Pediatric Lipodystrophy

WHAT PARENTS AND CAREGIVERS SHOULD KNOW

Lipodystrophies are a group of rare diseases characterized by the unexpected loss of body fat from various regions of the body.

Lipodystrophy can be inherited (genetic) or caused by other illnesses or drugs (acquired).

Genetic lipodystrophies: These are caused by alterations in genes and can manifest soon after birth or later in life depending on the subtype and specific genetic alterations. Congenital generalized lipodystrophy and familial partial lipodystrophy are the two main subtypes of inherited lipodystrophy; the other subtypes are extremely rare.

Acquired lipodystrophies: These usually occur during childhood, adolescence or adulthood and are associated with autoimmunity, panniculitis or can be due to unknown reasons (this is called "idiopathic"). Common types of acquired lipodystrophy include acquired generalized lipodystrophy (Lawrence syndrome) and acquired partial lipodystrophy (Barraquer-Simons syndrome).

Diagnosis of lipodystrophy is based on a detailed medical history and a thorough clinical exam to evaluate body fat loss.

In patients with acquired lipodystrophy, there are special lab tests that can be ordered, including serum complement levels and autoantibodies. These tests help can help with the diagnosis of four major types of lipodystrophies:

- Congenital generalized lipodystrophy
- Acquired generalized lipodystrophy
- Familial partial lipodystrophy
- Acquired partial lipodystrophy

Patients with suspected lipodystrophy typically need specialized testing and they should be referred to a lipid specialist. Once the diagnosis of lipodystrophy is made, clinicians should investigate whether the lipodystrophy is generalized, partial, or localized. In generalized forms, total or near-total loss of fat underneath the skin can be observed over the entire body. In partial forms, fat loss affects large areas, particularly the arms, and legs, but fatty tissue may build up in areas such as the abdomen, face, and neck. Localized forms of lipodystrophy are limited to small body areas.

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What Options Are There for Treatment of Lipodystrophy in Children and Adolescents?

Most patients need a balanced macronutrient diet with 50–60% of calories from carbohydrates, 20–30% from fat, and approximately 20% from protein; however, patients with elevated triglycerides will need an extremely low-fat diet with < 15% of calories from dietary fat.

Single Day Diet Example:

BREAKFAST

- Scrambled egg *(1 whole egg + 2 egg whites)*
- 1 slice whole wheat toast
- 8 oz glass nonfat milk

LUNCH

- Sandwich *(whole wheat bread, uncured deli turkey meat, reduced fat or fat-free cheese, lettuce/tomato)*
- 1 small apple
- Carrot sticks with nonfat Ranch dressing

SNACK

- 1 cup fruit

DINNER

- Spaghetti *(ground turkey breast, marinara sauce, and chickpea pasta or whole wheat pasta)*
- Side salad

Medication Options

Metreleptin therapy with a macronutrient diet described above should be considered for generalized lipodystrophy. Metformin and insulin therapy are usually needed for patients with diabetes. Triglyceride-lowering therapies like fibrates and fish oil are needed for patients with hypertriglyceridemia.

Metreleptin is an injectable medication that mimics the body's natural leptin processes that signal to the brain to create more fat tissue. It is used by patients with lipodystrophy who may not be producing fat tissue appropriately. The specific dose of the medication is dependent on the weight of the child and not on the age and should be defined by the Healthcare Team.

For all of these medications, it is important to talk to your Healthcare Team before starting any new medications or supplements to ensure they are safe to take with the prescribed medications.

To find a lipid specialist in your area, use the "find a clinician" tool on ***learnyourlipids.com***.

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