Very long-chain acyl-CoA dehydrogenase deficiency (VLCADD) is a rare genetic disorder of fatty acid metabolism that is transmitted as an autosomal recessive trait. It occurs when an enzyme needed to break down certain very long-chain fatty acids is missing or not working properly. VLCADD is one of the metabolic diseases known as fatty acid oxidation (FOD) diseases. In the past, the name long-chain acyl-CoA dehydrogenase deficiency (LCADD) was applied to one such disease, but today it is clear that all cases once thought to be LCADD are actually VLCADD. VLCADD was originally described in 1992 and is now recognized as having an incidence of 1:40,000 babies. The introduction of heel-stick tandem mass spectrometry for the early diagnosis of VLCAD in newborns has markedly increased the number of infants in which the disorder is detected.