



IgA Nephropathy

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What is IgA Nephropathy?

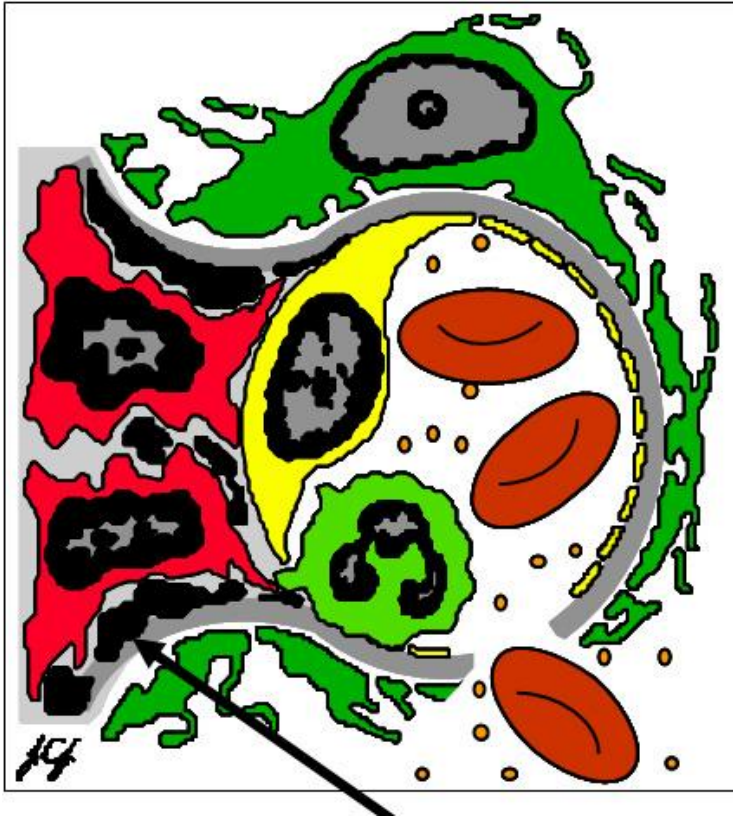
IgA Nephropathy is a relatively common kidney disease affecting millions of people worldwide. It is a disease that affects the filters, or *glomeruli*, of the kidneys. IgA is characterized by the *hematuria* it causes, which just means blood in the urine. This blood may be either *macroscopic* (visible to the naked eye) or *microscopic* (only seen under a microscope). Over time this disease can damage the kidney's ability to clean the blood properly, although most people with IgA Nephropathy lose kidney function very slowly, if at all.

IgA Nephropathy is named for the deposits of IgA that can be seen lodged in the kidney filters when viewed under a microscope. IgA is an *immunoglobulin*- a normal component of a healthy immune system. Immunoglobulins normally attach themselves to infection in the body and trigger the immune response, which works to eliminate the infection. Unfortunately in IgA Nephropathy, a defective form of IgA gets bound to another IgA molecule (instead of an infection) forming an *immune complex*. This *immune complex* can become lodged in the kidney, where it activates the immune system just like it would were it fighting off infection. The immune system activation causes inflammation and damage to the kidney itself. Though IgA Nephropathy may be suspected from certain symptoms (see below), a diagnosis of IgA Nephropathy requires a kidney biopsy. The sample of kidney can then be viewed under a microscope and the diagnosis confirmed.

What does it look like?

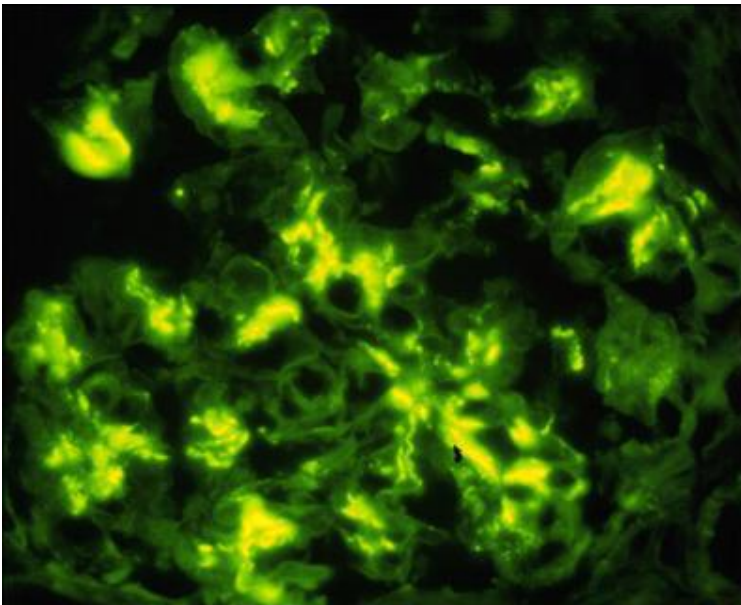
First a quick overview of the kidney- most people have two kidneys, one on each side of their lower back. All of the blood in your body passes through your kidneys many times during the day, and each time blood goes through some of it gets filtered by the glomeruli (singular = glomerulus). This filtering is how your body cleans the blood and removes extra water, and this filtered blood becomes your urine. Urine isn't red (like blood) because the red blood cells, which give blood its color, are too big to fit through the filters. A glomerulus is just a tiny bag of blood vessels through which blood gets filtered, and all of the filtered out blood (urine) runs into tubes (ureters) that eventually lead to your bladder.

