Congenital Aneurysms Adjacent to the Anuli of the Aortic and/or Mitral Valves*

Elliot Chesler, M.D.; Abdul S. Mitha, M.D.; and Jesse E. Edwards, M.D., F.C.C.P. (Hon)

The unusual occurrence of non-infected submitral and aortic aneurysms developing sequentially in an adult prompted us to review the pathogenesis of congenital aneurysms adjacent to the aortic and mitral valves. The findings support the suggestion that subvalvular aneurysms, like aortic sinus aneurysms, are a result of a congenital defect at the valve anulus. Submitral aneurysms occur only subjacent to the posterior leaflet.

Among left ventricular aneurysms, the subvalvular types are the least common. Such aneurysms have been thought to be a result of a congenital defect between the ventricular wall and the valvular anulii. These have been likened to congenital aortic sinus aneurysms where the accepted cause is a deficiency between the aortic media and the anulus of the aortic valve. This suggestion is supported by our encounter of a case in which an aortic sinus aneurysm developed three years after surgical repair of a submitral aneurysm (Fig 1 and 2). The combination of these rare anomalies prompted us to review the pathogenesis of congenital aneurysms related to the aortic and mitral valve anulii.

Subvalvular Aneurysms

Subaortic and submitral aneurysms have been collectively referred to as subvalvular aneurysms. They occur in two constant anatomic positions. Subaortic aneurysms occur under the intermediate portion of the left cusp of the aortic valve and submitral aneurysms under the posterior leaflet of the mitral valve. Submitral aneurysms are more common than subaortic aneurysms.

The clinical presentation is one of heart failure, angina pectoris, systemic embolism, or recurrent ventricular tachycardia. The murmur of aortic insufficiency may be present in the subaortic type and that of mitral insufficiency in the submitral type. Because submitral aneurysms are usually larger, the thoracic roentgenogram frequently reveals an abnormal contour. Subaortic aneurysms are smaller and contained within the cardiac silhouette and therefore are not usually visualized roentgenographically.

Whereas aortic sinus aneurysms may arise from any of the three sinuses, subaortic aneurysms occur only under the intermediate portion of the left aortic sinus. In the absence of rupture, the clinical presentation of these aneurysms results from valvular insufficiency or compression of the left coronary artery or of the conduction system.

Figure 1. Submitral aneurysm. Posterobasal view of left ventricle. A Lillehei-Kaster valve is in place in the mitral valve; another has been removed from the aortic position. A wedge-shaped defect representing the osium of a surgically excised aneurysm is occluded by an endothelialized Dacron patch under the posterior portion of the mitral anulus. LV = left ventricle.

*From the Cardiovascular Section, Veterans Administration Medical Center, Minneapolis; Wentworth Hospital, Durban, South Africa; and the Departments of Pathology, United Hospitals, St. Paul and the University of Minnesota, Minneapolis.

Supported by Public Health Service Research Grant 5 RO1 HL05694 from the National Heart, Lung and Blood Institute.

 Manuscript received January 20; revision accepted March 25.
Reprint requests: Dr. Jesse Edwards, United Hospitals, 333 North Smith, St. Paul, Minnesota 55102.
Figure 2. Aortic sinus aneurysm. Coronal section through the aorta, ventricular septum and right ventricle. A probe passes from a defect in the base of the right aortic sinus into a loculus of a large imperforate aneurysm within the ventricular septum. Ao = aorta; RV = right ventricle; VS = ventricular septum. Histologic study demonstrated discontinuity between the aortic media and the ventricular septum (same patient as in Fig 1).

Graphically. These aneurysms are frequently calcified, and the electrocardiogram may show evidence of myocardial infarction.

