

Anti-Glomerular Basement Membrane (GBM) Disease (Goodpasture's Syndrome)

What is it?

Goodpasture's Syndrome is a type of vasculitis (inflammation of blood vessels), which affects the kidneys and the lungs.

What causes it?

The body normally produces antibodies to fight off infection and disease. In this case, your body makes an antibody that can attack and damages a membrane in your kidneys and lungs.

What symptoms might I have?

You may feel short of breath and cough up blood.

The kidney damage may cause blood-stained or frothy urine or actual kidney failure.

Is it serious?

Without treatment, the condition can be life-threatening. In some cases, it may be too advanced for treatment to save the kidneys and dialysis will be necessary. However, powerful treatment is now very successful in saving lives and kidney function.

Inpatient treatment

As soon as diagnosis is made (on blood test or kidney biopsy), treatment will start with *Prednisolone* (steroid) and *Cyclophosphamide* (immunosuppressant).

- *Prednisolone* 1mg/Kg of body weight (max 60mg)
- *IV Cyclophosphamide*
- Plasma exchange daily for 14 days or until antibody negative

Discharge medication [Week One]

Prednisolone	Inpatient dose
<i>Lansoprazole</i>	30mg daily
<i>Alendronate</i> (non-dialysis)	70mg weekly
<i>Nystatin</i>	1ml four times a day
<i>Seprin</i>	480mg daily

Outpatient treatment

The condition is dangerous, requiring powerful treatment that can have serious side effects. You will be seen often and monitored carefully. Your blood will be checked for its white cell count (WCC) to check how it would respond to infection. Visits will initially be weekly and gradually become less often as drug doses are reduced.

Week 2	<i>Prednisolone</i> 45mg	
Week 3	<i>Prednisolone</i> 30mg	
Week 4	<i>Prednisolone</i> 25mg	Stop Nystatin
Week 5	<i>Prednisolone</i> 20 mg	
Week 6	<i>Prednisolone</i> 20 mg	
Week 9	<i>Prednis</i> 20/15 alt day)	
Week 12	<i>Prednisolone</i> 15mg	Stop <i>Seprin</i> & <i>Lansoprazole</i>
Mth 4	<i>Prednisolone</i> 10mg	
Mth 5	<i>Prednisolone</i> 5mg	Stop <i>Alendronate</i>
Mth 6	Stop <i>Prednisolone</i>	

Some possible side effects of the treatment:

Prednisolone:

- Infection - Steroids make you more prone to 'opportunistic infection' (so-called, because they only affect vulnerable people). You will be given antibiotics (*Seprin* and *Fluconazole*), to protect against thrush and pneumonia. If you have had previous TB exposure, you will be given *Isoniazid*. If you develop fever, cough, sore throat, contact us immediately.
- Osteoporosis - There is a risk that higher doses of steroids may weaken bones; you will be given *Alendronate* to prevent this.
- Stomach irritation - You will be given *Lansoprazole* to prevent irritation to the lining of your stomach.
- Appetite - Steroids nearly always increase appetite. To avoid gaining weight, you may need to be careful about the amount you eat. (Advice available from Dietician in Renal Clinic)
- Diabetes - High doses may affect your body's ability to handle sugar. Up to 20% of patients may develop temporary diabetes, some requiring treatment.
- Skin and muscles - Steroids may make the skin thinner and more likely to bruise and the larger muscles weaker.
- Blood pressure - A degree of fluid retention may cause the blood pressure to rise.
- Mood - Some patients find steroids cause mood disturbance- mood change, psychosis, insomnia.

Azathioprine

- Anaemia.
- Low white cell count with risk of severe infection.
- Temporary liver disorders.
- Major interaction with *Allopurinol* (given for gout) - danger of severe infection.
- Hair may become thinner / alopecia.

Cyclophosphamide

- Low white cell count with risk of severe infection.
- Thrombocytopenia (reduced number of platelets, which can result in bleeding/bruising).
- Inflammation and bleeding from the bladder.
- Hair may become thinner / alopecia.
- Nausea and occasional vomiting.
- There may be an increased risk of cancers (such as bladder and bone marrow) depending on the total dose. However, current doses are considerably less than prescribed in the past.
- Pulmonary fibrosis.
- If the total dose is greater than 50 mg /kg, there is a risk of reduced fertility. Sustained amenorrhoea (no periods) in long course women over 31yrs.

Kidney Care Nurses 0118 322 7899 Victoria Ward 0118 322 7476

This document can be made available in other languages and formats upon request.

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