The methylmalonic acidemias are organic acidemias caused by an enzymatic defect in the metabolism of four amino acids (methionine, threonine, isoleucine and valine). This results in an abnormally high level of acid in the blood (academia) and body tissues. In the acute form, drowsiness, coma, and seizures may occur. Mental retardation is a long-term consequence. The disorder may be caused by a deficiency of one or more of the enzymes methylmalonyl CoA mutase, methylmalonyl racemase, or adenosylcobalamin synthetic enzymes. Excretion of methylmalonate, a product of amino acid metabolism, in the urine is abnormally high and therefore is a marker of the disorder. All known organic acidemias are inherited as autosomal recessive traits. The Methylmalonic Acidemias occur at a rate of 1 in 50,000 to 1 in 100,000 live births.