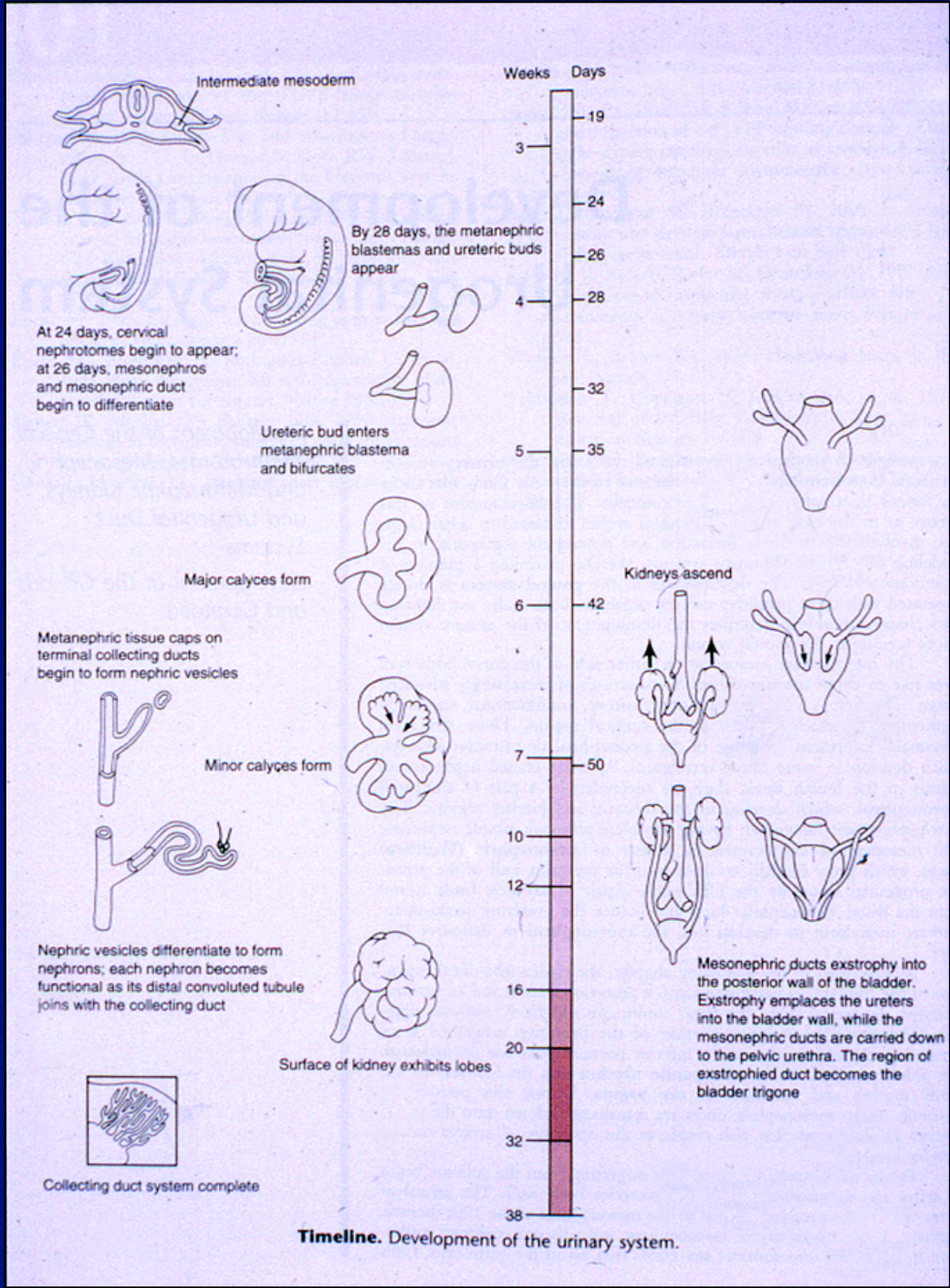
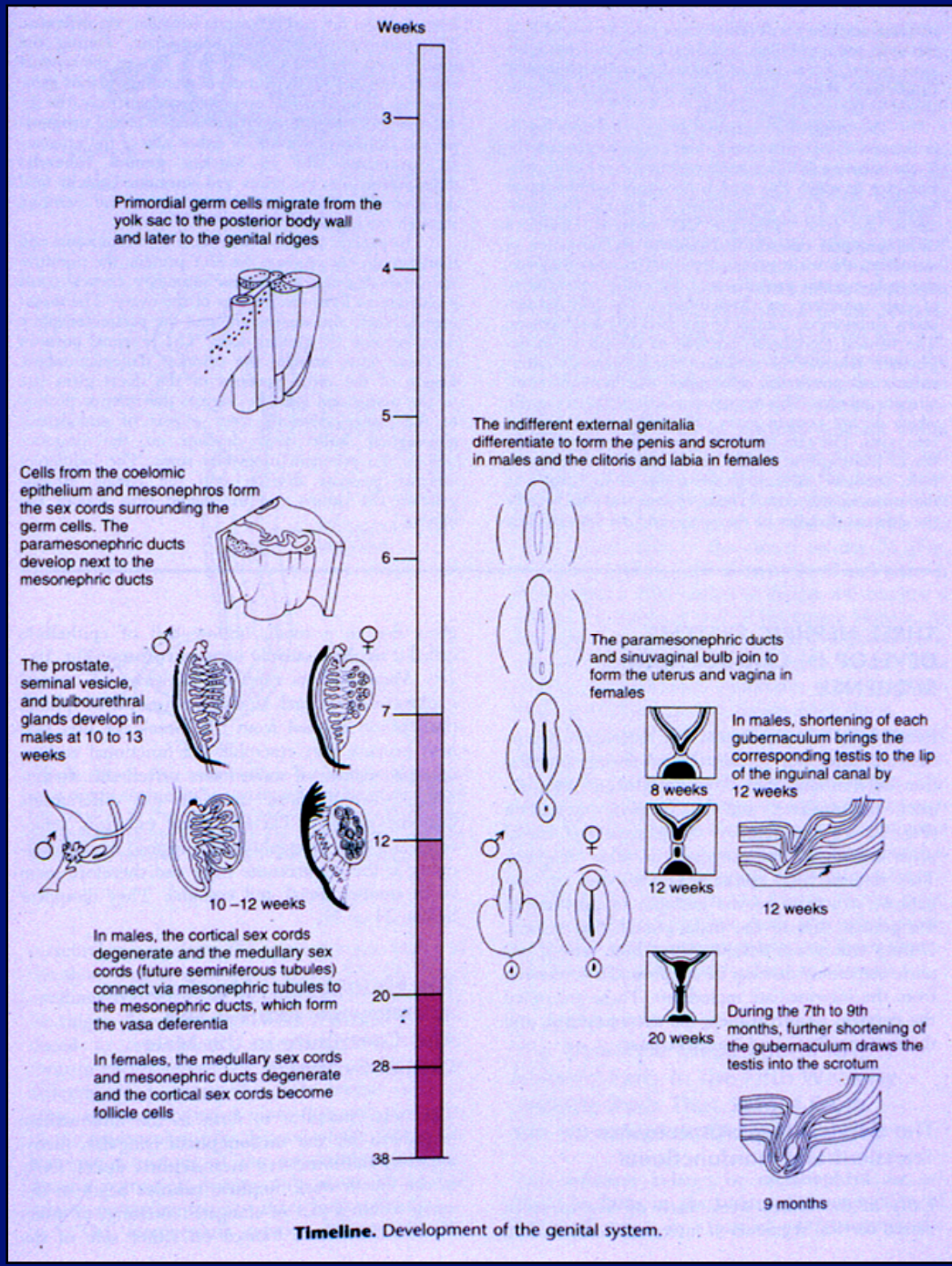


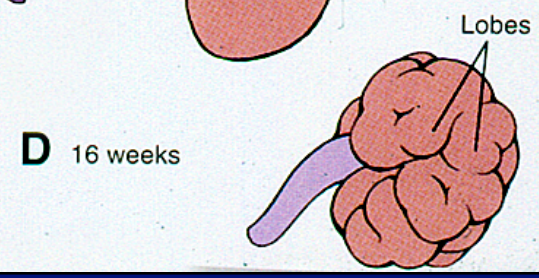
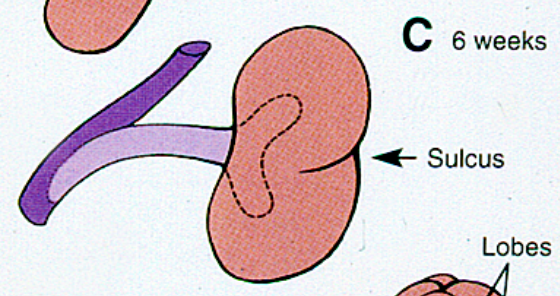
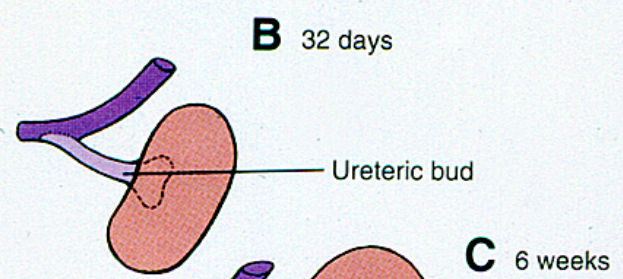
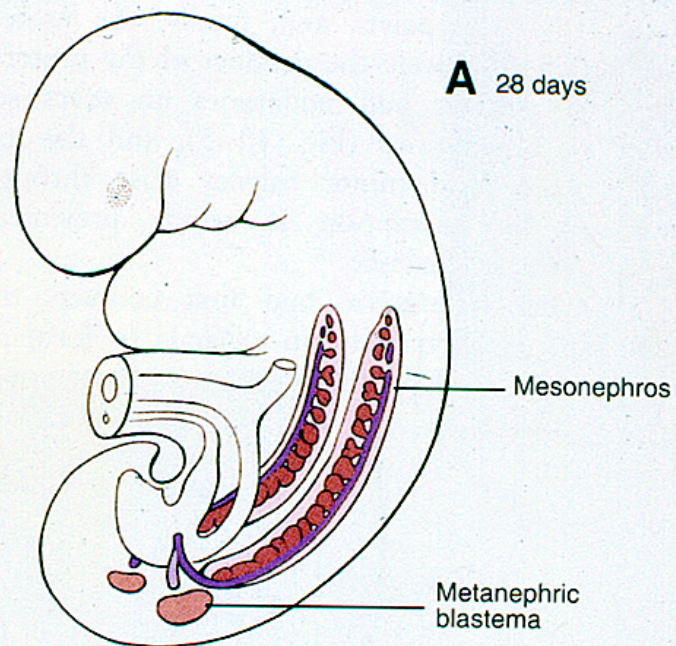


*Normal and Abnormal Development of
the Upper and Lower Urinary Tract*

Terry W.Hensle MD
Given Foundation Professor of Urology
Vice Chairman Dept of Urology
Columbia University College of P&S







RENAL DEVELOPMENT

- **WT-1** Gene located at 11p13 the “master switch” for gonadal development and renal morphogenesis
- **GDNF** Glial derived neurotropic factor implicated prominently in renal morphogenesis
- **RET** receptor tyrosine kinase is the receptor for GDNF and WT-1 at ureteral bud

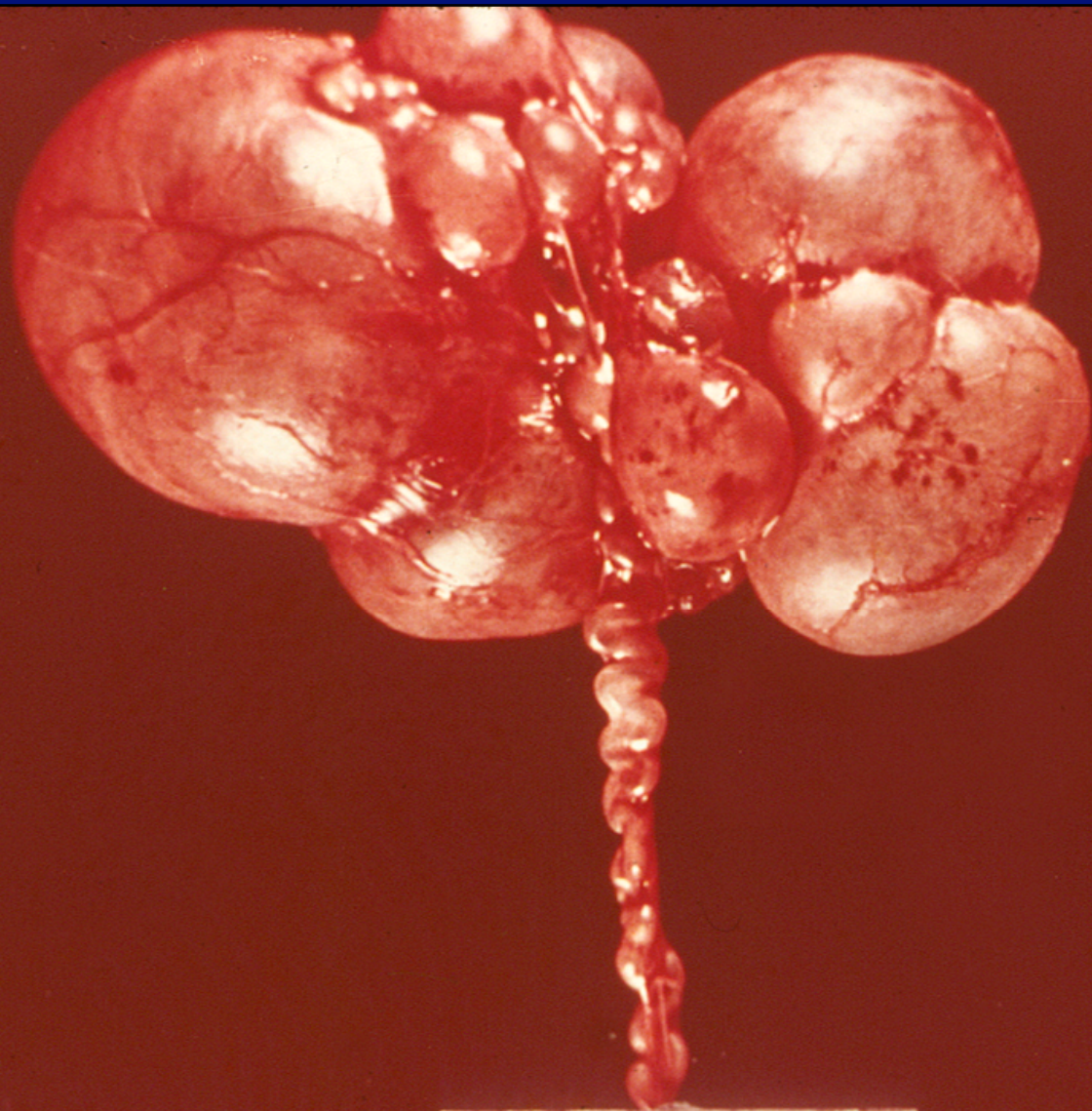
RENAL DEVELOPMENT

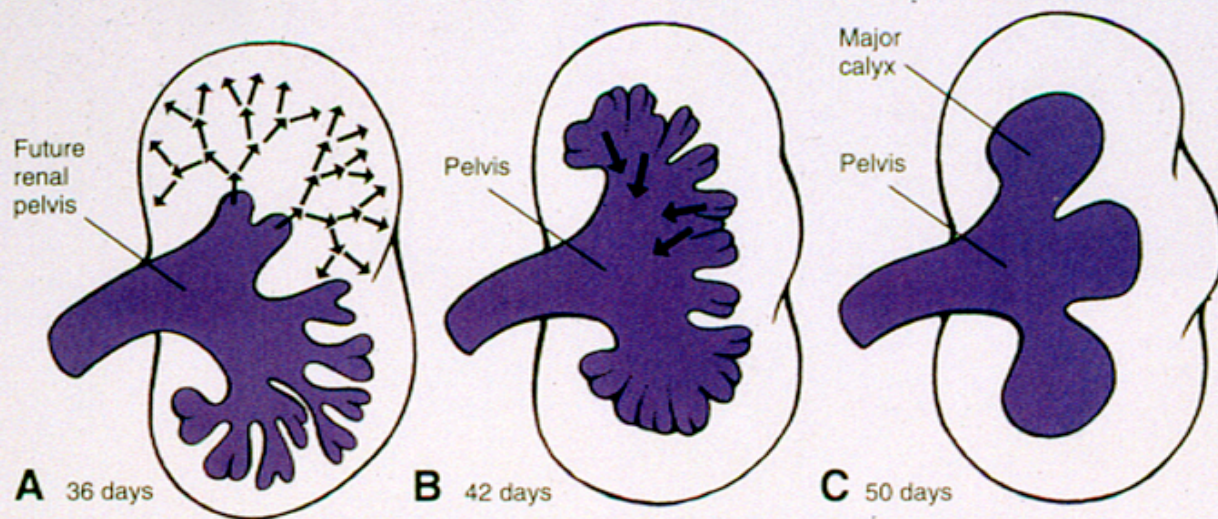
- BOTH WT-1 AND GDNF ARE NEEDED IN THE METANEPHRIC BLASTEMA FOR INDUCTION OF THE URETERAL BUD
- RET ENABLES GDNF AND WT ACTION AT THE MESONEPHRIC DUCT TO INDUCE THE URETERAL BUD

