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Rare Juvenile Primary Systemic Vasculitis

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7. PRIMARY ANGIITIS OF THE CENTRAL NERVOUS SYSTEM

7.1 What is it?

Primary Angiitis of the Central Nervous System (PACNS) in childhood is an inflammatory brain disease targeting small or medium blood vessels of the brain and/or spinal cord. Its cause is unknown, although in some children, previous exposure to varicella (chickenpox) raises the suspicion that there is an infection-triggered inflammatory process.

7.2 How common is it?

It is a very rare disease.

7.3 What are the main symptoms?

The onset may be very sudden weakness of limbs (stroke), movement disorder, difficult-to-control seizures, severe headaches, and confusion. Sometimes more diffuse neurological or psychiatric symptoms, such as mood and behaviour changes, may be presenting symptoms. Systemic inflammation causing fever and elevated blood inflammatory markers are commonly absent.

7.4 How is it diagnosed?

Blood tests and cerebrospinal fluid analysis ("lumbar puncture") are non-specific and are mainly used to exclude other conditions that might present with neurological symptoms such infections, other non-infectious brain inflammatory diseases or blood clotting disorders. Brain

or spinal cord imaging techniques are the main diagnostic investigations. Magnetic resonance angiography (MRA) and/or conventional angiography (X-rays) are commonly used to detect involvement of medium and large arteries. Repeated investigations are needed in order to assess disease evolution. When artery involvement is not detected in a child with progressive unexplained brain lesions, small vessel involvement should be suspected. This can be eventually confirmed by a brain biopsy.

7.5 What is the treatment?

For post-varicella disease, a short course (about 3 months) of corticosteroids is usually sufficient to halt disease progression. If appropriate, an anti-viral drug is also prescribed (acyclovir). Such a course of corticosteroids may only be needed for the treatment of angiography-positive non-progressing disease. If the disease progresses (i.e. brain lesions are getting worse), then intensive treatment with immunosuppressive drugs is vital to prevent further brain damage. Cyclophosphamide is used most commonly in the initial acute disease and then is replaced by maintenance treatment (e.g. azathioprine, mycophenolate mophetil). Drugs that affect blood clot formation (aspirin or anticoagulants) should be considered.