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EDITORIAL

# Brugada syndrome, early repolarization syndrome/j-wave syndromes, and a subtype of idiopathic ventricular fibrillation: microstructural cardiomyopathies

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Open acess

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## Abstract

Brugada Syndrome is an inherited cardiac channelopathy with a high incidence of ventricular fibrillation and sudden cardiac death in patients with structurally normal hearts. Diagnosis is based on a characteristic electrocardiographic pattern (coved type ST-segment elevation ≥2 mm followed by a negative T-wave in ≥1 in the right precordial leads V1-V2) combined with an absence of gross structural abnormalities and several other criteria. The cornerstone of BrS diagnosis and definition, is its characteristic ECG pattern that can be present spontaneously or unmasked by drugs. This entity was described by the Brugada brothers in 1992 and belongs to a group of diseases known as inherited primary arrhythmia syndromes. The prevalence varies among regions and ethnicities, affecting mostly males. Despite several genes identified, SCN5A seems to be the most affected gene related BrS (≈ 30% of patients). The current main therapy is an implantable cardioverter-defibrillator, but radiofrequency catheter ablation has been recently reported as an effective new treatment.

**Keywords:** Brugada Syndrome, sudden cardiac death.

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The Brugada Syndrome (BrS): is a hereditary clinical-electrocardiographic arrhythmic entity with a low prevalence worldwide (0.5 per 1,000 or 5 to 20 per 10,000 individuals), however, endemic in Southeast Asia (prevalence of 3.7 per 1,000). BrS has male/female ratio of 9:1 in Southeast Asia and 3:1 among Caucasians. Males are more often symptomatic than females, probably by the influence of sex hormones on cardiac arrhythmias and/or ion channels, and a different distribution of ion channels across the heart in males versus females.

The BrS is caused by alterations in the structure (microstructural cardiomyopathy) and function of certain cardiac ion channels and reduced expression of Connexin 43 (Cx43) predominantly in the Right Ventricular Outflow Tract (RVOT) causing electromechanical abnormalities. The reduced and heterogeneous expression of Cx43 produces functionally significant electrophysiological heterogeneity in the ventricular wall and may promote transmural dispersion of repolarization. BrS was considered an autosomal dominant mendelian entity in ≈ 25% of cases or sporadic. It is currently thought that BrS most likely is an oligogenic disorder, rather than a Mendelian condition, affecting several loci, and influenced by environmental factors.

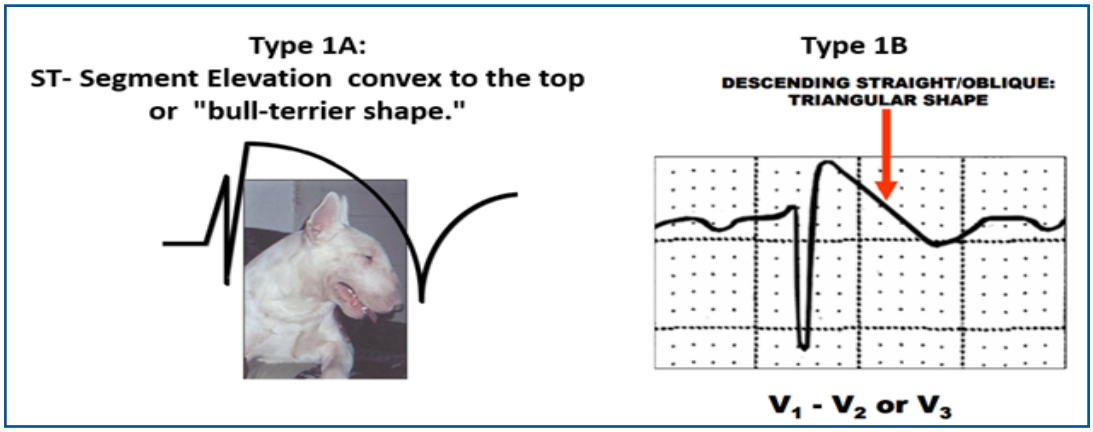
A family history of SCD at <45 years old, documented polymorphic ventricular tachycardia (PVT)/ ventricular fibrillation (VF), increased risk of syncope during sleep, and or nocturnal agonal respiration at the early morning hours. (a differential diagnosis is required with the other causes of syncope, such as vasovagal syncope.) Also, large meals and alcohol consumption can trigger arrhythmic events.

