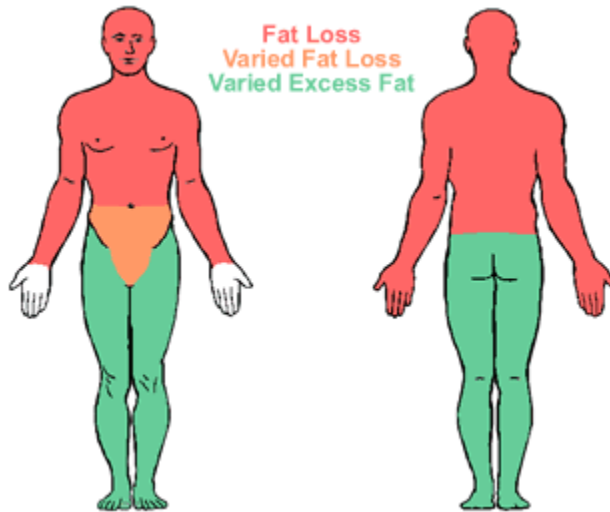


Acquired: Partial Lipodystrophy (Barraquer-Simons Syndrome)



The onset of acquired partial lipodystrophy usually occurs around 8-10 years of age and is usually preceded by an episode of acute viral infection. It is rare -approximately 250 patients of various ethnic origins have been described. It is characterized by the loss of fat from the face, extending to involve the neck, shoulders, arms, forearms, thoracic region and upper abdomen occasionally extending to the groin or thighs. Usually legs and hips are spared. After puberty, Women may accumulate disproportionately large amount of fat in the hips and legs. Fat loss usually occurs over 18 months but can occur periodically during several years.

Patients with acquired partial lipodystrophy usually do not have metabolic abnormalities associated with insulin resistance such as elevated lipid levels, acanthosis nigricans (dark velvety pigmentation of the skin), hirsutism (increased body hair) or menstrual abnormalities. Females are affected three times more often than males. Approximately one-fifth of these patients develop a kidney problem called Membranoproliferative Glomerulonephritis. It usually occurs more than 10 years after the onset of lipodystrophy. Patients have low levels of complement C3 (a factor that plays a role in immune response) in their blood (hypocomplementemia). They also have an antibody in their blood called the C3 nephritic factor. Acquired partial lipodystrophy is also associated with autoimmune disorders like systemic lupus erythematosus (SLE), dermatomyositis, hypothyroidism, pernicious anemia, celiac disease, dermatitis herpetiformis, rheumatoid arthritis, temporal arteritis and leukocytoclastic vasculitis. Some patients also develop drusen or macular degeneration later in life.

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