Two-dimensional echocardiography in diagnosing tricuspid atresia¹

Differentiation from other hypoplastic right heart syndromes and common atrioventricular canal

SHINTARO BEPPU, YASU HARU NIMURA, MASAHICO TAMAI, SEIKI NAGATA, HIROHIDE MATSUO, YASUNARU KAWASHIMA, TAKAHIRO KOZUKA, AND HIROSHI SAKAKIBARA

From the Research Institute, the Department of Medicine and the Department of Radiology, National Cardiovascular Center, and the First Department of Medicine and the First Department of Surgery, Osaka University Medical School, Osaka, Japan

SUMMARY Echocardiographic studies were performed in 8 patients with tricuspid atresia, 3 patients with other types of hypoplastic right heart syndromes, and 5 patients with common atrioventricular canal. Echocardiographic searching was performed mainly along the horizontal section at the level of the membranous septum of the heart. Conventional and two-dimensional (cross-sectional) echocardiograms were recorded. The findings were compared with those from the echocardiograms of 25 healthy subjects, 17 patients with mitral valve disease, and 3 patients with congestive cardiomyopathy who served as a control group as they had normal arrangements of the intracardiac structures.

In the control group, the annular ends of the anterior mitral and tricuspid valve leaflet echoes were the interatrial septum and the right anterior wall of the heart echoes, respectively.

In tricuspid atresia, a thick echo was detected in the tricuspid area. This echo did not open during diastole and was interpreted as indicating the atretic part of the tricuspid orifice. The anterior leaflet of the detectable single atrioventricular valve echo was attached to the interatrial septal echo.

In hypoplastic right heart syndromes other than tricuspid atresia, findings such as a minute right ventricle and a large left ventricle resemble those in tricuspid atresia. However, the tricuspid valve echo was detected and indicated to be open on the two-dimensional echocardiogram.

In common atrioventricular canal, the anterior leaflet of the detectable single atrioventricular valve echo was attached to the right anterior wall echo of the heart. The echo of the interventricular septum was detected in the apical area in 2 patients and not detected at all in the others.

As mentioned above, the echocardiographic features along the horizontal section were different among the control group, those with tricuspid atresia, hypoplastic right heart syndromes apart from tricuspid atresia, and common atrioventricular canal, and are thought to correspond to the anatomical differences.

Tricuspid atresia is defined as congenital complete absence of the right atrioventricular valve with hypoplasia of the right ventricle (Edwards and Burchell, 1949; Astley *et al.*, 1953; Keith *et al.*, 1967). As half of the patients die in the first 6 months of life (Keith *et al.*, 1967; Dick *et al.*, 1975), it is

desirable to diagnose this malformation exactly as soon as possible in the early days of life. While the conventional non-invasive examinations such as electrocardiography and chest x-ray film offer diagnostic information for this malformation, there are some cases in which these conventional examinations do not produce the characteristic findings (Kroop and Grishman, 1950; Gamboa *et al.*, 1966; Dick *et al.*, 1975). Therefore, the exact diagnostic method is angiocardiology. However, as an

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invasive examination carries some risk to the newborn, a new non-invasive method for diagnosing cardiac malformations is needed.

Echocardiography has proved to be a useful aid for diagnosing congenital heart diseases (Lundström and Edler, 1971; Dillon *et al.*, 1973; Nimura *et al.*, 1974; Popp *et al.*, 1974; Solinger *et al.*, 1974; Matsumoto *et al.*, 1975; Murphy *et al.*, 1975). In tricuspid atresia, a small right ventricle, a large left ventricle, and an absent tricuspid valve echo are noticed to be the characteristic features in the echocardiogram (Chesler *et al.*, 1970; Meyer and Kaplan, 1972; Godman *et al.*, 1974). However, as an absent tricuspid valve echo does not always mean an absent tricuspid valve, it is difficult to distinguish tricuspid atresia from other hypoplastic right heart syndromes (Godman *et al.*, 1974). Moreover, unless the ventricular septal echo is identified on the echocardiogram, the findings resemble those in cases of a single ventricle with common atrioventricular canal (Chesler *et al.*, 1970). The aim of this study is to differentiate tricuspid atresia from other malformations based on anatomical points using M-mode scan and two-dimensional echocardiography.

