

Loeys-Dietz Syndrome: Allergies

Many individuals with Loeys-Dietz syndrome (LDS) report having food allergies, sinus disease, asthma, seasonal/environmental allergies, eczema and/or ear/respiratory infections more than other individuals. Because each person with LDS is different, it is important to talk with your doctor about what care is right for you.

WHAT ARE THE MAIN ALLERGY CONCERNS FOR PEOPLE WITH LDS?

- **Food allergies** - People with LDS are more likely to have food allergies than the general population. The most common food allergies are egg, milk, soy, peanut, and wheat, but a variety of food allergies have been reported. Symptoms range from mild to severe. Some patients have acute, life-threatening reactions that typically begin shortly after they eat the food and includes hives, swelling, trouble breathing, cough, vomiting, and low blood pressure. Other patients have more chronic GI symptoms, such as abdominal pain, abnormal bowel movements, poor weight gain, reflux, vomiting, esophageal spasm or trouble swallowing, and decreased appetite. These chronic symptoms may indicate the presence of eosinophilic gastrointestinal disease, which has been found to be associated with LDS (see the LDSF GI fact sheet for more information).
- **Eczema** - LDS is also associated with eczema, a chronic inflammatory skin condition that is associated with a scaly itchy rash. In some children, certain foods can trigger their eczema, but many other factors, including changes in humidity, viral infections, temperature changes, etc., can also cause eczema to flare
- **Sinus Disease** - The sinuses are cavities or empty spaces in the head. The lining of the sinuses protect the body from dust, pollen, other types of foreign matter, and germs in the air we breathe. Breathing is dependent on clear sinuses. Sinus disease refers to any kind of inflammation or irritation of the sinuses. Usually the main signs of a sinus infection are a stuffy nose and drainage from the nose. People may experience headaches with sinus infections as well. Because of differences in the facial anatomy of people with LDS, patients may not be able to clear germs or other material from their sinuses as effectively as others.

- **Environmental Allergies** - Some people with LDS have increased environmental allergies (tree and grass pollen, dust, etc.). Allergies arise when our body's defense system begins to react to everyday things as foreign invaders. These reactions cause symptoms like allergic rhinitis (runny nose), itchy eyes, sneezing, and congestion. Allergies can also lead to mucus dripping into the throat and windpipe, causing cough and irritation.
- **Asthma** - Asthma (a chronic condition in which the airways become inflamed and constrict) may also be exacerbated by chronic sinus infections or allergies. Asthma can be triggered by many different environmental exposures or illnesses. Symptoms include wheezing, shortness of breath, chest tightness and coughing. Infection from the sinuses may also be carried into the lungs triggering inflammation and asthma; therefore, aggressive treatment of sinus infections in some patients can improve their asthma.
- **Otitis media (ear infections)** - There can also be differences in the anatomy of the ear canals in people with LDS. This can cause excess fluid to build up behind the ear drum which, like the sinuses, can become infected with bacteria. Symptoms include ear pain, fever or irritability.

WHAT IS THE TREATMENT AND MANAGEMENT OF THESE ALLERGY PROBLEMS?

Food allergies - Individuals with suspected food allergies should be seen by an allergist, who typically recommends skin and/or blood testing to diagnose specific allergies. Initial treatment usually involves avoiding foods that are suspected to be a problem. An allergist can be helpful in deciding which foods to avoid based on results from lab testing and an individual's history of reactions. In some cases, foods are then re-introduced gradually. In rare cases, feeding tubes (either through a tube through the nose (Nasogastric) or directly into the stomach (Gastrostomy or G-tube)) and special formulas may be necessary for children with LDS who are not able to get enough calories for growth because of their food allergies and/or GI symptoms.

Many children with food allergies are prescribed an Epi-Pen to use if they develop a life-threatening reaction after being exposed to a food. Epi-Pens release epinephrine, which works by constricting blood vessels to increase blood pressure, relaxing smooth muscles in the lungs to reduce wheezing and improve breathing, and stimulating the heart (increasing heart rate). The rapid constriction of blood vessels may be dangerous to an individual with LDS because of their underlying vascular disease. Thus, it is important to discuss with your cardiologist and/or allergist when to use an EpiPen during an allergic reaction.

Families should work with their child care providers, schools and peer families to ensure that a child with food allergies is kept safe, but is also not excluded from activities involving food due to their allergies. It is important for a child to not feel

isolated from peer activities. There are many resources available that provide recipes for snacks, cakes, etc. that can help provide some normalcy in school and other social settings. The Food Allergy and Anaphylaxis Network (www.foodallergy.org) can be a helpful resource for families dealing with food allergy.

Eczema - It is important to aggressively moisturize the skin of children with eczema, and talk with an allergist and/or dermatologist about topical creams and ointments that may help control the disease.

Sinus Disease & Seasonal/Environmental Allergies - There are different medications that are used to treat nasal congestion. Decongestants can increase blood pressure, thus the use of antihistamines is preferred in an LDS patient. Sinus infections should be treated aggressively with antibiotics. Patients with symptoms should see an Immunologist or Allergist.

Asthma - Bronchodilators (a type of medication that relaxes and dilates muscles in the airways and increases airflow to the lungs) should be avoided by patients with LDS in long-term management of asthma, but may be necessary for a severe exacerbation. A side effect of bronchodilators is increased blood pressure, which is to be avoided in people with LDS because of their vascular issues. Other medications, such as inhaled corticosteroids, can help decrease the frequency of asthma attacks. Individuals with LDS may be prescribed a beta-blocker (common type of high blood pressure medicine), which can worsen a pre-existing asthma predisposition. It is important to discuss this type of medication with your physician.

Otitis media (ear infections) - Ear infections should be treated aggressively with antibiotics. Some individuals may require tympanostomy tube (ear tube) placement for recurrent or persistent ear infections. This is also true for individuals who may be experiencing hearing loss as a result of infections. An ear tube is a small tube inserted through the ear canal into the eardrum to prevent the build up of fluid. Ear tubes are placed by an ear, nose and throat (ENT) doctor as an outpatient surgical procedure and stay in the ear for about a year before being removed or falling out themselves. Complications can include drainage from the ear, cholesteatoma (skin growth behind the eardrum) or perforation of the ear drum.

** It is important to note that the increased risk of sinus and ear infections in LDS is not associated with deficiencies in the immune system, but rather related to differences in craniofacial anatomy.*