BrS is frequently associated with the following complains: palpitations, precordial pain, seizures, nocturnal agonal respiration, Pre-syncope and/or Sudden Cardiac Death (SCD) secondary to PVT/VF, unexplained SCA or documented PVT/VF at rest and paroxysmal atrial fibrillation (AF) tendency in the absence of macroscopic or apparent structural heart disease, electrolyte disturbance, use of certain drugs or coronary heart disease(CHD) and/ or fever.

Predominantly, manifestations occur between 30 and 50 years of age[1]

**Table 1:** ST-T abnormalities in the different types of Brugada syndrome[1]

| | Type I | Type II | Type III |
|---|---|---|---|
| J-Wave amplitude | ≥ 2mm V1-V3 | ≥ 2mm V1-V3 | ≥ 2mm V1-V3 |
| ST-Segment configuration | Coved type “convex to the top” or rectilinear oblique descendent: 1A and 1 B | Saddleback | Saddleback |
| ST- Segment (terminal portion) | Gradually descending | Elevated ≥ 1mm | Elevated < 1mm |
| T wave polarity | Negative in ≥1 of the right precordial leads | Positive or biphasis | Positive |




Other causes of ST segment elevation in right precordial leads[2].

1.Arrhythmogenic Cardiomyopathy (ACM) refers to an arrhythmogenic cardiomyopathy not secondary to ischemic, hypertensive, or valvar heart disease. In other words, is a cardiomyopathy characterized by substitution of the ventricular myocardium by fibrous or fibrofatty scar tissue, predisposing to malignant ventricular arrhythmias and sudden cardiac death (SCD). The entity, affects the RV, the LV or both[3].

Arrhythmogenic left ventricular cardiomyopathy; Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)[4].

2.Global Early Repolarization Syndrome or type 3 ERS. An early repolarization pattern (ERP) is categorized as Type 3, when it is registered in inferior, lateral and anterior wall. This variant carry the highest risk of malignant ventricular arrhythmia with electrical storm.

3.Highly trained young subject and which is, in most cases, a benign condition.

4.Brugada phenocopies Type 1 BrEP has a coved ST segment elevation ≥ 2 mm followed by negative T wave and no isoelectric separation of T wave[5].

5.Acute pulmonary embolism in patient presenting with recurrent syncope[6].

6.Acute myocarditis[7].
7.Takotsubo cardiomyopathy[8].
8.Athlete's heart[9].
9.Left ventricular aneurysm[10].
10. Anteroseptal Acute myocardial ischemia or infarction[11].
11. Mechanical compression of the right ventricle: pectus excavatum[12].
12. Right bundle branch block[13].
13. LBBB[14].
14. Hyperkalemia[15].
15. Severe hypercalcemia[16].

Although the BrS diagnosis necessarily requires the presence of pattern 1 on the ECG, the Shangai score has diagnosis utility (Table 1). Its attributes a specific score to each sign and symptom, from the sum of which a diagnosis can be hypothesized as probable/certain (score ≥ 3.5), possible (score 2–3) or non-diagnostic (<2)[17].

Diagnostic criteria: probable/definite ≥ 3.5 points, possible 2–3 points, nondiagnostic < 2 points.

**Table 2:** Shangai Score in The Brugada Syndrome[17].

| Shangai Score | | |
|---|---|---|
| Fever induced type 1 ECG Brugada pattern | | 3 |
| Type 2-3 ECG that converts to type 1 with provocative test | | 2 |
| Clinical history | | |
| A) | Unexplained SCA or documented VF/polymorphic VT | 3 |
| B) | Nocturnal agonal respirations | 2 |
| C) | Suspected arrhythmic syncope “Fainting” or passing out. | 2 |
| D) | Syncope of unclear etiology | 1 |
| E) | AF/flutter age < 30 years without clear etiology | 0.5 |
| Family history | | |
| A) | First- or second-degree relative with definite BrS | 2 |
| B) | Suspicious SCD(fever, nocturnal, Brugada-aggravating drug) in a first- or second-degree relative | 1 |
| C) | Unexplained SCD age <45 years in first- or second-degree relative with negative autopsy | 0,5 |
| Genetic test | | |
| (A) | Probable pathogenic mutation in BrS susceptibility gene | 0,5 |

