The Clinical Significance of Familial Heart Block in the Perinatal Period

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Introduction: Familial abnormalities of heart pulse generation and heart conduction system, both symptomatic and asymptomatic, are sometimes overlooked in daily medical practice. Familial (and congenital) Heart Block (FHB) may be associated with other diseases such as: Hypertrophic and Dilated Cardiomyopathies, Familial Dysautonomia, Emery-Dreifuss dystrophy and Charcot-Marie-tooth disease. Previous reports have documented the dominant (or x-linked) transmission with variable expression of FHB, and its association with collagen disease in the mother. It may appear at any stage of life.
Purpose: To present the clinical and genetic findings, and to emphasize the clinical significance of familial heart block, especially when symptoms exist, even if they are sporadic and minor and are registered during the perinatal period.
Methods used: The members of three families (9 individuals in 3 generations) with symptomatic heart block are evaluated: eight of them required permanent cardiac pacemaker therapy. Last year, a female baby was born to one of our patients. Pre-Natal Fetal Arrhythmia had been noticed and 2nd degree Atrio-Ventricular (AV) block (Mobitz I & II) was observed on 24-hours Holter monitoring.
Conclusions: Careful follow-up is suggested for all family members in whom FHB is present, especially when symptoms exist, and even in the perinatal period.