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Rare Juvenile Primary Systemic Vasculitis

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1. WHAT IS VASCULITIS

1.1 What is it?

Vasculitis is inflammation of the blood vessel walls. Vasculitides include a wide group of diseases. The term "primary" means that the blood vessel is a major disease target with no other underlying disease. The classification of vasculitides depends mainly on the size and type of blood vessels involved. There are many forms of vasculitis, ranging from mild to potentially life-threatening. The term "rare" refers to the fact that this group of diseases is very uncommon in childhood.

1.2 How common is it?

Some of the acute primary vasculitides are quite common paediatric diseases (e.g. Henoch-Schönlein purpura and Kawasaki disease), while the others described below are rare and their exact frequency is unknown. Sometimes, parents have never heard the term "vasculitis" before the child is diagnosed. Henoch-Schönlein purpura and Kawasaki disease are covered in their own sections.

1.3 What are the causes of the disease? Is it inherited? Is it infectious? Can it be prevented?

Primary vasculitides do not usually run in the family. In the majority of cases, the patient is the only one affected in a family and it is very unlikely that siblings get the same disease. It is most likely that a combination of different factors plays a part in causing the disease. It is believed that various genes, infections (acting as triggers) and

environmental factors may be important for the disease development. One exception is a very recently described form of vasculitis, called "DADA2", but this is very rare.
These diseases are not infectious and cannot be prevented or cured, but they be controlled - meaning the disease is not active and its signs and symptoms go away. This state is called "remission".

1.4 What happens to the blood vessel in vasculitis?

The blood vessel wall is attacked by the body’s immune system, causing it to swell and resulting in structural disruption. Blood flow is impaired and blood clots may form in the inflamed vessels. Together with the swelling of the vascular walls, this effect may contribute to vessel narrowing or occlusion.
The inflammatory cells from the blood stream gather in the vessel wall, causing more damage to the vessel and to the surrounding tissue as well. This can be seen in tissue biopsy samples.
The vessel wall itself becomes more "leaky", allowing the fluid from within the blood vessels to enter the surrounding tissues and causing swelling. These effects are both responsible for the various types of rashes and skin changes seen in this group of diseases.
Decreased blood supply through narrowed or blocked vessels or, less frequently, vessel wall rupture with bleeding, may damage the tissues. Involvement of the vessels supplying vital organs like the brain, kidneys, lungs or heart can be a very serious condition. Widespread (systemic) vasculitis is usually accompanied by extensive release of inflammatory molecules, causing general symptoms like fever, malaise, as well as abnormal laboratory tests detecting inflammation: erythrocyte sedimentation rate (ESR) and C- reactive protein (CRP). The abnormalities of the vessel shape in the larger arteries can be detected through angiography (a radiological investigation procedure that allows us to see the blood vessels).