Chapter 6

RENAL ANOMALIES

RENAL AGENESIS

Bilateral renal agenesis is incompatible with prolonged extra uterine life, occurs twice as often in males as in females and is present in between 0.2-0.4% autopsies performed in infants stillborn or dying soon after birth.

Potter facies with large low set ears deficient in cartilage, wide set eyes, beaked nose and micrognathos is seen at birth.

Other congenital anomalies particularly bilateral pulmonary hypoplasia are often associated with renal agenesis.

Embryologically condition results from lack of ureteric budding from mesonephric duct.

For diagnosis of bilateral renal agenesis intravenous urogram can be undertaken in anuric neonate with progressive uraemia.

Ultrasonography, nephrography and computerized tomography are useful.

Unilateral renal agenesis occurs in 0.1% of neonates. More common in males. Associated anomalies of external ear on ipsilateral side, agenesis or atresia of ureter of corresponding kidney and congenital abnormalities in ipsilateral leg. Contralateral kidney is hypertrophied in compensation. This may be more easily accidentally damaged because of its larger size.

Unilateral renal agenesis is result of lack of formation of ureteric bud or its inability to stimulate differentiation of nephrogenic mesoderm.

Possibility of unilateral renal agenesis should always be excluded before contemplating nephrectomy or percutaneous kidney biopsy and before deciding that kidney is non functioning eg in suspected renal vein thrombosis.

Ultrasonography helps detect nonfunctioning kidney.

POLYCYSTIC KIDNEY

Usually bilateral in contradistinction to unilateral multi cystic kidney disease.

Infantile form presents at birth with bilateral renal masses and is rapidly lethal. It may present in late infancy. There may be siblings with same condition but no affliction of earlier generation.

Pathologically cysts are fusiform in shape affecting cortex and medulla and radially oriented with kidney retaining its general uniform shape.

On urography there is delay in appearance of dye. Nephrogram may show streaky pattern and delay of hours or days in loss of radio opaque material.

Lack of communication between nephrons and collecting ducts with dilatation of nephrons is seen. Hypertrophy and cystic degeneration of interstitial portions of collecting ducts occurs. These children die during first month of life but may survive for years with varying degree of chronic renal insufficiency.

Adult type may appear even in neonates although it characteristically gives rise to symptoms of renal insufficiency in middle age.

Inheritance is autosomal dominant.

Kidneys become distorted by roughly circular and randomly distributed cysts. Radiologically seen as 'spider kidney'.

Both types of polycystic kidneys may be associated with cysts of liver and pancreas.

In some patients renal cystic disease is associated with hepatic fibrosis. These juvenile patients present with features of portal hypertension such as ascites, splenomegaly and haemetemesis.

Familial bilateral cystic dysplasia of kidney rarely affects liver. Radiologically in these cases blunted renal calyces are seen.

Progressively increasing renal insufficiency is to be anticipated in all types of polycystic kidney disease. There are episodes of haematuria and recurrent urinary tract infection.

Kidney may be so large as to obstruct delivery of baby.

Treatment is symptomatic to correct fluid and electrolyte disturbance induced by renal failure and systemic hypertension. Surgical relief of portal hypertension by porta caval shunt has been found useful.

Renal transplantation is definitive treatment.

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