MERRF (Myoclonus Epilepsy with Ragged-Red Fibers) syndrome is an extremely rare disorder that begins in childhood and affects the nervous system and skeletal muscle as well as other body systems. The distinguishing feature in MERRF is myoclonus, consisting of sudden, brief, jerking spasms that can affect the arms and legs or the entire body. In addition, individuals with MERRF syndrome may have muscle weakness (myopathy), an impaired ability to coordinate movements (ataxia), seizures, and a slow deterioration of intellectual function (dementia). Short stature, degeneration of the optic nerve (optic atrophy), hearing loss, cardiomyopathy and abnormal sensation from nerve damage (peripheral neuropathy) are also common symptoms. Abnormal muscle cells are present and appear as ragged red fibers (RRF) when stained with the modified Gomori trichrome and viewed microscopically. MERRF is caused by mutations in mitochondrial DNA (mtDNA). MERRF syndrome is a rare disorder that affects males and females in equal numbers. Some researchers believe that mitochondrial myopathies may go unrecognized and underdiagnosed in the general population, making it difficult to determine the true frequency of disorders like MERRF syndrome.