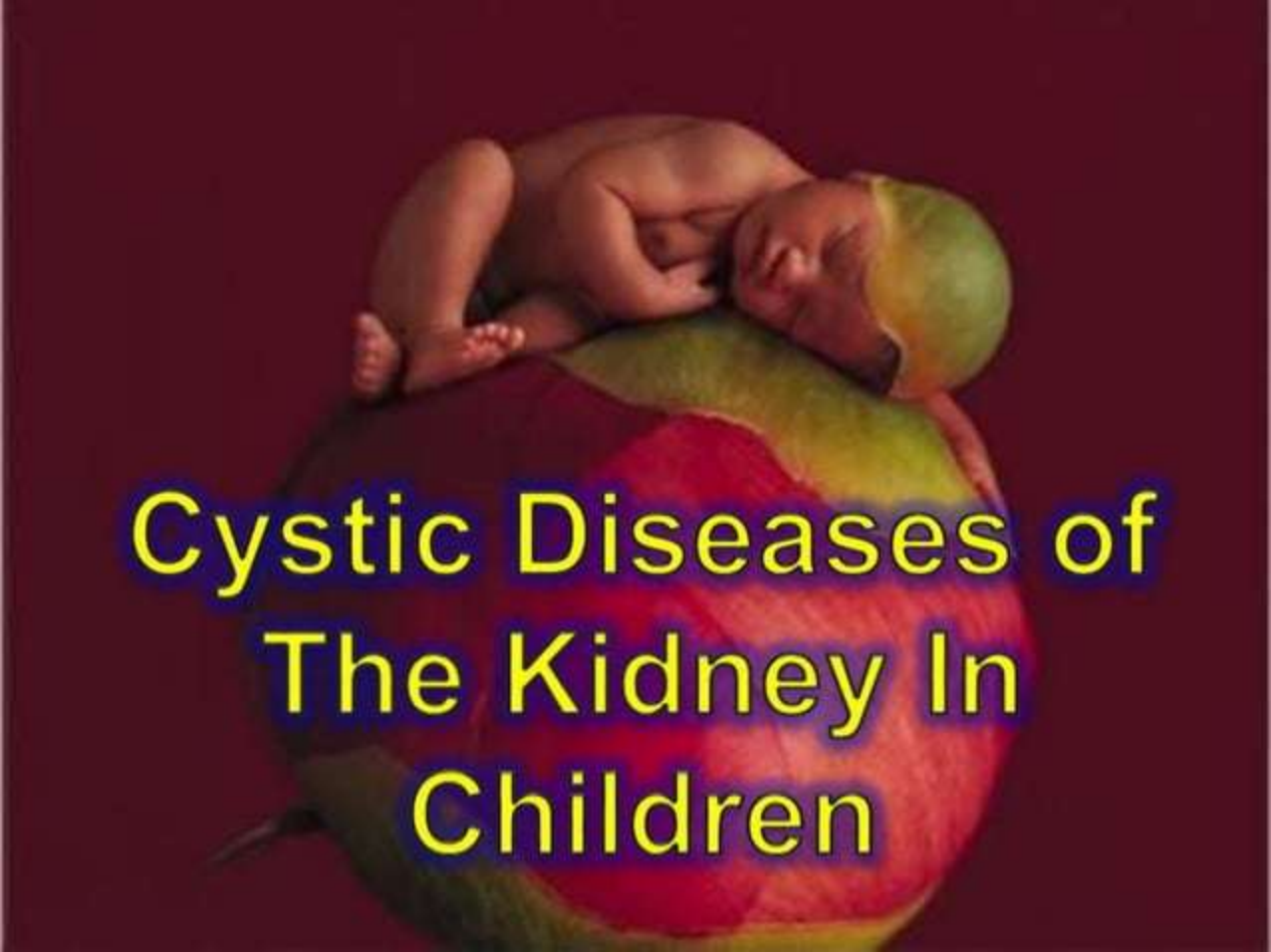

Cystic Diseases of
The Kidney In
Children

Cystic Diseases of The Kidney In Children

- Cystic diseases of the kidney in infants and children include a large number of clinicopathological disorders that may be inherited or sporadic in origin.

Cystic Diseases of The Kidney In Children

Genetic:

1. Autosomal recessive polycystic kidney disease (ARPKD)
2. Autosomal dominant polycystic kidney disease (ADPKD)
3. Juvenile nephronophthisis and medullary cystic disease complex
4. Glomerulocystic kidney disease

Non-genetic:

1. Simple cysts
2. Multicystic dysplastic kidney (MCDK)
3. Multilocular cysts
4. Acquired renal cysts
5. Caliceal cysts and diverticula
6. Cystic dysplastic kidney

Cystic Diseases of The Kidney In Children

- Some of these lesions can be detected on antenatal ultrasound.

