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Isolated acquired pulmonary valve regurgitation
Report of long-term follow-up

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SUMMARY A patient is reported with isolated pulmonary valvular regurgitation acquired probably as a result of endocarditis at age 22 years. The patient remained essentially asymptomatic with physical findings compatible with haemodynamically significant pulmonary regurgitation until death at age 85 years. A necropsy confirmed the presence of isolated severe pulmonary valvular destruction. This long-term follow-up lends further support to the concept that significant volume overload of the right ventricle from pulmonary regurgitation is well tolerated and usually does not require surgical intervention.

Since isolated acquired pulmonary valvular regurgitation is an unusual lesion (Abbott, 1936), the long-term consequences of the volume overload on the right ventricle are not well established in humans and the indications, if any, for surgical intervention remain unclear.

The following case report is a long-term follow-up of a case of isolated pulmonary regurgitation presumably occurring after bacterial endocarditis.

Case history

The patient was an 85-year-old white man. In 1912 he was admitted to hospital in Chicago, Illinois, with a prolonged febrile illness diagnosed as subacute bacterial endocarditis. After a prolonged stay in hospital and treatment of unknown type, he was discharged as cured, but was told that he had developed a heart murmur. He was advised to retire at the age of 22 years and to lead a quiet life; this he did. He was essentially asymptomatic from this point onward except for mild exertional dyspnoea.

In 1945, at age 54 years, he was first seen in Atlanta, Georgia, by the late Dr James Edgar Paullin. The cardiac physical findings described by Dr Paullin were as follows: 'At the apex, there is a soft systolic murmur, and a very faint diastolic murmur ... the first heart sound has a distinct click to it as though there was a moderate amount of sclerosis of the mitral or tricuspid valve. As one approaches the base of the heart, the diastolic murmur is heard best to the left of the sternum and very indistinctly over the right side in the 3rd and 4th interspaces. His pulse is regular in force and rhythm, equal and synchronous. It does not have a typical Corrigan's character. No capillary pulse. No pistol shot femorals. The patient was able to hold his breath for 10 seconds on several occasions without accelerating his pulse ...' Dr Paullin's conclusion was that he probably had mitral and aortic regurgitation.

In 1969 he was found to have chronic lymphocytic leukaemia which was successfully treated with chlorambucil. The patient was admitted in 1975 because of relapse of his leukaemia. After a prolonged hospital course he succumbed with an intestinal ileus. During his hospital course his cardiac examination revealed a heart rate of about 100/min. There was no jugular venous distension. All peripheral pulses were present and normal. Praecordial examination disclosed no abnormal pulsations. There was a prominent right ventricular systolic pulsation felt and seen in the epigastrium. On ausculation the first and second heart sounds were normal with inspiratory splitting of S_1. At the upper left sternal border a grade 2/6 systolic crescendo-decrescendo murmur and a grade 2/6 low-pitched, rumbling, crescendo-decrescendo early diastolic murmur starting soon after P_1 was heard. Third and fourth heart sounds were audible and both increased slightly with inspiration. His electrocardiogram showed sinus tachycardia, left axis deviation, and left bundle-branch block. His

