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ORIGINAL ARTICLE / KLİNİK ÇALIŞMA

Prevalence of Jervell-Lange Nielsen syndrome in children with congenital bilateral sensorineural hearing loss

Konjenital bilateral sensörinöral işitme kaybı olan çocuklarda Jervell-Lange Nielsen sendromu prevalansı

Yakup Ergül, M.D. [1], **Hasan Candaş Kafalı, M.D.** [1], **Erman Cilsal, M.D.** [1], **Bekir Yükçü, M.D.** [1], **İbrahim Yaman, M.D.** [2], **Filiz Çetinkaya Işık, M.D.** [2], **Alper Güzeltaş, M.D.** [1], **Mehmet Ertürk, M.D.** [3]

[1]Department of Pediatric Cardiology, University of Health Sciences İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey
[2]Department of Nursing, University of Health Sciences İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey
[3]Chief of Hospital, University of Health Sciences İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey

ABSTRACT

Objective: Long QT syndrome (LQTS) is an inherited cardiac ion channel disorder (channelopathy) that is characterized by prolonged QT intervals on the electrocardiography (ECG) and possess the risk of sudden cardiac death (SCD). Jervell-Lange Nielsen syndrome (JLNS) is a specific subtype of LQTS that is accompanied by congenital sensorineural hearing loss, inherited autosomal recessively, and higher risk of SCD. In this study, we aimed to investigate JLNS prevalence in deaf children attending special schools for hearing loss, located in our province.

Methods: An ECG screening program was conducted in 6 special schools for children with hearing loss in İstanbul and a total of 440 students between 6 and 18 years old were included. Corrected QT interval (QTc) was calculated using the Bazett formula. Notably, 51 students, detected with any abnormal finding on ECG, were invited to our center for a comprehensive examination.

Results: A total of 8 patients were found with a prolonged QT interval. JLNS was diagnosed in 4 (0.9%) patients. In addition, 2 students had already been diagnosed with JLNS at another center earlier. The other 2 students, being siblings, were newly diagnosed with JLNS; and appropriate treatment was initiated. Genetic testing revealed a pathological homozygous mutation in *KCNQ1* gene. The younger sibling (Case 1), who possessed a QTc of greater than 500 ms and a history of syncope, which was very suspicious for SCD, was implanted an implantable cardioverter-defibrillator. Propranolol treatment was initiated for both siblings.

Conclusion: JLNS should be carefully considered and screened, especially in patients with a history of congenital deafness.

ÖZET

Amaç: Uzun QT sendromu (LQTS), elektrokardiyografide (EKG) ölçülen QT mesafesinde uzama ile kendini gösteren ve ani kardiyak ölüm (AKÖ) riski barındıran, genetik geçişli/ailevi bir kardiyak iyon kanal defekti (kanalopati) hastalığıdır. Jervell-Lange Nielsen sendromu (JLNS) ise doğuştan sensörinöral sağırlık ile seyreden, otozomal resesif geçişli, ve AKÖ riski daha yüksek, özel bir LQTS tipidir. Bu çalışmada ilimiz sınırları içerisinde işitme engelliler okullarına devam eden öğrenciler arasında JLNS prevalansı araştırıldı.

Yöntemler: İstanbul il sınırları içerisinde hizmet veren 6 adet işitme engellilere özel okulda, 6 ila 18 yaş arasındaki 440 öğrenciyi içeren bir EKG taraması gerçekleştirildi. Düzeltilmiş QT değeri (QTc) Bazzet formülüne gore hesaplandı. EKG'sinde herhangi anormal bir bulgu saptanan toplam 51 öğrenci ayrıntılı bir değerlendirme için merkezimize davet edildi.

Bulgular: EKG okul taramasında toplam 8 hastada uzun QT saptandı. Dört hastada JLNS tanısı konuldu (%0.9). Bu hastalardan 2 tanesi daha önceden tanı aldıkları merkezde takibe devam edilmek üzere yönlendirilirken, yeni tanı alan iki hasta merkezimizde takip ve tedavi altına alındı. Genetik test sonucu *KCNQ1* geninde patolojik homozigor mutasyonu gösterdi. Aynı zamanda kardeş olan bu iki hastadan küçük olanın (Vaka 1) 500 ms aşan QTc değerleri olması ve öyküsünde şüpheli AKÖ düşündüren senkop öyküsü olması nedeniyle medikal tedavi dışında ayrıca implante edilebilir kardiyoverter defibrilatör implante edildi. Her iki kardeşe de medikal tedavi olarak propranolol başlandı.

