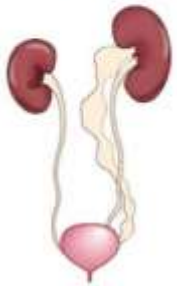
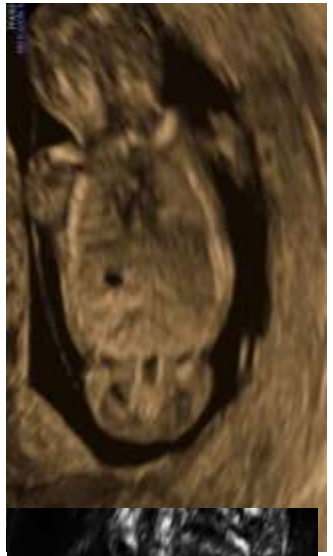


Importance of fetal scan in CAKUT management



Dr.Indrani Suresh

Fetal Medicine Consultant

Mediscan Systems

Nephkids 14 th sept 2025

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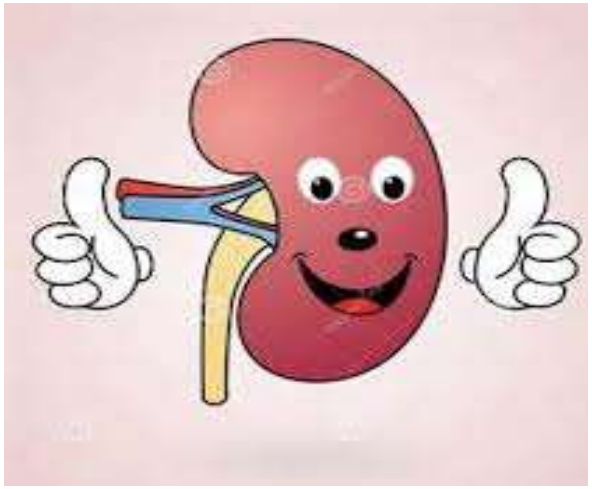
THANK YOU
Nephkids team .



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Congenital abnormalities of the kidney and urinary tract - CAKUT

- 20–30% of all major birth defects
- 1 in 500 live births.
- 1/3 of renal anomalies are detected by prenatal USG
- About 60% of children having surgery for renal or urinary tract problems in their first five years of life are identified by prenatal ultrasound

Semin Fetal Neonatal Med. 2017 April ; 22(2): 58–66. doi:10.1016/j.siny.2017.01.001.

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Plan of this talk

- Fetal renal system assessment
- Anomalies of renal system
- Approach to single kidney with preserved renal function
- Role of fetal intervention
- Genetic testing
- conclusion

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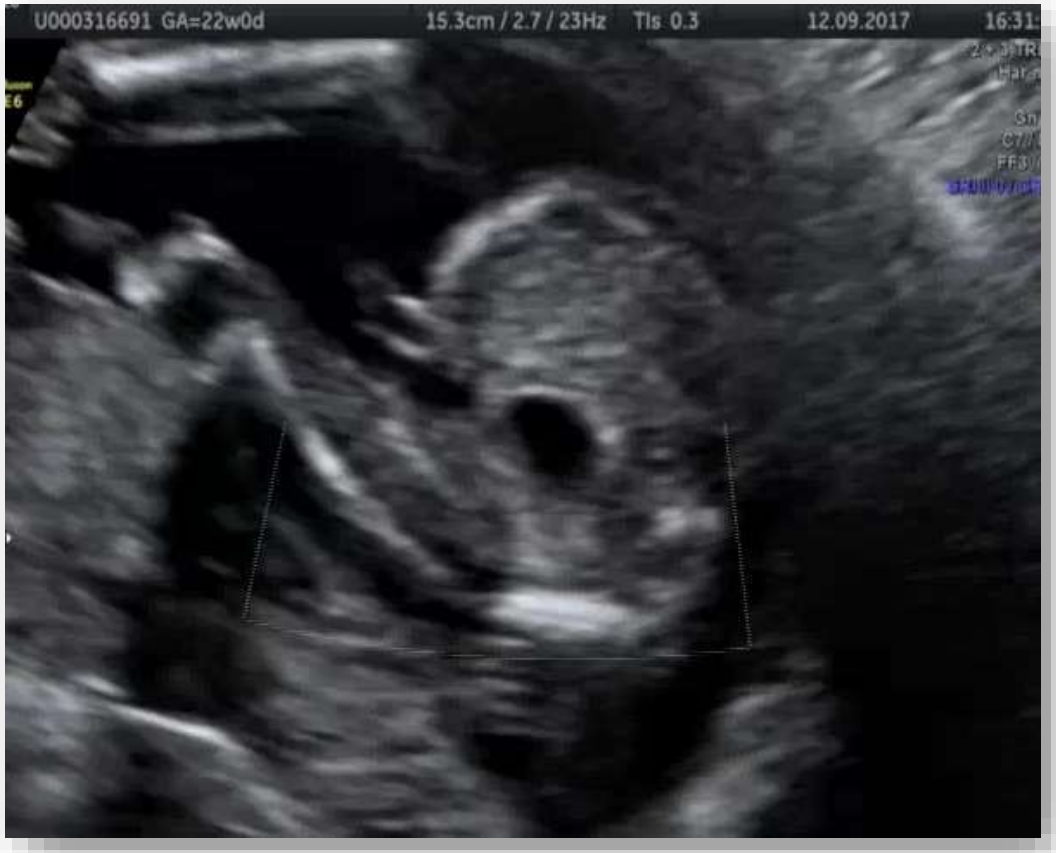
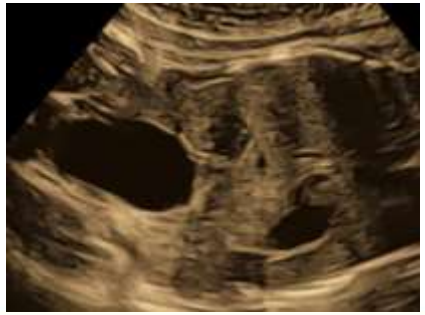
Index of renal function: liquor, Bladder

16 wks



- Transverse section of pelvis
  - Bladder and 2 Umbilical arteries
  - Bladder can be seen from 10 wks
  - Fetal urine contribution for liquor – 16 wks
  - The normal thickness of the bladder wall is about 2 mm
    Bladder filling & emptying 25 to 30min
  - Toward the end of the pregnancy, this cycle decreases, especially in female fetuses, due to hormonal effects



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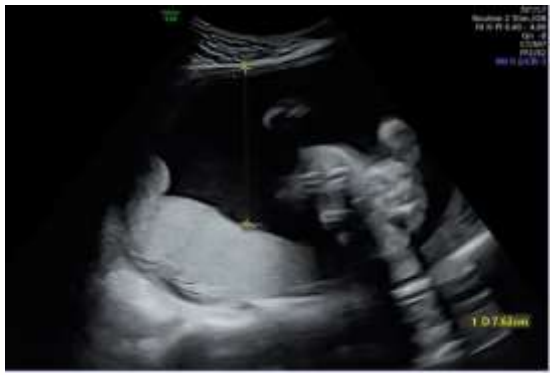
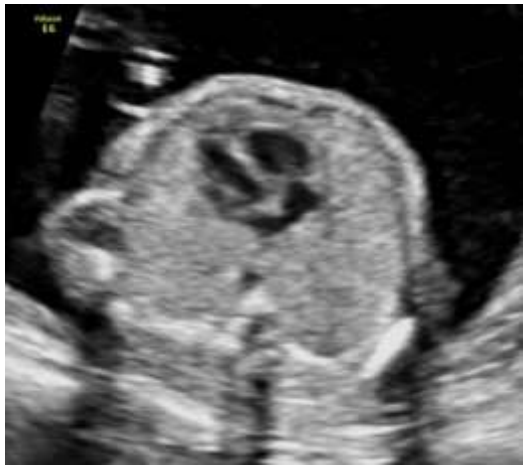
Renal function

- Fetal kidneys receive less than 5% of cardiac output (neonatal - 15%)
- Tubular immaturity, low sensitivity of the collecting duct to arginine vasopressin (AVP)- hypo-tonic urines (100–250 mOsm/kg H2O) leads to the production of large amniotic fluid essential for fetal well-being and lung growth
- Decline in hourly fetal urine production after 40 weeks' gestation due to maturity of of the tubular function





Springer International Publishing AG 2017.G. Buonocore et al. (eds.), Neonatology, DOI 10.1007/978-3-319-18159-2_261-1

Amniotic Fluid: Physiology and Assessment. Fischer, R, *Glob. libr. women's med*.,
(ISSN: 1756-2228) 2008; DOI 10.3843/GLOWM.102008

Fetal and neonatal physiology -4 th edition-Richard.A.Polin,William.W.Fox

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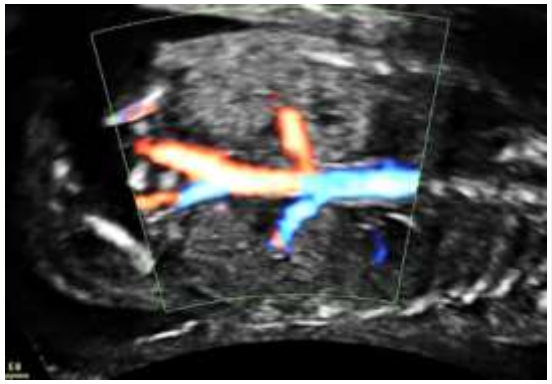
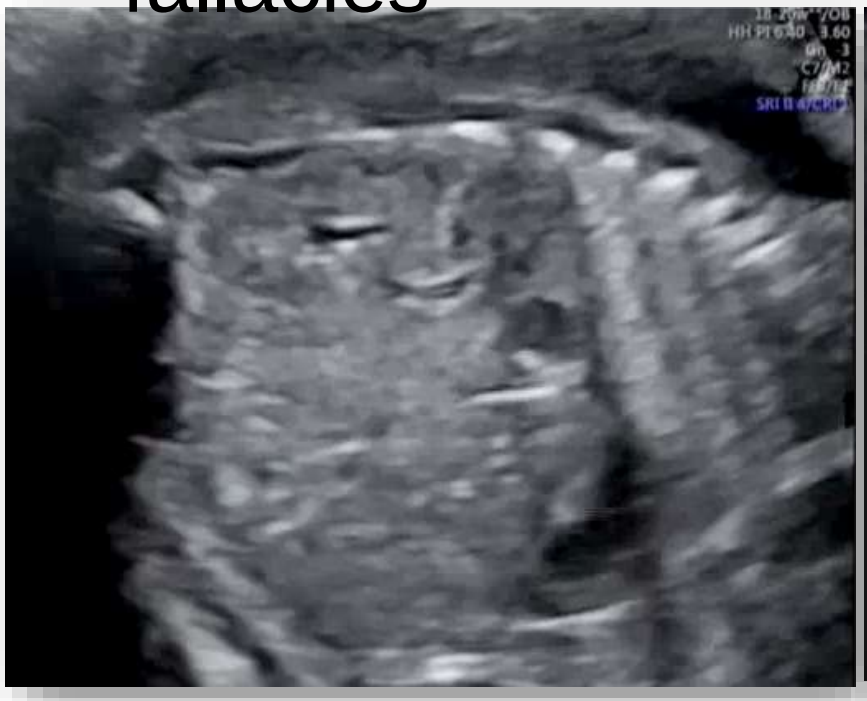
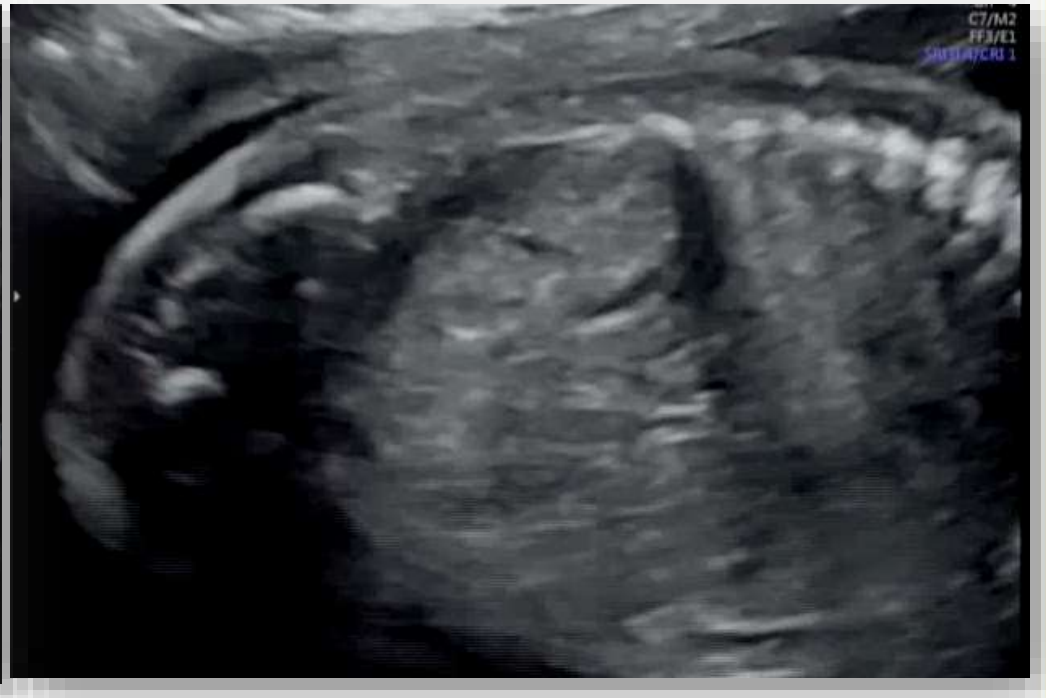
Assess the kidneys in 2 planes to avoid fallacies







- **Limitation – small bowel can mimic kidney**
- Fluid in Renal pelvis natural contrast
- Renal artery (pitfall Sup. Mes. Art)

Renal length = Gestational age in wks

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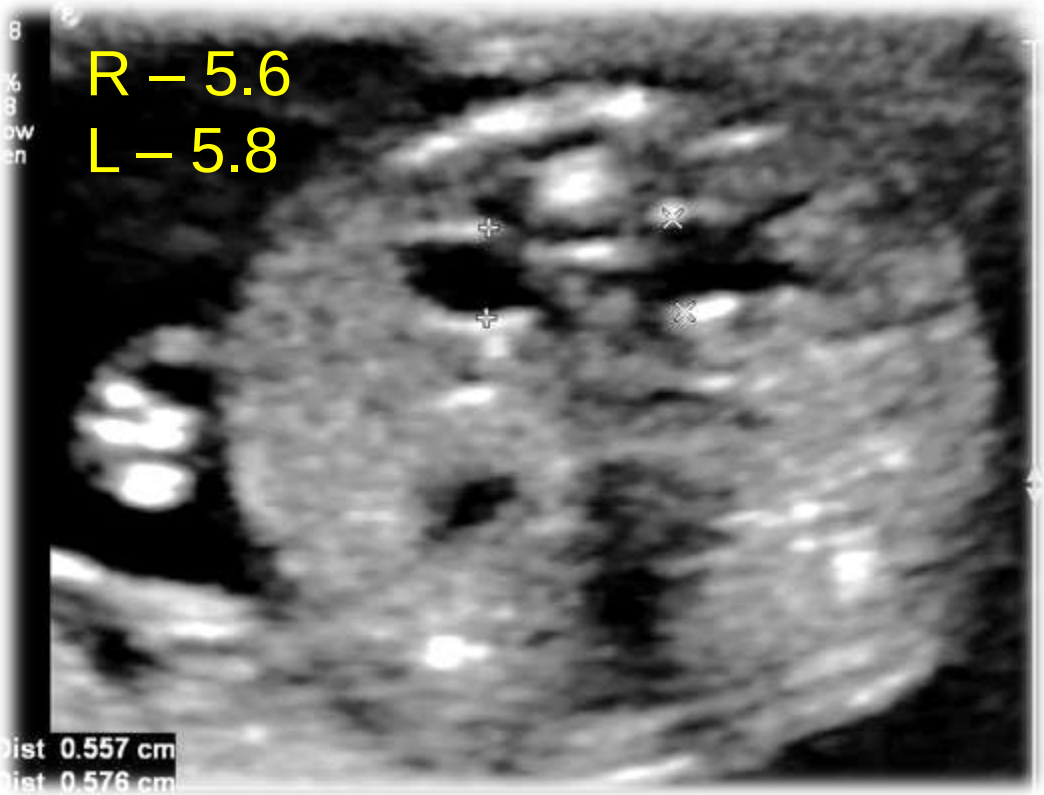
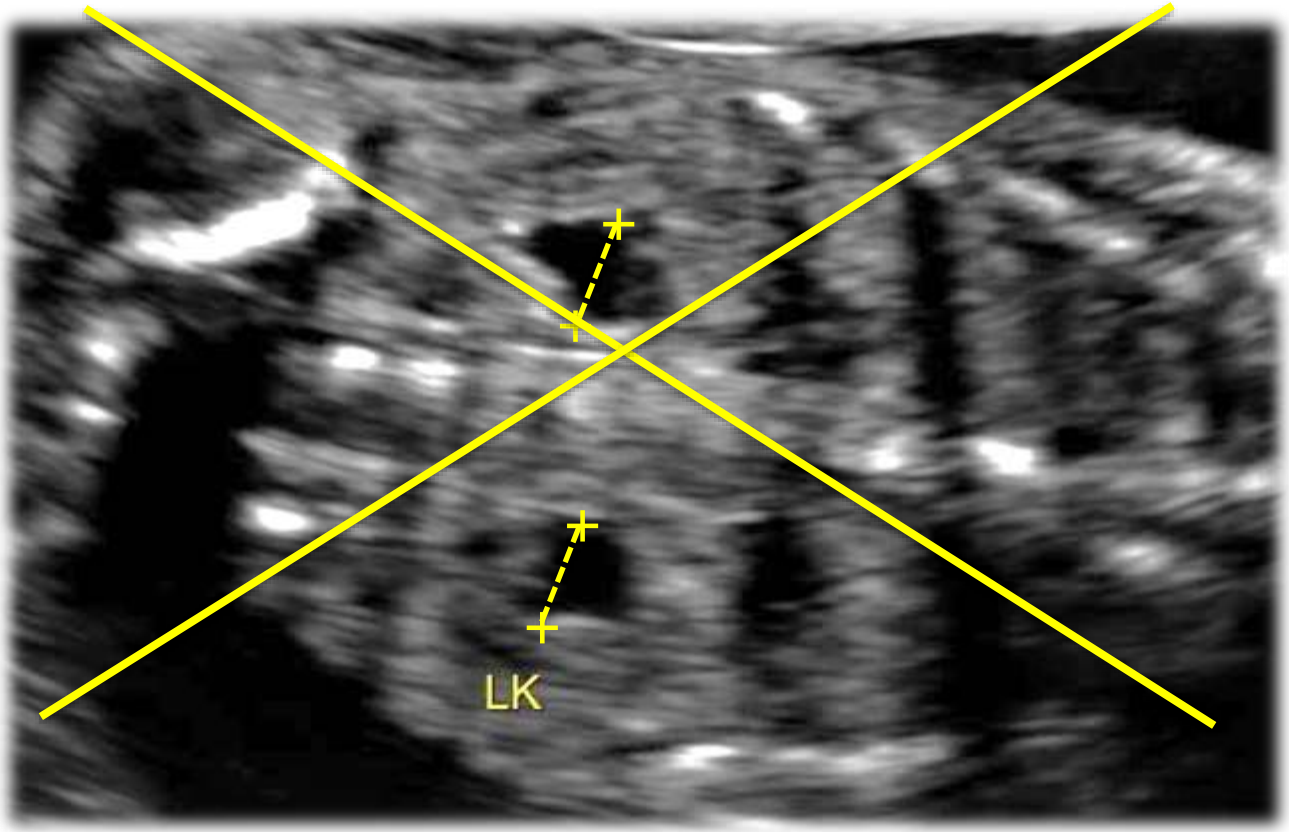
ASSESSMENT OF RENAL PELVIS