Subjects and methods

Eight patients with tricuspid atresia, aged from 2 months to 22 years, were examined. The diagnoses were confirmed by angiocardiology, showing that 5 patients were type Ib and the other 3 patients were type Ia, Ic, and IIc, respectively (Keith *et al.*, 1967). The atrial septal defect was large in all patients. All were treated by a shunt operation except for one. The patient with type IIc had a comparatively large right ventricle for tricuspid atresia.

Patients with hypoplastic right heart syndromes apart from tricuspid atresia, and those with common atrioventricular canal, and certain other subjects were also examined for comparison. There were 3 patients with hypoplastic right heart syndromes without tricuspid atresia; 2 of them, ages 6 months and 7 years, had pulmonary atresia with normal aortic root, and the other, age 22 years, had pulmonary stenosis, tricuspid stenosis, and atrial septal defect. The diagnoses were confirmed by catheterisation and angiocardiology. There were 5 patients with common atrioventricular canal, ranging in age from 6 months to 6 years; angiography showed that 3 of them had no interventricular septum. As controls, the other subjects, without intracardiac malformations, were 25 healthy children and adults, 17 patients with mitral valve disease, and 3 patients with congestive cardiomyopathy.

A commercially available ultrasonograph, an Aloka SSD-30B, was used with a 2.25 MHz transducer, 10 mm in diameter, with a pulse repetition rate of 1500 Hz. This equipment allows one to record both conventional and two-dimensional echocardiograms by switching modes of display. The two-dimensional echocardiogram was obtained by an electrocardiograph-triggered B-Mode method (Ebina *et al.*, 1967; King, 1973). This method has been described previously by Beppu *et al.* (1976).

The patients were examined in the supine position, during relaxed respiration. As the scan was a manual compound one, the echocardiographic searching could be made from various places along the selected section plane in the chest wall, even from different interspaces or from the right sternal border (Fig. 1). The transducer was usually placed in the left intercostal space and also on the right interspace and the sternum, if necessary, for the echoes to be obtained clearly. The horizontal section at the level of the membranous septum of the heart was selected for the echocardiographic scans, as previously reported (Beppu *et al.*, 1976).

Fig. 1 The transducer is attached to the end of the guide arm giving reference to its position and angle. As the scan is a manual compound one, the transducer can be placed in various sites on the chest wall to record echoes of a wide area of the heart.

Fig. 2 The normal relation between intracardiac structures in the horizontal section at the level of the membranous septum of a 63-year-old woman with congestive cardiomyopathy. Left, a two-dimensional echocardiogram, viewed from caudal, in late systole. The interventricular and interatrial septa, and the mitral and tricuspid valves are seen. The anterior mitral leaflet continues into the interatrial septum without interruption. The anterior tricuspid leaflet echo runs from the right anterior to the membranous portion. Right, a schematic drawing of the horizontal section of the heart. IVS, interventricular septum; IAS, interatrial septum; TV, tricuspid valve; AMV, anterior mitral valve leaflet; MEMB SEPT, membranous septum; RV, right ventricle; RA, right atrium; LV, left ventricle; LA, left atrium. (From Beppu et al., 1976, *British Heart Journal*, 38, 911.)

Results

The two-dimensional echocardiographic features at the level of the membranous septum in cases without malformations were the same as those previously described (Beppu et al., 1976).

In cases without malformations, the horizontal two-dimensional echocardiogram showed the inter-

ventricular and the interatrial septa and the mitral and the tricuspid valve echoes (Fig. 2). Each echo seemed to lead towards a central site. The site where these echoes meet is the membranous septum, but the echo of this part was not usually recorded clearly. Because the membranous septum lies almost perpendicular to the chest wall and is almost parallel to the ultrasound beam, the ultrasound is

Fig. 3 The two-dimensional echocardiogram of a 7-year-old girl with surgically repaired atrial septal defect; the right ventricular cavity is still large, viewed from the caudal, in a case with normal relations between intracardiac structures. Left, during systole, the tip of the anterior tricuspid valve leaflet echo attaches to the membranous portion. Right, during diastole, the tip of the anterior tricuspid valve leaflet echo separates from the membranous portion.

