A perfect storm: Wolf Parkinson White syndrome, Ebstein’s anomaly, biventricular non-compaction, and bicuspid aortic valve

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A 14-year-old African boy presented for routine clinical assessment to evaluate a systolic murmur.

The surface electrocardiogram showed sinus rhythm and the left postero-septal accessory pathway of Wolf Parkinson White syndrome (Panel A). Transthoracic echocardiography demonstrated features of Ebstein’s anomaly (Panel B and see Supplementary data online, Video S1): the downward (apical) displacement of septal leaflets of the tricuspid valve was 45 mm (28.8 mm/m²) with mild regurgitation at colour flow Doppler, leading to atrialization of the right ventricle (RV). There was extensive thickening and trabeculation of the apical half of the left ventricle (LV). Colour Doppler displayed a flow within the deep intertrabecular recesses. The LV was dilated with moderately depressed global systolic function. The RV appeared to be more heavily trabeculated than usual, but there was no dilatation. It had normal systolic function and the pulmonary artery systolic pressure was only 18 mmHg. The findings were suspected for non-compaction of the ventricular myocardium (Panel C): patient’s family refused magnetic resonance imaging making this diagnosis just alleged. Finally, we reported a congenital bicuspid aortic valve with mild-to-moderate eccentric regurgitation (Panel D and see Supplementary data online, Video S2) and normal dimension of the aortic root.

Medline search revealed few similar reported cases in the literatures. To our knowledge, the present case is the first reporting these concomitant pathological findings.

Conflict of interest: none declared.

Supplementary data are available at European Heart Journal — Cardiovascular Imaging online.

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