ISOLATED CONGENITAL PULMONARY INCOMPETENCE

BY

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Acquired incompetence of the pulmonary valve is relatively common and is usually due to pulmonary hypertension (Graham Steel, 1888) or following valvotomy for pulmonary stenosis. Endocarditis, rheumatic disease, and syphilitic infection may occasionally produce this lesion. Congenital pulmonary incompetence is rare. In 1000 cases of congenital heart disease, Abbott (1936) had 8, and 6 of these were due to idiopathic dilatation of the pulmonary artery while 2 were due to valvar deformity. Both these conditions may occur without pulmonary incompetence (Segel *et al.*, 1957; Kissin, 1936). When reviewing the subject in 1961, Price found that few cases had been diagnosed at cardiac catheterization. This paper presents a patient with isolated pulmonary incompetence, believed to be congenital in origin, in whom the diagnosis was made at right heart catheterization by intracardiac phonocardiography and dye dilution studies.

Case Report

The patient, a Maori man aged 46, was referred for assessment of a cardiac murmur known to have been present for 23 years when he was examined for military service. A regular soldier, he was leading an active life which included participation in sport and he had no cardiac symptoms. There was no family history of heart disease and his past history was non-contributory.

The patient was a well-developed muscular man, 5 ft. 8 in. (172 cm.) in height and weighing 164 lb. (74.4 kg.). On examination of the cardiovascular system the pulse was normal and the blood pressure 135/95 mm. Hg. The venous pressure was not raised and the jugular venous pulse was normal. The cardiac impulse was not palpable through his thick muscular chest. On auscultation the second heart sound was faint in all areas and appeared single. In the fourth left intercostal space there was a coarse crepitant systolic murmur of grade 1/4 intensity occupying most of systole and starting just after the first heart sound. A low-pitched musical decrescendo diastolic murmur was present separated from the audible element of the second heart sound by a short interval. It was not audible in the pulmonary area and no respiratory variation was noticed with either murmur. The chest radiograph showed a cardiac index of 0.52 and fullness of both hilar shadows, presumably due to large pulmonary artery branches. Fluoroscopy showed conspicuous pulsation in the region of the main pulmonary artery with increased hilar pulsation, but no definite expansile pulsation in the secondary branches. The electrocardiogram (Fig. 1) showed partial right bundle-branch block with clockwise rotation, and slight right ventricular hypertrophy. These findings suggested either isolated pulmonary or tricuspid valve disease, and right heart catheterization was performed to establish the diagnosis. A No. 8 size double lumen catheter was inserted at the right superficial brachial vein and advanced so that the distal lumen was lying in the main pulmonary artery and the proximal lumen in the right ventricle. Simultaneous pressures were recorded from these two regions using equal sensitive pressure transducers adjusted to identical baselines. Identical systolic and diastolic pressures were found in right ventricle and pulmonary artery and repeated withdrawals did not demonstrate any change nor was there any gradient across the pulmonary artery branches. Pressures in pulmonary artery and right ventricle were 33/3 mm. Hg, while mean pressures of wedge pulmonary artery and right atrium were 3.5 and 3 mm. Hg, respectively. The cardiac index as measured by the Fick method was 3.8. Injection of indocyanine

FIG. 1.—Electrocardiogram showing partial right bundle-branch block and right ventricular hypertrophy.

FIG. 2.—Sound records of intracardiac phonocatheter in right ventricle and right atrium at high and mid frequency (HF and MF).

green into pulmonary artery and sampling via the proximal lumen in right ventricle showed a considerable quantity of instantaneously appearing dye in the right ventricle, thus confirming regurgitation. Simultaneous pressures were also taken across the tricuspid valve and these established that there was no end-diastolic gradient, nor was there any instantaneously appearing dye in the right atrium following right ventricular injection. An intracardiac phonocatheter revealed a delayed crescendo-decrescendo diastolic murmur and a faint systolic murmur in the right ventricle (Fig. 2) but there were no murmurs in the right atrium. The external phonocardiogram at the fourth left interspace and pulmonary artery area showed the same murmur maximal at the fourth left interspace (Fig. 3). Though it was recorded in the pulmonary area, it could not be heard by several observers. The sound recording showed no evidence of pulmonary valve closure. Dye curves recorded from the vena cava, right atrium, and right ventricle and a recording at the left ear showed a lag in disappearance phase, but a recirculation was present, while curves from left and right pulmonary arteries were normal; this also confirmed pulmonary regurgitation. The dye curves and saturation figures excluded a shunt at any level or any anomalous pulmonary venous return.

Comment

This patient, known to have had a murmur since the age of 23 years, had nothing in his past history to suggest endocarditis, specific infection, or rheumatic fever. There is no evidence of any other valvar involvement, and it is reasonable to postulate a congenital cause for the anomaly.

