Anti-GBM (Goodpasture's) Disease

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What is anti-GBM disease?
Anti-glomerular basement membrane (GBM) disease, also known as Goodpasture’s disease, is a rare condition that causes inflammation of the small blood vessels in the kidneys and lungs.

Who are affected?
This disease tends to affect two age groups - young people aged 20 to 30, and older people in their 60s and 70s. Men are affected slightly more often than women.

What is the aetiology (cause)?
The disease occurs when the body’s immune system attacks the lining (known as the basement membrane) of the small blood vessels in the kidneys and lungs. It is not currently known why this occurs, although people with a particular "tissue type" seem to be at greater risk. Lung involvement is more common in cigarette smokers, and it is thought smoking may also be a trigger for developing the disease.

What are the symptoms?
Anti-GBM disease usually develops suddenly, over a period of a few weeks. The kidneys, the lungs, or both may be affected. About 50 per cent of patients have lung disease in addition to kidney disease, and isolated lung disease is very rare. Kidney involvement may not cause symptoms at first, although blood and protein may be detected in the urine. As the disease progresses, patients may develop signs of kidney failure such as tiredness, poor appetite, decreased urine production, breathlessness and leg swelling. When the lungs are involved, patients may have severe breathless, dry cough, or coughing up blood.

Making a diagnosis
The diagnosis of anti-GBM disease usually relies on a blood test to identify anti-GBM antibodies. Often a kidney biopsy (taking a small sample of kidney tissue with a needle) is required to demonstrate that the kidneys are involved. This may also confirm the diagnosis, by showing deposition of anti-GBM antibodies in the kidney. Other tests (such as X-rays or CT scans) are often used to show if the lungs are involved.
Treatment

Treatment requires a process called plasma exchange, which involves the use of a machine to remove anti-GBM antibodies from the bloodstream. This is often done daily for 2 weeks. In addition, immunosuppressive drugs such as steroids and cyclophosphamide, are used to suppress inflammation and stop further antibody production. Treatment usually continues for 6 months after the diagnosis is made.

For information on plasma exchange:
- Plasma exchange or plasmapheresis

Drugs and Side Effects

For information on the main drugs prescribed for Anti-GBM (Goodpasture's) disease see:
- Cyclophosphamide
- Steroids

For information on other drugs used in the treatment of vasculitis see Glossary of drugs and side effects.

Prognosis

When treatment is started promptly, before the need for dialysis, the majority of patients recover to have normal kidney and lung function. However, patients already on dialysis are unlikely to recover kidney function. Unlike some other forms of vasculitis, it is very rare for anti-GBM disease to relapse, and thus it does not usually require long-term treatment. Smoking is thought to be a possible trigger for causing relapses, and should be avoided. In patients remaining on dialysis, kidney transplantation is possible once anti-GBM antibodies have become undetectable.

Key points

- Anti-GBM disease is a rare form of vasculitis
- It causes damage to small blood vessels in the kidneys and lungs
- Diagnosis is confirmed by detecting anti-GBM antibodies in blood or deposited in the kidney
- Treatment with plasma exchange (to remove anti-GBM antibodies) together with prednisolone and cyclophosphamide is usually successful

Additional note

by Dr S McAdoo and Prof C Pusey

Anti-GBM and ANCA ‘double-positives’

It is recognised that some patients with anti-GBM disease are also found to have ANCA, which are antibodies usually associated with other forms of small vessel vasculitis such as Granulomatosis with Polyangiitis and Microscopic Polyangiitis (see individual diseases for more information). We believe these “double positive” patients should receive initial treatment as for anti-GBM disease, including plasma exchange, at the time of diagnosis. Our experience suggests that, despite having both types of antibodies, the response to this treatment is similar to patients with anti-GBM disease alone. However, unlike patients with isolated anti-GBM disease, they are at risk of future relapses related to their ANCA, and so we recommend that they continue on longer-term maintenance treatment (like those patients with isolated ANCA-related disease.)
Further reading

- The Kingston Whig - Rare condition will cost ailing teenager her kidneys

Personal story

The personal story of a young man with Anti GBM (Goodpasture's Disease) can be viewed at: Gareth's Story