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## Pulmonary Artery Aneurysm Coexisting With Dysplastic Pulmonary Valves

*To the Editor:*

We read with utmost interest the report by Saedi and colleagues[1] of 3 cases of isolated congenital dysgenesia of a pulmonary valve cusp associated with pulmonary artery aneurysm (PAA). In our view, this observation expands our understanding of this pathological association and might improve the nosologic classification and prognostic significance of PAA. Indeed, if one excludes what are usually peripheral "false aneurysms" of numerous causes (infectious, inflammatory, tumoral, congenital, traumatic or iatrogenic origin), then the PAA with pulmonary hypertension (ie, associated with cardiac disease [eg, left-to-right shunts, post-stenotic dilation], hypoxemic lung disease, and thromboembolic disease) and last aneurysm in the context of connective tissue diseases (Marfan, Loeys-Dietz) or inflammatory angiitis (eg, Takayasu, Behçet, Horton), the remaining consists of so-called low-pressure PAAs, as proposed by Veldtman and colleagues in 2003.[2] This subset includes true idiopathic aneurysms (therefore considered as such after exclusion of any known causes), which are exceedingly rare. The recent literature associated with 3 cases recently treated in our institution suggests that most of these "idiopathic PAAs" or low-pressure PAAs are associated with anomalies of the pulmonary valves within a spectrum extending from absent, to quadricuspid valves including not only bicuspid, but also dysplastic valves accounting for mild stenosis or insufficiency or, as in this report, dysgenesia of leaflets.[1] In this context, it is our contention that all these forms represent a nosologic entity in which abnormal morphogenesis of the outflow of the heart and arterial valves (linked to the role of neural crest cells from hindbrain and second heart field cells population)[3,4] is pivotal. The etiology is therefore congenital, with a development and an evolution that might result from interaction between intrinsic parietal weakness (media cells' apoptosis or cystic medial degeneration) and increased undue parietal stress due to valve dysfunction and related hemodynamics. These PAAs without pulmonary hypertension are unlikely to complicate abruptly. The surgical indication in this pathological association is decided most often by ongoing pulmonary valve regurgitation and right ventricular volume overload, rather than by PA size.

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## Isolated Pulmonary Artery Aneurysm Is Associated With Pulmonary Valve Leaflet Anomalies

*Reply*
*To the Editor:*

We thank Radermecker and colleagues[1] for their interest in our article[2] and highlighting several important points in their letter to the editor. Isolated pulmonary artery aneurysm without systemic involvement is a rare and clinically challenging abnormality. Guidelines are deficient regarding the best management strategy, and the standard treatment is controversial. Pulmonary arterial hypertension and vascular inflammatory processes are known to lead to arterial wall fragility, pulmonary dilation, and occasional dissection, but in the absence of specific arteriopathies, ascertaining the risk of complications in pulmonary artery dilation is more difficult. In our institute, we have observed that anomalies in the formation of the pulmonary valve leaflets are associated with pulmonary dilation and aneurysm formation. These anomalies are often hard or impossible to appreciate by transthoracic or even transesophageal echocardiography. Imaging modalities with the potential to image the pulmonary leaflets in an en face and focused view and 3-dimensional reconstruction including computed tomography angiography and cardiac magnetic resonance imaging are the best methods to detect the abnormal morphology of the pulmonary valve. Currently surgical recommendations for repair of the pulmonary artery aneurysm are similar to the ascending aorta aneurysm, and patients with aneurysm diameters of above 5.5 cm are thought to be at a greater risk for dissection and rupture. However, if the aneurysm is secondary to abnormal valvular leaflet morphology, there is no arterial wall weakening or remarkable cystic media degeneration and the dilation is the consequence of pulmonary flow disturbances. Patients with congenital valvular pulmonary stenosis have long been known to have post stenotic pulmonary artery dilation. The management in these patients consists of relief of the stenosis and clinical follow-up for the poststenotic dilation.[2] We believe that the same rule applies to patients with isolated pulmonary artery dilation due to valvular leaflet dysgenesia. If there are no compressive effects on adjacent structures by the aneurysm, the timing of surgery could be determined based on the guideline recommendations for correction of concomitant pulmonary valve regurgitation or stenosis.

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