

RENAL AGENESIS/ HYPOPLASIA

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Renal Agenesis/Hypoplasia
(Name no longer used: Potter Syndrome)

Common Associated Conditions

Condition Description

Renal agenesis is the name given to a congenital absence of one or both kidneys. The kidneys develop between the 5th and 12th week of fetal life, and by the 13th week they are normally producing urine. When the embryonic kidney cells fail to develop, the result is called renal agenesis. It is often detected on fetal ultrasound because there will be a lack of amniotic fluid (called oligohydramnios). When bilateral (both kidneys are absent) this condition is not compatible with survival; in fact, 40% of babies with bilateral renal agenesis will be stillborn, and if born alive, the baby will live only a few hours.

Babies with bilateral renal agenesis will have a number of unique characteristics: dry loose skin, wide-set eyes, prominent folds at the inner corner of each eye, sharp nose, and large low-set ears with lack of ear cartilage. They will typically have underdeveloped lungs, absent urinary bladder, anal atresia, esophageal atresia and unusual genitals. The lack of amniotic fluid causes some of the problems (undeveloped lungs, sharp nose, clubbed feet) and other problems occur because the kidneys and those affected structures are formed at the same time of fetal life (such as the ears, genitals, esophagus).

Prevalence

Bilateral renal agenesis occurs in 1 of 4500 live births and is usually found in boys. Unilateral renal agenesis occurs in 1 of 1000-2000 live births. Usually there is no family history of renal agenesis, but in 20-36% of cases, there is a genetic cause. The risk of recurrence in future pregnancies is 3% unless one parent has unilateral renal agenesis, in which case the risk is about 15%. Women with uncontrolled diabetes in pregnancy may deliver a baby with bilateral renal agenesis.

Babies with unilateral renal agenesis (absence of one kidney) may have no other symptoms at all. Unilateral renal agenesis is more common with intrauterine growth retardation (poor growth during pregnancy) and often results in premature birth. It is also more common when a mother is carrying more than one baby (multiple gestations, such as twins or triplets). Babies' ears may be located lower than typical on their heads (called low-set ears) so if ears are in this lower placement the baby should be examined for kidney problems. This is because the ears and kidneys are formed at the same time in fetal development. The ureters (small tubes that drain the urine from the kidney to the bladder) may also be abnormal and must be carefully examined early in life (by x-ray or ultrasound) so the kidney function of the one remaining kidney will be preserved.



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Short-term Treatment and Outcomes

Bilateral renal agenesis is fatal. If one kidney is present (unilateral renal agenesis) the child will develop normally. Many times the absence of a kidney is detected only incidentally when an older child or adult has an abdominal x-ray for some other reason. The remaining kidney, if properly functioning, can very effectively remove the wastes from the blood and keep the body entirely healthy.

Once detected, families where renal agenesis has occurred will be offered genetic counseling because of the possibility of recurrence in future pregnancies.

Long-term Treatment and Outcomes

Once diagnosed, children with one kidney (also called solitary kidney) will be encouraged to protect the remaining kidney from infection or injury. They will receive examinations of the kidney periodically and require prompt treatment of any urinary tract infection. They may be counseled to avoid contact sports where the kidney could be injured. Blood pressure monitoring will be essential throughout life since its elevation can cause kidney damage.

Common Complications

Children with unilateral renal agenesis may have frequent urinary tract infections, high blood pressure, or kidney stones, and thus they should be seen periodically by a kidney specialist (urologist or nephrologist) to detect any abnormalities. X-ray studies to detect problems of the ureter (the tube that drains urine from the kidney into the bladder) will typically be done. Ureters that do not drain the kidney effectively (called reflux) can cause back-pressure into the kidney and damage to the kidney can occur (called hydronephrosis).

Protecting the kidney function is very important. Sometimes children will be prescribed a low dose of an antibiotic to take once a day to prevent kidney infection and damage. Blood pressure should be carefully monitored and elevations treated. Dialysis or kidney transplant are the only options to treat children whose solitary kidney has ceased to function.

Implications for Children's Development

Children with unilateral renal agenesis will generally live normal lives with no developmental effects; in fact, many times the solitary kidney is only detected incidentally when x-rays are done for other purposes. The remaining kidney will enlarge to carry out the function normally done by two kidneys.

Children should be taught simple information about their condition and treatment, using terms that children can understand. Books and dolls can help children understand how their bodies work. They should be encouraged to ask questions about their bodies and the treatments and tests. Children do not need a great deal of detailed information but rather they require reassurance and explanations that are tailored to their age and understanding.

They need to be helped to understand that blood tests and uncomfortable procedures are necessary to help them feel better, even though they experience temporary pain or discomfort. Play therapy can assist young children to express their fears and feelings and can reveal misunderstandings they may have about their condition or treatment.

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