Case reports

British Heart Journal, 1972, 34, 859-861.

Calcific tricuspid incompetence in childhood

Malcolm Clarke¹

From the Department of Cardiology, Hospital for Sick Children, Toronto, Ontario, Canada

An unusual case of calcific tricuspid incompetence is reported. The calcification was present at the age of 10 years and was not associated with a left-to-right shunt or increased right ventricular pressure. The underlying disorder may represent congenital tricuspid incompetence, a rare condition diagnosed in only 2 of 30,000 children examined in the Cardiology Department of this hospital.

Calcification of the tricuspid valve is rare. Previously reported cases have been associated with increased pressure in the right ventricle or a left-to-right shunt proximal to the tricuspid valve. Furthermore, the calcification has not been evident until the fifth decade of life or later.

A case is reported here in which moderately severe tricuspid valve calcification was present at 10 years of age. There was isolated tricuspid incompetence, in itself an uncommon disorder, with normal right ventricular pressure and no intracardiac shunting.

Case report

An otherwise normal girl was found to have a soft systolic murmur at 11 months of age. When aged 10 years, a systolic murmur was again detected and investigated by right and left heart catheterization. Pulmonary and left ventricular angiograms were performed, but no abnormalities were noticed at that time. In 1970, when aged 13 years, a routine chest x-ray showed slight cardiac enlargement. Subsequent screening revealed intracardiac calcification (Fig. 1). Review of the cine-angiograms taken 3 years earlier showed that this had, in fact, been present then.

She was asymptomatic. Her past history included thrombocytopenic purpura in infancy, infective hepatitis when 10, and a cholecystectomy at 13 for a single cholesterol gallstone. She was a healthy, normally-developed girl with no cyanosis, clubbing, or heart failure. The jugular venous pressure was slightly raised. Arterial pulses were normal with a right brachial blood pressure of 110/80 mmHg. In addition to normal first and second heart sounds, there was a grade 2/4 pansystolic murmur, maximal just medial to the apex,

which increased in intensity during inspiration. There was no diastolic murmur. The rest of the clinical examination was unremarkable.

Chest x-ray showed slight cardiac enlargement with normal pulmonary vascularity. The electrocardiogram showed sinus rhythm with normal atrial loading and an incomplete right bundle-branch block pattern.

FIG. 1 Oblique view of the chest taken during fluoroscopy. The intracardiac calcification is arrowed.

¹ Present address: Department of Cardiology, St. Bartholomew's Hospital, London.

Because the previous cardiac catheterization had not clearly demarcated the right ventricle, further studies were performed. The right ventricular pressure was 25/6 mmHg, the right atrial 'a' wave was 6 mmHg, and the 'v' wave 6 mmHg. The mean right atrial pressure was 5 mmHg. No shunts were seen by oximetry or angiography. Cineangiograms indicated moderate regurgitation through the tricuspid valve, which was calcified (Fig. 2). There was normal attachment of the valve leaflets and some enlargement of the right atrial and right ventricular cavities. The great vessels were normally related.

Among other investigations, her blood count and sedimentation rate were normal, serum calcium, inorganic phosphates, alkaline phosphatases, and cholesterol were within normal limits, and the dye test for toxoplasmosis was negative.

She remains asymptomatic and is receiving no medication.

Discussion

Calcified aortic and mitral valves are common, but involvement of the tricuspid valve by calcification is rare, even where severe damage due to rheumatic valvulitis has occurred (Hollman, 1957). Other authors' experiences of tricuspid calcification are summarized in Table 1. It is of interest to note that in all of these patients the calcification was associated with congenital heart disease of at least 40 years' standing. Two commonly reported features are raised pressure in the right ventricle and increased flow across the tricuspid valve secondary to a left-to-right shunt proximal to it. Neither of these factors was present in the patient reported here, in whom the only demonstrable abnormality was tricuspid incompetence.

The aetiology of tricuspid valve disease in

FIG. 2 Frame from cineangiogram, right anterior oblique view. Contrast has been injected into the right ventricle (RV) and has regurgitated into the right atrium (RA). There is a central jet of regurgitation which is unrelated to the position of the catheter through the tricuspid valve. PA = pulmonary artery.