Renal Agenesis



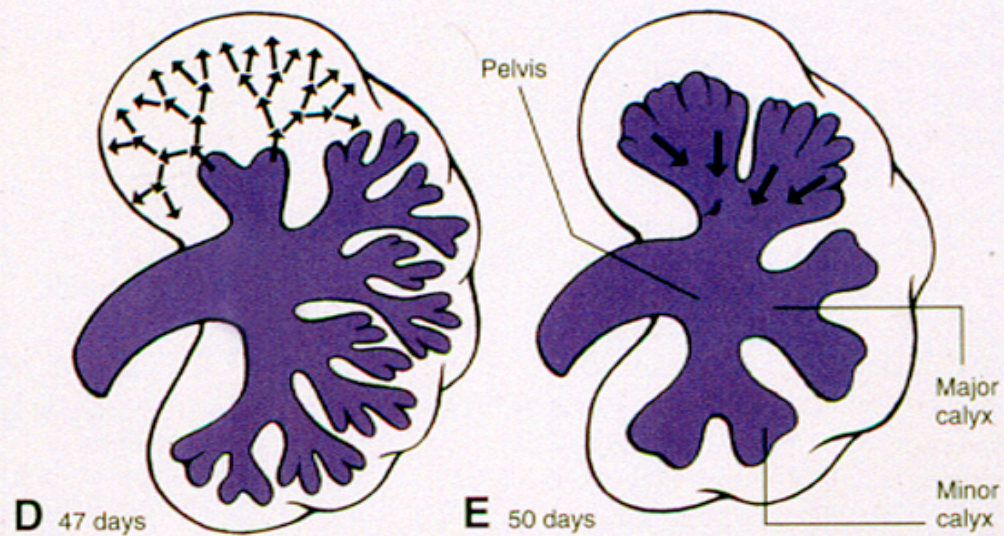
Multicystic Kidney





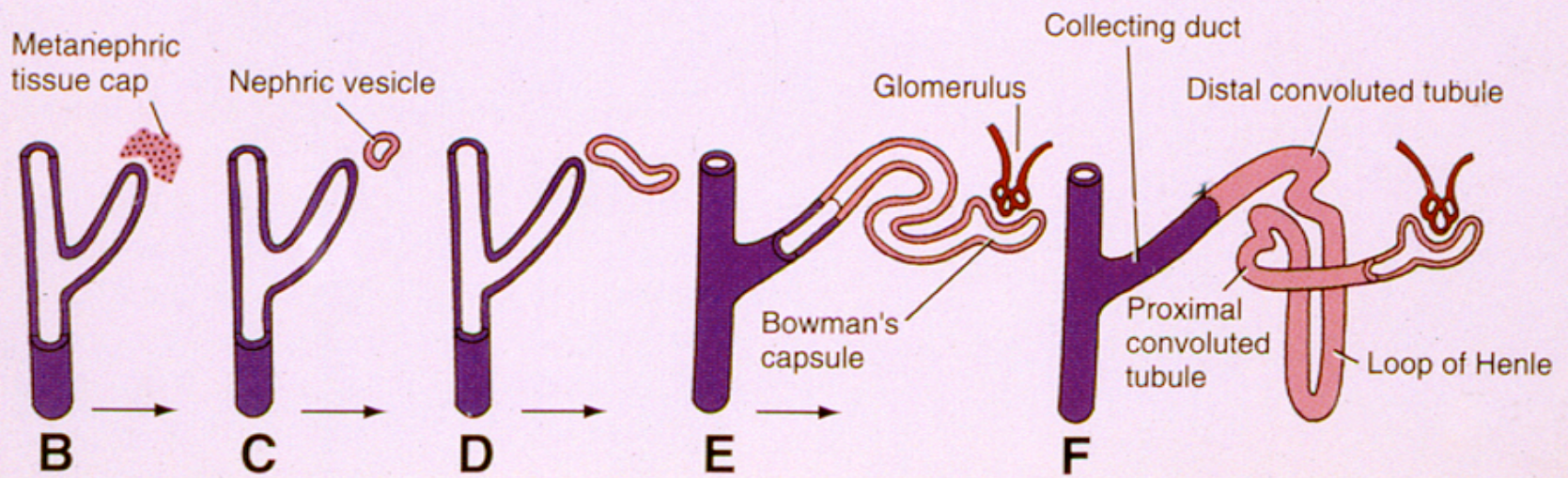
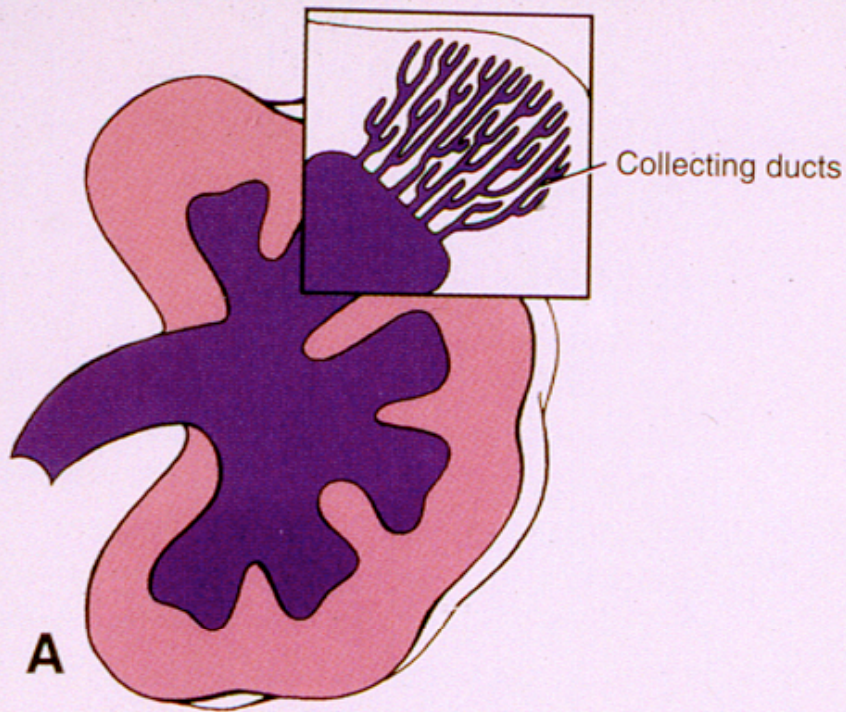
← Four generations of bifurcations →

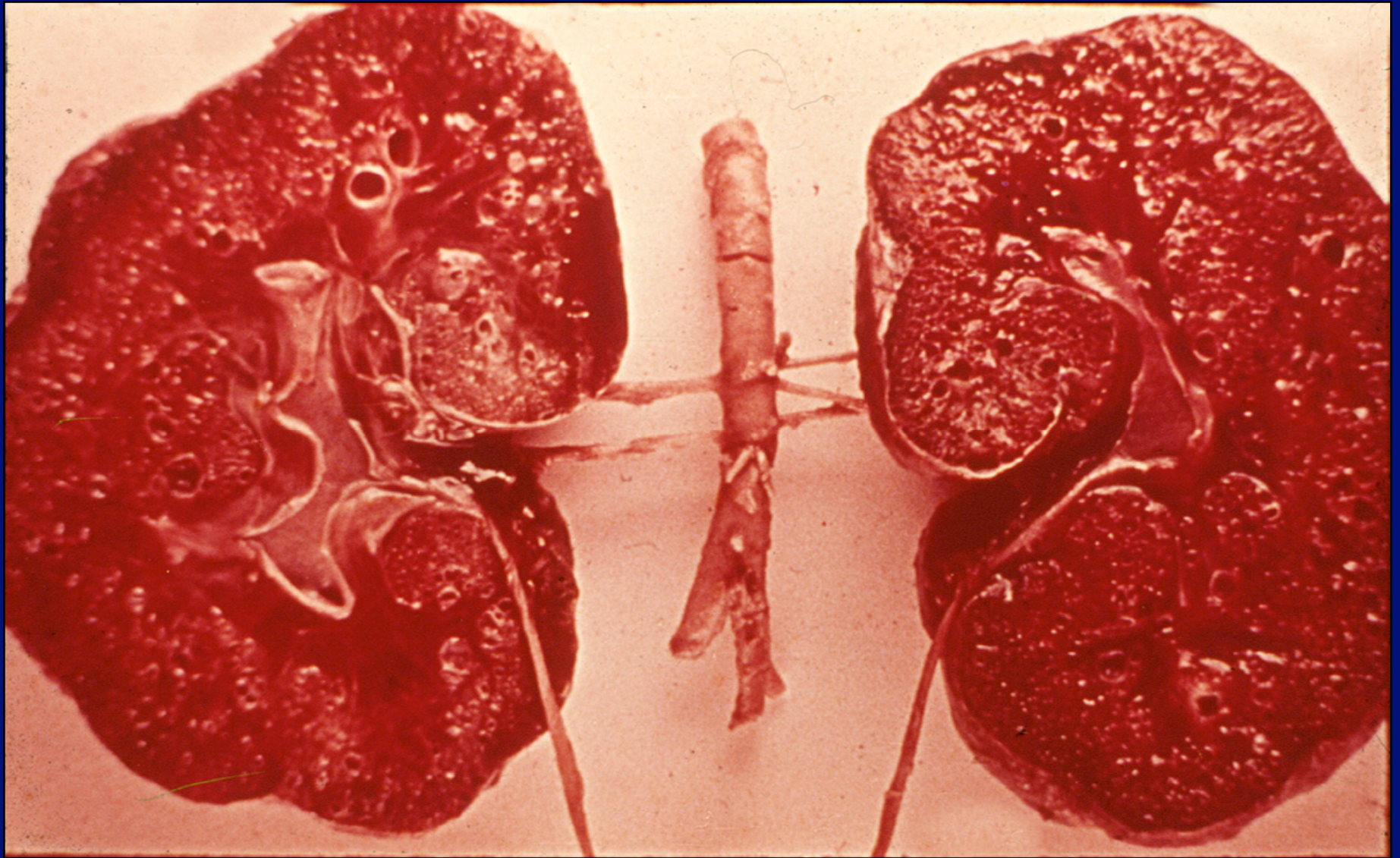
← Resorption →



← Branchings →

← Resorption →





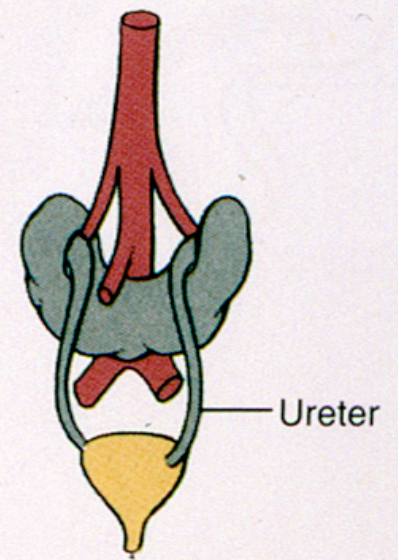
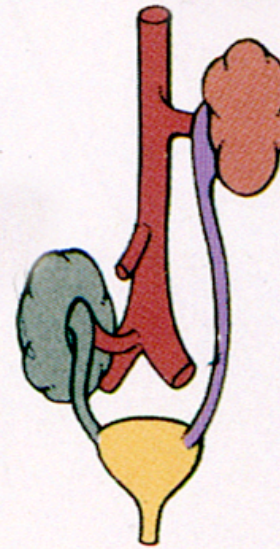
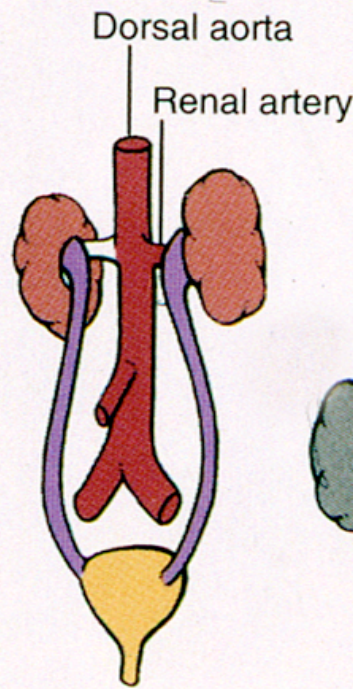
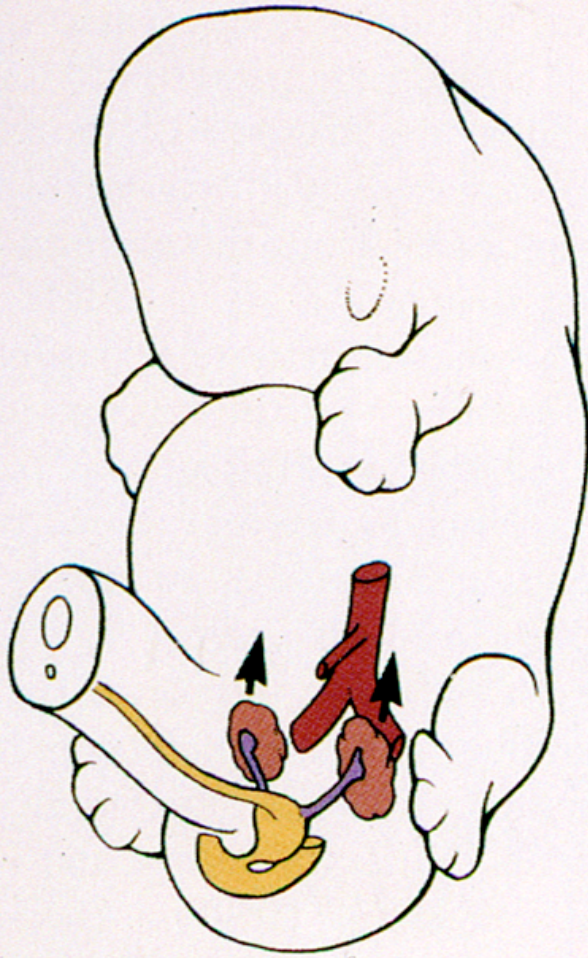


Renal Maldevelopment –Genetic Renal Cystic Disease

- AN EXAMPLE OF AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE. (ARPKD)
- ASSOCIATED WITH NEARLY UNIFORM INFANT MORTALITY
- ASSOCIATED WITH PKHD1 ON CHROMOSOME 6

Failure of Assent

Pelvic Kidney (Ectopic)
Horseshoe Kidney

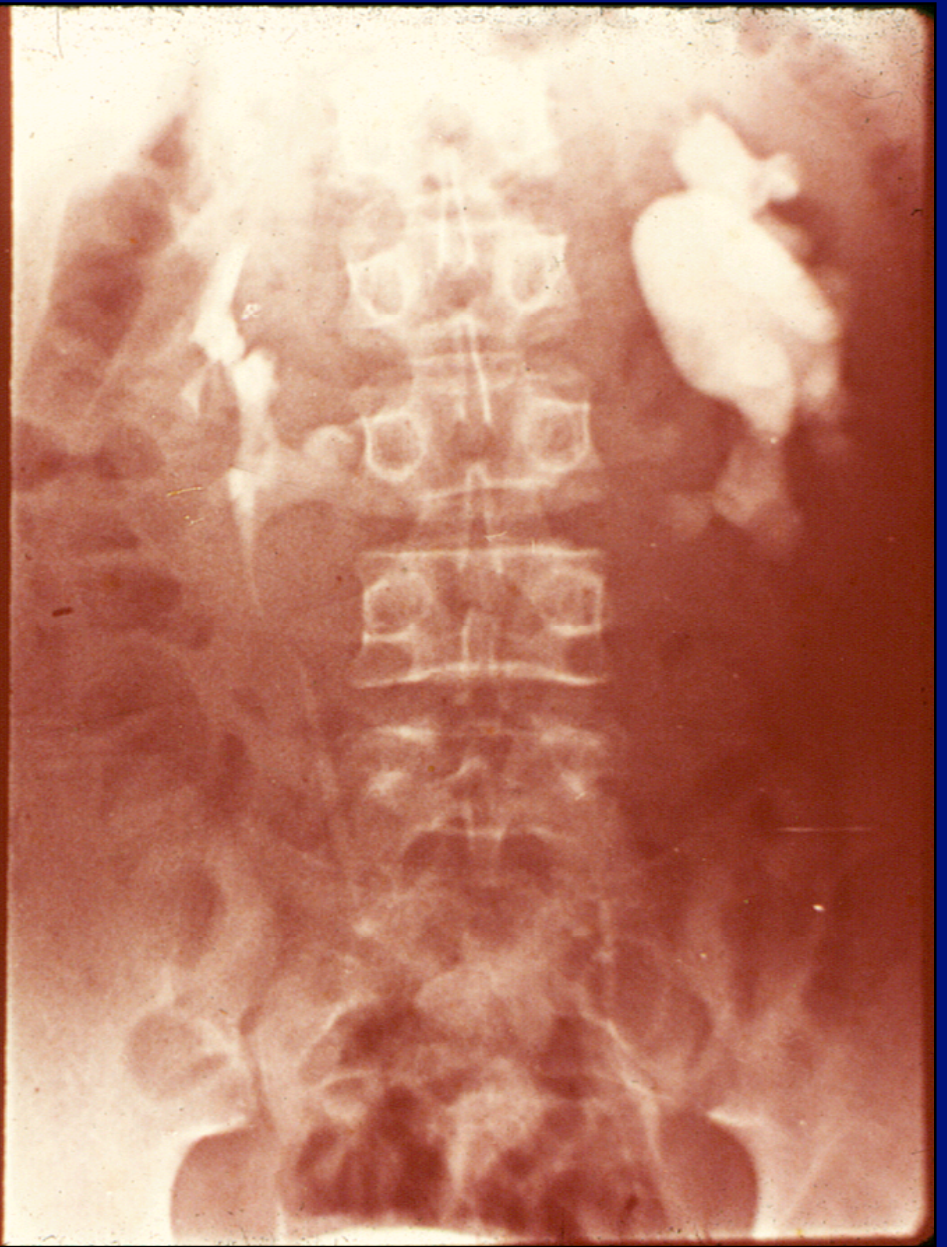


A 6th week

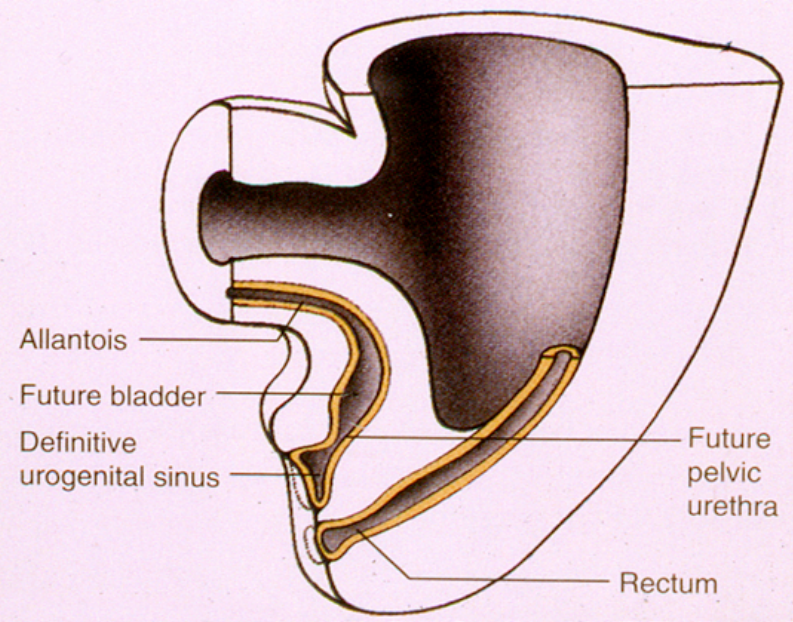
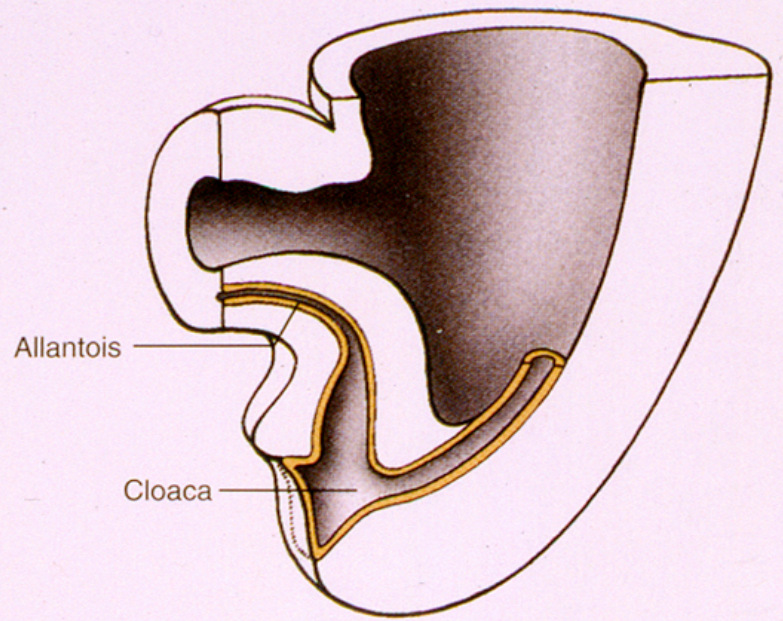
B Normal