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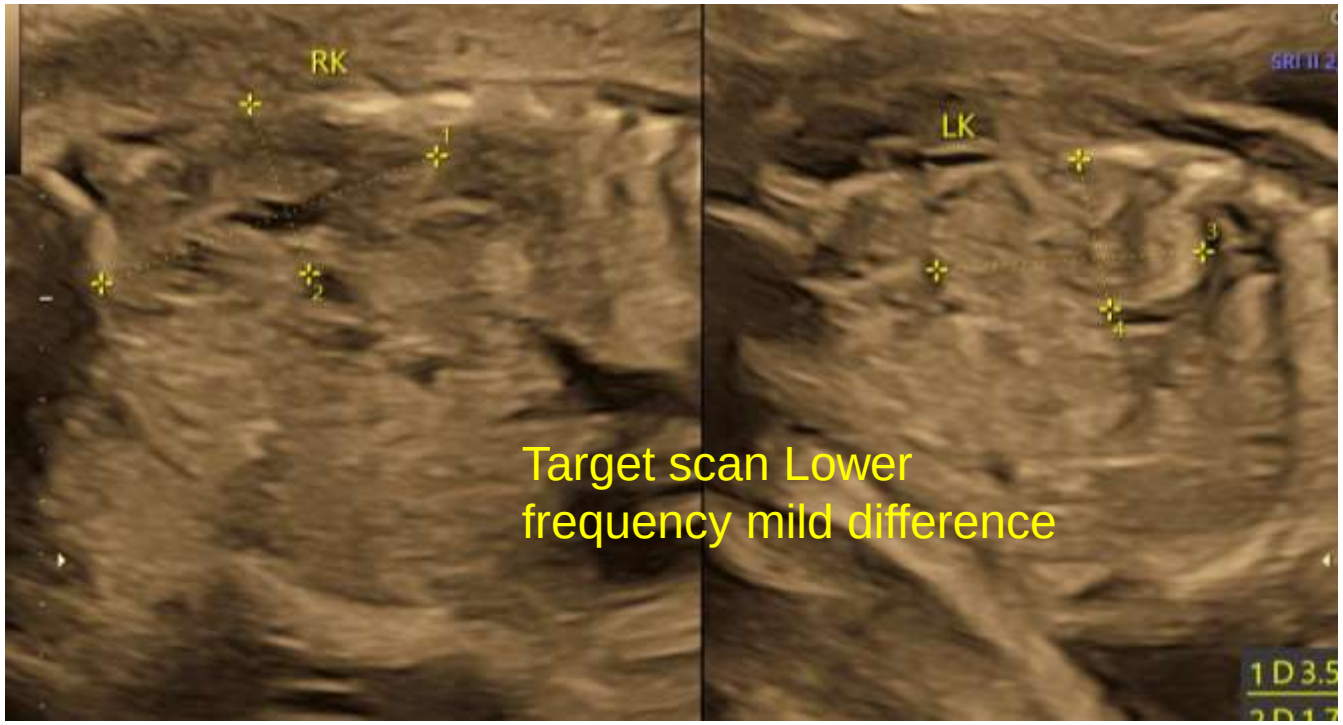
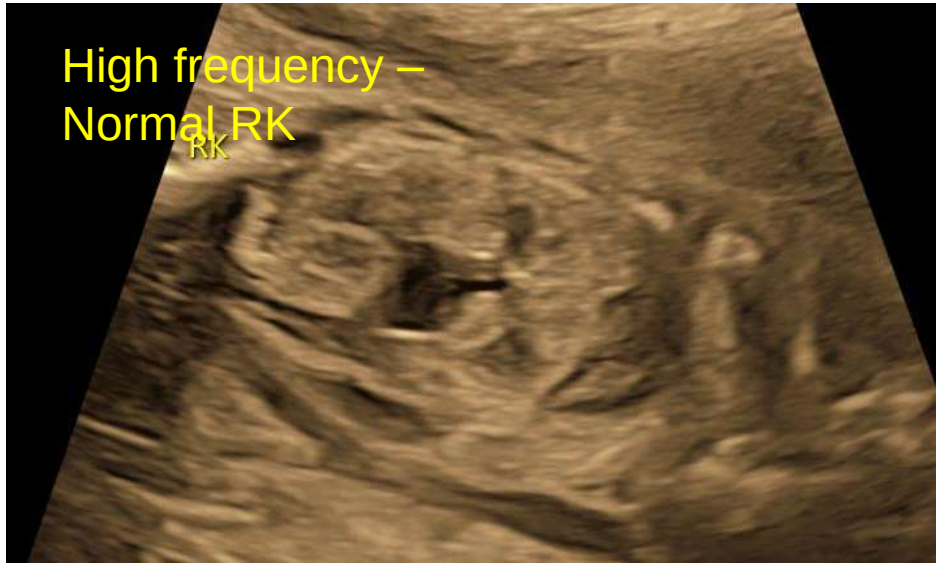
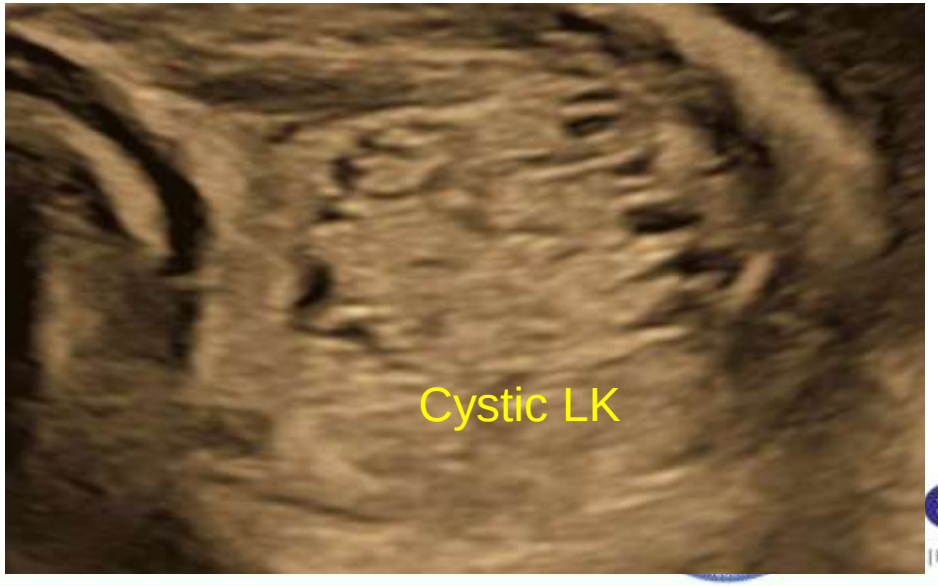
Importance high resolution scan in looking at morphology







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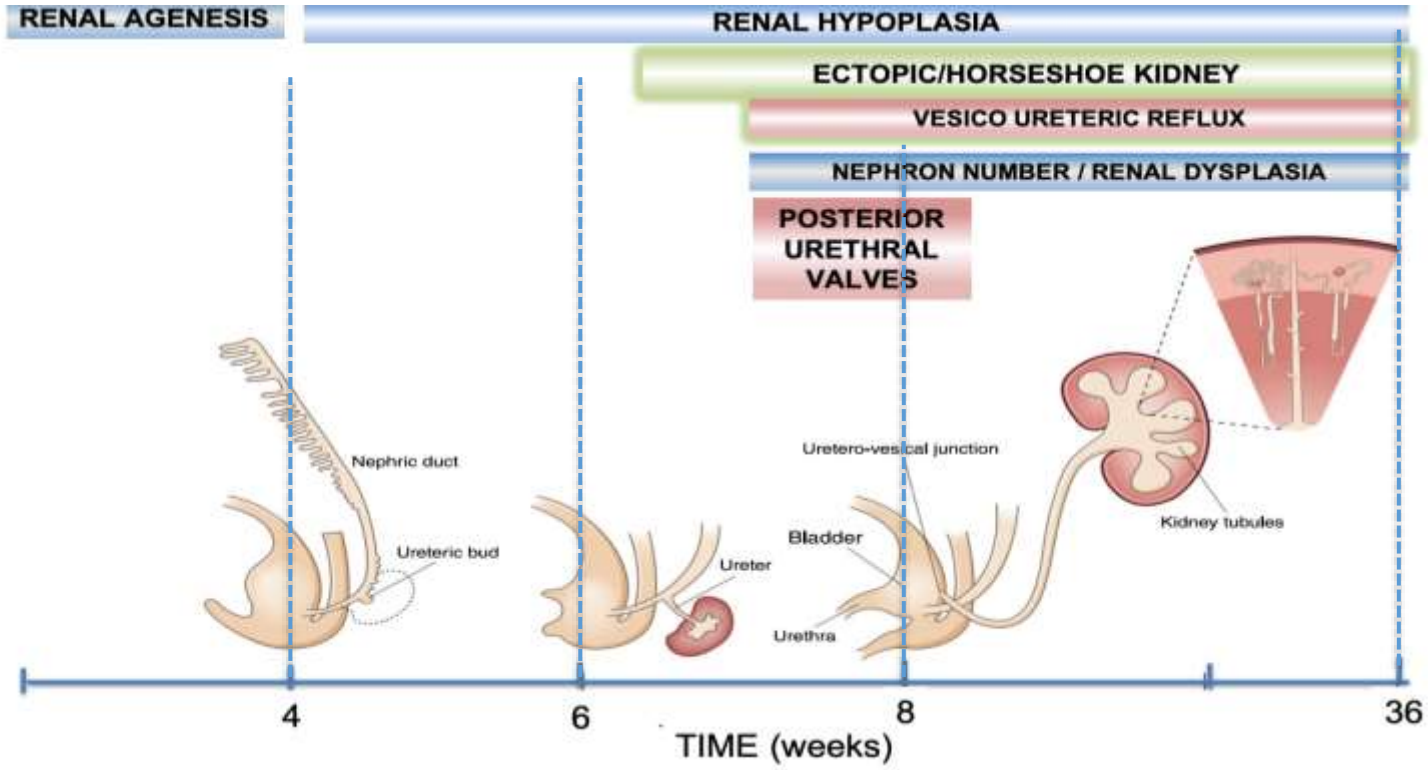
CAKUT

- Embryonic disorders arising during development
- Spectrum of defects involving Kidneys, ureters, bladder and urethra (outflow tracts)

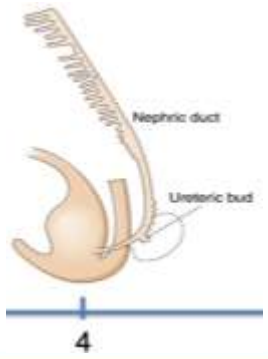


Vasikar Murugapoopathy[1] and Indra R. Gupta[1,2]

KIDNEY NUMBER

Abnormalities

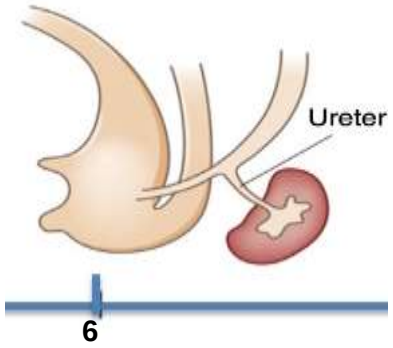
- Failure of formation of the kidney/s and outflow systems
- Renal agenesis
  - Unilateral
  - Bilateral



SIZE & MORPHOLOGY

Abnormalities

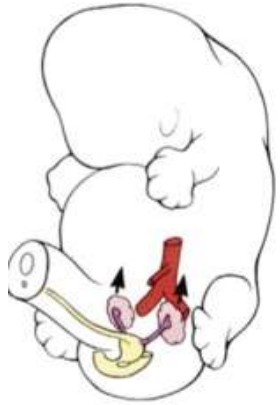
- Hypoplasia
- Dysplasia
- Multicystic dysplastic kidneys



KIDNEY POSITION

Abnormalities

- Low placed kidney
- Horse shoe kidney
- Ectopic kidney

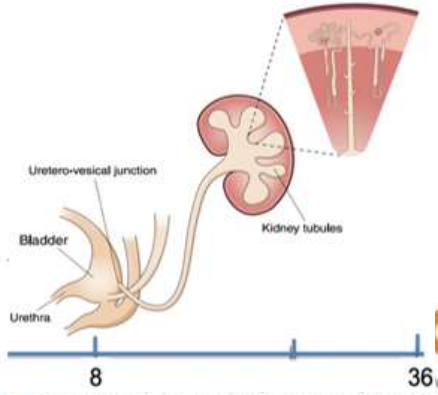


Ascent starts by 6 weeks

OUTFLOW ABNORMALITI

Abnormalities

- PUJ
- Ureterocele
- Megaureters
- Vesico ureteric reflux
- Posterior urethral valves
- Urethral atresia



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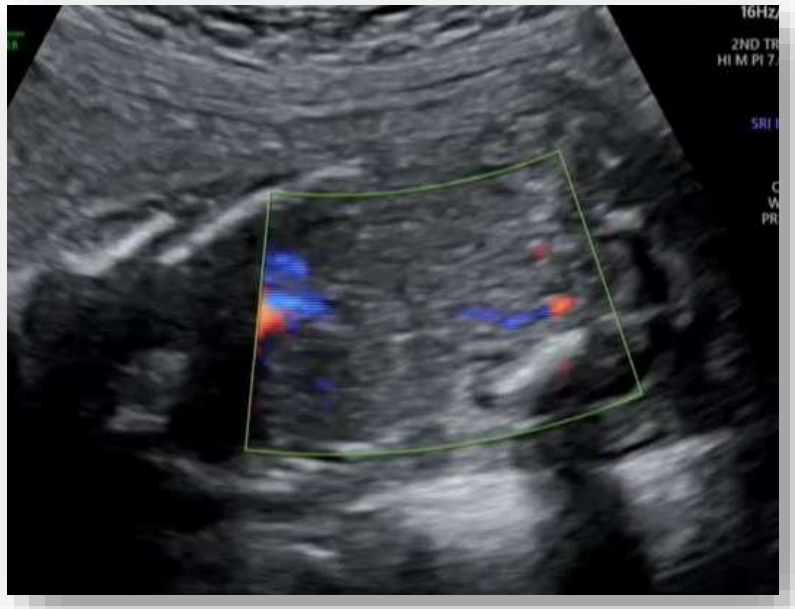
Teach Teach
an
al Pathology

Oligoamnios – ask history of maternal medication

- Administration of COX inhibitors NSAID, ACE inhibitors decreases renal blood flow, impairs renal function, and induces oligohydramnios.
- Abnormal morphogenesis (cystic dilatation of nephrons)



Antonucci R, Zaffanello M, Puxeddu E, Porcella A, Cuzzolin L, Dolores Pilloni M, Fanos V. **Use of non-steroidal anti-inflammatory drugs in pregnancy: impact on the fetus and newborn. Current drug metabolism.** 2012 May 1;13(4):474-90

Buttar HS. An overview of the **influence of ACE inhibitors on fetal-placental circulation & perinatal development.** Mol Cell Biochem.1997 Nov;176(1-2):61-71.

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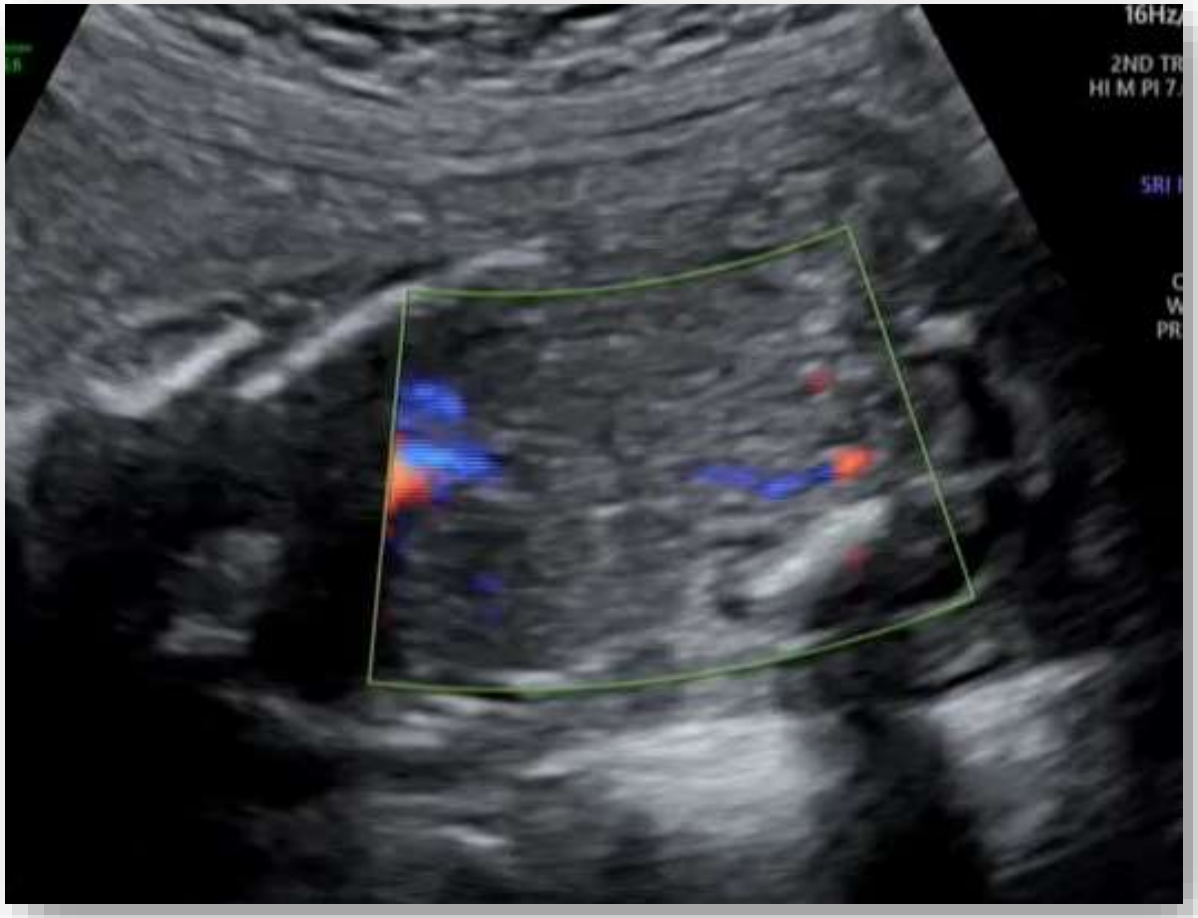
Bilateral Renal agenesis



- Diagnosis
  - 15-16 wks
- Anhydramnios (>15 wks)
- Absent bladder
- Empty renal fossae
- Bil lying down adrenals
- No renal arteries on colour
- Look for associated anomalies

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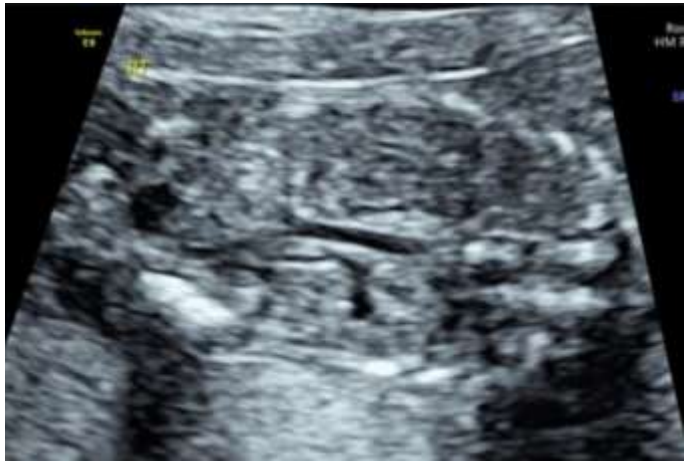
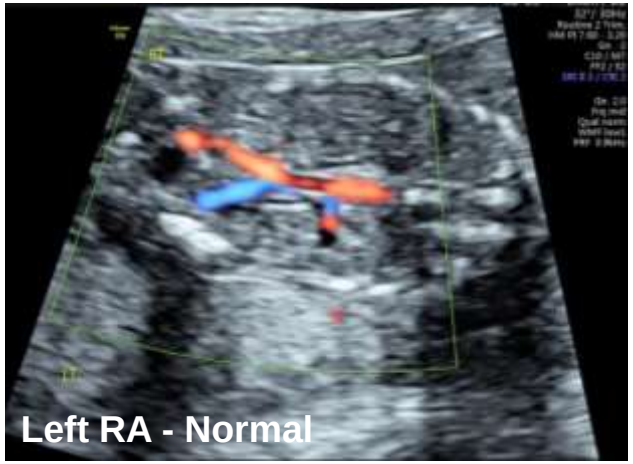
Empty renal fossa one side – ?unilateral renal agenesis

