Quadricuspid Pulmonary Valve

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Quadricuspid pulmonary valve is a rare finding which is usually discovered *post mortem* (Hudson, 1965). Its incidence is variously reported from 1 case in 20,000 necropsies (Simonds, 1923) to 5 cases in 5000 necropsies (Dagnini, 1930). Most other reports suggest, however, that the latter figure is the more accurate (Simpson, 1898; Thilo, 1909; de Vries, 1918; Houck, 1929). In reviewing the subject, Kissin (1936) noted that in many of the previously reported cases, quadricuspid pulmonary valves were found to be associated with other congenital abnormalities of the heart. As an isolated finding, this anomaly seems, therefore, to be very rare. The following is an account of four cases of isolated congenital quadricuspid pulmonary valve, and a further one in which the anomaly was associated with rheumatic mitral valve disease. In each instance, the anomaly was an incidental finding at necropsy, and there had been no clinical evidence of a pulmonary valve lesion during life. Four of these cases were encountered within the six months November 1964 to May 1965 in patients who had resided in the Bristol area. In 2 of the 5 patients, there was morbid anatomical evidence of pulmonary regurgitation. In one, there was evidence of pulmonary hypertension, but this was the patient with mitral valve disease. There was no other cardiac abnormality in the other 4 patients.

CASE REPORTS

Case 1 (PM 3860/TFH)†. A 73-year-old woman died in hospital in December 1953 as a result of barbiturate poisoning. Clinically, the cardiovascular system was normal. At necropsy, the heart was small and atrophic (250 g.) but otherwise normal except for a

small accessory cusp in the pulmonary valve. The endocardium of the right ventricle was normal.

Case 2 (PM 8903/BAE). A 43-year-old woman died in hospital in November 1964 of cardiac failure due to rheumatic heart disease. Clinically, there was evidence of mitral stenosis and incompetence and tricuspid incompetence. At necropsy, the heart was enlarged (610 g.) due to biventricular hypertrophy and dilatation. The pulmonary valve comprised four cusps, three of equal size and one smaller fenestrated cusp (0.5 cm. across) interposed between the septal and the right atrial cusps. The endocardium of the right ventricle was normal.

Case 3 (PM 9099/BAE). A 74-year-old woman died in hospital in March 1965 as a result of carcinoma of the colon. Clinically, the cardiovascular system was normal. At necropsy, the heart (355 g.) showed calcification of the aortic valve cusps. The pulmonary valve had four cusps, an accessory cusp (0.5 cm. across) being interposed between the septal and the right atrial cusps. The other three cusps were of equal size. Immediately beneath the accessory cusp was a plaque of endocardial fibrosis (0.8 cm. diameter).

Case 4 (PM 9193/BAE). A 73-year-old woman died in hospital in May 1965, following amputation of the right leg for arteriosclerotic gangrene. Clinically, the heart was normal. At necropsy, the heart was normal (323 g.) except for the pulmonary valve which contained four cusps, a fenestrated accessory cusp (0.5 cm. across) being interposed between the septal and the right atrial cusps (Figure). The other three cusps were of equal size. Immediately beneath the accessory cusp was a plaque of endocardial fibrosis (0.5 cm. diameter).

Case 5 (PM 9225/BAE). A 76-year-old man died in hospital in May 1965 following cerebral infarction. He had diabetes mellitus, severe generalized arterial disease, and high blood pressure. At necropsy, the heart was enlarged (504 g.) due to left ventricular hypertrophy. The pulmonary valve had four cusps, three of equal size and an accessory cusp (0.3 cm. across) interposed between the septal and the right atrial cusp. The endocardium of the right ventricle was normal.

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† Necropsy reference numbers quoted are those of the University of Bristol Department of Pathology.

FIGURE.—Pulmonary valve of Case 4 showing fenestrated accessory cusp and plaque of endocardial fibrosis beneath.

DISCUSSION

Longworth (1878) stated that the pulmonary valve cannot operate at maximum efficiency in the presence of four cusps. However, in only three of the cases reviewed by Kissin (1936) was there clinical evidence of pulmonary regurgitation during life. This was subsequently confirmed at necropsy by the finding of dilatation of the right ventricle and pulmonary artery. Pathological evidence of regurgitation was found more frequently, but to what extent is not clear.

