ARPKD: Autosomal Recessive Polycystic Kidney Disease

What is ARPKD?

ARPKD is a serious kidney disease in which the kidneys have many small **cysts**. Cysts are collections of fluid. Many children with ARPKD also have a liver disease called **congenital hepatic fibrosis**.

ARPKD is different in each person. Some children have kidney failure and others do not. Some babies with ARPKD have extremely large kidneys. Such kidneys do not leave enough room for the lungs to breathe. These babies usually have very bad breathing problems. Other babies with ARPKD do not have breathing problems. Some babies may have difficulty feeding well enough to grow.

What causes ARPKD?

Genes cause ARPKD. Genes are the "instructions" on how to build your body. Genes control many things, such as the color of your hair or eyes. Children get genes from both of their parents. Genes also control some kidney diseases. Some people have **one** gene for ARPKD. They do not have the disease. You must have **two** genes for ARPKD to get the disease. If a man and a woman **both** have one gene for ARPKD, there is a 1 in 4 chance that their baby will get **two** genes for ARPKD. A person with two genes will have the disease. ARPKD is **not** contagious. You can't "catch" it from someone else. ARPKD is **not** a form of cancer.

How is ARPKD treated?

No treatment can make the cysts go away. Doctors can only treat the problems related to the disease. There is not one "standard" treatment, because every child is unique. Talk with your child's doctor. He or she will develop a treatment plan especially for your child. These are some of the treatments available:

- Babies with breathing problems need a lot of medical care. They usually have kidney failure at birth. Sometimes doctors remove one or both kidneys. They do this to give the lungs more room to expand.
- People with ARPKD often have high blood pressure. Low sodium diet, exercise and medicines can help treat blood pressure.
- Most children with ARPKD develop kidney failure by late childhood or adolescence. The treatments for kidney failure are **dialysis** and **a kidney transplant**.
- Sometimes if infants have problems breathing or feeding, one kidney can be surgically removed to make room for the lungs and stomach.

Can my other children have ARPKD?

If you have a child with ARPKD, there is a **25% (1 in 4) chance** that each child from the same parents will have ARPKD.

- *If you have other children*: Ask your doctor if they should be look at for ARPKD.
- *If you want to have more children in the future:* Ask your doctor for advice about the risks of ARPKD.

Where can I get more information?

Your kidney doctor is your first source for information. Be sure to ask any questions you have. You may want to write down questions so you can ask them at the next visit. The PKD Foundation provides information and support for people with ARPKD.

PKD Foundation
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