

## **Pulmonary vasculitides**

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**Introduction:** The vasculitides are a diverse group of disorders that are characterised by inflammation of the blood vessels including arteries, veins or capillaries. When such inflammation occurs it may cause structural changes in the walls of blood vessels leading to blockage or narrowing and destruction of the wall (a process often referred to as necrotising vasculitis). Pulmonary vasculitis is an uncommon condition that can occur in persons of all ages and of diverse racial background.

**Causes:** There are over 15 categories of vasculitis affecting the small, medium and large sized blood vessels. Despite their relative differences each of the pulmonary vasculitides appear to share a common theme of immune system alteration.

### **Classification:**

- Behcet's disease
- Churg-Strauss syndrome
- Cryoglobulinaemia
- Giant cell arteritis (also known as temporal arteritis)
- Henoch-Schonlein
- Hypersensitivity vasculitis (leukocytoclastic vasculitis)
- Kawasaki disease
- Microscopic polyangiitis
- Polyarthritides nodosa
- Polymyalgia rheumatica
- Wegener's granulomatosis
- Takayasu's arteritis
- Lymphomatoid granulomatosis
- Vasculitis as a result of underlying connective tissue disorders (rheumatoid arthritis, SLE, polymyositis, dermatomyositis, scleroderma)
- Secondary to other disorders including sarcoidosis

Although the diseases listed above share many of the same characteristics, symptoms and potential treatment, each is different and patients are encouraged to learn as much as possible about their individual disease.

**Clinical Presentation:** In general, initial symptoms of pulmonary vasculitis may include those outside of the chest such as fever, lethargy, sore joints or arthralgias, myalgias and a skin rash. Involvement of specific organs will lead to symptoms referable to that particular organ. For example, with pulmonary vasculitis shortness of breath and cough frequently occur and may involve the upper airways to include sinus pain and bleeding from the nasal passages. Coughing up of blood is reported in these disorders as is chest pain and a history of asthma.

**Investigations:** A range of investigations and diagnostic procedures will be undertaken in order to confirm the diagnosis of vasculitis and further differentiate the condition into a particular entity.

**Procedures will include:**

- blood tests including biochemistry cell count and inflammatory markers
- analysis of urine both for culture, chemistry and cell counts
- radiological tests including chest x-rays, CT scan, ultrasound and MRI scan
- a bronchoscopy or telescope inspection of the wind pipe and airways to identify any inflammation or areas of narrowing
- tissue biopsy of involved organs to confirm typical pathological changes within the blood vessels or other characteristic findings such as granulomas (biopsied organs may include the sinuses, nasal septum, throat, joints, lungs, skin, muscle, peripheral nerves or kidney biopsy)
- further blood tests to screen for abnormal antibodies present in the blood (eg a positive ANCA blood result may be highly suggestive of Churg-Strauss syndrome or Wegener's granulomatosis)

**Treatment:** The treatment of the pulmonary vasculitides is highly dependent on the individual type of vasculitis present. Some forms of vasculitis may be self limiting and respond to simple removal of the offending antigen (eg some cases of hypersensitivity vasculitis which may be secondary to several drugs including penicillin). Normally for other forms of vasculitis a trial of corticosteroids or prednisone is required and highly effective therapy following diagnosis. Long term remissions can be induced and maintained with medication with very close monitoring of patients. Following achievement of remission a low dose of prednisone will be desirable to maintain remission and this is known as a maintenance dosage. Commonly when steroids are withdrawn patients may experience a relapse or alternatively will require a second drug known as a steroid sparing agent to induce a longer term remission.

These drugs known as immunosuppressive agents may include Imuran (azathioprine), cyclophosphamide, cyclosporin, methotrexate and mycophenolate mofetil. Bactrim or other formulations are often prescribed with these immunosuppressants to help minimise the risk of infection. Occasionally, other non-drug treatments are required in patients with resistant disease and include immunoglobulin or plasmapheresis.

Immunoglobulin is a human derived antibody obtained from random blood donors which may neutralise circulating proteins or antibodies within the blood that may contribute to the disease. Plasmapheresis is a form of treatment where blood is removed from the patient and filtered in order to remove similarly offending proteins and antibodies before being returned to the patient. There is emerging evidence that a patient with vasculitis may experience a relapse in response to a number of non-specific triggers including infection, physical and emotional stress, trauma and surgery.

## Useful Links

*Vasculitis Foundation:* [www.vasculitisfoundation.org](http://www.vasculitisfoundation.org)







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