Parry-Romberg syndrome is a rare, acquired disorder characterized by slowly progressive shrinkage (atrophy) of the skin and soft tissues of half of the face (hemifacial atrophy). In rare cases, both sides of the face are affected. In some people, atrophy may also affect the limbs usually on the same side of the body as the facial atrophy. The severity and specific symptoms of Parry-Romberg syndrome are highly variable from one person to another. Additional symptoms can potentially develop in some people including neurological abnormalities or abnormalities affecting the eyes or teeth. Parry-Romberg syndrome usually becomes apparent during the first decade of life or early during the second decade but does occur in adulthood. The majority of individuals with Parry-Romberg syndrome experience symptoms before the age of 20 years. The exact cause of Parry-Romberg syndrome is unknown and appears to occur randomly for unknown reasons (sporadically). Parry-Romberg syndrome is a rare disorder. The true incidence is unknown. Because the disorder often goes undiagnosed or misdiagnosed determining the true frequency of Parry-Romberg syndrome in the general population is difficult. Physicians studying the disorder have estimated that Parry-Romberg may affect as many as 1 in 250,000 people in the general population. Parry-Romberg syndrome appears to affect women slightly more often than men but proper studies of the population are lacking. Parry-Romberg syndrome typically becomes apparent during the first or early during the second decade of life, with the majority of affected individuals experiencing symptoms before the age of 20 years. However, the disorder has been described in infants and individuals more than 50 years of age. Parry-Romberg syndrome was originally described in the medical literature in 1825 (C.H. Parry) and 1846 (E. Henoch and H.M. Romberg). There are anecdotal reports of Parry-Romberg syndrome worsening in some pregnant women, either during pregnancy or shortly after childbirth. A diagnosis of Parry-Romberg syndrome is made based upon identification of characteristic symptoms, a detailed patient history, a thorough clinical evaluation and a variety of specialized tests. The specific tests that are used depend on which symptoms are present and which symptoms occur first. For example, magnetic resonance imaging (MRI) may be used in individuals with neurological symptoms. An MRI uses a magnetic field and radio waves to produce cross-sectional images of particular organs and bodily tissues. Surgical removal and microscopic examination (biopsy) of affected skin tissue may be used in individuals with linear scleroderma en coup sabre.