Sonuç: Doğuştan sensörinöral sağırlık öyküsü olan hastalarda JLNS özellikle dikkate alınması ve ayrıntılı araştırılması gereken bir hastalıktır.

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Correspondence: Yakup Ergül, M.D. Department of Pediatric Cardiology, University of Health Sciences İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey
Tel: +90 212 692 20 00 e-mail: yakupergul77@hotmail.com

Congenital long QT syndrome (LQTS) is genetic arrhythmia, characterized by a prolonged QT interval on electrocardiography (ECG), and may result in recurrent episodes of syncope and sometimes even sudden cardiac death (SCD). SCD is associated with torsades de pointes (TdP), a specific type of polymorphic ventricular tachycardia. The molecular pathology is in the ion channels of the myocardium, called channelopathy.[1-3] The most common form of LQTS is the Romano-Ward syndrome, following an autosomal dominant pattern of inheritance.[4] A rare, but more deadly form, called Jervell-Lange Nielsen syndrome (JLNS) is inherited as autosomal recessive and is associated with bilateral congenital sensorineural hearing loss.[5] Prevalence was reported to be 0.21% in children with congenital deafness earlier.[6] Although it can be easily diagnosed with a 12-lead ECG, by measuring the QT interval, it possess a high risk for SCD if not diagnosed and treated in time. In this study, we aimed to detect the prevalence of JLNS in children attending special schools for the hearing impaired, located in our province.

METHODS

Patients and methodology

This study was conducted as a social responsibility project at our center, obtaining the necessary permissions from the relevant authorities. The study was designed in a prospective manner and children 6 to 18 years of age, attending the 6 special schools for the hearing impaired located in the İstanbul province were included in the study.

The local ethics committee in our center approved the study with the decision number of 2018/30. A questionnaire was conducted in these schools with a total of 762 students, and informed consent was obtained from the parents of the students. Demographic characteristics and a short medical and family history were obtained from all the students attending these 6 schools using the questionnaire. On the basis of the results of the questionnaire, 440 patients with congenital bilateral sensorineural hearing loss were included in the study. A team of specialist nurses and technicians visited each school and conducted an ECG screening [using Hewlett Packard® 4745A Electrocardiograph (HPE, Houston, TX, USA) under the supervision of their teachers. A standard 12-lead ECG with a 25 mm/s paper speed and 1 mV/cm amplitude was obtained from each of the participants in addition to arterial blood pressure measurements. The electronically recorded ECGs were transferred to the digital ECG data and analysis system (Muse® Cardiology Information System, 9.0 SP-7 12-SL, GE Medical Systems Information Technologies, Inc, Milwaukee, WI, USA) used at our pediatric cardiac center. QT intervals were measured digitally by 2 experienced pediatric cardiologists/electrophysiologists conducting the study. Extremity lead II or precordial lead V5 was preferred for measurement, and the digital calipers were set at the beginning of Q wave and end of T wave (the point where the tangent drawn to the steepest slope crosses the isoelectric line), using full scale magnification. Corrected QT (QTc) values for heart rate were calculated automatically according to the Bazett formula.[7]

Abbreviations:

ECG	*Electrocardiography*
ICD	*Implantable cardioverter-defibrillator*
JLNS	*Jervell-Lange Nielsen syndrome*
LQTS	*Long QT syndrome*
QTc	*Corrected QT interval*
SCD	*Sudden cardiac death*
TdP	*Torsades de pointes*

In case of sinus arrhythmia, the maximum and minimum QTc values were averaged. Patients with a QTc value higher than normal (≥440 ms for boys and ≥450 ms for girls) or with any other abnormality diagnosed on the ECG were called for a detailed examination to our center. During the hospital visit, the patients were assessed with a detailed medical and family history, a complete physical examination, control ECG, standard transthoracic echocardiography, and 24-hour Holter ECG monitoring and treadmill stress test when required.