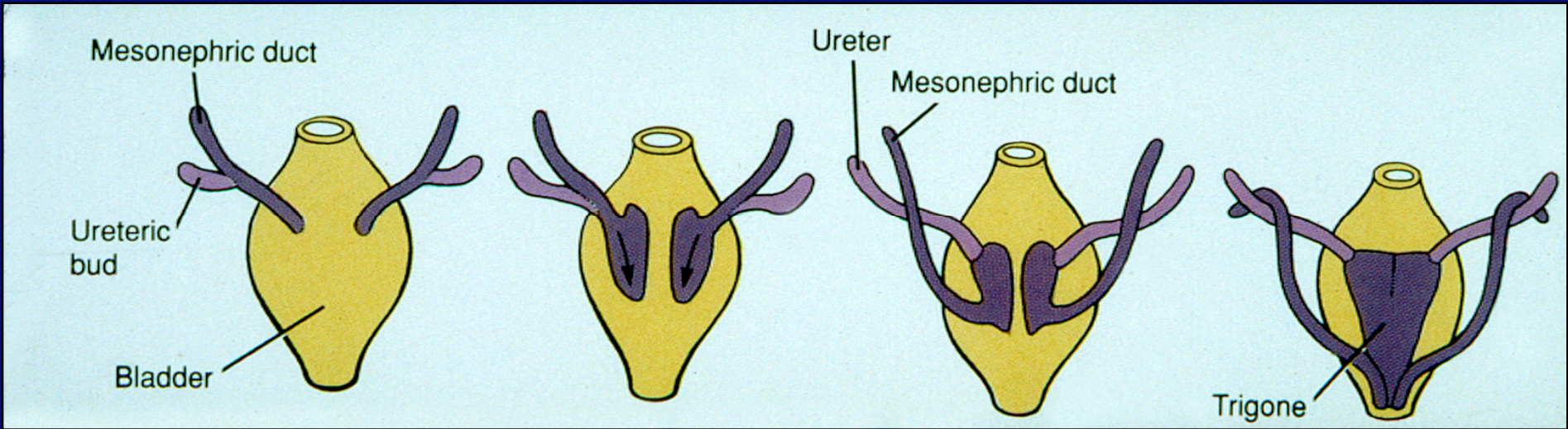
C Pelvic kidney

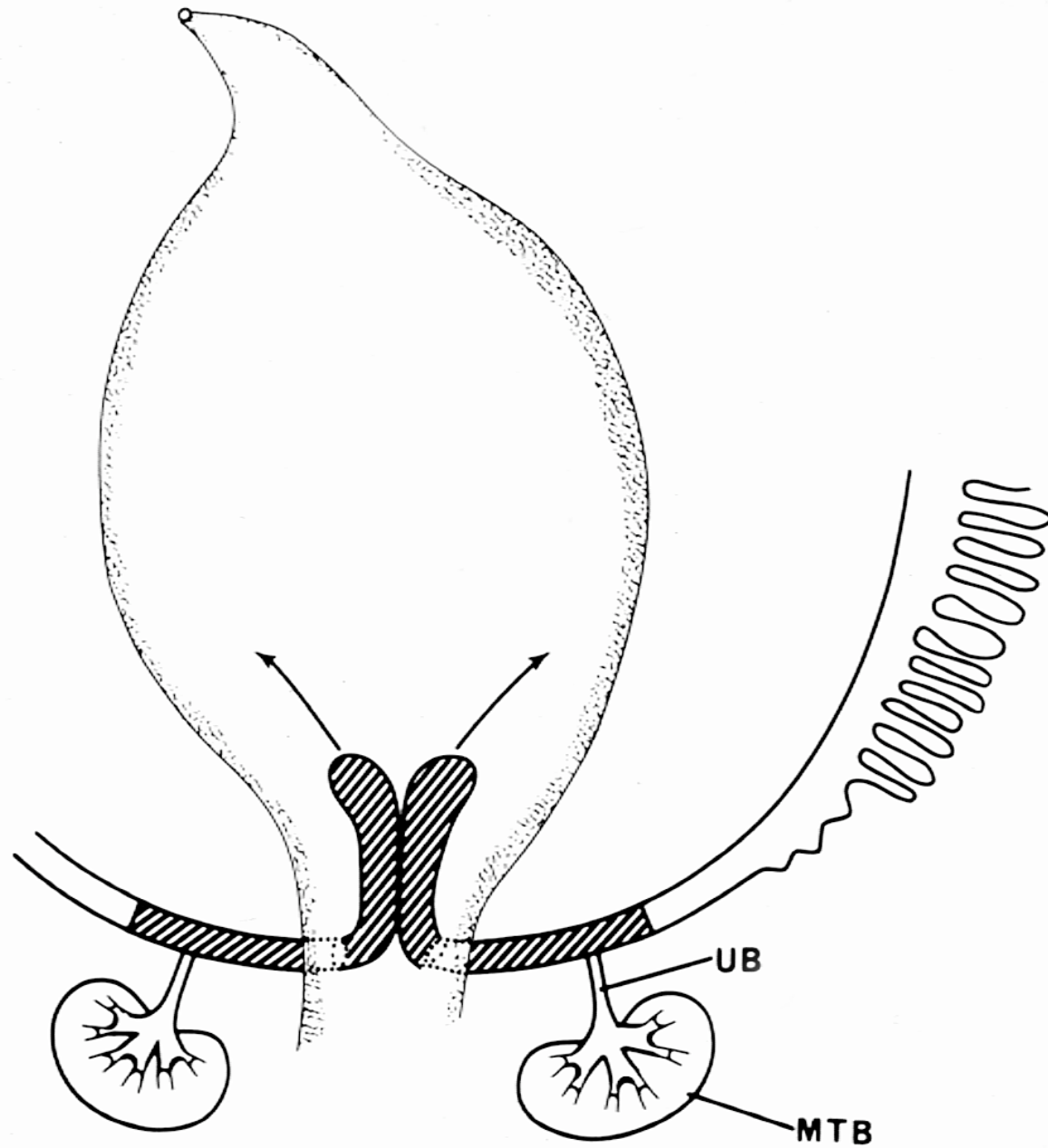
D Horseshoe kidney

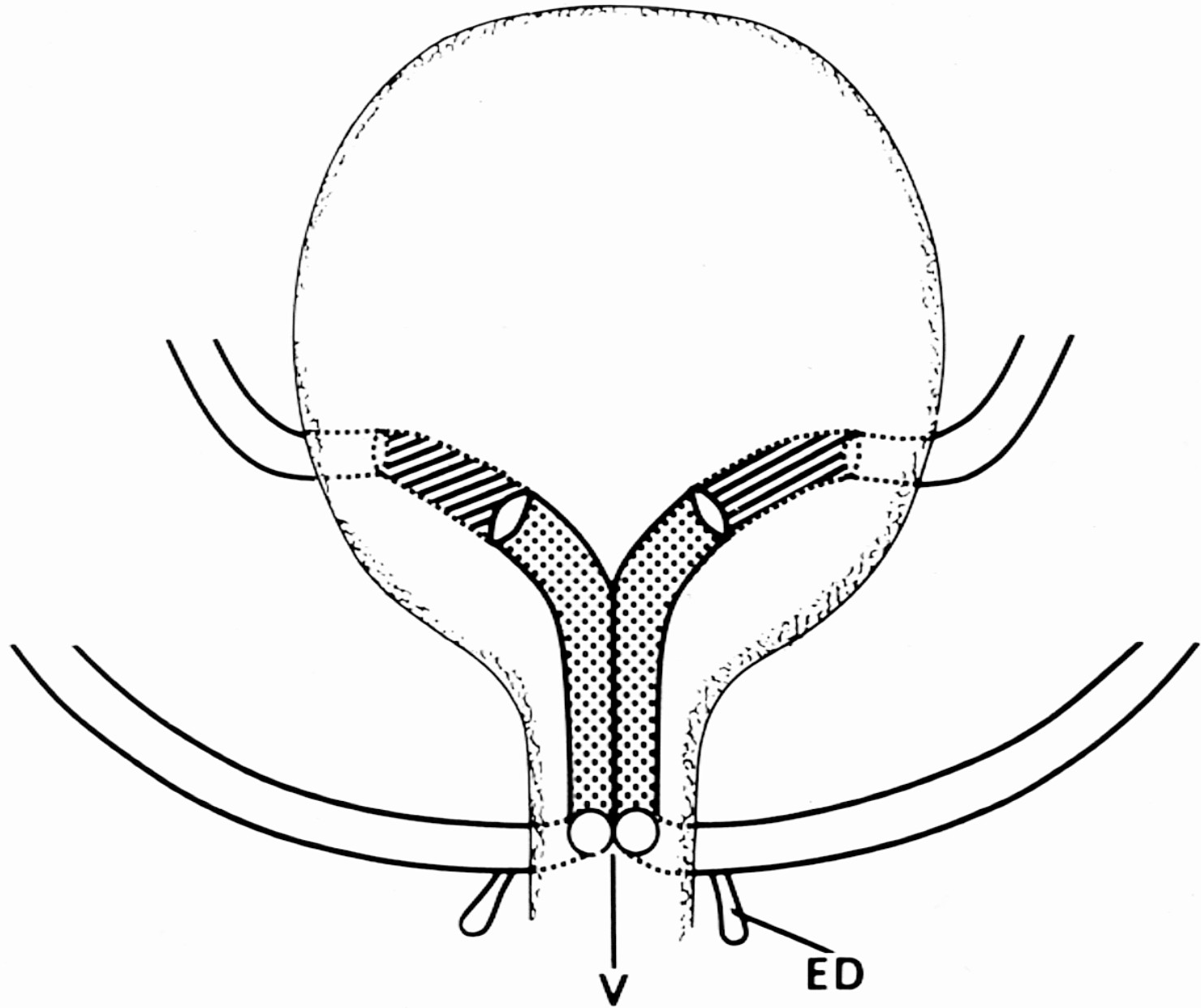


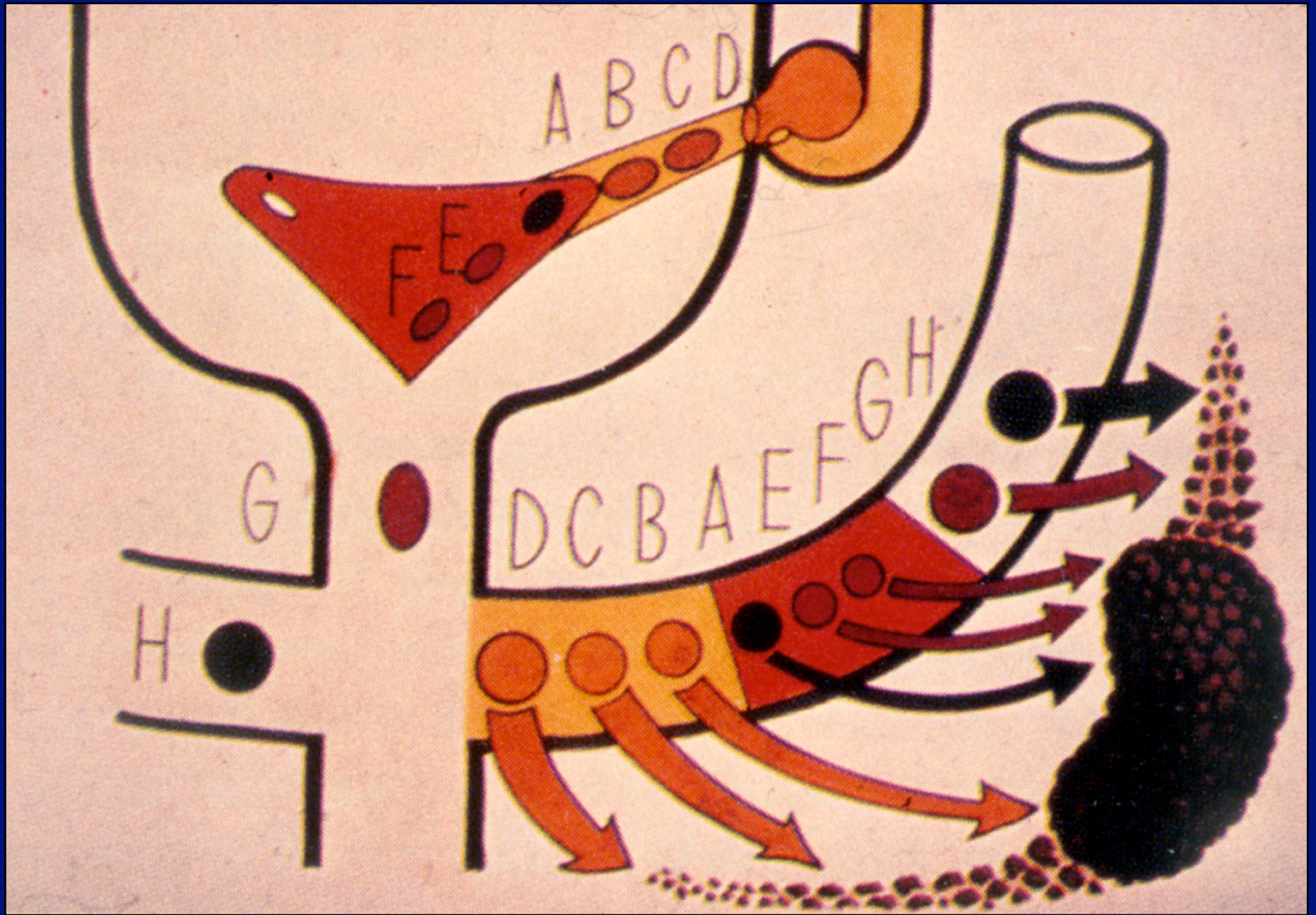
*Developmental Abnormalities of the
Bladder*











Abnormalities of Ureteral Bud Development

URETERIC BUD DEFECT

ASSOCIATED RENAL ANOMALY

Failure of bud

Renal agenesis

Abnormal position of bud

Ureterocele, ectopic ureter
diverticulum

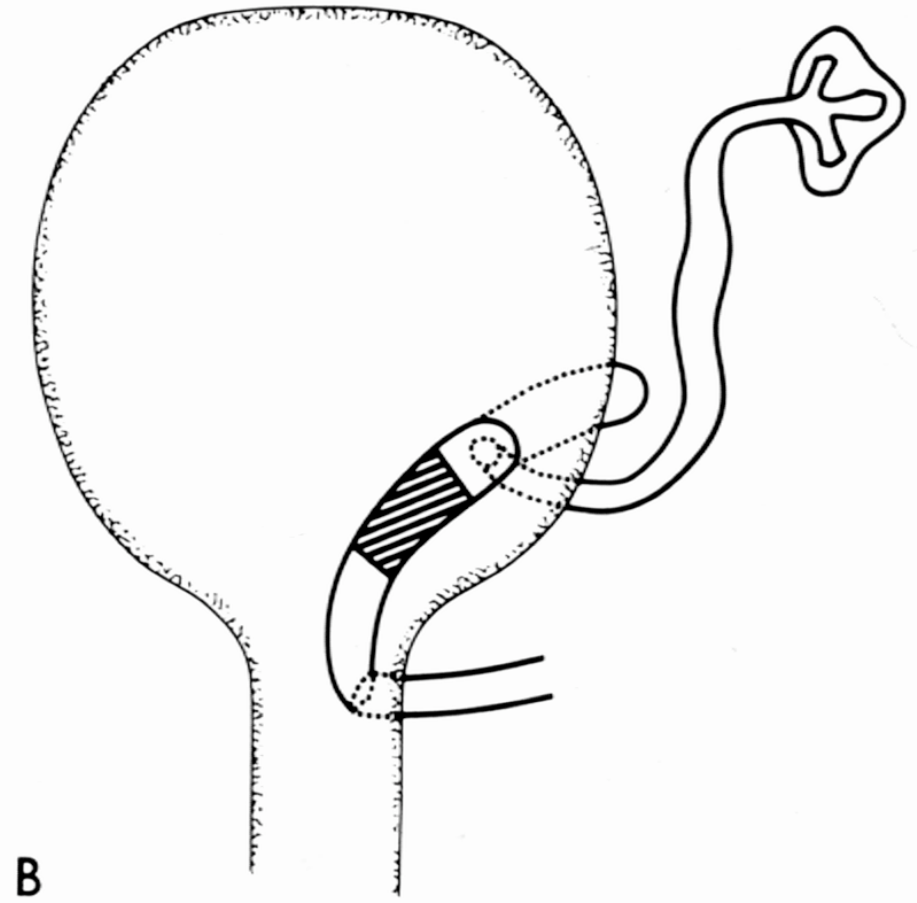
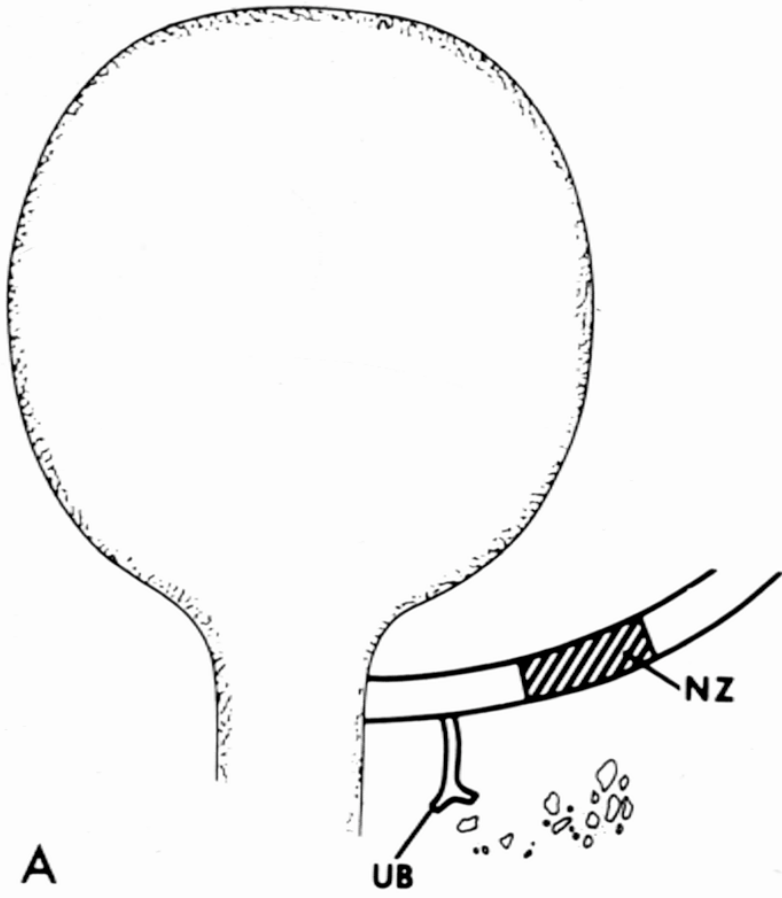
Splitting of bud

Duplication anomaly

Excess absorption of bud

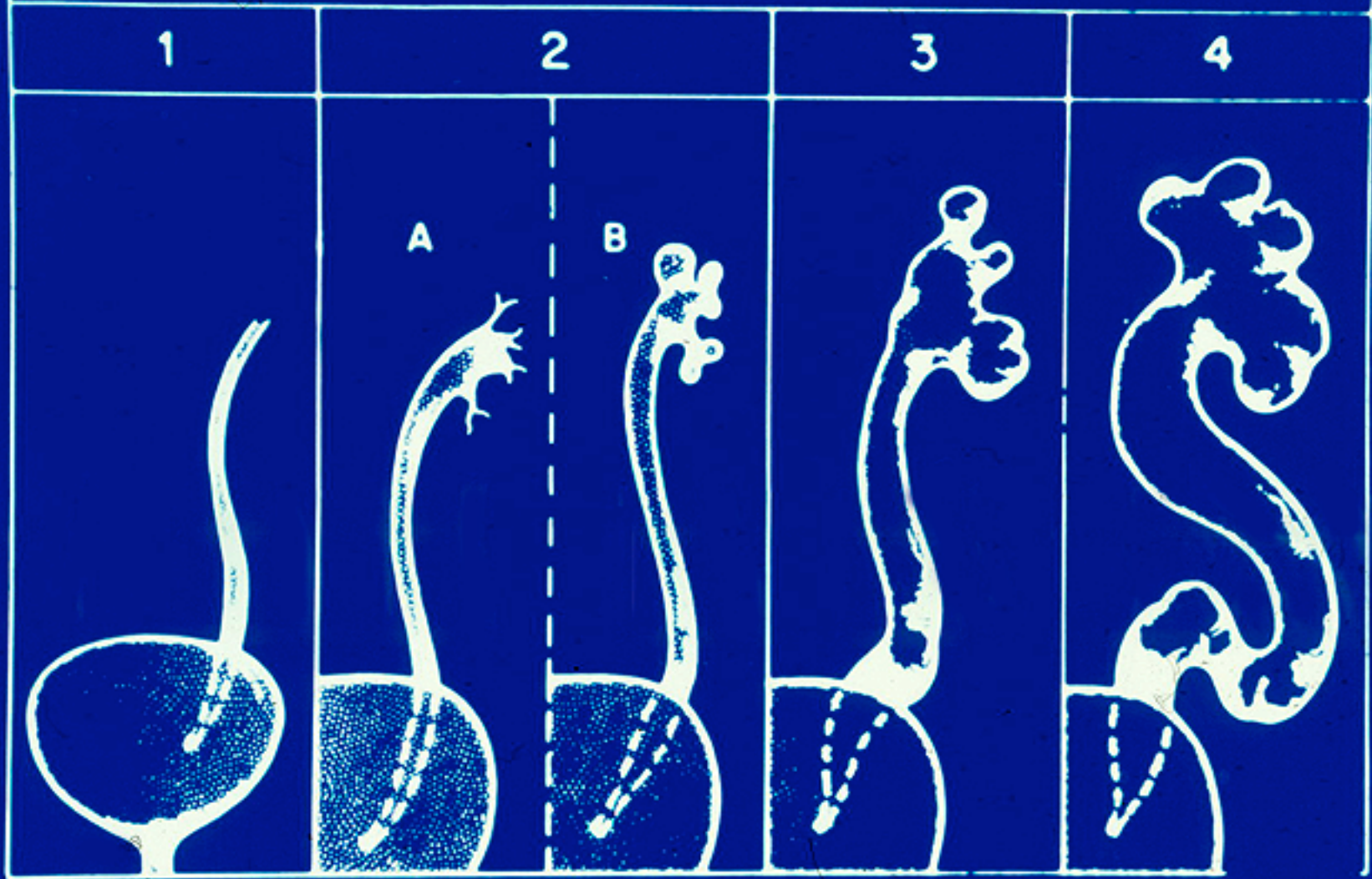
Refluxing orifice

Vesico Ureteral Reflux (VUR)





