
DPU
DR. D. Y. PATIL VIDYAPEETH, PUNE
Dr. D. Y. PATIL MEDICAL COLLEGE
HOSPITAL & RESEARCH CENTRE

# RARE ADULT BLADDER EXTROPHY EPISPADIASIS COMPLEX


Presented by-

DR SHIVAM SINGH

Mch Urology Resident

DR DEEPAK MANE

Assoc. Professor Urology

DR V P SABALE

HOD & Professor Urology

DYP MEDICAL COLLEGE AND HOSPITAL

- Patient 18 years old gentlemen ,

- With Chief complaint of - -Defect over Lower Abdomen since Birth
  -Urine Leakage from defect since Birth
  -Small Sized penis with upward deviation since Birth.

- H/O Present Illness – Patient complaints of defect over lower abdomen since birth associated with urine leak from defect .

- No history of any previous surgery . Pt has not taken any surgery advise till date.

- No history of any co-morbidity.

- Unmarried.

- No addictions.
- No h/o Similar complaints in family.

- On General Examination :-
- Patient conscious ,well oriented to time place and person
- Moderately Built
- BP= 120/80 mm Hg
- Pulse = 68 / min
- Spo2 = 99% on room air

Systemic Examination :-

- CVS - S1S2 Present
- CNS - GCS 15/15
- P/A –Soft Non-Tendor , No palpable organomegaly.
  Umblicus - Absent

# Local Examination

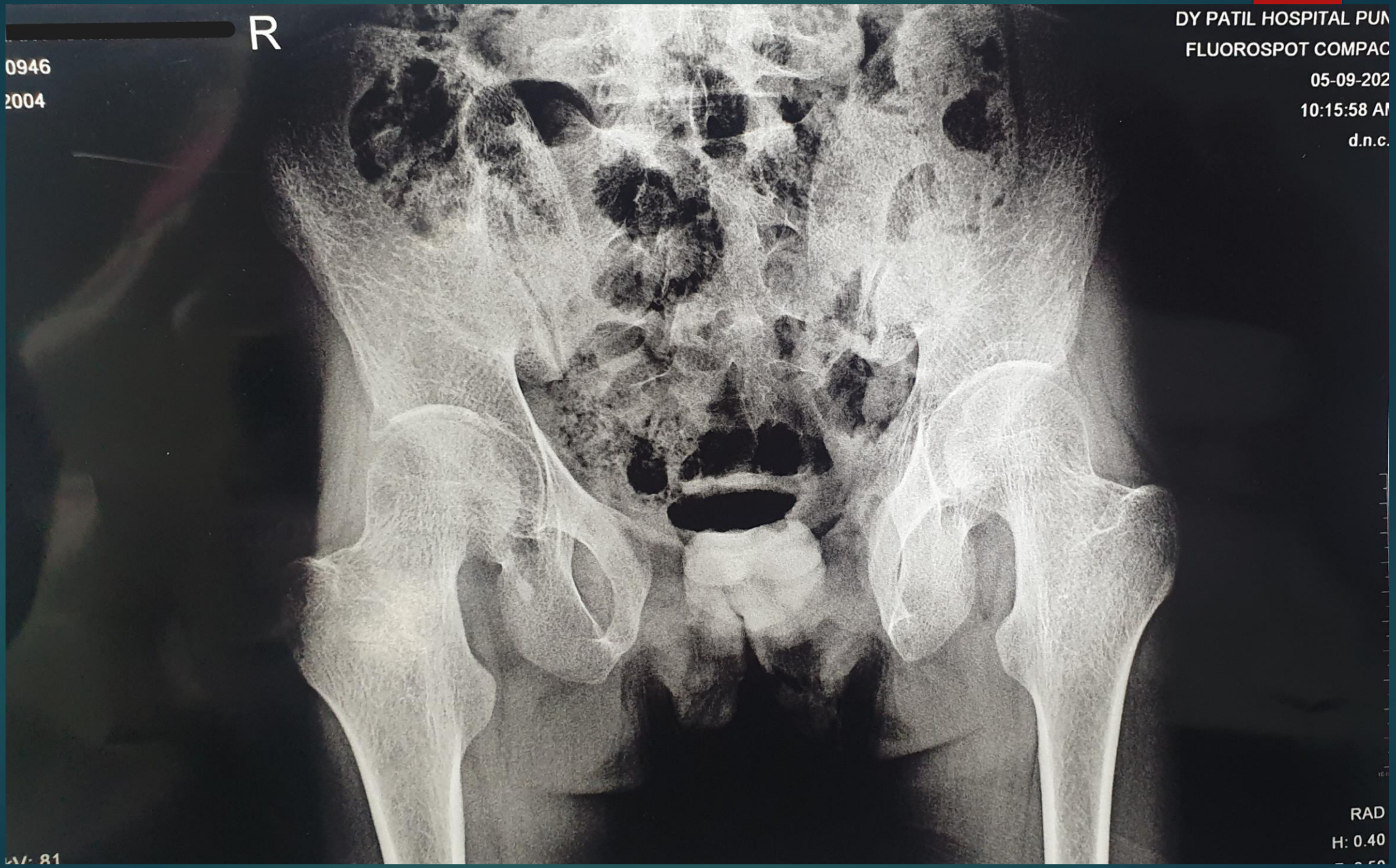
- Defect of size approx. 8 x 10 cms present in hypogastrium with visible ureteric orifices per abdomen with urinary leakage.

- Ill formed short penis With Dorsal Chordee.

- B/L Ureteric Orifices seen dorsally in the wound on the abdominal wall

- Bilateral testis Palpable with Rugae Present over the Scrotum.

- Pubis Symphysis Not Palpable with Presence of Pubic Diastasis with a gap of approximately 5 cms.


R
0946
2004
DY PATIL HOSPITAL PUN
FLUOROSPOT COMPAC
05-09-202
10:15:58 AM
d.n.c.
RAD
H: 0.40

# Ultrasound Abdomen

Rt Kidney – 8.3 x 1.4 cms
Normal in size and echotexture.
CMD Well maintained.
No Calculus or hydronephrosis.

