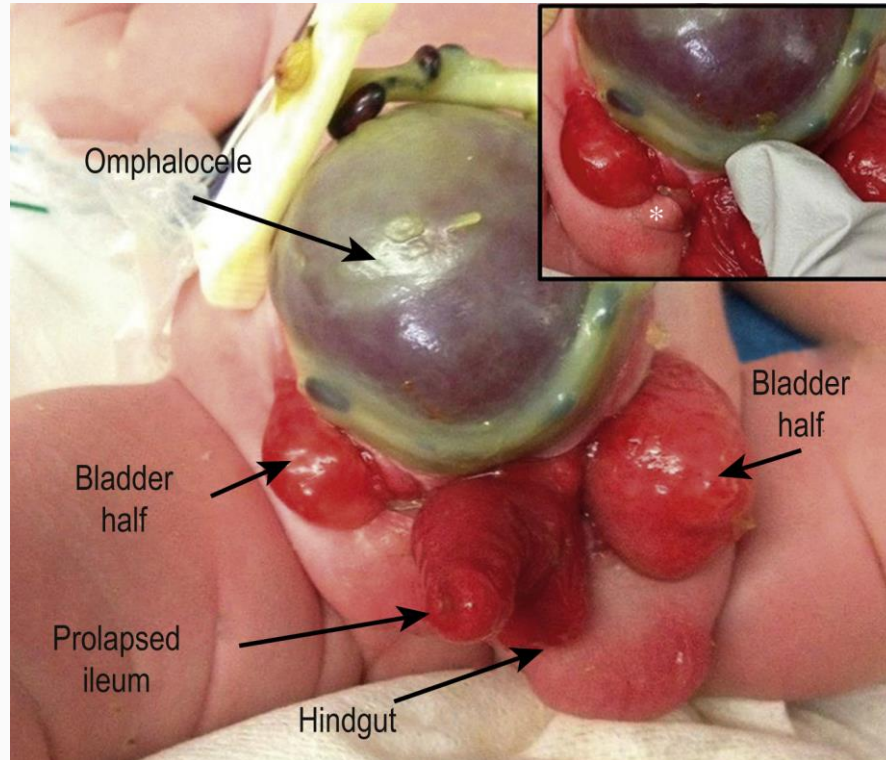


02

# Cloacal Exstrophy



# Cloacal exstrophy



# Cloacal exstrophy

- Incidence 1 : 300,000 (extremely rare)
- Most severe form of EEC
- Male = Female
- Often sporadic
- OEIS complex (omphalocele-exstrophy-imperforate anus-spinal defect)
- C677T have a strong association
- Embryology and pathogenesis: failure of mesodermal tissue migration to the lateral fold

# Associated anomalies

- Renal anomalies (48%) (more common than BE)
  - Single kidney
  - Abnormal kidney locations (pelvic kidney, crossed fused renal ectopia, horseshoe kidney)
  - Renal agenesis
  - UPJO
- Genital anomalies
  - Male
    - Separated two hemiphalluses
    - Undescended testes
  - Female
    - Uterine didelphys and fusion anomalies of the Mullerian structures (85%)
    - Vaginal agenesis (33%)

# Associated anomalies

## Gastrointestinal anomalies

- Ileal intussusception
- Exposed appendices
- Imperforate anus
- Midgut foreshortening
- Bowel duplication
- Malrotation
- Intestinal atresia
- Meckel diverticulum

## Skeletal anomalies

- Congenital hip dislocation
- Talipes equinovarus
- Hip dysplasia (16%)
- Varieties of limb deficiencies

# Associated anomalies

## Fascial anomalies

- Omphalocele
- Rupture omphalocele requires immediate attention

## CNS anomalies

- Spinal dysraphism 85-90%
- Intracranial anomalies 33%  
(Chiari malformation, hydrocephalus, craniosynostosis)

# Perioperative management

- Need experienced team
- ASAP after birth, cover the bladder and hindgut with plastic wrap
- Ligate the umbilical cord with a suture (Preventing umbilical clamp to injure the bladder or the hindgut)
- Optimal hydration and nutrition

