https://printo.it/pediatric-rheumatology/GB/intro

Rare Juvenile Primary Systemic Vasculitis

Version of 2016

4. POLYARTERITIS NODOSA

4.1 What is it?

Polyarteritis nodosa (PAN) is a form of vessel wall-destroying (necrotising) vasculitis affecting mainly medium and small arteries. The vessel walls of many ("poly") arteries - polyarteritis - are affected in a patchy distribution. Inflamed parts of the artery wall become weaker and under the pressure of the blood stream, small nodular outpouchings (aneurysms) form along the artery. This is the origin of the name "nodosa". Cutaneous (skin) polyarteritis affects predominantly skin and musculoskeletal tissue (sometimes also muscles and joints) and not the internal organs.

4.2 How common is it?

PAN is very rare in children, with an estimated number of new cases per year of one per million. It affects boys and girls equally and is more commonly seen in children around 9-11 years of age. In children, it may be associated with streptococcal infection or much less frequently also with hepatitis B or C. Recently, a genetic form of PAN has been described called DADA2. The gene can be tested for in (very few) specialist centres, and is not always routinely available currently.

4.3 What are the main symptoms?

The most common general (constitutional) symptoms are prolonged fever, malaise, fatigue and weight loss.
The variety of localised symptoms depends on the organs affected.

Insufficient blood supply to the tissue causes pain. Therefore, pain at various sites may be a leading symptom of PAN. In children, muscle and joint pain is as frequent as abdominal pain, which is due to the involvement of gut-supplying arteries. If the vessels supplying the testes are affected, scrotal pain may also occur. Skin disease can present as a wide range of changes from painless rashes of various appearance (e.g. spotty rash called purpura or purplish skin mottling called livedo reticularis) to painful skin nodules and even ulcers or gangrene (complete loss of blood supply causing damage to peripheral sites including fingers, toes, ears or the tip of the nose). Kidney involvement can result in the presence of blood and protein in urine and/or raised blood pressure (hypertension). The nervous system can also be affected to a variable degree and the child may have seizures, stroke or other neurological changes.
In some severe cases, the condition can worsen very quickly.
Laboratory tests usually show marked signs of inflammation in the blood, with high white blood cell counts (leukocytosis) and a low level of haemoglobin (anaemia).

4.4 How is it diagnosed?

To consider a diagnosis of PAN, other potential causes of persistent fever in childhood should be excluded, such as infections. The diagnosis is then supported by the persistence of systemic and localised manifestations despite antimicrobial treatment, which is usually administered to children with persistent fever. The diagnosis is confirmed by the demonstration of vessel changes through imaging (angiography) or by the presence of vessel wall inflammation in a tissue biopsy.
Angiography is a radiological method where blood vessels that are not seen on ordinary X-rays are visualised by contrast fluid that has been injected directly into the blood stream. This method is known as conventional angiography. Computed tomography can also be used (CT angiography).

4.5 What is the treatment?

Corticosteroids remain the mainstay of treatment for childhood PAN. The mode of administration for these drugs (often directly into veins

when the disease is very active, later in tablets) and the dose and duration of treatment are tailored individually according to a careful assessment of disease extent and severity. When the disease is limited to the skin and musculoskeletal system, other drugs suppressing immune functions may not be necessary. However, severe disease and vital organ involvement requires early addition of other medication, usually cyclophosphamide, in order to achieve disease control (so-called induction therapy). In cases with severe and unresponsive disease, other drugs including biologic agents are sometimes used but their efficacy in PAN has not been formally studied.

Once disease activity settles, it is kept under control with maintenance therapy, usually with azathioprine, methotrexate or mycophenolate mophetil.

Additional treatments used on an individual basis include penicillin (in case of post-streptococcal disease), drugs that dilate blood vessels (vasodilators), blood pressure lowering agents, drugs against blood clot formation (aspirin or anticoagulants), painkillers (non-steroidal anti-inflammatory drugs, NSAIDs).