

Developmental disorders of renal disease

Psychology, Psychotherapy



Some of the common developmental kidney disorders include polycystic kidney disease, congenital nephrotic disease, nephroblastoma, renal agenesis, duplication anomalies, fusion anomalies, malrotation, multicystic dysplastic kidney disease, renal dysplasia, renal hypoplasia, etc. Congenital nephrotic disease is an inherited disorder that may present at birth in which the infant has proteins present in the urine (proteinuria) along with swelling of the body (oedema). The condition is rare and is usually found in children born in Finnish families. Children born with the disorder have a protein found in the urine, known as ‘nephryn’.

Several substances such as proteins, fats, blood proteins, etc are excreted in the urine. The individuals develops several symptoms including swelling, low birth weight, malnutrition, kidney failure, poor appetite, infections, presence of blood in the urine, poor general health, cloudy appearance of the urine, etc (Charytan, 2006). Nephroblastoma or ‘Wilm’s tumour’ is a condition characterised by the formation of a malignant tumour in the kidney. It commonly occurs in infants and children. Wilm’s tumour is a very frequent tumour that develops in the abdomen in children.

The condition is frequently related to other birth defects such as urinary tract abnormalities, enlargement of one half of the body, missing iris, etc. As the condition is more frequent in identical twins, it is considered to have a genetic link. The tumour seldom spreads to the other parts of the body. One in every 200, 000 children develop this disorder. The child may develop several symptoms including abdominal pain and swelling, presence of blood in the urine, fever, loss of appetite, nausea, vomiting, malaise, hypertension, constipation, cloudiness of the urine, etc (Nanda, 2006).

Polycystic renal disease (Cystic renal development disorder) is a familial condition in which the affected individuals develop cysts in the kidney. The condition is an autosomal dominant condition and the symptoms less frequently develop in childhood. One in every 1000 develops the symptoms of polycystic renal disease. In childhood, an autosomal recessive version of polycystic renal disease can also develop. The child may develop severe symptoms along with renal failure with a fatal outcome. Lung function insufficiency is another frequent complication that can result in death.

The common symptoms of polycystic renal disease include abdominal pain, abdominal swelling, presence of blood in urine, flank pain, excessive passage of urine, drowsiness, hypertension, joint pain and swelling, nail defects, cysts in other portions of the body such as testis, liver and pancreas, colon defects, swelling of the kidneys, bile duct defects, portal hypertension, fibrosis, brain abnormalities, kidney stones, anaemia, frequent urinary tract infections, renal failure, liver failure, rupture and bleeding of the cysts, etc. The child has a positive family history of polycystic renal disorder.

The exact manner in which multiple cysts are formed in the kidney is not understood clearly. However, a genetic cause has been outlined. Once the kidney cysts are formed, they tend to swell, resulting in deterioration of the kidney function. The individual develops several symptoms (Silberberg, 2007). Renal agenesis is a condition in which the kidneys fail to develop. It can occur unilaterally as well as bilaterally. In the bilateral form, several other conditions such as pulmonary hypoplasia, oligohydramnios, facial defects, limb abnormalities, etc, occur resulting in fatal outcomes.

In the unilateral form, the individual develops trigone and ureteral orifice defects, absence of the ureter, etc. This form is less severe compared to the bilateral version. The individual can survive provided the kidney function is managed appropriately (Merck, 2005). Duplication anomalies are conditions in which the individual develops extra collecting systems. This may affect one kidney or both, and may involve the ureter, calyx, ureteral orifice and the renal pelvis. These conditions have to be treated very carefully depending on the extent to which function is affected (Merck, 2005).

Fusion anomalies are conditions in which the kidneys are united to one another. However, the ureters are separate and enter the bladder on either side. Several conditions such as vesicoureteral reflux, congenital renal cystic dysplasia, etc, tend to occur more frequently with fusion anomalies. A condition known as ‘horseshoe kidneys’ is characterised by the fusion of the renal parenchyma bilaterally. It is one of the most common kidney fusion abnormalities. The ureters tend to function normally. Sometimes the point at which the renal pelvis is united to the ureter is abnormal resulting in urinary obstruction.

The second most common kidney union anomaly is ‘crossed fused renal ectopia’ in which the kidneys are present on one of the body, and the ureter crosses the midline of the body and empties into the bladder of the either side. Pancake kidney or ‘fused pelvic kidney’ is a condition in which the kidney is single or fused and is emptied into 2 ureters and collecting systems (Merck, 2005). In multicystic dysplastic kidney, the kidneys consist of multiple cysts, cartilage, primitive tubules, and multiple cysts. The individual develops several symptoms such as infection, swelling, hypertension, etc.

Renal dysplasia is a condition in which the renal tubules, vasculature, collecting tubules, etc, develop abnormally, resulting in distortion of the normal kidney function. In renal ectopia, the kidneys are not placed in their exact anatomical position resulting in several problems such as obstruction. Renal hypoplasia is a condition in which the ureteral bud gives rise to an underdeveloped and small kidney. However, the size of the nephrons is normal. The individual can develop hypertension (Merck, 2005). Ureterocele is a condition in which the ureter is enlarged at the point it is inserted into the bladder.

At this point, the flow of urine into the bladder is seriously affected resulting in obstruction and hydronephrosis. In neurogenic bladder, due to a defect in the spine at the lumbosacral region, the nerves that supply the bladder are affected resulting in improper drainage of the bladder. The individual is at a risk of developing several problems such vesicureteral reflux, infection, renal hypertension, scarring, renal failure, etc. Hydronephrosis is a condition in which the ureter and the collecting system are dilated (Conley, 2007).

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