Persistent right sinus venosus valve

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SUMMARY A 13-year-old girl presented with clinical features of pulmonary stenosis and regurgitation. Haemodynamic studies suggested the presence of a right ventricular tumour. M-mode and two dimensional echocardiograms indicated one or probably two soft thin walled structures originating from the right atrium. At operation a persistent right sinus venosus valve was removed.

One earlier case report described the M-mode echocardiographic features of this condition in a neonate who died shortly after operation. This report illustrates that a large persistent right sinus venosus valve may present clinically years after birth. Echocardiography played an important role in making the diagnosis.

At one stage during the life of the embryo the right valve of the sinus venosus almost divides the right atrium into two chambers. Normally this structure regresses some time between the ninth and 15th week. Its cephalic portion remains as the crista terminalis; its caudal portion, being divided by the left horn of the sinus venosus, remains as a valve of the inferior vena cava (Eustachian valve) and the valve of the coronary sinus (Thebesian valve).

The appearance of the valves can vary from a simple muscle bar to a fenestrated membrane or to a Chiari's network. These variations have been described extensively by Kauffman and Andersen¹ and Yater.² Large membranes in the right atrium, however, are rare. Persistence of the right sinus venosus valve can be associated with hypoplasia or atresia of the tricuspid and/or pulmonary artery orifice.¹⁻⁴ One case described by Gerlis and Anderson⁵ also had an imperforate Ebstein malformation of the tricuspid valve. Echocardiographic M-mode features of this phenomenon have been reported once.⁶ The appearance in diastole of a cloud of echoes in the tricuspid valve orifice and in the right ventricular outflow tract has also been reported.⁶

This report describes the haemodynamic, echocardiographic, and surgical findings in a case of persistent right sinus venosus valve presenting with unusual clinical findings of pulmonary stenosis and regurgitation.

Case report

A 13-year-old girl was known to have a systolic murmur since birth. In 1975 she was catheterised and

valvular pulmonary stenosis and regurgitation were diagnosed.

She was admitted to hospital in 1979 to evaluate her progressively limited exercise capacity. On examination she was a thin girl, 144 cm in height and 29 kg in weight. Her lips were mildly cyanosed. The peripheral pulses were normal and peripheral oedema was not present. Her blood pressure was 115/80 mmHg, with a heart rate of 80 beats per minute. The first sound was closely split. A grade 4/6 rough mid-systolic murmur was audible in the second intercostal space at the left sternal border. In addition, a grade 3 protodiastolic murmur was heard best in the third intercostal space. The second heart sound was split. These findings were confirmed by phonocardiography.

Chest x-ray film showed normal heart size, with dilatation of the pulmonary conus. Electrocardiography showed a PR interval of 0.18 second and evidence of right atrial and ventricular hypertrophy.

The M-mode echocardiogram, performed with the Echocardio Visor single element, showed multiple "blurred" echoes in diastole posterior to the anterior leaflet of the tricuspid valve (Fig. 1A). These echoes were also seen in the right ventricular outflow tract. There was no interval between the tricuspid valve opening and the appearance of these "blurred" echoes. At the level of the pulmonary valve a discrete echo structure lying anterior to the valve was detected. This echo structure moved posteriorly with the onset of systole, shortly after the opening of the pulmonary valve (Fig. 1B, see arrow).

When the heart was examined with a dynamically focused Multiscan system,⁷ an abnormal thin walled

Fig. 1 *M-mode echocardiogram of the persistent membrane. (A) shows a cloud of echoes in the tricuspid valve (TV) orifice (arrows). At the level of the pulmonary valve (B) one single echo structure could be detected, being anterior to the pulmonary valve (PV), which moves posteriorly in systole after a short interval (arrow).*

soft tumour could be seen attached to the inner surface of the right atrium. As seen by the four chamber view, this tumour entered the right ventricle during diastole. Using the sagittal right ventricular outflow tract view, multiple echoes were seen crossing the pulmonary valve during systole (Fig. 2A).

