

The world's largest family with familial atrio-ventricular nodal reentry tachycardia

Rodziny napadowy częstoskurcz węzłowy — obserwacja wielopokoleniowej rodziny

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Typical atrioventricular nodal reentrant tachycardia (AVNRT) is one of the most common forms of paroxysmal supraventricular tachycardia (SVT) referred for radiofrequency catheter ablation (RFCA). Dual atrioventricular nodal pathways, which are the substrate for AVNRT, are rarely occurring congenital abnormalities. We report a three-generation family from the southeastern region of Poland (Podkarpackie voivodship) with 6 women having normal hearts and presenting with a history of paroxysmal tachycardia with onset of symptoms in adulthood (Fig. 1). Recordings of clinical SVT, dual AVN electrophysiology, induction of typical AVNRT (Fig. 2), and results of RFCA are available for 5 of the affected women: the proband (57 years old), her 2 sisters (aged 51 and 47 years), proband's daughter (21 years old) and her cousin (26 years old). They underwent RFCA with modification of a slow pathway. The proband's mother (aged 80 years) reported a 30-year history of paroxysmal, regular, and rapid palpitations, usually lasting for several minutes and terminated by vagal manoeuvres. At the time of the evaluation she suffered from advanced lung cancer, and refused regular check-ups and electrophysiological study, so an electrocardiography with documented paroxysmal tachyarrhythmia was not available. Two asymptomatic young women from this family (the second proband's daughter (30 years old) and niece (26 years old) refused an evaluation of dual AVN electrophysiology and inducibility of AVNRT. Interestingly, having never been trained by the doctor, the proband's mother herself found a way to terminate the palpitations and explained to all of her daughters how to perform the Valsalva manoeuvre, so the youngest women in this family were taught by their mothers how to do it, long before the first episode of palpitations occurred. Of note, the symptoms of paroxysmal arrhythmia were not present in males. During 8-year follow-up a second RFCA procedure was required in the proband. The other members of this family have not reported palpitations or arrhythmias requiring invasive evaluation and treatment. There are only limited reports on the familial form of AVNRT (FAVNRT) with up to 4 first-degree relatives within two generations, with no genetic substrate identified so far. To our knowledge, this is the largest family with FAVNRT reported until now, and the first in which inheritance through the maternal line is reported, due to probable autosomal dominant, X-linked dominant, or mitochondrial mode of transmission. Up to now we have observed more than 20 new families with FAVNRT, and further studies are planned to confirm the presumed higher incidence of familial AVNRT in Podkarpackie voivodeship and to evaluate its genetic background. Finally, the history of this family shows the significance of a careful and detailed collection of medical history, and points towards the importance of family screening in AVNRT patients.

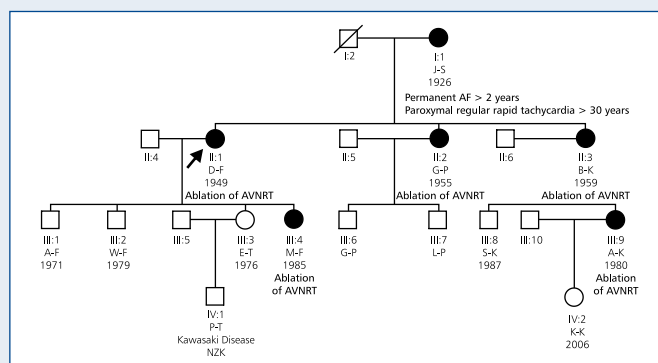


Figure 1. Pedigree tree; arrow indicates a proband, squares — men, black circles — affected women



Figure 2. Typical induction of slow-fast atrioventricular nodal reentrant tachycardia after pacing from proximal coronary sinus in the proband

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