East African Scholars Journal of Medical Sciences
Abbreviated Key Title: East African Scholars J Med Sci
ISSN 2617-4421 (Print) | ISSN 2617-7188 (Online) |
Published By East African Scholars Publisher, Kenya

OPEN ACCESS

Volume-2 | Issue-5 | May -2019 |

Letter to the Editor

Value of electro-anatomic mapping of the right cardiac chambers in myotonic dystrophy type 1

Josef Finsterer, MD[1] and Claudia Stöllberger, MD[2]

[1]Krankenanstalt Rudolfstiftung, Messerli Institute, Austria
[2]Medical Department, Krankenanstalt Rudolfstiftung, Vienna, Austria

*Corresponding Author
Univ.Doz. DDr. J. Finsterer

Keywords: myotonic dystrophy, electro-anatomic mapping, cardiac involvement.

With interest we read the article by Dello Russo *et al.,* (2006) who interpreted altered electro-anatomic patterns of the right cardiac chambers in 13 patients with myotonic dystrophy-1 as myocardial involvement in the disease (Russo, A. D. *et al.,* 2006). The study raises the following concerns.

Which were the clinical, ECG and echocardiographic findings in the 13 patients? Which cardiac medication did these patients take? Did the authors find left ventricular hypertrabeculation/noncompaction (Finsterer, J. *et al.,* 2001) in any of their patients?

From which type of supra-ventricular tachycardia suffered the control subjects? To which degree did these rhythm abnormalities influence the results?

It is speculated that low-amplitude potentials reflect the presence of either fibrous tissue or fatty infiltration. How can one be sure that low-amplitude potentials resulted from affection of the myocardium and not from affection of the conduction system, from changes of the electric properties of the endocardium. focal myocarditis, atrophic myocardiocytes or from desynchronisation of the stimulus propagation velocity (Pelargonio, G. *et al.,* 2002)?

Assuming slowed impulse propagation in all areas of the right atrium, one would also expect low amplitude and prolonged P-waves on standard ECG.

Did 24h-ECG indicate a propensity towards ventricular arrhythmias?

According to tables 2 and 3 mean values of the potential amplitudes were all within the normal range (i.e. ≥0.5 mV in the right atrial and ≥1.5 mV in the right ventricular myocardium). How many patients actually had decreased values?

The weak correlation between cardiac involvement and the CTG-repeat size in the present and previous studies can be partially explained by the fact that the mutation is dynamic and differentially distributed in various tissues. Even within a tissue the CTG-repeat length can vary between different cells. The number of CTG-repeats may be different in lymphocytes and myocardiocytes and thus may pretend a correlation where there is none. Determination of the CTG-repeat length in cardiomyocytes would be more helpful in this respect.

How to explain the differences between the values of the atrium and the ventricle? From the presented data it appears that the right ventricle was less severely affected than the right atrium. How to explain?

Clarified shoud be the discrepancy between the statement “there is predominant involvement of the inter-atrial septum” and the finding that potential amplitudes were the highest along the inter-atrial septum.

Quick Response Code

Journal homepage:
http://www.easpublisher.com/easjms/

Article History
Received: 15.04.2019
Accepted: 30.04.2019
Published: 19.05.2019

Was the transformation of the myocardium into fibrous tissue confirmed by any other technique, like tissue Doppler echocardiography or cardiac MRI? Myocardial involvement has been repeatedly demonstrated in the hearts of MD1 patients (Finsterer, J. *et al.*, 2001).

From the presented data it can not be concluded that application of electro-anatomic mapping is an adequate tool to estimate the arrhythmogenic risk in MD1-patients. No results of a long-term follow-up, which would allow assessing the risk for fatal rhyhtm abnormalities, are provided.

Though mentioned as a heading, it is not clear from the presentation, which the clinical implications of these findings are.

Overall, electro-anatomic mapping actually lacks a clinical relevance. The reproducibility and inter-observer variability of the technique has to be determined before a final assessment of the results and its discussion is feasible.

REFERENCES

1. Russo, A. D., Pelargonio, G., Parisi, Q., Santamaria, M., Messano, L., Sanna, T., ... & Giglio, V. (2006). Widespread electroanatomic alterations of right cardiac chambers in patients with myotonic dystrophy type 1. *Journal of cardiovascular electrophysiology*, *17*(1), 34-40.
2. Finsterer, J., Stöllberger, C., Wegmann, R., Jarius, C., & Janssen, B. (2001). Left ventricular hypertrabeculation in myotonic dystrophy type 1. *Herz*, *26*(4), 287-290.
3. Pelargonio, G., Russo, A. D., Sanna, T., De Martino, G., & Bellocci, F. (2002). Myotonic dystrophy and the heart. *Heart*, *88*(6), 665-670.
4. Finsterer, J., Stollberger, C., Blazek, G., & Spahits, E. (2001). Cardiac involvement in myotonic dystrophy, Becker muscular dystrophy and mitochondrial myopathy: a five-year follow-up. Can J Cardiol, 17, 1061-9.