Histiocytoid Cardiomyopathy

National Cancer Institute

Source

National Cancer Institute. Histiocytoid Cardiomyopathy. NCI Thesaurus. Code C45745.

A hamartomatous lesion of the sinoatrial node, atrioventricular node, and Purkinje fibers of the cardiac conducting system. It occurs predominantly in the first two years of life. Most patients present with arrhythmias and electrical disturbances. It is characterized by the presence of multifocal, poorly defined islands of large polygonal cells with a granular eosinophilic cytoplasm, a small round to oval-shaped nucleus, and occasional nucleoli. The cytoplasmic appearance is due to extensive accumulation of mitochondria. If left untreated, this condition is usually fatal. However, the outcome has improved over the past two decades due to developments in surgical intervention, electrophysiological mapping, and ablation of the arrhythmogenic foci, with a survival rate of approximately 80%. (WHO 2015)