Cystic Diseases of The Kidney In Children

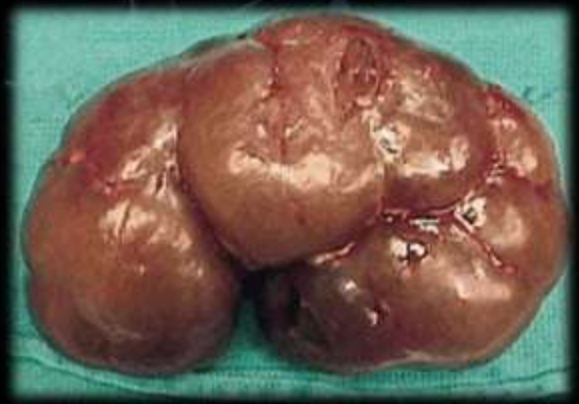
- Ultrasound is the primary imaging modality in the diagnosis of most cystic renal diseases, and recourse to other modalities is rarely required.

Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- 1 in 50 000 of the normal population.
- Recessive mode of inheritance.
- Generalised dilatation of the collecting tubules affecting both kidneys.

Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- The kidneys are filled with tiny little cysts. Occurrence of macrocysts is rare.

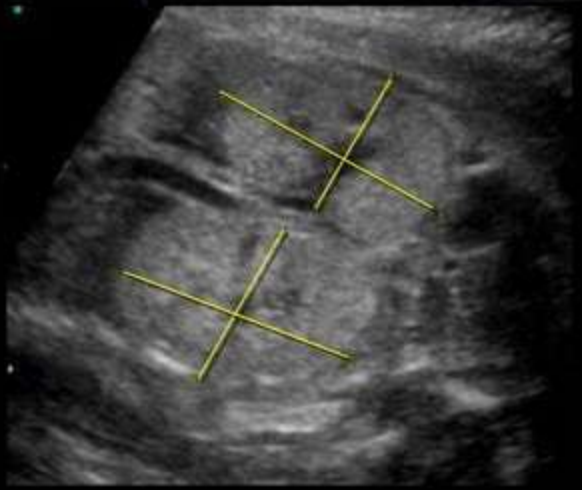


Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- ***ARPKD appears to be a spectrum of abnormality, with the renal disease and hepatic fibrosis varying inversely:***
- Children presenting at birth or in the neonatal period have predominantly renal disease.
- Those diagnosed later in childhood or adolescence, liver disease with hepatomegaly and splenomegaly secondary to portal hypertension predominates, with comparatively milder renal involvement.

Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- ARPKD may be suspected on an antenatal ultrasound when markedly enlarged echogenic kidneys, without visible cysts, are seen in the third trimester.
- Oligohydramnios may also be evident.

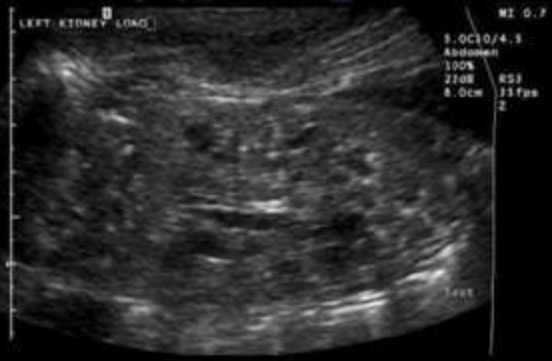


Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- Presentation with respiratory distress from pulmonary hypoplasia is commonly encountered in the neonatal period as a result of oligohydramnios.

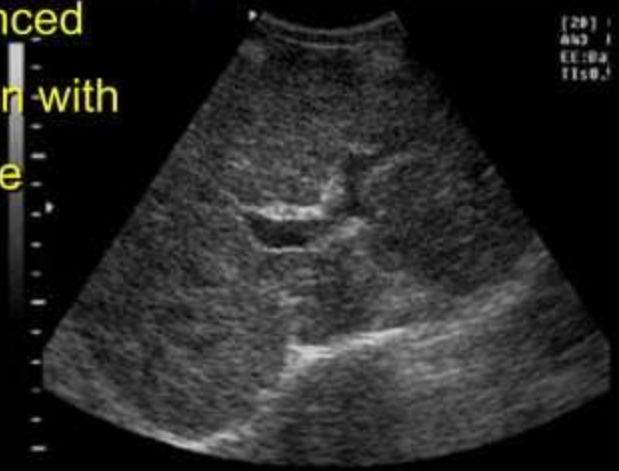
Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- Bilaterally enlarged and diffusely echogenic kidneys with maintained reniform shape, without corticomedullary differentiation. Often with a sonolucent rim.
- The kidneys appear bright.
- Both kidneys are involved.



Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- In older children with advanced disease, portal hypertension with oesophageal varices maybe seen.



Autosomal Dominant Polycystic Kidney Disease (ADPKD)

- ADPKD is more common than ARPKD, occurring in approximately 1 in 1000 of the population.
- It is a disease of adults, usually manifesting in the third to fourth decade of life.

Autosomal Dominant Polycystic Kidney Disease (ADPKD)

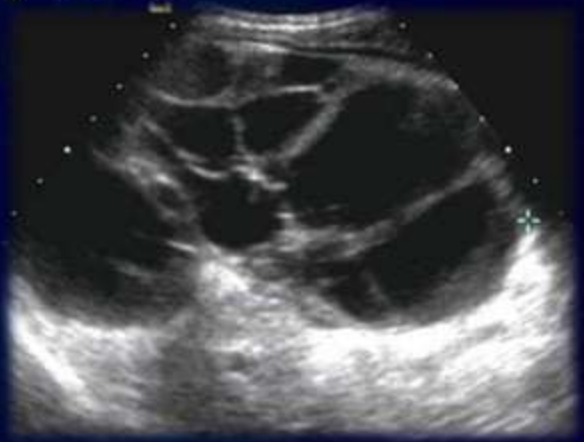
- A positive family history or the affection of one of the parents is one of the major criteria for the diagnosis of ADPKD in childhood.

Autosomal Dominant Polycystic Kidney Disease (ADPKD)

- Hepatic and pancreatic cysts may sometimes be found, but in contrast to ARPKD there is no significant periportal fibrosis.

Autosomal Dominant Polycystic Kidney Disease (ADPKD)

- Enlarged often echogenic kidney with varying number of cysts and normal intervening renal parenchyma.

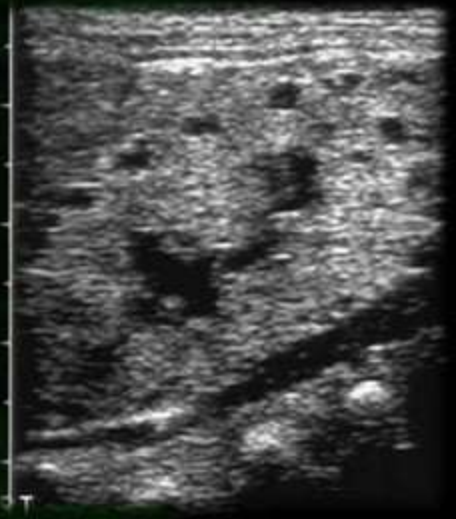


Glomerulocystic Kidneys

- Glomerulocystic kidneys is an inherited disorder that is typically sporadic without any familial pattern.
- Pathologically it is characterized by cystic dilatation of Bowman's space of the glomeruli.

Glomerulocystic Kidneys

- The ultrasound appearance is often indistinguishable from ARPKD. The kidney is usually echogenic but may contain cysts of variable size.
- The diagnosis is made by clinical symptoms, history, biopsy and ultrasound.

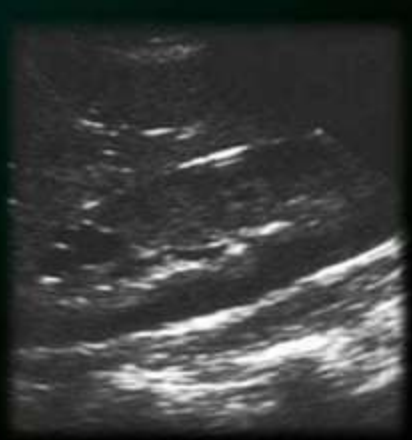


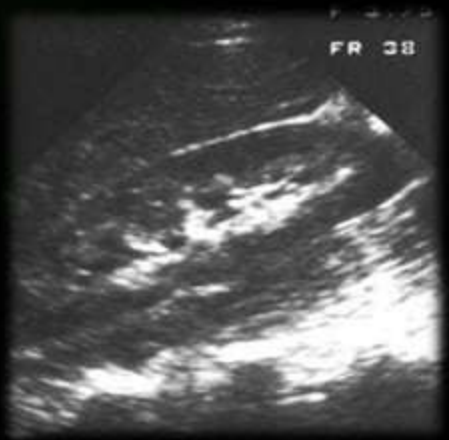
Juvenile Nephronophthisis

- Juvenile nephronophthisis is a recessive inherited disorder.
- Clinically, polyuria, polydypsia, salt wasting and progressive uraemia

Juvenile Nephronophthisis

- Ultrasound demonstrates loss of corticomedullary differentiation, with increased echogenicity and a variable number of cysts in the medulla or at the corticomedullary junction.





