



# IgG4ward!

## Understanding Retroperitoneal Fibrosis (RPF) and IgG4-related disease (IgG4-RD)

*A reading companion to the IgG4ward! Fireside Chat with Dr. John Stone and Dr. James Tomlinson*

### What is Retroperitoneal Fibrosis (RPF)?

Retroperitoneal Fibrosis (RPF) is a condition where a thick, inflamed tissue forms deep inside the abdomen, in a space called the retroperitoneum. This area includes important organs like the **kidneys, ureters** (the tubes that carry urine from the kidneys to the bladder) and **major blood vessels**.

In people with IgG4-related disease (IgG4-RD), the immune system can create inflammation in this area, which may turn into **fibrosis** (scar tissue). When the ureters get trapped or blocked by this tissue, it can lead to serious problems like **urine backup, kidney damage, pain** and even kidney failure if left untreated.

### What You Will Learn in the Fireside Chat

This conversation includes both **medical expertise** and **real patient experiences**. You will hear from:

- **Dr. John Stone** (Rheumatologist) and **Dr. James Tomlinson** (Nephrologist), who explain how RPF presents, how it is diagnosed and what treatment looks like.
- **Patients and caregivers** who share their lived experiences with RPF, including delayed diagnosis, stents, surgeries and emotional stress.

### Key Takeaways:

#### 1. Symptoms Aren't Always Obvious – and That Can Delay Diagnosis

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RPF symptoms can be hard to recognize. They often come on slowly and may be brushed off as other, more common health issues. For example, back pain might be mistaken for a muscle strain, or bloating might be attributed to digestion.

## Common symptoms include:

- Dull pain in the lower back, abdomen or sides
- A feeling of fullness or bloating
- Reduced urination, or pain when urinating
- Swelling in legs or feet
- Fatigue, sometimes extreme
- High blood pressure

For many people with RPF, these symptoms are caused by pressure from inflammation and scar tissue in the retroperitoneum. If the **ureters are compressed**, urine can't flow properly from the kidneys to the bladder, leading to serious complications like kidney damage.

In the Fireside Chat, several stories reflected how symptoms were present for a long time but were overlooked until serious organ damage occurred. Early awareness and imaging can make a big difference.

## 2. RPF is Often Mistaken for Cancer

One of the most emotional aspects of the IgG4-RD & RPF journeys is that many are first told they may have cancer. This is because imaging scans (like CT or MRI) often show masses or tissue growths that look similar to cancerous tumors. Doctors will usually want to rule out malignancy quickly, which may mean referrals or recommendations for biopsy.

That period of uncertainty can cause a lot of fear. But once cancer is ruled out, patients are sometimes left without clear next steps until they are referred to a physician familiar with IgG4-RD. This highlights why education about rare disease is so important: when providers recognize patterns consistent with IgG4-RD, they can help patients move toward treatment faster and with less emotional trauma.

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## 3. Diagnosis Involves Imaging – And Sometimes Biopsy

**RPF is typically found on imaging scans such as:**

- CT scans, which show blockages or tissue thickening
- MRI, which may better show soft tissue inflammation
- PET scans, used to detect areas of active inflammation

**Depending on the case, physicians may also:**

- Order a biopsy to confirm inflammation and rule out malignancy
- Check bloodwork, including IgG4 levels (although these can be normal in some people)
- Monitor kidney function through urine and blood tests

Importantly, not everyone with IgG4-RD has elevated IgG4 levels – so a “normal” result does not rule out the disease. A diagnosis is often made by imaging, labs, symptoms and sometimes a biopsy.

In the Fireside Chat, Dr. Tomlinson, emphasized how early imaging can reveal the disease even before major symptoms appear, especially in at-risk patients. However, since RPF is rare, many people see multiple doctors before someone makes an accurate diagnosis of RPF.

## 4. Treatment is Often Combination of Medicine and Procedure

Treatment for RPF depends on the severity, the level of fibrosis (scar tissue) and whether the kidneys or ureters are at risk.

**Medical treatment options include:**

- **Steroids** to reduce inflammation
- **B-cell depletion** therapies, such as *Rituximab* or *Inebilizumab*, which target immune cells that drive the disease

**Procedures or surgeries may include:**

- **Ureteral stents** to keep the ureters open
- **Nephrostomy tubes** if urine needs to drain directly from the kidneys

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- **Ureterolysis**, a surgery to remove or move the ureters away from fibrotic tissue

Some people respond well to medications alone, while others need surgical interventions. Many patients, live for years with stents and must manage the challenges of pain, infection and uncertainty.

Dr. Stone and Dr. Tomlinson emphasized the importance of ongoing monitoring, since scar tissue may persist even after inflammation goes down. It is also important to work with a team of specialists- rheumatologists, urologists, nephrologists – to ensure care is coordinated.

## Questions to Ask Your Physician

- Are my kidneys or ureters affected?
- Is a stent or nephrostomy tube necessary for me? (If yes, please explain why)
- Would B-cell therapy be appropriate?
- How will we monitor if the disease is active?
- Should I be referred to a urologist or nephrologist?
- What follow-up imaging or labs will I need

## Did You Know?

- RPF can appear on scans before symptoms start
- Scar tissue may still appear even when the disease is no longer active
- Patients may need stents if inflammation or scarring remains

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