

The Triangle of Koch in the Setting of Ebstein's Malformation

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In this issue of *Revista Española de Cardiología* Sánchez-Quintana et al¹ provide another wonderful morphologic study of cardiac features of clinical significance. As we have come to expect from their previous contributions, their illustrations are quite superb, with unique scanning electron micrographs supplementing their histological images.

As they explain, the heart serving to provide the phenotype of the lesion now known as Ebstein's malformation came from a young adult. The young man was admitted on June 28, 1864, to the Department of Medicine in Breslau, now known as Wrocław, and belonging to Poland, but at that time located in the kingdom of Prussia. The patient, named Joseph Prescher, was deeply cyanosed, with multiple heart sounds emanating from his malformed heart. Sadly, he died after no more than 10 days in hospital. Wilhelm Ebstein, then a young attending physician, recorded the results of the autopsy, his description being superbly illustrated by Dr Weiss.² In the heart of Joseph Prescher, the septal leaflet of the deformed tricuspid was represented by a fibrous excrescence. This was described by Ebstein as being the size of the *drei groschen* coin then used in Prussia. This description has subsequently been interpreted, in various translations of the original publication, as corresponding to a quarter, a nickel, and a dime. These potential differences almost certainly reflect the effects of inflation, rather than changes in size of the abnormal excrescence.

The phenotypic findings described by Ebstein were the displacement of the hinge of the mural leaflet of the tricuspid valve, along with the remnants of the septal leaflet, away from the

atrioventricular junction. The illustrations of Dr Weiss also show clearly how the narrowed keyhole orifice of the abnormal valve is displaced so as to point towards the right ventricular infundibulum. Over time, these changes have usually been interpreted as representing "downward" displacement, but this is yet another indication of the unfortunate habit of morphologists, including myself, of removing the heart from the body, and describing it as though positioned on its apex. In reality, there is rotational displacement of the hinges of the abnormal right-sided valve in Ebstein's malformation, and it usually closes in bifoliate rather than trifoliate fashion.³ Fortunately, Sánchez-Quintana et al avoid the pitfall of inappropriate description, and rightly photograph their gross specimens in attitudinally appropriate orientation,⁴ albeit still illustrating their histological findings in "Valentine" fashion.

Sánchez-Quintana et al then show exquisitely how the anticipated normal position of the hinge of the septal leaflet of the tricuspid valve is represented in the abnormal hearts by a fibrous shelf. They are in error, however, when they suggest that the muscular atrioventricular septum is diminished in size in the setting of Ebstein's malformation. In reality, there is no true atrioventricular septum in the normal heart. The area that they emphasise as occupying a horizontal rather than a more vertical position is better described as the atrioventricular muscular sandwich.⁵ As can be seen in their Figure 1A,¹ a superior extension from the inferior atrioventricular groove interposes between the atrial and ventricular muscle masses in this area, reaching superiorly to the level of the central fibrous body. The artery to the atrioventricular node extends through this inferior pyramidal space, with the anatomy of this crucial area having previously been described by Sánchez-Quintana working with a group of collaborators that included myself.⁶ As can be seen from Figure 1 of the current work,¹ nonetheless, and when considered from the stance of pressures, then a muscular atrioventricular septum does exist in the setting of Ebstein's malformation. This area is bounded in the right ventricle by the fibrous crest on the one hand, and

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by the rotationally displaced hinge of the septal leaflet of the tricuspid valve on the other hand. Anatomically, of course, it is part of the muscular ventricular septum, since as described by Sánchez-Quintana et al, the atrioventricular junction is marked by the fibrous shelf. In physiologic terms, nonetheless, this part of the muscular septum can be considered atrioventricular, being interposed between the atrialised component of the right ventricular septum and the left ventricle. The area is well seen in the illustrations provided from 3 of the autopsied hearts.¹

Apart from this minor solecism, the account provided by Sánchez-Quintana et al is well constructed.¹ It should serve to elucidate the morphology of Koch's triangle for those seeking to ablate arrhythmias in the setting of Ebstein's malformation.

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