

Hereditary lymphedema affects females more often than males. The estimated prevalence of these disorders is 1 in 6,000 individuals within the general population. Hereditary lymphedema type II (Meige syndrome) is the most common form accounting for approximately 80 percent of cases. The prevalence of hereditary lymphedema type I (Milroy disease) is unknown. Approximately 200 cases have been reported in the medical literature. The diagnosis of hereditary lymphedema may be confirmed by a thorough clinical evaluation and a variety of specialized imaging tests including lymphoscintigraphy, ultrasound, and magnetic resonance imaging (MRI). During lymphoscintigraphy, a radioactively labeled colloid substance is injected intradermally into either the hands or feet. The time required for the tracer to be transported from the point of injection to the regional lymph nodes is recorded. In congenital lymphedema, the tracer may move sluggishly or not move from the site of injection. During an ultrasound, reflected sound waves create an image of the developing fetus. An ultrasound is used to rule out other conditions. A Doppler ultrasound can evaluate venous conditions such as varicose veins and venous blood clots. An MRI uses a magnetic field and radio waves to produce cross-sectional images of particular organs and bodily tissues. An MRI is used to detect findings characteristic of hereditary lymphedema including swelling (edema), a mass surrounded by a sac containing lymph fluid (lymphocele), and the formation of fibrous tissue (fibrosis).