# Multidisciplinary Approach to the Management of Lymphatic Malformations of the Head and Neck

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**KEYWORDS**

• Lymphatic malformations • Head • Neck • Multimodal treatment • Sclerotherapy • Surgery

**KEY POINTS**

- Lymphatic Malformations (LMs) may present clinically as low, intermediate, or high grade lesions.
- The typical presentation of a LM is a flesh colored or bluish noncompressible mass.
- They are characterized by episodes of exascerbation and remission.
- Multimodal treatment provides significantly improved outcomes for patients.

Lymphatic malformations (LMs) occur in 2.8 to 5:100,000 live births.[1,2] Most involve the head and neck and they are equally common in males and females.[3–5] They are developmental anomalies of unknown cause, although recent evidence suggests that an upregulation of the mammalian target of rapamycin (mTOR) pathway may be a causal factor leading to the overproduction of abnormal lymph vessels.[6] These vessels are likely dilated lymphatic sacs sequestered from the lymphatic and venous systems.[7,8] This overproduction results in the accumulation of lymph in dilated cystic spaces, which in turn results in the clinical features of a LM.

## CLINICAL PRESENTATION

In about 50% of cases, the condition is apparent at birth and, in 90% of cases, symptoms are present by 2 years of age.[9] Routine prenatal ultrasonography has led to a

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higher incidence of prenatal diagnosis.[10,11] However, not all prenatally diagnosed LMs are present at birth. Some are associated with significant cardiac, renal, and skeletal deformities and do not proceed to parturition, aborting either spontaneously or electively. Longstreet and colleagues[12] recently proposed an anatomic classification of fetal nuchal lymphatic anomalies. They recognized 3 groups of fetal LMs.

### *Nuchal Thickening*

Nuchal thickening is a hypoechoic region exceeding the 95th percentile between skin and soft tissues bordered by the fetal spine and occiput. About 30% of these lesions are associated with chromosomal abnormalities, such as trisomy 21, trisomy 18, and Turner syndrome. A large percentage (80%) of these resolve by birth.

### *Dorsal Lymphatic Malformations*

Dorsal LMs are septated, fluid-filled, multilocular cavities extending along the entire dorsal length of the fetus. These cavities are associated with a high percentage of other abnormalities, such as cardiac, renal, and skeletal. Many of these abort spontaneously or electively. Of those that did survive to parturition, 50% had resolved spontaneously.

### *Ventral Lymphatic Malformations*

Ventral LMs were defined as those occurring on the anterior and/or lateral cervical region of the fetus. None of these were associated with other congenital anomalies and all were present at birth. Ventral LMs are at risk for airway obstruction and may necessitate an ex utero intrapartum treatment (EXIT) procedure.

The typical presentation of an LM is a noncompressible flesh-colored or bluish mass involving the head and/or neck (**Fig. 1**). The location and extent of involvement are

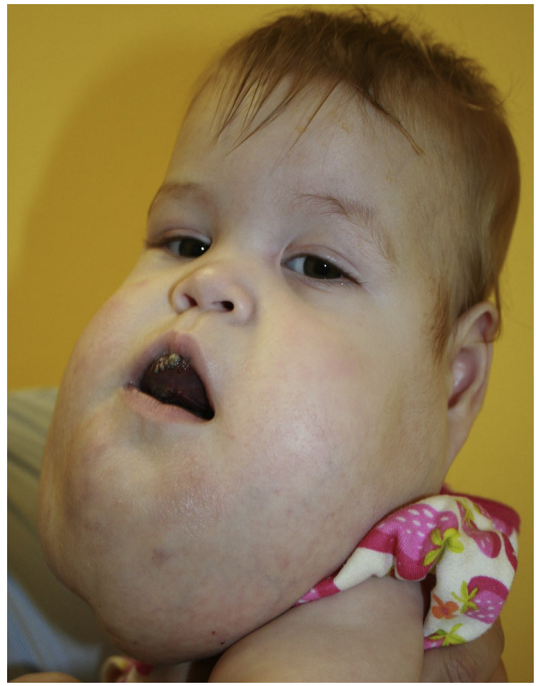

**Fig. 1.** A child with a large bilateral cervicofacial LM.