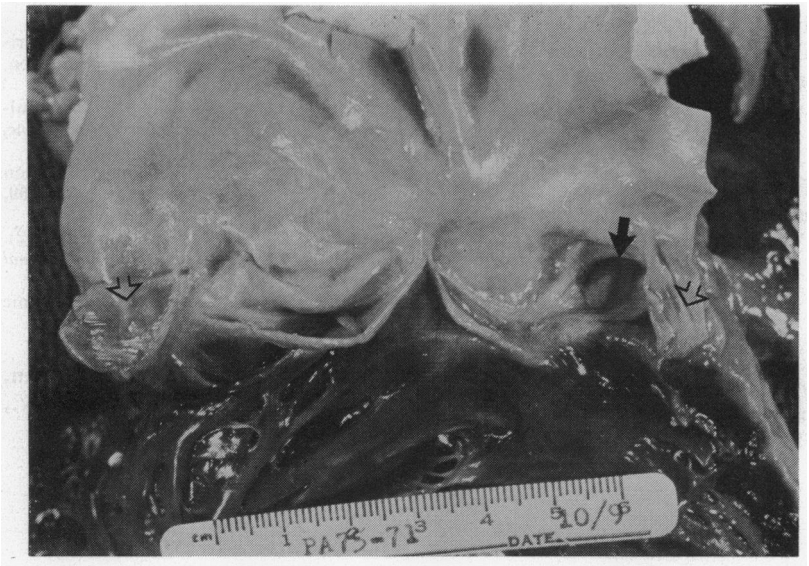

Figure *Necropsy specimen showing the opened right ventricle and pulmonary artery. The open arrows point to the single intact leaflet. The closed arrow points to the 'aneurysm' in the ring area probably representing a healed abscess. Note the vestiges of the other two leaflets.*

radiographic heart size was at the upper limits of normal. In addition, there were some chronic pulmonary changes with hilar lymphadenopathy, scarring, and fluid on the right side. An echocardiogram showed a dilated right ventricular chamber with strikingly paradoxical septal motion. The degree of paradoxical septal motion was thought to result from the combined effects of right ventricular overload and left bundle-branch block. A pulmonary valve echo was not recorded.

At necropsy the heart weighed 425 g. The right ventricle was moderately dilated and hypertrophied (7 mm free wall thickness). The pulmonary valve was grossly deformed (Figure). The left anterior leaflet was grossly intact and appeared normal apart from slight thickening. Half of the right anterior leaflet was absent, with a thickened residual leaflet partially attached to a muscle band below it. The posterior leaflet was absent except for a tiny rolled up raphe along the attachment to the valve ring. There was a small blind outpouching of the pulmonary ring area near the posterior leaflet which appeared to be a healed abscess. The surface of the deformed pulmonary valve was glistening and free of thrombotic material. The pulmonary artery was slightly dilated but otherwise normal. The rest of the heart including other valves was normal. Professor R. E. B. Hudson reviewed the heart and concluded that the lesion was in all probability caused by healed bacterial endocarditis.

Discussion

Presumably this patient had infective endocarditis involving his pulmonary valve in 1912 at the age of 22. A diastolic murmur compatible with pulmonary regurgitation was described in 1945 and an echocardiogram in 1975 documented a volume overload on his right ventricle. At necropsy the pulmonary valve was severely damaged and right ventricular hypertrophy was present. Though we cannot state with certainty that his murmur in 1912 resulted from pulmonary regurgitation, the necropsy did not show another reason for a diastolic murmur other than the deformed, virtually absent pulmonary valve. Despite the presence of pulmonary regurgitation for at least 30 years and possibly 63 years, he was never symptomatic and had no signs of right ventricular failure though he did have right ventricular hypertrophy and dilatation.

Previous reports of patients with congenital and acquired pulmonary regurgitation have shown that the haemodynamic effects are well tolerated as long as the pulmonary arterial pressure is low and there are no associated lesions (Price, 1961; Laneve *et al.*, 1962; Holmes *et al.*, 1968). In experimental animals, removal of one of the three pulmonary leaflets produced no long-term haemodynamic effects, whereas removal of 2 of 3 leaflets or creation of a shunt lesion in addition produced congestive heart failure in some, but not all animals (Kay and Thomas, 1954; Fowler and Duchesne, 1958; Ellison *et al.*, 1970). This situation may be analogous to our patient who had only one intact pulmonary valve leaflet but no clinical heart failure.

This case adds further support to the evidence that isolated pulmonary regurgitation in man can be tolerated for lengthy periods with no significant

symptoms or disability though compensatory right ventricular hypertrophy and dilatation usually occur over a period of time. Management in such cases requires no more than prophylaxis for bacterial endocarditis.

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