Fig. 4 The horizontal echograms in tricuspid atresia in a 3-year-old boy. Left, during systole, the anterior mitral valve leaflet echo was in the closing position. The echo of the anterior mitral valve leaflet continued into the interatrial septum echo as in normal cases. The interatrial septum echo ended abruptly in the atrial area, demonstrating atrial septal defect (indicated by the white arrow on the left). From the membranous portion to the right anterior wall of the heart, a thick echo was recorded. In this patient, the echo of the interventricular septum was not recorded near the membranous septum, demonstrating ventricular septal defect (indicated by the white arrow on the right). Right, during diastole, the anterior mitral valve leaflet was in the opening position. However, a thick echo in the area corresponding to the tricuspid orifice did not move or separate from the membranous portion. AT, atretic part of the tricuspid orifice.

not reflected sufficiently. The echo of the anterior mitral valve leaflet was always continued into the echo of the interatrial septum. The echo of the anterior tricuspid valve leaflet ran from the anterior wall of the heart to the medial end of the interventricular septum echo. The two-dimensional echocardiogram during diastole showed the tip of the anterior tricuspid leaflet separated (or opened) anteriorly from the membranous portion (Fig. 3).

(1) TRICUSPID ATRESIA

Echocardiography indicated a small right and a large left ventricle in all patients except the one with type IIc. Two-dimensional echocardiography along the horizontal section at the level of the membranous septum showed that the echo of the anterior mitral leaflet continued into the interatrial septum as in normal subjects (Fig. 4). A thick echo, between the annular end of the anterior mitral valve leaflet

Fig. 5 M-mode scan from right to left at the level of the membranous septum of the heart. On the right side of the heart, the anterior wall of the heart moved posteriorly during diastole. Scanning leftward, this echo separated from the anterior wall increasing its amplitude. Behind this echo, the interatrial septum echo suddenly appeared, joining up with the anterior mitral valve leaflet echo. The echo recorded from the right anterior wall of the heart to the annular end of the anterior mitral valve leaflet did not show the normal anterior movement of the tricuspid valve during diastole.

Fig. 6 In 2 patients with tricuspid atresia, the echo of the atretic part of the tricuspid orifice showed a tiny two-peaked pattern during diastole.

and the right anterior wall of the heart, was detected. The anatomical site of this echo corresponded to that of the anterior tricuspid leaflet in normal subjects. However, the echo did not separate from the membranous portion during diastole (Fig. 4, right). The echo of the interatrial septum ended abruptly in the atrial area, indicating atrial septal defect (Matsumoto *et al.*, 1975; Beppu *et al.*, 1976).

M-mode scan along the horizontal section showed the same findings as were seen in two-dimensional echocardiography. On the right, the anterior wall of the heart moved posteriorly during diastole. From there to the annular attachment of the anterior mitral valve leaflet, a thick echo with a wavy pattern was recorded. This echo

corresponded to the tricuspid valve in a normal heart; however, it did not move like the atrio-ventricular valve. Behind this echo, the interatrial septum echo appeared abruptly and continued to the left to the anterior mitral valve leaflet echo (Fig. 5). The echo between the right anterior wall and the membranous portion showed no anterior movement during diastole in all except 2 patients with tricuspid atresia, and in these 2 patients, a tiny two-peaked pattern was seen during diastole (Fig. 6).

In the patient with type IIc, whose right ventricle was fairly large, standard echocardiograms showed that the sizes of both the ventricles were comparatively balanced, as seen in healthy subjects (Fig. 7). In the right ventricular cavity, an echo moving anteriorly during diastole and posteriorly during systole was detected (Fig. 7). This motion was identical to that of an atrioventricular valve leaflet. Therefore, this echocardiographic finding apparently suggested that the tricuspid valve did exist and function in this patient. However, the horizontal two-dimensional echocardiograms showed the same findings among all the patients (Fig. 8). A thick echo was detected in the area corresponding to the tricuspid annulus and did not open during diastole. The echo moving like a tricuspid leaflet, mentioned above, was recorded when the ultrasound beam was directed to the middle portion of the right ventricle. As this patient had a large ventricular septal defect, a subvalvular apparatus, like the chordae tendineae, was being moved by the blood through this defect, not through the tricuspid orifice.