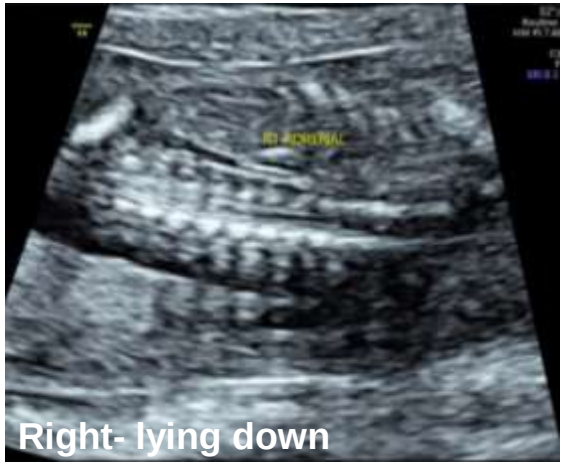
17+3 weeks

LK- Normal

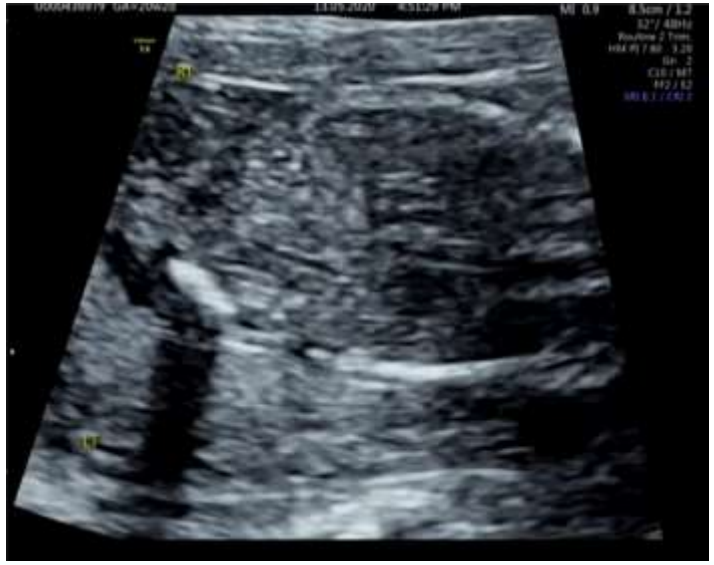




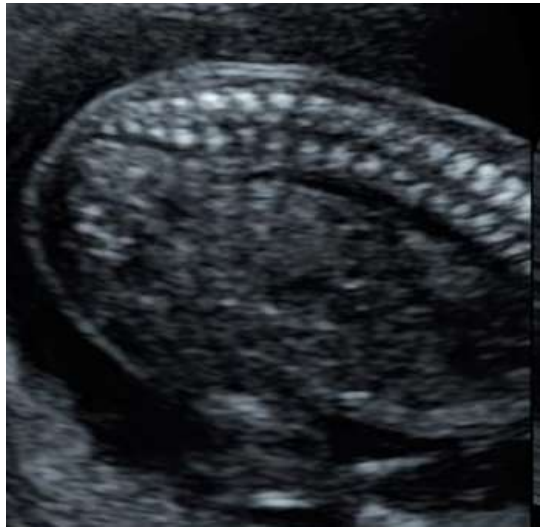
Left RA - Normal

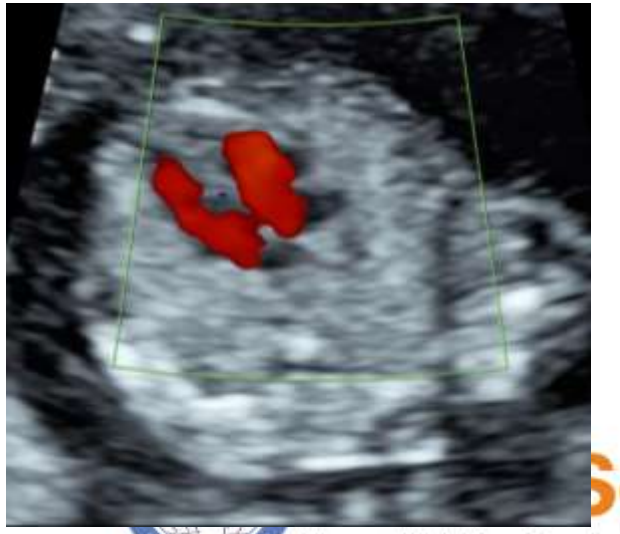


Right- lying down



Abdominal Aorta Bif Angle - 44°





Scan
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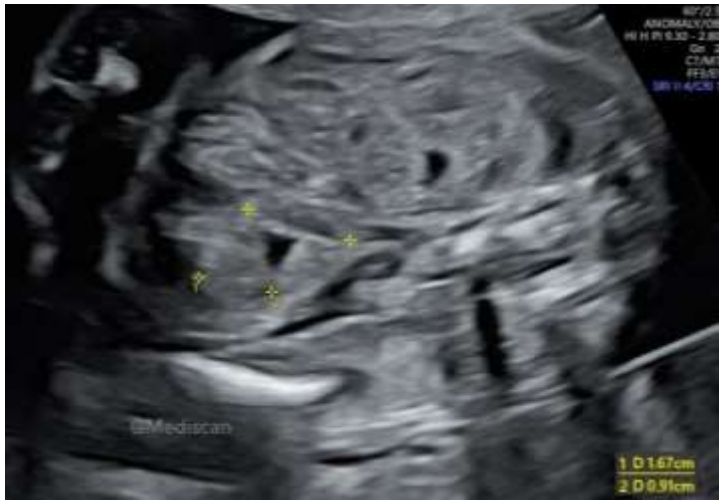
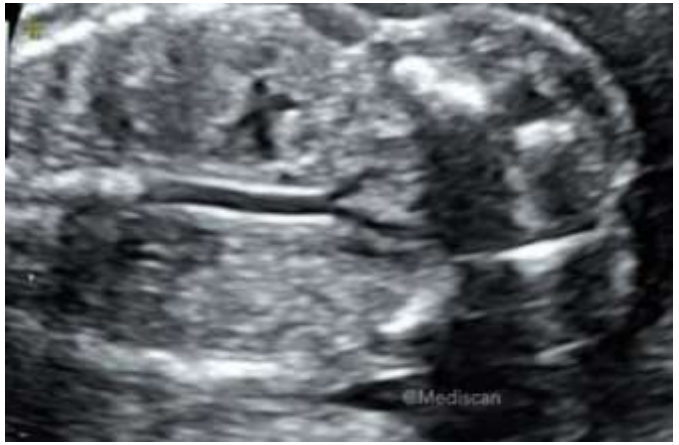
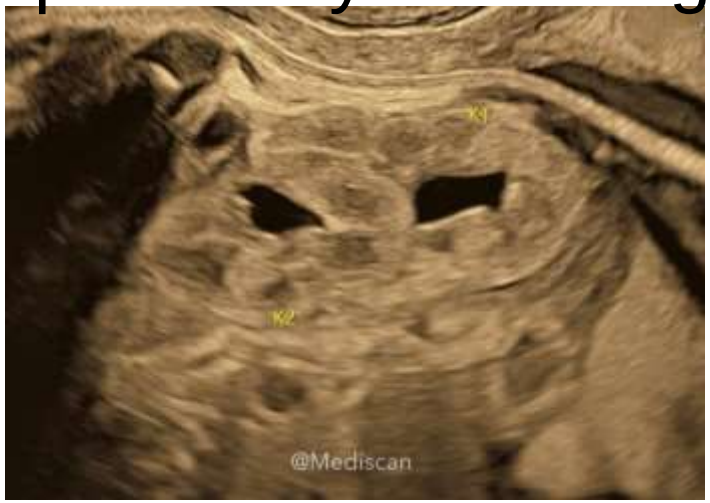
Empty renal fossa unilateral look for ectopic kidney as fetus grows



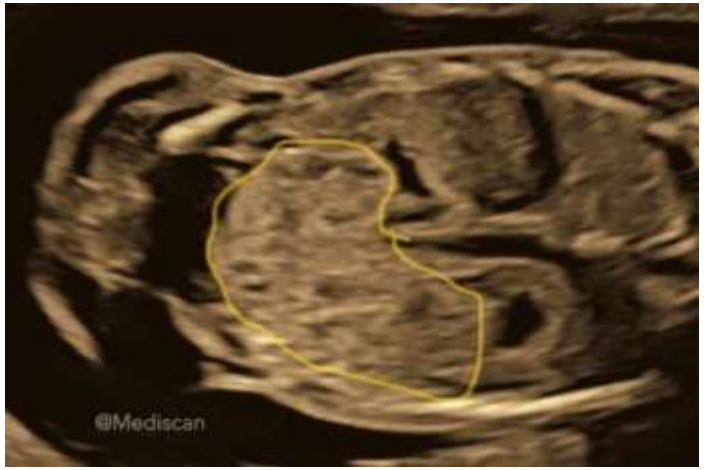
PELVIC KIDNEY



UNILATERAL RENAL AGENESIS



CROSSED ECTOPIA



HORSESHOE KIDNEY

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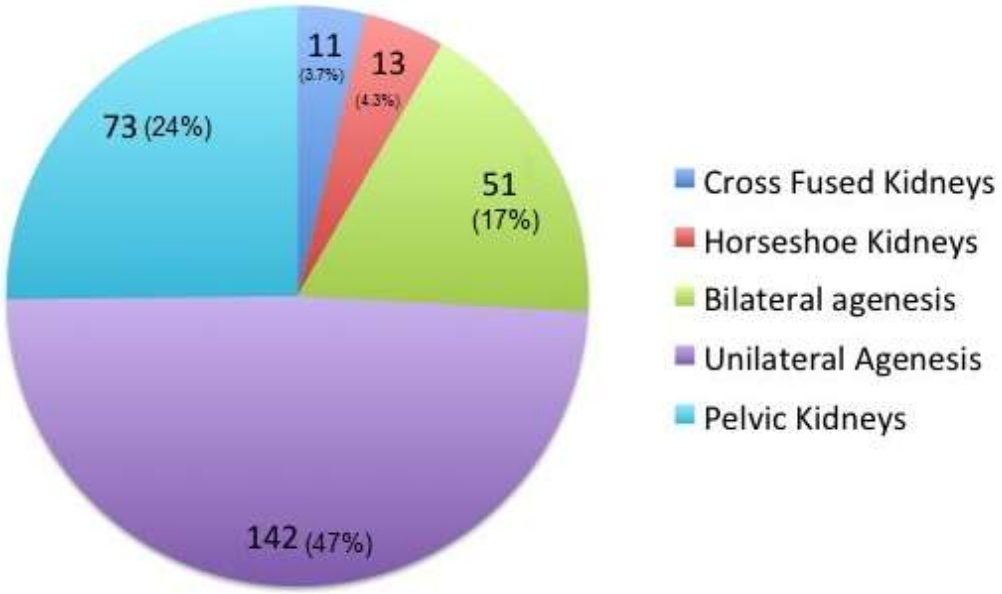
Empty renal fossa (Mediscan 2014-15) n= 299



- Isolated 60% (180)
- Assoc. anomalies 40% (119)
- Most diagnosed in the 2nd and 3rd trimester

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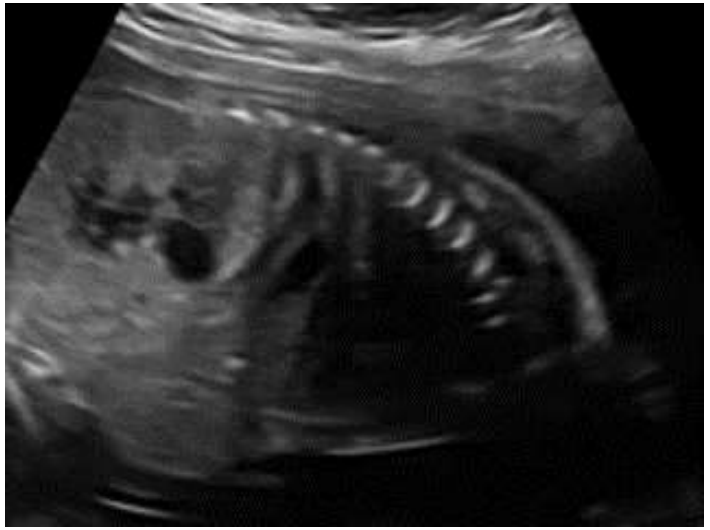
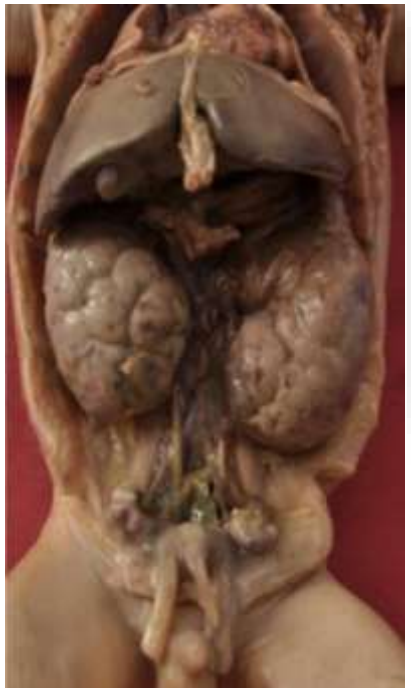
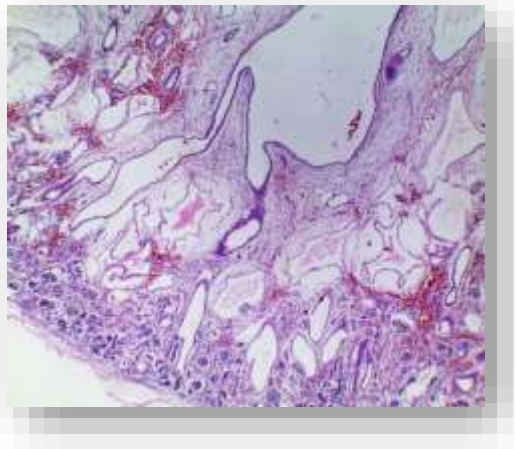
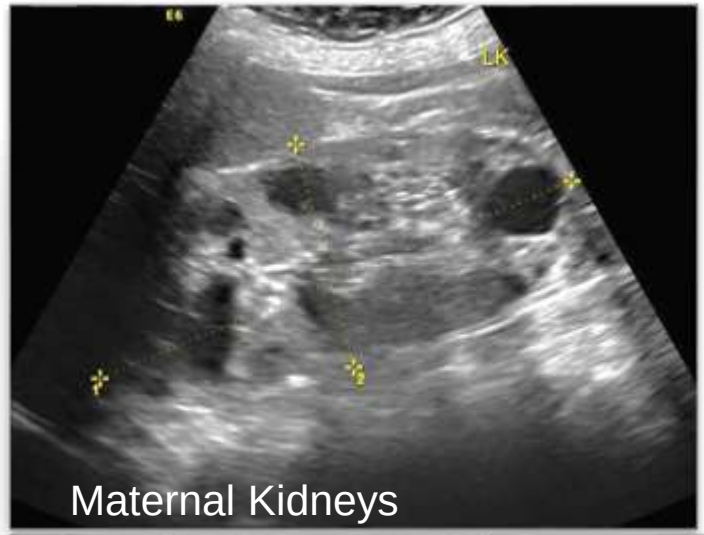
ECHOGENIC KIDNEYS - 1

- Primigravida
- For routine target scan 23 wks
- Liquor – normal

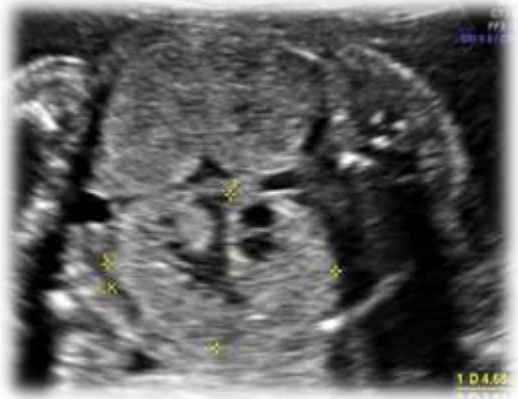


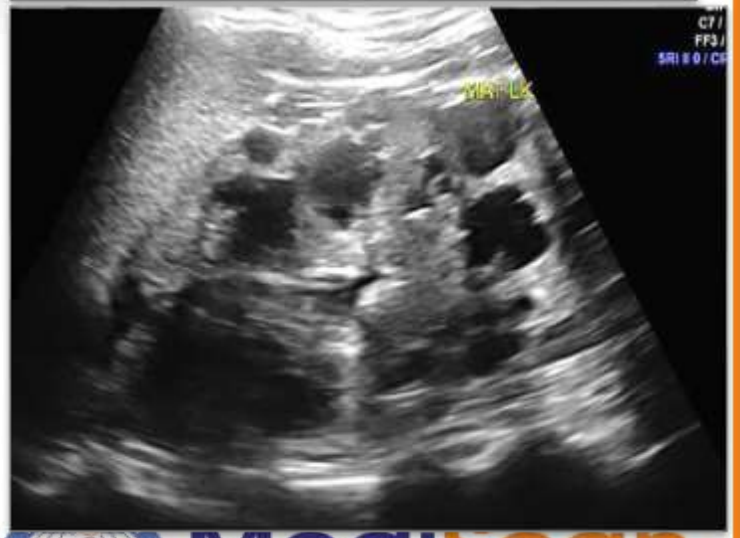






Maternal Kidneys





Autosomal Dominant
Polycystic Kidney disease
PKD1 gene

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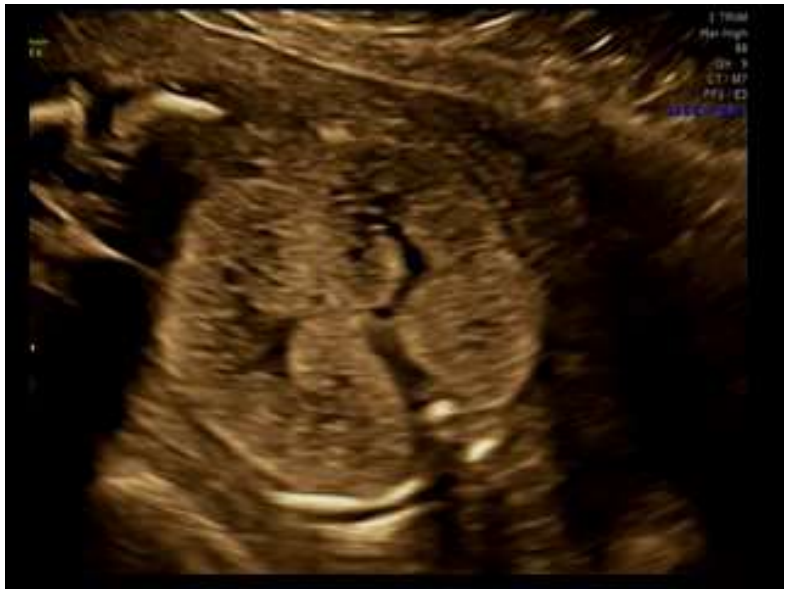
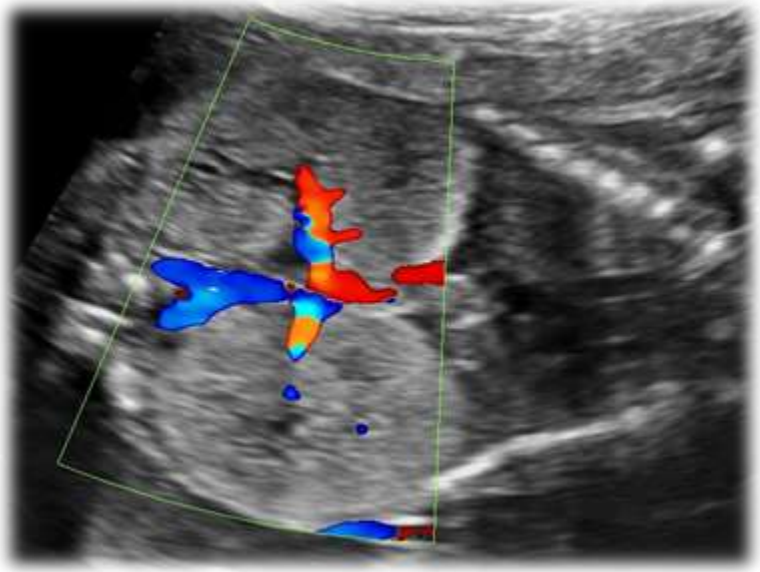
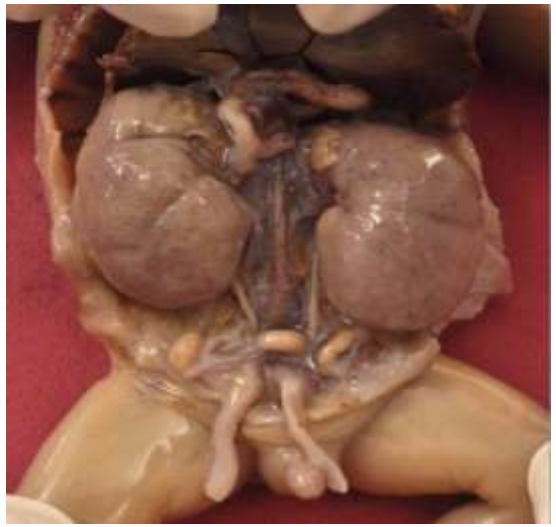
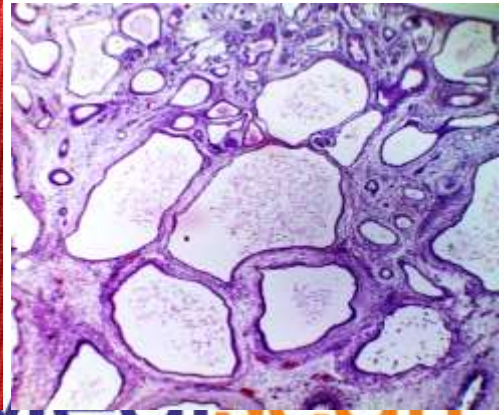
ECHOGENIC KIDNEYS - 2



