CASE REPORT

Echocardiographic Diagnosis of Ruptured Aneurysm of Sinus of Valsalva
Report of Two Cases

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SUMMARY Echocardiographic features of two cases of ruptured congenital aneurysm of Valsalva sinus with (case 1) and without (case 2) a supracristal ventricular septal defect were studied before and after surgery by standard echocardiography, M-mode scan and two-dimensional echocardiography. Discontinuity was observed in the echo from the aneurysm wall of the Valsalva sinus in case 1 with ruptured aneurysm, but was not observed in a similar case of unruptured aneurysm. Herniation of the right coronary cusp of the aortic valve into the right ventricular outflow tract was also observed in case 1. After surgical correction the echo from the aneurysm wall and from the herniated right coronary cusp of the aortic valve disappeared. Discontinuity in the echo from the anterior aortic wall and the interventricular septum also disappeared. In case 2, discontinuity in the echo from the anterior aortic wall, and tricuspid flutter with an abnormally low early diastolic peak were observed. These abnormalities disappeared after surgery. The ability of echocardiography to detect ruptured Valsalva aneurysm is discussed.

THE CHARACTERISTIC ONSET and auscultatory findings in a ruptured aneurysm of the sinus of Valsalva have led to successful surgical repair. The differential diagnosis may at times be difficult and echocardiography, as in other types of congenital heart disease, has proved useful. In the present report, the echocardiographic manifestations in two surgically repaired cases of ruptured congenital aneurysm of the sinus of Valsalva of two different types are described.

Materials

Two cases of ruptured congenital aneurysm of Valsalva sinus with and without a supracristal ventricular septal defect were studied.

Case 1: K.S., a 25-year-old man. The ruptured aneurysm, situated in the right coronary Valsalva sinus, was associated with a supracristal ventricular septal defect 15 mm in diameter and aortic regurgitation of type Ic. The rupture in the top of the aneurysm measured during surgery was about 5 mm in diameter and opened into the right ventricular outflow tract. At surgery after the ligation of the rupture, the ventricular septal defect was closed from both sides of the septum and the aortic wall, using a teflon patch and pledget.

Case 2: T.Y., a 36-year-old woman. This case revealed at surgery a ruptured congenital aneurysm of the right coronary Valsalva sinus without an associated ventricular septal defect. The size of the aneurysm was 15 mm in length and 10 mm in diameter at its base. The rupture in the top of the aneurysm was 5 mm in diameter and opened into the right ventricle. The base of the Valsalva aneurysm was closed using a teflon patch and pledget from both the aortic and right ventricular sides. An associated ostium secundum atrial septal defect 3 cm in diameter and tricuspid regurgitation were noted. This atrial septal defect was closed without patch.

Method

Two types of commercially available echocardiographs were used, Aloka SSD-30B (manufactured by Japan Radiation and Medical Electronics Co., Mitaka City, Tokyo) for the first case and Toshiba SSA-03B (manufactured by Tokyo Shibaura Electric Co. Ltd., Kawasaki City, Japan) for the preoperative recordings of the second case. The frequency of ultrasound used was 2.25 MHz at a pulse repetition rate of 1500 Hz in the Aloka SSD-30B and 2 MHz at a pulse repetition rate of 400 Hz in the Toshiba SSA-03B. Transducers were round and flat with a diameter of 10 mm. The experimental azimuth resolving power measured with nylon lines and plastic plates in Aloka SSD-30B was 3 mm at a distance of about 5 cm from the surface of the transducer. Patients were in a supine position breathing normally during the examination and a contact method was adopted for the recording of standard and two-dimensional echocardiography. The surface of the transducer was brought into contact with the skin surface by echo transmission jelly.
For the recording of echocardiograms of case 1, the fast time constant circuit, which enhances echoes from rapidly moving structures, was used, but the time varied gain sensitivity setting, equivalent to sensitivity time control, was not used. In case 2 the sensitivity time control was used so as to clearly visualize the echo from the anterior aortic wall.

The M-mode scan was performed by sector scans, and it was recorded on Polaroid film. The phase-selected two-dimensional echocardiograms were recorded using the electrocardiogram gated circuit. We set the brightening period of the display cathode ray tube at 30 msec. Manual scan was performed with the aid of a guide arm. We held the transducer at one position during the brightening period in each heart beat, and thus showed each brightening period as a single line. About 100 heart beats were required to record a satisfactory picture of the entire heart by this technique. The details of recording these two-dimensional echocardiograms were described in our previous reports.10-12

Results

Echocardiographic examination was carried out in both cases before and after surgical repair. In the first case both standard and two-dimensional echocardiography were performed and in the second case only the standard echocardiography.

