Oxford Kidney Unit

IgA Nephropathy

Information for patients
If you have been told by the doctor looking into your kidney problem that you have IgA nephropathy, this leaflet is designed to help you. It gives you information about IgA nephropathy, about its cause and its effects, about treatments you may be recommended, and about the likely effect on your kidney function in the future.

What do the kidneys do?

The kidneys are the specialised organs that filter and purify the blood, ridding the body of excess water, salts and waste products.

The blood entering the kidneys is filtered through a series of coils of small blood vessels, which under the microscope look like balls of wool. This filter, called the glomerulus, allows water and chemicals to pass but holds back proteins and cells. The filtered fluid is then processed as it passes down a long tube (called the tubule) so that only the excess water, salts, acids and waste products are left by the time it reaches the end of the tube.

This fluid, now called urine, dribbles into the pelvis of the kidney and flows down a long muscular tube called the ureter into the bladder.

The other important function of the kidneys is to produce three essential messenger substances: (1) active vitamin D which is necessary for calcium absorption from the gut and healthy bone formation; (2) renin, which controls the blood pressure; and (3) erythropoietin (epo), which stimulates the bone marrow to produce red blood cells.
IgA Nephropathy

Most nephrologists call this condition IgA nephropathy, but you may hear it called IgA Disease or Berger’s Disease (after the man who first described it). These names all describe the same condition.

IgA nephropathy (which just means IgA diseases of the kidney) is the commonest of a group of conditions which have the general name glomerulonephritis.

Glomerulonephritis means inflammation of the kidneys which particularly affects the glomeruli, the microscopic filters of the kidney. IgA nephropathy is given that name because an antibody called IgA is deposited in the glomeruli.

How is it diagnosed?

Although your doctor may have suspected you had IgA nephropathy because of your symptoms (more of this later), the diagnosis could only be confirmed when you had your kidney biopsy. When the pathologist examined the biopsy, IgA was found in the glomeruli. Because IgA nephropathy (like all forms of glomerulonephritis) affects both kidneys equally, a small biopsy from one kidney will provide the answer.

What is IgA?

IgA is an antibody, part of our natural defences against infection. We all have plenty of IgA in our blood and also in the inner lining of our intestine and airways where it works in our defence. If you have IgA nephropathy, a very small proportion of all the IgA in your body sticks in your kidneys and sets off inflammation which damages the kidneys. The rest of the IgA in your body works normally so you have normal resistance to infection.

Despite a lot of research we do not know why the IgA deposits in the kidney, so we do not yet know the fundamental cause of IgA nephropathy.
What are the common symptoms of IgA nephropathy?

The commonest way that IgA nephropathy comes to medical attention is because of blood in the urine (haematuria). Typically there are episodes of visible blood in the urine and these most often coincide with a sore throat or respiratory infection. Less commonly the episodes seem to be brought on by a bowel infection or even by exercise. The urine may go bright red but more likely it will be brown (like tea without milk), or if there is less blood it may just look a bit smoky. Blood clots are not usually passed. The haematuria is usually painless although occasionally there is discomfort in the kidneys while the haematuria is visible.

Between these episodes there will still be invisible (microscopic) haematuria, which can be found if the urine is tested with a special ‘stick’ which detects blood (stick tests of the urine are frequently performed at a GP’s surgery or hospital outpatient clinic). Some people never have visible haematuria. The problem may therefore only be discovered when a urine sample is checked at a routine medical examination, for example on joining a GP’s list, starting a new job or when seeking life insurance.

Some other types of glomerulonephritis may produce the same symptoms, so the kidney biopsy is needed to make the diagnosis. (There are many other causes of haematuria other than glomerulonephritis and these will also be needed to be investigated).

As well as blood there may also be proteinuria (protein in the urine). Usually this will only be found on a ‘stick’ test, but just occasionally there will be so much loss of protein in the urine that fluid retention occurs (usually first spotted as ankle swelling). This is known as the ‘nephrotic syndrome’ but it happens less often in IgA nephrotic syndrome than in many other forms of glomerulonephritis.
High blood pressure (hypertension) often occurs as it does in most other kidney diseases.

**Who is likely to get IgA nephropathy?**

The condition is three times more common in males than females – but this difference is unexplained. It can be diagnosed at any age but most commonly in children and young adults who are likely to have typical symptoms of episodes of visible haematuria.