- Autosomal Recessive polycystic kidneys (ARPKD)
- Always bilateral
- Associated with hepatic fibrosis
- Oligohydramnios







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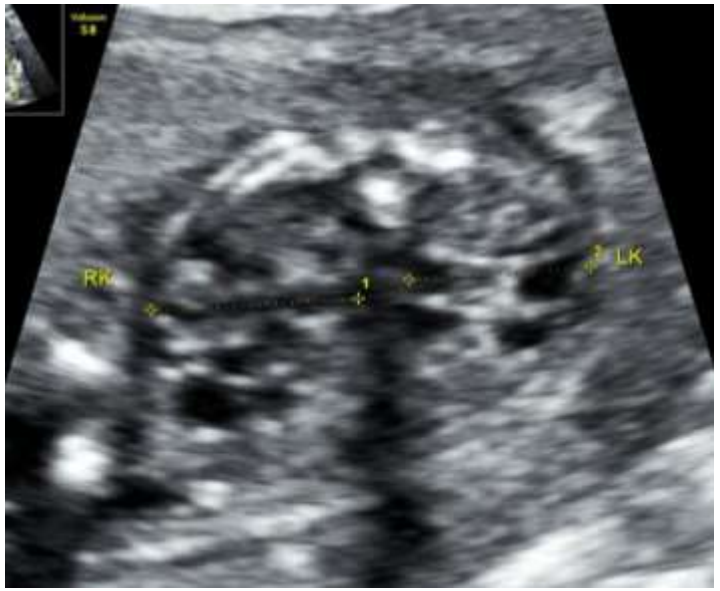
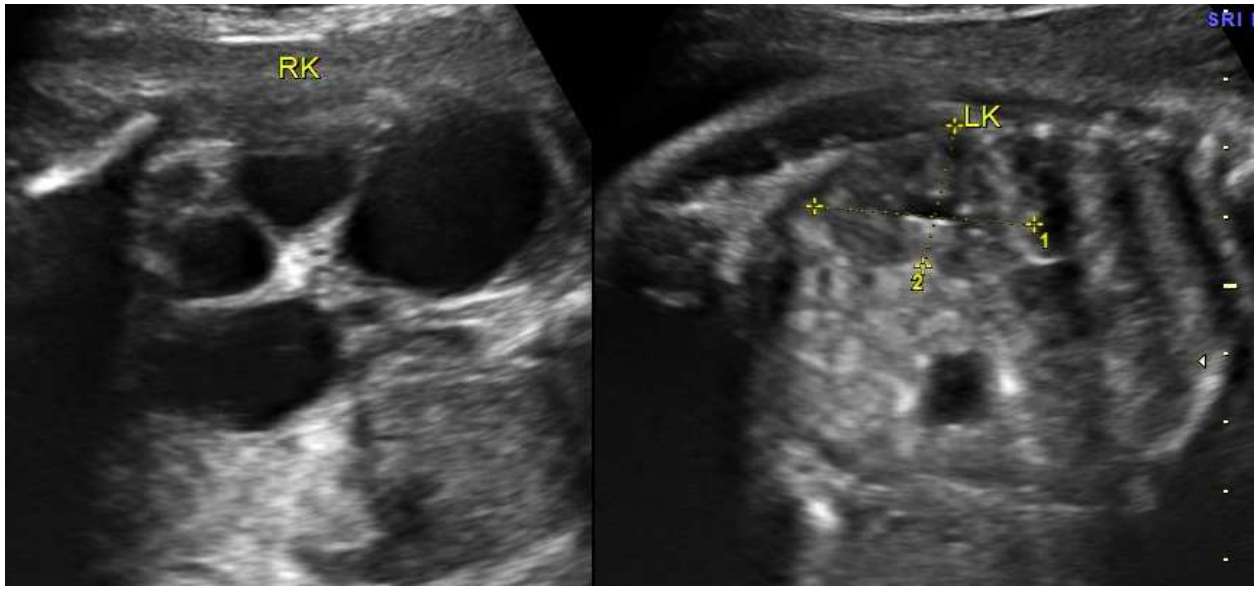
MCDK



MCDK – Bilateral



MCDK – Unilateral

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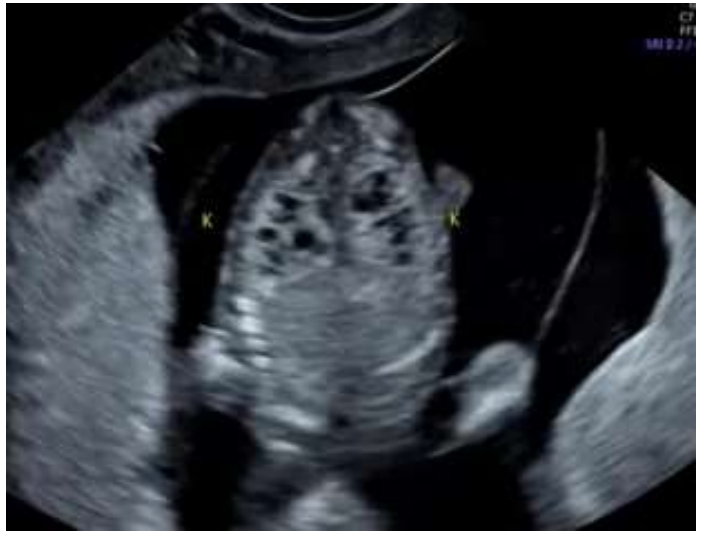
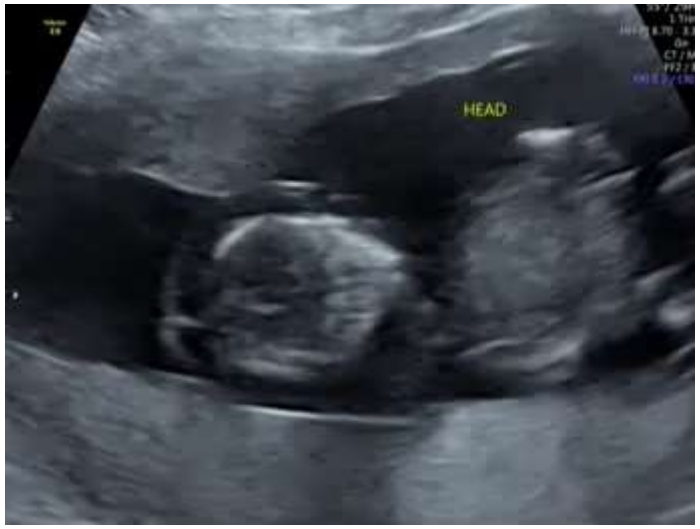
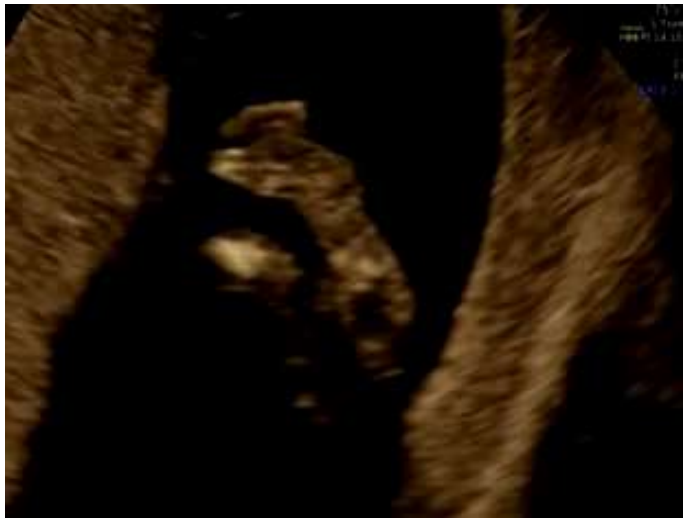
CYSTIC RENAL DYSPLASIA- SYNDROMIC







Cystic kidneys Encephalocele Polydactyly

Meckel Gruber Syndrome

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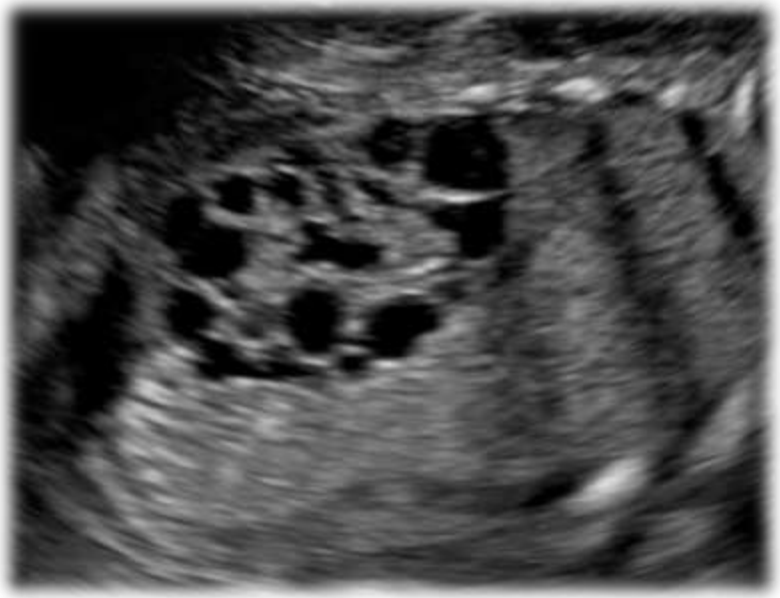
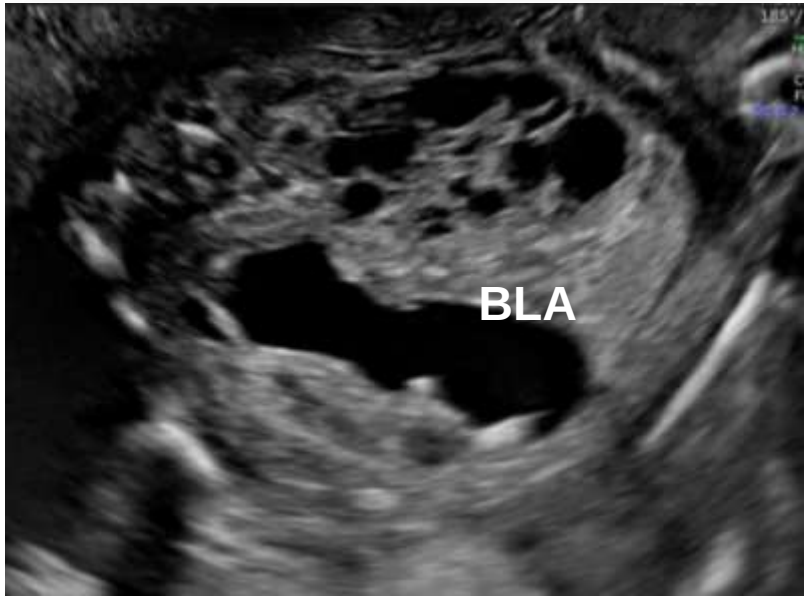
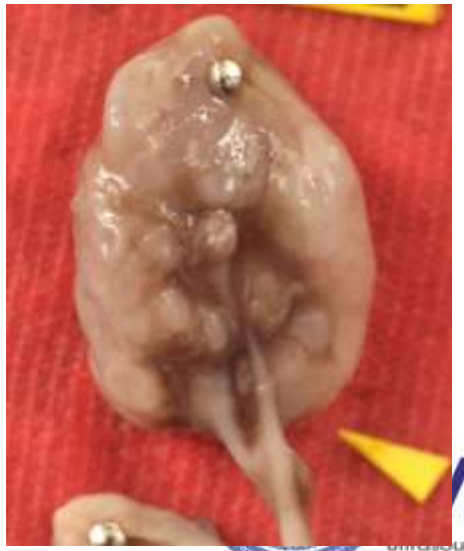
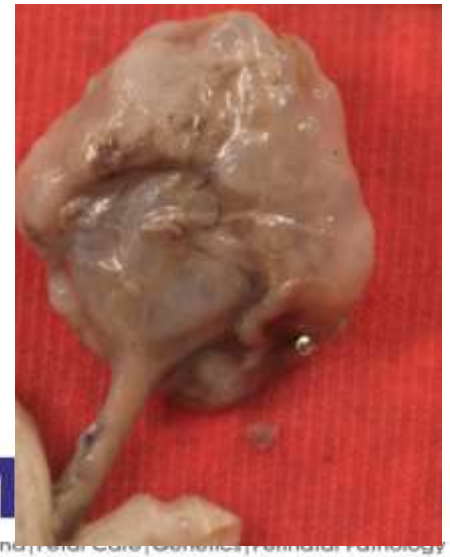
CYSTIC RENAL DYSPLASIA- OBSTRUCTIVE





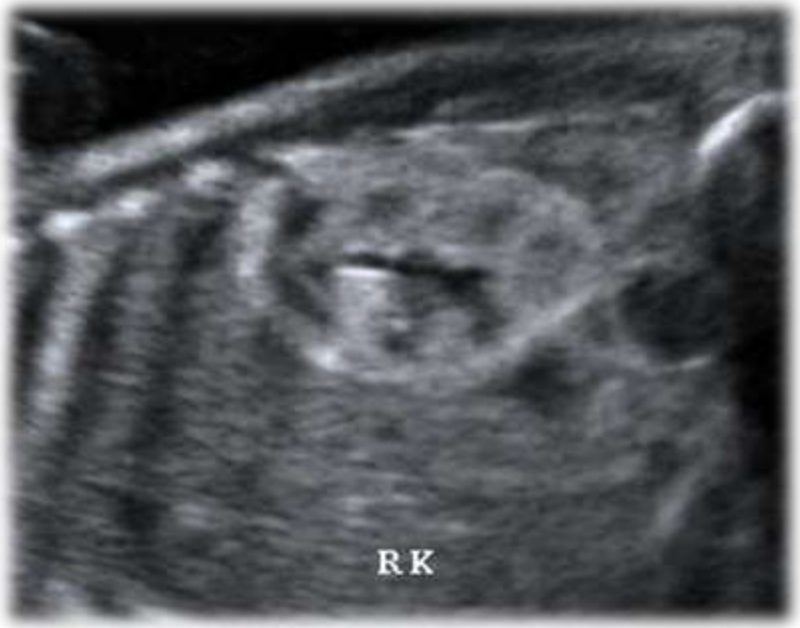
- Lower urinary tract obstruction
- Cystic changes in the kidney due to obstructive dysplastic changes
- Non functioning kidneys





Isolated mildly echogenic normal size kidney preserved function -? Normal variant

- Looking at complete phenotype …Extra renal anomalies and family history important in planning genetic testing and counselling



- Serial fetal scan to asses renal function and postnatal examination

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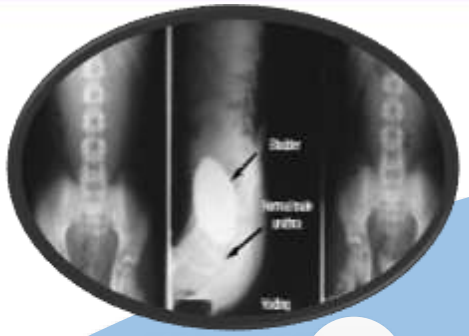
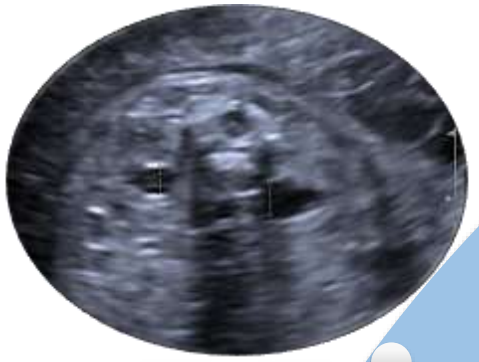
1. UTD - 1-5% of pregnancies



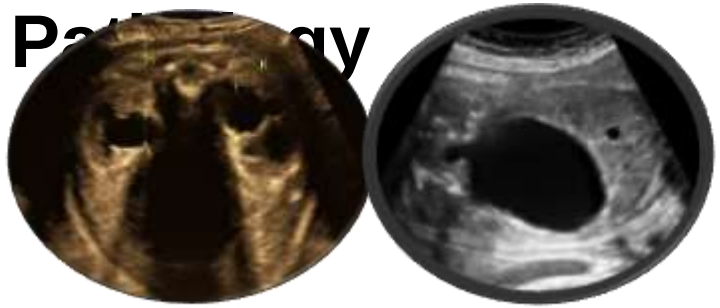
Undetermined



Transient / Physiological

80%

Obstructive Pa[illegible]gy



- Dilatation is NOT always Obstruction!
- Dilatation is a dynamic process

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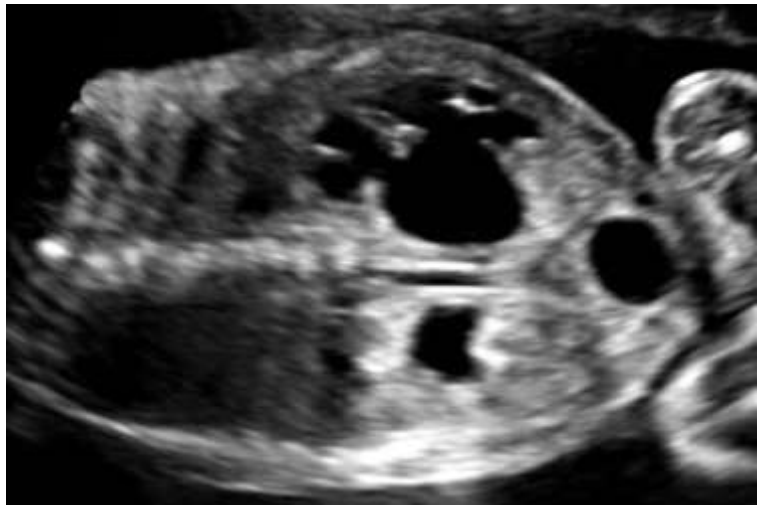
2. Standardizing nomenclature



- Hydronephrosis
- Pyelectasis
- Pelviectasis,
- Uronephrosis
- Hydroureteronephrosis
- UT fullness or prominence
- Pelvic fullness

URINARY TRACT DILATATION (UTD)

A1 **A2 –A3**

Journal of Pediatric Urology (2014) 10, 982–999

ELSEVIER

Journal of Pediatric urology

Multidisciplinary consensus on the classification of prenatal and postnatal urinary tract dilation (UTD classification system)

Hiep T. Nguyen [d,f,*], Carol B. Benson [h,a], Bryann Bromley [b], Jeffrey B. Campbell [d,f], Jeanne Chow [g], Beverly Coleman [a,h], Christopher Cooper [d,f], Jude Crino [e], Kassa Darge [g], C.D. Anthony Herndon [d,f], Anthony O. Odibo [e], Michael J.G. Somers [c], Deborah R. Stein [c]

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Post-natal risk

Severity	Second trimester	Third trimester	Risk of PN pathology
MILD	4-<7mm	7-<9mm	**11.9 %**
MODERATE	7-10mm	9-15mm	**45.1 %**
SEVERE	>10 mm	>15mm	**88.3 %**

Exceptions

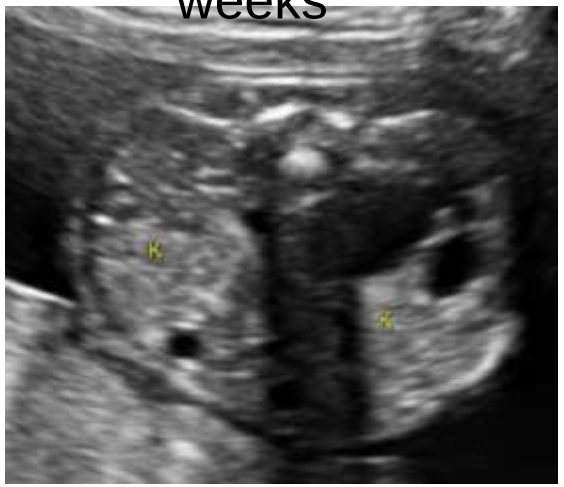
1. **VUR**
2. **Distal Urethral Obstruction**

Lee RS, Cendron M, Kinnamon DD, Nguyen HT. Antenatal hydronephrosis as a predictor of postnatal outcome: a meta-analysis. Pediatrics 2006;118:586.