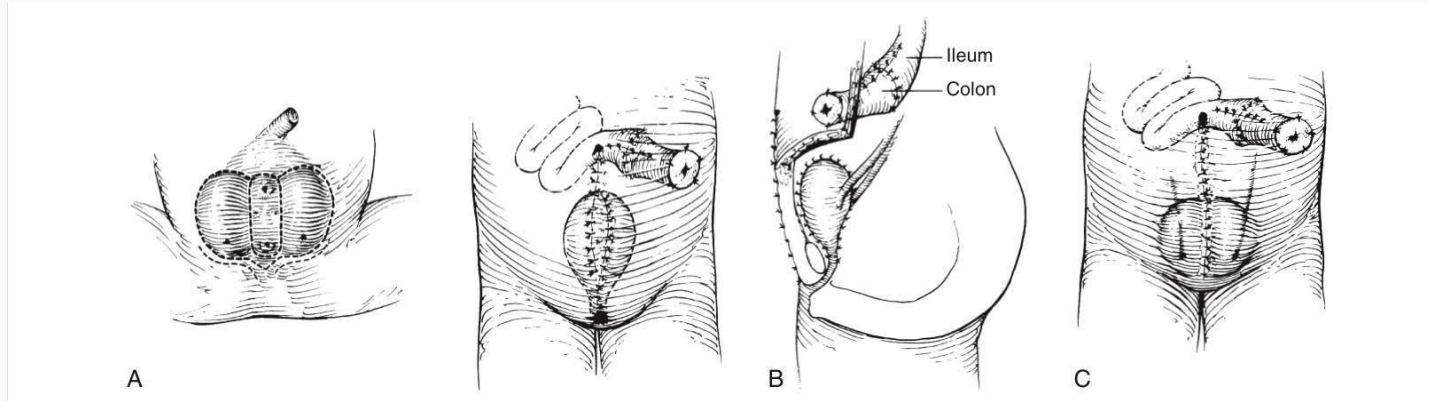
## Preoperative studies

- Ultrasound (upper urinary tract, internal genital structures, spinal cord)
- MRI if the anatomy remain unclear
- Abdominal films
- Karyotyping

# Cloacal exstrophy closure techniques

## General plan

- Closure of omphalocele with re-approximation of the posterior bladder halves, and tubularization of the cecum with incorporation of the hindgut into the gastrointestinal tract via an end colostomy (BE like anatomy)
- Repair of the exstrophic bladder and genitalia along with approximation of the pubic diasthesis





# Cloacal exstrophy closure techniques

- Initial reconstruction can be performed within the first 72 hrs of life if medically stable
  - 90% have spina bifida that need urgent closure
  - 90% have omphalocele that may need immediate attention to avoid rupture
- There are hindgut duplication in some cases. Must be preserved unless absolutely unusable
- Appendix must be preserved for possible use of urinary reconstruction
- Candidate for anal pull-through procedure : Children with anocutaneous fistula and preserved innervation

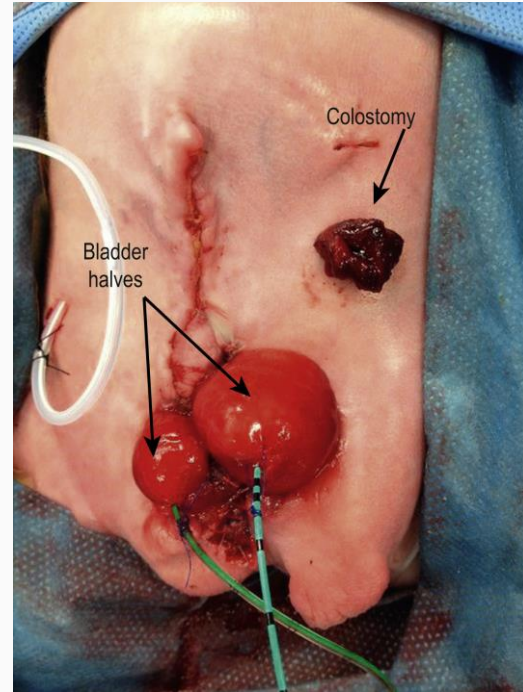
# Cloacal exstrophy closure techniques

- Initial reconstruction can be performed within the first 72 hrs of life if medically stable
  - 90% have spina bifida that need urgent closure
  - 90% have omphalocele that may need immediate attention to avoid rupture
- There are hindgut duplication in some cases. Must be preserved unless absolutely unusable
- Appendix must be preserved for possible use of urinary reconstruction
- Candidate for anal pull-through procedure : Children with anocutaneous fistula and preserved innervation

