of the reticuloendothelial system. It is inherited as an autosomal recessive and to date 10 patients have been described. In all patients the tonsils or tonsillar remnants show a pathognomonic yellow-orange colouration and in four patients, including the one we have studied, peripheral neuropathy has been present. Death from presumed myocardial infarction has been reported in a 48-year-old man. Serum cholesterol and phospholipid levels are decreased but triglyceride is increased and the serum is turbid even in the fasting state. In our patient we have demonstrated that the proportion of triglyceride is increased in all the lipoprotein fractions including the small amount of alpha-lipoprotein. LEVY *et al.* [1] have shown that in this condition serum triglyceride can be influenced by dietary carbohydrate and in our patient long-term carbohydrate restriction resulted in some reduction in serum triglyceride levels (from 400 mg/100 ml to about 200 mg/100 ml). In an attempt to lower triglyceride levels further chlorophenoxyisobutyrate was given; triglyceride levels were not lowered but there has been a significant increase in total cholesterol concentration (from 50 mg/100 ml to 111 mg/100 ml) with a proportionate increase in beta-lipoprotein cholesterol (from 10 mg/100 ml to 45 mg/100 ml). It is not known whether this rise is due to mobilisation of cholesterol from tissues; there has been no marked change in the clinical features over a 2-year period.

1. LEVY, R.I.; LEES, R.S. and FREDRICKSON, D.S.: J. clin. Invest. *45:* 531 (1966).

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