1. Echocardiographic Findings in Case 1

PREOPERATIVE ECHOCARDIOGRAPHY (PERFORMED ON NOVEMBER 19, 1973)

Standard Echocardiography

In standard echocardiograms (figs. 1-B,C,D) recorded near the left sternal border in the third intercostal space involving the aortic root, the following abnormalities were observed. With the transducer directed posteriorly and slightly inwards, disappearance of the echo from the anterior aortic wall in diastole and an abnormal thin echo in the right ventricular outflow tract moving almost parallel with the aortic walls from early systole to late diastole were observed. This abnormal thin echo was connected to that of the right coronary cusp of the aortic valve between the echoes from the anterior and posterior aortic walls via rapid anterior excursion of the echo from the right coronary cusp into the right ventricular outflow tract (fig. 1-B). The disappearance of the echo from the anterior aortic wall in diastole was judged to occur when the ultrasound beam passed through the supracristal ventricular septal defect due to the cephalad shift of the base of the heart in diastole (fig. 1-A). The abnormal echo in the right ventricular outflow tract was judged to be that of the aneurysm wall of the Valsalva sinus (fig. 1-A). The mean distance between echoes from the anterior and
posterior aortic walls was 27 mm and that between the aneurysm wall of the Valsalva sinus and the posterior aortic wall was 37 mm. The difference between the two values seemed to approximate the size of the aneurysm. The echoes from the anterior aortic wall and interventricular septum disappeared when the ultrasound beam entered the defect of the supracristal interventricular septum (fig. 1-C). Discontinuity, as indicated by white arrows, was observed in the echo from the aneurysm wall in the right ventricular outflow tract anterior to the right coronary cusp. This discontinuity was judged to arise from the rupture in the aneurysm wall. With a slight caudal rotation of the probe the echo from the interventricular septum was recorded only in diastole and was interpreted as representing the lower margin of the supracristal ventricular septal defect (fig. 1-D). The supracristal ventricular septal defect at the base of the heart in diastole was retracted towards the apex in systole, and the ultrasound beam came to pass through the defect itself. At this moment the right coronary cusp and the aneurysm wall protruding into the right ventricular outflow tract through the ventricular septal defect were also recorded (figs. 1-A, D).

M-mode Scan

An M-mode scan was performed in the directions of the three echocardiograms in figure 1 (fig. 2). The transition of each echo and the discontinuity between the anterior aortic wall and the interventricular septum were well visualized. The recording of an echo from the aneurysm wall of the Valsalva sinus was intermittent; the herniation of the right coronary cusp into the right ventricular outflow tract was visualized.

Two-dimensional Echocardiography

a) Horizontal section: A horizontal two-dimensional echocardiogram was recorded in the left third intercostal space in diastole (fig. 3-A). The prolapsed Valsalva sinus into the right ventricular outflow tract and the supracristal ventricular septal defect (VSD) were visualized. Discontinuity which seemed to represent the rupture in the Valsalva aneurysm was recorded as shown by the white arrow.

b) Section along the long axis of the heart: A two-dimensional echocardiogram along the long axis of the heart was recorded in mid-systole with a probe placed at the left sternal border in the third intercostal space (fig. 3-B). The supracristal ventricular septal defect (VSD), and the aneurysm wall of the Valsalva sinus of the right coronary cusp protruding into the right ventricular outflow tract through this septal defect, were visualized. An interruption in the echo (indicated by a white arrow) was observed on the top of the aneurysm and this was judged to represent the rupture.

POSTOPERATIVE ECHOCARDIOGRAPHIC EXAMINATION
(PERFORMED ON MARCH 23, 1974)

The postoperative echocardiographic examinations were performed as near the preoperative examination sites as possible.

Standard Echocardiography

The preoperatively observed abnormal echo in the right ventricular outflow tract and the discontinuity in the echo from the anterior aortic wall disappeared, and the echocardiogram became simpler as a whole (fig. 4-A). The echo from the anterior aortic wall under the same gain sensitivity setting as in the preoperative recording became stronger than the preoperative recording.

M-mode Scan

In the M-mode scan from the aortic root toward the apex the transition of each echo became simpler. The echo from the anterior aortic wall continued smoothly into that of the interventricular septum without discontinuity, as in healthy persons (fig. 4-B).
Two-dimensional Echocardiography

a) Horizontal section: The right ventricular outflow tract became narrower than before surgery. The echo from the anterior aortic wall showed no discontinuity and was more intense than before surgery (fig. 4-C).

b) Section along the long axis of the heart: The discontinuity between the echoes from the anterior aortic wall and the interventricular septum, and the enlargement of the right ventricular outflow tract, disappeared (fig. 4-D).