Aortic Sinus Aneurysms

These may arise from any of the three sinuses. Aneurysms of the posterior and right aortic sinuses present into and may rupture into the right atrium, right ventricle, or both, producing an aortocardiac fistula. Much less commonly, these aneurysms may produce the same clinical picture following an initial dissection into the ventricular septum before actually perforating. Rarely, aortic sinus aneurysms may burrow into the ventricular septum but remain intact; this variety has the potential to compress the conduction system and produce complete heart block (Fig 2 and 3). The intermediate portion of the left sinus is the only portion of the aortic root not related to a cardiac chamber, its immediate external relationship being the epicardium containing the left main coronary artery. Aneurysms of the

Figure 3. Diagrammatic representation of an imperforate aneurysm above the right aortic sinus burrowing into the ventricular septum and presenting into the right ventricle. In this and in succeeding illustrations, Ao = aorta; LC = left coronary artery; LV and RV = left and right ventricles, respectively; RC = right coronary artery; VS = ventricular septum.

Figure 4. Diagrammatic representation of an aortic sinus aneurysm above the aortic anulus in close proximity to the left coronary artery.
Congenital Aneurysms (Chester, Mitha, Edwards)

Pathogenesis of Aortic Sinus and Subvalvular Aneurysms

The pathogenesis of noninfected spontaneous subvalvular and aortic sinus aneurysms (so-called congenital aneurysms) appears to be similar. As demonstrated in Figure 2 and previously, an aortic sinus aneurysm is a consequence of a deficiency of the normal continuity between the aortic media and anulus. Similarly, subvalvular aneurysms demonstrate a lack of continuity between the myocardium and the anulus of the aortic or mitral valve (Fig 1).

In an aortic sinus aneurysm the wall is formed by aortic adventitia, whereas the wall of a subvalvular aneurysm is formed by thickened epicardium. Both types are, therefore, "false aneu-
Subvalvular aneurysms. The subvalvular aneurysms that have been described were universally located in those portions of the aortic or mitral valve anuli which are directly related to the epicardium and where there is a muscular-fibrous junction. Whereas defects above the aortic anulus are responsible for aortic sinus aneurysms (Fig 4), defects below the anulus are responsible for subaortic aneurysms (Fig 5). Defects below the anulus of the right and posterior sinuses are not uncommon and involve the muscular or membranous portions of the ventricular septum producing a ventricular septal defect. It is theoretically possible, however, that a partial defect in the muscular septum could produce non-fistulous types of subaortic aneurysms in these positions (Fig 6).

Because of the anatomy of the mitral anulus, congenital aneurysms occur only below the posterior leaflet. Two-thirds of the mitral anulus is related to the posterior leaflet which is attached to the myocardium of the left ventricle by anular tissue ("mitral ring"). The immediate external relationship of the mitral ring is the epicardium in the atrioventricular groove. A dehiscence of this muscular-fibrous union will result in a fibrous-walled, submitral aneurysm below the posterior leaflet (Fig 7). Morphologically identical aneurysms may occur in the same position following surgical replacement of the mitral valve.10 The remaining one-third of the mitral anulus is formed by the fibrous union between the aortic and mitral valves through the so-called "mitral-aortic intervalvular fibrosa." Perforation of the latter structure has been described only as a consequence of spread of infective endocarditis from the contiguous aortic and mitral valves. This results in a mycotic aneurysm situated in the epicardial wedge between the base of the aorta and the left atrium (Fig 8).11

The exact nature or cause of the defect in subvalvular aneurysms is uncertain. Virtually all the cases have been described in black people, but a subaortic aneurysm has been documented in a white female.5 The predilection for constant anatomic sites in the absence of evidence of coronary atherosclerosis, infection or trauma would favor a congenital cause. Subvalvular aneurysms have not hitherto been reported in association with another congenital cardiac anomaly. As far as we are aware, the concurrence of an aortic sinus and mitral subvalvular aneurysm is unique and lends support to the hypothesis that subvalvular aneurysms are also of congenital cause. While their pathogenesis appears to be similar, it is uncertain, however, whether the dehiscence between the anulus and related structures is a primary failure of union or a later spontaneous separation.

REFERENCES

3 Chesler E, Tucker RBK, Barlow JB. Subvalvular and apical left ventricular aneurysms in the Bantu as a source of systemic emboli. Circulation 1967; 35:1158-62
8 Fishein MC, Obma R, Roberts WC. Unruptured sinus of Valsalva aneurysm. Am J Cardiol 1975; 35:918-22