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Editorial

The surgical treatment of Ebstein's malformation

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THE MOST APPROPRIATE SURGICAL TREATMENT FOR Ebstein's malformation and, indeed, the very nature of the lesion itself, remain contentious issues. The long experience of the group from Hôpital Broussais has proved invaluable in directing surgeons towards the best techniques for conservative repair and in pointing anatomists towards the most appropriate description of the deranged valvar structure. This is exemplified by the advances made in the understanding of the left atrioventricular valve in hearts with deficient atrioventricular septation ("atrioventricular canal malformations"). It was the beautifully explicit diagrams of Carpentier[1] which, for me at least, first focused attention on the unequivocal trifoliate arrangement of this left atrioventricular valve, although the information required was also provided by Van Mierop and his colleagues in their earlier studies.[2,3] The impact of all these excellent investigations, however, was dissipated by continued use of the adjective "mitral" to describe the trifoliate valve, together with arguments which continued concerning the optimal surgical treatment of the zone of apposition between the edges of the two leaflets of the trifoliate valve which bridge the ventricular septum (the "commissure" or "cleft"). It is now becoming appreciated that it is better to avoid nominative terms for this structure and simply describe the zone of apposition between the leaflets.[4]

In the accompanying paper,[5] Chauvaud and his colleagues describe the continuing evaluation of their surgical management of Ebstein's malformation. At the same time, (for me, at any rate) they continue to clarify the morphology of this fascinating lesion. I had thought that I understood well the anomaly, having published several accounts of its structure which, over time, illustrated my own increasing appreciation of its morphology.[6-8] When preparing a chapter[9] from a presentation for a meeting also attended by both Chauvaud and Carpentier, however, elements of my continuing ignorance were highlighted by the comments of these skilled surgeons. Much of the impetus for my own chapter had itself been provided by an editorial comment by Becker[10] on another study with which I was involved, which had focused on the histologic make-up of the ventricular myocardium.[11] Having studied the initial commentary, I was unsure of the focus of Becker's dissatisfaction. Only after a Letter to the Editor[12] did it emerge that Becker[13] considered that the nomination "Ebstein's malformation," if used in isolation, should apply only to the index case, or else its very close cousins. In cases with more severe associated malformations, such as pulmonary atresia, Becker considers it necessary to add "of the tricuspid valve" so as to clarify any potential confusion. I presume from this response that Becker agrees that the essence of Ebstein's malformation of the tricuspid valve is the finding of some part of the proximal hinge point of the valvar leaflets within the inlet component of the right ventricle rather than at the atrioventricular junction.

Chauvaud and his colleagues[5] now emphasize that, within this abnormal anatomic arrangement, some features are more important surgically than others. Of these, the feature which I had tended to ignore was the reduced mobility of the anterosuperior leaflet, despite having emphasized the spectrum of abnormality of distal tethering of this leaflet from focal to linear attachment in one of our earlier studies.[7] I had also concentrated my attention on the mural as well as the septal leaflets without appreciating fully that the tongue of tissue joining septal and anterosuperior leaflets in severely malformed hearts is, in reality, the displaced mural leaflet. In those hearts with severe disease, the effect of the congenital derangement is to produce a circlet of valvar tissue not at the atrioventricular junction but at the junction of the inlet and apical trabecular components of the right ventricle (Figure 1). The precise arrangement of this valvar mechanism is not always evident to the anatomist studying previously prosected hearts, since the initial dissection of the heart can grossly distort the true morphology. Examination of two hearts from the carefully preserved collection of Dr. Leon Gerlis, however, exemplifies the anatomy as described by the surgeons in the operating room. These specimens also show how, with almost identical anatomy at the junction of inlet and apical trabecular compo-

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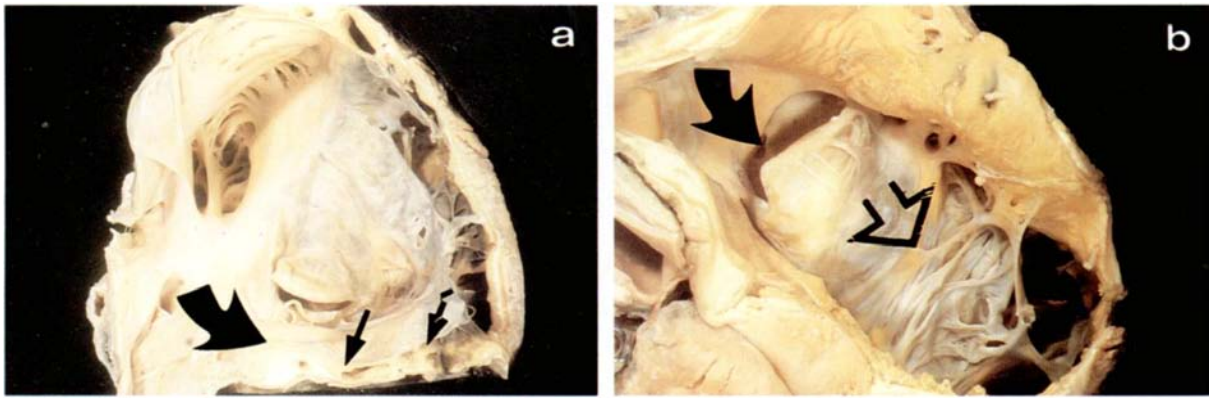