Older people are less likely to have visible haematuria; they are more likely to have protein as well as blood in the urine at the time of diagnosis and to have raised blood pressure and other evidence of more advanced kidney damage. We presume that these older people have in fact had IgA nephropathy for many years but because they never had visible haematuria or a routine urine test, it was not diagnosed until a much later stage.

**What will happen to my kidneys if I have IgA nephropathy?**

Although the function of the kidneys of many patients does not deteriorate, we now know that about a quarter of all people with IgA nephropathy will eventually develop impaired kidney function followed by kidney failure. This is usually a very slow process and may take twenty years or more.
Can you tell in each individual case about the risk of kidney failure?

We can identify those patients who are already a good way along that path. If there is protein as well as blood in the urine, if the blood pressure is raised, if the blood creatinine level (which measures kidney function) is already raised – then eventual kidney failure is much more likely.

The other information comes from your kidney biopsy. If haematuria is the only clinical sign of the disease, the biopsy may show very little damage to the kidneys other than the deposits of IgA – in this case the risk of kidney failure is small. If there is already proteinuria and hypertension, scarring of the kidney will be more obvious in the biopsy specimen; this scarring of the kidneys makes eventual kidney failure more likely.

Although we can make a good general prediction for each individual of what may happen, we cannot be certain. That is why it is vital to keep an eye on all patients with IgA nephropathy. Even if the only sign is haematuria and the biopsy does not show damage, there is still a small long term risk of kidney failure. For that reason you will be kept under long term review in the Nephrology Clinic or by your GP.

Does IgA nephropathy ever get better?

Yes it can do. In some people even the invisible haematuria will slowly disappear, leaving no clinical evidence of the disease. We know that the IgA can still be in the kidney and haematuria may come back, although this is unlikely. So regular check-ups are still necessary.
What happens if I have repeated attacks of visible haematuria?

Do not worry. Many people with IgA nephropathy have quite a few attacks of visible haematuria. These gradually get less frequent and usually stop altogether after a few years – although of course the blood will still be found on ‘stick’ testing of the urine.

You might think that with each attack more damage would be done to the kidneys but in fact there is not a close link between the number of attacks of haematuria and the chance of developing kidney failure later on.

Just occasionally an attack of haematuria will cause a sudden deterioration in kidney function, which usually goes back to normal in a week or two, but this is unusual.

Does IgA nephropathy run in families?

Very occasionally a family will have more than one member affected by IgA nephropathy, but this is very unusual. For the great majority of patients with IgA nephropathy only one person in the family is affected, so there is no need for the rest of the family to be checked.
Does IgA nephropathy affect any part of the body other than the kidneys?

No, it does not, but there is a condition called Henoch-Schönlein purpura (often shortened to HSP) which is related to IgA nephropathy.

HSP gets its name from the two German doctors who originally described it over a hundred years ago. We now know that the kidney problem in HSP is very similar to IgA nephropathy; but patients with HSP also get a skin rash and may have aches and pains in their joints and attacks of abdominal pain. IgA is found in the skin as well as in the kidneys in HSP. The skin, joint and abdominal effects of HSP often go away in a few weeks or months, but the kidney problem can stay much longer, and may eventually cause kidney failure. HSP can occur at any age but is commonest in young children.

Should I change my lifestyle?

No, you should not.

- **Diet** – there are no foods known to cause IgA nephropathy or to provoke attacks of visible haematuria. You should continue with normal healthy eating.

- **Alcohol** – alcohol has no effect on kidney disease. You may continue to drink alcohol in moderation.

- **Exercise** – if you are one of the few patients who notice that vigorous exercise produces visible haematuria it may be sensible to avoid the activities you know do this. Otherwise you can continue to enjoy all physical activities.
What treatment can be used in IgA nephropathy?

There is no cure for IgA nephropathy so treatments are prescribed to lessen its effects or to slow its progress.

1. **Treatment for the attacks of visible haematuria.** When visible haematuria occurs for the first time it can be dramatic and alarming. There is no specific treatment. Although it may seem you are losing a lot of blood it only needs a relatively small amount to colour the urine, so it is very unusual to become anaemic from haematuria in IgA nephropathy. The haematuria will settle down on its own in a few days.

   If the attacks are started off by a sore throat or other airway infection, the infection should not be treated any differently because you have IgA nephropathy. If you have a bacterial sore throat you may need antibiotics, but many of these infections are caused by viruses and will get better on their own. For many years patients were given a small dose of penicillin every day to prevent attacks but this makes no difference to the kidney disease.