# Cloacal exstrophy closure techniques

## Two-stage CE closure

- Insert urethral catheter in the ureteral orifices and secure with chromic 5-0
- Repair omphalocele
- Ligate umbilical vessels
- Separate bladder plate from the adjacent skin
- Separate cecal plate and tubularized
- Reapproximate the bladder halves in midline
- Repair inguinal hernias if found



# Cloacal exstrophy closure techniques

## One-stage CE closure

- Closure the omphalocele
  - Cecal plate tubularization
  - Hindgut terminal colostomy
  - Bladder and urethral closure
  - Reconstruction of the external genitalia
  - Osteotomies
- 
- Perform only under optimal anatomic conditions
  - Caution : Abdominal compartment syndrome
  - May require secondary urologic procedure to achieve continence  
(BNR, bladder neck closure, bladder augmentation, conduit)

# Postoperative management

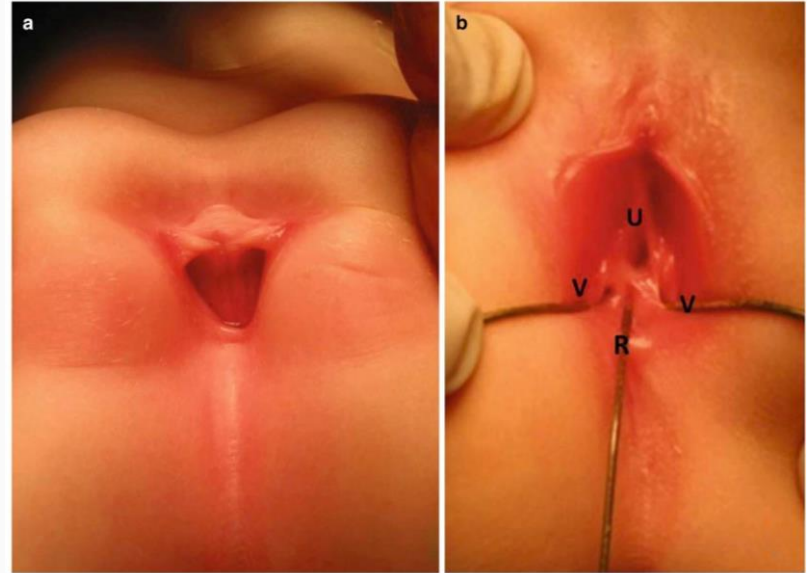
- Close monitoring
- Immobilization 4-6 wk
- Avoid abdominal distention
- Adequate pain control
- Prophylactic antibiotics

# Gastrointestinal Reconstruction

- Short bowel syndrome usually present even in those with normal bowel length
- Hindgut should be constructed and placed in continuity with the intestine
  - Promote nutritional and water absorption
  - Promote growth of hindgut tissue for later reconstruction of urinary tract or vagina if needed

# Covered cloacal exstrophy

- Cloacal exstrophy but intact abdominal skin
- Looks like cloacal malformation with very large single orifice
- Low umbilical cord implantation
- Two separate pubic prominence
- Need high index of suspicion for diagnosis
- Functional prognosis similar to CE



# Gender of rearing

- According to genetic karyotype



# Sexual function and quality of life

- Male : phallic inadequacy
  - Total phallic reconstruction
- Female
  - Vaginal reconstruction / replacement

# Thank you for your attention

Do you have any questions?

Download handout at

<http://www.pedsurgery.md.chula.ac.th/th-activities.html>

**CREDITS:** This presentation template was created by [Slidesgo](#), and includes icons by [Flaticon](#), and infographics & images by [Freepik](#)

Please keep this slide for attribution