REFLUX GRADES



URETER
ONLY

1

MILD
DIL

2

MOD
DIL

3

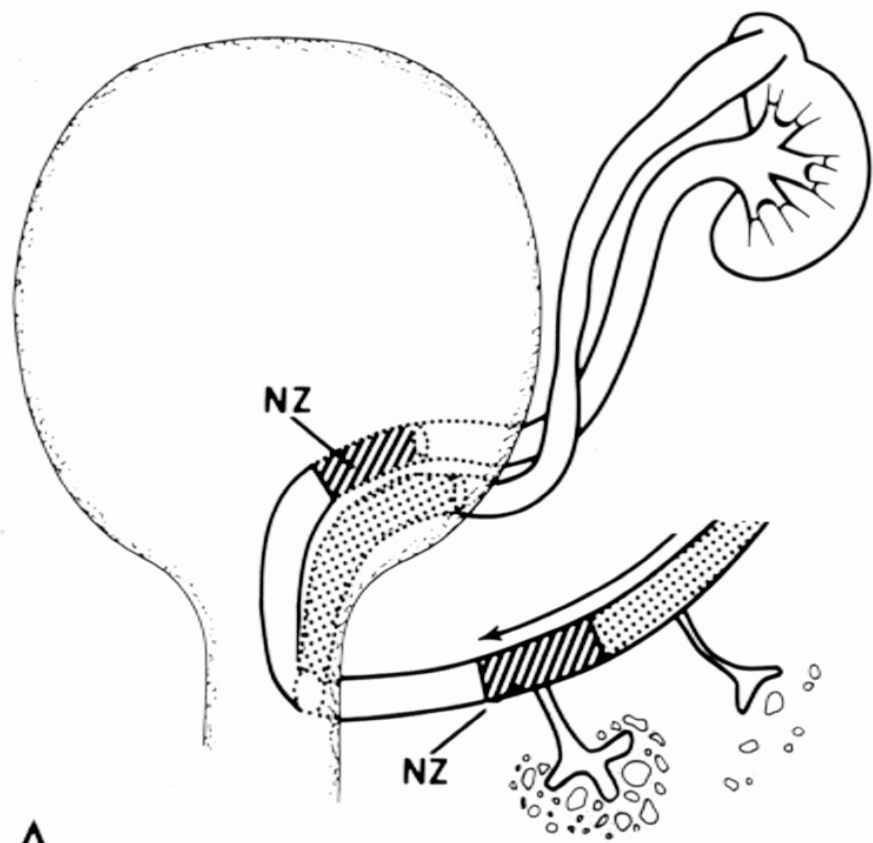
SEVERE

4

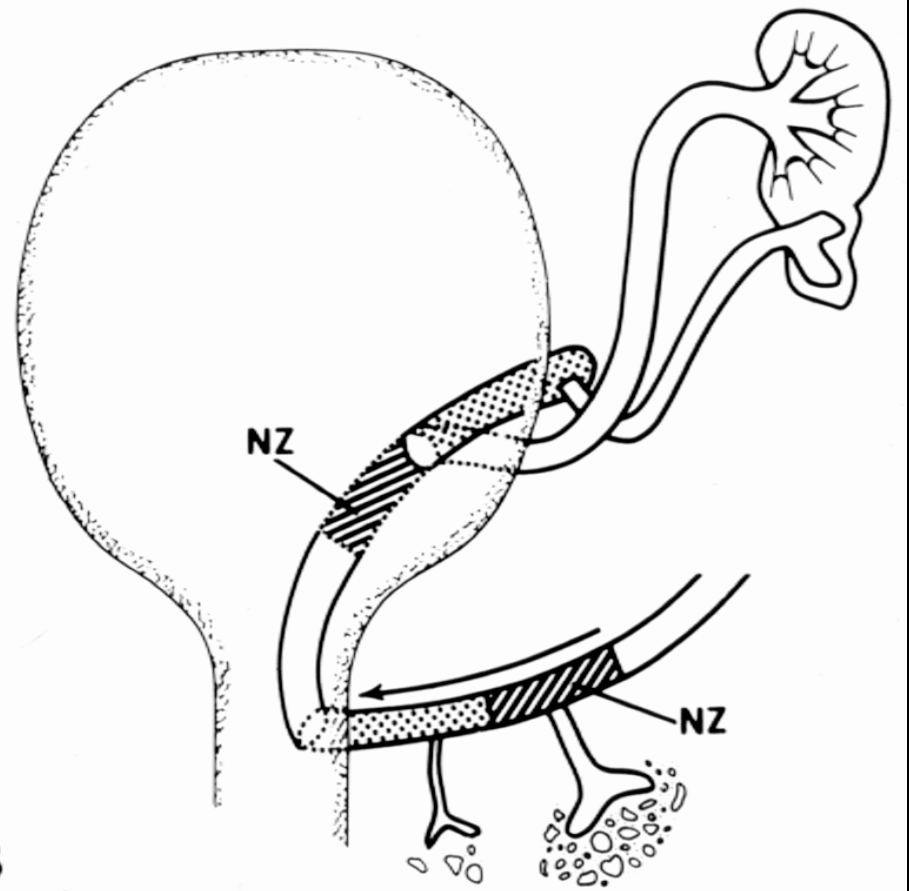
MASSIVE

5

Ureteral Duplication

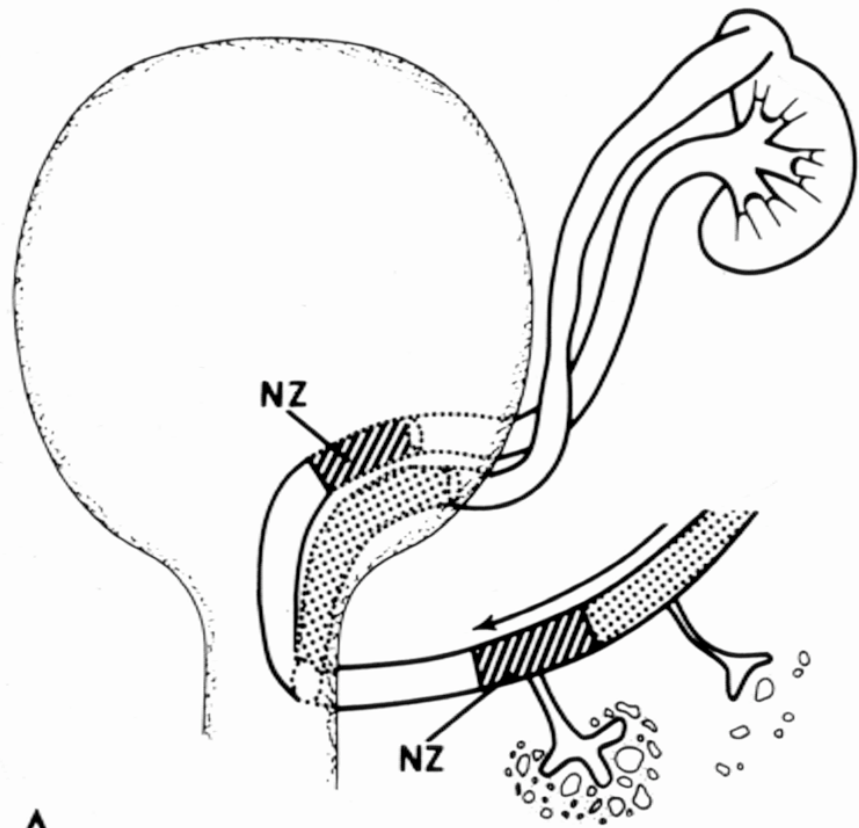


A

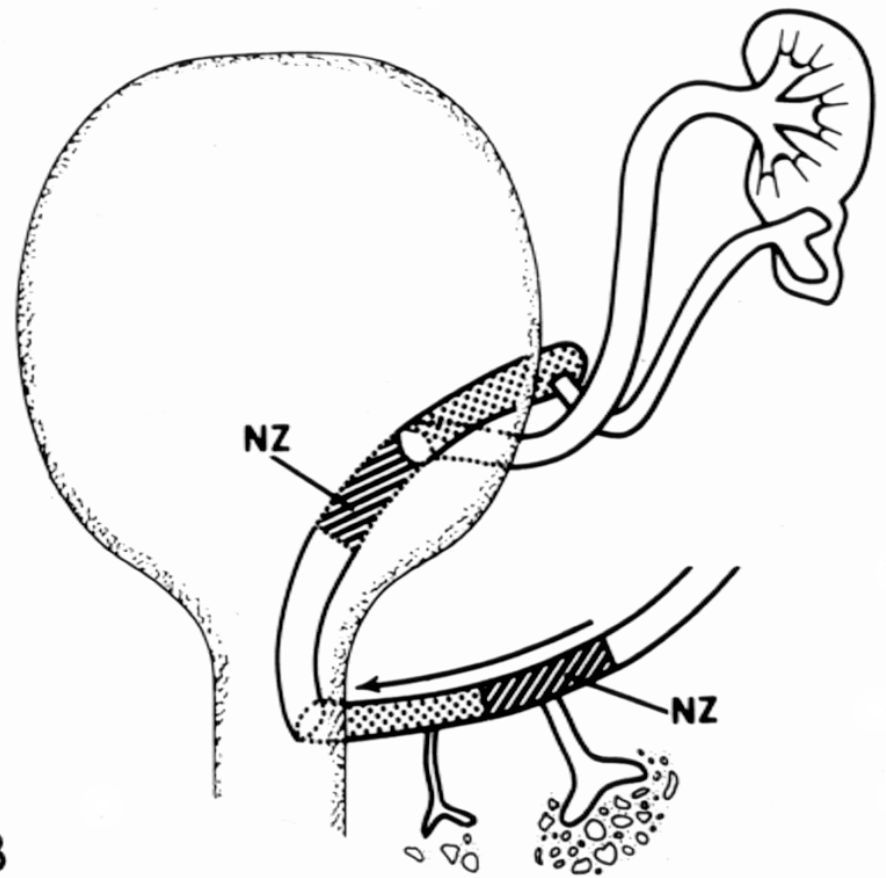


B

Ectopic Ureter

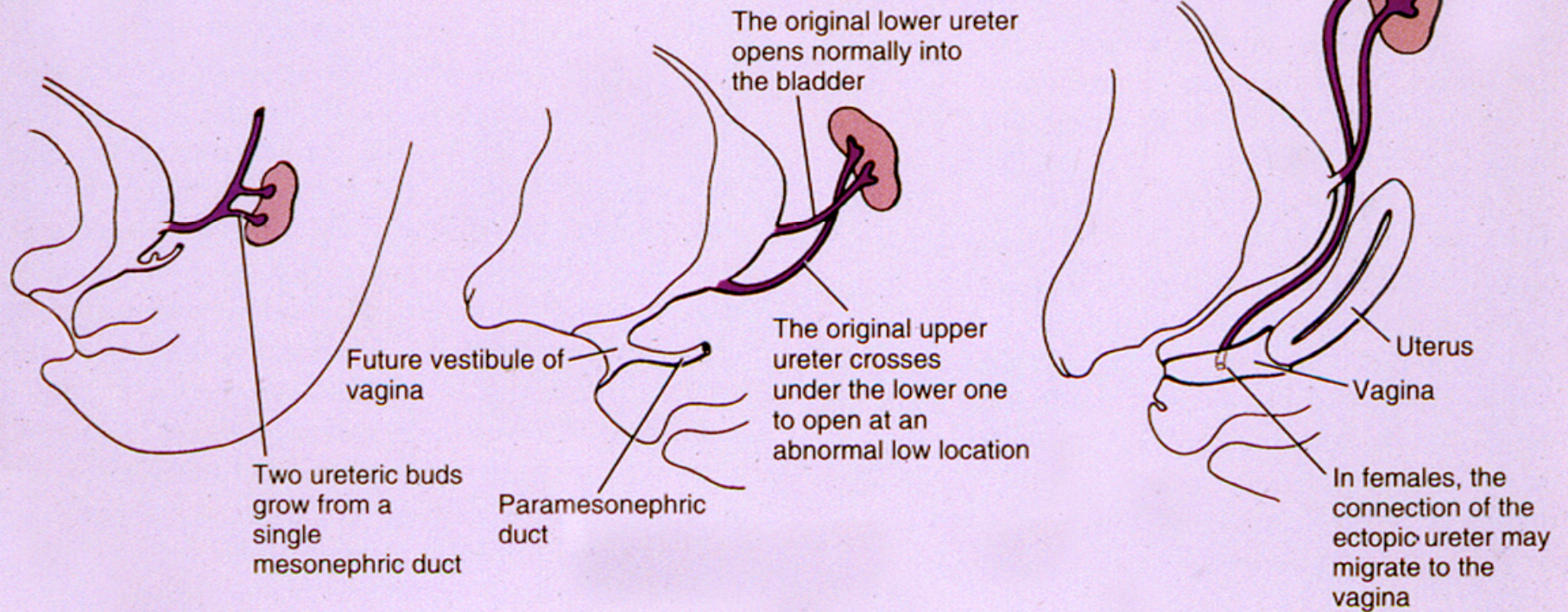


A



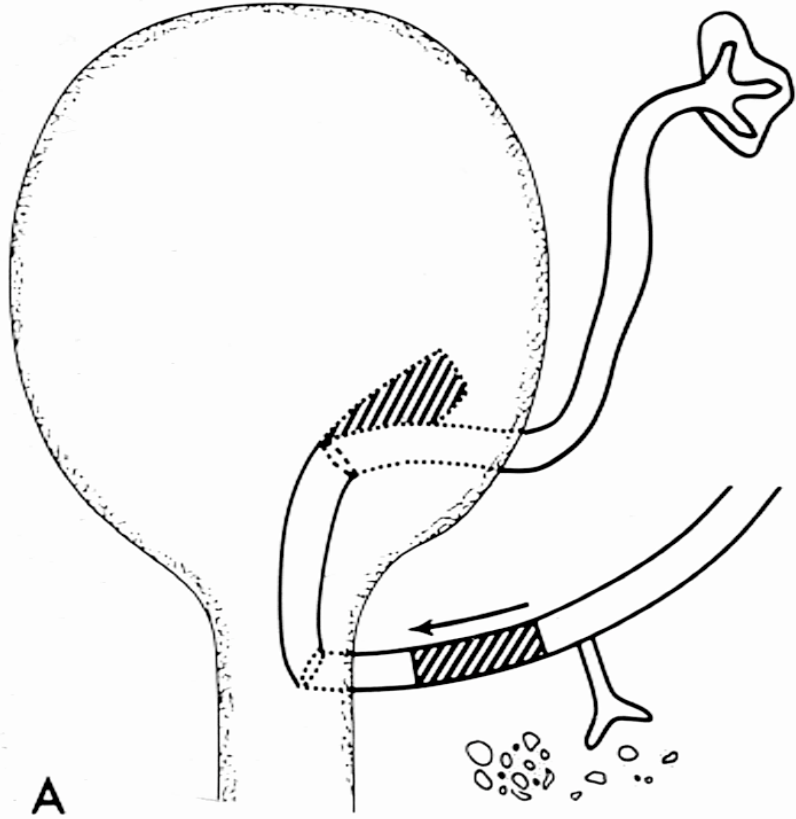
B

The ureters invert their positions as they exstrophy into the bladder (the Weigert-Meyer rule):

