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Brugada Syndrome; Preventive Screening Measures to Decrease Associated Cardiac Deaths

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Abstract

- Brugada syndrome is a sodium channel deficiency in the myocardium that can cause arrhythmias and sudden cardiac death. The myocardium deficient sodium channels can be inherited or affected by environment factors such as fever or medications.
- Screening for this syndrome can be challenging because many times the first symptom of Brugada syndrome is cardiac arrest. Though incidents of sudden cardiac deaths are low, sudden cardiac death can impact families and communities.
- In Italy, screening ECG's is claimed to have reduced sudden cardiac events by 85% (Maron, 2014). Preventing sudden cardiac death in these patients through sensible, reasonable preventive practices such as screening electrocardiograms and patient education is the goal.
- Performing 12 lead electrocardiograms (ECGs) in a screening nature and pairing it with medical history and physical exam can help increase discovery of Brugada syndrome and decrease associated cardiac events.
- The American Heart Association and the American College of Cardiology currently do not recommend screening ECGs. The International Olympic Committee and the European Society of Cardiology recommend screening ECGs for athletes.
- The search was limited to articles that were published within the last 10 years. Databases searched included Pubmed, CINAHL, and SPORTDiscus. Several articles were found to be of significance to the topic.
- **Key terms:** Brugada syndrome, sudden cardiac death, risk stratification, and type 1 ECG changes

Introduction

Brugada syndrome is a sodium channel deficiency in the myocardium that can cause ECG changes and sudden cardiac death. The myocardium deficient sodium channels can be inherited or affected by environment factors such as fever or medications. Screening for this syndrome can be challenging because many times the first symptom of Brugada syndrome is cardiac arrest. Though incidents of sudden cardiac deaths are low, sudden cardiac death can impact families and communities. Preventing sudden cardiac death in these patients through sensible, reasonable preventive practices is the goal.

Statement of the Problem

Brugada syndrome's worse and sometimes only symptom is sudden cardiac arrest. Cardiac arrest is often deadly and for those that survive can have lifelong disabilities. Incidents of events are low, however, one event can have a profound impact on families and communities. The goal is to find screening tools and techniques that can help determine an individual's risk.

Research Question

- Does physical exam and personal medical history provide an accurate risk assessment to prevent sudden cardiac death in Brugada syndrome compared to using electrocardiogram alone? Is routine ECG cost effective?

Literature Review

- It is estimated that four percent of all sudden cardiac deaths are related to Brugada syndrome with 20% of those sudden cardiac deaths having no structural heart abnormalities (Okamura,2015).
- There seems to be a male and Asian decent dominance. Dynamed (Dynamed, 2016) has a reported prevalence in Asia is about 1 per 1000 people. Prevalence of Brugada syndrome (BrS) in the general population is estimated to be 5/10,000 people and may be lower in Europe and the United States.
- Clinical history of familial sudden cardiac arrest has traditionally been used to identify risk in patients. Studies suggest that family history alone in those that have Brugada syndrome is not a reliable predictor of sudden cardiac arrest (Rivard,2016).
- Increased risk of sudden cardiac arrest can be associated with a group of symptoms. They are type 1 ECG changes associated with syncope and reproducible upon electrophysiology studies. This mostly affects the male gender and younger than 45 years of age.

Discussion

- Patients diagnosed with Brugada syndrome have a low relative risk of sudden cardiac death when that is the single criteria. When syncope is added with type 1 ECG changes that risk increases.
- Our screening practices of asking about family history stands to be an ineffective tool. A meta-analysis review of 27 studies looking at cardiac events in patients with suspected Brugada syndrome was performed.
- One area that helped identify risk was subjects that had Brugada syndrome and had either an aborted cardiac arrest or syncope were at 4.97 fold increase risk of sudden cardiac death. Those subjects that had Brugada syndrome with type 1 ECG changes had 2.78 fold increase risk of sudden cardiac death (Wu, 2016). An area that did not show significance was family history of the disease, genetic mutation, or being male.
- Standard ECG screening has been performed in other countries. In Italy, screening ECG's is claimed to have reduced sudden cardiac events by 85% (Maron,2014). Performing ECG screening has been met by some resistance in the United States.
- Another interesting note was that most providers in another survey also did not cover all the American Heart Association recommended 12 points of physical exam and medical history (Patel,2016).
- The European Society of Cardiology and the International Olympic Committee recommend ECG screening for athletes. The screening is viewed to be justifiable from an ethical, legal, and medical bases. There should be another step to not just limit ECG screening to athletes.

