155- Epileptic seizures and Brugada syndrome in Tunisian patients

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INTRODUCTION

Patients having Brugada Syndrome (BS), a rare inherited channelopathy, with an increased risk of arrhythmia, syncope and sudden cardiac death, may present seizures as uncommon clinical manifestations.

Patients presenting with convulsions are sometimes incorrectly treated for epilepsy, as these symptoms may be manifestations of underlying cardiac disease.

OBJECTIVES

The aim of this study was to report seizures phenotypes in BS patients who presented to our genetic counselling for SCN5a gene molecular exploration..

METHODS

We selected through our genetic counselling reports of 2014, BS patients.

We collected information about seizures.

1. Synonyms:

- Pokkuri Death Syndrome
- SUNDS: Sudden Unexpected Nocturnal Death Syndrome

2. Na channelopathy:

Familial clustering, autosomal dominant inheritance

3. Manifestations:

Syncope, VT, VF, sudden death at sleep

4. Treatment:

Brugada Syndrome

.Na dysfunction

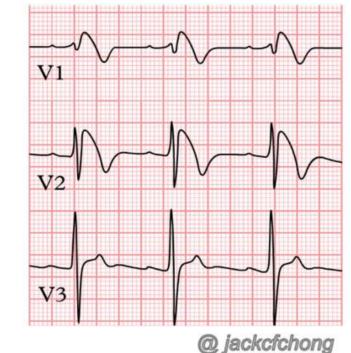
Epicardium

Transmural Voltage Gradient

 ICD (implantable cardioverterdefibrillator), quinidine

5. Diagnostic ECG

(coved type STE & TWI in V1~3)



Coved type ST-segment in V1-V2

Loss of function of sodium channels

Normal

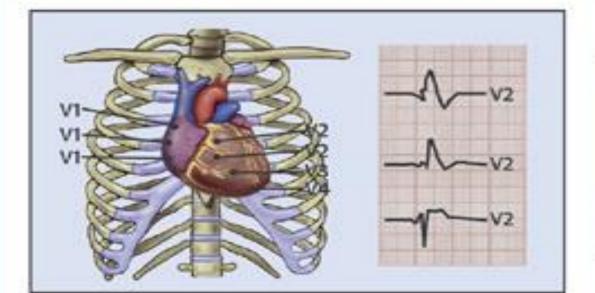
Notch in Phase 1

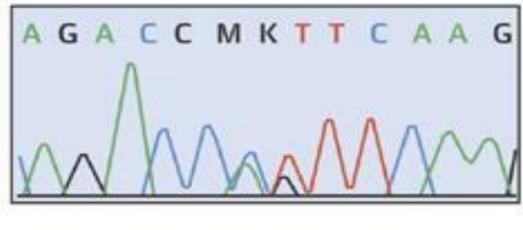
ICD is standard therapy, epicardial radiofrequency ablation a promising one

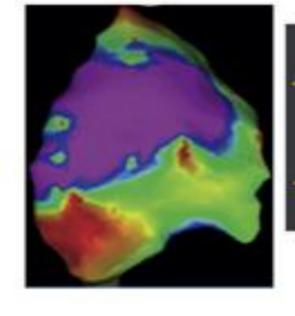
Diagnosis

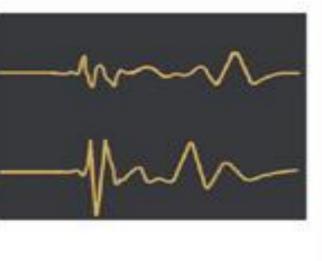
Pathophysiology

Management









Brugada, J. et al. J Am Coll Cardiol. 2018;72(9):1046-59. (1)

RESULTS

Five probands/families with BS were included.

Only one patient presented a generalized convulsive crisis.

He was a 24-year-old man from Sfax town, born from a consanguineous couple without any family history of sudden cardiac death.

He was diagnosed at the age of 20 years when he visited the emergency due to nocturne tonic-clonic generalized seizures.

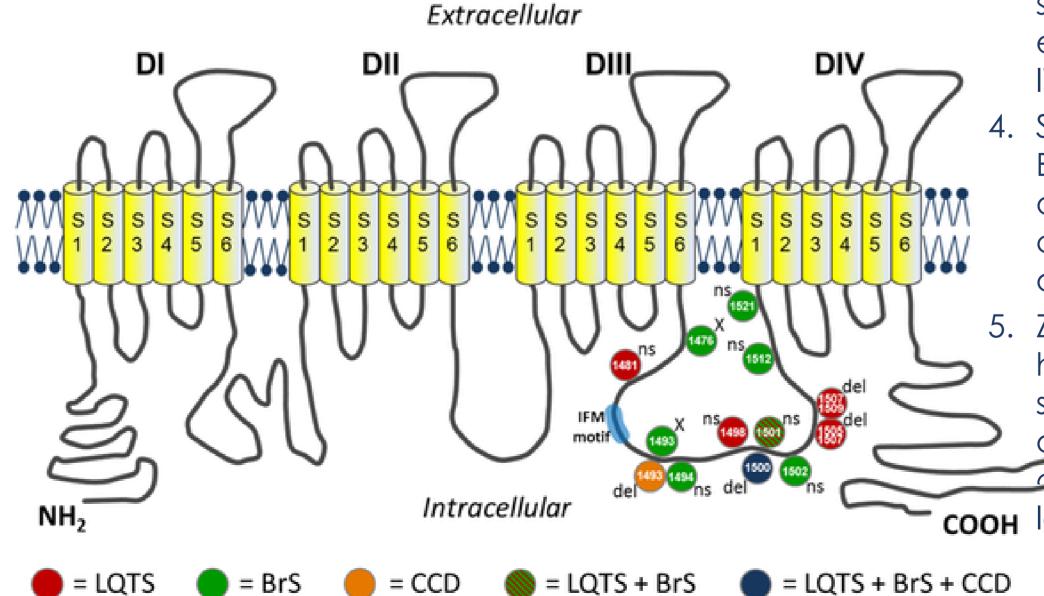
The patient had no medical history.

Physical examination, biological assessment, chest X-ray and electroencephalogram were normal.

However, the electrocardiogram patterns were compatible with BS type 1.

No SCN5a mutation was revealed.

SCN5a mutations in BS (5)



CONCLUSION

BS patients may experience typical symptoms such as ventricular arrhythmias and syncope or may be asymptomatic.

Symptoms that are more atypical may reveal the disease such as sudden nocturnal awakening, occasional nocturnal enuresis, nocturnal convulsions or nocturnal agonal breathing.

This study highlights BS presented with epileptic seizures, that must be considered to prevent sudden cardiac death.

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