

## 6. IgA nephropathy

### A GUIDE FOR PATIENTS

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#### IgA nephropathy

IgA (immunoglobulin A) nephropathy is another name for the most common form of primary glomerulonephritis worldwide.

##### What is the cause of IgA nephropathy?

The exact cause of IgA nephropathy is unknown. Whenever we have an infection such as a sore throat, our body attacks the invading viruses or bacteria, in part by producing compounds called immunoglobulins. Immunoglobulins inactivate the invading organism, allowing the body to eliminate it. There are three main classes of immunoglobulins, namely IgG, IgA and IgM. The principal immunoglobulin produced when there is an infection of the throat, gastrointestinal tract (stomach or bowel) or breast is immunoglobulin A or IgA. In patients with IgA nephropathy, the IgA also deposits in the tiny filters of the kidney, the glomeruli, causing inflammation (not infection) and this results in blood and protein leaking into the urine. Occasionally, inflammation leads to progressive damage and renal failure.

Usually, IgA nephropathy is a primary diagnosis, that is, it is not associated with other disease processes. It may however sometimes be seen in association with certain skin conditions (dermatitis herpetiformis), some forms of liver disease (alcoholic liver disease, chronic active hepatitis), some bowel conditions (coeliac disease, certain inflammatory bowel diseases) or be part of a condition known as Henoch Schonlein purpura.

##### How do I know if I have IgA nephropathy?

IgA nephropathy is twice as common in men than in women and is usually diagnosed when the patient is relatively young (average age 30 years), although any age group can be affected.

Patients with IgA nephropathy often have a **family history** of nephritis. It is therefore important that all family members are screened. This is best done by urinalysis **testing the urine** for the presence of blood and protein or having a sample of the urine examined by a pathologist, specifically by microscopy, looking for an increased number of red and white blood cells.

**Symptoms** may have often been present for a long time before a definitive diagnosis is made. In over 40% of cases there has been recurrent passing of blood in the urine, often accompanied by loin pain (pain in the back) and generally feeling unwell. This passage of blood often occurs within 48 hours of an upper respiratory tract infection, gastrointestinal or other infection. In the majority of patients, testing of the urine reveals the persistent presence of blood, often accompanied by protein. High blood pressure is very common in IgA nephropathy, present in 25% of patients when first diagnosed and developing over time in a further 25%. This high blood pressure is a very important factor, because if it is not well controlled it can accelerate the rate of deterioration of kidney function.

#### What tests may be necessary to confirm IgA nephropathy?

**Microscopy of urine** usually shows an increased number of red blood cells, often accompanied by an increased number of white blood cells.

The **24-hour urine protein** loss is often raised and in a small number of patients (approximately 10%), is very large (greater than 3 grams per day).

**Blood creatinine** concentration (the level of substance coming from protein breakdown) is abnormal in approximately 20% of patients when they are first seen by a doctor, indicating significant impairment of kidney function. A more specific test of kidney function is provided by the **creatinine clearance**, which involves a blood sample and a timed urine collection. The creatinine clearance is reduced in 35% of patients at the time of first being seen by their doctor. A **renal biopsy**, if performed, will provide an accurate diagnosis of the condition and an indication of the severity of the nephritis and help determine which form of treatment is likely to be beneficial.

Temporary decreases of renal function are frequently seen in patients with IgA nephropathy (greater than 10% change in over 70% of patients).

These changes in renal function do not mean that there will be a permanent or progressive change.

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### ***IgA nephropathy continued...***

#### **What are the long-term prospects if I have IgA nephropathy?**

For the majority of patients with IgA nephropathy the long-term outcome is usually good. We have treated and followed more than 200 patients with IgA nephropathy for many years. Over this time less than 10% of patients have progressed to end-stage renal failure. Fifteen years after the onset of symptoms, 88% of patients will still have good renal function and only 12% will have progressed to end stage renal failure.

Many patients with IgA nephropathy may not be firmly diagnosed with renal biopsy, especially if they have a trace of blood in their urine and their kidney function is normal. If we take these patients into account as well as the figures in the paragraph above then it is clear that overall long-term outcome is good for this condition.

In addition to this overall good long term outcome, a **complete remission** of disease may be seen in approximately 7% of patients with IgA nephropathy.