What is CNS Vasculitis?

CNS is short for “Central Nervous System,” which includes the brain and spinal cord. Vasculitis is what doctors call it when the immune system attacks blood vessels (arteries or veins), causing them to get inflamed and swollen. Putting these together, CNS Vasculitis is an inflammatory disease directed against blood vessels in the brain and/or spinal cord. When these blood vessels get inflamed, it can be hard for blood to get to parts of the brain.

Primary CNS Vasculitis is when inflammation is limited to brain and spinal cord with no other cause. It is also sometimes called Primary CNS angiitis of childhood (cPACNS). Secondary CNS Vasculitis is when inflammation develops due to another disease or infection. It is important to rule out these causes to guide treatment.

CNS Vasculitis occurs in both adults and children, but only recent attention has been given to this condition in children.

This disease can be reversed to some extent with treatment, but continued monitoring is needed because sometimes inflammation can occur again (relapse).

How is CNS Vasculitis diagnosed?

The most helpful tests involve MRI of the brain, angiography (a picture of the blood vessels in the brain), or using a needle to collect fluid from the spine (called a lumbar puncture or spinal tap). However, brain biopsy (a surgery that removes a tiny piece of brain) may be required to diagnose small vessel vasculitis.

To make the diagnosis, doctors use the Calabrese Criteria that includes 1) a new neurological deficit, 2) imaging/biopsy evidence of CNS Vasculitis, and 3) no evidence of other diseases that attack blood vessels.

What are the symptoms for children with CNS Vasculitis?

The most common symptoms are headaches and neurologic problems such as movement problems (such as hemiplegia – trouble moving parts of the body on one side), seizures, decreased alertness and changes in thinking skills.

Other symptoms are more variable and depend on the size and location of blood vessels affected and how long the disease has been happening. When small vessels are affected, symptoms are often a very bad headache, seizures and/or specific neurological deficits. When large vessel affected, symptoms often look more like “classic” effects of a stroke (e.g., loss arm or leg movement, loss of speech).

How is CNS Vasculitis treated?

Children are assessed and treated urgently in hospitals where treatments often include powerful drugs to decrease the immune system’s response. This “immunosuppressive therapy” might involve high dose steroids to get rid of the inflammation as well as specific medications (such as cyclophosphamide).

Children with CNS Vasculitis sometimes need extra help before coming home (e.g., physical, occupational and speech therapy). A neuropsychological assessment is also helpful to children and their families as they return to school. Neuropsychologists are specially trained to understand how brain illnesses, like CNS Vasculitis, affect a child’s thinking, learning, moving, feeling, and behaving. Assessments help understand a child’s strengths, weaknesses and needs. Neuropsychologists work with parents, the school and therapists to help set up support in the school and help families understand how their child’s learning skills were affected by their illness. Many neuropsychologists work with children and families over time, guiding services or providing help themselves as children recover. To find a neuropsychologist, check the link at the bottom of the next page.
What happens to children with CNS Vasculitis over time?

Not a lot is known about the long-term effects of CNS Vasculitis on children's thinking skills or behaviors. Some children make very good recoveries but sometimes they can have difficulties problem solving, or with attention and/or memory. Because it is hard to predict who will recover well, neuropsychological assessments and follow up are important parts of monitoring and support for children and youth with CNS Vasculitis.

Where can I find online resources?

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<td>Vasculitis Foundation</td>
<td><a href="http://www.vasculitisfoundation.org/education/forms/central-nervous-system/">http://www.vasculitisfoundation.org/education/forms/central-nervous-system/</a></td>
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Are there articles that give the “big picture” for professionals?


Version 1.1, updated 1-17-2014 by Tricia Williams, Ph.D., ABPP

Additional Information:

What is board certification in clinical neuropsychology?

Believe it or not, most states allow licensed psychologists to call themselves neuropsychologists without showing they have any special training on how to care for people who have neurological or neurodevelopmental disorders. Clinicians who are board-certified in clinical neuropsychology have proven, through a rigorous evaluation, that they are fully competent. That evaluation is conducted by the largest certification group in psychology, the American Board of Professional Psychology (ABPP; http://www.abpp.org), and its subspecialty board, The American Board of Clinical Neuropsychology. The goal of ABPP is to protect the public by examining and certifying psychologists who demonstrate competence in approved specialty areas.

How do I find a board-certified clinical neuropsychologist?

These are listed by name and by location on the web site of the American Academy of Clinical Neuropsychology (AACN; http://theaacn.org). Once you find a neuropsychologist near you, click on their name for more information, including the kinds of people they work with and how to contact them.

What is pediatric neuropsychology?

[Click here](http://www.div40.org/resources.htm) to download a pamphlet that explains what makes pediatric neuropsychology unique, and what to expect from an evaluation by a pediatric neuropsychologist.

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