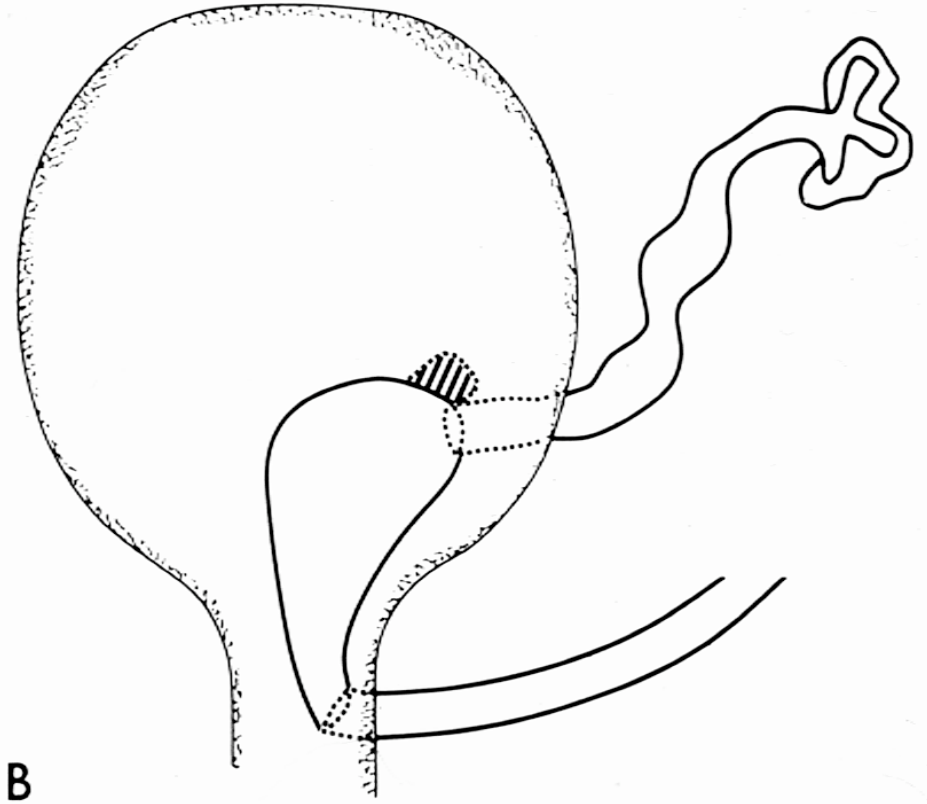




Ureterocele



A

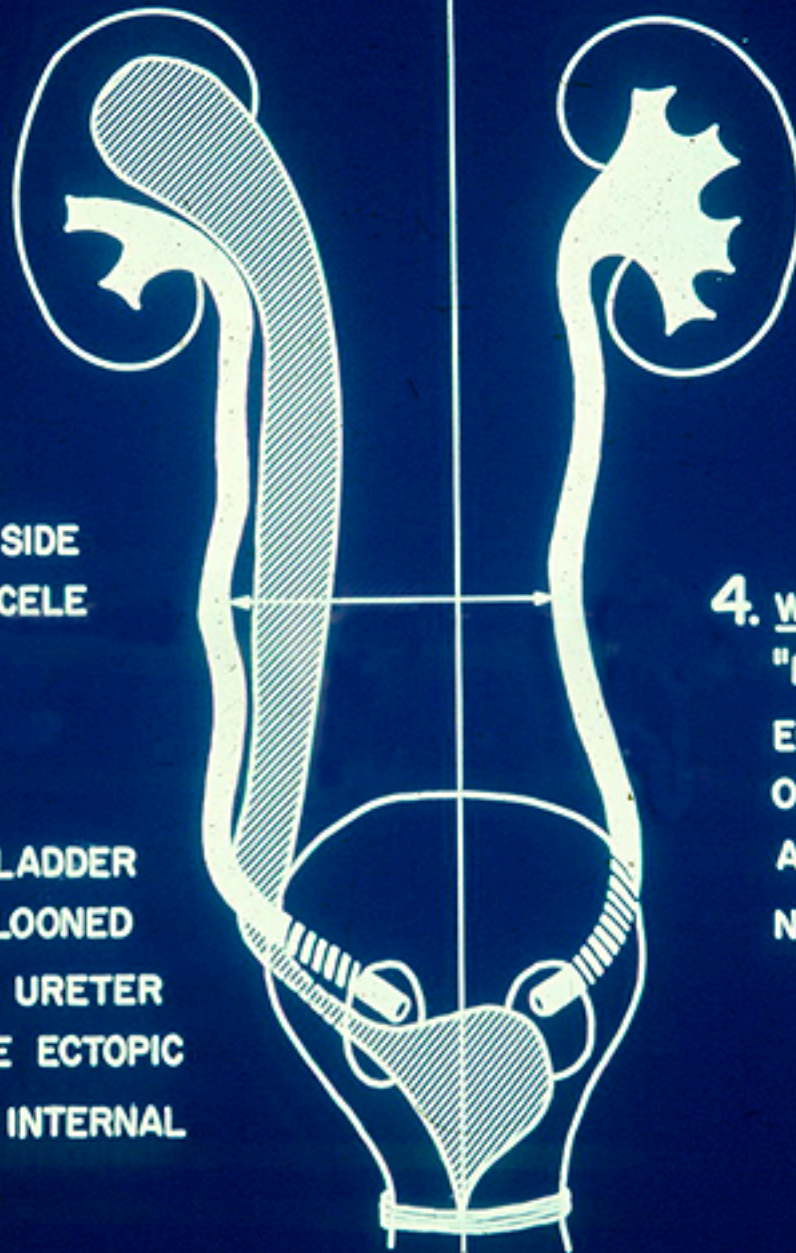


B

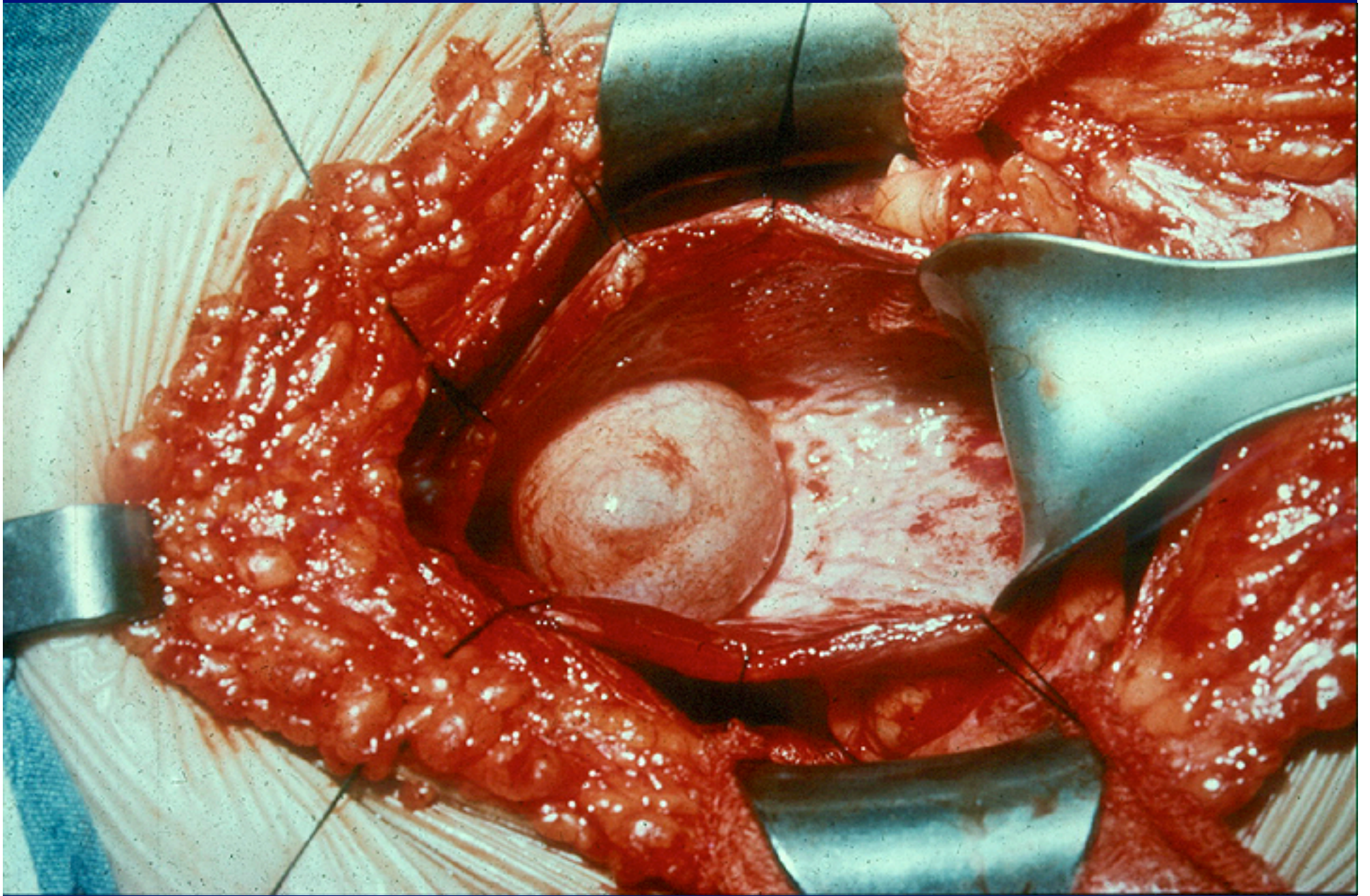
1. DROOP
OF
LOWER
CALYCES

2. LOWER POLE
URETER ON SIDE
OF URETEROCELE
DISPLACED
LATERALLY

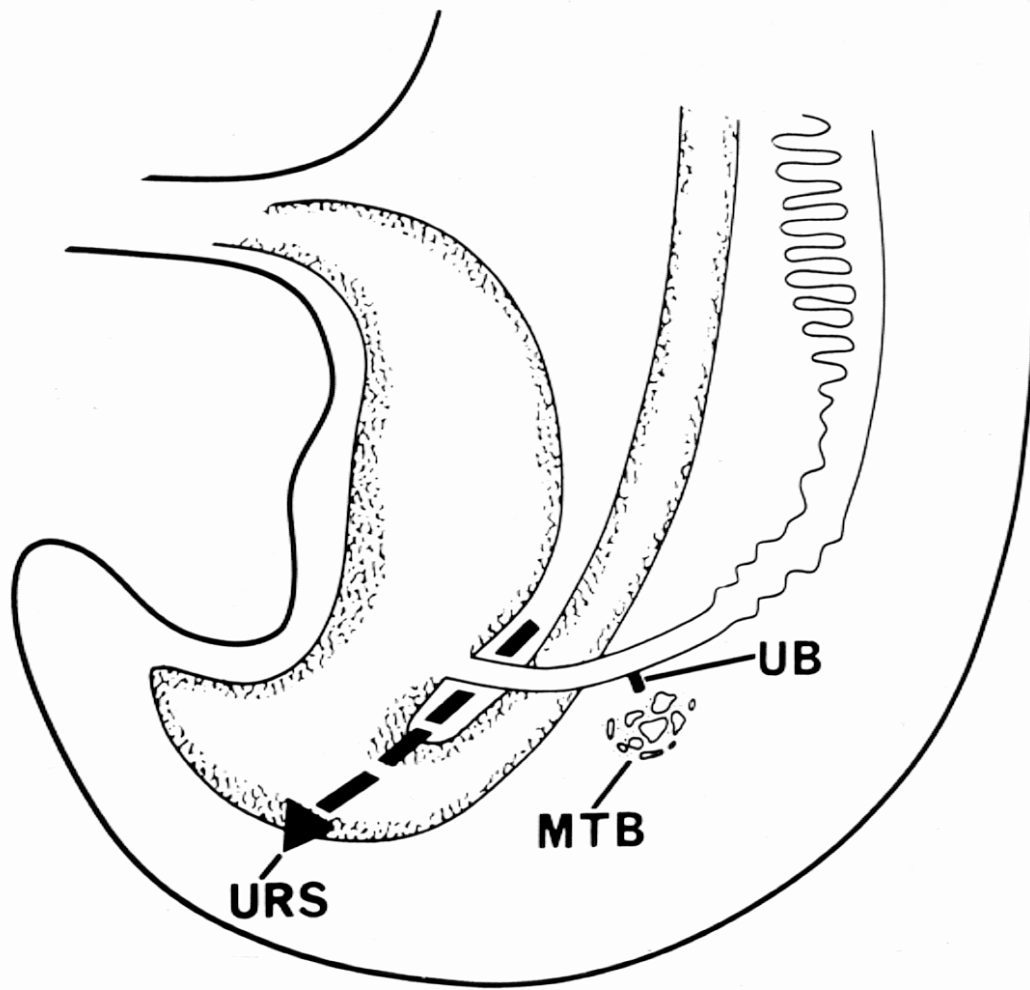
3. "MASS" IN BLADDER
DUE TO BALLOONED
UPPER POLE URETER
WITH ORIFICE ECTOPIC
IN ZONE OF INTERNAL
SPHINCTER

