

Table 1: Complex congenital lymphatic anomalies

Gorham-Stout Disease (GSD):	A rare complex vascular anomaly syndrome characterized by the infiltration of lymphovascular channels in bone and the surrounding soft tissues.[42] Also referred to as Gorham Stout syndrome, massive osteolysis, or vanishing bone disease, the remarkable feature of this condition is progressive osteolysis and cortical bone destruction without adequate signs of repair. The clinical features can vary greatly depending on the site of disease involvement and is often described as focal vs regional (multiple contiguous bones) involvement.
Generalized Lymphatic Anomaly (GLA)	(Previously referred to as lymphangiomatosis, cystic angiomatosis, generalized cystic lymphangiomatosis) Multifocal dilated, cystic lymphatic malformation in association with multifocal bone lytic skip lesions. Bone lesions preserve the cortex, are generally stable and more often associated with the appendicular skeleton. Skin, superficial soft tissue, spleen, intestine, liver, lung can also be involved.
Central Conducting Lymphatic Anomaly (CCLA)	(Previously referred to as lymphangiectasia) Enlarged lymphatic channels with dysmotility of flow and distal obstruction that inhibits adequate clearance of lymph. Obstructed and static flow causes retrograde reflux of lymphatic fluid. Often involves dysfunction at the level of the thoracic duct or cisterna chyli.
Kaposiform Lymphangiomatosis (KLA)	A rare complex lymphatic anomaly with predominance of intrathoracic and extrathoracic effusions. Dilated lymphatic channels contribute to dysmotility, stasis, and reflux similar to CCLA. Lymphatic channels can be blood-filled. Effusions more commonly demonstrate frank hemorrhage. Histologic analysis demonstrates clusters of spindle endothelial cells without nuclear atypia; a pattern seen in Kaposiform hemangioendothelioma. The combination of both CCLA features and Kaposiform features results in an aggressive, proliferative complex malformation that manifests as refractory hemorrhagic effusions. Extrathoracic disease can manifest as pericardial effusions, cystic lesions of the spleen, and bone lesions. Bone involvement tends to present as multiple non-contiguous lytic lesions that spare the cortex most often involving the thoracic spine.
Waldmann's disease	The presence of dilated lymphatic vessels or Primary Intestinal lymphangiectasia resulting in leakage of lymph into the peritoneal cavity. Clinically presents with peripheral, symmetric pitting edema and moderates serious effusions.