Lt Kidney – 8.6 x 1.5 cms
Normal in size and echotexture.
CMD Well maintained.
No Calculus or hydronephrosis.

Urinary bladder was seen over the abdominal wall through a defect.

B/L testis were normal in size and texture

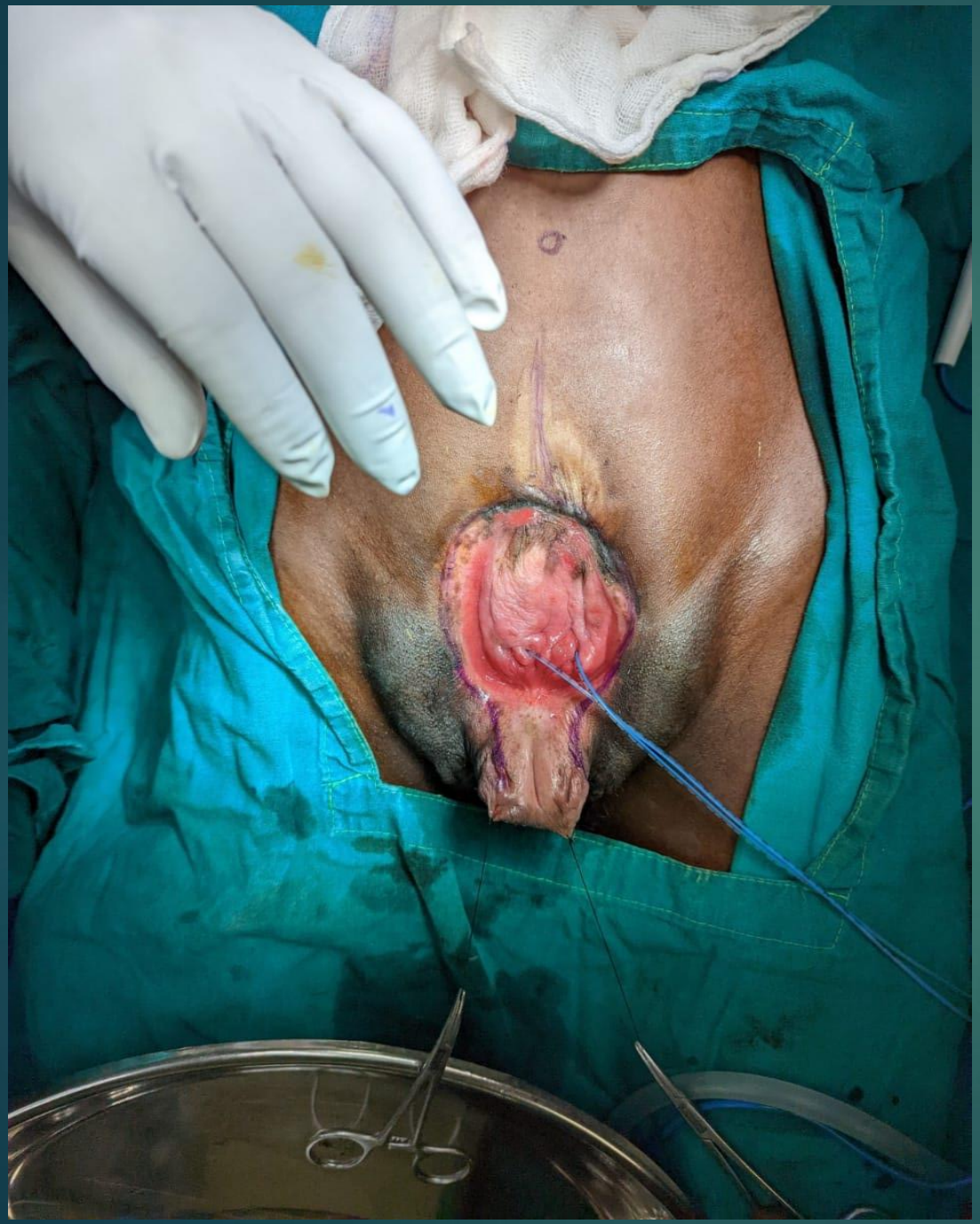
# Classical Bladder Extrophy with Epispadiasis Complex