FIG. 3.—Sound recordings of external phonocardiogram on high frequency over pulmonary artery and at the fourth left intercostal space.

pulmonary valve in this case; simultaneous arterial sampling was not done and it is not therefore possible to determine the degree of regurgitation. From the curves obtained however it was felt that the regurgitation was significant. In the absence of dilatation of the pulmonary artery it is considered that regurgitation was due to a primary valvar anomaly. Little is known of the prognosis in isolated pulmonary valvular regurgitation. Experimentally produced pulmonary valvular incompetence in the dog is thought to be well tolerated (Ellison *et al.*, 1955). However, Fowler and Duchesne (1958) have demonstrated that this lesion is not entirely without consequence as right ventricular dilatation was observed twelve to eighteen months after production of the lesion. Clinically this lesion has always been postulated as benign when there is no accompanying pulmonary hypertension and is haemodynamically equivalent to an atrial septal defect in that both lesions are associated with an increase in right ventricular output. In this case the cardiac index was within normal limits and symptomatically there was no limitation. A patient previously described by Ford *et al.* (1956) died suddenly at the age of 44 years after progressive right heart failure and the authors stressed that this was not always a benign lesion. In the present instance, in which the data obtained argued for appreciable regurgitation, the lesion has been well tolerated.

Summary

A patient with isolated pulmonary valvular regurgitation is described. In the absence of relevant heart illnesses and with the long history of a cardiac murmur it is assumed that the valve defect is congenital; as there is no dilatation of the pulmonary artery the lesion must be valvar. Clinical features are stressed with particular reference to the cadence, timing, and situation of the diastolic murmur.

The diagnosis was established at catheterization by the instantaneous appearance of dye in the
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There is frequently an interval between the second sound and the onset of the diastolic murmur which may be of the crescendo-decrescendo type; in the absence of pulmonary hypertension it is usually low-pitched (McKusick, 1958). The localization of the murmur to the fourth left interspace is of considerable interest, but has been mentioned once before (Collins, Braunwald, and Morrow, 1960). Presumably the thick muscular chest is responsible for its failure to be detected clinically in the pulmonary area even though it was detected phonocardiographically. An intracardiac phonocatheter is useful for the diagnosis of this condition and its simplicity is appealing. Diastolic murmurs recorded in the right ventricle represent stenotic or flow murmurs across the tricuspid valve or regurgitant across the pulmonary, and as there was no abnormality at tricuspid level this was due to pulmonary regurgitation. Other writers (Price, 1961; Collins *et al.*, 1960) comment on similar pressure pulses in the right ventricle and pulmonary artery as a diagnostic feature of this condition and mention that a late or absent diastolic notch and a steeply sloping diastolic limb are characteristic features of the pulmonary artery pressure curve. The method as described by Bajec *et al.* (1958) of injection distally and sampling proximally to the valve successfully localized the regurgitation to the

right ventricle when it was injected into the pulmonary artery, and confirmed by identical pressure recordings in both sites and also by the presence of the typical diastolic murmur in the right ventricle. This lesion was not associated with any symptoms.

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References

- Abbott, M. E. (1936). *Atlas of Congenital Cardiac Disease*. American Heart Association, New York.
- Bajec, D. F., Birkhead, N. C., Carter, S. A., and Wood, E. H. (1958). Localization and estimation of severity of regurgitant flow at the pulmonary and tricuspid valves. *Proc. Mayo Clin.*, 33, 569.
- Collins, N. P., Braunwald, E., and Morrow, A. G. (1960). Isolated congenital pulmonic valvular regurgitation. *Amer. J. Med.*, 28, 159.
- Ellison, R. G., Brown, W. J., Jr., Hague, E. E., and Hamilton, W. F. (1955). Physiologic observations in experimental pulmonary insufficiency. *J. thorac. Surg.*, 30, 633.
- Ford, A. B., Hellerstein, H. K., Wood, C., and Kelly, H. B. (1956). Isolated congenital bicuspid pulmonary valve. *Amer. J. Med.*, 20, 474.
- Fowler, N. O., and Duchesne, E. R. (1958). Effect of experimental pulmonary valvular insufficiency on the circulation. *J. thorac. Surg.*, 35, 643.
- Kissin, M. (1936). Pulmonary insufficiency with a supernumerary cusp in the pulmonary valve. *Amer. Heart J.*, 12, 206.
- McKusick, V. A. (1958). *Cardiovascular Sound in Health and Disease*. Williams and Wilkins, Baltimore.
- Price, B. O. (1961). Isolated incompetence of the pulmonic valve. *Circulation*, 23, 596.
- Segel, N., van Lingen, B., Resnekov, V., and McGregor, M. (1957). Isolated congenital pulmonary incompetence. *S. Afr. med. J.*, 31, 1157.