Corneal manifestations of systemic disease related to metabolic disorders are frequent and may be the first indication of disease. Conditions are typically rare due to recessive inheritance, bilateral, and result in loss of corneal clarity due to deposition of various substances in the different layers of the cornea.

**LIPID METABOLISM DISORDERS**

**Hyperlipoproteinemias**

Ocular manifestations of hyperlipoproteinemias result from extracellular deposits of cholesterol, cholesterol esters, phospholipids, and triglycerides. They are common and often manifest as xanthelasma and arcus. Though arcus is common in older patients, presentation in a patient 35 to 40 years old or asymmetric presentation warrants investigation to rule out a lipid abnormality. Arcus may be associated with hypercholesterolemia, xanthelasma, alcohol, elevated blood pressure, cigarette smoking, diabetes, increasing age, and coronary heart disease. It remains unclear whether corneal arcus is an independent risk factor for coronary heart disease.  

Schnyder crystalline corneal dystrophy (crystalline stromal dystrophy) is thought to be a localized defect of lipid metabolism, which is covered in detail in Chapter 6.

**Hypolipoproteinemias**

Lecithin-cholesterol acyltransferase (LCAT) facilitates removal of cholesterol from the liver. Its deficiency leads to accumulation of unesterified cholesterol in the tissues, causing atherosclerosis, renal insufficiency, arcus, and nebular corneal clouding due to focal lipid deposition. Five disorders are related to abnormally low serum lipoprotein levels: LCAT deficiency, Tangier disease, fish eye disease, familial hypobetalipoproteinemia, and Bassen-Kornzweig syndrome.