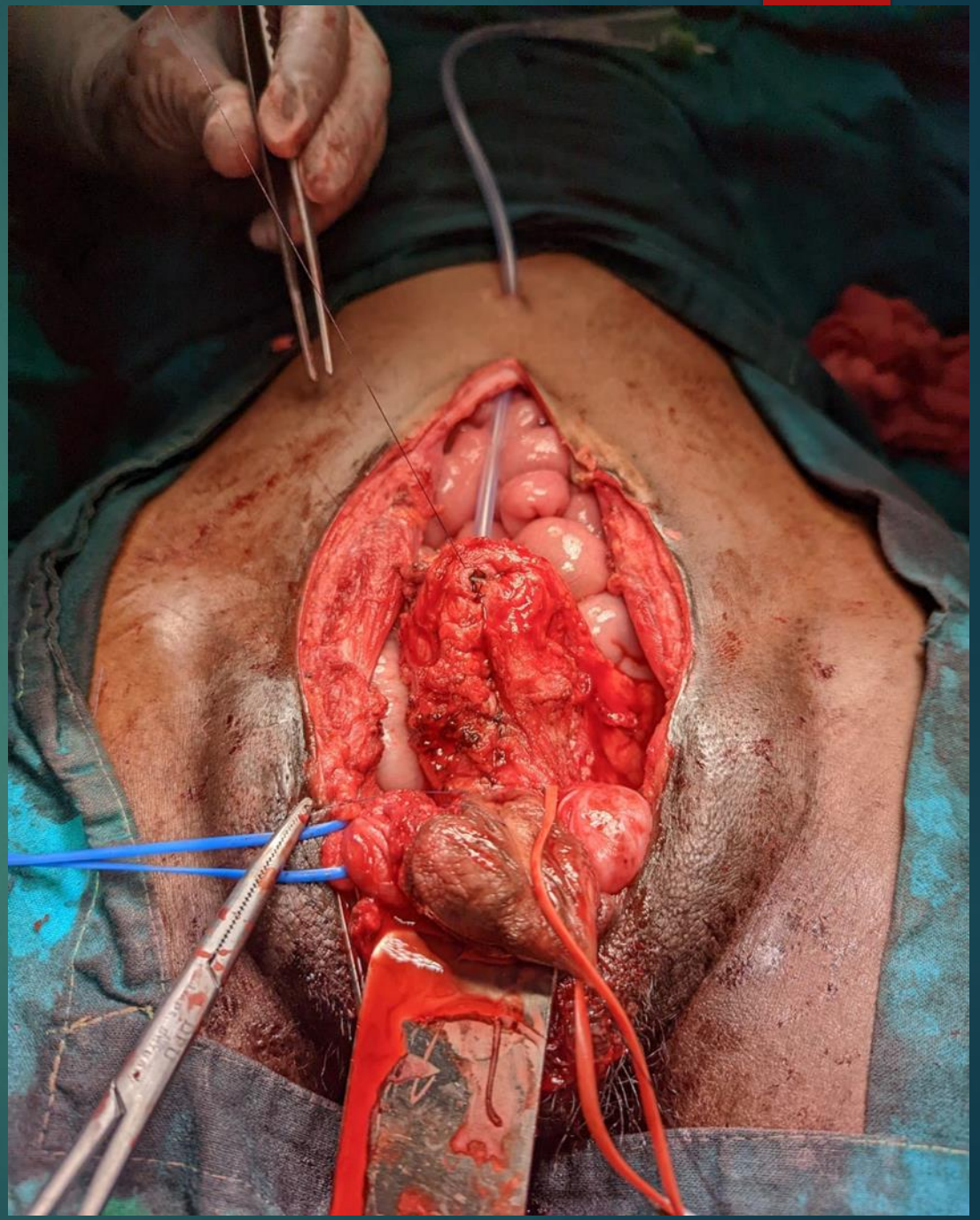
# Management

-Orthopaedic Consultation

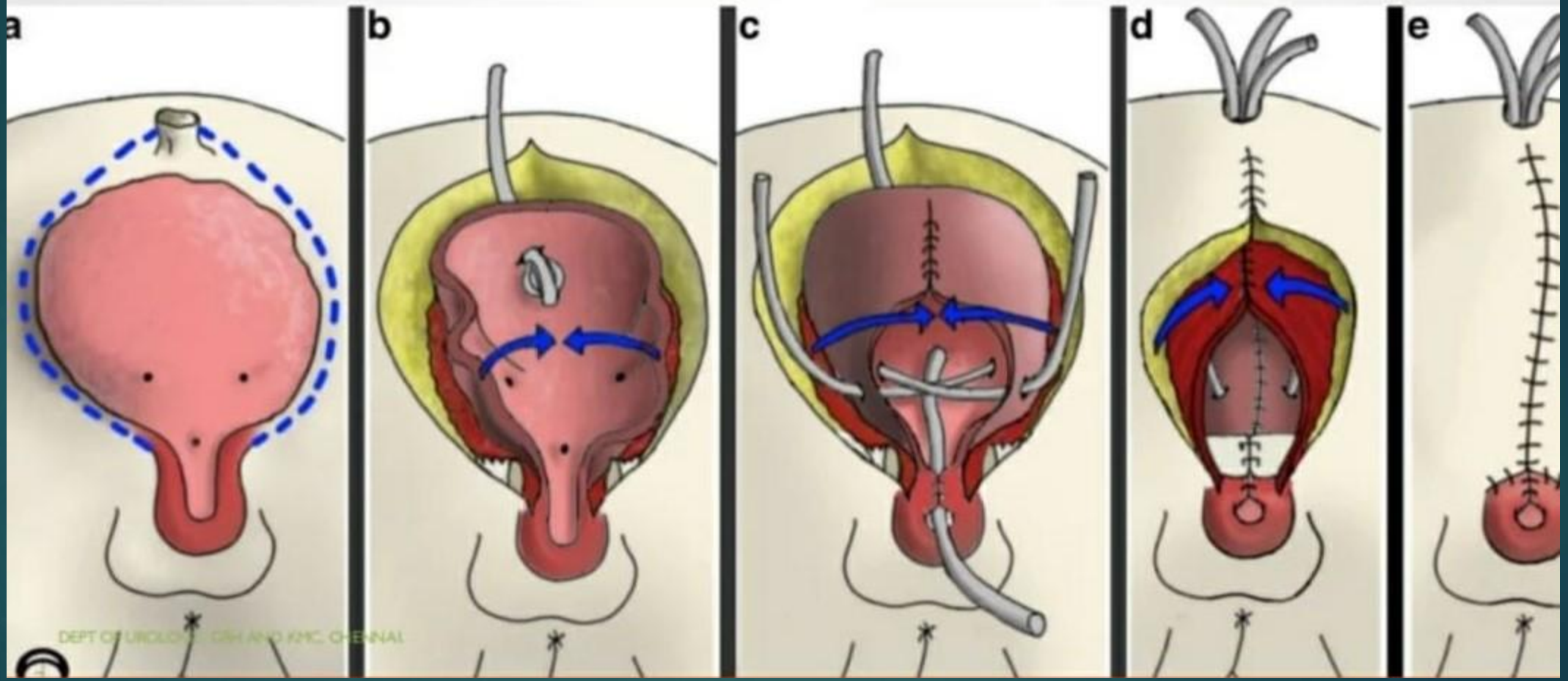
-Surgical Options

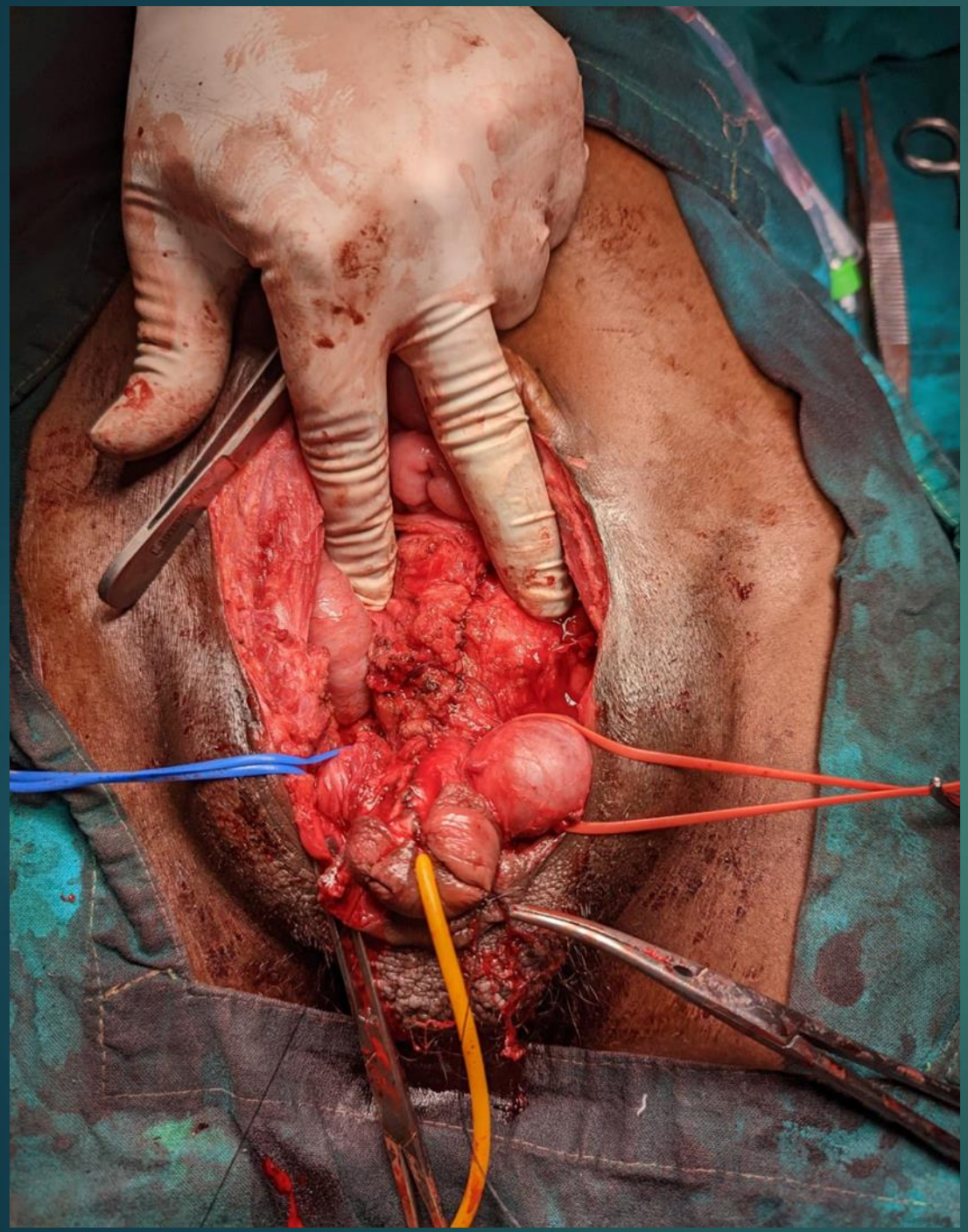
-Plastic Surgery Consultation

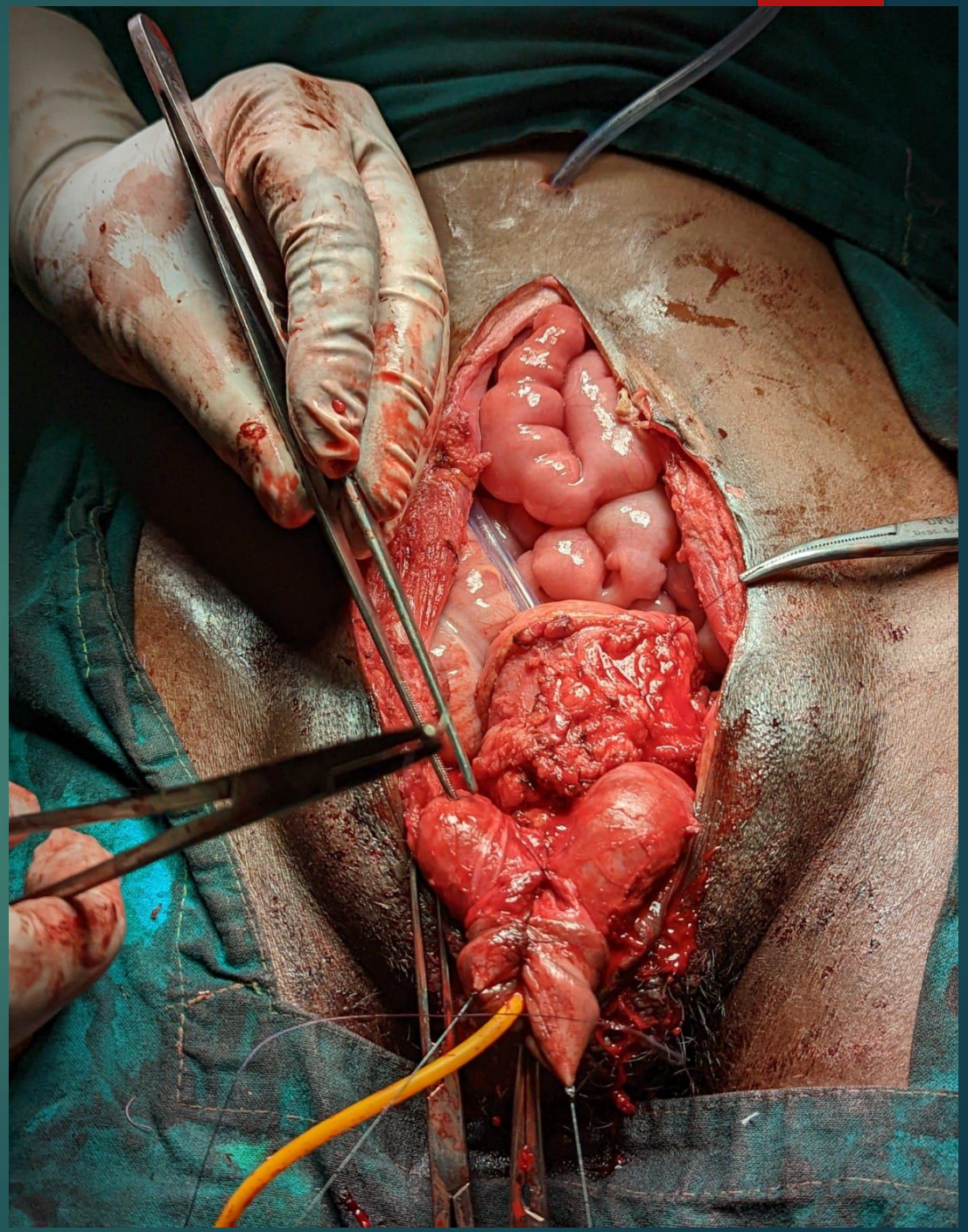