The above echocardiographic features along the horizontal section were detected in all patients examined except one whose membranous portion could not be recorded because of the interference from the sternum or the lung.

Fig. 7 Conventional echocardiogram in a patient with tricuspid atresia type IIc. The right ventricle was comparatively large. In this cavity, an echo moving anteriorly during diastole and posteriorly during systole was detected ("TV?" in figure). This motion was identical to that of the atrioventricular valve leaflet, and, therefore, this echo apparently suggests the tricuspid valve.

Fig. 8 Horizontal echograms in a patient, a 22-year-old man, with tricuspid atresia type IIc (same patient as in Fig. 7). Left, during systole, the echo of the anterior mitral valve leaflet was closing. Between the membranous portion and the right anterior wall of the heart, a thick echo was detected. Right, during diastole, this echo did not move, as in other patients with tricuspid atresia. The conventional echocardiogram of Fig. 7 was recorded when the ultrasound beam was directed at the middle portion of the left ventricle, indicated by the white line.

(2) HYPOPLASTIC RIGHT HEART SYNDROMES OTHER THAN TRICUSPID ATRESIA

In hypoplastic right heart syndromes without tricuspid atresia, a minute right ventricle and a large left ventricle were recorded by conventional echocardiography. Those features were identical to those in tricuspid atresia. Scanning toward the right of the heart, however, an echo of the anterior tricuspid valve leaflet was detected (Fig. 9, left).

Its motion was so tiny that it was difficult to know whether the valve functioned and opened or not, since even in tricuspid atresia, the atretic part of the tricuspid orifice moved anteriorly slightly during diastole in 2 patients. The two-dimensional echocardiogram along the horizontal section, however, showed that the tip of the anterior tricuspid leaflet echo was separated from the medial end of the interventricular septal echo during diastole (Fig. 9,

Fig. 9 Echograms in a 6-year-old girl with atrial septal defect and persistent ductus arteriosus in addition to pulmonary atresia with normal aortic root. Left, an echo of the anterior tricuspid valve leaflet was recorded. It moved anteriorly slightly during diastole. Right, horizontal echogram at early diastole. A white line indicates the beam direction by which the conventional echocardiogram on the left was recorded. The tip of the anterior tricuspid valve leaflet echo separated from the medial end of the interventricular septal echo, indicating that the tricuspid valve opened during diastole. The interatrial septal echo ended abruptly in the atrial area, indicating the atrial septal defect.

Fig. 10 Horizontal echocardiograms in a 5-year-old girl with single atrium, persistent ductus arteriosus, and pulmonary atresia in addition to common atrioventricular canal. The echocardiograms on the left and the right are those at early systole and at mid diastole, respectively. This patient had the interventricular septum near the apical area and its echo was recorded on the echogram. The attachment site of the anterior leaflet of the only detectable atrioventricular valve was the right anterior wall of the heart. CommV, common atrioventricular valve; SA, single atrium.

right). The anterior mitral leaflet echo was connected to the interatrial septal echo as in normal subjects. The echo of the interatrial septum ended abruptly in the atrial area, indicating atrial septal defect (Matsumoto *et al.*, 1975; Beppu *et al.*, 1976) (Fig. 9).

(3) COMMON ATRIOVENTRICULAR CANAL

In common atrioventricular canal, the conventional echocardiogram showed a large ventricle and a single atrioventricular valve echo. In the patients with an interventricular septum, the septal echo was recorded only in the apical area. On the two-dimensional echocardiograms along the horizontal section at the level of the membranous septum, however, the anterior leaflet echo of the sole atrioventricular valve was attached to the anterior wall of the heart directly (Fig. 10). These echocardiographic features were detected in 3 of the 5 patients with this malformation. In the others, the attachment site of the anterior leaflet echo was not detected because of interference from the sternum and/or lung.

