

## **AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPKD)**

**(also known as infantile polycystic disease)**

*The National Kidney Federation cannot accept responsibility for information provided. The above is for guidance only. Patients are advised to seek further information from their own doctor.*

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[www.kidney.org.uk](http://www.kidney.org.uk)

*The NKF - the largest Kidney Patient charity in the UK*

*A federation of more than fifty patient charities, supporting kidney patients & carers*

NKF(National Kidney Federation)  
The Point, Coach Road, Shireoaks, Worksop, Nottinghamshire, S81 8BW  
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[www.kidney.org.uk](http://www.kidney.org.uk)

**HELPLINE 0800 169 09 36**



## **Autosomal Recessive Polycystic Kidney Disease (ARPKD)**

**(also known as infantile polycystic disease)**

**Infantile polycystic disease is a rare condition that should be diagnosed by a paediatrician. It is distinct from having a small number of normal cysts in the kidney and some other conditions where multiple cysts are found in the kidneys. It is also distinct from adult polycystic kidney disease.**

Incidence is estimated to be from 1:10,000 to 1:40,000. The disease is inherited as a recessive condition, ie two copies of the mistaken gene are required to cause the problems; one copy makes you a carrier. Thus both parents have to be carriers, and any brother or sister of an affected child has a 1:4 chance of having it. Both parents are carriers without symptoms or signs.

Small cysts appear on the tubules of the kidney. The greater number of tubules involved, then the more likelihood of renal damage. The liver is often affected which may mean a transplant of both organs may be needed.

Most cases are diagnosed in infancy. Some may be suspected by foetal ultrasound while others are diagnosed at birth.

The main complication in newborn babies is severe difficulty breathing. Mechanical ventilation may be needed. High blood pressure is common as is some degree of kidney failure.

The rate at which kidney function is lost varies widely, but the majority of babies who survive the first month of life do not develop severe kidney failure until later childhood or adolescence.

Elevated blood pressure and urinary tract infections need to be treated. Erythropoietin can be used to treat anaemia. Where renal failure occurs, dialysis will be needed. With transplantation, the outcome is good.

### **Other Information/Support groups**

The PKD Charity

Helpline : 0300 111 1234

E-mail : [info@pkdcharity.org.uk](mailto:info@pkdcharity.org.uk)

Website : [www.pkdcharity.org.uk](http://www.pkdcharity.org.uk)

*AND*

*Infokid.org.uk*

*a specialist website for paediatric kidney diseases, tests and procedures*