**Figure 1:** Brugada Syndrome Candidate Genes[18].

| **Brugada Syndrome Candidate Genes** | | |
|---|---|---|
| ABCC9 | KCNE2 | SCN2B |
| ACTC1 | KCNE3 | SCN3B |
| AKAP9 | KCNE5/KCNE1L | SCN4B |
| ANK2 | KCNH2 | SCN5A |
| CACNA1C | KCNJ2 | SCN10A |
| CACNA2D1 | KCNJ5 | SEMA3A |
| CACNB2 | KCNJ8 | SNTA1 |
| CASQ2 | KCNQ1 | TMEM43 |
| CAV3 | RANGFR / MOG1 | TNNI3 |
| DSC2 | MYBPC3 | TNNT2 |
| DSG2 | MYH7 | TPM1 |
| DSP | MYL2 | TRPM4 |
| FLNC | MYL3 | LMNA |
| GPD1L | PKP2 | PLN |
| HCN4 | PLN | CBL |
| JUP | NOS1AP | tRNA-Ala |
| KCND3 | RYR2 | tRNA-Gln |
| KCNE1 | SCN1B | tRNA-Met |

I. Genes that encoding Na+ channels: SCN5A (sodium voltage-gated channel alpha subunit 5), SCN10A sodium voltage-gated channel alpha subunit 10, SCN1B Sodium channel subunit beta-1, SCN2B Sodium Voltage-Gated Channel Beta Subunit 2, and SCN3B Sodium Voltage-Gated Channel Beta Subunit 3,

II. Genes that encoding K+ channels HCN4, KCND2, KCND3, KCNE3, KCNE5, KCNH2, and KCNJ8. Observation The human genome contains ≈ 80 K+ channel genes of which 40 genes encode voltage-gated K+ channel pore-forming subunits that fall into 12 subfamilies.

III. Genes that encoding Ca++ channels CACNA1C, CACNA2D1, CACNB2, PLN, and TRPM4[18, 19, 20].

IV. BrS with sarcomeric mutations that modified Ca++ signaling [21-25].

V. Co-expression of ion channels in the heart and brain, leading to cardiac arrhythmia and epilepsy[26,27].

Brugada Syndrome is an inherited cardiac channelopathy with a high incidence of ventricular fibrillation and sudden cardiac death in patients with structurally normal hearts. Diagnosis is based on a characteristic electrocardiographic pattern (coved type ST-segment elevation ≥2 mm followed by a negative T-wave in ≥1 in the right precordial leads V1-V2) combined with an absence of gross structural abnormalities and several other criteria.

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**Resumo**

A Síndrome de Brugada é uma canalopatia cardíaca hereditária com alta incidência de fibrilação ventricular e morte súbita cardíaca em pacientes com corações estruturalmente normais. O diagnóstico é baseado em um padrão eletrocardiográfico característico (elevação do segmento ST tipo côncava ≥2 mm seguido por uma onda T negativa em ≥1 nas derivações precordiais direitas V1-V2) combinado com ausência de anormalidades estruturais macroscópicas e vários outros critérios. A pedra angular do diagnóstico e definição da SBr é o seu padrão característico de ECG, que pode estar presente espontaneamente ou ser desmascarado por medicamentos. Esta entidade foi descrita pelos irmãos Brugada em 1992 e pertence a um grupo de doenças conhecidas como síndromes de arritmias primárias hereditárias. A prevalência varia entre regiões e etnias, afetando principalmente homens. Apesar de vários genes identificados, o SCN5A parece ser o gene relacionado à SBr mais afetado (≈ 30% dos pacientes). A terapia principal atual é um cardioversor-desfibrilador implantável, mas a ablação por cateter de radiofrequência foi recentemente relatada como um novo tratamento eficaz.

**Palavras-chave:** Síndrome de Brugada, morte súbita cardíaca.