2. A Contrasting Echocardiographic Study for Case 1

As a contrasting study for case 1, an M-mode scan (fig. 5-A) and a standard echocardiogram (fig. 5-B) were recorded at the left sternal border in the third intercostal space in a 30-year-old woman with unruptured aneurysm of the Valsalva sinus associated with a supracristal ventricular septal defect. In the M-mode scan interruption in the echo of the interventricular septum and herniation of the right coronary cusp into the right ventricular outflow tract were observed. In the standard echocardiogram a very wide aortic root (compared with the left ventricular outflow tract) and a thin echocurve of the aneurysm wall of the Valsalva sinus (compared with the posterior aortic wall) were recorded. But no discontinuity like that shown in figures 1 and 2 was observed in the echo from the aneurysm wall of the Valsalva sinus in this case.

Figures 3

Two-dimensional echocardiograms of case 1. A) A horizontal section in the left third intercostal space in diastole. B) A section along the long axis of the heart with a probe placed in the third intercostal space at the left sternal border in midsystole. LV = left ventricle. The arrow indicates the portion which was judged as the rupture of the Valsalva aneurysm.
3. Echocardiographic Findings in Case 2

PREOPERATIVE STANDARD ECHOCARDIOGRAPHY (PERFORMED ON NOVEMBER 25, 1971)

Aortic Root

In the standard echocardiogram of the aortic root recorded in the fourth intercostal space slightly lateral to the left midclavicular line with the probe directed posteroinferiorly and medially, a discontinuity was observed in the echo from the anterior aortic wall near the Valsalva sinus, but herniation of the aortic valve was not observed (fig. 6). This discontinuity in the echo from the anterior aortic wall was judged to reflect the introitus of the aneurysm of the right coronary Valsalva sinus and this was confirmed at surgery.

Anterior Tricuspid Leaflet

In the standard echocardiogram of the anterior tricuspid leaflet recorded in the fifth intercostal space between the left midclavicular and parasternal lines with the probe directed posteriorly, enlargement of the right ventricular outflow tract and an abnormal motion pattern of the anterior tricuspid leaflet were recorded (fig. 7). In the echo from the anterior tricuspid leaflet the early diastolic peak was recorded at a lower position than the late diastolic peak, and oscillation was observed in systole and early diastole on the display cathode ray tube (this is difficult to recognize in the photograph due to overexposure). These two findings were judged to reflect the influence of the jet stream from the rupture of the Valsalva aneurysm against the anterior tricuspid leaflet or its subvalvular apparatus.

POSTOPERATIVE ECHOCARDIOGRAPHY (PERFORMED ON MARCH 25, 1974)

In this recording, made two-and-a-half years after surgical correction, the echo from the anterior aortic wall showed no discontinuity (fig. 8-A) and the echo from the anterior tricuspid leaflet was almost normal (fig. 8-B).

Discussion

Cardiac catheterization and retrograde aortography are considered necessary to diagnose ruptured aneurysm of Valsalva sinus and to differentiate it from such other diseases as coronary arteriovenous fistula, atrial septal defect,

Figure 4 Postoperative echocardiograms of case 1. A) A standard echocardiogram corresponding to the preoperative standard echocardiogram of figure 1-C. B) An M-mode scan from the aortic root to the interventricular septum and anterior mitral leaflet corresponding to the preoperative M-mode scan of figure 2. C) A horizontal two-dimensional echocardiogram in the third left intercostal space corresponding to the preoperative echocardiogram of figure 3-A. D) A two-dimensional echocardiogram along the long axis of the heart corresponding to the preoperative echocardiogram of figure 3-B. The abnormal echo and the herniated right coronary cusp of the aortic valve in the right ventricular outflow tract disappeared as did the discontinuity between the echoes from the anterior aortic wall and the interventricular septum.
supracristal ventricular septal defect associated with aortic regurgitation, and ruptured aneurysm of the interventricular septum into the right atrium. Because of our recent echocardiographic studies we now feel that the echocardiographic diagnosis of the ruptured aneurysm of the Valsalva sinus and its differentiation from other diseases are possible.

Furthermore, there are several reports in the literature on echographic abnormalities of the aortic root\textsuperscript{15, 16} and the Valsalva sinus.\textsuperscript{15, 16} In the recent report by Cooperberg et al.,\textsuperscript{16} similar findings to those of our first case are reported except for the detection of the rupture itself.

Congenital aneurysms situated in the right coronary sinus are frequently associated with a defect in the ventricular membranous septum.\textsuperscript{1} The presence of the supracristal ventricular septal defect can be recognized by detecting discontinuity between the anterior aortic wall and the interventricular septum by M-mode scan or two-dimensional echocardiography.