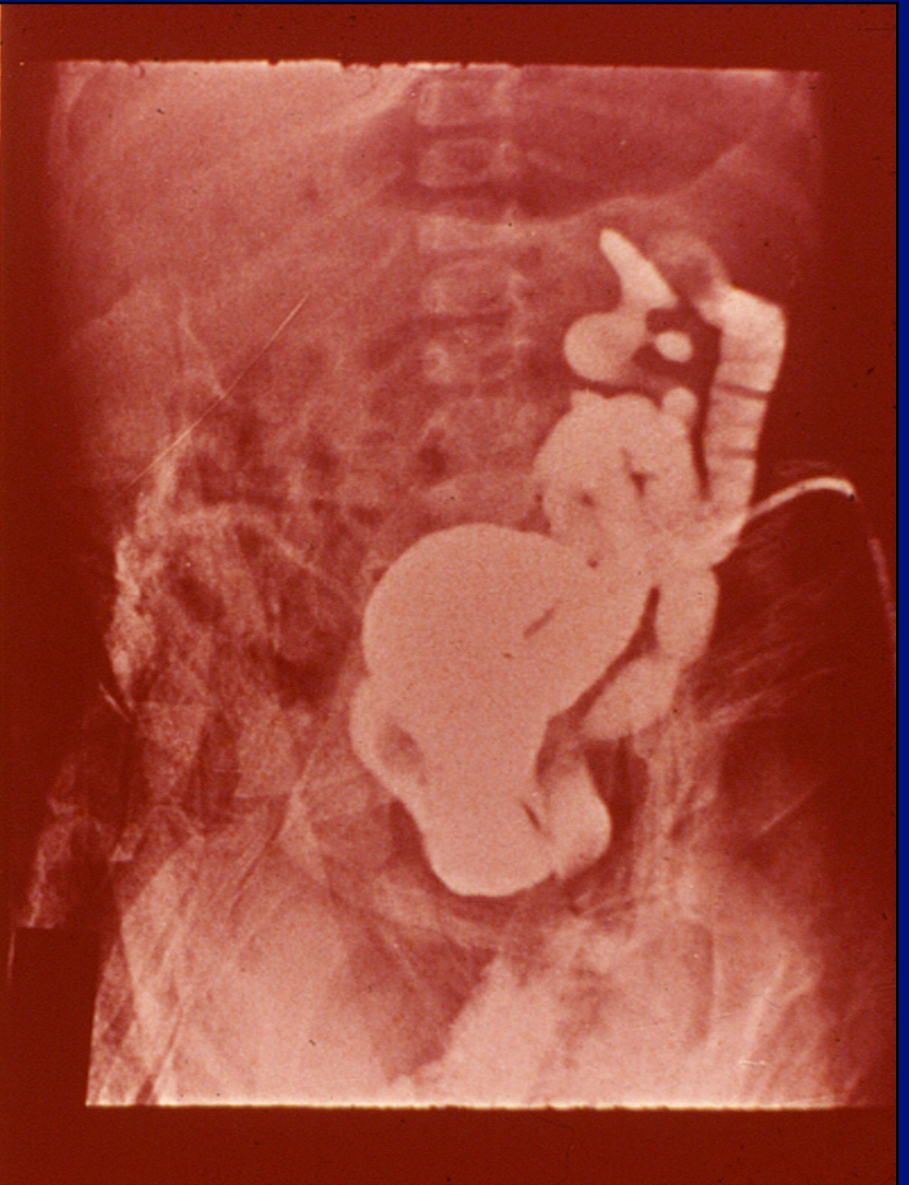


4. WEIGERT-MEYER
"RULE" —
ECTOPIC URETERAL
ORIFICE MEDIAL
AND CAUDAL TO
NORMAL ORIFICE

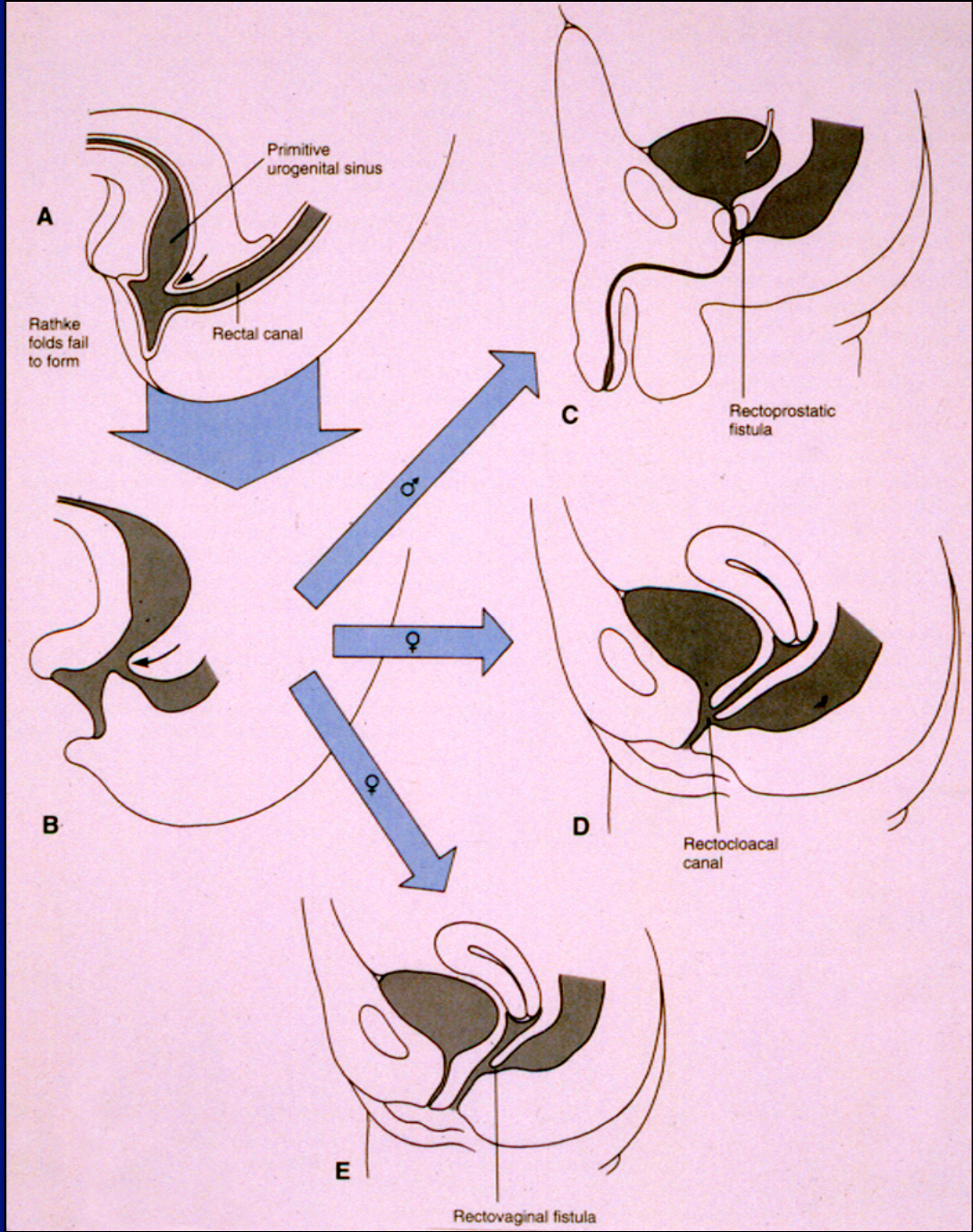


Cloacal Anomaly

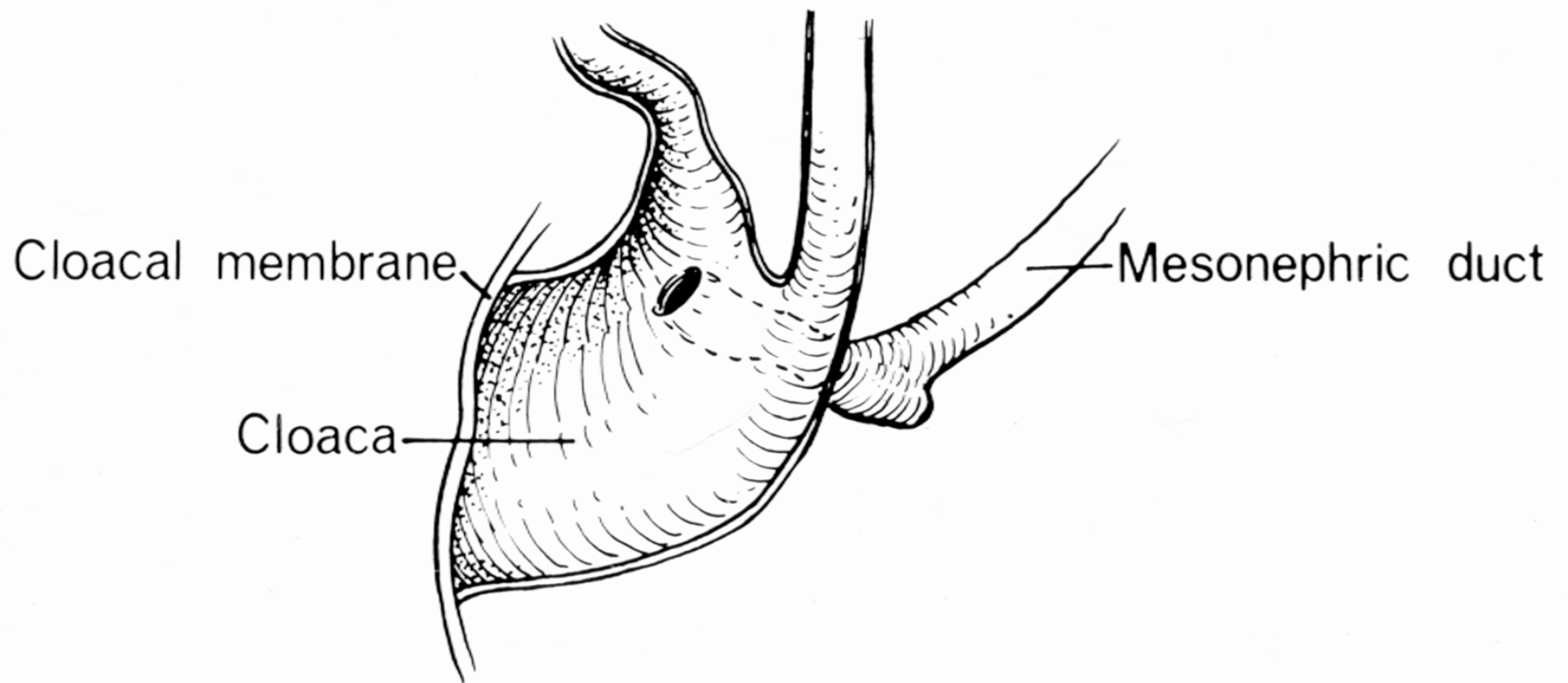




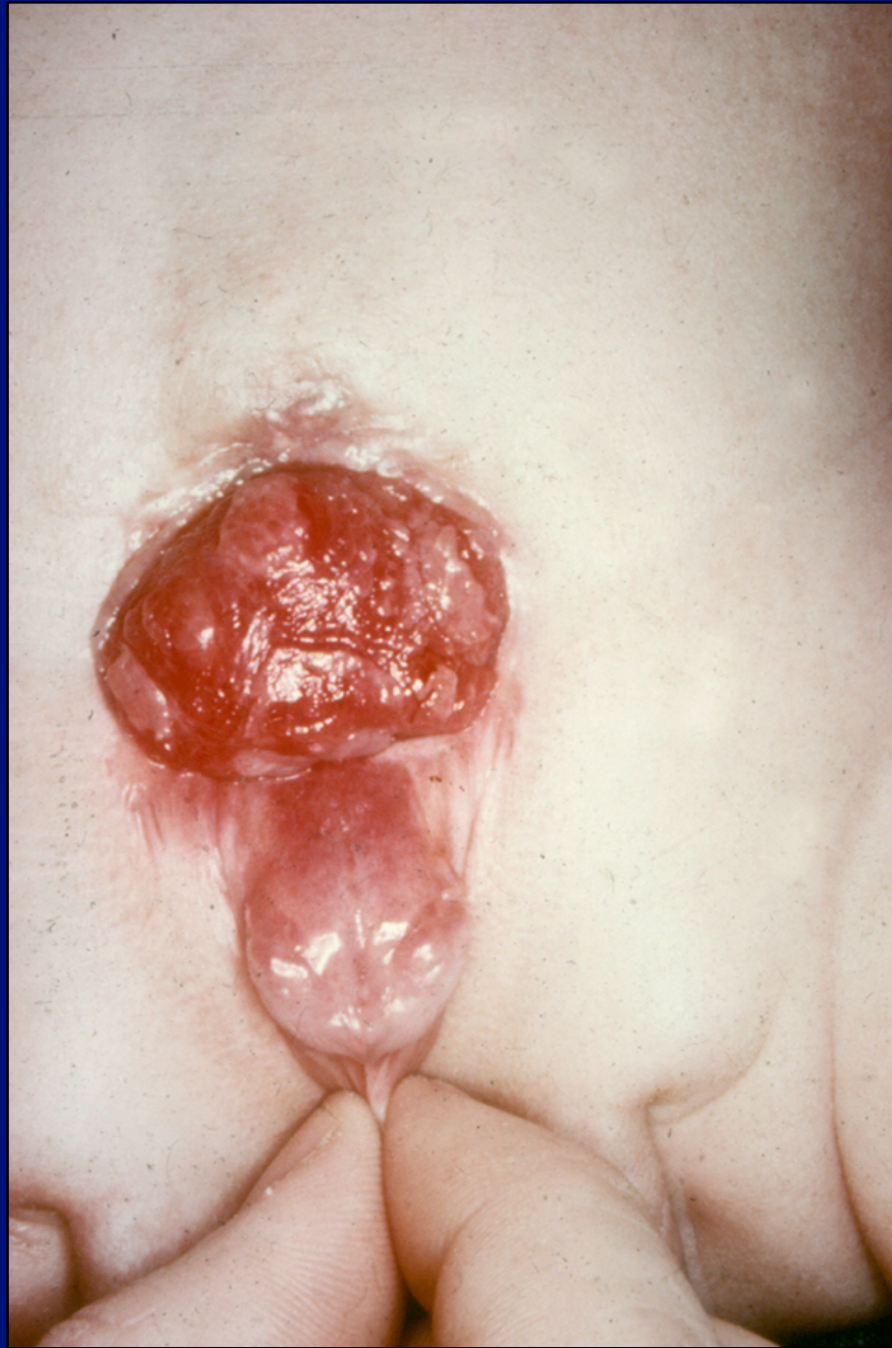
Imperforate Anus

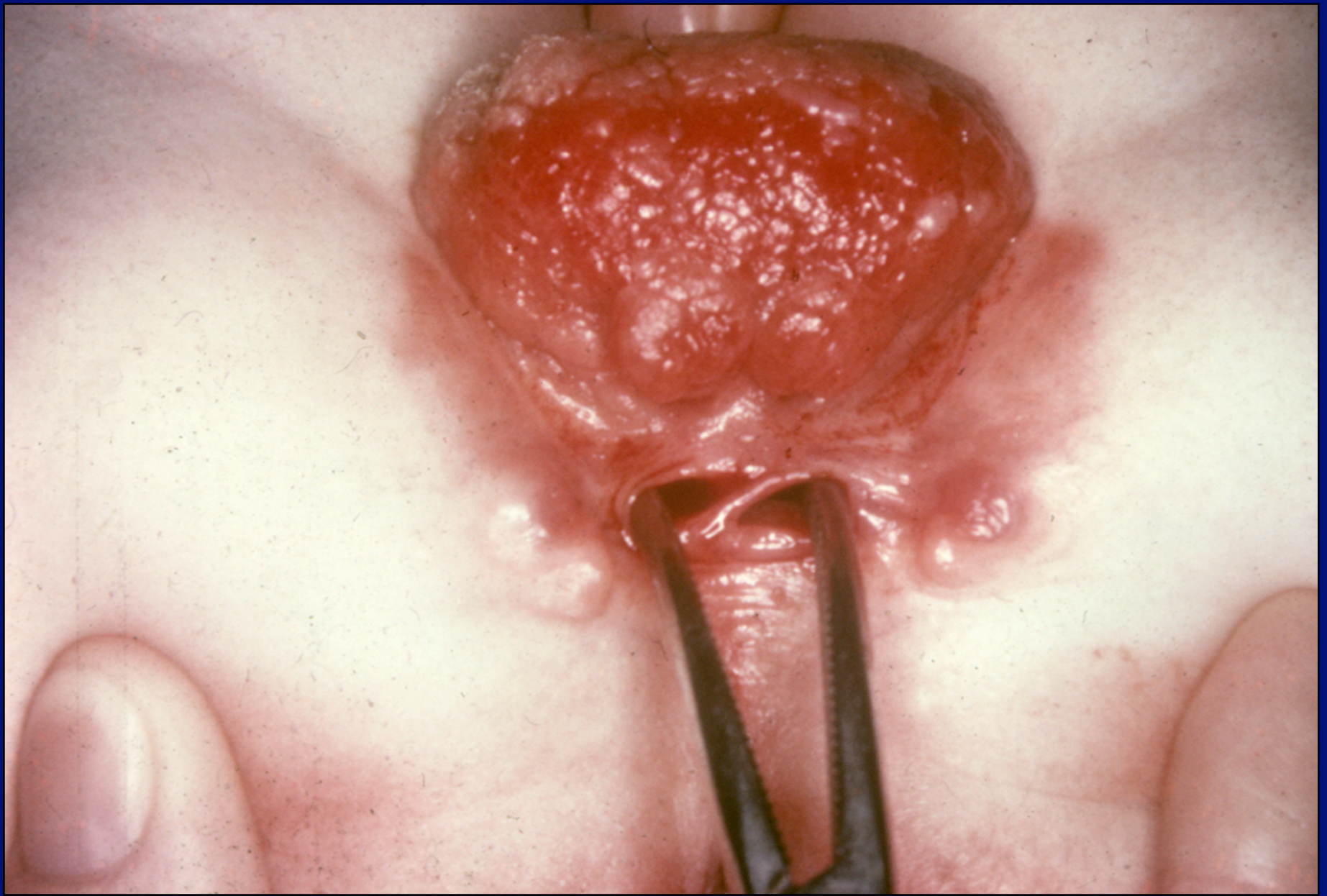


Exstrophy - Epispadias Complex

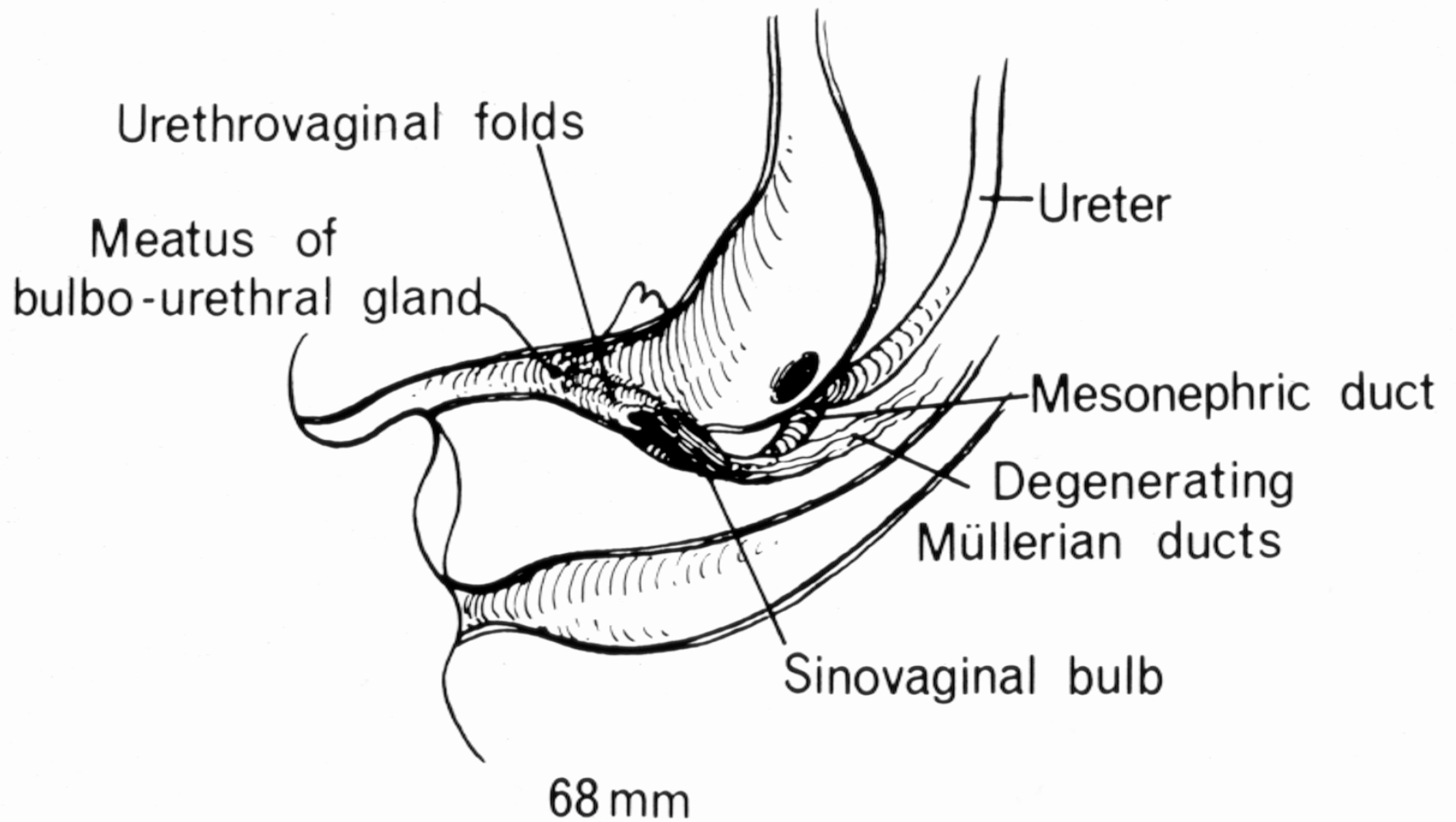


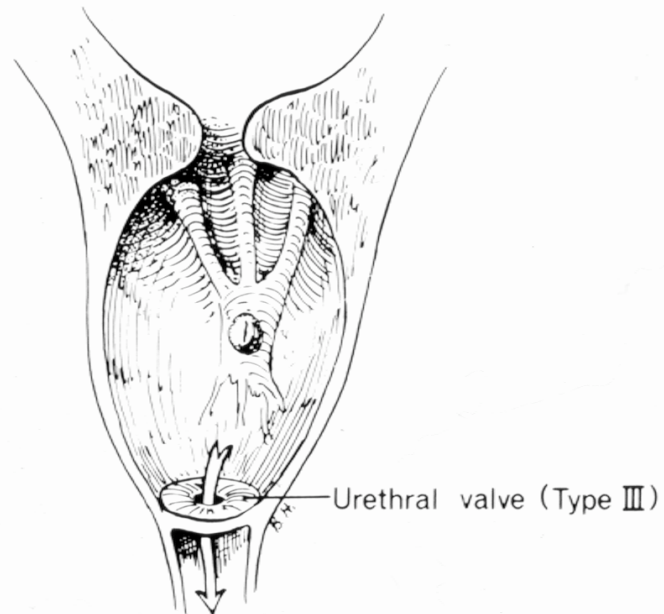
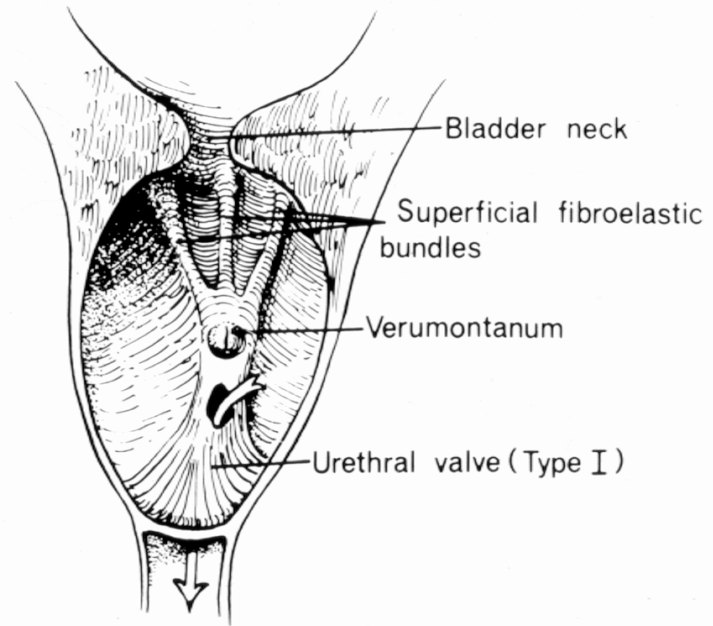
4 mm





