



HCSP FACT SHEET

HCV ADVOCATE

• EXTRAHEPATIC MANIFESTATION OF HCV •

Essential Cryoglobulinemic Vasculitis

Written by: Alan Franciscus, Editor-in-Chief

Foreword

Vasculitis refers to a group of conditions that are characterized by inflammation of the walls of blood vessels; these include the veins, arteries and capillaries. Essential mixed cryoglobulinemia is a blood disorder that is caused by abnormal proteins in the blood called cryoglobulins that precipitate or clump together when blood is chilled and then dissolve when re-warmed. These proteins can be deposited in small and medium-sized blood vessels, which can lead to restricted blood flow to joints, muscles, and organs.

There are many classifications of vasculitis such as Takayasu arteritis, Giant cell arteritis, polyarteritis nodosa, Wegener's granulomatosis, Kawasaki disease, Isolated CNS vasculitis, Churg-Strauss arteritis, microscopic polyarteritis/polyangiitis, hypersensitivity vasculitis, Henoch-Schonlein purpura and *essential cryoglobulinemic vasculitis*.

This fact sheet will focus on the type of vasculitis that is associated with HCV-related essential mixed cryoglobulinemia. It should be noted that cryoglobulinemic vasculitis is **uncommon or rare** in people with hepatitis C. In addition, the prevalence of vasculitis varies based on geographic location – the condition is more common in Southern Europe than in Northern Europe or Northern America.

Important note: Everyone with hepatitis C should be evaluated and receive HCV treatment. Current treatment is very expensive, and some insurance companies and Medicaid/Medicare are restricting HCV treatment to people with the most severe HCV disease. One of the conditions that qualify people for HCV treatment is extrahepatic manifestations. Discuss any symptoms with your medical provider and have them recorded in your medical records. If you are not approved for the drugs to treat hepatitis C, you may qualify for free drugs or co-pay assistance through a pharmaceutical patient assistance program. More information is available at the end of this fact sheet.

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**EXECUTIVE DIRECTOR,
EDITOR-IN-CHIEF,
HCSP PUBLICATIONS**
Alan Franciscus

DESIGN
*Leslie Hoex,
Blue Kangaroo Design*

PRODUCTION
C.D. Mazoff, PhD

CONTACT INFORMATION

Hepatitis C Support Project
PO Box 15144
Sacramento, CA 95813
alanfranciscus@hcvadvocate.org

The information in this fact sheet is designed to help you understand and manage HCV and is not intended as medical advice. All persons with HCV should consult a medical practitioner for diagnosis and treatment of HCV.

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Essential Cryoglobulinemic Vasculitis

Vasculitis is sometimes referred to as a ‘hurting disease’ because it is commonly associated with pain. Vasculitis can affect almost every organ in the body.

The more common symptoms and conditions produced by vasculitis include:

- **Skin** – rashes characterized by purplish red spots that are usually found on the legs
- **Joints** – joint aches, and arthritis that includes swelling
- **Lungs** – Shortness of breath, cough, and lung infiltrates
- **Kidneys** – clumps of red blood cells in the kidneys, and loss of protein through the urine
- **Gastrointestinal tract** – abdominal pain, and bloody diarrhea
- **Blood** – anemia and/or elevated white blood cell counts
- **Sinuses and nose** – chronic sinus congestion and infections, hearing problems, and inflammation of nasal tissues
- **Eyes** – damage to the blood vessels of the eye
- **Brain** – headaches, difficulty with coordination, changes in mental status, and strokes (rare)
- **Nerves** – shooting pains in the arms and legs, as well as numbness and weakness

Other symptoms include fever, itching welts, fatigue, weight loss, muscle ache and pain, enlarged lymph nodes, and peripheral neuropathy.

There are no standardized diagnostic tests or criteria for vasculitis. Vasculitis is usually diagnosed by a variety of tests that includes testing for cryoglobulins, antinuclear antibody test (that can detect underlying connective tissue disorders), skin or tissue biopsy, electromyography (detecting electrical signals in muscle cells), arteriography (pictures of blood vessels), and by the clinical manifestations or the symptoms listed above.



Pictures of the hand from the same patient at different times. The image on the left is normal and the one on the right shows the patient in the midst of a flare of cryoglobulinemic vasculitis.

<http://vasculitis.med.jhu.edu/typesof/images/tattoo.gif>

Treatment

Treatment of vasculitis is multi-faceted approach depending on the severity of disease progression.

Since hepatitis C related essential cryoglobulinemic vasculitis is caused by hepatitis C it is generally recommended to treat the underlying cause (HCV). Unfortunately, HCV therapy does not always result in long term resolution of vasculitis and could potentially exacerbate underlying peripheral neuropathy—a condition that is associated with vasculitis.

Talk with your medical provider to find out if treatment for hepatitis C is right for you.

Plasmapheresis

Plasmapheresis is a treatment that removes the blood, separates the blood cells from plasma, and return the blood cells to the body. This therapy is usually combined with other immunosuppressant medications.

Cryofiltration

An addition to plasmapheresis. The blood is removed by plasmapheresis then chilled in a refrigeration unit and the cryoglobulins are filtered and removed. The filtered plasma is re-warmed and combined with the blood cells and returned to the body. This procedure is rarely performed since it may also filter out other components of blood.

Rituximab

Rituximab is an immunosuppressant drug that is FDA approved to treat some forms of non-Hodgkin’s lymphoma. Treatment with rituximab has been found to

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the most effective treatment with long term remission in some patients. There is some evidence that it may be helpful when combined with HCV therapy.

Steroids

In severe cases prednisone and cytotoxic drugs (such as cyclophosphamide and chlorambucil) may be used, however, the use of these drugs in people with hepatitis C is generally discouraged.

Conclusion

In summary, essential cryoglobulinemic vasculitis is a rare condition in people infected with hepatitis C. It is a difficult condition to diagnose because there are no established diagnostic criteria, and current treatment strategies have had limited success. Fortunately, there is much research into the cause and treatment of vasculitis as well as new drugs to treat hepatitis C.

Related publications:

- **Patient Assistance Programs**
http://hcvadvocate.org/hepatitis/factsheets_pdf/Patient_Assistance.pdf
- **Cryoglobulinemia**
http://hcvadvocate.org/hepatitis/factsheets_pdf/Cryo.pdf
- **An Overview of Extrahepatic Manifestations**
http://hcvadvocate.org/hepatitis/factsheets_pdf/Extrahepatic.pdf

For more information

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| <ul style="list-style-type: none"> • Americans with Disabilities Act
www.ada.gov • Centers for Disease Control and Prevention
www.cdc.gov | <ul style="list-style-type: none"> • National Institute of Arthritis and Musculoskeletal and Skin Diseases, Toll free: 877-226-4267
www.niams.nih.gov • National Fibromyalgia Partnership
www.fmpartnership.org |
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