White patches of endocardial thickening are well recognized at sites of abnormal blood flow beneath an incompetent valve (Hudson, 1965), and this so-called 'jet lesion' was evident in Cases 3 and 4. It seems likely, therefore, that some degree of regurgitation had occurred in the region of the accessory cusp in these two patients. This was insufficient to give rise to dilatation of the right ventricle or pulmonary artery, or to the clinical signs of a valve lesion during life. It is apparent, therefore, that a quadricuspid pulmonary valve rarely gives rise to clinical evidence of incompetence.

Isolated pulmonary incompetence, when diagnosed clinically, does not, however, necessarily connote an underlying congenital anomaly of the valve. In a pathological study of 1000 cases of congenital heart disease, Abbott (1936) found 8

with isolated pulmonary incompetence. Of these, 6 were due to idiopathic dilatation of the pulmonary artery, and 2 were associated with a valve deformity. The first report of isolated pulmonary incompetence diagnosed during life was by Kezdi, Priest, and Smith (1955). Up to 1961, 15 such cases had been reported in the English literature (Price, 1961), and necropsy in 2 of these revealed a congenital deformity of the pulmonary valve. More recently, there have been a few clinical reports of isolated congenital pulmonary incompetence (Sloman and Wee, 1963; Sanyal *et al.*, 1964; Kelly, 1965), but the presence of an underlying pulmonary valve anomaly in these cases is conjectural.

The most commonly reported arrangement in quadricuspid pulmonary valve is that of a rudimentary accessory cusp interposed between three cusps of equal size. The accessory cusp is frequently deformed, shrunk, or fenestrated. Less common findings are the combination of two large and two small cusps, four cusps of equal size, or four cusps each of different size (Kissin, 1936).

It is emphasized that in none of the cases reported here was the presence of a quadricuspid pulmonary valve a factor contributing to death. The finding was unsuspected, the valve being inspected only as part of the routine dissection of the heart. The anomaly may possibly be more common than a review of the reports suggests.

SUMMARY

Five cases of congenital quadricuspid pulmonary valve are recorded. In all, the anomaly was an incidental finding at necropsy. Four of the five patients were women. In two there was evidence of regurgitation in the region of the accessory cusp.

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REFERENCES

- Abbott, Maude E. (1936). *Atlas of Congenital Cardiac Disease*. American Heart Association, New York.
- Dagnini, G. (1930). Anomalie di numero delle valvole polmonari. *Cuore e Circol.*, **14**, 363.
- de Vries, W. M. (1918). Über Abweichungen in der Zahl der Semilunarklappen. *Beitr. path. Anat.*, **64**, 39.
- Houck, G. H. (1929). The incidence of cardiac anomalies at the Massachusetts General Hospital. *J. techn. Meth.*, **12**, 167.
- Hudson, R. E. B. (1965). Endocardial fibrosis. In *Cardiovascular Pathology*, Vol. I, p. 863. Edward Arnold, London.
- Kelly, D. T. (1965). Isolated congenital pulmonary incompetence. *Brit. Heart J.*, **27**, 777.
- Kezdi, P., Priest, W. S., and Smith, J. M. (1955). Pulmonic regurgitation. *Quart. Bull. Northw. Univ. med. Sch.*, **29**, 368.
- Kissin, M. (1936). Pulmonary insufficiency with a supernumerary cusp in the pulmonary valve; report of a case with review of the literature. *Amer. Heart J.*, **12**, 206.
- Longworth, L. R. (1878). Why are the segments of the semilunar valves three in number? *Clinic, Cincinnati*, **14**, 73.
- Price, B. O. (1961). Isolated incompetence of the pulmonic valve. *Circulation*, **23**, 596.
- Sanyal, S. K., Hipona, F. A., Browne, M. J., and Talner, N. S. (1964). Congenital insufficiency of the pulmonary valve. *J. Pediat.*, **64**, 728.
- Simonds, J. P. (1923). Congenital malformations of the aortic and pulmonary valves. *Amer. J. med. Sci.*, **166**, 584.
- Simpson, F. O. (1898). Congenital abnormalities of the heart in the insane. *J. Anat. Physiol. (Lond.)*, **32**, 679.
- Sloman, G., and Wee, K. P. (1963). Isolated congenital pulmonary valve incompetence. *Amer. Heart J.*, **66**, 532.
- Thilo, L. (1909). *Zur Kenntnis der Missbildungen des Herzens*. Inaug. Diss. B. Georgi, Leipzig.