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## Retroperitoneal fibrosis

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### Summary

- Retroperitoneal fibrosis is the abnormal growth of tissue on and around abdominal structures, including blood vessels and ureters.
  - Complications may include blockages in the urinary tract, and kidney failure.
  - Diagnosis can be tricky, especially in the earlier stages of the disease. This is because many symptoms are non-specific and common to other conditions.
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Retroperitoneal fibrosis (RPF) is the abnormal growth of white and 'woody' tissue on and around the structures of the abdomen that are outside the peritoneum. Occasionally, the tissue can be cancerous. RPF is also known as Ormond's disease.

The peritoneum is the membranous tissue that lines the abdominal cavity and covers the abdominal organs.

Abdominal structures located outside of the peritoneum, known as retroperitoneal structures, include blood vessels such as the aorta (the artery that carries blood from the heart to the body) and the tubes that connect the kidneys to the bladder (ureters).

Retroperitoneal fibrosis is a rare condition that develops in about one in 200,000 people. While it can affect anyone at any age, those at highest risk (for reasons that are not clear) are men aged between 40 and 60 years.

In the later stages of the disease, the person may develop complications, including blockages within the urinary tract, and kidney failure.

### Cause of retroperitoneal fibrosis

It is thought that RPF might be a type of autoimmune disorder (where the body's immune system mistakenly attacks and destroys healthy body tissues). In about seven out of 10 people with RPF, no underlying cause is found – this is called 'idiopathic RPF'.

Doctors still don't know why these fibrous masses grow. Inflammation can be present in some cases, which suggests the involvement of immune system cells. Some people with RPF have pre-existing autoimmune disease such as Hashimoto thyroiditis, rheumatoid arthritis or systemic lupus erythematosus (SLE).

In the remaining cases, doctors can trace the development of RPF to a specific trigger such as:

- prescription drugs, including certain beta-blockers (used to treat heart and blood pressure conditions) and an ergot derivative used in the past for migraine
- illegal drugs, including cocaine
- abdominal aortic aneurysm (abnormal swelling of the aorta)
- urinary tract injury
- infection
- cancer in any of the retroperitoneal structures – the resulting RPF is known as 'malignant RPF'
- cancer therapy, including chemotherapy.

### Symptoms of retroperitoneal fibrosis

The symptoms of RPF tend to worsen with time and may include:

- abdominal pain
  - pain in nearby areas such as the back or scrotum
  - circulation problems in the legs, which can cause pain and skin discolouration
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- fever
- malaise (a general, non-specific, feeling of unwellness)
- weight loss
- nausea and vomiting
- urination problems such as bloody urine, decreased urination, excessive urination or needing to urinate during the night
- excessive thirst
- inability to urinate, which is a medical emergency.

### **Complications of retroperitoneal fibrosis**

Problems involving the urinary tract are common complications of RPF. The kidneys filter waste products and excess fluid from the blood to make urine. Urine leaves the kidneys through tubes called ureters and is stored temporarily in the bladder. Urine leaves the bladder through a tube called the urethra.

Obstructive uropathy is a blockage of the ureter. RPF can cause obstructive uropathy of either one (unilateral) or both (bilateral) ureters. The inability to urinate at all is known as 'anuria'. This is a dangerous condition and needs immediate medical attention. Without medical attention, the kidneys may fail and the build-up of waste products in the blood may cause symptoms such as vomiting and mental confusion.

Retroperitoneal fibrosis can also cause other complications, depending on which structure is blocked or compressed by the fibrous growths.

Some examples of the affected areas and the complications that can occur are:

- blood vessels – can cause high blood pressure (hypertension)
- bowel – can cause constipation
- bile duct – can cause jaundice
- lymphatic vessels – can cause swelling of the leg or legs.

### **Diagnosis of retroperitoneal fibrosis**

Diagnosis can be tricky, especially in the earlier stages of the disease. This is because many RPF symptoms are non-specific and are common to other conditions. To complicate matters further, RPF looks similar on scan images to some other conditions, such as pancreatic cancer or abdominal aortic aneurysm.

Tests used in the diagnosis of RPF may include:

- physical examination
- medical history
- blood tests
- urine tests
- x-ray examination
- contrast examination, such as barium meal x-ray
- kidney ultrasound
- abdominal CT scans
- abdominal MRI scans
- exploratory surgery, including biopsy (removal of a tissue sample for examination in a laboratory).

### **Treatment for retroperitoneal fibrosis**

The treatment used for RPF depends on the underlying cause, the severity of the condition and the degree of damage caused by any complications. Options may include:

- addressing the known trigger – for example, stopping the particular medication if the RPF is medication-related
- treatment of underlying condition – for example, cancer therapy

- medications, including anti-inflammatory medications such as corticosteroids
- medications that suppress the activity of the immune system, such as cyclophosphamide
- drainage of the kidneys using a thin tube inserted through the skin (percutaneous nephrostomy)
- surgery to remove the fibrous masses
- surgery to insert hollow tubes (stents) into the blocked ureters to prop them open
- treatment for complications – for example, chronic kidney failure may need dialysis or organ transplant.

With appropriate and timely medical care, a person with benign (non-cancerous) RPF has a positive long-term outlook. However, in the case of malignant (cancerous) RPF, outcomes can vary and death is possible.

### **Where to get help**

- In an emergency, call triple zero (000)
- Emergency department of your nearest hospital
- Your **GP (doctor)**
- **Kidney Health Australia** helpline Tel. **1800 454 363**

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