   If you have many attacks of haematuria due to tonsillitis you may be advised to have your tonsils out. There is no proof that this will reduce the chance of kidney failure. So the decision should be based on the amount of trouble your tonsils are giving you, rather than the fact that you have IgA nephropathy.

2. **Steroids and other treatments to suppress your immunity.** Because there is something slightly wrong with the immune system which causes the IgA to stick in your kidneys in IgA nephropathy, a number of treatments which suppress your immune system have been tried. These include steroids (such as prednisolone), cyclophosphamide and azathioprine. A course of treatment may be beneficial in patients whose kidneys are already damaged but this has
not been conclusively proved. In a small number of patients whose kidney function is deteriorating rapidly this type of immunosuppressive treatment may be useful; but it has potential for considerable side-effects so your nephrologist will carefully discuss the pros and cons.

There is one exception. If you have a lot of protein in the urine (nephrotic syndrome) a course of steroids may help to reduce this rapidly, helping you to feel better and perhaps reducing the chance of kidney failure. This treatment is only appropriate for a few patients with IgA nephropathy.

3. **Fish Oil** sounds an unlikely treatment for kidney disease, but it does have a number of small effects which together seem to reduce the inflammation and scarring which cause the kidney failure. There is some evidence that fish oil reduces the chance of kidney failure when given to those whose kidneys have already deteriorated quite a lot; although not all nephrologists are convinced by the evidence. There is no reason to consider this treatment if your IgA nephropathy is mild. You could get the same effect from eating oily fish but you would have to eat such vast amounts it would not be possible; so the fish oil is given in capsules. Cod liver oil tablets which you can buy at a chemist are not suitable; the fish oil capsules must be prescribed by your doctor.

Fish oil may be a good treatment because it does not have all the potential risks associated with steroids and other immunosuppressive treatment. However it may slightly reduce the efficiency of your blood clotting system although this is not usually a practical problem. It does also give some people a fishy taste in their mouth and some tummy upset which means they cannot take it.

4. **Blood Pressure.** Raised blood pressure is particularly common in patients with IgA nephropathy and may develop quite early on when the kidney damage is very mild. It is vital to control the blood pressure with tablets for two reasons. Firstly, it is
important for your general health; we know that you are more likely to have a heart attack or stroke at a younger age if you have high blood pressure, but if the blood pressure is controlled that risk is greatly reduced. Secondly, it is very important for your kidneys. Once your kidneys have been damaged they are sensitive to the effects of raised blood pressure and will deteriorate more rapidly.

Treating raised blood pressure is the one thing which definitely helps delay kidney failure in IgA nephropathy. Keeping blood pressure normal with tablets is therefore the single most important way of reducing the risk of kidney failure from IgA nephropathy. There is a debate at present about the best blood pressure treatments to use. Most doctors believe that ‘ACE inhibitors’ or ‘HRBs’ are the best choice for blood pressure treatment if you have IgA nephropathy.

5. Treatment for kidney failure. If you have IgA nephropathy your kidneys may eventually fail. This happens to about a quarter of all patients and usually occurs very gradually, often over twenty years or more. If your kidneys fail, you will require treatment to replace kidney function, just as will be the case with other causes of kidney failure. Various forms of dialysis may be offered to you according to your personal circumstances. For many patients the best treatment, if it can be achieved, will be a kidney transplant. The fact that it was IgA nephropathy that caused your kidney failure does not have any effect on dialysis treatment.
What will happen if I have a transplant? Will IgA nephropathy damage my new kidney?

We know that IgA may stick in the glomeruli of your new kidney if you have a transplant. The immunosuppressive treatment which you receive to stop you rejecting your kidney does not prevent this happening. But this is not as serious as you might think. Although IgA gets into the kidney it usually does not cause much in the way of inflammation or damage to the kidney. A few patients have lost their transplant as a result of recurrence of IgA nephropathy but, if it does occur, it is nearly always a very slow process as it is in the patient’s own kidneys. Overall the chance of a successful kidney transplant is not reduced if your original kidney problem was IgA nephropathy.
We hope this information has helped you to understand more about IgA nephropathy.

If you have any more questions or concerns you should ask the nephrologist who treats you.

**The Consultants in the Renal Clinics are:**

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