Gottron syndrome (GS) is an extremely rare inherited disorder characterized by the appearance of premature aging (progeria), especially in the form of unusually fragile, thin skin on the hands and feet (distal extremities). GS is described as a mild, nonprogressive, congenital form of skin atrophy due to the loss of the fatty tissue directly under the skin (subcutaneous atrophy). Other findings may include abnormally small hands and feet with unusually prominent veins on the chest; small stature; and/or abnormally small jaw (micrognathia). From infancy on, children with Gottron syndrome appear older than their actual age. The skin is unusually thin, taut, and parchment-like on the hands and feet (distal extremities). The hands and feet remain abnormally small into adulthood. The veins on the chest are very visible and prominent due to diminished amounts of fat under the skin (subcutaneous fat). It is believed that Gottron syndrome may affect more females than males. Approximately forty cases have been reported in the medical literature. Treatment for Gottron syndrome is symptomatic and supportive. Genetic counseling may be of benefit for patients and their families.