Figure 1. *This heart with severe Ebstein's malformation of the tricuspid valve is seen (a) from the inlet and (b) from the infundibular aspects. The inlet view shows how the septal and mural leaflets are displaced away from the atrioventricular junction (large arrow), with tethering of the mural leaflet (small arrows) to the parietal ventricular wall. The effective valvar orifice (large arrow) (b) points to the pulmonary valve because of the linear attachment of the leading edge of the anterosuperior leaflet (open arrow).*

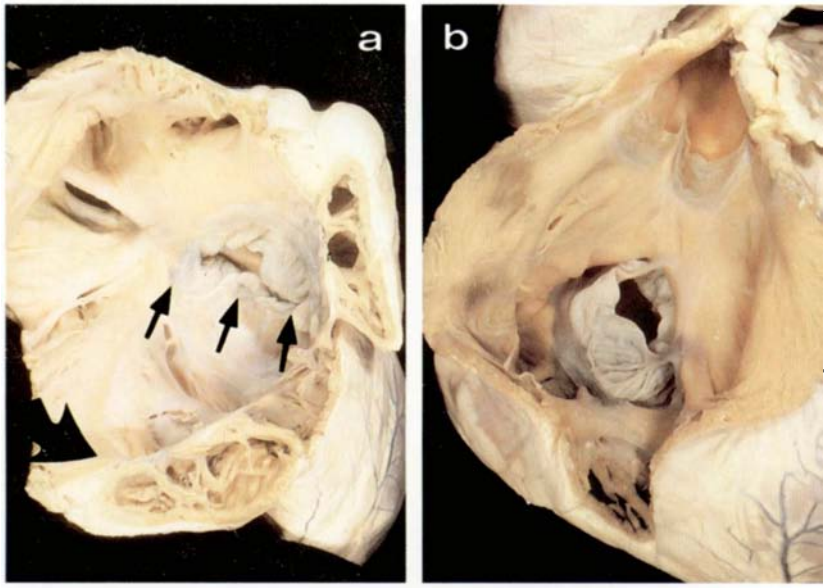

Figure 2. *An additional heart with Ebstein's malformation of the tricuspid valve viewed from the inlet (a) and the infundibular (b) aspects. In this heart, the mural and septal leaflets (small arrows) have their hinge point well away from the atrioventricular junction (curved arrow). Note the muscle in the relatively thin wall of the inlet component.*

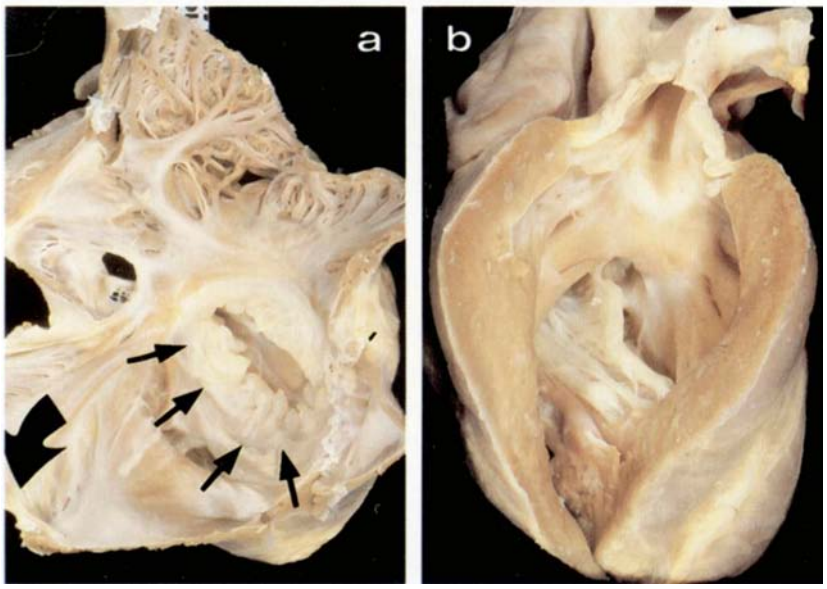

Figure 3. *In this specimen, comparable to Figure 2, there is extreme thinning of the wall of the atrialized inlet of the right ventricle (between curved and small arrows).*

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nents, there can be marked differences in the extent of muscular thinning and annular dilation of the inlet component of the right ventricle (Figures 2, 3). This emphasizes still further the extremes of the clinical spectrum produced by Ebstein's malformation of the tricuspid valve.

The surgical experience of the group from Hôpital Broussais, therefore, shows what can be achieved by those who understand fully the anatomy as presented to them in the operating room. It was surely an appreciation of this anatomy which led them to propose the longitudinal, as opposed to transverse, plication of the dilated inlet component of the right ventricle. As they indicate, it is this procedure which, at one stroke, restores the volume of the right ventricle and reduces the dilation of the atrioventricular junction. At the same time, nonetheless, one presumes that the liberated anterosuperior leaflet still coapts distally with the displaced septal leaflet, and the coaption occurs at the junction of the inlet with the apical trabecular component, since the inlet component of the right ventricle is effectively obliterated by the approach taken from the group at Hôpital Broussais. As Chauvaud and his colleagues also emphasize, their approach is not difficult to apply, since Quaegebeur and his colleagues[14] used the Broussais technique of longitudinal plication with equally good results.

Once again, therefore, the careful anatomic observations of Chauvaud, Carpentier and their colleagues have led to an increased understanding of the structure of congenitally malformed hearts. Such understanding, and application, of complex anatomy continues to be an essential part of the evolution of surgical techniques as we seek to achieve the magical zero mortality.

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