Infarction of Papillary Muscles and Mitral Insufficiency Associated with Congenital Aortic Stenosis

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In 21 infants, each with symptomatic congenital aortic stenosis and left ventricular cavity of nearly normal size studied clinically, evidence for mitral insufficiency was frequently encountered. In five of the patients left ventriculography was done and, in each, mitral insufficiency was demonstrated. This background of experience led us to study, in particular, the mitral valve in specimens of heart from infants with congenital aortic stenosis.

This study led to the conclusion that there is a common association between congenital aortic stenosis, on one hand, and infarction of the papillary muscles of the mitral valve, with resulting mitral insufficiency, on the other. It is the purpose of this report to describe the pathological features of this combination of lesions as a means of emphasizing its occurrence.

Observations

Specimens of heart were available for study from 11 infants with congenital aortic stenosis and congestive cardiac failure who had died when 3 days to 6 months old. Four of the patients had died in the early postoperative period following an aortic valvotomy.

The following features were present in each case: The coronary arteries arose normally from the aorta. The aortic valve exhibited a form of severe congenital stenosis, and the left ventricular cavity was of about normal size while its wall was hypertrophied. None of the cases exhibited so-called hypoplastic left ventricle.

Among the 11 specimens, three forms of stenosis of the aortic valve were observed. In each of nine cases a unicommissural unicupid type of aortic valve was exhibited. In the tenth case, a bicuspid aortic valve was present, while in the eleventh the aortic valve was membranous with a central orifice. Endocardial fibroelastosis frequently coexisted. In eight of the 11 cases, the wall of the left ventricular cavity was covered by a thick, pale gray layer which on histological examination was composed of elastic and collagenous fibrous tissue.

In each case, atrophy was present in one or both papillary muscles of the mitral valve. When one papillary muscle was involved, it was usually the posteromedial one. The involved papillary muscles were thin, flattened, and darker than the adjacent myocardium (fig. 1). Histological examination revealed that in each case the anterolateral and posteromedial papillary muscles of the left ventricle were the sites of varying degrees of replacement of myocardial fibers by collagenous scar tissue (fig. 2). The posteromedial papillary muscle exhibited more extensive scarring than did the anterolateral one. In addition, in two instances, calcification was present in the posteromedial papillary muscle. In cases of severe degrees of scarring of the papillary muscles there were, as well, focal areas of scarring in the subendocardial region of the free wall of the left ventricle.

Secondary manifestations of mitral insufficiency in the form of left atrial enlargement

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together with "rolling" of the free edge of the anterior leaflet of the mitral valve were seen in six infants. No other lesions of the leaflets of the mitral valve were observed although the chordae appeared shorter than normal. A jet lesion was present in the left atrium in one case.

Comment

There is ample evidence that dysfunction of the papillary muscles may be responsible for incompetence of the mitral valve.\(^3\)

In symptomatic young infants with congenital aortic stenosis, the frequent association of mitral insufficiency appears to be related to the common finding of infarction of the papillary muscles of the left ventricle.

Noren and associates\(^4\) from our institutions reported the occurrence of infarction of the left ventricular papillary muscles in cases of anomalous origin of the left coronary artery from the pulmonary trunk. In that condition the lesions of the papillary muscles are like those in the current series with congenital aortic stenosis. In each, the common finding of mitral insufficiency is considered to result from involvement of the papillary muscles by a process of infarction.

In anomalous origin of the left coronary artery, the basis for myocardial ischemia sufficient to cause infarction may be readily explained as related in one way or another to the fact that one coronary artery arises from the pulmonary trunk.\(^5\)

Others\(^6-8\) have demonstrated the occurrence of ischemic myocardial necrosis in congenital aortic stenosis. Our studies confirm these observations and emphasize the particular tendency for the infarction to involve the left ventricular papillary muscles with the greater effect in the posteromedial papillary muscle.

In aortic stenosis it is probable that the altered hemodynamics characteristic of the valvular disease and perhaps left ventricular hypertrophy together represent the basis for myocardial ischemia. The greater susceptibility to infarction of the posterior papillary muscle is not immediately explained.

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**Figure 1**

*From infants with severe degrees of congenital aortic stenosis. (a and b). From a 4-week-old infant. (a). Left atrium and left ventricle. The anterolateral (A) and the posteroomedial (P) papillary muscles show atrophy. The endocardium of the left ventricle is thickened with fibroelastosis. (b). Close-up view of the posteroomedial papillary muscle (P). Atrophy of one segment of the muscle is evident. (c). From a 7-month-old infant. The posteroomedial papillary muscle (P) shows two segments: one is atrophic; the other segment is stout, long, and connected with the mitral valve through short chordae. (d). From a 4-day-old infant. The anterolateral (A) papillary muscle is atrophic while the posteroomedial papillary muscle (P) is normal.*

**Figure 2**

*Photomicrographs of papillary muscles in infants with congenital aortic stenosis. (a). From the 4-day-old infant whose heart is illustrated in figure 1 (d). The anterolateral papillary muscle shows focal scarring of relatively minor degree. Elastic tissue stain; \(\times 4\). (b). From a 3-month-old infant. The posteroomedial papillary muscle shows scarring alternating with intact muscle. The adjacent free wall of the left ventricle shows minimal scarring of the myocardium in the subendocardial region. Endocardial thickening. Elastic tissue stain; \(\times 4\). (c). From a 5-month-old infant. The posteroomedial papillary muscle shows extensive loss of myocardial fibers and replacement by scar tissue. In addition, there are multiple areas of calcification, the latter considered to represent calcification of infarcted myocardial bundles. H and E; \(\times 40\). (d). The posteroomedial papillary muscle from the 7-month-old infant whose heart is illustrated in figure 1 (c). There is extensive calcification. H and E; \(\times 40\).*

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Summary
Study of the papillary muscles in 11 infants dying in the first year of life with congenital aortic stenosis and normal coronary arteries demonstrated infarction of the left ventricular papillary muscles in each. Lesions were more extensive in the posteromedial than in the anterolateral papillary muscle.
Infarction of papillary muscles is considered the basis for the common occurrence of mitral insufficiency in symptomatic infants with congenital aortic stenosis. Congenital aortic stenosis, infarction of left ventricular papillary muscles, and mitral insufficiency are so interrelated as to constitute a syndrome.

References

Peacock on the Function of the Papillary Muscles:
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There is, perhaps, in the animal frame no more beautiful example of the adaptation of structure to the function to be accomplished than is afforded by the auriculo-ventricular valves. The insertion of the cords into the fleshy columns, instead of directly into the muscular walls, is apparently not to give greater power of resistance to the pressure of the blood during the systole, but to furnish a means of shortening the attachments of the curtains, when, with the contraction of the ventricle, the walls are more closely approximated. Were it not for this arrangement, the free fold of the mitral, for instance, would fall back towards the auricle during the systole, and the two curtains not being properly adjusted, the blood would flow into the auricular cavity. By the action of the muscular columns, however, the cords are drawn upon as the parietes of the ventricle approximate, and the curtains are kept in apposition and tightly stretched across the aperture, so as effectually to close it.—Quoted from Ian H. Porter: The Nineteenth-Century Physician and Cardiologist Thomas Bevill Peacock (1812-82). Med Hist 6: 246, 1962.
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