

## Author's response

Firstly, we would like to express our appreciation of Dr. Riera's comments about our article entitled 'Ebstein's anomaly with Brugada-like pattern: a critical view'.

We agree with Dr. Riera as to the title, which we think would have been better if it had been 'Ebstein's anomaly with **atypical** Brugada-like pattern: a critical view', so as not to mislead readers regarding the ECG shown.

However, a step-by-step analysis of his comments leads us to disagree in the following regards:

1. Our colleague probably made a mistake with respect to his comments on "idiopathic ventricular fibrillation", which was never mentioned in our article.
2. Our keywords did not include "Brugada type I ECG pattern", but only "Brugada syndrome".
3. The similarity between the ECG shown and some ECGs reported as 'atypical Brugada' is a fact, and this makes up its own consistency. Therefore, the observation of a "clear inconsistency" does not hold.
4. The "audacious" diagnosis of LPFB may, and in fact must, be made, even in the presence of RVH or in patients with hearts in the vertical position (asthenic habitus), with less evident S waves in V5 and V6 than would be expected for such a degree of RBBB. This is because of the competing forces: LPFB essentially downward and those from RBBB to the right and upward. The concomitance of delays from both RBBB and LPFB involves a greater coincidence of initial forces from RBBB with the final forces from LPFB, thus giving rise to tall, monophasic R waves, with a slurred descending branch in the inferior leads and absence of S waves in those same leads. Different from an isolated RBBB, the association with LPFB yields smaller r waves in aVr. Although rare, this RBBB/LPFB association in Brazil has its primary cause in Chagas' cardiomyopathy, the secondary cause being coronary artery disease.
5. Nowadays, we know that certain genetic mutations affect not only the cardiac morphogenesis, but also a proper development of the conduction system. Sánchez-Quintana et al. obtained anatomopathological evidence that, in hearts with Ebstein's anomaly, the body of the AV node has a different shape and the His bundle is significantly shorter in length than in control subjects; it would occur as if "Ebstein's anomaly not only causes an abnormality in the development of the muscular AV septum, but also of the central fibrous body and its contents (His bundle)." Thus, in any type of congenital cardiopathy, diagnosis of lesions of the conduction system, independent of any atrial or ventricular hypertrophies existing, should be made as early as possible, since such lesions precede advanced AV blocks, and eventually lead to sudden cardiac death.
6. Most cardiology services worldwide do not have vectorcardiography equipment available. Therefore as much information as possible should be drawn from a basic and widely-known tool such as the ECG. Whenever diagnosis can be obtained from ECG data, VCG may be unnecessary.
7. We think that Dr. Riera's reference 2, of the work by Elizari MV et al., 2007, sounds somewhat odd. Some 40 years ago, investigators used to talk about hemiblocks. However it is now agreed that the His bundle left branch has three fascicles, thus damages affecting them are called 'fascicular blocks'.

### Recommended bibliography

1. Rosenbaum MB, Elizari MV, Lazzari TO, Halpern MS, Ryba D. QRS patterns heralding the development of complete heart block, with particular emphasis on right bundle branch block with left posterior hemiblock. In: Sandoe E, Flensted-Jensen E, Olesen KH eds. Symposium on Cardiac Arrhythmias, Elsinore, Denmark 1970.
2. Moffa PJ, Sanches PCR eds. O Eletrocardiograma e o Vetorcardiograma nos Bloqueios das Divisões do Ramo Esquerdo do Feixe de His. In: Eletrocardiograma Normal e Patológico. 7 Ed. São Paulo, Roca 2001: 413-461.
3. Sánchez-Quintana D, Picazo-Angelín B, Cabrera A, Murillo M, Angel Cabrera J. El triángulo de Koch em la anomalia de Ebstein. Rev Esp Cardiol, 2010; 63: 660-667.

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