Haemodynamic studies disclosed an increased right ventricular pressure of 70 mmHg (50 mmHg in 1975). Pulmonary regurgitation of 27 per cent, measured with a flow velocity probe, was present. No intracardiac shunts were detected. An angiogram of the pulmonary artery showed filling of the pulmonary artery and right ventricular outflow tract. In addition, a pendular structure of low density could be seen moving up and down in the right ventricle (Fig. 2B). A contrast injection in the right ventricle disclosed a heavily trabeculated ventricle. Situated close to the tricuspid valve there was a round tumour which appeared to be freely mobile. In addition, the valve proved to be mildly regurgitant. Review of the angiographic data obtained in 1975 showed that the tumour was already present, but considerably smaller in size.

It was concluded, therefore, that a benign flexible moving tumour was present, probably originating from within the right atrium.

At operation a large membrane representing a persistent right sinus venosus valve was found and resec-

ted. The pulmonary valve was normal. The white finger-like membrane measured 7 by 3 cm. Three broad-stalked cut edges were recognised; they were fenestrated, as were the free edges between them. Histological examination showed that the membrane consisted of a collagenous connective tissue covered by thin endocardium.

Discussion

The appearances of a persistent right sinus venosus valve vary considerably. Usually a simple muscle bar is present, as large membranes are rare. Associated abnormalities, such as hypoplasia or atresia of the tricuspid and or pulmonary valve, are also rare.

This report illustrates the clinical and echocardiographic features of a persistent right sinus venosus valve without associated cardiac anomalies.

The M-mode echocardiographic features of a persistence of the right sinus venosus valve seen in a newborn infant have been reported on only one occasion.⁶ These features, as with those obtained in patients with right atrial myxoma, appear as a cloud of echoes penetrating the right ventricle during diastole.

Usually these echoes appear posterior to the anterior leaflet of the tricuspid valve. There is a slight time interval between the opening of the valve and the prolapse of the tumour into the tricuspid valve orifice.

Fig. 2 Two dimensional systolic image with diagram illustrating the right ventricular long axis view (A) which correlates with the angiocardigram of the right ventricle (B). In (A) a cloud of echoes representing the tumour (T) was seen moving up and down in the right ventricular outflow tract (RVOT). In (B) the tumour was seen as a pendular low contrast structure in the RVOT and crossing the pulmonary valve in systole. Arrows indicate the top of the tumour. (RV, right ventricle; PA, pulmonary artery.)

In our patient, however, this interval could not be recorded on M-mode echocardiography. Moreover, the cloud of echoes was not only present in the tricuspid valve orifice, but also appeared in the right ventricular outflow tract. The tumour was also seen passing the pulmonary valve as, in diastole, a single echo structure was present anterior to the pulmonary valve, which moved posteriorly shortly after its opening.

The real-time two dimensional four chamber view clearly showed a thin walled tumour attached to the inner surface of the right atrial wall. It crossed the tricuspid valve during diastole. Using the right ventricular long axis view a pendular tumour mass was seen in the right ventricular outflow which traversed the pulmonary valve. It was uncertain from these echocardiographic findings whether one or two soft thin walled tumours were present. Haemodynamic and angiographic data in 1975 had established a diagnosis of valvular pulmonary stenosis and regurgitation but the final studies in 1979 confirmed the presence of a soft tumour in the right ventricular cavity, which, in itself, caused the pulmonary obstruction.

At operation a large persistent right sinus venosus valve was removed successfully. Explanation for the absence of the interval on the M-mode echocardiogram between the appearance of the blurred echoes and the opening of the tricuspid valve must be related to the size of the persistent valve. This valve probably remained present within the right ventricular cavity and did not return to the right atrium in systole.

Our observations indicate that echocardiography is helpful in diagnosing this rare right atrial anomaly. The important features are the presence of multiple echoes which on the M-mode tracing appear posterior to the anterior tricuspid valve leaflet, while two dimensional echocardiography may visualise the structure at its site of atrial attachment.

It is fair to conclude, however, that the final diagnosis, and, in particular, its differentiation from other atrial "tumours", can only be established at operation.

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