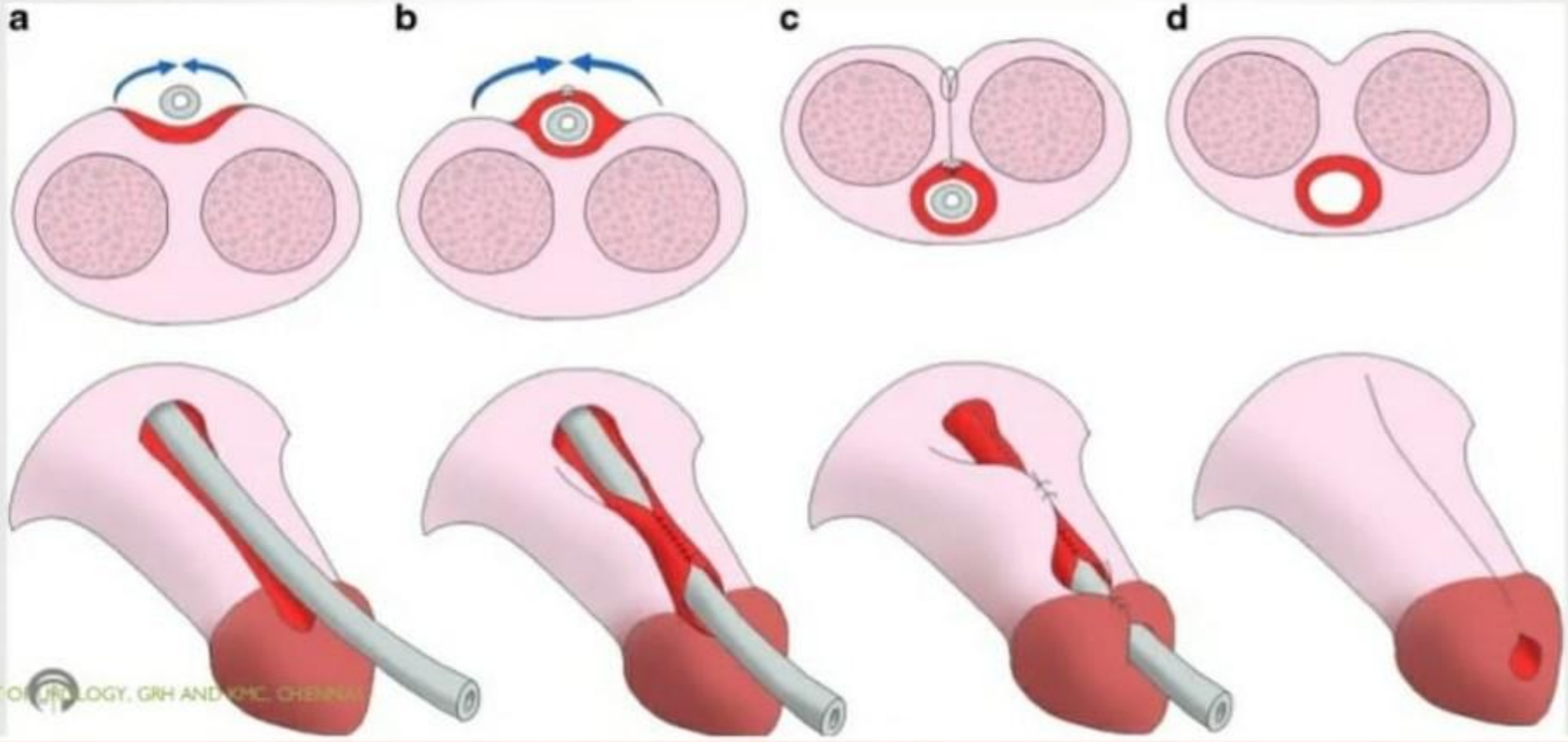
# MSRE STAGE I

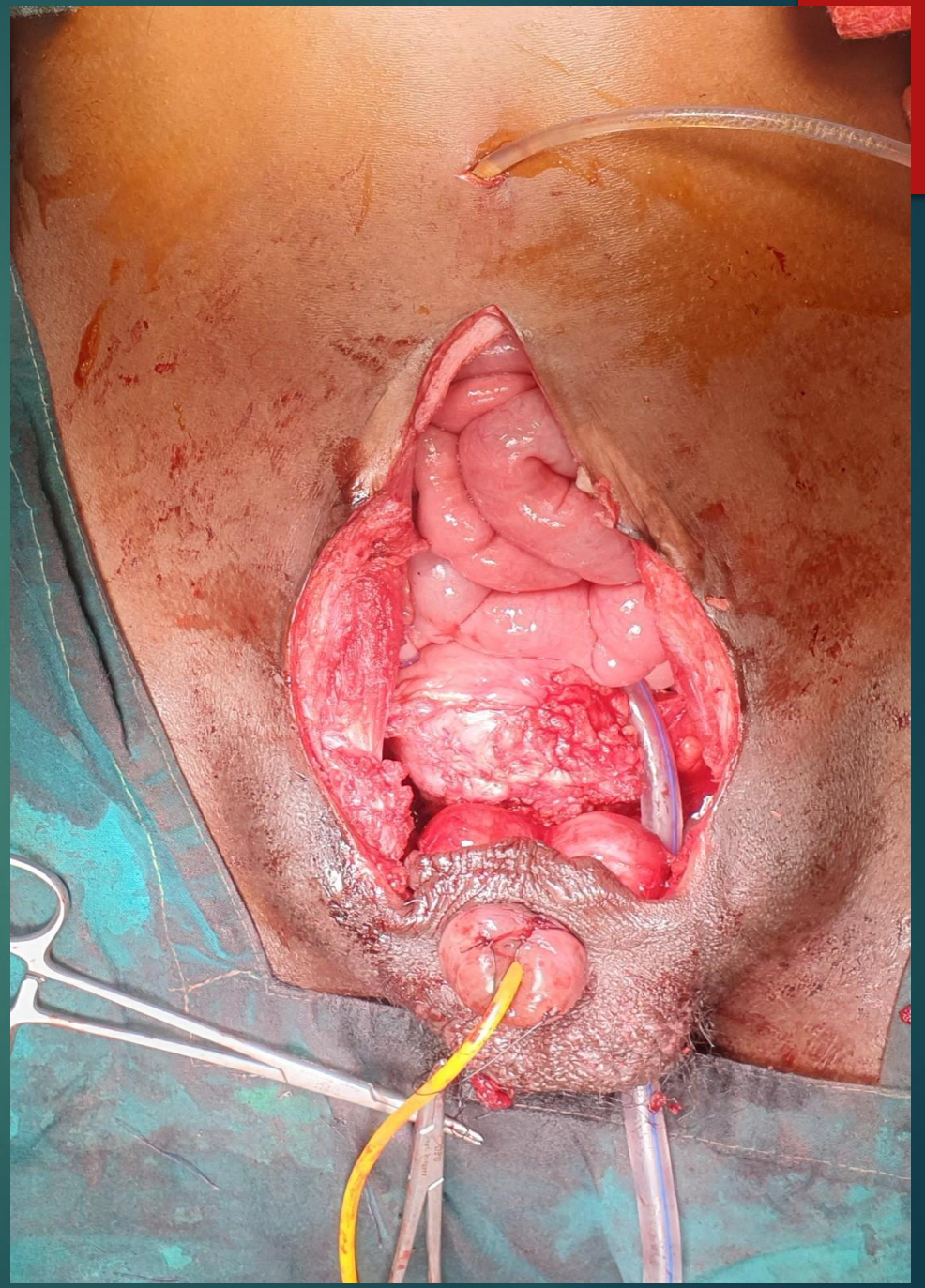


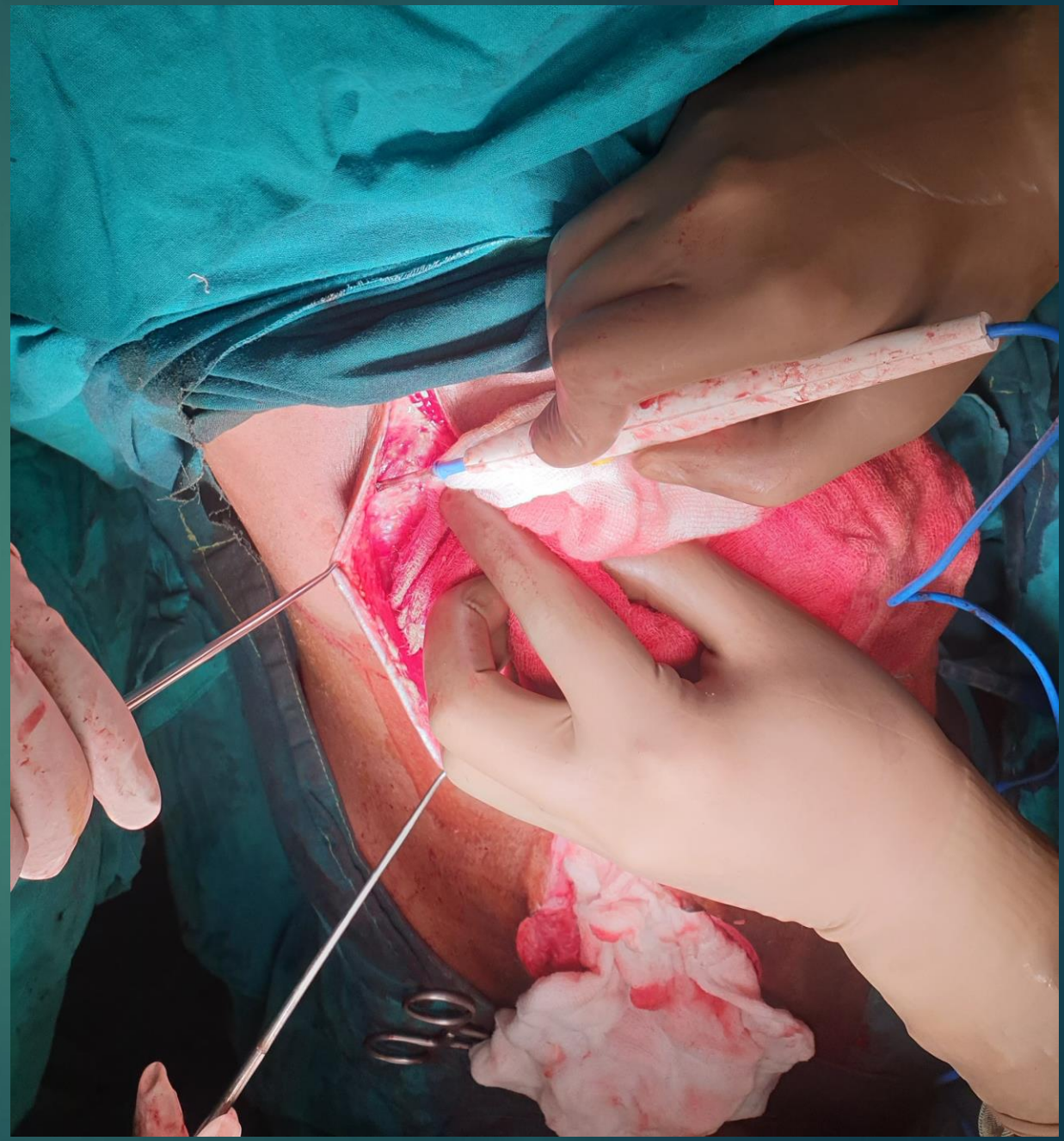




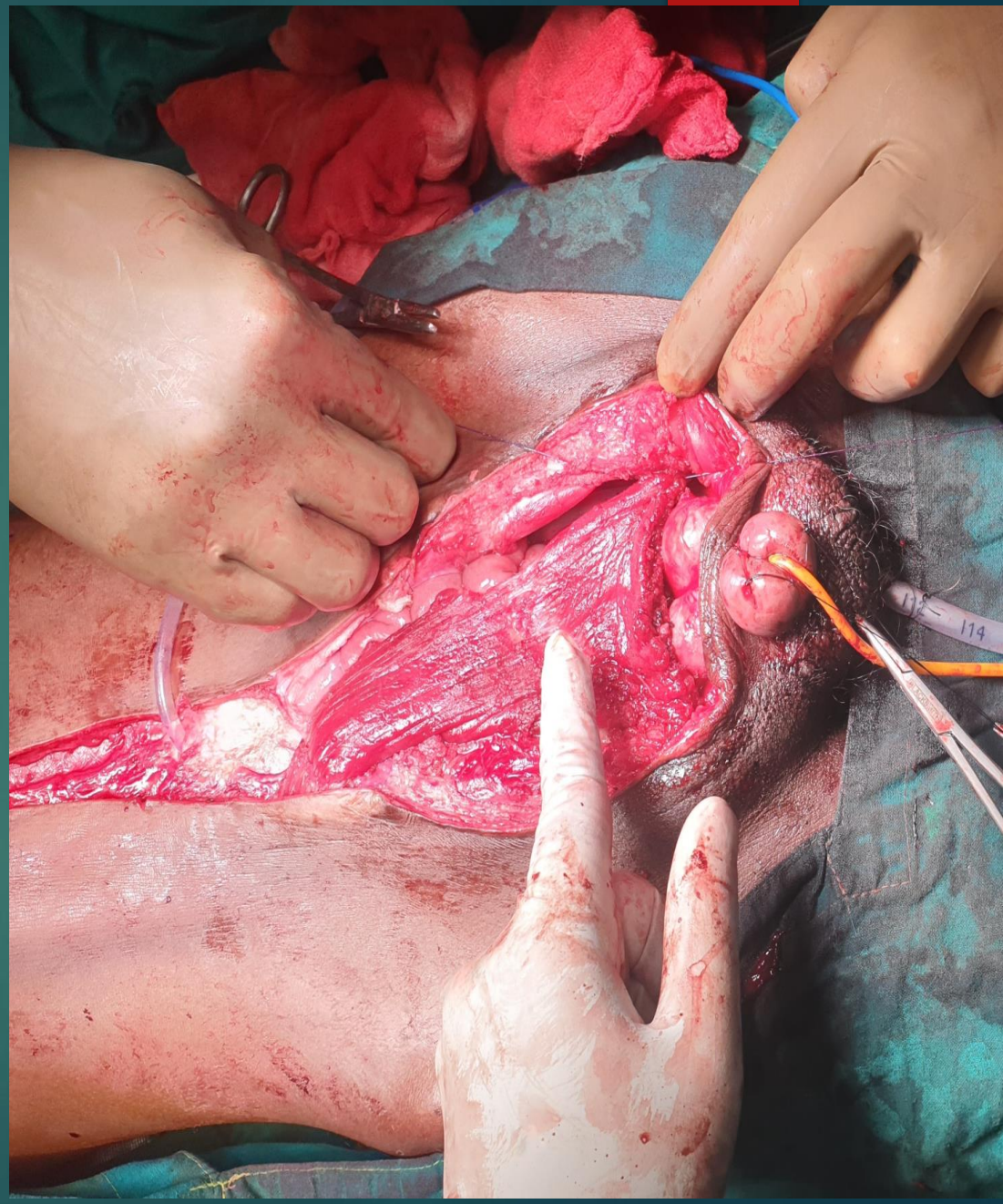
