

Therapy & Management

The efficacy of various drugs, such as sulfonylureas, biguanides and thiazolidinediones has not systematically been investigated, although they have been used to control blood glucose levels in patients with lipodystrophies. Insulin is commonly given to control hyperglycemia. Extremely high doses of insulin are often required to control levels of blood glucose. In such cases, concentrated preparation of insulin containing 500 units/ml can be helpful.

What are the treatments available to improve insulin sensitivity?

Treatment is based on improving glucose control and preventing complications, especially cardiovascular disease. Diet is a mainstay of treatment along with exercise and weight loss. A low calorie diet reduces insulin resistance in days even before much weight loss takes place. Ten to twenty pounds (5-10 kg) substantially helps glycemic control and a loss of 16% of body weight improves glucose metabolism by a 100%.

Medications such as metformin (Glucophage®), troglitazone (Rezulin®), and acarbose (Precose®), alone or in combination, have been used to improve insulin sensitivity mainly by reducing plasma glucose by different mechanisms. All of these treatments can be employed to prevent the development of Type 2 diabetes.

Therapy for High Blood Cholesterol and Triglycerides

Patients with extreme high blood triglyceride levels should be treated with fibrates (gemfibrozil, clofibrate, bezafibrate, fenofibrate) and omega-3 polyunsaturated fatty acids (omega-3 fatty acids) from fish oils. Occasionally patients may require a combination of drugs including fibrates and statins (Lovastatin, simvastatin, atorvastatin), however, risk of muscle toxicity may limit their use. Niacin should be avoided since it may exacerbate hyperglycemia. Oral contraceptives and postmenopausal hormone replacement therapy should be avoided in females as the estrogens in these preparations may increase the blood level of triglycerides.

Management of Lipodystrophy

Cosmetic disfigurement due to fat loss is a major cause of concern for patients. Besides fat loss and excessive muscular appearance and hirsutism (increased body hair), dark colour pigmentation and thickening of the skin in the neck, armpits, groin and the trunk due to acanthosis nigricans may pose cosmetic problems. Diabetes is usually severe, and may produce short and long-term complications, such as kidney disease, retinal (eye) disease, heart disease and neuropathy (nerve damage). Hypertriglyceridemia (high levels of blood fats) may cause acute pancreatitis (acute pancreatic injury with severe abdominal pain) and can potentially further contribute to early-onset coronary heart disease.

Cosmetic Management

Several surgical methods have been used including facial reconstruction with transplanted tissue flaps, transposition of facial muscles, facial implants and collagen injections in the cheeks, however the results are variable. Liposuction and lipectomy for removal of excessive facial or neck fat have been carried out in several patients with familial partial lipodystrophies. The decision to undergo any such modes of treatment must be undertaken with the advice of the physician. Areas of excess fat accumulation may reduce with weight loss.