

Ventricular Tachycardia Associated Hereditary Prolonged Q-T Interval

JAVED IQBAL*

SUMMARY:

Ventricular Tachycardia has been described in association with a variety of congenital and acquired causes of Q - T prolongation. Among the acquired causes, most important are anti arrhythmic drugs like Quinidine, Pronestyl, Calcium antagonists like Segontin, Herbesser. Among other acquired causes are Hypokalemia, Hypocalcaemia, Hypomagnesemia, etc.

Regarding the congenital causes of prolonged Q - T, two syndromes are described. These are:

- i) Jervell and Lange Neilsen Syndrome;
- ii) Romano-Ward Syndrome.

HEREDITARY PROLONGED Q - T INTERVAL

Jarvell and Lange Neilsen Syndrome is characterized by congenital deafness, prolonged Q - T interval in ECG, Syncope and sudden death. It is inherited autosomal recessive disease. It was first described by Jarvell and Lange Neilsen in 1957 (1). Incidence is one per 300,000 population.

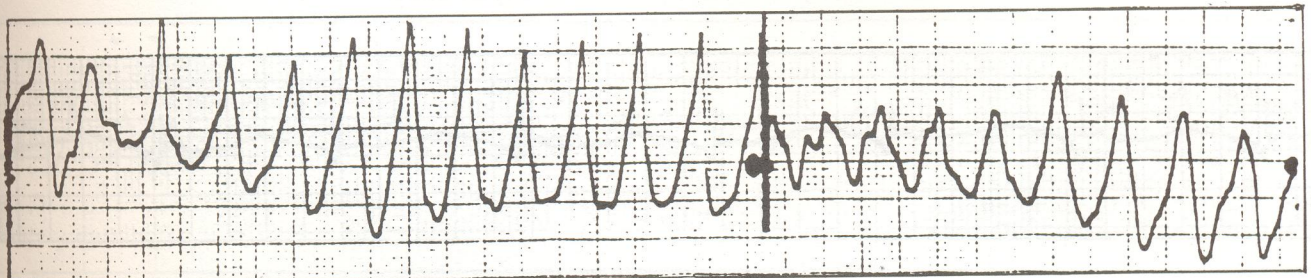
Romano Ward syndrome which is an autosomal dominant disease is characterized by prolonged Q - T interval in ECG, syncope and sudden death without congenital deafness (2).

Syncope and death are due to ventricular arrhythmias, often during emotional or physical stress. The commonest type of ventricular

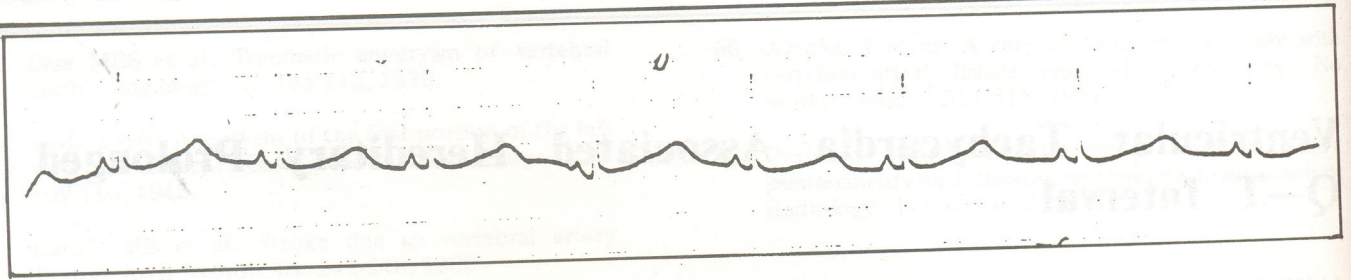
tachycardia seen during syncope in these syndromes is of Torsade de pointes (4). It is form of polymorphous ventricular tachycardia in which polarity of QRS complex exhibits phasic alternation in both axis and rate.

CASE REPORT - I

A deaf and dumb girl of nine years, named Robina presented on 1st of March, 1986 with history of syncopal attacks for the last seven years. She used to have these attacks five or six times in a year. For the last one week or so she had frequent syncopal attacks. According to the story told by her mother and observation made by us in the ward, she cried during the attack, then became unconsciousness. Attacks were self-terminating. Episodes were not like epileptic fits or hysterical fits. On clinical examination, no obvious cause was found of these syncopal attacks. Laboratory investigations revealed no abnormality. ECG recorded during such an attack showed ventricular tachycardia of Torsade de pointes type.



* Mayo Hospital Lahore



ECG recorded during normal state showed prolonged U-1 interval (0.60 sec.) (Fig. 2.) Echocardiography was done which revealed no cardiac abnormality. The girl was diagnosed as a case of Jarvell and Lange Neilsen Syndrome.

CASE REPORT - II

A child, Usman Ali, three and half years old presented to us on 8th of October, 1986 with history syncopal attacks for the last two years. Physical examination and laboratory investigation revealed no abnormality. ECG was done and it showed prolonged Q - T interval. (Fig.3.)

