



Airway Clearance Indications: Autoimmune Disorders

Wegener's Granulomatosis

Wegener's granulomatosis (sometimes called Wegener granulomatosis) is a disorder in which certain blood vessels become swollen and inflamed and inflammatory tissues called granulomas destroy normal tissue. Inflammation of the blood vessels, a condition called vasculitis, restricts blood flow to affected areas, further compounding tissue damage. Wegener's granulomatosis primarily affects the medium-sized blood vessels in the upper respiratory tract including the nose, sinuses, ears, lungs and kidneys, although other areas may be involved. Wegener's granulomatosis is very rare, with approximately 500 new cases diagnosed in the United States each year. However, the incidence of Wegener's granulomatosis and related vasculitic diseases appears to be on the rise. Whether this is due to an actual increase in the number of affected individuals or an increase in the identification of existing disease is not known. Wegener's granulomatosis affects both genders and generally presents in the 4th or 5th decade of life. The majority of diagnosed patients are Caucasian.

The underlying cause of Wegener's granulomatosis is not definitively known, but it is generally classified as an autoimmune disorder. Autoimmune disorders are characterized by an abnormal response in which the body's immune system mistakenly attacks and destroys its own healthy tissue.

What Happens in Wegener's Granulomatosis?

Over the course of the disease 90% of Wegener's patients will experience lung involvement usually secondary to the development of granulomatous lesions and scar tissue in the parenchyma, bronchi and trachea. Up to 60% of patients will develop permanent airway abnormalities including tracheal strictures, focal or diffuse wall thickening and airway narrowing interfering with effective cough. Without adequate cough clearance, airway secretions may become trapped in narrow rigid airways, contributing to the development of respiratory infections including pneumonia. Recurrent respiratory infections and pneumonia further trigger the hyper-reactive autoimmune response creating a vicious cycle of inflammation, development of granulomas, mucus plugging, infection and more inflammation.

How Airway Clearance Therapy Can Help with Wegener's Granulomatosis

There is currently no known cure for patients with Wegener's granulomatosis. The goal of treatment is to maximize function and maintain or improve quality of life. Keeping the airways clear of excess secretions and thereby reduce the incidence of inflammation and/or infection and is crucial to maintaining respiratory health. Airway clearance therapy using High Frequency Chest Wall Oscillation (HFCWO) has been demonstrated by clinical study to promote excess mucus clearance and improve bronchial drainage. Shear forces are created by HFCWO treatment that mechanically releases adhered secretions from the walls of the pulmonary tract. HFCWO has also been shown to reduce the viscosity of secretions which significantly improves mobilization of



excess mucus. By replicating cough, HFCWO can effectively mobilize pulmonary secretions from smaller airways to larger airways where they can be coughed out, swallowed or suctioned.

Symptoms of Wegener's Granulomatosis

- Sinusitis
- Nose bleeds, pain and sores around the nose
- Night sweats
- Fatigue
- Weight loss
- Blood in urine
- Cough
- Shortness of breath
- Joint pain
- Hemoptysis (blood in sputum)

For More Information on Wegener's Granulomatosis

1. The Vasculitis Foundation: <https://www.vasculitisfoundation.org/wegenersgranulomatosis>
2. The National Institute of Allergy and Infectious Diseases (NIAID) fact sheet on Wegener's granulomatosis: <http://www3.niaid.nih.gov/healthscience/healthtopics/wegener/default.htm>
3. Mayo Clinic fact sheet on Wegener's granulomatosis:
<http://www.mayoclinic.com/health/wegeners-granulomatosis/DS00833>
4. Cleveland Clinic fact sheet on Wegener's granulomatosis:
http://my.clevelandclinic.org/disorders/wegener_granulomatosis/hic_wegeners_granulomatosis.a.spx