CONDITION

Retroperitoneal fibrosis

Retroperitoneal fibrosis is inflammation and scar-like tissue that can block the tubes carrying urine from the kidneys.

WHAT IT IS
Inflammatory scar-like tissue behind the abdomen

BODY SYSTEM
Urinary system and tissues around major abdominal blood vessels

MAIN SYMPTOMS
Back or side pain, abdominal pain, reduced urine, fatigue

WHAT CAUSES IT
Usually unknown or immune-related; sometimes another condition or treatment

HOW IT IS DIAGNOSED
CT or MRI imaging, with biopsy when needed

MAIN TREATMENT
Steroids, urine drainage, and sometimes immune medicines or surgery

CAN IT BE CURED?
Sometimes - treatment can produce lasting remission

IS IT AN EMERGENCY?
Sometimes - little or no urine from blocked ureters

LONG-TERM OUTLOOK
Often manageable, but relapse and kidney damage can occur

Overview

Retroperitoneal fibrosis is a rare condition in which inflamed, scar-like tissue develops deep inside the abdomen. This area, called the retroperitoneum, lies behind the lining of the abdominal cavity.

The tissue usually forms around the aorta, the body's largest artery, and nearby blood vessels. It can surround or squeeze the ureters. These are the tubes that carry urine from each kidney to the bladder.

If a ureter becomes narrow or blocked, urine can build up in the kidney. This is called hydronephrosis. A long-lasting blockage can reduce kidney function, sometimes without causing obvious urinary symptoms.

The condition is not itself cancer. However, some cancers and other conditions can produce a similar appearance. Careful imaging and, in some cases, a tissue biopsy are needed to tell them apart.

Treatment aims to relieve any urinary blockage, stop active inflammation and protect the kidneys. Long-term follow-up matters because the condition can return after it has improved.

Symptoms

Symptoms often develop gradually. The most common is a dull, constant pain in the lower back, side or abdomen. It may be difficult to point to one exact location. Pain may spread toward the groin or upper thigh.

Some people first seek care because they feel generally unwell rather than because of urinary symptoms. Others have no symptoms, and the condition is found during imaging for another reason.

Possible symptoms include:

A blocked ureter may not hurt. One kidney can sometimes keep making enough urine to hide a blockage on the other side. If both ureters become severely blocked, urine output may fall sharply and kidney function can worsen quickly.

Symptoms alone cannot confirm retroperitoneal fibrosis. Back pain, fatigue and urinary changes have many more common causes.

- Persistent back, side, pelvic or abdominal pain
- Tiredness, reduced appetite, nausea or unintended weight loss
- Less urine than usual or difficulty passing urine
- Swelling of a leg or the scrotum from pressure on nearby blood vessels
- Fever or a general feeling of illness during active inflammation

Causes and risk factors

In many cases, no separate cause is found. This is called idiopathic retroperitoneal fibrosis. "Idiopathic" means that the cause is not known.

The immune system appears to drive much of the inflammation. Some cases are part of IgG4-related disease, an immune-mediated condition that can cause inflammation and scar-like tissue in several organs. It may also affect the pancreas, salivary glands, kidneys or

major blood vessels.

Less often, retroperitoneal fibrosis develops in connection with another factor. These secondary cases have been linked to certain cancers, infections, abdominal surgery, radiotherapy and some medicines. Older ergot-derived migraine medicines and certain dopamine-acting medicines are among those reported in medical literature.

Do not stop a prescribed medicine because of a possible link. A clinician can review the timing, benefits and risks and decide whether an alternative is appropriate.

The condition is diagnosed more often in men and most often during middle or later adulthood, although anyone can develop it. Smoking and asbestos exposure have also been associated with idiopathic retroperitoneal fibrosis. An association does not prove that either exposure directly caused an individual case.

Diagnosis

Diagnosis usually begins with blood tests and imaging. Blood tests check kidney function and may show inflammation or anaemia, which means a reduced number of red blood cells. Urine tests may help assess kidney health or look for infection.

Inflammatory markers, such as C-reactive protein and the erythrocyte sedimentation rate, may be raised. They can also be normal and are not specific to this condition. A blood IgG4 level may support a diagnosis of IgG4-related disease, but it cannot confirm or exclude it by itself.

A CT scan usually shows where the abnormal tissue is, whether it surrounds the blood vessels or ureters, and whether urine is backing up into a kidney. MRI can provide similar information and may be used when it is a better option for the individual.

An ultrasound can show hydronephrosis, but it does not usually define the retroperitoneal tissue as clearly as CT or MRI. PET imaging is sometimes used to look for active inflammation or involvement elsewhere, although its findings are not unique to retroperitoneal fibrosis.

A biopsy removes a small tissue sample for examination under a microscope. It may be recommended if the scan appearance is unusual, cancer or infection is a concern, or the condition does not respond as expected. Not everyone needs a biopsy.

The assessment may also look for an underlying cancer, infection, medicine exposure or signs of IgG4-related disease.

Treatment and management

Treatment is planned around two immediate questions: whether the kidneys are obstructed and whether the inflammation is active.

A severe ureter blockage may need to be drained promptly. A thin internal tube called a ureteral stent can hold the ureter open. Another option is a nephrostomy, in which a tube drains urine directly from the kidney through the skin. These measures protect kidney function but do not treat the underlying inflammation.

Glucocorticoids, often called steroids, are commonly used to reduce inflammation in idiopathic retroperitoneal fibrosis. Treatment usually starts under specialist supervision and is reduced gradually rather than stopped suddenly.

Other medicines that alter immune activity may be considered when steroids like prednisone or hydrocortisone are not suitable, cause troublesome effects, or do not control the condition. They may also be used to reduce steroid exposure or treat IgG4-related disease. The choice depends on the suspected cause, other health conditions and response to earlier treatment.

