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<https://reachmd.com/programs/cme/whim-syndrome-a-chronic-neutropenic-disorder/24551/>

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WHIM Syndrome: A Chronic Neutropenic Disorder

Announcer:

Welcome to CME on ReachMD. This episode is part of our MinuteCE curriculum.

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Dr. Newburger:

Hello. I'm Dr. Peter Newburger. I'm Professor of Pediatrics and Molecular, Cell and Cancer Biology at UMass Chan Medical School and Attending in Hematology at Boston Children's Hospital. I will now discuss WHIM Syndrome: A Chronic Neutropenic Disorder.

WHIM syndrome is a disorder in which the poorly functioning immune system is present from birth. As you can see from this graph of the time course in which different features of the WHIM syndrome present, neutropenia and infection are the earliest and most consistent findings in WHIM syndrome. Lymphopenia develops a little later, hypogammaglobulinemia subsequently, and warts, which take some time to develop, usually do not appear until the second or third decade of life.

In this different form of the graph, you can see again that neutropenia is the earliest and most consistent finding. Whereas lymphopenia, represented by the absolute lymphocyte count, is not present early in life; once it develops, it's usually consistently found.

So WHIM syndrome should be considered in the differential diagnosis for chronic neutropenia with onset in early infancy or chronic neutropenia with recurrent infections; that is to say, features consistent with likely congenital neutropenia. Definitely test for WHIM in chronic neutropenia with lymphopenia, chronic neutropenia with hypogammaglobulinemia, or any patient with a family history of WHIM, regardless of clinical findings.

Genetic testing is the most direct approach, and early diagnosis is paramount to prevent irreversible tissue damage and long-term sequelae. Early diagnosis also informs genetic counseling and family planning for affected families.

WHIM syndrome is the subject of multiple research programs worldwide, including an active program at the Severe Chronic Neutropenia International Registry. SCNIR.org, of which I'm co-director. We study WHIM syndrome in the context of other chronic neutropenia disorders, and provide advice to physicians, patients, and families who are trying to diagnose a chronic neutropenia disorder or understand one that has already been diagnosed.

Thank you for your attention.

Announcer:

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