ДВУХКАМЕРНАЯ КАРДИОСТИМУЛЯЦИЯ ПРИ СЕМЕЙНОЙ ПОЛНОЙ АТРИОВЕНТРИКУЛЯРНОЙ БЛОКАДЕ, РАЗВИВШЕЙСЯ ВО ВЗРОСЛОМ ВОЗРАСТЕ

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Ключевые слова: семейная полная атриовентрикулярная блокада, двухкамерный кардиостимулятор, генетическое сцепление.

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The familial complete atrioventricular block, is an inherited disease, may appear in adult period [1–3]. A 39-year-old-woman had symptomatic (two episodes of near syncope during exertion) complete atrioventricular block (QRS wave ≤0,12 second on the surface electrocardiography) (Fig. 1) and was successfully treated with permanent dual chamber (DDDR) pacemakers. Her father, a 66-year-old-, had also complete atrioventricular block who was successfully treated with DDDR pacemaker in the another hospital about 7 years ago. Her other family members had normal sinus rhythm. She and her father had not any disease such as hypertension, diabetes mellitus, rhumatologic disease and valvular heart disease. Their laboratory tests such as complete blood count, urea, creatinine, potassium, magnesium, thyroid function tests, antinuclear antibody, romatoid factor and transthoracic echocardiogram were normal.

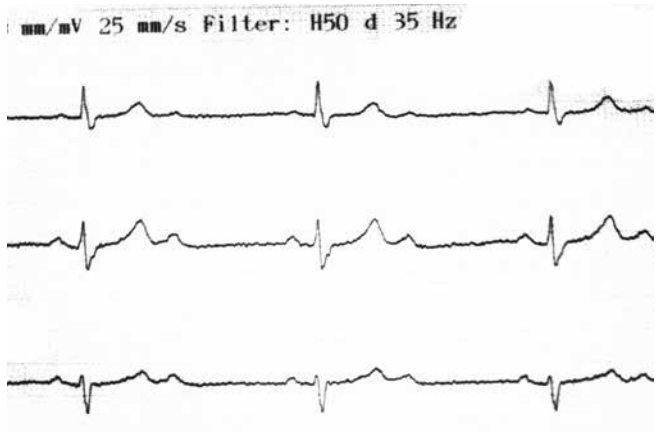

Figure 1. A 39-year-old-woman had complete atrioventricular block (QRS wave ≤0,12 second)

Familial heart block occurs in two forms, type I and type II (narrow QRS) [1]. Type I is an autosomal dominant bundle-branch hereditary disease which may progress to complete atrioventricular block [1, 2]. This form have a progressive feature; the risk to life appears to be greatest at soon after birth, during puberty and the early twenties [1]. The surface electrocardiography includes right bundle-branch block, left anterior hemiblock or left posterior hemiblock, or complete atrioventricular block with broad QRS complexes [1, 2]. Although type I form is bundle-branch disease, type II form is atrioventricular nodal disease with an atrioventricular block and an idionodal escape rhythm [1, 2]. Fernandez et al [3] also suggested that type II form is a an atrioventricular nodal disorder with clinical onset between the fourth and sixth decade, as in our cases. Treatment is implantation of a pacemaker because of symptomatic episodes, as in our cases. The pathophysiology of these diseases is unclear; but, linkage analysis and positional cloning offer a means of identification of disease-causing genes including chromosome 19q13.2–13.3 encoding myotonin protein kinase that is implicated as a cause of myotonic dystrophy [2]. Finally, because of these findings, it was thought a familial complete atrioventricular block of adult onset in these patients. Also, the clinicians' awareness of familial heart block might diagnose asymptomatic member of these families.

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Dual chamber pacemaker treatment in familial complete atrioventricular block of adult onset

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