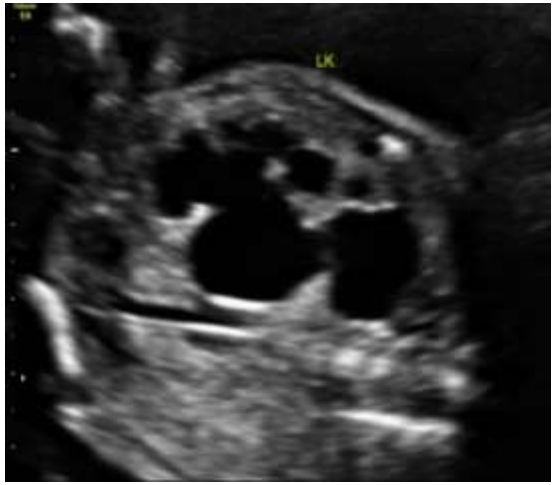
Signorelli M, Cerri V, Taddei F, Groli C, Bianchi UA. Prenatal diagnosis and management of mild fetal pyelectasis: implications for neonatal outcome and follow-up. Eur J Obstet Gynecol Reprod Biol 2005;118:154.

Unilateral upper tract dilatation likely PUJO

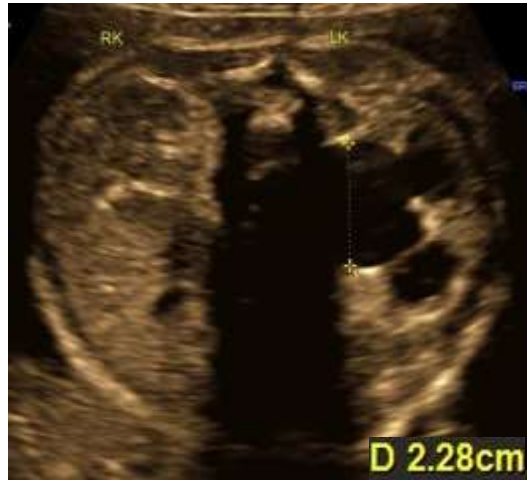
24 weeks

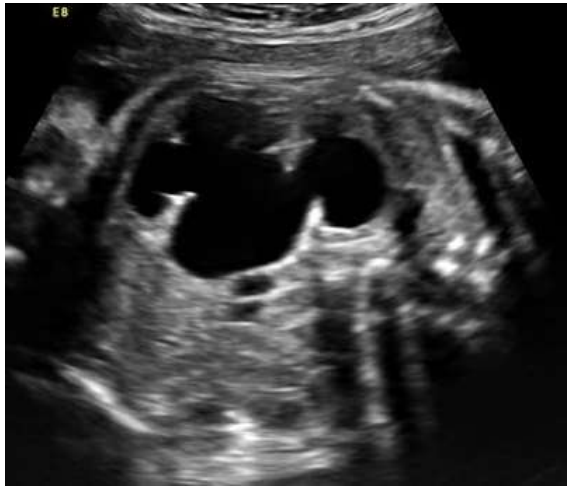


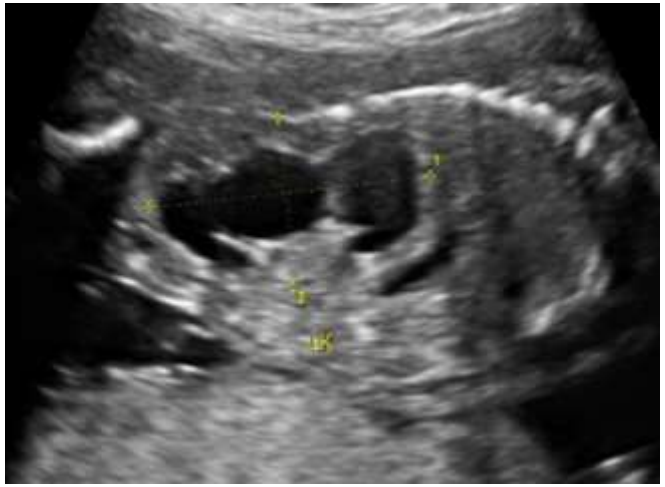
26 weeks

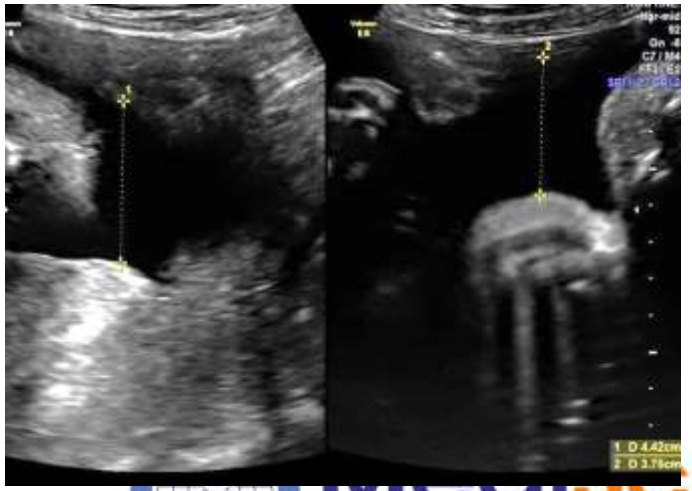


28 weeks









30 weeks

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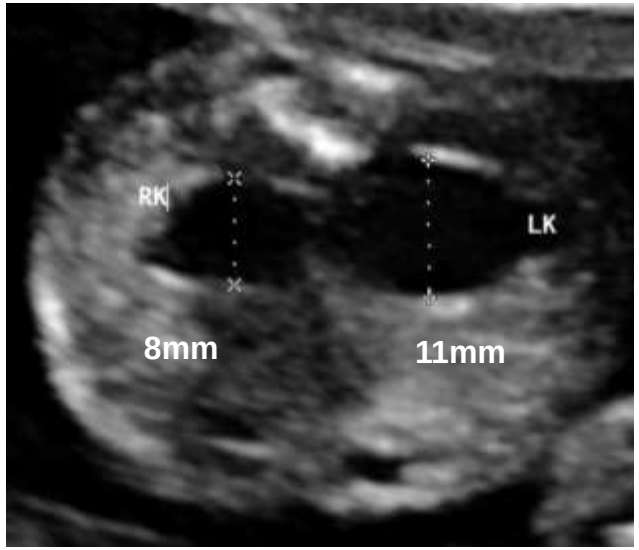
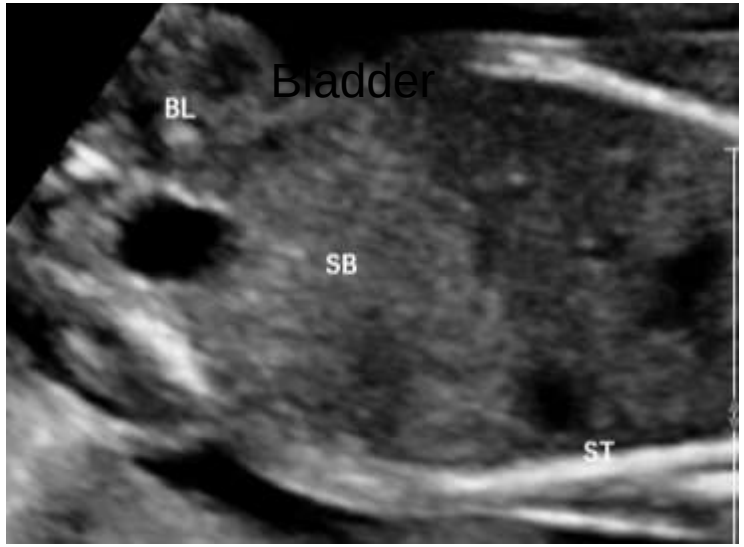
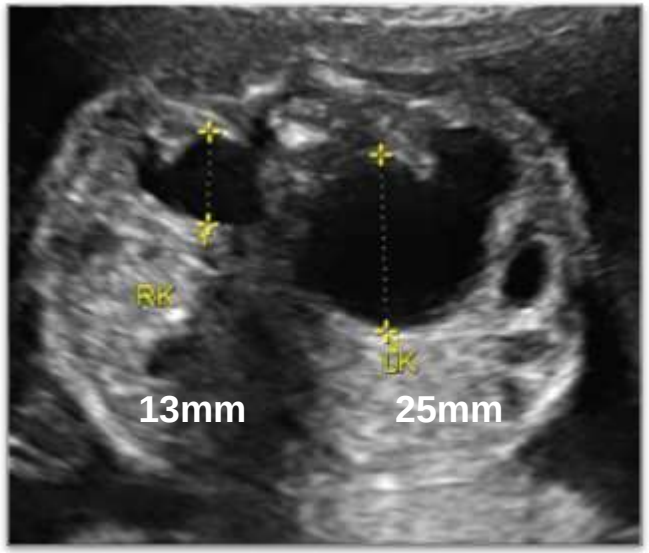
Bilateral uppertract dilatation Lt> RT

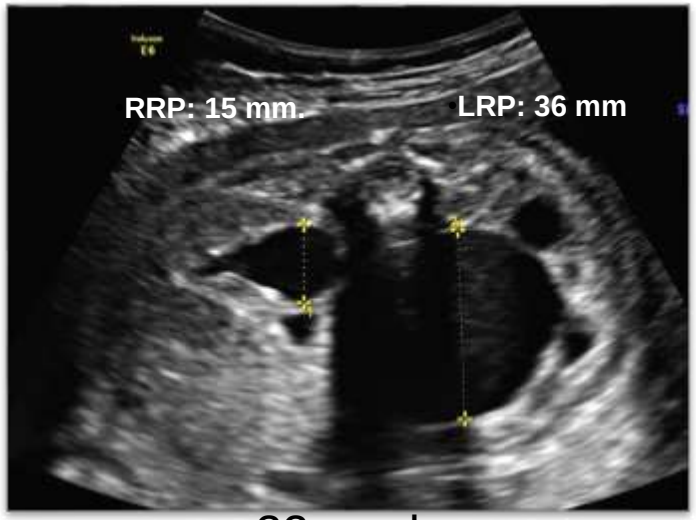
20 wks





26+2 wks





32 weeks

Bladder normal

Liquor normal

No cysts in the renal cortex

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Recommendation - Reporting

- Description of seven imaging parameters
  1. AP-Renal Pelvis Diameter
  2. Calyceal dilatation
  3. Parenchyma thickness
  4. Parenchymal appearance
  5. Ureters
  6. Bladder
  7. Liquor

<u>Impression section :</u>

- The specific UTD category (Normal, UTD A1, UTD A2-3, UTD P1, UTD P2, or UTD P3)
- Along with the suggested management scheme.
- Ideally, representative images should be provided with the report

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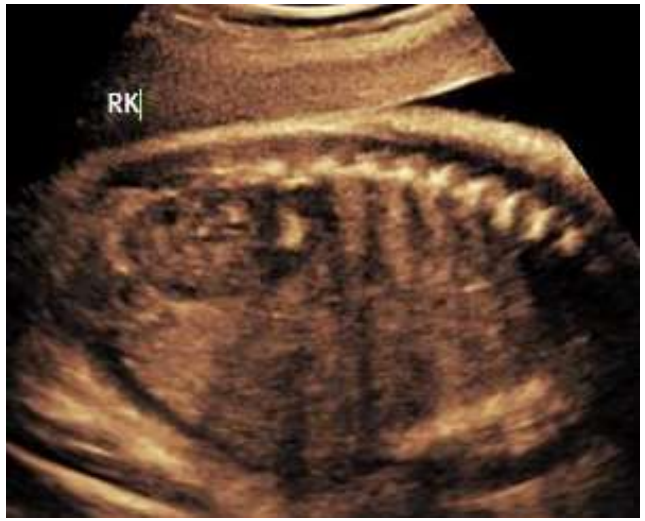
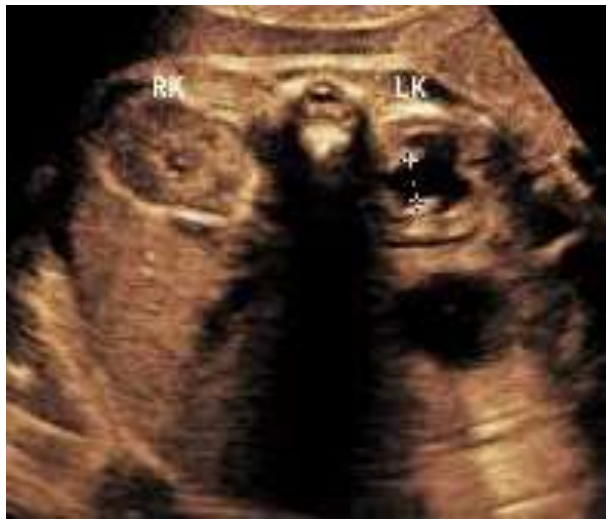
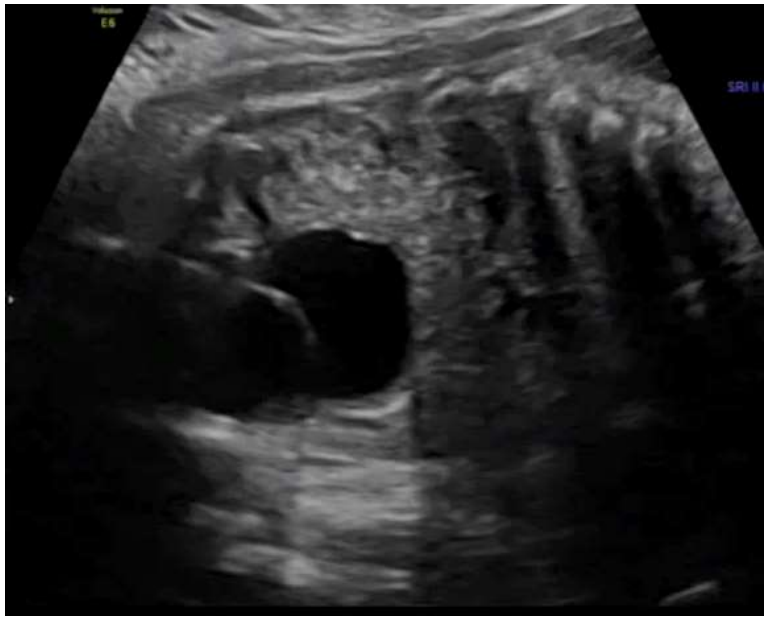
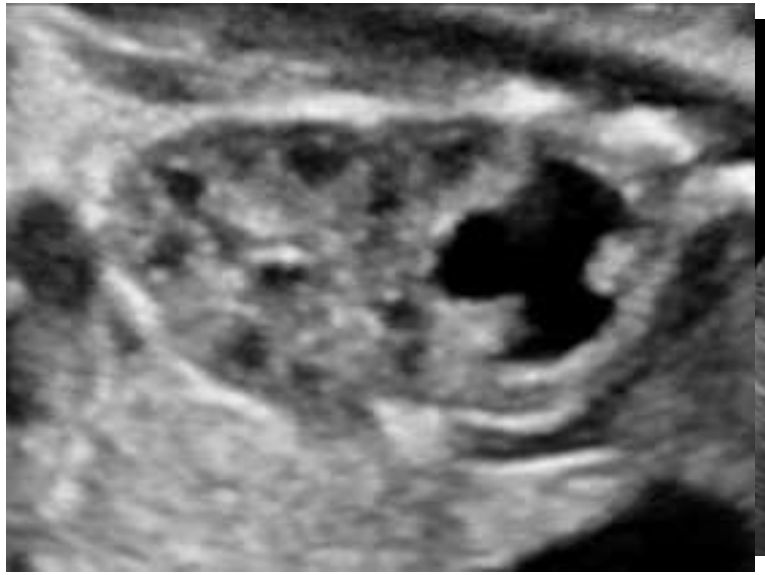
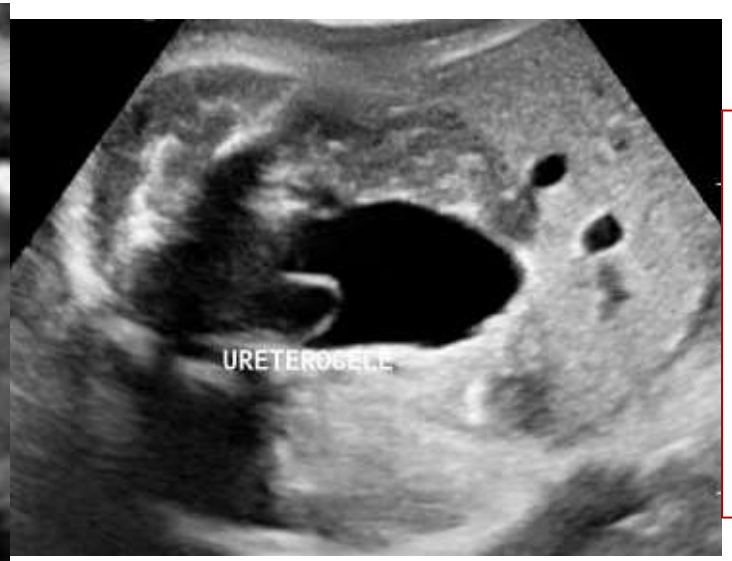
Uretero vesical obstruction - Ureterocele











Double moiety

Ureteroceles

- **Simple – ureteric orifice in the bladder trigone**
- **Ectopic - ureteric orifice,in the bladder neck or posterior urethra**

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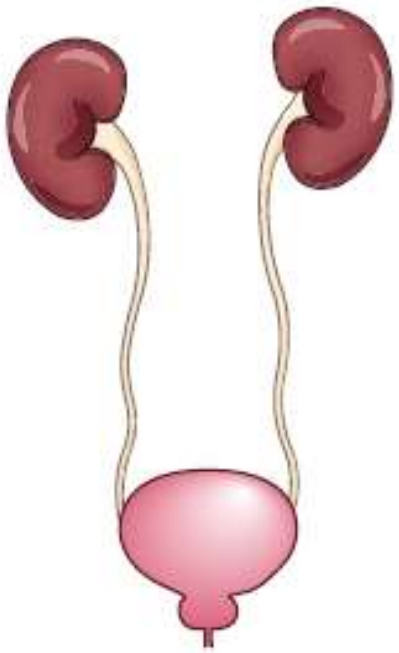
Lower Urinary Tract Obstruction



- PUV – 1:5000- 1:25,000
- Wide spectrum of prenatal presentation

- Other less common d/d
  - Anterior urethral valves
  - Urethral atresia
  - Urethral agenesis
  - Presacral mass

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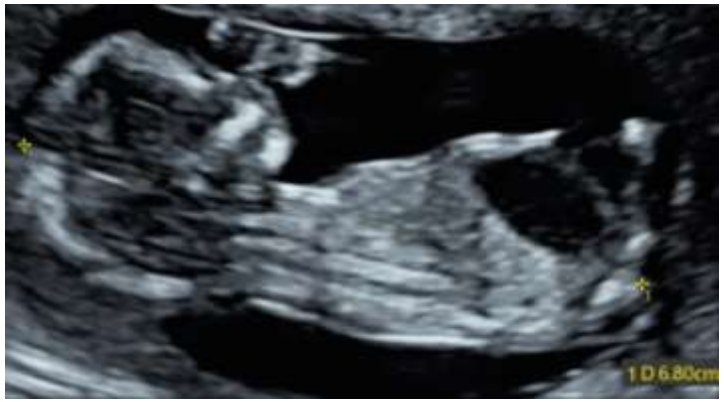
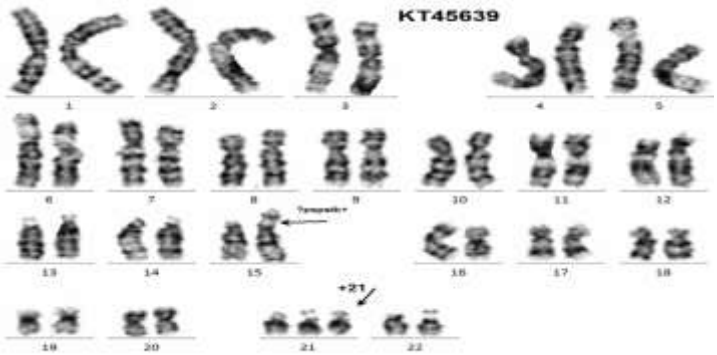
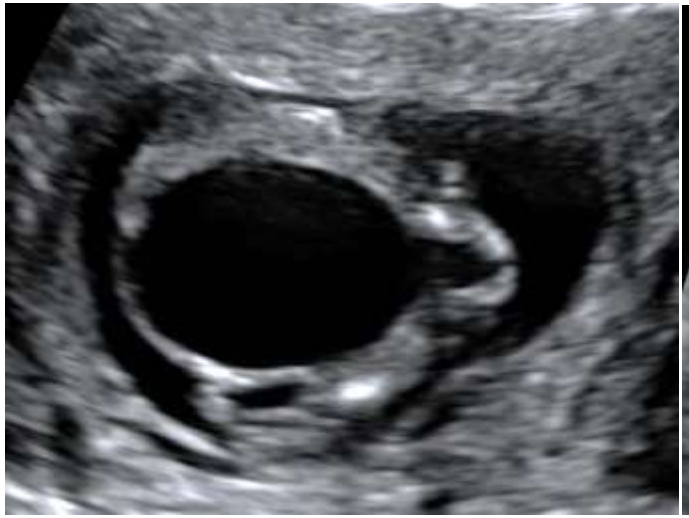
FIRST TRIMESTER LUTO