Any syncope attacks, drowning, and aborted cardiac arrest were investigated carefully in personal history and also in family history (among relatives younger than 35 years). Implanted ICD/cardiac pacemaker and a history of epilepsy (as a misdiagnosis made for syncope associated with genetic arrhythmia syndromes like JLNS) were also investigated in detail.

A 24-hour Holter ECG monitoring and a treadmill stress test was conducted in all the patients with a long QTc. Before starting the stress test, baseline ECG readings were obtained in the supine and standing positions. After conducting a modified Bruce protocol, achieving about 85% of the predicted maxi-

Table 1. Results of ECG screening in 6 schools for children with hearing loss and the detected JLNS cases with their clinical features

School number	Students screened, n	Abnormal ECG finding, n (%)	JLNS diagnosed, n	Clinical features of the JLNS cases, diagnosed
1	39	10 (25)	1	QTc 500 ms, already diagnosed before and during follow-up at another center.
2	83	8 (9.6)*	1	QTc 600 ms; already diagnosed before and during follow-up at another center. ICD implanted earlier.
3	36	3 (8.3)	0	-
4	59	4 (6.7)	1	Case 1:† QTc, 540-600 ms; new diagnosis, ECG, normal, 24-hour Holter: average QTc, 500 ms; max QTc, 600 ms.
5	80	8 (10)	1	Case 2:‡ QTc, 450 ms; new diagnosis; ECG, normal; 24-hour Holter: average QTc, 460 ms; max QTc, 500 ms. Treadmill test: max QTc of 520 ms at the fourth minute of the recovery phase.
6	143	16 (11)§	0	-
Total	440	49 (11.1)	4 (0.9)	

ECG: electrocardiography; JLNS: Jervell-Lange Nielsen syndrome; ICD: implantable cardioverter-defibrillator; max: maximum; QTc: corrected QT interval.
*Including 3 patients with a borderline long QTc.
†Evaluated as having high risk for sudden cardiac death and was implanted an ICD in addition to medical therapy with propranolol.
‡The older sibling of Case 1. Only medical therapy was initiated (propranolol).
§Including 1 patient with a borderline long QTc.

mum heart rate, or the 90th percentile of the expected effort time for age and sex, ECG was recorded at the 1, 3, 4, 5, and 7 minutes of the recovery phase. In 24-hour Holter ECG, numerous QTc measurements were made manually in addition to the automatic QTc calculator of the Holter system. All the records were carefully scanned for specific finding of LQTS, such as T wave alternance.

Statistical analysis

Data related to the patients were recorded on the electronic database system (FileMaker® Pro-12, FileMaker, Inc.; Santa Clara, CA, USA) used in our center and transferred to an Microsoft Excel sheet (Microsoft; Redmond, WA, USA) for basic statistics such as mean, median, and standard deviation.

RESULTS

Of 440 students included in the study, 255 (57.9%) were boys. Their average age was 14 years and 7 months±3 years and 9 months (range, 6-18 years). After the ECG screening conducted at their schools, 51 students were called for a detailed investigation. In 49 of them, suspected or abnormal ECG findings were detected; and in 2 of them, adequate ECG evaluation could not be performed owing to extensive artefacts (Table 1). QT prolongation was found in 8 (1.8%) patients, and JLNS was diagnosed in 4 (0.9%) patients.

Among the 8 patients with a prolonged QT interval, a marked prolongation ≥500 ms was detected in 3 patients. The patient with a QTc of 600 ms was already diagnosed with JLNS earlier and had an implanted ICD from another center. The patient with a QTc of 500 ms was also diagnosed with JLNS at another center and declined to be admitted for further investigation at our center. The other patient with a QTc of 500 ms was diagnosed for the first time (Case 1) and was a 9-year-old boy with bilateral sensorineural hearing loss and a history of a very suspicious syncope, explained by the family as near drowning. The other 5 patients with a prolonged QT interval had borderline values varying between 450 and 470 ms, one of them (Case 2, the sibling of Case 1) was a 16-year-old boy. He had neither a previous history or syncope nor any other symptoms; however, he had bilateral sensorineural hearing loss. He had a maximum QTc of 540 ms during his treadmill test, mea-

