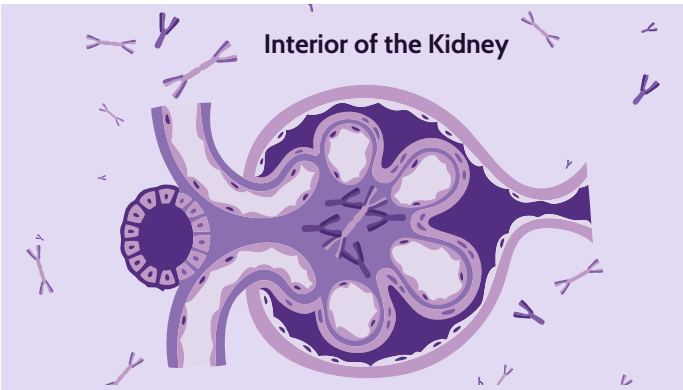
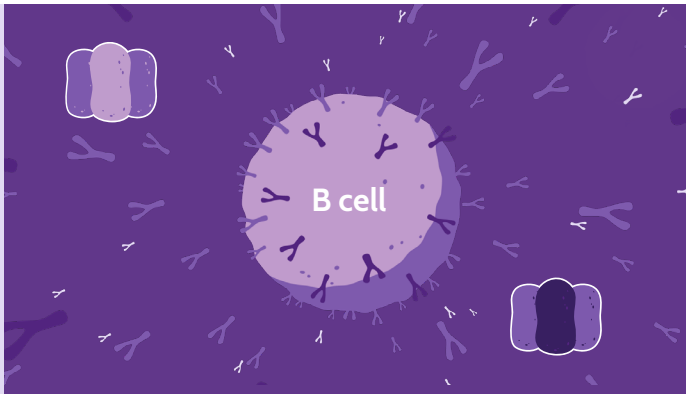
IgA Nephropathy:
Understanding This Serious Kidney Disease

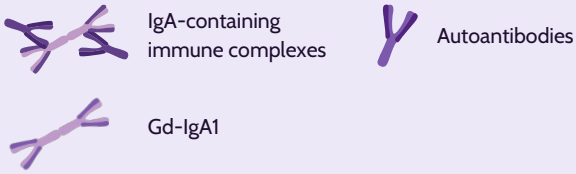
IgA nephropathy (IgAN) is a serious autoimmune kidney disease resulting from uncontrolled B cells that cause damage to the tiny blood vessel networks (glomeruli) responsible for filtering blood in the kidneys. In many cases, people with IgAN progress to end-stage kidney disease or kidney failure within 20 years of diagnosis.

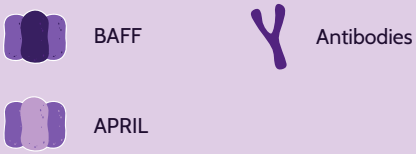


Immune complexes resulting from abnormal IgA deposit in the kidneys, triggering damage and inflammation



BAFF and APRIL play a role in uncontrolled B cell activity in IgAN





What causes IgAN?

In IgAN, the body produces an abnormal form of the IgA antibody called galactose-deficient IgA1 (Gd-IgA1). IgA normally helps fight infections, but instead of protecting the body, Gd-IgA1 triggers an abnormal immune response, causing the body to make antibodies against it (autoantibodies). The autoantibodies combine with the Gd-IgA1 antibodies to form clusters called immune complexes, which build up in the kidneys. Over time, this buildup leads to damage and inflammation, especially within the kidney filters (glomeruli), making it harder for the kidneys to remove waste from the blood.

How do uncontrolled B cells impact IgAN?

IgAN is an autoimmune disease that is driven by uncontrolled B cells. B cells are one of the key components of the body's immune system and play an important role in protecting it from foreign invaders (viruses, bacteria, etc.). In diseases driven by uncontrolled B cells, there is a mistaken attack on healthy tissue, and in the case of IgAN, the attack is on the kidney.

Specifically, two types of proteins called BAFF (B cell-activating factor) and APRIL (A proliferation inducing ligand) play key roles in the lifecycle of B cells and are elevated in people with IgAN. Uncontrolled B cells, driven by BAFF and APRIL, produce increased levels of disease-causing antibodies, which in turn, result in kidney damage.

The serious impact of IgAN

IgAN is the most common form of primary glomerulonephritis worldwide, and a high percentage of people with IgAN progress to end-stage kidney disease, often necessitating long-term dialysis or a kidney transplant. IgAN can recur after a kidney transplant.

Prevalence:
~160,000 diagnosed people with IgAN in the U.S.

Diagnosis:
Kidney function is assessed by testing the patient's blood and urine. A kidney biopsy is required to confirm a diagnosis of IgAN.

Median age at diagnosis:
IgAN can occur at any age, but is most commonly diagnosed when patients are between 20 and 40 years old.

Progression:
IgAN is a progressive disease, with up to 72% of people with IgAN progressing to end-stage kidney disease within 20 years of diagnosis.

Signs and symptoms

IgAN can be silent for years before diagnosis. When symptoms appear, they may include:

Proteinuria:
elevated protein in urine; may have a foamy appearance

Hematuria:
blood in urine, can be visible or microscopic

Fatigue:
worsens as kidney function declines

High blood pressure

Swelling or edema:
in hands, feet, or around the eyes

Progression and severity of IgAN can be measured via estimated glomerular filtration rate (eGFR), a key metric of kidney function that reflects the volume of blood the glomeruli filter per minute.