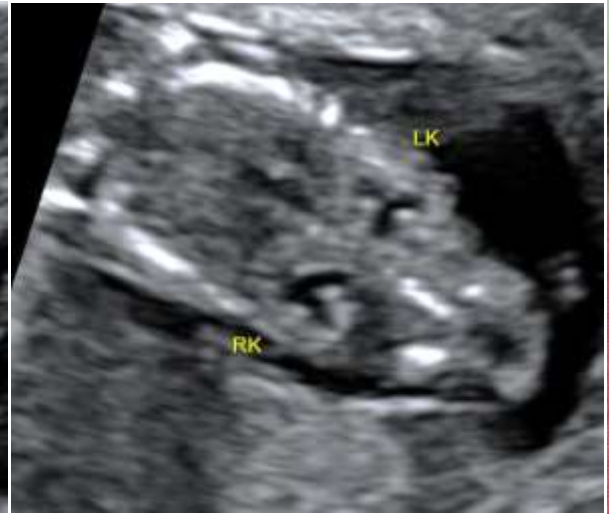
Bladder 7-15mm

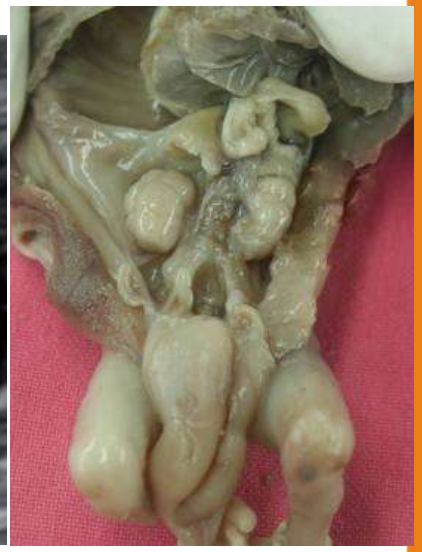




Bladder >15mm







MEGACYSTIS

7-15mm – Think aneuploidy

>15 mm – Think Obstruction

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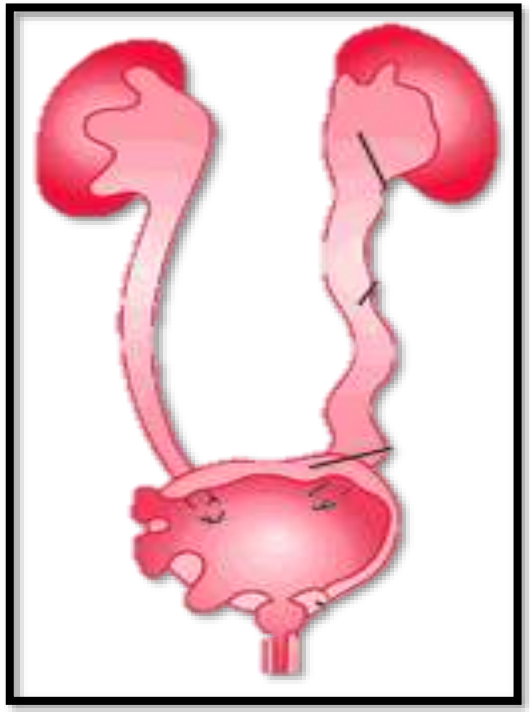
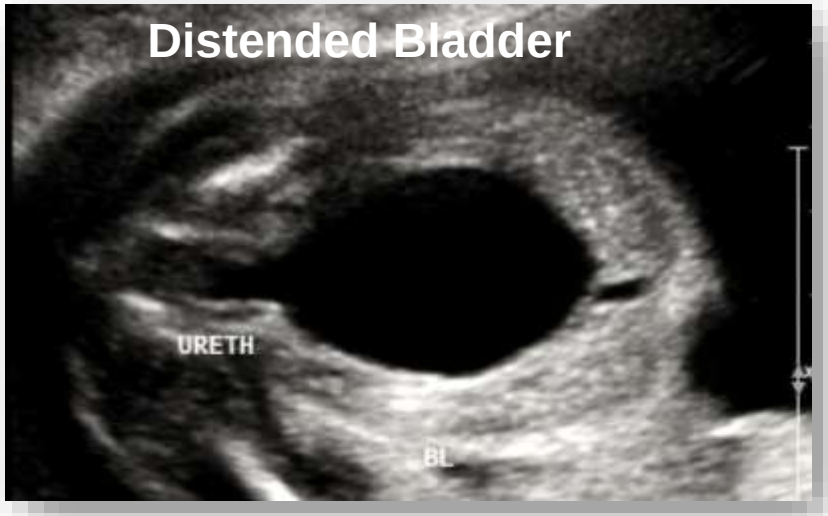
LUTO – 2ND, 3RD TRIMESTER PRESENTATION

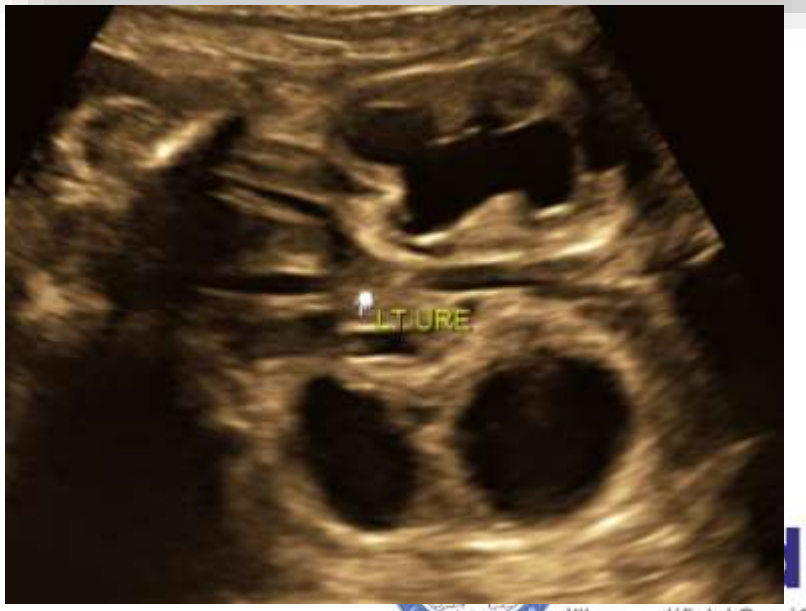
PRESENTATION:1



Male fetus - PUV

Dilated posterior urethra





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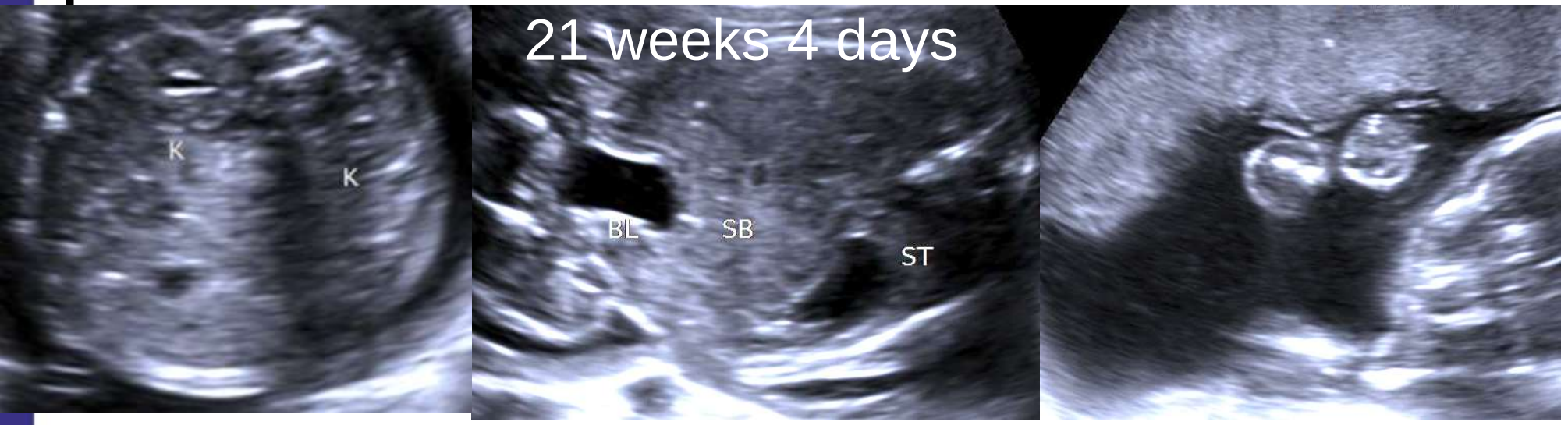
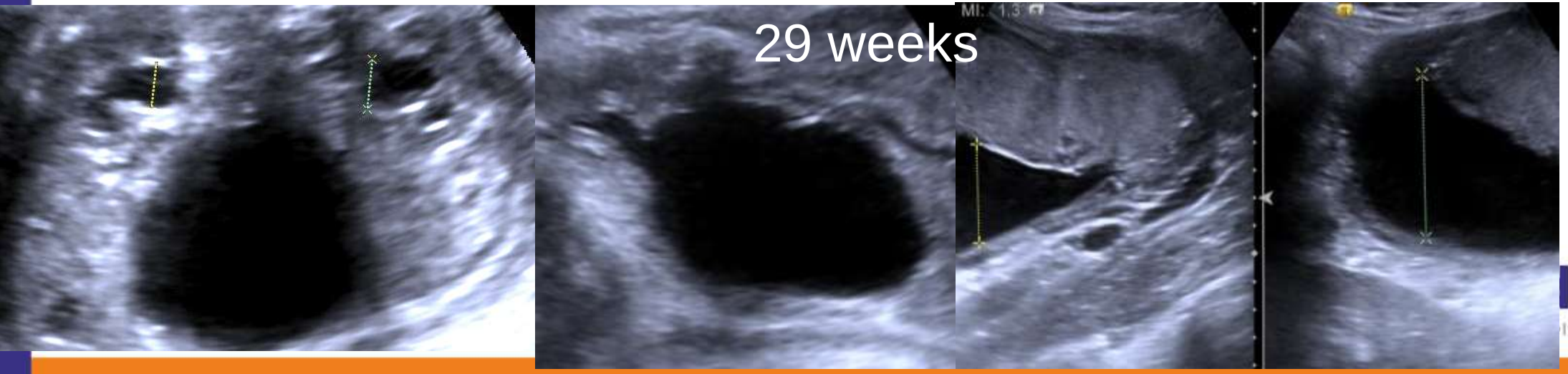
Evolving problem

20 years, Mrs C, Referred to us for routine target scan at 21 weeks 4 days





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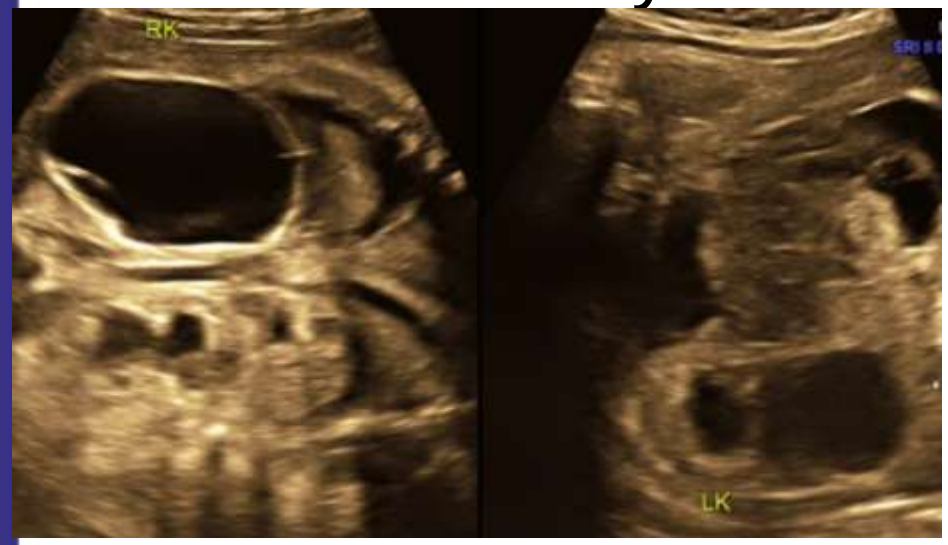
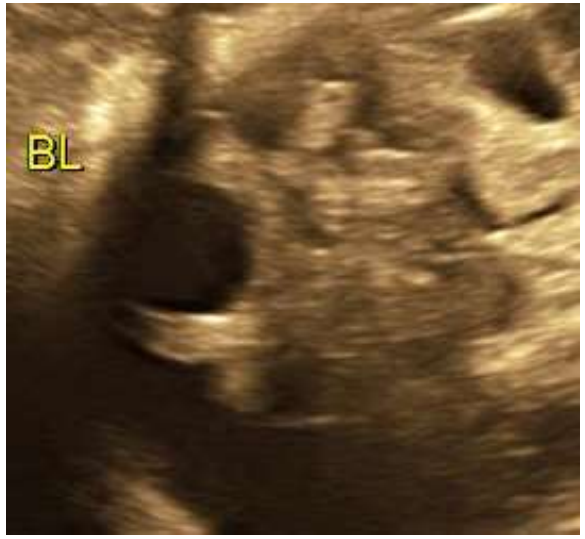
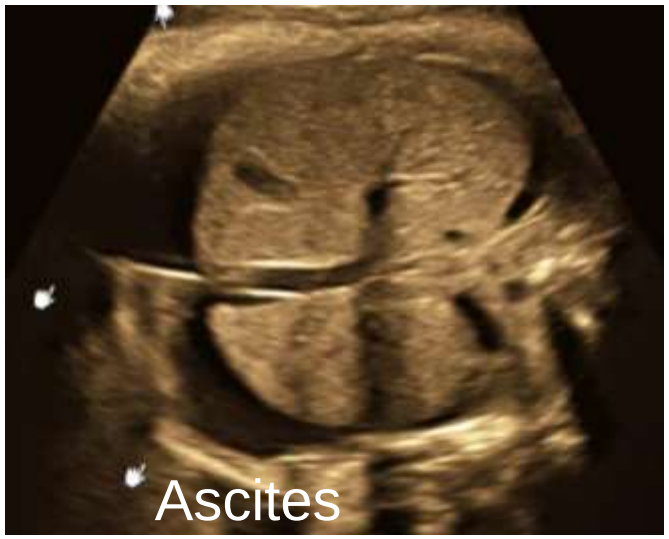
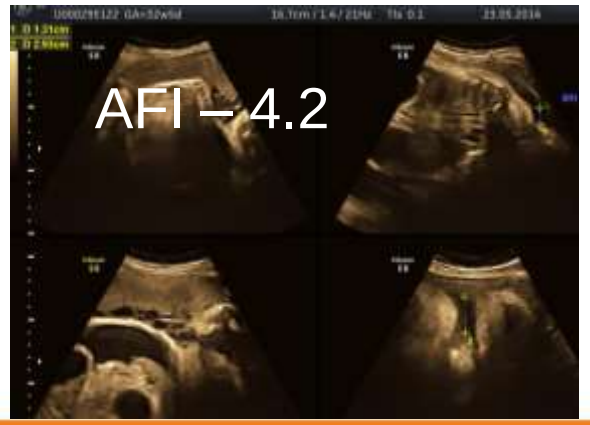
32 weeks 5 days







Bilateral hydroureteronephrosis
Right paranephric pseudocyst



EXPECTED OUTCOME ?

Delivered 3kgs male baby at 35 weeks

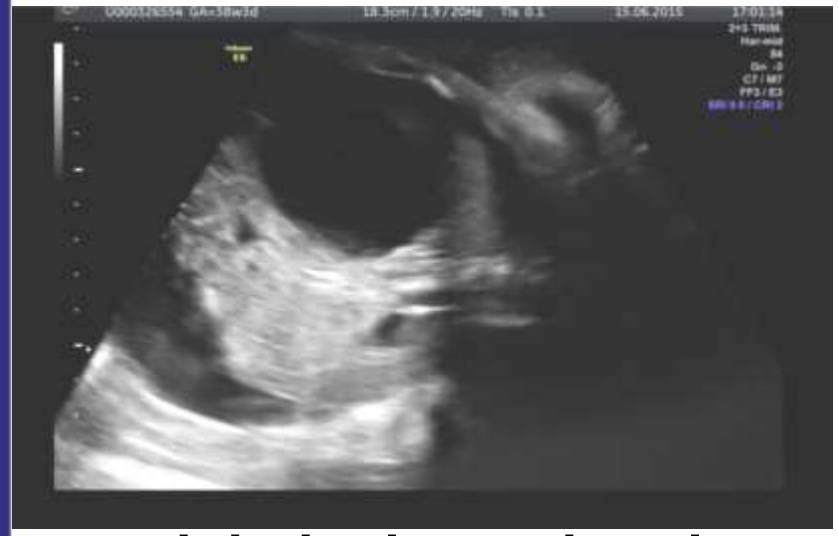
Baby was operated on day 5 of life for posterior urethral valve

Baby doing well @ 8 yrs!

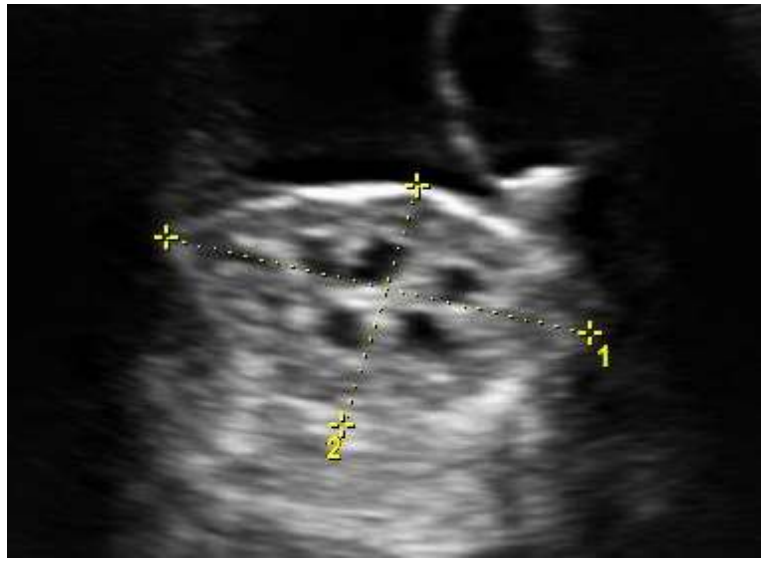
Did rupture protect the kidneys ?

