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Editorial Comment

When is the supravalvar mitral ring truly supravalvar?

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In the current issue of the Journal, Vaideeswar and colleagues[1] describe an amazing series of autopsied specimens with so-called "supravalvar mitral ring". As they point out, this lesion was one of the 4 malformations observed by Shone and his colleagues[2] when describing the constellation now known as the Shone syndrome. In the original description, it was the exception rather than the rule for all four malformations to be present at the same time. So it is with the series of hearts examined by the workers from Mumbai, with none of their patients possessing all 4 of the possible lesions. In their series, they also noted an association in some of their specimens with rheumatic disease, which is not surprising considering the ongoing prevalence of this destructive disease in their country. As far as I am aware, theirs is the largest series of autopsied hearts thus far described with this fascinating lesion. Despite the excellence of their descriptions, it remains the case that, for me at least, they have failed to clarify one of the ongoing problems with description of the entity.

This devolves on whether the stenosing ring is always "supravalvar". They conclude that the entity could simply be described as a supravalvar ring. If this is the case, then it should always be located within the left atrium, in other words on the left atrial aspect of the atrioventricular junction. In this respect, Vaideeswar and his associates[1] correctly emphasise the need to distinguish the lesions producing true supravalvar rings from the shelf that divides the left atrium in so-called "cor triatriatum sinister". One of their illustrations shows clearly that this is not difficult. As can be seen, in their specimens with the true supravalvar entity, the stenosing ridge is positioned within the left atrial muscular vestibule, being located on the atrial aspect of the hinge of the mitral valvar leaflets from the atrioventricular junction.

In only 9 of their 24 specimens, however, was the stenosing ring positioned within the muscular atrial vestibule of the mitral valve. In the remaining hearts, according to the authors, the stenosing ring was located in "annular" position. But, as was emphasized by Sullivan and his associates[3] in an important echocardiographic study, oftentimes the stenosing ring is located within the funnel created by the leaflets of the mitral valve, in other words on the ventricular aspect of the mitral valvar annulus. From the illustrations provided by Vaideeswar and associates,[1] it is difficult for the observer to be entirely sure of the location of their so-called "annular" lesions relative to the atrioventricular junction. Apart from this, it is also difficult, when seeking to establish relationship of the lesion on the aortic leaflet of the mitral valve, to be sure of the precise position of its hingeline, or "annulus". In at least one of their illustrations, their Figure 4, the lesion seems to be located on the ventricular aspect of the hingeline. In this specimen, the location of the lesion corresponds with the description as provided by Sullivan and colleagues[3] on the basis of their echocardiographic study.

It is a pity, therefore, that the workers from India did not provide a more precise account of the location of the stenosing ring relative to the atrioventricular junction when involving the mural leaflet of the mitral valve, or the hingeline when

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involving the aortic leaflet of the valve. This information would provide the much-needed anatomic proof concerning the true supravalvar location of the obstructing shelf. Even if located at the annulus of the valve, however, rather than within the funnel of the leaflets, the lesion should be described as valvar rather than supravalvar. This is because, as emphasized by Perloff and Roberts[4] in their superlative account, the annulus is an integral part of the mitral valvar complex. Lesions involving the annulus, therefore, should be considered to be valvar, rather than supravalvar. It could well be, therefore, that the stenosing ring obstructing flow across the morphologically left atrioventricular junction would better be divided into supravalvar and valvar categories. Vaisdeeswar and his colleagues would do us an even greater service if they returned to their specimens and clarified this ongoing conundrum.

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