# MSRE STAGE II
# (MODIFIED CANTWELL-RANSLEY REPAIR)








114


114

# Discussion

## Incidence

- Estimated to be around 2.15 cases per 1,00,000 live births.
- Risk of recurrence in a given family is approx. 1 in 100.

## Etiology

- 10 folds increase in exstrophy births to mother received large doses of progesterone in first trimester.
- Increased Maternal Age
- CASPR 3 Gene on Chromosome 9

# Anatomical Considerations

## Skeletal Defects -

External rotation of posterior and anterior pelvic segment by 12 and 18 degrees respectively

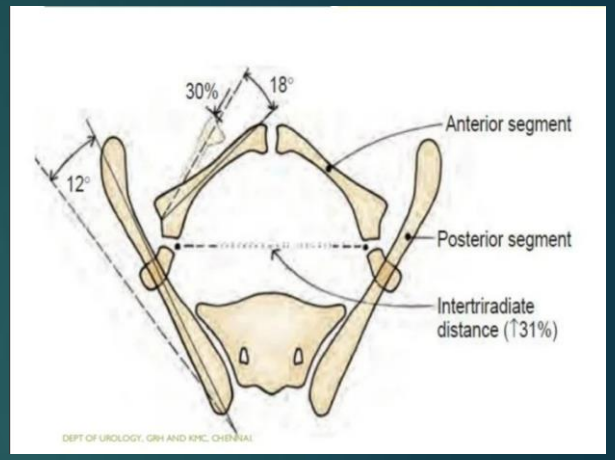
Inferior rotation of bony pelvis

Increased Pubic Diastasis

Increased inter triradiate cartilage distance

Shortened anterior pubic segment by 30 %.