Thick bladder

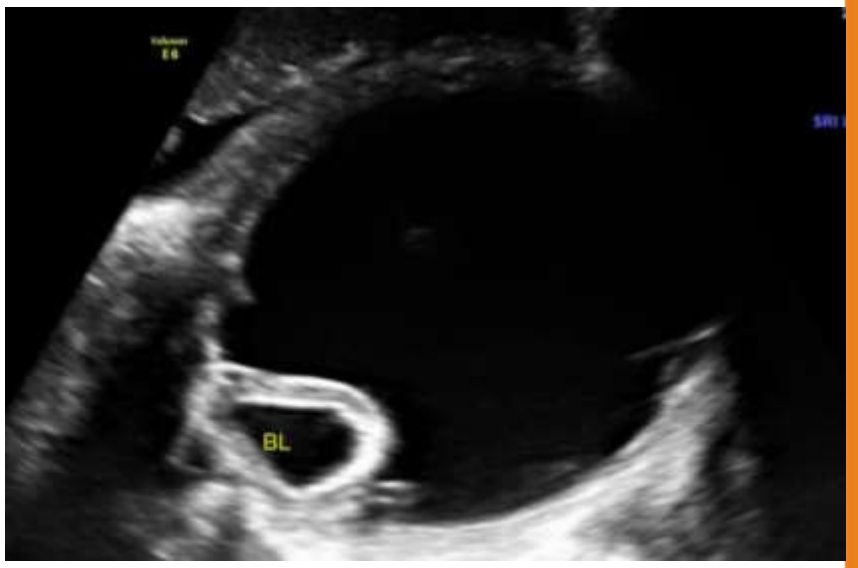
First referral@38 weeks
up to 28 wks reported normal



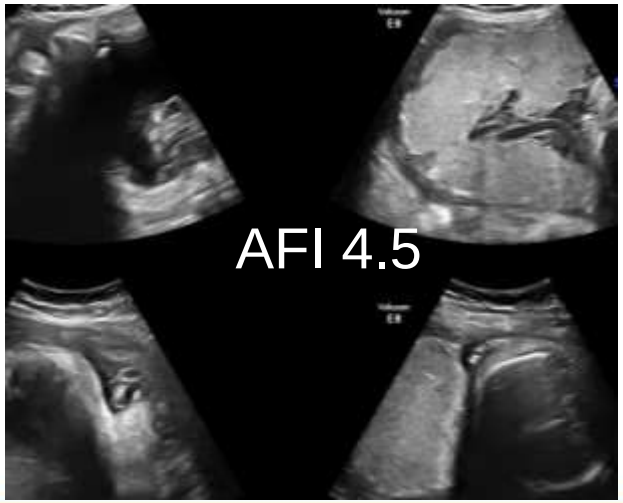
Right hydronephrosis with urinoma



Left mild hydronephrosis



Thickened bladder with ascites



ISSUES :
Distended abdomen – risk of dystocia at delivery

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OUTCOME

- Ascitic tap was done before LSCS and 210 ml of fluid drained
- Delivered male baby 2.9 kgs
- No cry at birth, shifted to NICU for 4 days
- PUV procedure done with primary fulgration
- 5 years Child has normal weight and developement

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What evidence says

Impact of prenatal urinomas in patients with posterior urethral valves and postnatal renal function

•*Stefanie Kleppe , Joachim Schmitt , Annegret Geipel , Ullrich Gembruch , Manfred Hansmann , Peter Bartmann , Ingo Franke and Axel Heep*

From the journal Journal of Perinatal Medicine

No association was found between renal function and urinoma formation in patients with PUV.

Fetal obstructive uropathy complicated by urinary ascites: outcome and prognostic value of fetal serum β-2-microglobulin

E. SPAGGIARI, S. DREUX, I. et al

Ultrasound Obstet Gynecol 2013; 41: 185–189

Fetal serum β-2-microglobulin predicts postnatal renal outcome in obstructive uropathy complicated by fetal urinary ascites, and occurrence of fetal ascites does not protect renal function

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Difficult situations in counselling

- Unilateral MCDK
- Unilateral empty renal fossa

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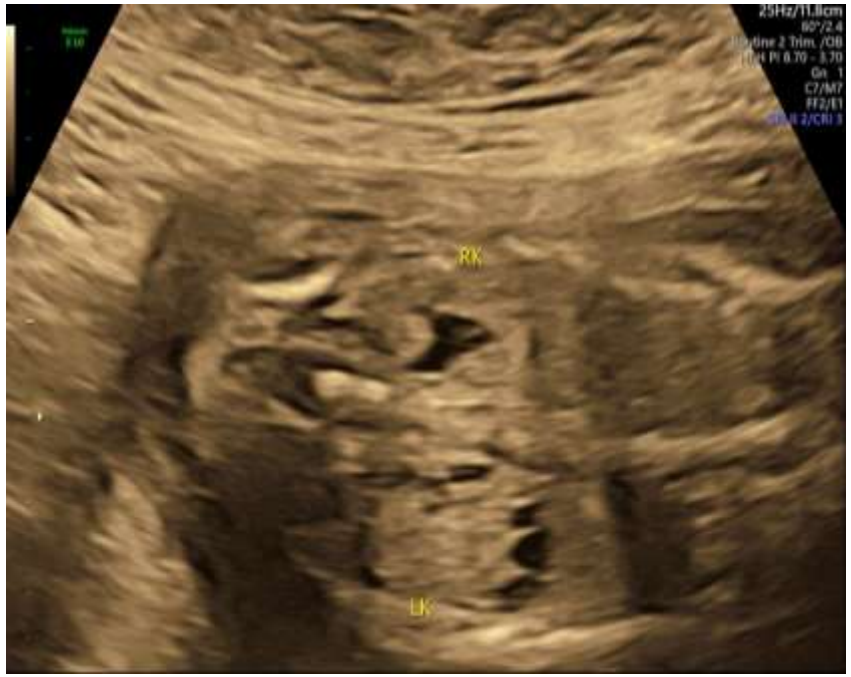
- Mrs. X Primigravida, married since 3 months , Non consanguinous marriage, RH positive pregnancy
- First trimester screening –LOW RISK . 20 WEEKS 4 DAYS



Right kidney 2.1 x 1.2 cm

Left Kidney enlarged 4.0 x 2.2 cm
Echogenic cortex with corticomedullary differentiation is indistinct ,with multiple cysts of varying sizes in renal parenchyma
Ureters not dilated
Liquor and bladder Normal
Rest of the fetus - OK

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- LEFT MULTICYSTIC DYSPLASTIC KIDNEY
- RIGHT KIDNEY AND BLADDER NORMAL

Pedigree highlights-Patient's niece diagnosed with left side reflux nephropathy / consulting ped nephro no surgery done / doing well

Suggested:

1. Direct testing by amniocentesis for QFPCR / Chromosomal micoarray/ DNA store.
2. Parental KUB scan
3. Repeat scan after 4 weeks for reassessment
4. Postnatal KUB

Patient opted for Amniocentesis -Normal QFPCR

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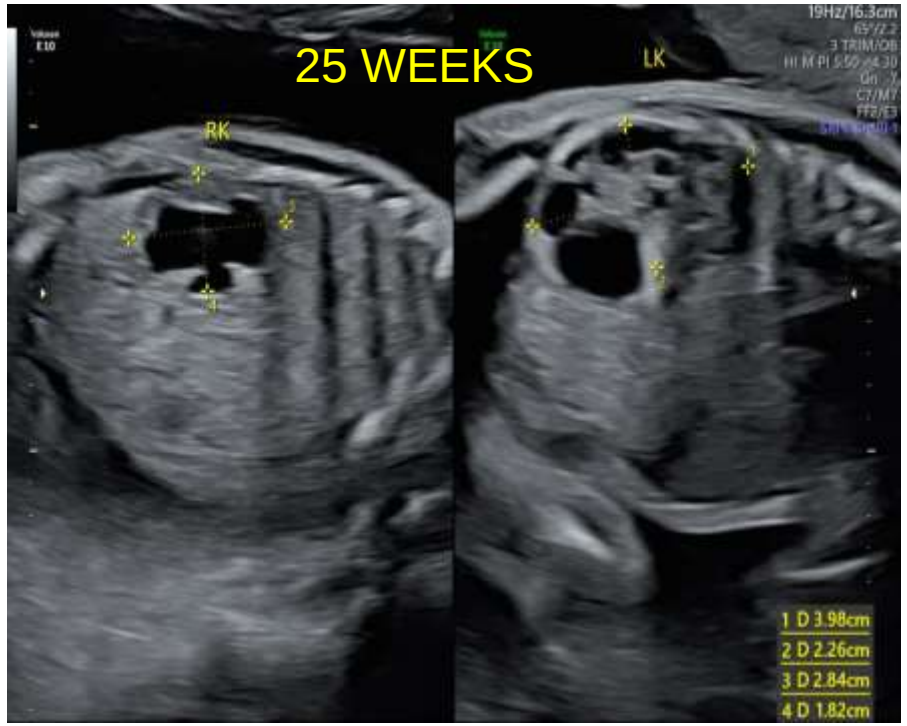
Patient consulted Pediatric Nephrologist

Adviced follow up after 4 weeks



Liquor normal,Bladder appeared normal

Right kidney2.8 x 1.8 cm

Cortex appeared Thinned out,renal pelvis appears dilated measuring 8.4 mm ,Central and peripheral calyceal dilatation seen ,Ureter not dilated

Left Kidney4.0 x 2.3 cm

MCDK

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LEFT MULTICYSTIC DYSPLASTIC KIDNEY

RIGHT RENAL PELVICALIECTASIS - URINARY TRACT DILATATION CLASSIFICATION UTD A2-3

BLADDER SEEN , LIQUOR NORMAL-Indication of preserved renal function .

Amnio – CMA normal

Recounselling about evolving change in Right kidney and hidden anomalies

couple decided to terminate pregnancy

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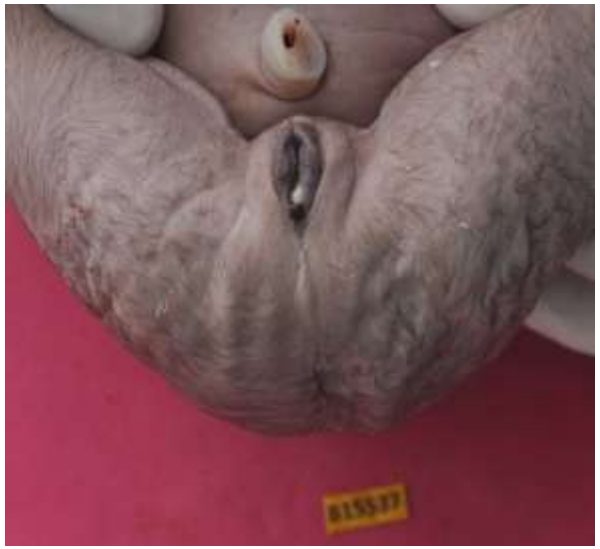
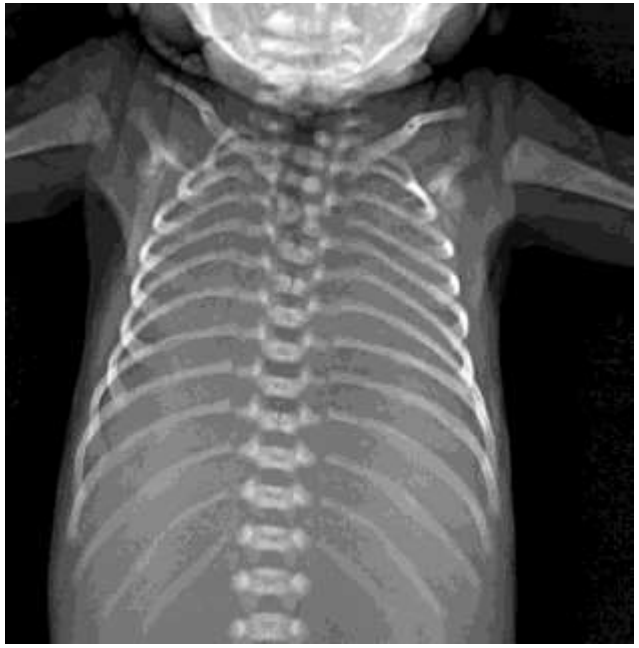
Patient opted for termination of pregnancy sent fetus for AUTOPSY



Imperforate anus

RK:HYDRONEPHROSIS WITH DILATED RIGHT URETER

LK:MULTICYSTIC DYSPLASTIC KIDNEY WITH HYPOPLASTIC URETER



Vertebral anomaly

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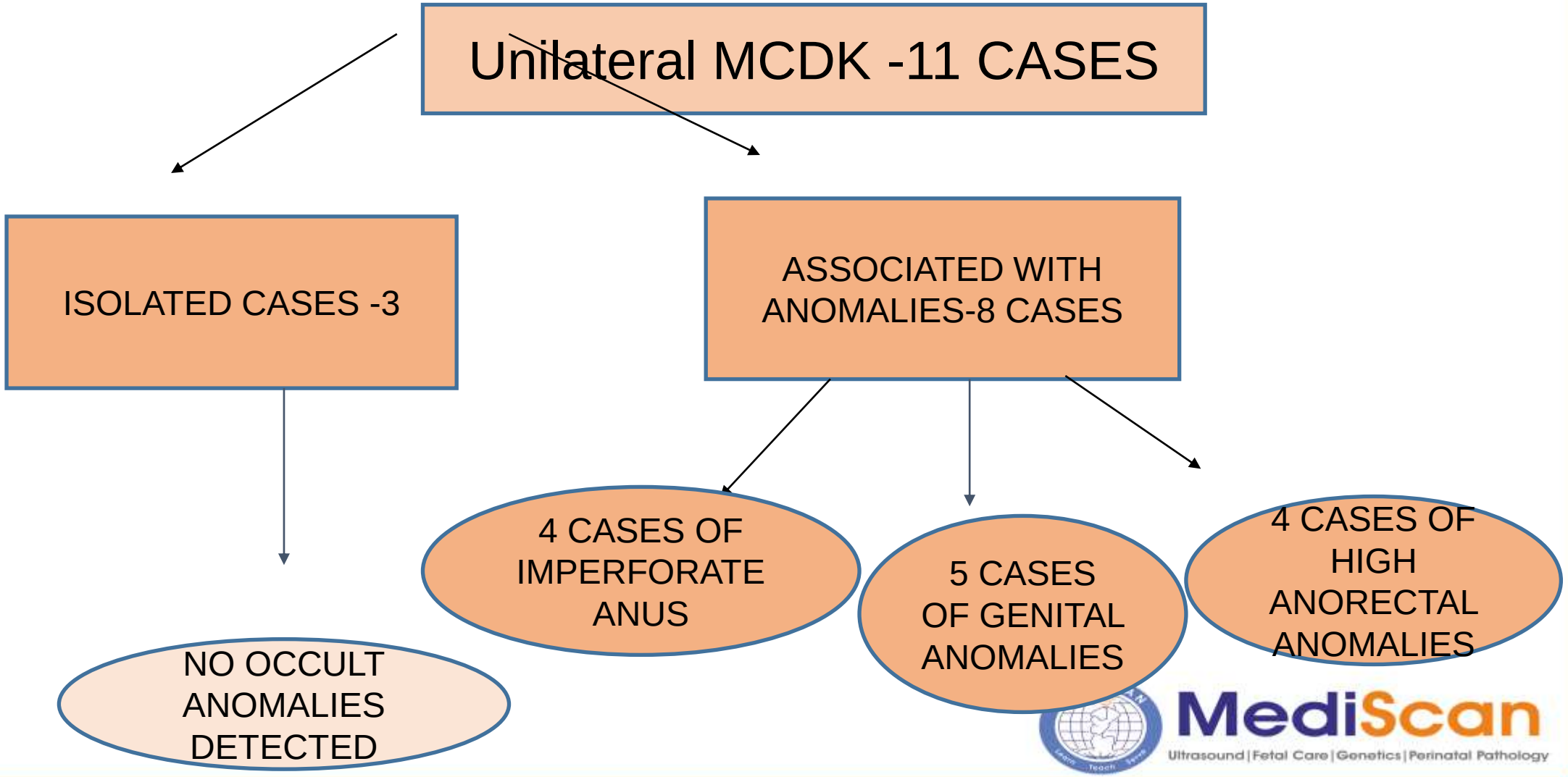
RETOSPECTIVE STUDY OF UNILATERAL MCDK diagnosed in the department of perinatal pathology in Mediscan from 2018 to 2023



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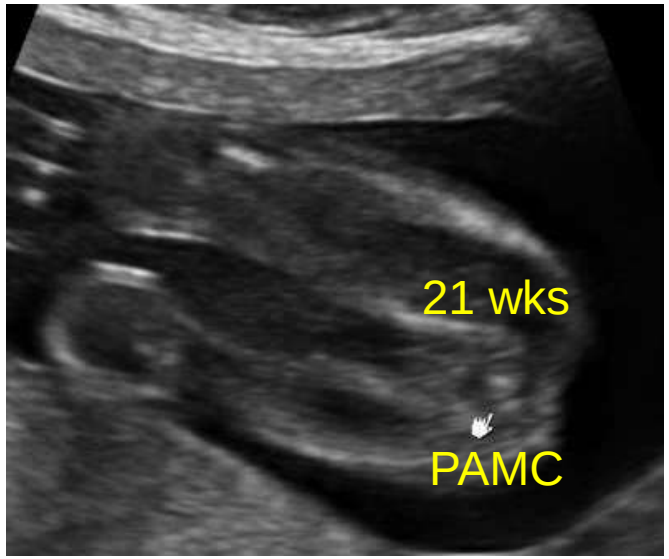
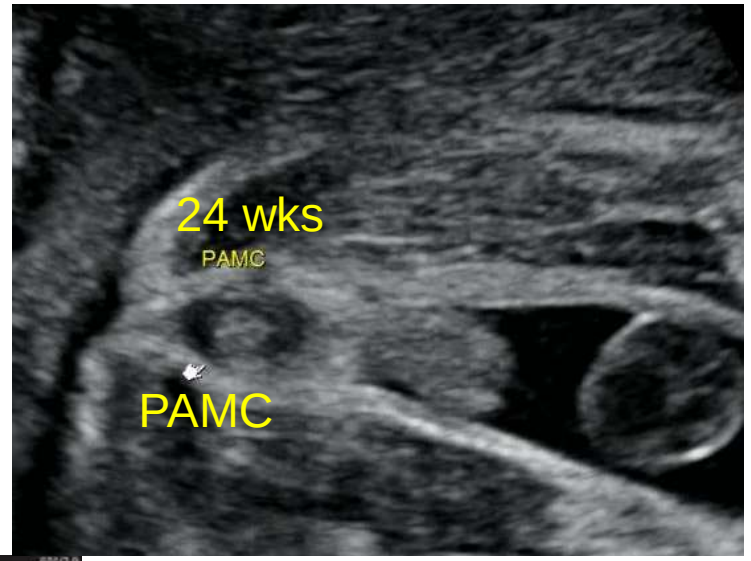
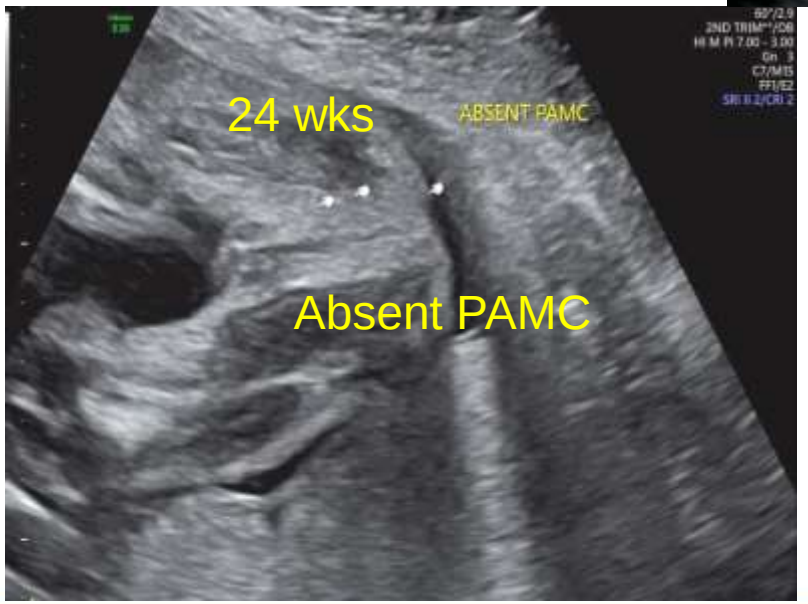
Absent PAMC – Marker of anorectal anomaly has variable sensititivity







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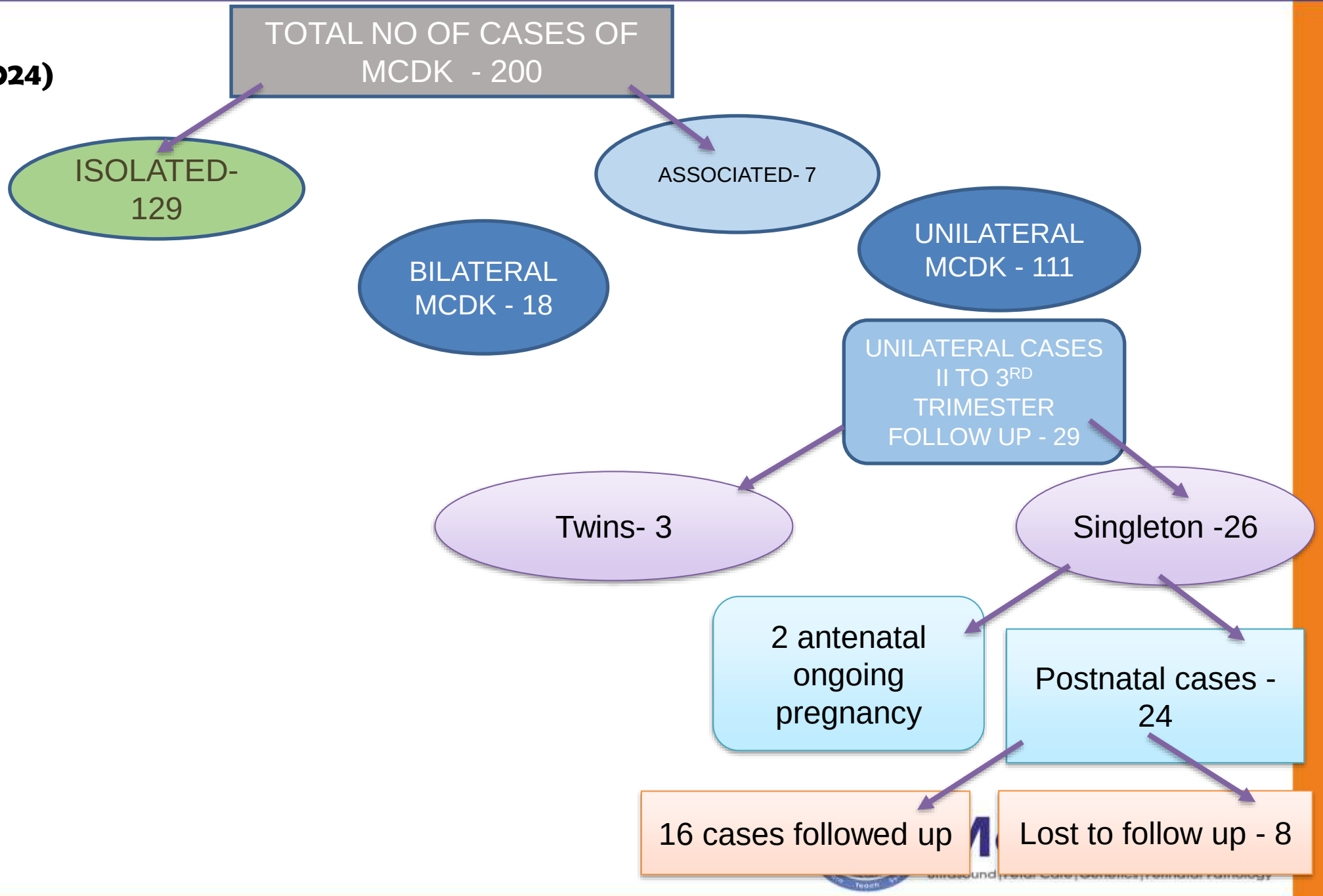
MEDISCAN DATA
(JAN 2022 – DEC 2024)



Prenatal diagnosis and outcome of unilateral multicystic kidney

Gurcan Turkyilmaz, Bilal Cetin, Emircan Erturk, Tugba Sivrikoz, Ibrahim Kalelioglu, Recep Has, Atıl Yuksel, Tayfun Oktar & Orhan Ziylan

METHODS:

•**Study Design:** Retrospective review of **144 fetuses** diagnosed with unilateral MCDK from **2010–2017**.

