

Distal myopathy (or distal muscular dystrophy) is a general term for a group of rare progressive genetic disorders characterized by wasting (atrophy) and weakness of the voluntary distal muscles. The distal muscles are those farther from the center of the body and include the muscles of the lower arms and legs and the hands and feet. Conversely, the proximal muscles are the muscles closest to the center of the body such as the muscles of the shoulder, pelvis, and upper arms and legs. Although age of onset can occur anytime from infancy to adulthood, most forms develop later in life and are slowly progressive. Inheritance is autosomal dominant or recessive. Since no distal myopathy has been linked to the X-chromosome, distal myopathies affect males and females in equal numbers. The exact incidence of the distal myopathies is unknown. Some forms have been identified with greater frequency in certain populations. Udd distal myopathy occurs with greater frequency in Finland where the prevalence is estimated to be 7 in 100,000 individuals. Welander distal myopathy occurs with greater frequency in Sweden where the prevalence is estimated to be 1 in 1,000 individuals. Approximately 220 cases of IBM2 have been identified in the medical literature. The muscular dystrophies as a whole are estimated to affect 250,000 individuals in the United States.