Table 2. Pathological ECG findings detected in the screening program and the results/endpoints

ECG finding	Number of students diagnosed on ECG screening	Number of students admitted for detailed exam at our center	Results
Prolonged QT interval	8	6	2 JLNS cases newly diagnosed 1 JLNS case diagnosed previously (other center) 3 cases with borderline prolonged QT, follow-up
Ventricular pre-excitation (WPW)	2	1	1 WPW case newly diagnosed, referred for EPS
Ectopic atrial rhythm	5	3	Benign, follow-up
Early repolarization	4	1	Benign
Supraventricular ectopy	2	1	Benign
First degree AV block	1	-	-
Junctional escape	1	-	-
Left axis deviation	5	3	Benign
Right axis deviation	1	-	-
LV hypertrophy	4	1	Echo: normal
RV hypertrophy	1	-	-
Incomplete RBBB	5	2	Benign
Right atrial dilatation	1	1	Echo: normal
Sinus bradycardia	5	1	Benign
Sinus tachycardia	1	1	Benign
Negative T wave	1	-	-
Low QRS voltage	1	-	-
Undetermined rhythm	1	-	-
Artefacts	2	1	Control ECG: normal
Total	51	23*	

AV: atrioventricular; ECG: electrocardiography; EPS: electrophysiological study; JLNS: Jervell-Lange Nielsen syndrome; LV: left ventricle; RBBB: right bundle branch block; RV: right ventricle; WPW: Wolff-Parkinson-White.
*Patients with an ECG finding who were admitted to our center after the screening program were investigated further in detail with echocardiography (n=23), ambulatory 24-hour Holter (n=7), and treadmill stress test (n=6).

sured at the fourth minute of the recovery phase. Of note, 3 patients with borderline QTc values, investigated further with 24-hour Holter ECG and treadmill tests, were found not to have JLNS. However, because of their borderline QTc values measured during the school screening, they were advised to follow-up for some time. One patient with a borderline QTc did not want to be admitted to our center for further investigation and could not be contacted again during the span of the study. As a result, 4 patients were diagnosed with JLNS (4/440), and the prevalence was found to be 0.9% within the study population.

Details of the various pathologic ECG findings other than QT prolongation are shown in Table 2. Echocardiography revealed no pathology in any of the patients, and the 24-hour Holter ECG in 2 patients and treadmill stress test in 2 patients were conducted for further investigation. Most of them were removed from follow-up after they were shown to have no significant pathologies. Invasive electrophysiological study was recommended for the asymptomatic patient with ventricular preexcitation (Wolff-Parkinson-White Syndrome). The 2 patients with newly diagnosed JLNS were evaluated in detail. Case 1 was a 9-year-old boy with congenital bilateral sensorineural hearing loss. He was using an external hearing aid, and there was a plan to implant a cochlear device in the future. The patient did not have any actual symptoms but had a very suspicious history of a syncope episode nearly 1 year earlier. He fell into

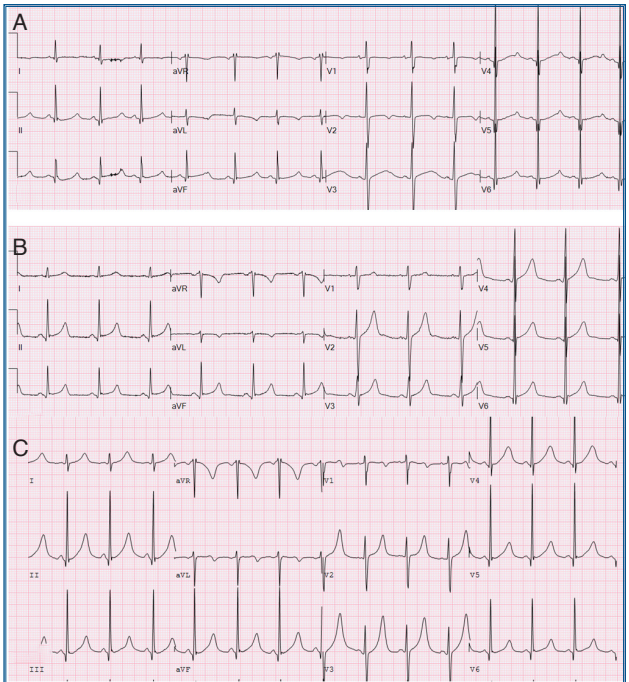

Figure 1. (A) ECG findings from Case 1 with a QTc measured as 540 ms. **(B)** Resting ECG from Case 2 during treadmill test with a QTc measured as 450 ms. **(C)** ECG findings from Case 2 during the fourth minute of the recovery phase on treadmill stress test, with a QTc measured as 520 ms.
ECG: electrocardiography; QTc: corrected QT interval.