Discussion

Echocardiography, especially the two-dimensional method, is useful in diagnosing congenital heart diseases, as it shows non-invasively the anatomical relations between the intracardiac structures (Tanaka *et al.*, 1971; King, 1973; Nimura *et al.*, 1974; Sahn *et al.*, 1974; Matsumoto *et al.*, 1975; Beppu *et al.*, 1976). In tricuspid atresia, a minute right

ventricle, a large left ventricle, and an absent tricuspid valve echo have been reported as characteristic features in the conventional echocardiogram (Chesler *et al.*, 1970; Godman *et al.*, 1974; Meyer and Kaplan, 1972). However, the first two findings are to be expected in hypoplastic right heart syndromes (Godman *et al.*, 1974), and the third finding does not always mean that the tricuspid valve does not exist. Moreover, if the interventricular septal echo is not recorded, it seems impossible to diagnose tricuspid atresia, because common atrioventricular canal with single ventricle offers similar findings (Chesler *et al.*, 1970).

On the other hand, in a specific case, as with the patient with type IIc who had a comparatively large right ventricle, the conventional echocardiogram showed that the right ventricle was not minute, and the echo apparently suggested that the tricuspid valve was recorded in the right ventricular cavity. Therefore, it seems to be impossible to diagnose tricuspid atresia from the criteria mentioned above.

In this study, echocardiographic differences between tricuspid atresia, hypoplastic right heart syndromes apart from tricuspid atresia, and common atrioventricular canal were investigated using two-dimensional echocardiography from the anatomical points of view (Fig. 11).

In cases with normal anatomical relations between intracardiac structures, the annular attachment of the anterior mitral and tricuspid valve leaflet echoes were the interatrial septum and the

Fig. 11 Schematic drawings of the horizontal section of the normal heart (NORMAL), tricuspid atresia, imperforate tricuspid valve (IMPERFORATE TV), hypoplastic right heart syndromes (HYPO RIGHT HEART), and common atrioventricular canal (COMMON AV CANAL). These drawings indicate the differences of the attachment site of the functioning atrioventricular valve among these malformations. In tricuspid atresia, including imperforate tricuspid valve, the functioning atrioventricular valve is the mitral valve and its anterior leaflet connects with the interatrial septum as in normal cases. The interatrial septum is defective in every case of tricuspid atresia. In hypoplastic right heart syndromes, the right ventricle is hypoplastic; however, the tricuspid valve does exist. In common atrioventricular canal, the anterior common valve leaflet is formed by fusion of the anterior part of the anterior mitral valve leaflet and a part of the tricuspid valve. Therefore, its leaflet attaches to the anterior wall of the heart. As common atrioventricular canal is the extreme form of endocardial cushion defect, there is no connection between the interatrial septum and any leaflet of the common valve. *, imperforate membrane; CV, common atrioventricular valve.

right anterior wall of the heart, respectively (Beppu *et al.*, 1976).

In tricuspid atresia, including imperforate tricuspid valve, anatomically, the functioning atrioventricular valve is the mitral valve. Therefore, echocardiographically, its anterior leaflet echo should connect with the interatrial septum echo, as in normal cases. In the present study, the echo of the anterior leaflet of the detectable single atrioventricular valve was attached to the interatrial septal echo as in normal cases. Thus, it was decided that this valve was not the tricuspid valve or the common atrioventricular valve, but rather the mitral valve. In the tricuspid area between the membranous portion and the right anterior wall of the heart, a thick echo was detected. Since it did not open during diastole, it was interpreted to be the structure between the right atrium and the ventricle, though this echo might include those of the atrioventricular part of the membranous septum and the ridge between the right atrium and the ventricle. Such echocardiographic findings obtained are considered to be specific to tricuspid atresia. Anderson *et al.* (1977), on the basis of their morphological study, emphasised that imperforate tricuspid valve should be distinguished from tricuspid atresia. It might be difficult clinically to distinguish imperforate tricuspid atresia from muscular atresia. However, in the patient with type IIc, the findings of the area corresponding to the tricuspid annulus were the same as those in the other patients examined. His right ventricle was fairly large and the conventional echocardiogram indicated the echo apparently suggesting the tricuspid valve in the right ventricular cavity. The