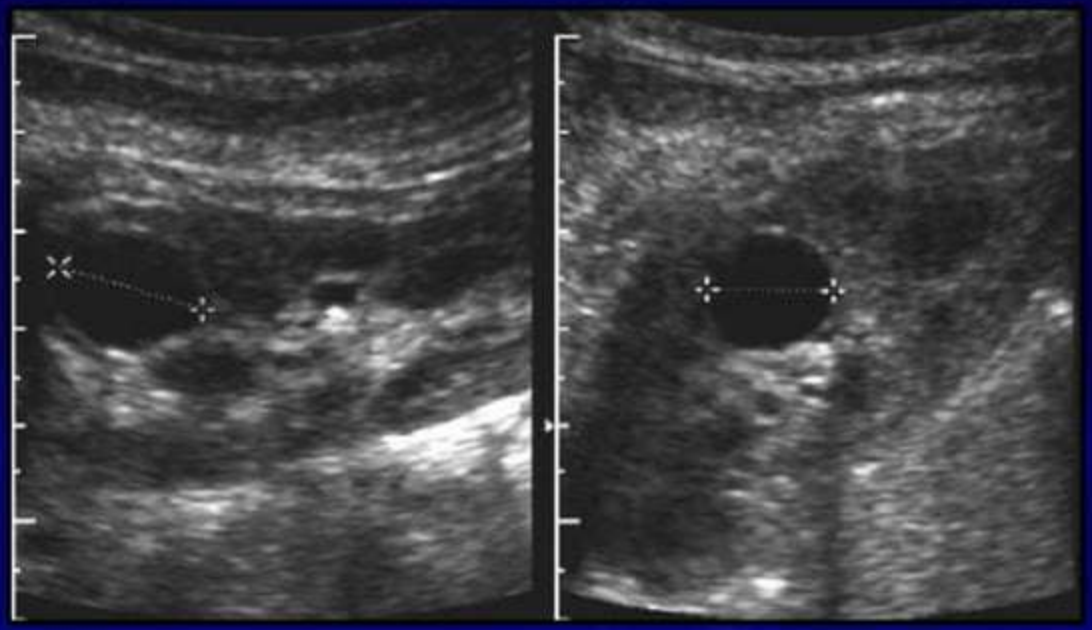
Juvenile Nephronophthisis

- The characteristic ultrasound feature of juvenile nephronophthisis is the presence of an echogenic but usually normal sized kidney even when the patient has an end-stage renal failure

Simple Cysts

- Simple cysts are rare in children.
- The incidence of cysts increases with the age of the child.
- They are usually detected incidentally and can be either unilateral or bilateral.

Simple Cysts

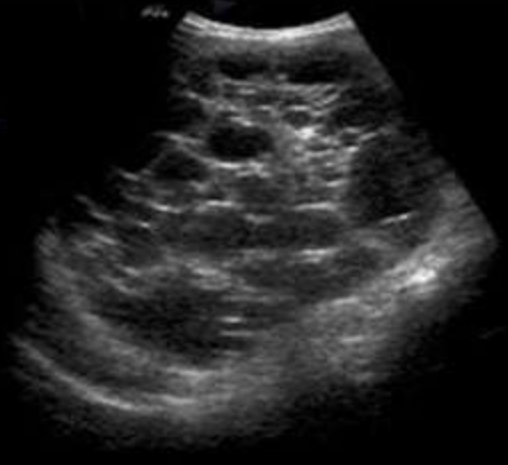


Multilocular Cystic Nephroma

- Multilocular cystic nephroma is an uncommon benign neoplasm.
- It occurs in children aged 3–5 years with increased incidence in boys.

Multilocular Cystic Nephroma

- On ultrasound it is characterized by the presence of cysts that have a well-defined capsule and a variable number of septa.
- There is no normal renal tissue.