shallow water when playing at a seaside and nearly drowned. His father performed first aid intervention (a short mouth-to-mouth resuscitation), although the father was not sure if the patient had a cardiac arrest. He recovered in a short time, and no further medical investigation was sought by the family. On his ECG obtained during the school screening, QTc was calculated as 540 ms (Figure 1A). On the control ECG obtained at our center, QTc was calculated as 600 ms; therefore, there was no need for a treadmill, which could be dangerous in this situation. Echocardiography revealed a structurally normal heart. His 24-hour Holter ECG revealed a very long QTc of >500 ms during the entire recording and up to 600 ms for short periods. Although not a typical T-wave alternance from beat to beat, some T wave alterations suggested a high risk for SCD. As a result, the patient was assessed as having a very high risk for SCD, and an epicardial ICD was implanted for prophylaxis. Propranolol was initiated with a dose of 3 mg/kg/day in 3 separate doses per oral. One month later, the medical therapy was changed to nadolol 1.5 mg/kg/day in 2 separate doses because of hallucinations as a drug side effect of propranolol. His complaints declined dramatically thereafter. Meanwhile, he was referred to a genetic center for a genetic study, revealing a pathologic homozygous mutation in *KCNQ-1* gene. Pedigree (family tree) drawn for this patient and his sibling (Case 2) is shown in Figure 2. There was a consanguineous marriage between his parents (first degree cousins) as expected. Although both his parents and his sister had no hearing loss, they had borderline prolonged QTc on their ECG, suggesting a heterozygous "carrier" state. Both parents were referred to our adult cardiology department, and propranolol therapy and other typical measures like sports restriction and avoidance of drugs causing QT prolongation were recommended for his 13-year-old sister. She was also referred for genetic study. The 2 early infant deaths seen in his cousins were noted. The patient is still having close follow-up under medical treatment at our pediatric cardiac arrhythmia unit, without any other problems.

Case 2 was a 16-year-old male patient with congenital bilateral sensorineural hearing loss. He was using an external hearing aid. He never had a complaint of syncope or palpitation before. QTc was calculated as 450 ms (Figure 1B), both on the ECG obtained at school screening and at our center during the detailed investigation. Echocardiography revealed a structurally normal heart. His 24-hour Holter ECG revealed a borderline QTc value between 440 and 460 ms; however, the QTc was prolonged up to 500 ms for only short periods; and therefore, a treadmill stress test was conducted and revealed a borderline QTc at rest and a marked prolongation up to 520 ms in the recovery phase at the fourth minute (Figure 1C). The result of his genetic study has not yet been released. Medical treatment with propranolol at a dose of 3 mg/kg/day in 3 separate doses was also initiated. The patient is still in close follow-up under medical treatment at our pediatric cardiac arrhythmia unit without any other problems.

DISCUSSION

During the clinical course of patients with LQTS, recurrent syncopes and even SCD owing to malignant TdP attacks can be observed.[8-10] At present, more than 15 subtypes of LQTS are detected genetically, most of them being inherited in an autosomal dominant pattern. Associated congenital bilateral sensori-