Surgery may be needed if ureter obstruction continues despite medical treatment or cannot be managed with stents. Ureterolysis is an operation that frees the ureters from surrounding fibrous tissue and moves or protects them to reduce the chance of further trapping. Surgery can relieve obstruction, but inflammation can still remain or return.

Follow-up commonly includes kidney blood tests, symptom reviews and repeat imaging. If a stent or nephrostomy is in place, it requires planned care and may need removal or replacement.

Complications

The main complication is kidney damage caused by blocked urine flow. Early relief of obstruction often allows kidney function to improve, but recovery may be incomplete if the blockage was severe or long-lasting.

Other possible complications include:

Treatment can also have unwanted effects. Long courses of steroids may affect bones, blood sugar, blood pressure, mood, sleep and infection risk. Other immune treatments have their own monitoring needs. These risks are balanced against the need to control inflammation and protect the kidneys.

- Chronic kidney disease
- Urinary infection, especially when drainage devices are present
- Repeated or persistent ureter obstruction

- Leg swelling or, rarely, a blood clot from pressure on veins
- Reduced blood flow from involvement around major arteries
- Relapse after treatment has ended

Prevention

Most idiopathic and IgG4-related cases cannot currently be prevented because their exact cause is not known.

Avoiding smoking and following workplace precautions around asbestos are sensible for overall health, but they cannot guarantee that retroperitoneal fibrosis will not occur.

Secondary cases may sometimes be reduced by addressing the cause, such as changing an implicated medicine or treating an infection. This should be done with medical guidance rather than by stopping treatment on your own.

Once retroperitoneal fibrosis has developed, regular monitoring can help prevent avoidable kidney damage by finding recurrent obstruction early.

Living with it and what to expect

During the first weeks or months, care often focuses on restoring urine flow and bringing inflammation under control. Pain and general symptoms may improve before scans show a major change. Kidney function may recover after drainage, although the amount of recovery is not predictable.

The tissue seen on a scan may not disappear completely. Residual scar-like tissue can remain even when the inflammation is no longer active. Clinicians therefore consider symptoms, kidney tests, inflammatory markers and imaging together.

Some people have one episode followed by a lasting remission. Others need treatment for a longer period or have relapses months or years later. A relapse may cause renewed pain or changes in kidney tests, but it can also be found on routine imaging before symptoms appear.

Keeping scheduled follow-up is important even when you feel well. Monitoring may become less frequent when the condition is stable, but long-term checks are often needed.

Most people can continue their usual activities as symptoms and kidney function improve. Temporary adjustments may be needed around procedures, fatigue or medicine effects. If kidney damage remains, ongoing kidney care may become part of long-term management.

When to get medical help

Seek emergency care if you pass little or no urine, especially if this is accompanied by severe weakness, vomiting, confusion, breathlessness or rapidly increasing swelling. These can be signs of serious kidney obstruction or kidney failure.

Fever or shaking chills together with side or back pain, reduced urine, or a urinary stent or nephrostomy also needs urgent assessment. An infection behind a blocked ureter can become serious and may require drainage as well as antibiotics like amoxicillin or doxycycline .

Contact your healthcare team promptly if you notice:

Routine appointments are the right place to discuss mild ongoing fatigue, stable discomfort, medicine concerns and monitoring results. Do not wait for symptoms before attending planned kidney tests or imaging.

- A clear reduction in urine
- New or worsening back, side or abdominal pain
- Blood in the urine
- New leg or scrotal swelling
- Problems with a stent or nephrostomy
- Significant side effects from steroids or other immune medicines

Common questions

Is retroperitoneal fibrosis cancer?

No. It is an inflammatory and fibrotic condition, not a cancer. Some cancers can look similar on a scan or can rarely cause secondary fibrosis, so a biopsy or further testing may be needed when the diagnosis is uncertain.

Can retroperitoneal fibrosis go away?

Active inflammation can enter remission with treatment, and the abnormal tissue may shrink. Some scar-like tissue may remain on scans. Lasting remission is possible, but relapse can occur.

Is it the same as IgG4-related disease?

Not always. Some cases are part of IgG4-related disease, while many are not. Blood tests, imaging, examination for other affected organs and sometimes a biopsy help make this distinction.

Will I need a ureteral stent forever?

Not necessarily. A stent may be temporary while treatment reduces inflammation. How long it is needed depends on whether the ureter remains narrowed and whether kidney drainage has improved. Stents left in place require scheduled review and replacement or removal.

Can retroperitoneal fibrosis come back after treatment?

Yes. Relapse can occur during treatment reduction or after treatment has ended. Regular kidney tests and imaging are intended to identify this before lasting damage develops.

Will I need surgery?

Many people are managed with urine drainage and medicines. Surgery may be considered when obstruction continues, repeatedly returns or cannot be managed adequately with a stent or nephrostomy.

Sources

- Vaglio A, Maritati F. Idiopathic Retroperitoneal Fibrosis . Journal of the American Society of Nephrology . 2016;27(7):1880-1889.
- Vaglio A, Salvarani C, Buzio C. Retroperitoneal fibrosis . The Lancet . 2006;367(9506):241-251.
- Scheel PJ Jr, Feeley N. Retroperitoneal fibrosis: the clinical, laboratory, and radiographic presentation . Medicine . 2009;88(4):202-207.
- Khosroshahi A, Wallace ZS, Crowe JL, et al. International consensus guidance statement on the management and treatment of IgG4-related disease . Arthritis & Rheumatology . 2015;67(7):1688-1699.
- National Organization for Rare Disorders. Retroperitoneal Fibrosis . Rare Disease Database . Accessed August 22, 2026.