Turkish perspective of Jervell and Lange-Nielsen syndrome

Sir,

The authors of "Jervell and Lange-Nielsen Syndrome (JLNS) masquerading as intractable epilepsy" claimed that JLNS is more prevalent in Norway and Turkey.[1] The prevalence of JLNS in Norwegian families are reported by Tranebjaerg *et al*,[2] the authors cited this study, but there is no citation about the prevalence in Turkey.

As far as we know there are just two data about the prevalence of clinically diagnosed JLNS in Turkey.[3,4] There are also some case reports and studies in the literature in Turkey, these cases are also clinically diagnosed as JLNS and not genetically confirmed.[5-9] There is one family, reported by Tyson *et al*,[10] resident of UK but Turkish, where JLNS has been genetically confirmed. There is another report by us about genetically confirmed Turkish JLNS family.[11] These two families have the same pathologic variant. Because of this reason, we would like to review the Turkish literature about long QT families and cases diagnosed as clinically and/or genetically JLNS.

Ocal *et al*,[3] investigated the prevalence of JLNS syndrome in a school for deaf children, evaluated by ECG 350 on congenitally deaf children with an age range of 6-19 years. They found eight children with a QTc interval > 440 ms were further studied by cardiac examination, repeat ECGs (three times), Holter monitoring, echocardiography, and exercise testing. They had assessed the families for a history of syncope and deafness and who underwent ECG evaluations regarding lengthened QTc interval. Among these eight children, only two girls aged 14 and 15 years were diagnosed as having LQTS according to Schwartz's criteria (0.57% of the 350 deaf children; 95% confidence intervals 0, $\leq P \leq 0.013$). At the end of this study, the authors reported that the prevalence of JLNS among these congenital deaf children was 0.21%.

After that study, Ilhan *et al*,[4] in their study pointed out the

diagnostic importance of the ventricular depolarization parameters and prevalence of JLNS among 132 children with congenital hearing loss (CHL). They found that patients with CHL and JLNS (n: 5; two family + 1 JLNS from the family pedigree, died before) had significantly longer mean values of QT, QTc, JT, and JTc intervals and dispersion values than those of CHL without JLNS (n: 127). In another study, the researchers analyzed the prognostic significance and effectiveness of 24-h Holter monitoring recordings in pediatric cardiac patients. They obtained 490 Holter monitoring recordings from 367 cases in Ege University Medical Faculty, Paediatric Cardiology Department, from 1999 to 2001 and these recordings were analyzed retrospectively. They found that 54% of the cases were normal whereas dysrhytmia of a wide spectrum was observed in 46% of the cases. The distribution of these findings were ventricular dysrhytmia 25%, supraventricular dysrhytmia 10%, supraventricular and ventricular dysrhytmia together 5%, complete A-V block 4%, and long QT syndrome was 2%. Among these 2% LQTSs, just one JLNS, two Romano Ward, and three sporadic cases were observed.[4]

In another study, Tutar *et al*,[6] examined the ECG traces of 397 deaf children, after exclusion of JLNS (n: 3) and compared them with those of 361 normal hearing counterparts.

Acet *et al*,[7] reported two sisters with Jervell-Lange-Nielsen Syndrome (JLNS). They reported about a 21-year-old woman who was referred to the emergency unit due to recurrent seizures. Her corrected QT interval was 491-506 ms, the patient died because of sepsis within 61 days of her hospitalization. The authors found out that she had a congenital sensori-neural hearing loss and one sibling of her died when she was 2 years old and had a sister with hearing loss from her family history. They evaluated the patient's 16-year-old sister and found out that her corrected QT interval was 474 ms.

Herguner *et al*,[8] also reported a case with congenital sensorineural hearing loss and syncopes mimicking seizures. They also found out that the case was born as the third child of consanguineous marriage. They also reported that in the pedigree of the case, there were six persons affected except the case. They also had hearing loss and died suddenly during their childhood.

In another case report, the authors described two siblings with JLNS. They reported a 3-year-old girl who was admitted to emergency service with respiratory arrest and loss of consciousness. Her electrocardiography showed that she had torsade de pointes type ventricular tachycardia. Defibrillation and anti-arrhytmic medications were failed and the patient died. The authors found from the retrospective history that she and her sister were deaf. They evaluated the patient's 5-year-old sister and found that her ECG showed the corrected QT interval of 480 ms.[8]

In the light of all these studies and reports, there were 13 cases clinically diagnosed as JLNS, 8 cases diagnosed again clinically from retrospective history of the family pedigree, and two genetically confirmed JLNS in Turkish population.

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