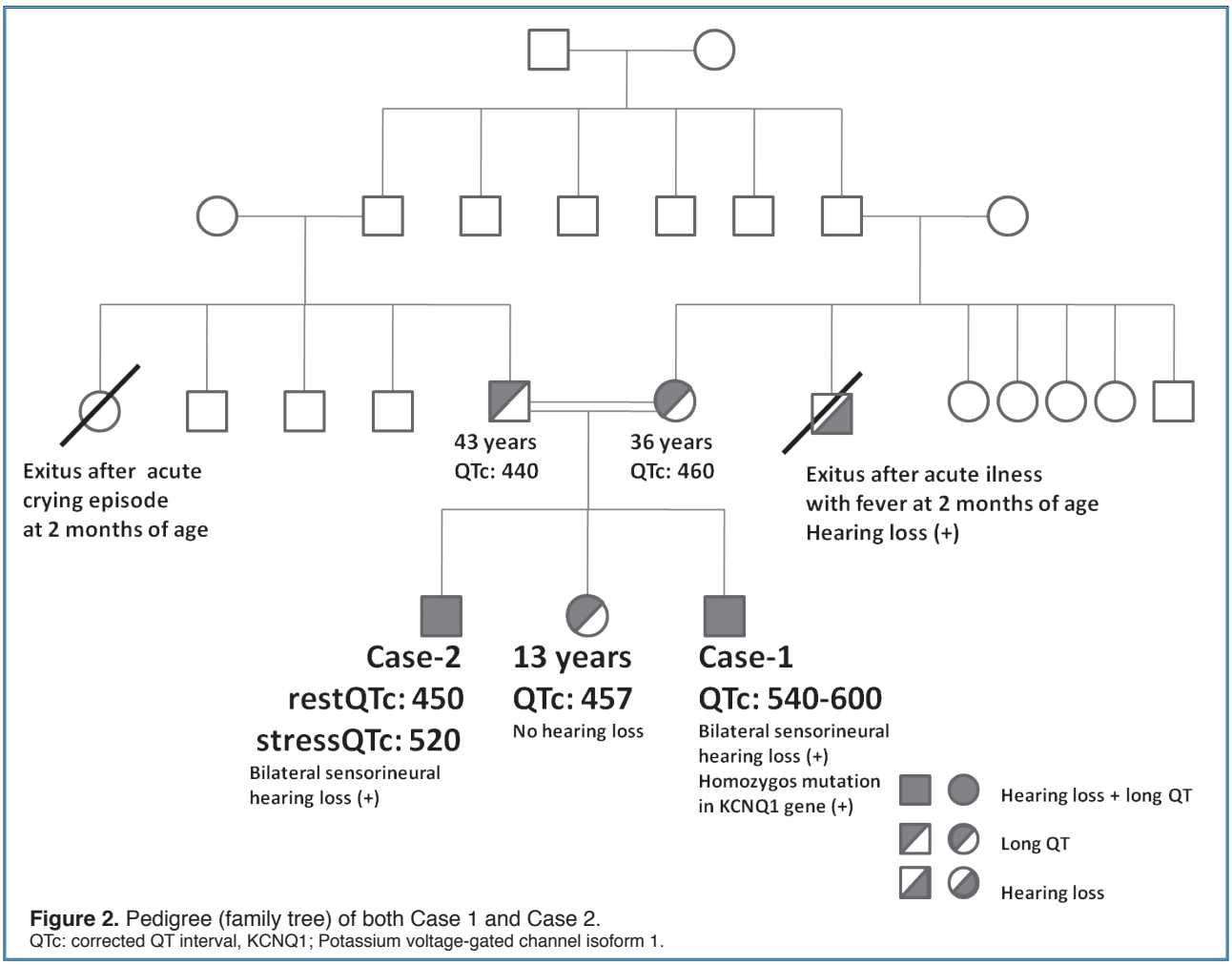

Figure 2. Pedigree (family tree) of both Case 1 and Case 2.
QTc: corrected QT interval, KCNQ1; Potassium voltage-gated channel isoform 1.

neural hearing loss is characteristic for JLNS, which is inherited in an autosomal recessive pattern.[11,12]

In patients with JLNS, the risk of SCD is higher than in other types of LQTS. According to many studies, a QTc value of >550 ms should indicate primary prophylaxis with an ICD implantation or left cardiac sympathetic denervation even though the patient is asymptomatic.[13]

The 24-hour Holter ECG monitoring is very useful for diagnosis and especially for risk assessment. Prolonged QT intervals occurring during any time of the day can be detected. Deep bradycardia periods, QTc values of >500 ms during most of the monitored time, and especially T-wave alternance are associated with high risk. In particular, T-wave alternance is an indication of electrical instability and serious ventricular arrhythmias that can develop in the near future.[4,5,9] In Case 2, we observed QTc values between 440 and 460 ms most of the time during the 24-hour monitoring; however, there were also times where QT was prolonged with an QTc value up to 500 ms. In Case 1, QTc values were >500 ms during the entire 24-hour monitoring period, reaching up to 600 ms, indicating a high risk for SCD. Some T-wave changes, although not typical T-wave alternance, and a history of syncope and near drowning, requiring first aid, were considered as high risk. Therefore, in addition to medical treatment, ICD implantation was recommended and performed at our center.