Echocardiography was done which revealed no cardiac abnormality. The child was diagnosed as a case of Romano-Ward syndrome.

The picture of a patient with primary prolongation of Q - T interval who has syncopal episodes in association with physical and emotional stress is so typical that the diagnosis is elementary if the physician is aware of syndrome. The sudden death in these patients requires a correct diagnosis so that proper and effective treatment can be instituted. For this reason, a proposal of diagnostic criteria was made in Texas on March, 24, 1984 in a symposium on "Prolonged Repolarization".

**TABLE I
DIAGNOSTIC CRITERIA***

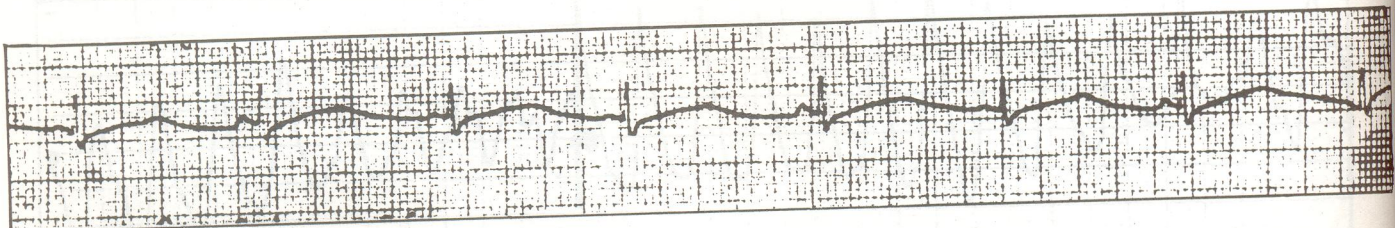
Major	Minor
- Prolonged Q - T interval (Q - T 440 msec.)	- Congenital Deafness
- Stress induced syncope.	- Episodes of T-wave alternans.
- Family members with LQTS.	- Low heart rate (in children)
	- Abnormal ventricular repolarization.

*The diagnosis should be made in the presence of two major or one major and two minor criteria.

Role of Autonomic Nervous System

Many authorities agree that ventricular tachycardia in these cases is caused by imbalance of right and left sympathetic tone. A strong relationship between this syndrome and sympathetic nervous system is suggested by several factors.

1. Ventricular arrhythmias can be triggered by stressful events known to increase sympathetic tone.



2. Efficacy in preventing the development of ventricular arrhythmias has been achieved by beta-adrenergic blocking drugs and by unilateral (left) cervicothoracic sympathetic ganglionectomy.
3. Prolongation of Q - T interval and alteration of T-waves can be produced by changes in sympathetic tone. This indeed happened with unilateral stimulation of the left stellate ganglion or with bilateral stimulation of both stellate ganglion, but with a greater intensity applied to the left.

These experiments suggested that patients with LQTS may have a congenital imbalance between right and left cardiac sympathetic innervation, with a left dominance. This would result in a prolongation of the Q - T interval both at rest, and often during episodes of alteration of the T-wave when the sympathetic activity increases. The electro-physical studies suggest that in these cases of LQTS, there is delayed and non-uniform ventricular repolarization. This non-uniform repolarization is basis of Torsade de pointes.

THERAPY.

In the treatment of the cases of Hereditary Prolonged Q - T Syndrome, remarkable efficacy in preventing malignant ventricular arrhythmias has been reported using propranolol with a decrease in mortality from 73% to 6%. Propranolol must be used in full beta-blocking dosage. The initial dose is 2mg/kg which is increased to 3mg/kg if necessary. It is important that no dose should be skipped. Some of the reported deaths in patients treated with propranolol occurred on the very first day or next-day after propranolol was stopped. Parents of such patients should be taught how to do cardio-pulmonary resuscitation. Exercise should be prohibited. Monitoring during surgical procedures is essential.

We also put both of our patients on propranolol. After propranolol was started, they had no syncopal attacks.

FOLLOW UP

Both patients visit our department monthly for followup. The girl of Jarvell and Lange Neilsen syndrome had no syncopal attack for five months when her parents stopped propranolol for her. She again started having syncopal attacks after four doses of propranolol were missed. Then they consulted us again and we advised them never to omit any dose. She is symptom free for last 4 months.

The mother of the child who has Romano-Ward disease, also missed the two doses of propranolol. The child had severe syncopal attacks. He was brought to our department and his ECG recording showed Torsade de pointes. (Fig.4)

Fig 4

I gave him double dose of propranolol and attacks stopped. I advised his mother never to omit even a single dose of propranolol. He is symptom free for last one month.

CONCLUSION

Firstly in the differential diagnosis of syncopal attacks in children LQTS must be considered as one of the possible causes. These children can be mis-diagnosed as epileptic or hysterical.

Secondly, ventricular tachycardia associated with prolonged QT Interval must not be treated with class-I anti arrhythmic agent whereas those arrhythmias associated with a normal QT Interval may be treated successfully with these drugs.