Pelvic floor defects .




## Abdominal wall Defects -

Triangular lower abdominal wall defect with post wall of bladder and posterior urethra visible.

B/L Indirect Inguinal hernias

Anal opening anteriorly displaced just behind urogenital diaphragm with imperforate anus.

## Male Genital Defects -

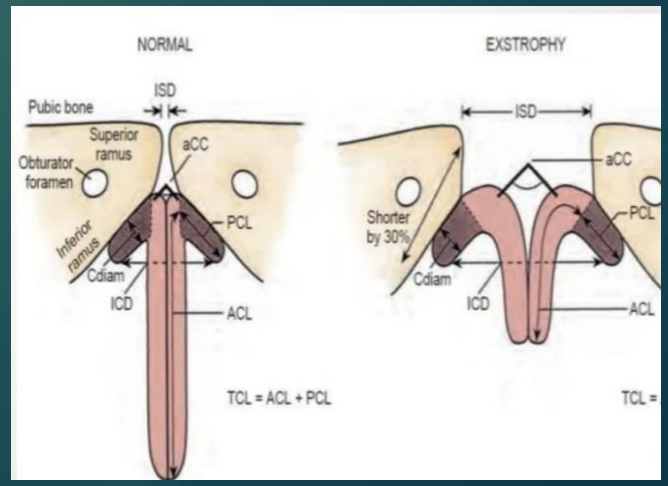
Anterior Corporal length decreased by 50%

Prostate/ Seminal Vesicles/ Vas deferens – Normal

Testis – Undescended

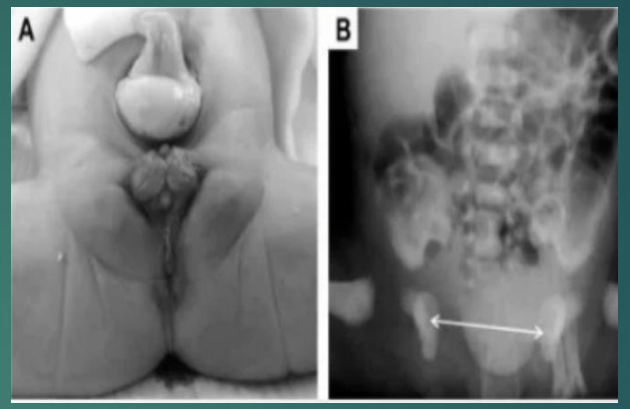
Cavernous Nerves laterally displaced.

Anterior Wall of bladder deficient but with Normal neurophysiological composition .

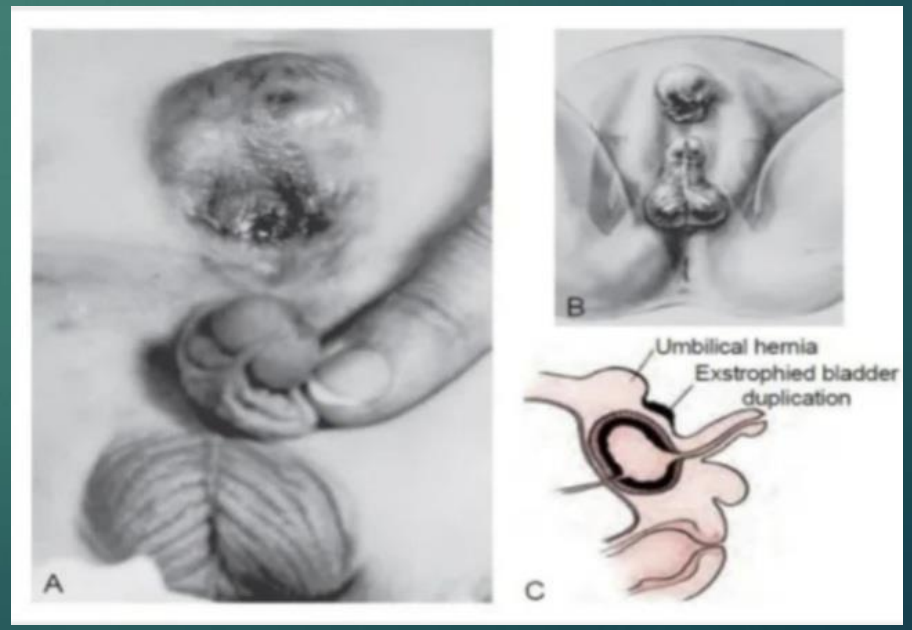


# Variants of CBE

- Pseudo-Extrophy
  - Musculoskeletal defects present
  - Urinary Tract defects absent




- Superior Vesicle Fissure Variant
  - Musculoskeletal defects present
  - Urinary tract defects – Only in uppermost part
    - Vesicostomy
- Duplicated Extrophy
  - Musculoskeletal defects present
  - Extrophied Bladder Present – Duplicated
  - Normal bladder also present in pelvis
- Covered Extrophy
  - Split Symphysis Variant
  - Musculoskeletal defects present
  - No significant urinary tract defect
- Female Epispadiasis




# Management

Modern Staged Repair of Extrophy (MSRE)

Complete Primary Repair of Extrophy (CPRE)

# Management

## Most Widely Used – MSRE

### 3 Stages

#### Stage 1 -Newborn

Bladder Template Closure ± Osteotomy.

B/L Anterior Innominate & Vertical Iliac Osteotomy.

#### Stage 2 – 6- 10 months

Epispadiasis Repair with correction of Chordee.

Increases Bladder Outlet resistance.

Improves Bladder Capacity.

#### Stage 3 – 4 to 5 Years of Age

Adequate Bladder Capacity necessary – 100 ml.

Bladder Neck reconstruction with anti-reflux procedure.

# Thank You