Lymphatic Malformation

What is Lymphatic Malformation?
A lymphatic malformation is a mass in the head or neck that results from an abnormal formation of lymphatic vessels. Lymphatic vessels are small canals that lie near blood vessels and help carry tissue fluids from within the body to the lymph nodes and back to the bloodstream. A lymphatic malformation is a congenital (present from birth) defect that occurs during early embryonic development when the lymphatic vessels do not properly form. The vessels may become blocked and enlarged as lymphatic fluid collects in the vessels, forming a mass or a cyst. 80 % of all LM’s are located in the head and neck. However, the malformations can form in any area of the body. Cause is unknown but it is believed that the error occurs during fetal development. MRI (magnetic resonance imaging) or CT (computed tomography) is typically used to make a diagnosis.

It is believed to affect 1 in 2,000 to 4,000.
It affects both sexes and all races.
LM patients live all over the world.

Complications of LM
Since many of LM’s are in the neck area, airway issues are the highest priority. LM’s can cause macroglossia, thickening of the tongue, which can block airway and cause speech/eating complications. Viral infections or sudden trauma can cause rapid but temporary swelling.

Many patients require a tracheostomy to provide a patent airway and a feeding tube for nutrition. However, once LM’s are treated and stable, many of the tracheostomy and feeding tubes can be removed.

Bone overgrowth can develop causing facial deformity. The overgrowth of the jaw line can be cosmetically repaired with surgery when the patient is in late teens and have reached their maximum growth.

Dental and oral hygiene can also be areas if concern.

Treatment
There are currently several types of treatments for LM’s. However, there is no one “best practice” which outlines the optimal approach as each LM patient presents and responds differently to treatment. Surgical removal still remains the best among surgeons surveyed. However, there are other treatments which can be used alone or with surgery. Sclerotherapy is an injection of an agent that shrinks the cyst. Macrocystic lesions respond the best to sclerotherapy. Some agents used are alcohol, doxycycline, or Bleomycin. OK-432 is another sclerosing agent that remains in a non-FDA approved clinical study. Laser therapy is typically used in conjunction to other procedures with good results and may require several treatments. Radio frequency ablation uses high-energy radio frequency sound waves to destroy lesions. Chemotherapy agents have also been used. Many patients can go through several surgeries and treatments during their youth. Therefore, it is strongly encouraged that they seek professional counseling to deal with any emotions and physical challenge.

www.liamsland.org
www.childrensenthouseton.com/lymphatic-malformations/
http://www.rarediseases.org/