

Ebstein's anomaly with Brugada-like electrocardiogram pattern: A critical view

Elisabeth Kaiser, Carlos Alberto Pastore, Paulo Jorge Moffa

Heart Institute (InCor), Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo, Brazil

Brugada syndrome (BS) is a chanelopathy, with no apparent heart disease, 'coved' or 'saddle-back' type ST-segment elevation in the right precordial leads of electrocardiogram (ECG) and propensity for life-threatening ventricular arrhythmias.

Recently, a variant (atypical) type of BS has been reported, with downsloping ST-segment elevation in the inferior leads, sometimes associated with myocardial infarct, cocaine abuse, mediastinal tumor, hyperkalemia or hypothermia. Some authors have reported this 'atypical variant' with early repolarization aspect, that disappears with exercise. We present a case of non-operated Ebstein's anomaly (Fig. 1) with left posterior fascicular block (LPFB) concomitant with RBBB, responsible for the variant (atypical) type of BS.

LPFB is almost invariably associated with RBBB. The initial QRS forces due to LPFB, caused by activation of the left ventricular anterolateral wall in superior and leftward direction, originate Q-waves in D2, D3 and aVF. The middle forces, also due to

LPFB, are caused by delayed activation of the left ventricular posteroinferior wall. The terminal forces are caused by RBBB. These two main forces have a similar direction, resulting in increased R voltage in inferior leads. The higher the QRS voltage in D2 and D3, the greater the likelihood that it is due to RBBB with LPFB. RBBB with right ventricular hypertrophy does not produce such tall R-waves. Two significant features of RBBB with LPFB are its rarity and its frequent association with heart block.

In conclusions, several physical/biochemical alterations may induce persistent or transient bundle-branch/fascicular blocks and repolarization modifications, expressing the severity of the associated condition, but these events should not be misinterpreted as BS.

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Figure 1. Non-operated Ebstein's anomaly in a 23 year-old female patient. The electrocardiogram shows 1st degree atrioventricular block, right atrial hypertrophy and rightward QRS axis. Tall R-waves and downsloping ST-segment elevation are seen in inferior leads, due to left posterior fascicular block. The signs of right bundle-branch block are better available by the negative terminal forces in D1 and aVL, and R-wave in aVR.

Address for correspondence: Elisabeth Kaiser, Heart Institute (InCor), Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo, Av. Dr. Eneas de Carvalho Aguiar, 44. 054023-000 São Paulo, SP, Brazil, tel: +55 11 3069 5598, fax: +55 11 3062 0343, e-mail: elisabeth.kaiser@terra.com.br

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