

IgA Nephropathy (AKF)

What is IgA nephropathy?

Glomerulonephritis is the term used for a group of diseases which damage the glomeruli (the filtering system of the kidney). In some of these the body's own immune system attacks the glomeruli and causes inflammation and scarring. There is still much to be discovered about the underlying causes of glomerulonephritis. It is the commonest cause of kidney damage and failure of kidney function.

There are many different types of glomerulonephritis but IgA nephropathy is the most common serious form in Australia. IgA nephropathy is a chronic kidney disease that may progress over a period of 10 to 20 years, and can lead to end-stage renal disease.

Why is it called IgA nephropathy?

As a French doctor, Jean Berger, was the first to describe the condition, IgA nephropathy is sometimes called Berger's disease. IgA refers to the antibody IgA (Immunoglobulin A) which is derived from the lining of the throat, air passages and intestine and is found in the kidney associated with kidney damage. Nephropathy simply means kidney disease.

What causes IgA nephropathy?

IgA is deposited into the mesangium - the substance between the kidney filters. This result in inflammation and kidney damage. How the IgA is deposited is uncertain, and is the subject of ongoing research. It is thought that in some people the body may produce

excess IgA antibody when fighting infections of the throat, lungs, tonsils and intestines. Some of the IgA lodges in the glomerulus causing inflammation.

Who gets IgA nephropathy?

Many people are not aware that they have the disease, or are only made aware of the disease when it is at a late stage. Found more often in males than females, the condition is usually diagnosed before the age of thirty. Roughly 10–30 percent of those with IgA nephropathy will suffer eventual kidney failure within 10–20 years. In Australia, about 10% of people on dialysis have IgA nephropathy. There may be a genetic link associated with IgA nephropathy. Certain families and certain parts of the world seem to have a higher incidence of the condition. Some evidence suggests that it is an autoimmune disorder.

What are the signs and symptoms of IgA nephropathy?

In some people, the presence of blood or protein in the urine is the only clue to kidney dysfunction. Signs and symptoms characteristic of IgA nephropathy include:

- Blood in the urine (haematuria – turns the urine bright red)
- Protein in the urine (proteinuria – causes foaming of the urine)
- High blood pressure
- Persistent low back pain (not associated with injury)
- Swelling of extremities (ankles, legs and wrists) and eye area (due to fluid retention)

These symptoms may be found in association with:

- Sore throat or respiratory infection
- Diarrhoea or vomiting

How is IgA nephropathy diagnosed?

Detecting blood and/or protein in the urine in association with other significant signs and symptoms may suggest a diagnosis of IgA nephropathy. However diagnosis of the condition can only be made by a kidney biopsy, whereby

under local anaesthetic, a small sample of kidney tissue is removed by a needle and examined microscopically for the presence of characteristic deposits of IgA in the glomeruli.

What is the treatment for IgA nephropathy?

At present, there is no cure for IgA nephropathy. Slowing the progress of the disease and preventing complications are key priorities. Treatment recommendations are made following careful review of an individual's symptoms, condition and kidney health history.

Control of high blood pressure is an important treatment for the condition, as high blood pressure over an extended period of time will have further adverse consequences on kidney health. Medications may also help preserve kidney function.

Recent studies indicate that fish oil supplements and steroids may help slow the loss of kidney function in some cases of IgA nephropathy. Consultation with a renal specialist will help determine whether these are useful treatment options for an individual case. Additional new treatments are being discovered which may prove even more effective than current therapies in preserving kidney function.

People with kidney failure due to IgA nephropathy usually eventually require dialysis treatment or a kidney transplant. The success rate of transplantation is good in people with IgA nephropathy. Although the IgA deposits can reappear in the transplanted kidney, the signs and symptoms of the disease remain mild. The immunosuppressant drugs taken to prevent transplant rejection may be responsible for the mild symptoms.

How does the disease progress?

The majority of people with the mild form of IgA nephropathy have a good outlook. With proper care and disease management, they may suffer from little more than blood in the urine, high blood pressure and the usual deterioration

of kidney function associated with age.

Those with more severe kidney damage may find their kidney function declines to the extent that dialysis or transplantation is necessary. It is very important that blood pressure and kidney function be closely monitored over time.

Should a person with IgA have regular follow-up and care?

Regular consultation with a doctor or kidney specialist is recommended for any person with IgA nephropathy, even in its mild form. Even healthy individuals with IgA nephropathy should make a point of having regular blood pressure checks and urine tests. This will help detect early signs of any potential problems or deterioration of their condition. Although the genetics of the disease are unclear, it is recommended that those with a family history of the condition include blood pressure and urine tests in their annual visit to the doctor.

Reference: Donadio, J.V., Grande, J.P., Bergstralh, E.J., Dart, R.A., Larson, T.S. and Spencer, D.C. (1999). The long-term outcome of patients with IgA nephropathy treated with fish oil in a controlled trial. *Journal of the American Society of Nephrology* 1999 Aug; 10 (8): 1772 - 7

Further information:

The international IgA Nephropathy Network – www.igan.net