A treadmill stress test is also very helpful for diagnosis, especially in borderline cases; and it can also help to differentiate between some subtypes of LQTS.[14-18] Although the prolongation of QT interval during the recovery phase, especially at the fourth minute, is typical for type-1 LQTS, significant prolongation in the seventh minute of the recovery phase, in addition to the QT hysteresis, indicate a type-2 LQTS and is consistent with a dangerous electrical instability

Table 3. Prevalence of JLNS in children with congenital sensorineural hearing loss reported previously in the literature*

Study (authors)	Publication date	Patients, n	JLNS, n	JLNS, %
Fraser et al. (as cited in Schwartz et al.[6])	1964	2994	9	0.30
Puletti et al. (as cited in Schwartz et al.[6])	1967	211	0	0
James et al. (as cited in Schwartz et al.[6])	1967	367	0	0
Dupuis et al. (as cited in Schwartz et al.[6])	1969	182	0	0
Sanchez et al. (as cited in Schwartz et al.[6])	1969	511	1	0.19
Fay et al. (as cited in Schwartz et al.[6])	1971	1126	1	0.08
Dolara et al. (as cited in Schwartz et al.[6])	1971	262	1	0.38
Pernot et a. (as cited in Schwartz et al.[6])	1971	298	1	0.33
Gillet et al. (as cited in Schwartz et al.[6])	1972	231	1	0.43
Total (combined by Schwartz et al.[6])	1964-1972	6557	14	0.21
Schwartz et al.[6]	1975	375	0	0
Ocal et al.[22]	1997	350	2	0.57
Tuncer et al.[23]	2000	132	5	3.78
Sanecka et al.[24]	2016	1027	5*	0.48
Present study	2021	440	4	0.9

JLNS: Jervell-Lange Nielsen syndrome; LQTS: long QT syndrome.
*These patients were diagnosed with a dominant mutation, not JLNS, but LQTS.

during depolarization in the cardiac cell membrane.[18-20] In our study, the treadmill stress test was helpful to rule out a LQTS diagnosis in 3 patients with borderline QTc values and was also useful for support of the diagnosis in Case 2 (revealing a prolongation of QTc up to 500 ms at the fourth minute of the recovery phase).

Although the association between sensorineural hearing loss and LQTS is still not clear, Mulroy and Harrison[21] have found in their study that a small lesion occurring in the nodes and optic placode regions of the chick animal model during the early embryologic development caused prolongation in QT interval on the basis of the ECG. It was suggested that abnormalities in this region occurring during early stages of development play a role in the pathogenesis of the disorder.

In a study by Schwartz et al.[6] involving results from 9 previous studies, which included a total of 6557 patients, the prevalence of JLNS in children with hearing impairment was calculated as 0.21% (within range of 0%-0.43%) (Table 3). The prevalence of JLNS among children with hearing loss was found to be 0.9% in our study. These high values of prevalence underline the importance of ECG screening programs among such children.

In a study conducted in Turkey, Ocal et al[22] investigated the prevalence of this syndrome in a school for deaf children, evaluating 350 congenitally deaf children with an age range of 6 to 19 years using ECG. Of note, 8 children with a QTc interval >440 ms were further studied by cardiac examination, repeat ECGs (3 times), Holter monitoring, echocardiography, and exercise testing. Among these 8 children, only 2 (0.57%) girls aged 14 and 15 years were diagnosed as having LQTS according to Schwartz's criteria (95% confidence intervals 0, p≤0.013). They also concluded that JLNS should be considered in children with syncope or a family history of it, especially those with congenital deafness.