TABLE Tricuspid valve calcification

Authors	Age and sex of patients	Diagnosis	RV pressure (mmHg)	Pulmonary/systemic flow ratio	Detection of calcification
Hollman (1957)	46 M	Pulm. valve stenosis	Presumed increased	1:1	Necropsy
Sautter, Emanuel, and Doege (1965)	75 F	Pulm. valve stenosis; ventric. septal defect; tricuspid stenosis	Presumed increased	Not known	Necropsy
Rogers, Chandler, and Franch (1969)	51 F	Pulm. valve stenosis; atrial septal defect	69/9	'Small left-to-right shunt'	Radiography
"	42 M	Atrial septal defect	70/10	2.8:1	Radiography
"	51 M	Pulm. valve stenosis	165/10	1:1	Radiography
"	51 F	Pulm. valve stenosis; atrial septal defect	Presumed increased	Not known	Radiography
Cooksey, Parker, and Weldon (1970)	44 M	Atrial septal defect	35/10	2.5:1	Radiography, confirmed at operation
Present case	13 F	Tricuspid incompetence	25/6	1:1	Radiography

this patient is uncertain. No evidence was found to suggest systemic disease such as carcinoid syndrome, systemic lupus erythematosus, rheumatic heart disease, or previous bacterial endocarditis. Nor was there a history of severe blunt chest injury to implicate a traumatic causation for the tricuspid incompetence (Jahnke, Nelson, and Aaby, 1967). The appearances were unlike those of endomyocardial fibrosis, in which calcification of the outflow tract of the right ventricle may occur (Abrahams, 1962). Ebstein's anomaly, even in a mild form, was felt to be excluded by the angiogram. No abnormality of calcium metabolism was detected, though this does not necessarily reflect the status at the time the calcification occurred some years earlier. It could be argued that the valvar incompetence is secondary to distortion resulting from calcification under an undetermined stimulus, but the presence of a cardiac murmur in infancy suggests that the basic lesion is congenital in origin.

Isolated congenital tricuspid incompetence is usually due to underdevelopment of the subvalvar apparatus; the abnormally short chordae tendineae hold the valve open during ventricular systole. It may present in varying forms, from severe heart failure with death within hours of birth (Jordan and Taylor, 1966) to complete lack of symptoms for many years (Barritt and Urich, 1956). It is an uncommon disorder: Kincaid *et al.* (1962) reviewed 8 published cases and added 2 of their own. Two other examples have been recorded by Reisman *et al.* (1965), and one each by Jordan and Taylor (1966) and Antia and Osunkoya (1969). Valve calcification has not been described in this condition, the majority of reports being pathological studies on infants. Many mild forms of this disorder are probably unrecognized. However, in addition to the case reported above, only one other child out of 30,000 patients examined by the cardiologists of the Hospital for Sick Children, Toronto, has isolated tricuspid incompetence. This boy, now aged 8 years, underwent investigation at the age of 4 after the discovery of a systolic murmur which increased in intensity during inspiration. The electrocardiogram and chest x-ray were normal. At

cardiac catheterization, the right heart pressures were normal and no shunts were detected. Moderate tricuspid regurgitation, unrelated to the position of the catheter, was seen on the angiogram. There was slight enlargement of the right ventricle and right atrium. The valve leaflets were normally attached to the valve ring. He remains free from symptoms, and no calcification has been noted.

I should like to thank Dr. J. D. Keith for allowing me to report these patients, and Dr. T. Izukawa for his helpful advice.

References

- Abrahams, D. G. (1962). Endomyocardial fibrosis of the right ventricle. *Quarterly Journal of Medicine*, **31**, 1.
- Antia, A. U., and Osunkoya, B. O. (1969). Congenital tricuspid incompetence. *British Heart Journal*, **31**, 664.
- Barritt, D. W., and Urich, H. (1956). Congenital tricuspid incompetence. *British Heart Journal*, **18**, 133.
- Cooksey, J. D., Parker, B. M., and Weldon, C. S. (1970). Atrial septal defect and calcification of the tricuspid valve. *British Heart Journal*, **32**, 409.
- Hollman, A. (1957). The anatomical appearance in rheumatic tricuspid valve disease. *British Heart Journal*, **19**, 211.
- Jahnke, E. J., Jr., Nelson, W. P., and Aaby, G. V. (1967). Tricuspid insufficiency; the result of non-penetrating cardiac trauma. *Archives of Surgery*, **95**, 880.
- Jordan, J. D., and Taylor, W. T. (1966). Case of combined congenital tricuspid and mitral insufficiency. *American Journal of Diseases of Children*, **111**, 105.
- Kincaid, O. W., Swan, H. J. C., Ongley, P. A., and Tites, J. L. (1962). Congenital tricuspid insufficiency. Report of two cases. *Proceedings of the Staff Meetings of the Mayo Clinic*, **37**, 640.
- Reisman, M., Hipona, F. A., Bloor, C. M., and Talner, N. S. (1965). Congenital tricuspid insufficiency. *Journal of Pediatrics*, **66**, 869.
- Rogers, J. V., Chandler, N. W., and Franch, R. H. (1969). Calcification of the tricuspid annulus. *American Journal of Roentgenology, Radium Therapy, and Nuclear Medicine*, **106**, 550.
- Sautter, R. D., Emanuel, D. A., and Doege, K. H. (1965). Association of pulmonary valvular stenosis and muscular ventricular septal defect. Report of a case in a patient aged 75 years. *American Journal of Cardiology*, **16**, 743.

Requests for reprints to Dr. Malcolm Clarke, St. Bartholomew's Hospital, London E.C.1.