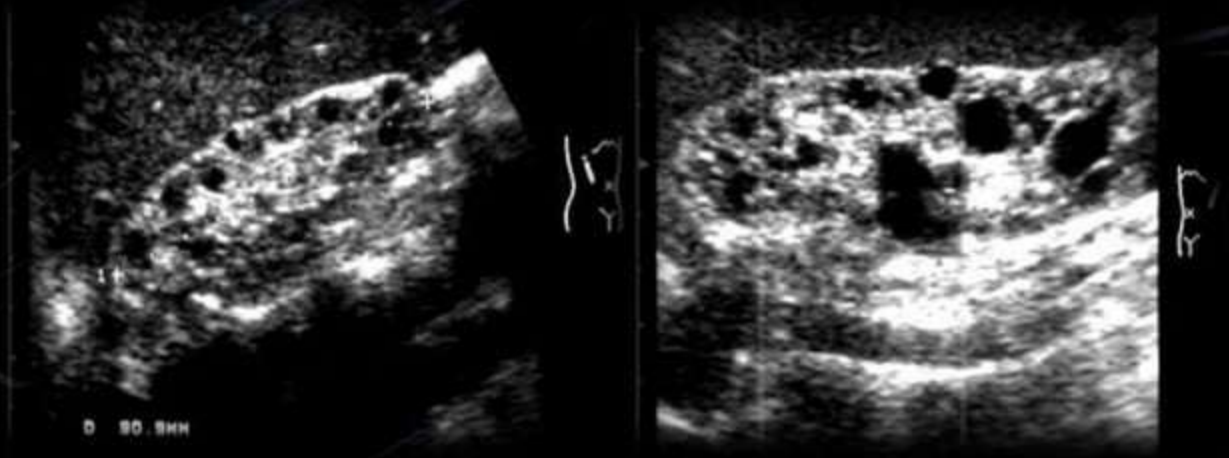
Multilocular Cystic Nephroma

- Multilocular cystic nephromas are reported to have a malignant potential and so are treated by surgery.
- Additional cross-sectional imaging with MRI or CT is warranted to assess the extent of the lesion, to confirm a normal contralateral kidney and to exclude metastatic disease.

Acquired Cystic Kidney Disease (ACKD)

- Up to 22% of patients with chronic renal failure develop cysts within their native kidneys.
- The incidence of cysts increases with the number of dialysis years, with 90% of those on dialysis for more than 10 years having visible cysts.

Acquired Cystic Kidney Disease (ACKD)



Acquired Cystic Kidney Disease (ACKD)

- After successful renal transplantation, cysts usually regress in size.
- These cysts can be complicated by haemorrhage, can become secondarily infected or rarely can undergo transformation to renal cell carcinoma in adulthood

Multicystic Dysplastic Kidney (MCDK)

- MCDK is the second most common cause of an abdominal mass in a newborn, after hydronephrosis.
- It is a developmental anomaly characterised by multiple cysts of varying sizes, without identifiable normal renal parenchyma.
- It is always unilateral, as bilateral disease is incompatible with life.

Multicystic Dysplastic Kidney (MCDK)

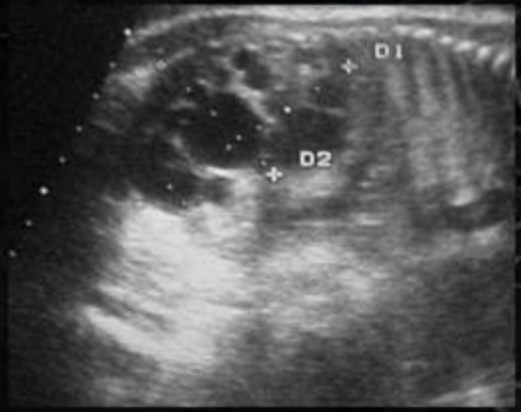
- The primary abnormality of MCDK is an atretic ureter with no connection between glomerulus and calices.

Multicystic Dysplastic Kidney (MCDK)

- MCDK can also rarely be secondary to ureteroceles that have caused complete obstruction.
- It can rarely be seen in one half of a duplex kidney or in other abnormalities such as a horseshoe kidney.

Multicystic Dysplastic Kidney (MCDK)

- The diagnosis is normally made at antenatal or neonatal ultrasound.
- The typical appearance of MCDK is of multiple large and small cysts.



Multicystic Dysplastic Kidney (MCDK)

- The contralateral kidney has a 20–30% chance of having an abnormality, mainly PUJO or ureteric stenosis.
- Based on this, MCDKs are classified into two types: (a) simple, which has a normal contralateral kidney; and (b) complicated, which has an abnormal contralateral kidney.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