echo in the right ventricular cavity should be the subvalvular tissue such as chordae tendineae. This finding indicates that this structure is an imperforate tricuspid valve (Anderson *et al.*, 1977). Thus echocardiography is considered to be advantageous in distinguishing the two types of tricuspid atresia from each other. If the feature of the atretic part of the tricuspid orifice is shown to be membranous and the annulus to be enough large for valve implantation, it might be possible for the atretic part to be excised and a new but normal circuit to be established. Two-dimensional echocardiography should offer useful information about surgical treatment.

Every case with tricuspid atresia has atrial septal defect. In the present study, the interatrial septal echo ended abruptly in the atrial area. This echo interruption is interpreted as indicating the atrial septal defect (Matsumoto *et al.*, 1975; Beppu *et al.*, 1976). If the size of the defect of the atrial septum is small in tricuspid atresia, atrial septostomy is necessary immediately after birth. Horizontal two-dimensional echocardiography may give the necessary information.

In hypoplastic right heart syndromes apart from tricuspid atresia, echocardiographic features such as a small right ventricle and a large left ventricle were similar to those in tricuspid atresia. In our patients, the echo of the anterior tricuspid valve leaflet could be detected in the area of the tricuspid orifice by conventional echocardiography, and its motion was more definite than in tricuspid atresia. However, it could not be determined whether the tricuspid valve did open or not. The two-dimensional echocardiogram during diastole showed clearly

that the tip of the anterior tricuspid leaflet echo was opened, separated from the medial end of the interventricular septum echo.

In common atrioventricular canal, the conventional echocardiogram showed similar findings to those in tricuspid atresia in the point of a single atrioventricular valve echo. In order to differentiate the two, it is essential to identify the detectable single valve as either the mitral valve or the common valve. Common atrioventricular canal is an extreme form of endocardial cushion defect. In endocardial cushion defect, as the endocardial cushion is not developed, there is no continuity between the anterior mitral valve leaflet and the interatrial septal echoes (Beppu *et al.*, 1976). Moreover, anatomically, the anterior leaflet of the common atrioventricular valve is formed from the fusion of the anterior part of the anterior mitral valve leaflet and the anterior part of the tricuspid valve. Specifically, the anterior leaflet of the common atrioventricular valve attaches to the anterior wall of the heart, in the same way as the anterior tricuspid valve leaflet does in normal subjects (Fig. 11). There is no connection between the interatrial septum and any leaflet of the common valve. In this study, on the horizontal two-dimensional echocardiogram, the annular attachment site of the anterior leaflet of the common valve was shown to be the anterior wall of the heart. This configuration corresponds to the above-mentioned anatomy of this malformation.

In this study, the site of the annular attachment of the anterior leaflet echo could not be detected in one patient with tricuspid atresia and in two patients with common atrioventricular canal. In these patients, the sternum and/or lung disturbed the echocardiographic search for the site concerned. However, as the sternum is not calcified in newborns, the echocardiograms can be obtained satisfactorily through the sternum. Therefore, it should be easier to search for the attachment site of the detectable sole atrioventricular valve when the examinee is a newborn.

In summary, echocardiographic differentiation between tricuspid atresia and common atrioventricular canal is based on the difference of the attachment site of the anterior leaflet echo of the detectable single atrioventricular valve (Fig. 11). When it is the interatrial septum, the valve is the mitral valve and the malformation is tricuspid atresia. When it is the anterior wall of the heart, the valve is the common valve and the malformation is common atrioventricular canal. In hypoplastic right heart syndromes apart from tricuspid atresia, the tricuspid valve is recorded as opening during diastole. The above differentiation can be more clearly

performed by two-dimensional echocardiography than by conventional echocardiography.

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Requests for reprints to Dr Shintaro Beppu, National Cardiovascular Center, 5-125 Fujishirodai, Suita-city, Osaka 565, Japan.