

Renal Agenesis

BIRTH DEFECT RESEARCH FOR CHILDREN



What is Renal Agenesis?

Renal Agenesis is the congenital absence or severe malformation of one or both kidneys. The kidneys are part of the urinary system that also includes the bladder, the ureters, and the urethra. The kidneys filter out waste products from the blood and eliminate them as urine that flows through tubes called ureters to the bladder (the storage area) and then through the canal called the urethra. Bilateral Renal Agenesis is the absence of both kidneys (sometimes called Potter's Syndrome). It is associated with oligohydramnios, a deficiency of amniotic fluid in a pregnant woman. Because the amniotic fluid normally acts as a cushion, too little fluid can cause compression of the fetus resulting in further malformations and problems such as growth retardation; pulmonary hypoplasia (underdeveloped lungs); low-set ears; and a broad, flat nose.



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Unilateral Renal Agenesis (URA) is the absence of one kidney. The solitary kidney enlarges to compensate for the absent one and maintains normal kidney function. The ureter on the affected side may also be absent or abnormal. Abnormality of the reproductive tract on the affected side is sometimes associated with URA (more often in females than males).

How many children have Renal Agenesis?

Bilateral Renal Agenesis affects one to two out of every 10,000 births and is 2 1/2 times more frequent in males. Unilateral Renal Agenesis is more common, occurring in one out of every 750-1,000 births. It is more common in males, and the left kidney is more frequently absent.

How do you know if your child has Renal Agenesis?

Some cases of Bilateral Renal Agenesis are detected through prenatal ultrasound while other cases are not evident until birth. The ultrasound examination may reveal the absence of amniotic fluid, the absence of kidneys, and possibly the absence of the bladder. Babies with Unilateral Renal Agenesis most often appear normal at birth. URA is usually discovered incidentally through x-ray or ultrasound imaging of the abdomen for other reasons.

What causes Renal Agenesis?

Renal Agenesis usually occurs sporadically. But 20-36% of the bilateral cases present a familial recurrence, occurring more commonly in infants with a parent who has a kidney malformation, especially Unilateral Renal Agenesis. There is no known prevention for Renal Agenesis and genetic counseling is recommended.

How can you help a child who has Renal Agenesis?

Because of the pulmonary hypoplasia associated with Bilateral Renal Agenesis, kidney dialysis and kidney transplantation are usually not considered, and no treatment is administered. With Unilateral Renal

Agenesis, the solitary kidney usually grows faster and becomes larger and heavier than normal.

Therefore, the kidney becomes more susceptible to injury. Children with Unilateral Renal Agenesis should avoid heavy contact sports. It is also important for them to have urine tests and blood pressure checks annually and kidney function checks every few years (or more frequently if the other tests are abnormal). Treatment may become necessary for kidney problems such as obstruction or infection. Kidney transplantation may be necessary if the kidney becomes severely diseased, and dialysis or transplantation may be necessary to treat kidney failure. Patients receiving transplantation require anti-rejection medication and regular medical follow-up.

What's in the future for a child with Renal Agenesis?

Bilateral Renal Agenesis is fatal. Approximately 40% of the cases result in stillbirths. Infants rarely survive more than two days and usually die of respiratory failure within a few hours of birth. Children with Unilateral Renal Agenesis usually have few or no problems in the first few years if the solitary kidney is healthy and there are no other anomalies. They do have a greater chance of developing high blood pressure and may experience a mild decrease in kidney function in later life. Sterility has been noted in some individuals with URA. Otherwise, these children usually live a normal life span.

Fact Sheet by:

Birth Defect Research Children,
Inc.

www.birthdefects.org