Two-dimensional echocardiography is suitable for defining the course and spread of echoes and the interrelations of neighboring echoes from cardiac structures, while standard echocardiography is suitable for recording only the movement of the echoes from cardiac structures. M-mode scan is a good technique for demonstrating continuity and movement of echoes from the heart, but it has two shortcomings: scanning at varying speeds produces inconsistent visualizations of the spread of cardiac structures, and the angulation of the probe causes the depth of the structures reflecting the echoes (with respect to the chest wall) to be overestimated.
Real-time two-dimensional echocardiographs using mechanical high speed scanning seem to provide a dynamic visualization of the ruptured aneurysm of Valsalva sinus. Phase-selected (stop-action) two-dimensional echocardiography is also useful in visualizing the locations of cardiac structures at a desired phase of the cardiac cycle. The echocardiographic diagnosis of the rupture of the aneurysm of the Valsalva sinus required considerable care. Especially in detecting defects in cardiac structures, differentiation between technical drop-outs and real structural discontinuities is important. For this differentiation the compound scan is important. In this method the ultrasound beam comes from many different angles and positions and technical drop-outs can be avoided. This study can be facilitated by comparing cases with defects demonstrated by other methods with cases without defects under the same condition. Comparing the preoperative echocardiograms with postoperative ones is often also helpful. We also have to carefully use the gain control circuit, such as the time varied gain sensitivity settings, i.e. sensitivity time control, and the fast time constant circuit (FTC), in order to avoid artifacts in the echo. The FTC is used to enhance the echoes from rapidly moving structures.

In diagnosing the ruptured aneurysm of Valsalva sinus, for example, excessive use of near gain control should be avoided, and repeated recordings of the echo tracing of the aneurysm wall at slightly different positions are required. If the periodic discontinuity in the echo tracing of the aneurysm wall is a constant finding over many cardiac cycles, it can be judged to indicate the rupture in the aneurysm wall. In this instance the size of the rupture must be larger than the azimuth resolving power of the equipment at that depth. The experimental azimuth resolving power of the present equipment is 3 mm and the diameter of the rupture in each of the present cases was 5 mm. There have been other reports of ruptures with diameters exceeding 5 mm. In two reports the longer diameter exceeded 15 mm; such ruptures should be easily detectable by echocardiography.

The same is true in detecting ventricular septal defects. King et al.9 have also referred to the technical drop-outs in the diagnosis of ventricular septal defect by two-dimensional echocardiography. They emphasized that recognition of the significance of an absent echo signal in a given location requires a thorough understanding of cardiac anatomy and physical and technical principles of ultrasonic imaging. They also pointed out that most larger ventricular septal defects occur in the septum immediately below the posterior semilunar valve, the portion of the septum most easily visualized by cardiac ultrasonography.

The tricuspid valve flutter observed in our second case in systole and in early diastole was not observed in our first case. Nanda et al. have recently reported on the systolic flutter of the tricuspid valve in two cases with congenital left ventricular-right atrial communication17 and mentioned that its mechanism was not clear. We speculate that the deficiency in the tricuspid valve plays an important role in the production of this systolic flutter.

They have also mentioned the diastolic flutter of the tricuspid valve in pulmonary incompetence and large atrial septal defects. They explained this flutter as probably due to the regurgitant jet abutting the open leaflet. We also observed recently a clear diastolic flutter of the anterior tricuspid leaflet in pulmonary incompetence. But in our experience with 22 cases with ostium secundum atrial septal defect11 no obvious flutter was observed. At present we would like to attribute the systolic and early diastolic flutter of the tricuspid valve to the jet stream blowing directly against the closed and opening leaflet.

References
Right Ventricular Growth in a Case of Pulmonary Stenosis with Intact Ventricular Septum and Hypoplastic Right Ventricle

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SUMMARY Adequate growth of the hypoplastic right ventricle in a patient with severe pulmonary stenosis with an intact ventricular septum was documented after pulmonary valvotomy in infancy. It is postulated that the growth of the ventricular chamber is largely the result of pulmonary regurgitation resulting from successful pulmonary valvotomy. Based on this and the observations of others on the growth of the hypoplastic right ventricle in pulmonary atresia (with intact septum) cases, an organized approach to eventual total surgical correction is recommended.

SIGNIFICANT PULMONARY STENOSIS with an intact ventricular septum is usually associated with a hypertrophied right ventricle. Only rarely is this lesion associated with a hypoplastic right ventricle. The long-term prognosis in these cases, and in cases with pulmonary atresia with an intact ventricular septum and hypoplastic right ventricle, depends upon initial successful palliation and ultimate growth of the right ventricular cavity to a reasonable size, so as to handle an adequate cardiac output. The purpose of this communication is to illustrate the growth of the hypoplastic right ventricle in a case of severe pulmonary stenosis with an intact ventricular septum, to discuss its therapeutic implications in the management of hypoplastic right ventricle (with pulmonary stenosis or atresia), and to comment on an interesting physiologic finding in this case, namely interatrial right-to-left shunting in the presence of a normal right ventricular pressure.

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