

# J-Wave syndromes expert consensus conference report: Emerging concepts and gaps in knowledge

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## Preamble

The J-wave syndromes (JWSs), consisting of the Brugada syndrome (BrS) and early repolarization syndrome (ERS), have captured the interest of the cardiology community over the past 2 decades following the identification of BrS as a new clinical entity by Pedro and Josep Brugada in 1992.<sup>1</sup> The clinical impact of ERS was not fully appreciated until 2008.<sup>2–4</sup> Consensus conferences dedicated to BrS were held in 2000 and 2004,<sup>5,6</sup> but a consensus conference specifically focused on ERS has not previously been convened other than that dealing with terminology, and guidelines for both syndromes were last considered in 2013.<sup>7</sup> A great deal of new information has emerged since. The present forum was organized to evaluate new information and highlight emerging concepts with respect to differential diagnosis, prognosis, cellular and ionic mechanisms, and approaches to therapy of the JWSs. Leading experts, including members of the Heart Rhythm Society (HRS), the European Heart Rhythm Association (EHRA), and the Asian-Pacific Heart Rhythm Society (APHRS), met in Shanghai, China, in April 2015. The Task Force was charged with a review of emerging concepts and assessment of new evidence for or against particular diagnostic procedures and treatments. Every effort was made to avoid any actual, potential, or perceived conflict of interest that might arise as a result of outside relationships or personal interest. This consensus report is intended to assist health care providers in clinical decision-making. The ultimate judgment regarding care of a particular patient, however, must be made by the health care provider based on all of the facts and circumstances presented by the patient.

Members of this Task Force were selected to represent professionals involved with the medical care of patients with the JWSs, as well as those involved in research into the mechanisms underlying these syndromes. These selected experts in the field undertook a comprehensive review of the literature. Critical evaluation of methods of diagnosis, risk stratification, approaches to therapy, and mechanistic insights was performed, including assessment of the risk-to-benefit ratio. The level of evidence and the strength of the recommendation of particular management options were weighed and graded. Recommendations with class designations are taken from HRS, EHRA, APHRS, and/or European Society of Cardiology (ESC) consensus statements or guidelines.<sup>8,9</sup> Recommendations without class designations are derived from unanimous consensus of the authors. The consensus recommendations in this document use the commonly used Class I, IIa, IIb, and III classifications and the corresponding language: “is recommended” for a Class I consensus recommendation; “can be useful” or “is

reasonable” for a Class IIa consensus recommendation; “may be considered” for a Class IIb consensus recommendation; and “is not recommended” for a Class III consensus recommendation.

## Introduction

The appearance of prominent J waves in the electrocardiogram (ECG) have long been reported in cases of hypothermia<sup>10–12</sup> and hypercalcemia.<sup>13,14</sup> More recently, accentuation of the J wave has been associated with life-threatening ventricular arrhythmias.<sup>15</sup> Under these circumstances, the accentuated J wave typically may be so broad and tall as to appear as an ST-segment elevation, as in cases of BrS. In humans, the normal J wave often appears as a J-point elevation, with part of the J wave buried inside the QRS. An early repolarization pattern (ERP) in the ECG, consisting of a distinct J-wave or J-point elevation, or a notch or slur of the terminal part of the QRS with and without an ST-segment elevation, has traditionally been viewed as benign.<sup>16,17</sup> The benign nature of an ERP was challenged in 2000<sup>18</sup> based on experimental data showing that this ECG manifestation predisposes to the development of polymorphic ventricular tachycardia (VT) and ventricular fibrillation (VF) in coronary-perfused wedge preparations.<sup>15,18–20</sup> Validation of this hypothesis was provided 8 years later by Haissaguerre et al,<sup>2</sup> Nam et al,<sup>3</sup> and Rosso et al.<sup>4</sup> These seminal studies together with numerous additional case-control and population-based studies have provided clinical evidence for an increased risk for development of life-threatening arrhythmic events and sudden cardiac death (SCD) among patients presenting with an ERP, particularly in the inferior and inferolateral leads. The lack of agreement regarding the terminology relative to early repolarization (ER) has led to a great deal of confusion and inconsistency in reporting.<sup>21–23</sup> A recent expert consensus report that focused on the terminology of ER recommends that the peak of an end QRS notch and/or the onset of an end QRS slur be designated as J<sub>p</sub> and that J<sub>p</sub> should exceed 0.1 mV in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG for ER to be present.<sup>24</sup> It was further recommended that the start of the end QRS notch or J wave be designated as J<sub>o</sub> and the termination as J<sub>t</sub>.

ERS and BrS are thought to represent 2 manifestations of the JWSs. Both syndromes are associated with vulnerability to development of polymorphic VT and VF leading to SCD<sup>1–3,15</sup> in young adults with no apparent structural heart disease and occasionally to sudden infant death syndrome.<sup>25–27</sup> The region generally most affected in BrS is the anterior right ventricular outflow tract (RVOT); in ERS, it is the inferior region of the left ventricle (LV).<sup>2,4,28–32</sup> As a consequence,

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