•**Assessment:** Prenatal ultrasonography was used for diagnosis and follow-up. Postnatal evaluations included renal imaging and functional testing.

•**Genetic Testing:** Karyotyping was performed in 24 cases (16.6%); chromosomal microarray in 2 cases.

Anomaly/Outcome	Number of Cases
Total Contralateral Anomalies	38
- Vesicoureteral Reflux (VUR)	20
- Ureteropelvic Junction Obstruction	12
- Duplex Kidney	4
- Renal Agenesis	2
- Ureterocele	7

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This is to certify that this dissertation titled **"Perinatal outcomes in fetuses with unilateral empty renal fossa"** is a Bonafide record of work done by Dr. **SIDDHI SHREE DIXIT,** in the department of Fetal Medicine, MediScan Systems, Chennai- 600004, under my guidance and supervision, during the period of he fellowship for the award of Fellowship in Fetal Medicine, July 2022 – June 2025.

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PERINATAL OUTCOMES IN FETUSES WITH UNILATERAL EMPTY RENAL FOSSA

Study Design: Retrospective cohort study

Study Period: 6 years from 2018- 2024

- **Primary objective** -To evaluate the postnatal outcomes of fetuses diagnosed with unilateral empty renal fossa on prenatal ultrasound
- **Secondary objective-**
- To identify the associated structural anomalies in fetuses with a prenatal diagnosis of unilateral empty renal fossa and assess the spectrum of diagnoses.
- To determine the feasibility and accuracy of first and second trimester ultrasound in detecting unilateral empty renal fossa and its correlation with second/third trimester findings

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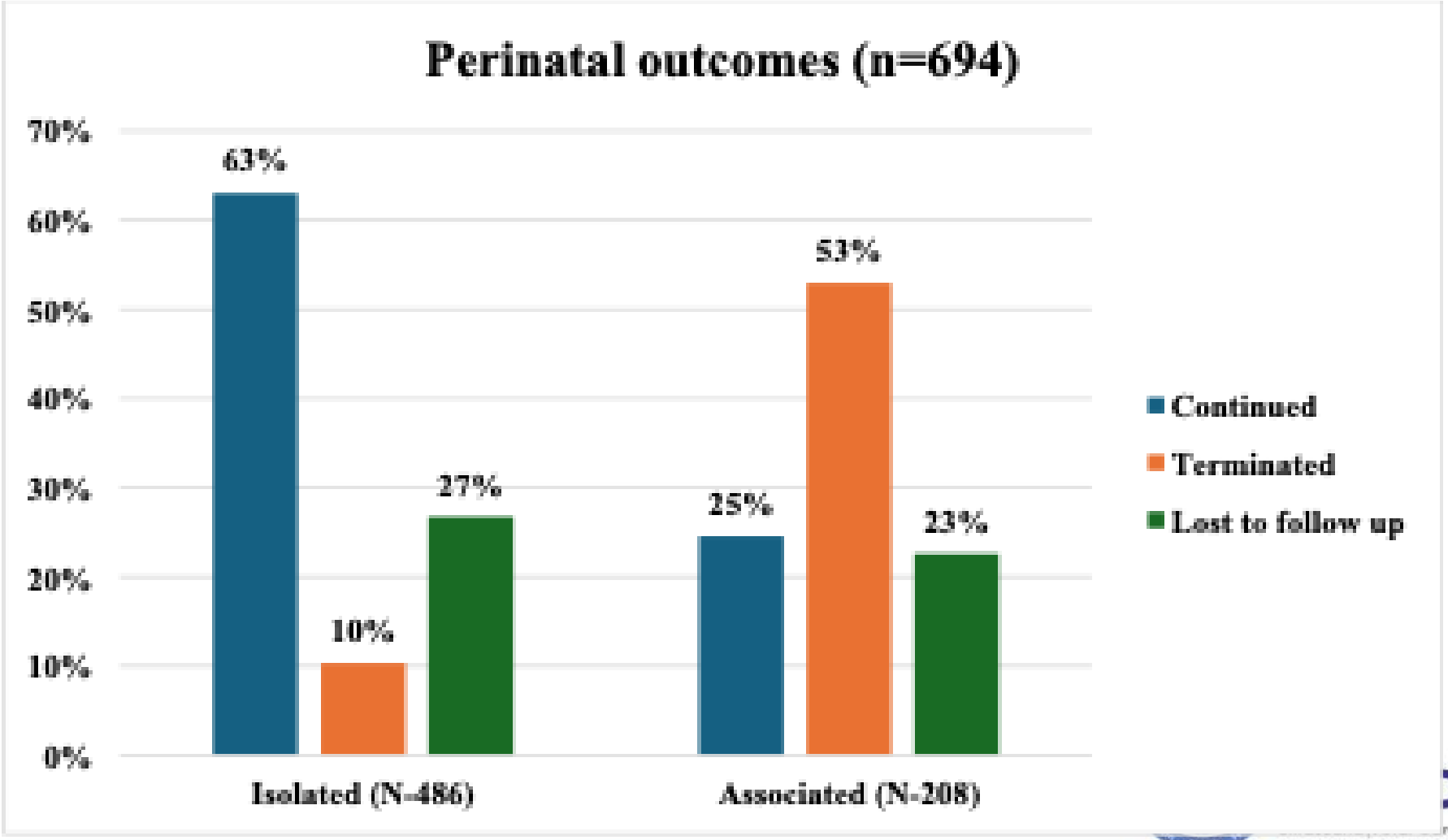
- **Outcomes in groups with isolated and associated anomalies**



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ISOLATED CASES THAT UNDERWENT TOP

- 49/486 isolated cases underwent TOP -10%. Of these 8 were sent for autopsy.
- **In the 8 cases sent for autopsy**
- **Prenatal USG findings:**
  - 5 –CAKUT abnormality in contralateral normally placed kidney (MCDK or UTD)
  - 2- Normal contralateral kidney
  - 1-Evolving FGR in second trimester follow up visit
- **On autopsy –**
  - occult anomalies were noted in 7 out of the 8 cases- anal anomaly in 6 and genital anomaly (bicornuate uterus) in 1.
  - Only 1 case had no occult anomaly. This baby prenatally had solitary MCDK.

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HNF1B alterations associated with congenital anomalies of the kidney and urinary tract

Makiko Nakayama · Kandai Nozu · Yuki Goto
Koichi Kamei · Shuichi Ito · Hidenori Sato · Mitsuru Emi
Koichi Nakanishi · Shigeru Tsuchiya · Kazumoto Iijima

HNF1B mutations broad spectrum of renal diseases-→
1)renal cystic malformations
2)MCDK
3)Glomerulocystic kidney disease
4) Autosomal dominant tubulointerstitial kidney disease

- HNF1B is a DNA-binding transcription factor essential for normal kidney development-located in chromosome 17q12.

- Unilateral MCDK with normal function may still carry HNF1B deletions

- Recurrent 17q12 microdeletions were found to involve HNF1B and 14 other genes

- But HNF1B is likely the primary driver of renal phenotype.

Genetic testing- Basic KT/CNV/WES decided according to the anomaly

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Fetal intervention for LUTO

- Ruano Classification LUTO
- Stage II / Stage III
- Candidates for Intervention

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RENAL FUNCTION TESTING

- 2 TAPS of bladder : 48 hrs after 1st
- Fetal urine Biochemistry
- Fetal Urine beta-2 microglobulin

Urinary component	Favorable
Sodium (Na)	Less than 100 mEq per liter
Chloride (Cl)	Less than 90 mEq per liter
Osmolarity (Osm)	Less than 210 mEq per liter
Calcium (Ca)	Less than 2 mmol per liter
Beta-2 microglobulin	Less than 2 mg per liter

Proteomics analysis of fetal urine in fetuses with PUV (used as the prototypic LUTO)[46] showed a 12-peptide signature (12PUV), which is specifically expressed in fetuses who would develop ESKD before the age of 2 years[46]

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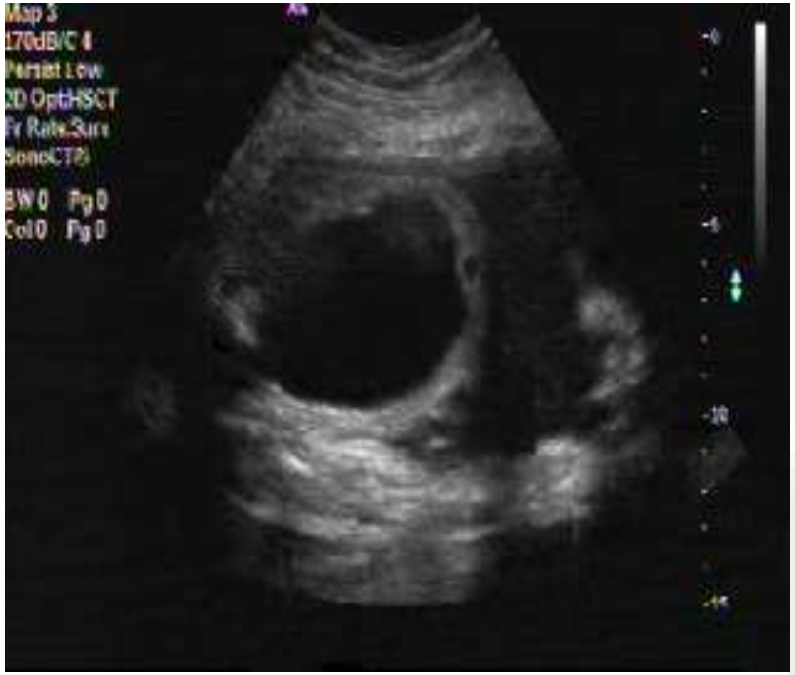
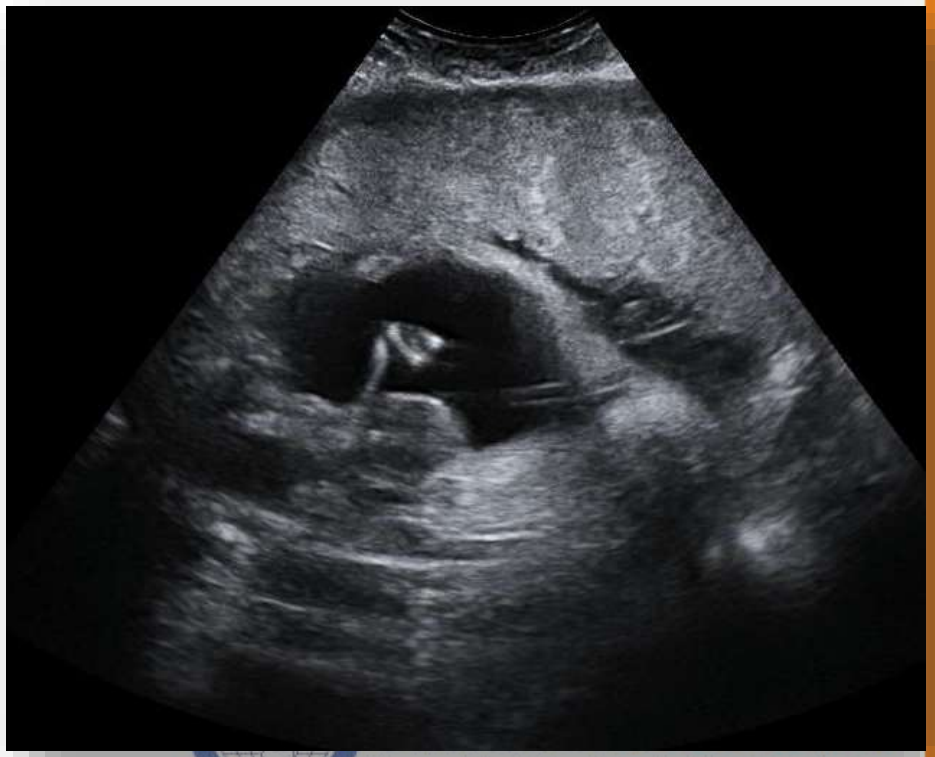
Fetal Intervention

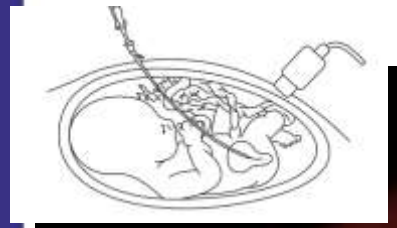


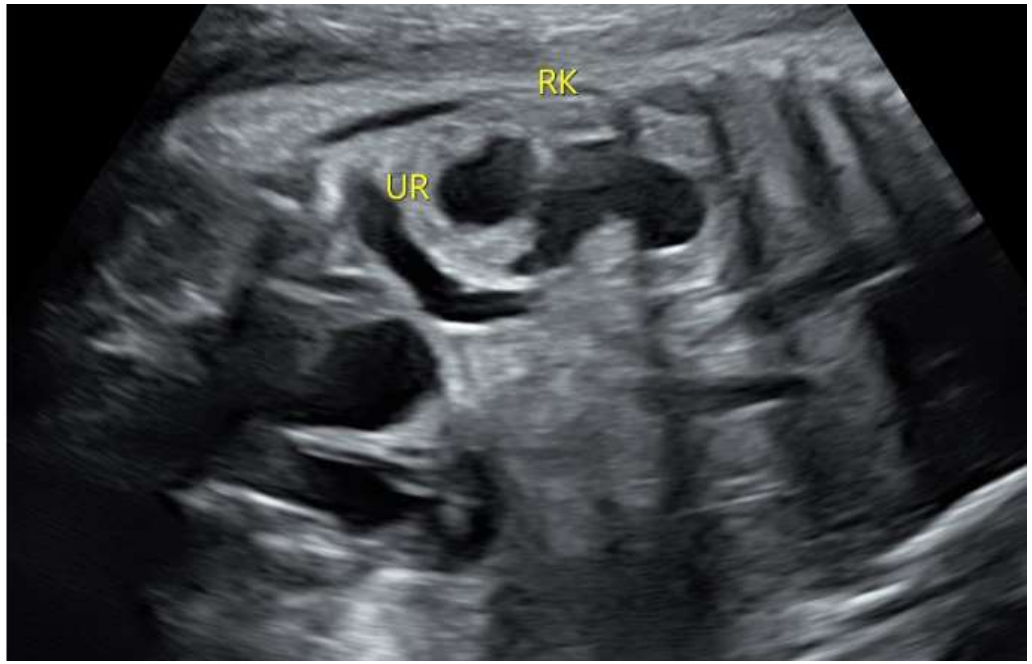
VA Shunt
Displacement
Migration
40%



- At 15 years
- Creatinine 1.1
- DOING WELL

In-Utero Laser Fenestration of Posterior Urethral Valves

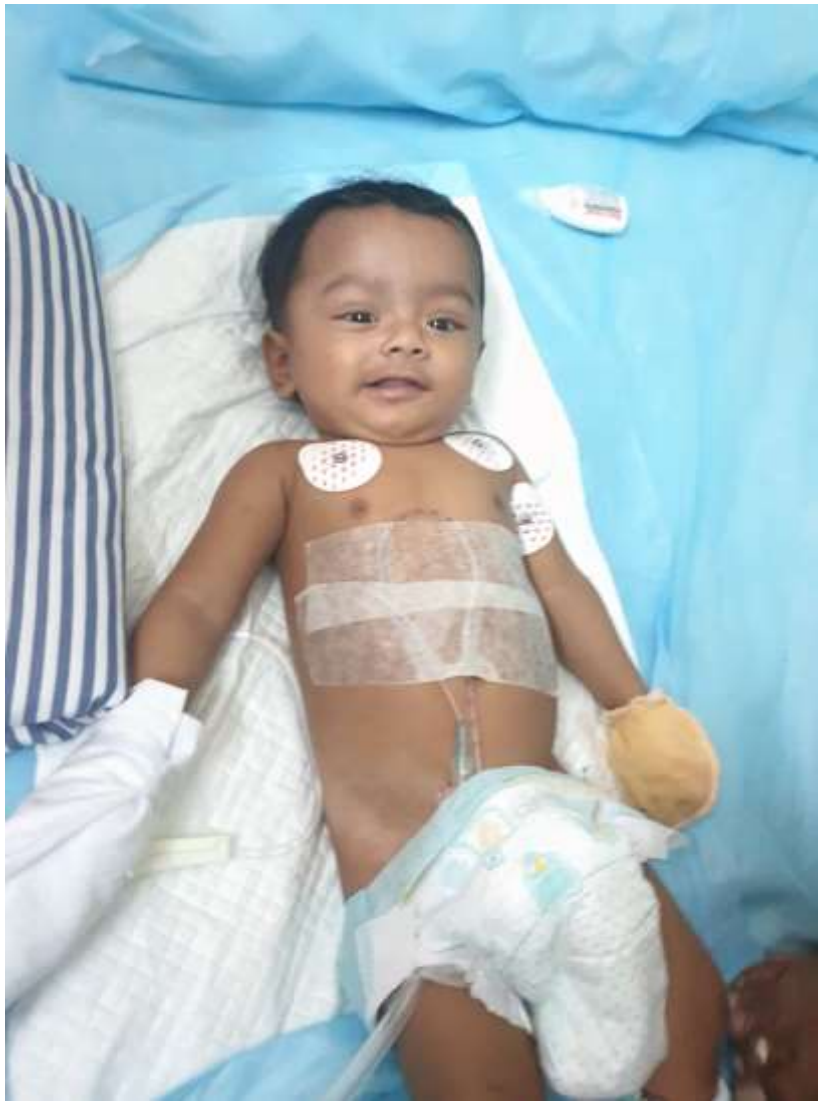




- 31 weeks - Bladder normal
- Filling and emptying seen
- Liquor normal
- Baby delivered at 36 weeks
- Postnatal fulguration done on Day 3

Followup

- Redo fulguration at 6 months
- Now child is 11 months old
- Voiding well
- No postvoid residue on ultrasound
- S.Creatinine 0.5
- Spot P/Cr = .41



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Conclusion

- Urinary system can be examined from 14 wks best time would be 20wks
- When a problem is suspected extended examination best possible transducer to be done. Take a detailed history and pedigree
- Bilateral Cystic kidneys can be syndromic extrarenal anomalies to be looked in scan /postnatal/autopsy
- Genetic testing to be offered
- Obstructive uropathy serial monitoring
- Interact with pediatric nephrologist / urologist
- Be supportive to the family.

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Robot Assistance
THAN
K YOU