Alopecia and periorificial dermatitis in biotin-responsive multiple carboxylase deficiency

Three siblings with infantile-onset biotin-responsive multiple carboxylase deficiency are described. Recognition of the characteristic dermatologic manifestations, alopecia and periorificial dermatitis, should result in early diagnosis and institution of potentially lifesaving therapy with biotin. Other metabolic disorders may present a similar clinical picture. Immunologic dysfunction and/or aberration in lipid or branched chain amino acid metabolism may be the common pathophysiologic link in some or all of these disorders.