The drawing below shows how IgA Nephropathy affects the kidney. Pictured is a single blood vessel from a glomerulus, shown in cross section. The red disks are red blood cells, and they normally should remain inside the vessel itself (shown as a ring of yellow, gray, and green).



Because the IgA deposits have activated the immune system and damaged the vessel wall, however, red blood cells (and protein, the tiny yellow dots) are spilling out the bottom of the vessel and into the urine.

Here is a picture from the actual kidney biopsy of someone with IgA Nephropathy. Shown is a single glomerulus, in which the actual IgA deposits have been stained fluorescent green.



How did I get it?

IgA is an *autoimmune disease*, a disease in which a person's own immune system mistakenly damages their own body. Though scientists know that the IgA immune complex (see above) causes the inappropriate immune response, it is not yet known what causes the defective IgA in the first place. It likely has both genetic and environmental components; a person is born with a predisposition for the disease, and then some sort of "trigger", for example an infection or food exposure, turns the disease "on".

While IgA commonly occurs in Caucasians and Asians, it is relatively uncommon in those of African descent. It is twice as common in males as females, and though it potentially affects any age group IgA Nephropathy is most commonly diagnosed in early and middle adulthood.

What are the symptoms?

Microscopic hematuria (blood in the urine), either constant or episodic, is the most common finding in people with IgA Nephropathy. Sometimes this blood can become visible as well (macroscopic), though when it does the urine typically appears more brown or "cola" colored than bright red. Bouts of macroscopic hematuria from IgA Nephropathy often occur during or immediately after other transient illnesses, such as an upper respiratory infection.

In addition to hematuria, people with IgA Nephropathy can have proteinuria (protein in the urine) as well. While the amount of proteinuria is generally less than 3.5g, or *sub-nephrotic* (see the "Nephrotic Syndrome" elsewhere on this site, it can sometimes result in significant leg swelling and fluid retention. Though IgA Nephropathy can be suspected from hematuria and other symptoms, it can only be diagnosed by way of a renal biopsy.

Treatment

Despite its prevalence, there is not a universally accepted treatment for IgA Nephropathy. This is in part because of its tendency to progress very slowly, if at all, and the toxicity of many of the drugs that could be used.

It is generally accepted that blood pressure control and limiting the amount of protein in the urine are of paramount importance. Both of these goals can often be accomplished with the use of two types of blood pressure medications- ACE-inhibitors (angiotensin converting enzyme inhibitors) and ARBs (angiotensin II receptor blockers). Another commonly used agent is Fish Oil; though studies are conflicting regarding its true benefit, its lack of serious side-effects lead many doctors to recommend it.

Since IgA Nephropathy is essentially an over-activation of the immune system, many *immunosuppressive* drugs have been tried with varying success. The most widely used are steroids, given either intravenously, by mouth, or both for at least 6 months.

Because of it often progresses so slowly, it is not inappropriate to simply follow some patients with normal renal function and minimal proteinuria without starting any therapy at all. It is the disease's generally slow progression that has made it difficult for doctors to decide on the one "best" treatment.

Prognosis

Though some patients spontaneously undergo a complete remission of symptoms and never experience renal insufficiency, a more typical course would be either stability of symptoms from diagnosis or a slowly progressive disease. On average, at 20 years (from diagnosis), 20 % of patients will have progressed to end stage renal disease, requiring either dialysis or a kidney transplant.

Rarely, a more rapidly progressive type of IgA Nephropathy is seen on biopsy. In these patients, immediate and aggressive immunosuppression is generally required.

Kidney Transplant in IgA Nephropathy

A portion of the patients diagnosed with IgA nephropathy will eventually progress to renal failure. Fortunately, kidney transplant is a treatment option for these patients.

For some general information about kidney transplant, click [here](#).

Will the IgA nephropathy come back in my kidney transplant?

Recurrence of IgA nephropathy is fairly common after a kidney transplant. If you did a kidney biopsy of all transplant patients with IgA nephropathy, about 60% of them would have evidence of the disease in the biopsy specimen, but not all would have symptoms or other abnormal test results. About 20-40% of patients actually have abnormal protein or blood detected in their urine due to recurrent IgA nephropathy. .

Recurrence usually happens, on average, around 2.5 years after the transplant, but can occur at any time.

Is there any treatment for IgA nephropathy that comes back in a transplant?

There are no specific treatments for IgA nephropathy in transplant patients. The treatment is similar to the treatment of IgA nephropathy in your original kidneys. This includes blood pressure control and immunosuppressive medications for more severe cases.

If the IgA nephropathy does come back, will it cause me to lose my kidney transplant?

Fortunately, renal function is usually good for several years after recurrence of IgA nephropathy in a transplant. After about 5 years, the rate of kidney loss due to the disease is increased. About 40% of patients who have recurrent IgA nephropathy will eventually lose their kidney because of the disease, but it can take up to 10 years or more for that to happen.



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