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Rare Juvenile Primary Systemic Vasculitis

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5. TAKAYASU ARTERITIS

5.1 What is it?

Takayasu arteritis (TA) affects mainly large arteries, predominantly the aorta and its branches and the main lung (pulmonary) artery branches. Sometimes the terms "granulomatous" or "large-cell" vasculitis are used, referring to the main microscopic feature of small nodular lesions formed around a special type of large cell ("giant cell") in the artery wall. In some lay literature, it is also referred to as the ‘pulseless disease’, since in some cases the pulses in the extremities may be absent or unequal.

5.2 How common is it?

Worldwide, TA is considered relatively frequent due to its more common occurrence in the non-white (mainly Asian) population. It is very rare in Europeans. Girls (usually during adolescence) are affected more frequently than boys.

5.3 What are the main symptoms?

Early disease symptoms include fevers, loss of appetite, weight loss, muscle and joint pain, headache and night sweats. Laboratory markers of inflammation are increased. As the artery inflammation progresses, signs of diminished blood supply are apparent. Increased blood pressure (hypertension) is a very frequent initial symptom in childhood disease due to the involvement of abdominal arteries affecting blood supply to the kidneys. Loss of peripheral limb pulses, differences in

blood pressure in different limbs, murmurs heard with the stethoscope over the narrowed arteries and sharp extremity pain (claudication) are common signs. Headaches, various neurological and eye symptoms may be a consequence of the disturbed blood supply to the brain.

5.4 How is it diagnosed?

Ultrasound examination using the Doppler method (for blood flow assessment) is useful as a screening or follow-up method to detect involvement of major arterial trunks close to the heart, although this method often fails to detect involvement of more peripheral arteries. Magnetic resonance (MR) imaging of blood vessel structure and blood flow (MR angiography, MRA) is the most appropriate method for visualising large arteries such as the aorta and its main branches. In order to see smaller blood vessels, X-ray imaging may be used, where blood vessels are visualised by contrast fluid (which is injected directly into the blood stream). This is known as conventional angiography. Computed tomography can be used as well (CT angiography). Nuclear medicine offers an examination called PET (Positron Emission Tomography). A radioisotope is injected into the vein and recorded by a scanner. Accumulation of the radioisotope at actively inflamed sites demonstrates the extent of arterial wall involvement.

5.5 What is the treatment?

Corticosteroids remain the mainstay of the treatment for childhood TA. Their mode of administration and the dose and duration of treatment are tailored individually according to careful assessment of disease extent and severity. Other agents suppressing immune functions are often used early in the disease course in order to minimize the need for corticosteroids. Frequently used drugs include azathioprine, methotrexate or mycophenolate mophetil. In cases of severe disease, cyclophosphamide is used first in order to achieve disease control (so-called induction therapy). In cases with severe, unresponsive disease, other drugs including biologic agents (such as TNF blockers or tocilizumab) are sometimes used but their efficacy in childhood TA has not been formally studied.
Additional treatments used on an individual basis include drugs that dilate blood vessels (vasodilators), blood pressure lowering agents,

drugs against blood clot formation (aspirin or anticoagulants) and painkillers (non-steroidal anti-inflammatory drugs, NSAIDs).