In another study conducted in Turkey, Tuncer et al.[23] have attempted to identify patients with JLNS patients among 132 deaf-mute school children. A total of 5 deaf-mute patients had JLNS (a prevalence of 3.78%). They have also determined that the deaf-mute children who did not meet the criteria for JLNS still had evidence of subtle de-repolarization abnormalities evidenced by intermediate prolongation of QTc, JTc, and the corresponding measures of dispersion, and they believed an ECG examination of

the deaf-mute children would reveal this potentially life-threatening syndrome.

In a recent study by Sanecka et al.[24] from Poland, published in 2016 and including 1,027 patients with hearing loss (mean age 21.8±19.9 years, including 441 children under 14 years) abnormal QTc was found in 13 (4.1%) women, 20 (7.3%) men, and 72 (16.3%) children. In the studied group, no recessive mutation of *KNCQ1* or *KCNE1* was found. In 5 patients, dominant mutations in *KCNQ1* (n=1), in KCNH2 (n=3), and in SCN5A (n=1), which were pathogenic for LQTS and 2 mutations of unknown clinical significance in SCN5A were found. Overall, of these 6 patients, LQTS was diagnosed in 3 patients who were asymptomatic, with abnormal QTc and in 2 patients with normal QTc, but who were previously treated for epilepsy. The authors concluded that JLNS is a very rare condition even in a population with hearing loss.

LQTS has a familial inheritance, thus 70% to 80 % of the "index" cases have actually other family members possessing a long QT (probands), being asymptomatic, and undiagnosed, although also at risk of SCD.[12] For patients with a borderline QTc value and modified Schwarz scores between 2 and 3.5, a final diagnosis for LQTS cannot be established; however, it is recommended to monitor these patients for a period of time. Occasionally, reference for genetic testing may be needed.[11,12] In our study, we examined 3 patients with borderline values between 440 and 470 ms on their screening ECG and modified Schwarz scores of 2 to 2.5 in detail, which revealed normal 24-hour Holter ECG and treadmill stress tests. Their family members were also screened with ECG, revealing normal QTc values. Therefore, these patients were not diagnosed as having LQTS but were recommended for a follow-up examination.

As a first-line treatment for LQTS, it is recommended to avoid physical stress and other triggering factors such as medications causing QT interval prolongation, and an appropriate beta-blocker treatment should be administered to slow down and control the heart rate.[5,12] Left cardiac sympathetic denervation and ICD implantation are the preferred options for high-risk cases for whom standard measures are not sufficient.[25-28] In our study, Case 1 was considered to be at high risk owing to his significantly long QT intervals and a history of syncope suggesting a suspicious cardiac arrest; and therefore, an ICD implantation was recommended.

At present, efforts are continuing for performing QTc screenings with routine ECGs in the newborn in certain countries and centers.[29] ECG scans may be performed at an affordable price at least for patients with JLNS, a high-risk group among patients with LQTS, and in children with congenital sensorineural hearing loss, especially when such patients may be accessed collectively, such as during the newborn hearing screening or as is the case in this study, in the special schools for the hearing impaired.[22-24]

Limitations

This study has several limitations. First, only single ECG records were obtained from each participant. We know that sometimes QT measurements can be found normal even in LQTS patients, so serial ECG recordings should be obtained from each patient. Serial ECG recordings on different days would diagnose more patients with prolonged QT intervals. Second, the sample size is relatively small, and neither all of the students visiting the six schools involved in this study agreed to participated in this study, nor all of the participants found to have positive ECG findings and called to our center for further examination joined the call.

Conclusion

It is very important to conduct ECG screening programs, at least for the populations possessing a higher prevalence of LQTS and a higher risk of SCD. Multiple cases of SCD can be prevented with simple measures like beta-blocker therapy, sports restriction, and avoidance of medications causing QT prolongation. In addition, a screening of the probands can diagnose many undetected cases. QT measurement should be a routine part of all pediatric ECG evaluations, regardless of having hearing loss or not. It is especially important for routine examinations before participating in sports, because in most types of LQTS, the risk of lethal arrhythmias and SCD increase with exercise.

Ethics Committee Approval: Ethics committee approval was received for this study from the Clinical Researches Ethics Committee of University of Health Sciences İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital (Approval Date: June 8, 2018; Approval Number: 2018/30).

Peer-review: Externally peer-reviewed.

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