Polyarteritis Nodosa Factsheet

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Polyarteritis nodosa (PAN) is a form of vasculitis that affects the medium blood vessels of the body. It used to be thought that microscopic polyangiitis – which affects small blood vessels - was another form of PAN but is now known to be a condition in its own right. A separate leaflet on microscopic polyangiitis is available.

A mild form of PAN which mainly affects the skin is known as cutaneous PAN.

Treatment is mainly with steroids and/or medicines to control the activity of the immune system.

Who gets PAN?

In the UK, PAN affects approximately 2 in a million people a year. It occurs throughout the world but is more frequent in areas where hepatitis B is common. It is mainly seen in people aged 40-60. Rarely, children can be affected, usually around the age of 10. It is commoner in men than women but in children both sexes are equally affected.

What causes PAN?

The basic mechanism is inflammation of the medium sized blood vessels. Hepatitis B infection is seen in some people who also have PAN leading to the theory that the inflammation results from the response of the body's immune system. Fewer such cases are seen in the UK now that hepatitis B immunisation has become commonplace. In people who do not have Hepatitis B it has been suggested that other infections may be involved such as hepatitis C or herpes zoster virus.

What are the symptoms of PAN?

At first you may just feel generally unwell and weak. You may lose weight or get aches and pains. PAN can affect any area of the body apart, but usually not the lungs and as it progresses you may get numbness and tingling in the hands or feet or tummy pain after eating. Cutaneous PAN causes a rash, bruising, nodules (small lumps) or skin changes due to poor blood circulation. Children may just feel ill and the disease may be difficult to spot in the early stages.

How is PAN diagnosed?

The diagnosis is usually based on your symptoms and the results of tests. In contrast with some other types of vasculitis, antineutrophil cytoplasmic antibodies (ANCA) are not seen in PAN. Removal of a little bit of tissue from a blood vessel (biopsy) is often performed. In PAN this will show inflammation. Scans may also be done to confirm the diagnosis and rule out other illnesses.

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What is the treatment for PAN?

All the treatments for PAN are directed at reducing the activity of the immune system and damping down inflammation. The main treatment is steroids such as prednisolone. Other medicines which can be used are cyclophosphamide and azathioprine. In people with hepatitis B-related PAN, antiviral medicines and plasma exchange are often used. Plasma exchange involves removing the person's plasma (the liquid that remains when cells are removed from blood), purifying it and then transfusing it back into the person's blood circulation. Mild cases of cutaneous PAN may be treated with anti-inflammatory medicines.

Does PAN get better?

Providing PAN is treated the symptoms are usually adequately controlled and the disease does not come back. In older people and people who develop tummy problems the outlook may be poorer. If the illness is left untreated, life-limiting complications of the kidneys, nervous system or heart may occur.

Further reading

Mohammad AJ, et al; Prevalence of Wegener's granulomatosis, microscopic polyangiitis, polyarteritis Rheumatology (Oxford). 2007 Aug;46(8):1329-37. Epub 2007 Jun 6.

http://www.ncbi.nlm.nih.gov/pubmed/19797309

Gullevin B et al Hepatitis B virus-associated polyarteritis nodosa: clinical characteristics, outcome, and impact of treatment in 115 patients. Medicine (Baltimore).2005 Sep;84(5):313-22.

Thatayatikom A et al, Pediatric Polyarteritis Nodosa, Medscape, 2011.

http://emedicine.medscape.com/article/1007428-overview#a0156

Puéchal X et al, Therapeutic immunomodulation in systemic vasculitis: Taking stock. Joint Bone Spine. 2013 Jul;80(4):374-9.

http://www.ncbi.nlm.nih.gov/pubmed/23237994

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