Table 2 Patients' characteristics according to LTA events during follow-up and predictive factors for LTA in univariate analysis (N ¼ 106)

Characteristic	LTA event (n ¼ 10)	No LTA event (n ¼ 96)	HR (95% CI)	P
Sex: male	6 (60)	52 (54)	0.79 (0.22–2.81)	.72
Age at diagnosis (y)	6.5 ± 7.0	11.6 ± 5.4	0.86 (0.76–0.96)	.009
Age at diagnosis >15 y	7 (70)	60 (62)	0.74 (0.19–2.89)	.66
Follow-up duration (mo)	42 (6–73)	55 (15–103)	—	—
Symptoms at diagnosis	7 (70)	14 (15)	9.65 (2.481–37.49)	.001
Spontaneous type 1 ECG pattern	8 (80)	28 (29)	9.1 (1.9–42.96)	.005
Family history of SCD	3 (30)	43 (45)	2.12 (0.55–8.21)	.28
ICD	5 (50)	17 (18)	3.85 (1.11–13.36)	.003
SCN5A mutations	9 (90)	49 (51)	—	.08*

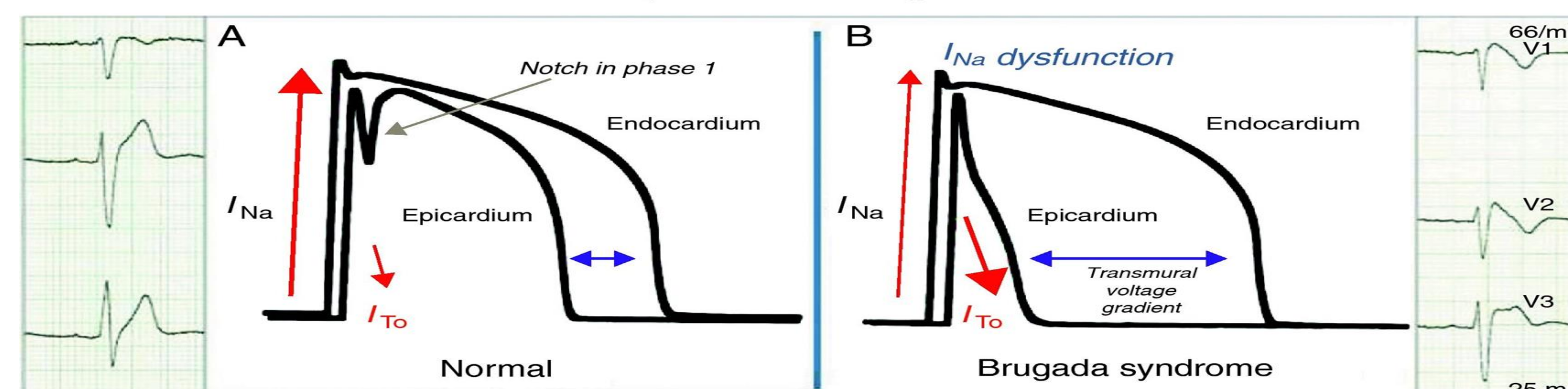
Values are presented as mean ± SD, as median (1st–3rd quartile), as n (%), or as otherwise indicated. CI ¼ confidence interval; ECG ¼ electrocardiographic; HR ¼ hazard ratio; ICD ¼ implantable cardioverter-defibrillator; LTA ¼ life-threatening arrhythmias; SCD ¼ sudden cardiac death.

*P value calculated using the log-rank test.

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Action potential in Brugada ECG



Applicability to Clinical Practice

Performing a 12 lead ECG is fairly inexpensive. It is anticipated that performing 12 lead ECG's more routinely and pairing it with medical history and physical exam can aid in helping diagnosis Brugada syndrome. The diagnosis of Brugada Syndrome can help a provider with patient education. There are risk stratification tools, however they require further research. Those with the syndrome can be given education in regards to further evaluation and treatment. That education is geared towards helping the patient understand their risk and how steps such as CPR and AED education can improve survivability should they experience a cardiac arrest event.

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