Posterior Urethral Valves

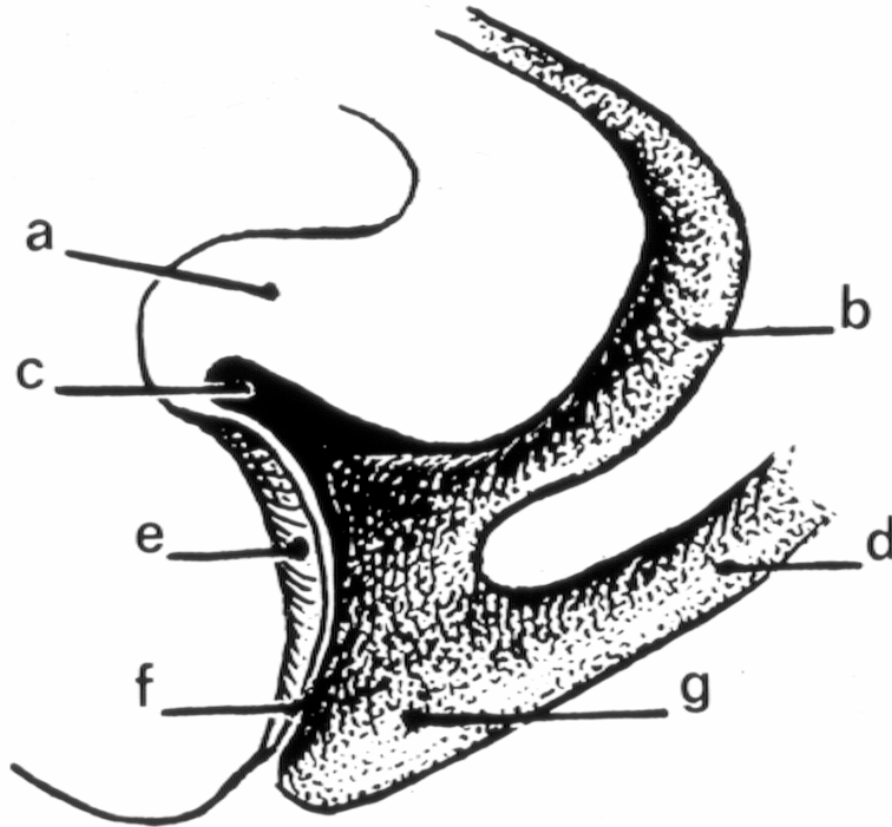




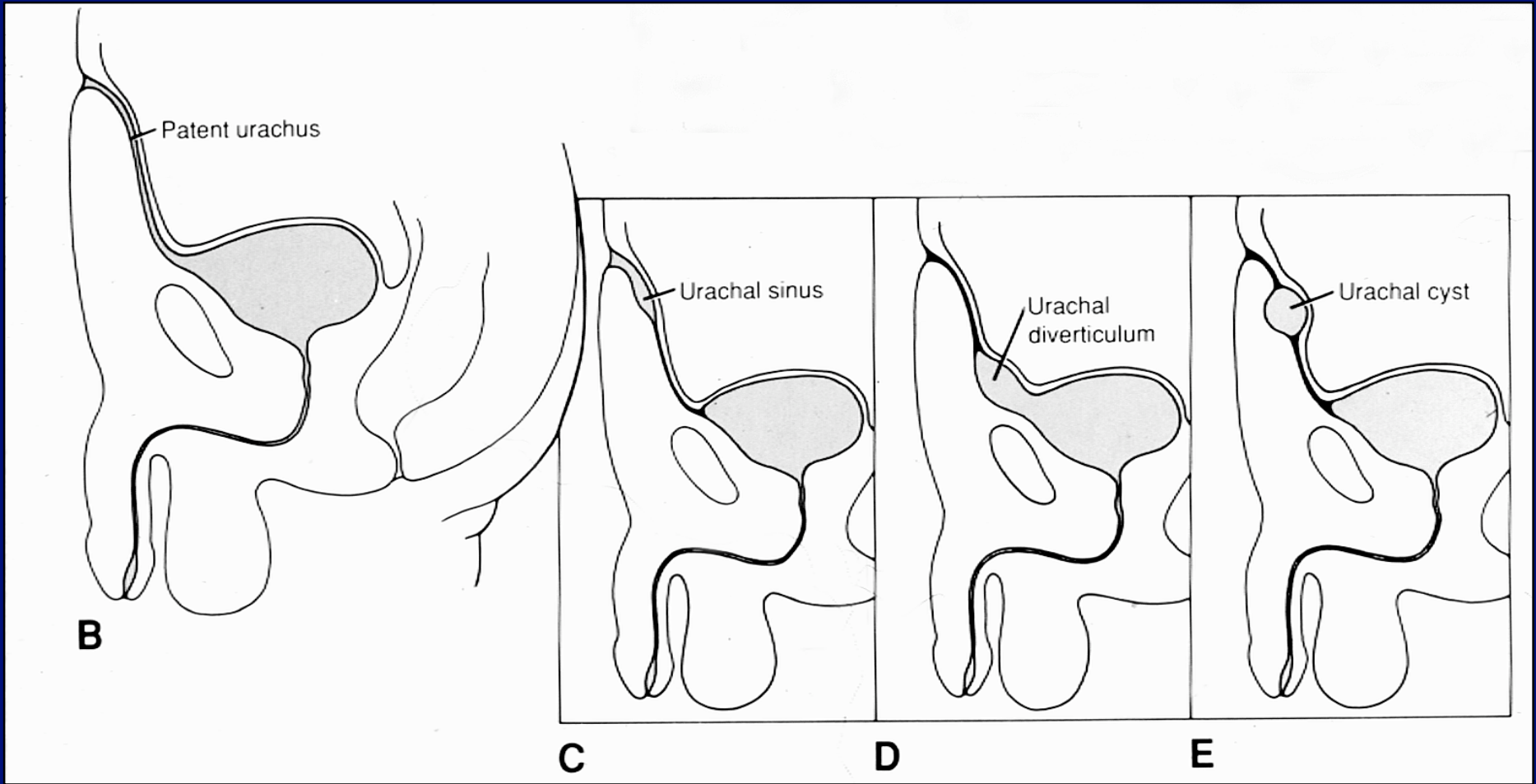


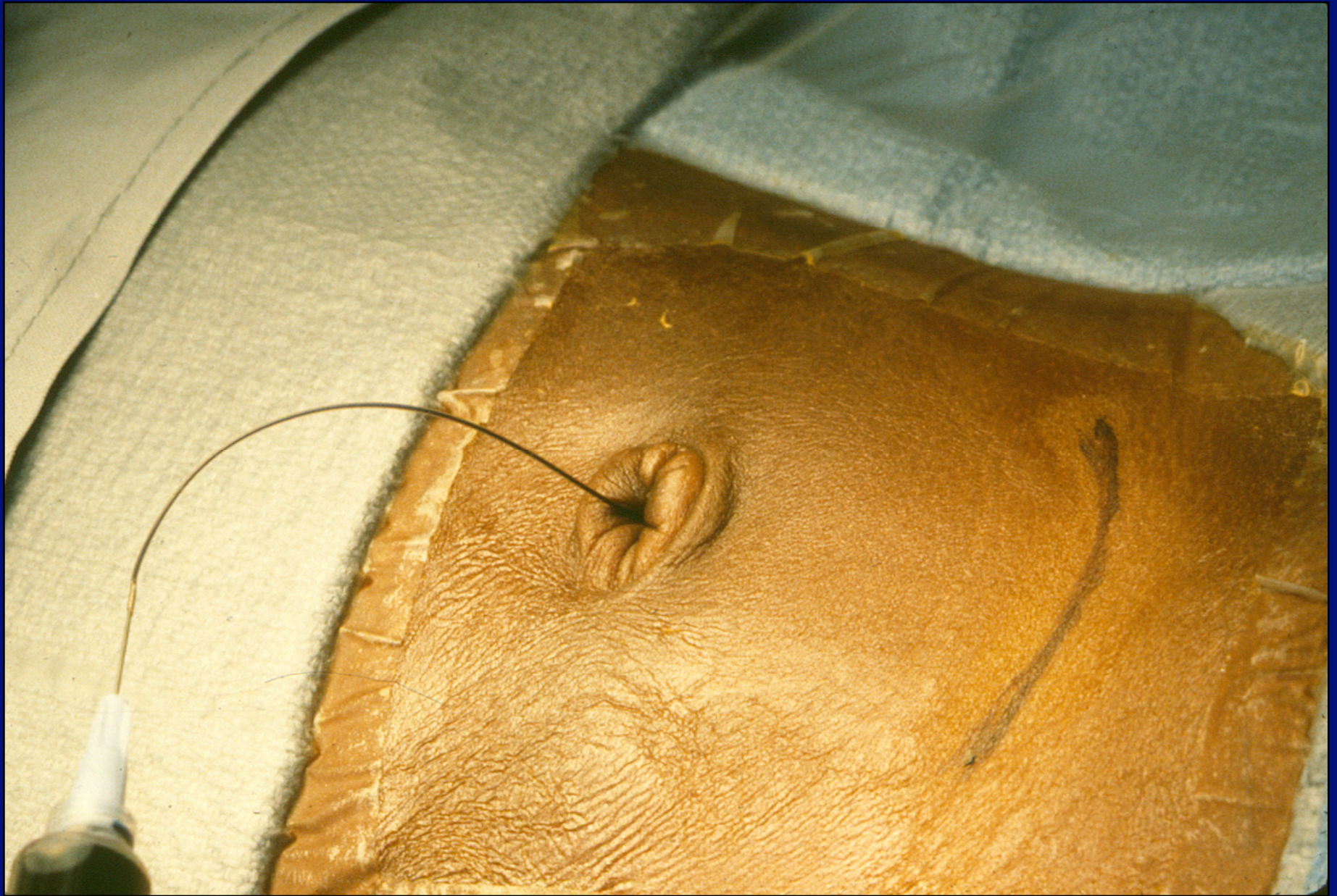


Urachal Abnormalities

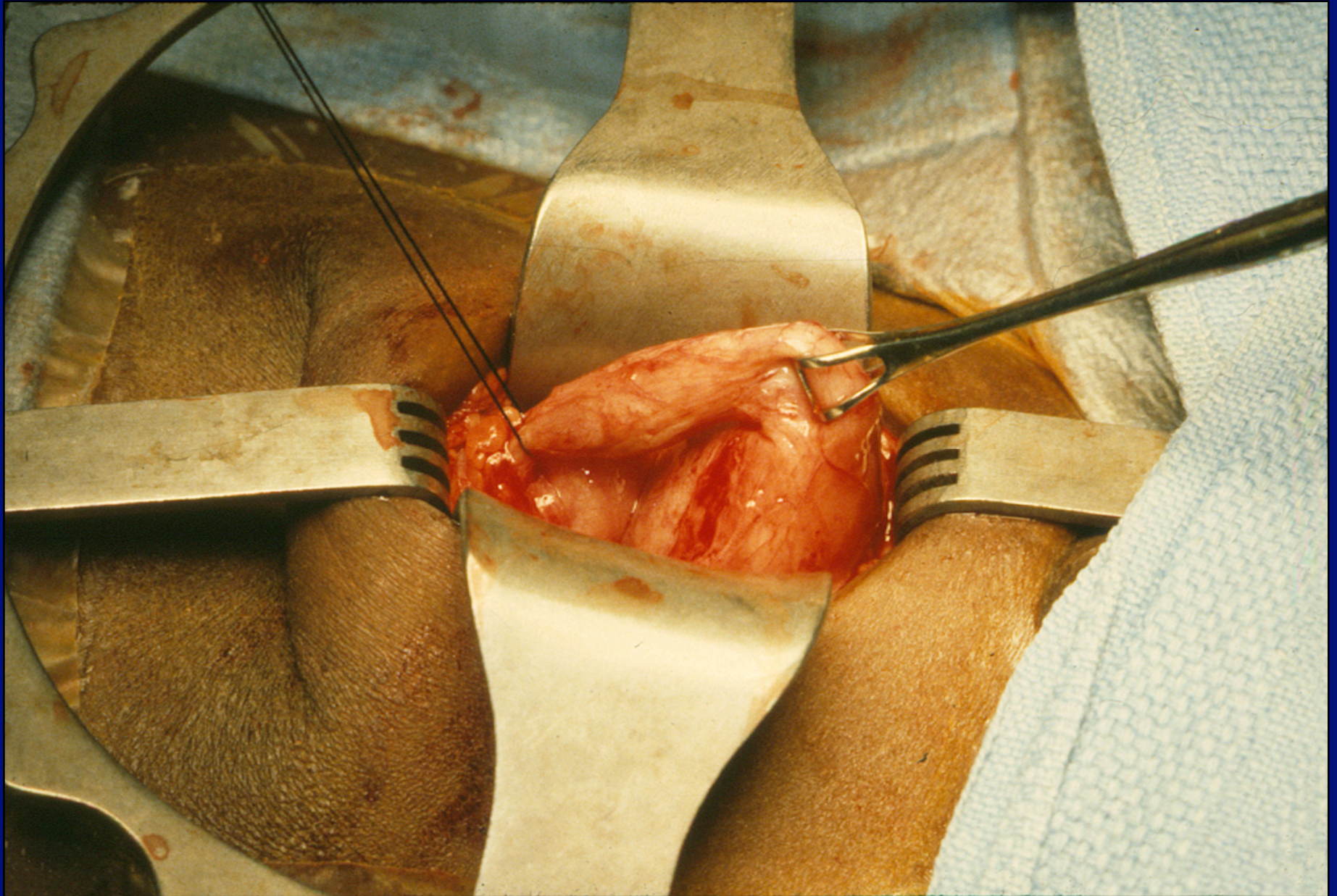


Cloacal endo-
derm penetrates inferior aspect
of genital tubercle to form ure-
thral plate: a, genital tubercle;
b, primitive urogenital sinus; c,
urethral plate; d, hindgut; e,
primitive urethral groove; f,
cloacal membrane; g, cloaca.

