

Relationship among myotonic dystrophy type I and Brugada Syndrome

Dr. Andrés R. Pérez Riera

We observe peculiar and many coincidences between both entities:

- 1) Both have autosomal dominant transmission;
- 2) Both are manifested predominantly initially in the productive time of life (between 20 and 30 years old)
- 3) Both present a higher incidence of atrial arrhythmias and paroxysmal AF;
- 4) Both present a higher incidence of ventricular arrhythmias;
- 5) Both may present an extreme deviation of the electric axis of QRS in the frontal plane;
- 6) Both present a frequent prolongation of the PR interval;
- 7) Both have involvement of the intraventricular His system with prolongation of the HV interval in the His electrogram;
- 8) Both present frequent complete RBBB pattern in ECG;
- 9) Our case shows that both can present upwardly convex ST segment elevation from V1 to V2;
- 10) Both present high prevalence of late potentials in signal averaged ECG (Nalos 1987; Milner 1991; Masaki 2000);
- 11) Both are considered channel diseases (Milner 1991)
- 12) Both can affect chromosome 3 (Masaki 2000; Balser 1999)
- 13) Both may present E point in QRS loop in VCG, which does not coincide with 0 point (**Mahadevan 1992**);
- 14) Both present tendency to sudden cardiac death;
- 15) Both present enhanced risk of arrhythmias with anesthetics.

References

Nalos PC, Gang ES, Mandle WJ, et al. The signal-average electrocardiogram as a screening test for inducibility of sustained ventricular tachycardia in high risk patients: a prospective study. *J Am Coll Cardiol* 1987;9:539-548

Milner MR, Hawley RJ, Jachim M, et al. Ventricular late potentials in myotonic dystrophy. *Ann Intern Med* 1991;115 (no. 8):607-613

Masaki R, Watanabe I, Sugimura H, et al. Role of signal averaged electrogram in the identification of high risk patients in the syndrome of right bundle branch block and ST segment elevation in leads V1-V3. *PACE* 2000;23:732

Balser JR. Sodium "channelopathies" and sudden death: Must you be so sensitive? *Circ Res* 1999; 85:872-874

Mahadevan M, Tsilfidis C, Sabourin L, et al. Myotonic dystrophy mutation: an unstable CTG repeat in the 3' untranslated region of the gene. *Science* 1992;255:1253-1255.