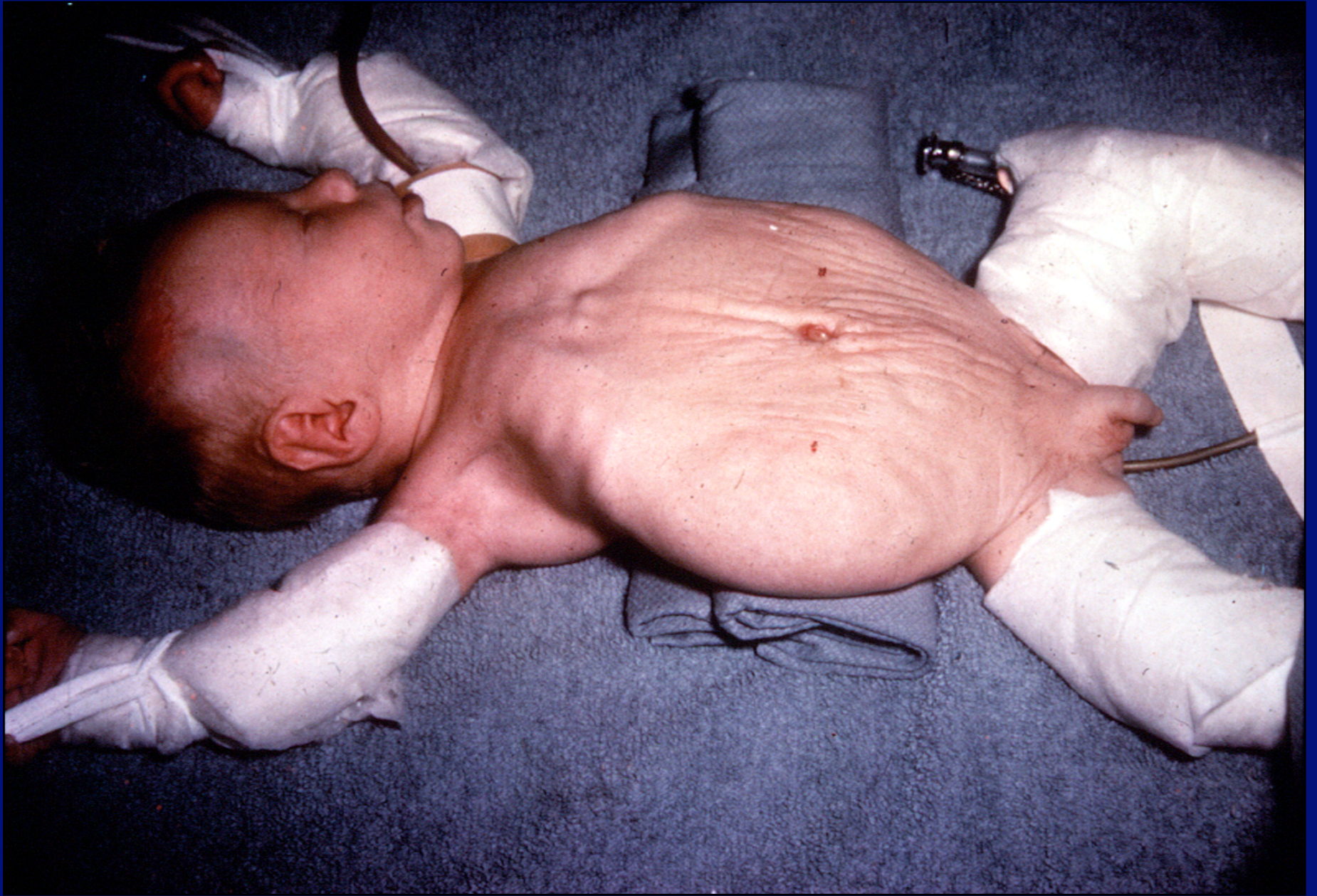








Prune Belly Syndrome



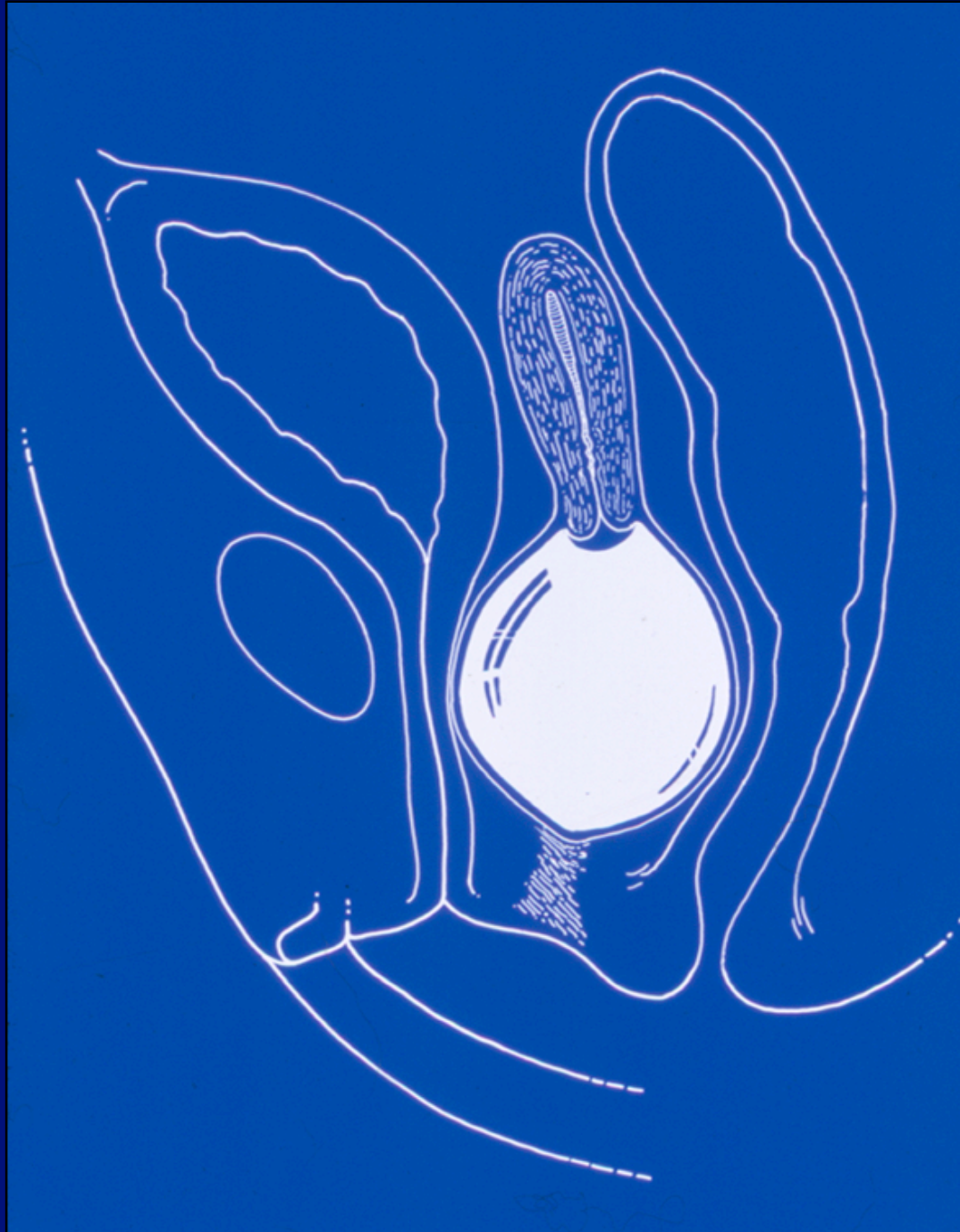
Anomalies of
Vaginal Plate Canalization



CANALIZATION OF
OF VAGINAL PLATE

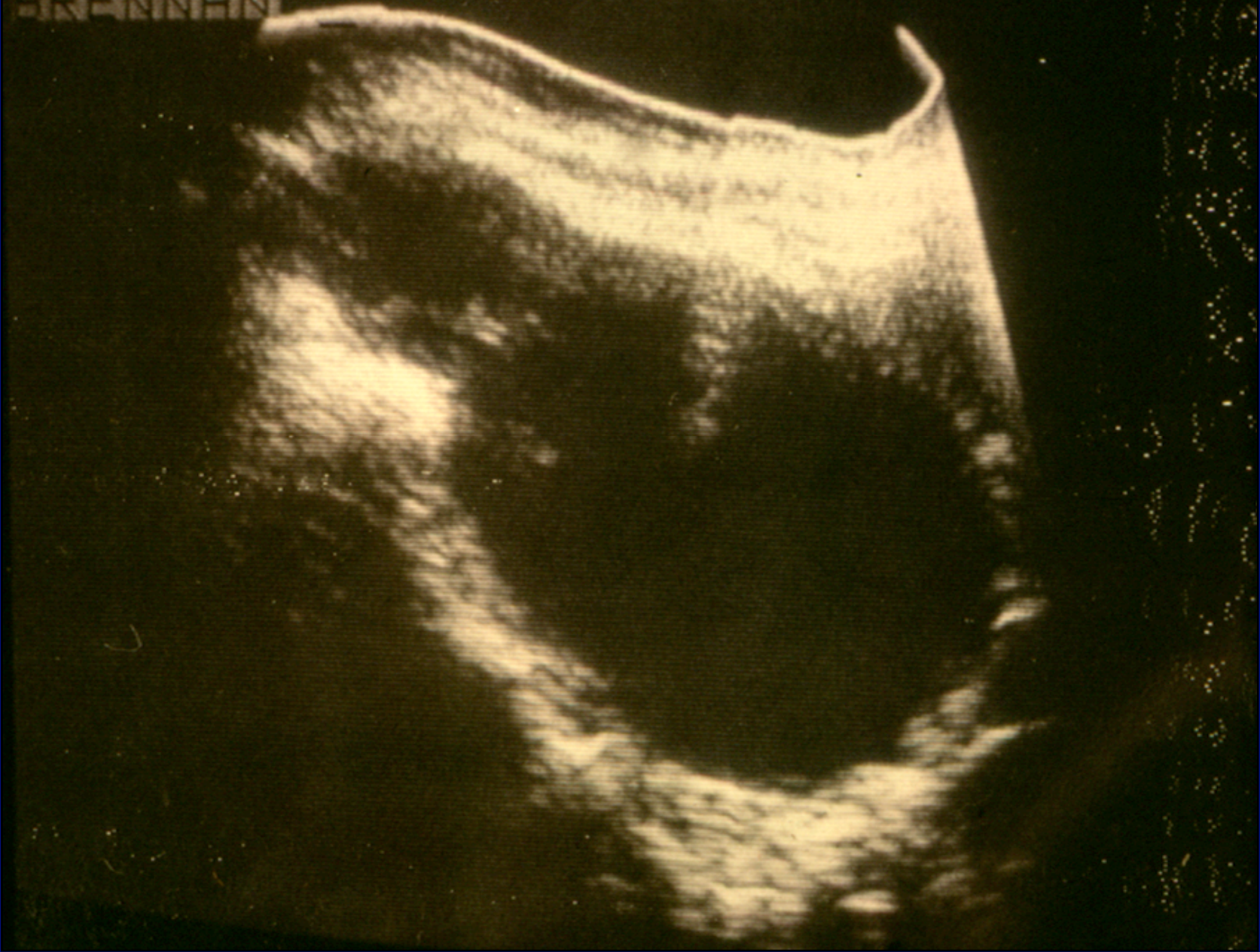




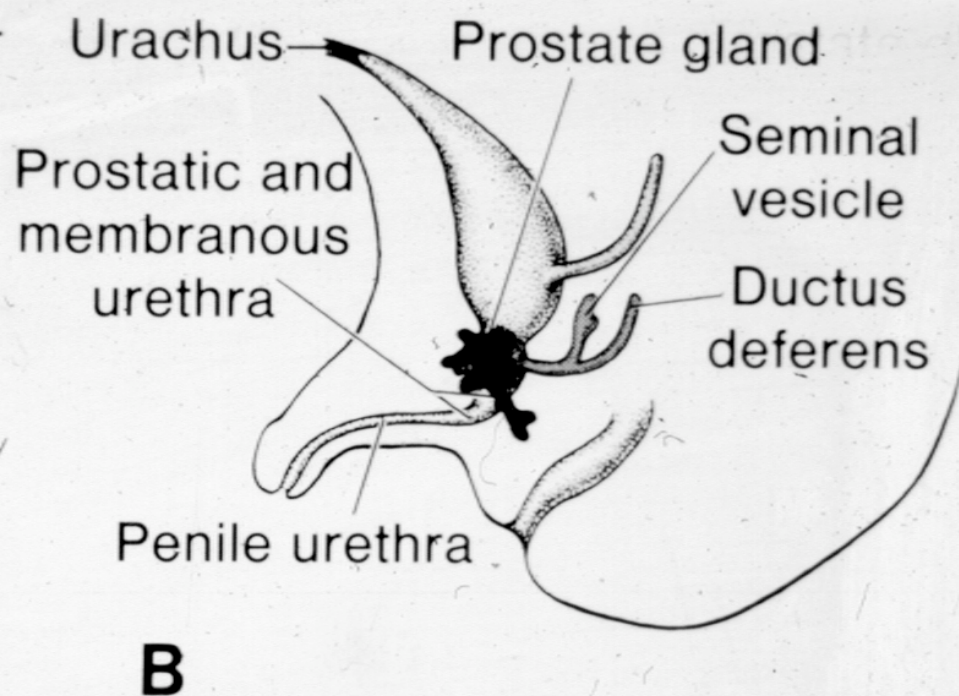
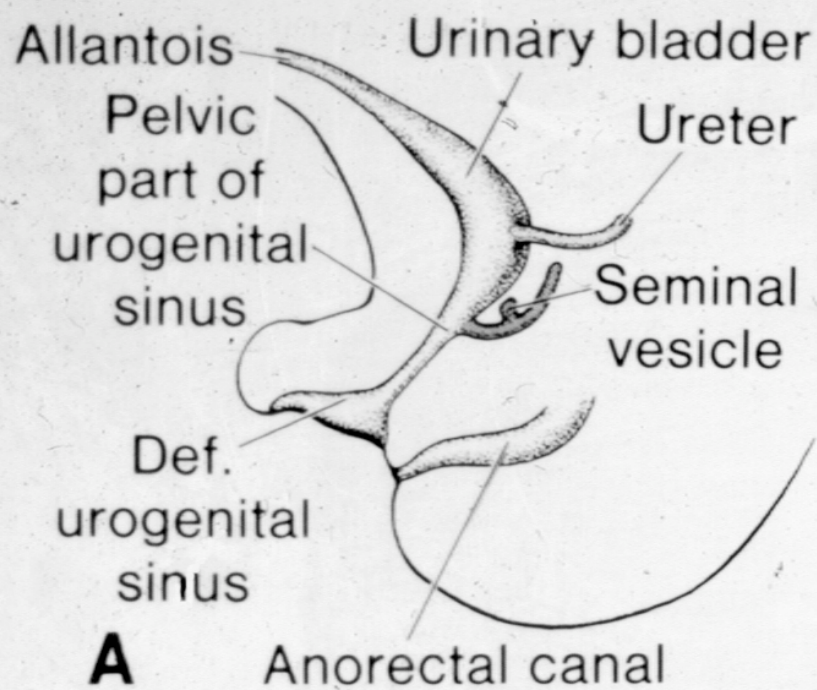


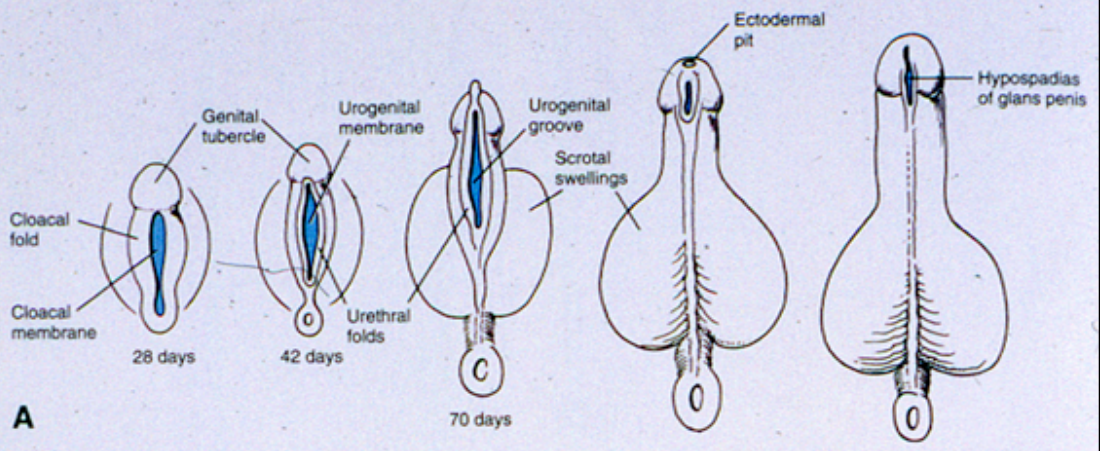


12-11-21

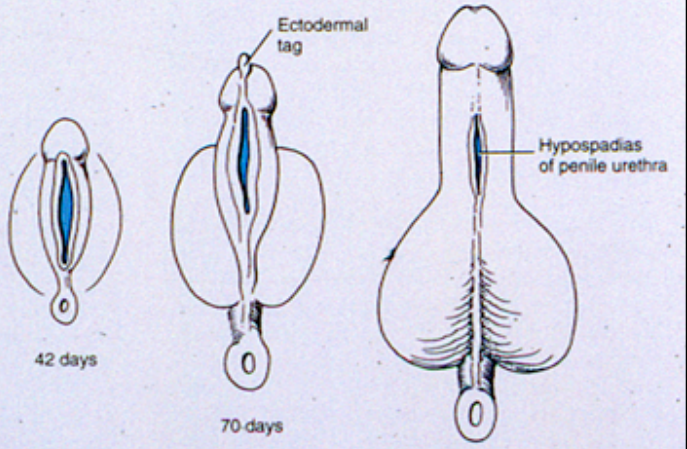
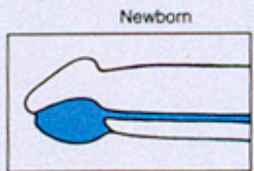


Anomalies of the Urethral Plate

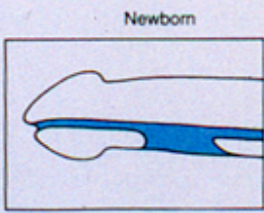




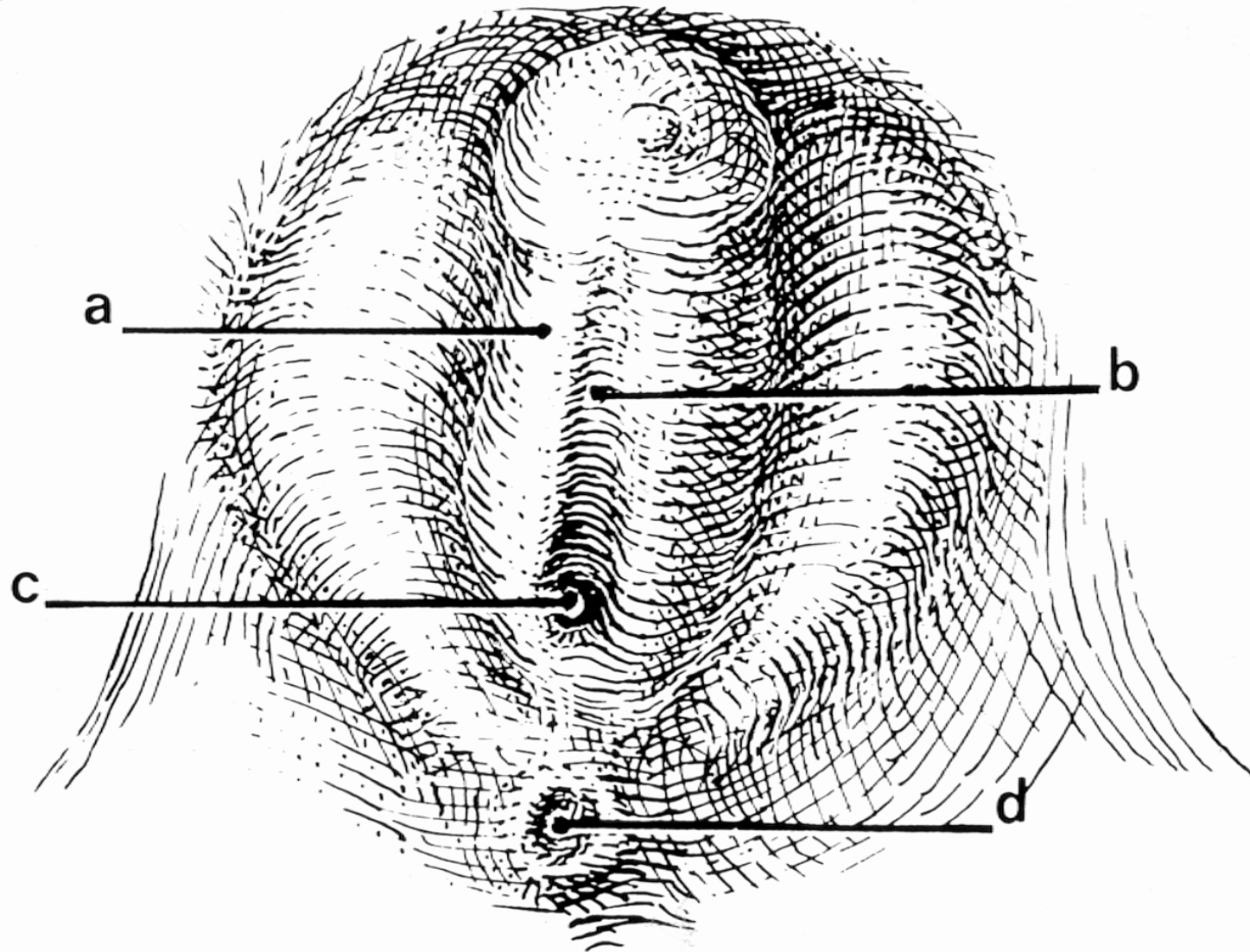
A



B



Hypospadias



External genitalia after cloacal division: a, urethral fold; b, primitive urethral groove; c, urogenital sinus opening; d, anal membrane.

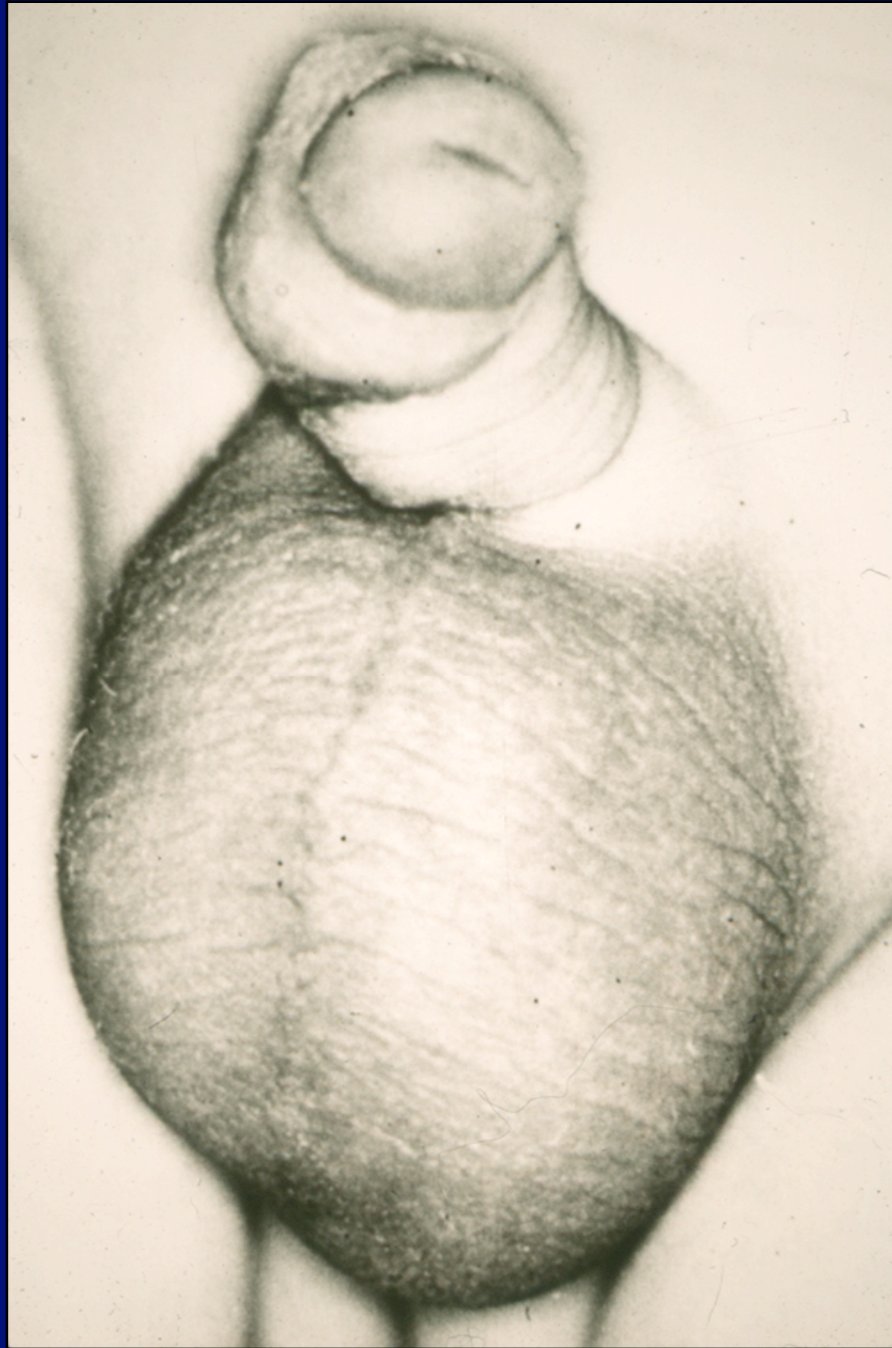








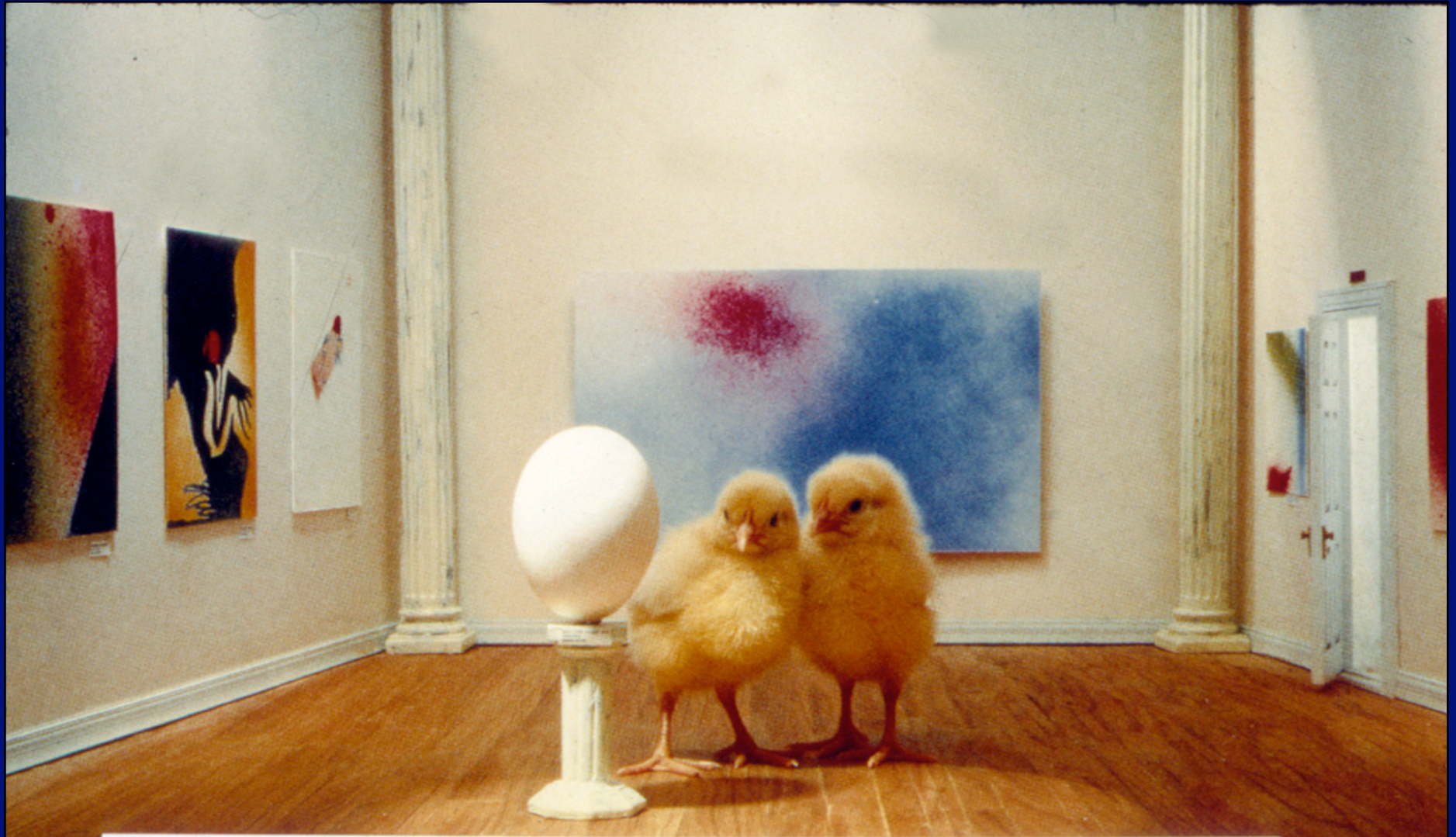
Other Genital Anomalies











WHICH CAME FIRST?

Urinary System Abnormalities

<u>ANOMALY</u>	<u>GESTATION</u>	<u>ETIOLOGY</u>
Bladder duplication	3 weeks	Notochord Duplication
Agenesis of Vas and Seminal Vesicles	3 - 4 weeks	Mesonephric duct abnormality
Agenesis of penis	4 weeks	Genital tubercle defect
Obstructive megaureter	4 weeks	Mesonephric duct abnormality
Uteral duplication	5 weeks	Ureteral bud defect

Urinary System Abnormalities

<u>ANOMALY</u>	<u>GESTATION</u>	<u>ETOLOGY</u>
Exstrophy- Epispadias	5 - 6 weeks	Cloacal membrane anomaly
Ectopic Ureter	5 - 6 weeks	Ureteral bud defect
Imperforate Anus	6 weeks	Failure of urorectal septum
Ureterocele	7 - 8 weeks	Persistence of Chwalle's membrane
Posterior urethral valves	8 weeks	Persistence of Muller's tubercle

Urinary System Abnormalities

<u>ANOMALY</u>	<u>GESTATION</u>	<u>ETOLOGY</u>
Hypospadias	8 weeks	Failure of urethral groove
Urachal Abnormalities	9 weeks	Persistent allantois
Prune Belly Syndrome	12 weeks	Secondary failure of mesoderm
Undescended Testis	7 months	Failure of end organ response