Echocardiographic Diagnosis of Endocardial Cushion Defects

Jyunichi YOSHIKAWA, M.D., Takane OWAKI, M.D., Hiroshi Kato, M.D., Yasuhiko Tomita, M.D., Kunizo Baba, M.D., and Kumeo Tanaka, M.D.

SUMMARY

Sixteen patients with endocardial cushion defects were studied by echocardiography and ultrasonocardiotomography. Three characteristic findings, common to both complete and partial forms, were observed. They were the ostium primum defect of the interatrial septum, the left ventricular outflow narrowing and the atrioventricular valve abnormalities which are the anatomic bases of endocardial cushion defects. Several additional findings were observed in patients with complete form. Most strikingly, the anterior leaflet echo locating in the left ventricle had the direct connection to that in the right ventricle without the interposition of the interventricular septum by echocardiogram. Ultrasonocardiogram, on the other hand, demonstrated beautifully the common anterior leaflet and the high interventricular septal defect. There were, in addition, the absence of the paradoxical motion of the interventricular septum and the difficulty in detecting the interatrial septum.

The intact interatrial septum continuous with the anterior mitral leaflet was demonstrated in patients with the interventricular septal defect of atrioventricular canal type, in which there were the typical abnormalities in the atrioventricular valves and the left ventricular outflow narrowing as well.

This study indicates that ultrasonic examination may provide a direct, yet noninvasive method in the diagnosis of various types of endocardial cushion defects.

Additional Indexing Words:
Endocardial cushion defects Ostium primum defect of the interatrial septum Atrioventricular valve abnormalities High interventricular septal defect Common anterior leaflet

Endocardial cushion defects (ECD) is not an uncommon congenital anomaly. Cardiac catheterization and angiography have been necessary to establish the definitive diagnosis of ECD. Ultrasonic techniques, however, have developed rapidly in recent years and have been well established

From the Department of Cardiology, Kobe Municipal Central Hospital, Kanocho 1-5, Ikutaku, Kobe.
Received for publication May 17, 1974.
as noninvasive methods in the diagnosis of congenital anomalies. The purpose of this report is to describe our experiences in the ultrasonic examination of patients with various types of ECD and to show the ultrasonic findings consistent with the ostium primum defect of the interatrial septum, the atrioventricular valve abnormalities and the left ventricular outflow narrowing. Furthermore, it is attempted to distinguish complete form from partial form by detecting the common anterior leaflet and the high interventricular septal defect.

**Materials and Methods**

Sixteen patients with ECD diagnosed by cardiac catheterization and angiocardiography were studied by echocardiography and ultrasonocardiotomography. Seven were males and 9 were females with the age ranging from 3 months to 32 years. Five patients had complete form, 4 partial form and 2 special form having the interventricular septal defect without the ostium primum defect of the interatrial septum. This special form is called generally the interventricular septal defect of atroventricular canal type (VSD of ECD type). Anatomic detail was obtained in all patients with partial form and special form at corrective surgery and in 2 with complete form at autopsy.

All ultrasonic examinations were carried out using echocardiograph Model Aloka SSD-90 and a 2.25 megacycle transducer. To obtain ultrasonocardiotomograms the stop action techniques utilizing an ultrasonic compound contact scanner with an unit for synchronization with an electrocardiogram was used. The mitral valve was first located from a left sternal position, utilizing the fourth intercostal space. Medial rotation of the transducer from the mitral valve position (M-mode transverse scan and transverse ultrasonocardiotomogram) passing the beam through the tricuspid valve and the interatrial septum originating from the anterior mitral leaflet was performed. The detection of the interatrial septum is easily possible in cases with the right ventricular dilatation including the ostium secundum defect of the interatrial septum. Emphasis was placed on detecting the continuity between the anterior mitral leaflet and the interatrial septum, and in addition between the anterior mitral and tricuspid leaflets.

**Results**

1) Partial Form

Atrioventricular valves and interventricular septum;

The most striking finding in mitral valve echoes was abnormal multiple diastolic echoes composed of 2 anterior and 1 posterior echoes (Fig. 1). To detect this finding, an intense examination was necessary because the beam direction was grossly restricted frequently. Abnormal anterior mitral leaflet proximity to the interventricular septum was observed. On the other hand,
Fig. 1. Echocardiogram (left panel) and ultrasonocardiogram (right panel) of a case with partial form of endocardial cushion defects. The ultrasonocardiogram was taken at mid-diastolic phase. Most importantly, abnormal multiple diastolic echoes of the mitral valve is observed. Note the mitral anterior leaflet proximity to the interventricular septum, the paradoxical motion of the interventricular septum and the right ventricular cavity dilatation. MV = mitral valve, TV = tricuspid valve, IVS = interventricular septum, LVPW = left ventricular posterior wall, LV = left ventricle, RV = right ventricle, La = left atrium.

the anterior tricuspid leaflet crossed the interventricular septum into the left ventricular cavity.

There was no direct connection between the anterior mitral and tricuspid leaflets. The anterior mitral leaflet was possible to connect with the anterior

Fig. 2. Echocardiographic M-mode scan of a case with partial form of endocardial cushion defects with the beam moving medially from the mitral valve position. The mitral anterior leaflet (MV) is not continuous with the tricuspid anterior leaflet (TV). The interatrial septum (IAS) is recorded, but not continuous with the anterior mitral leaflet.
tricuspid leaflet with the interposition of the interventricular septum (Fig. 2).

There were the dilated right ventricle, the paradoxical motion of the interventricular septum and increased excursion of the anterior tricuspid leaflet similar to those of the ostium secundum defect of the interatrial septum.

Continuity between the anterior mitral leaflet and the interatrial septum;

The distinct interatrial septum was detected in 5 cases in which the anterior mitral leaflet was not continuous with the interatrial septum (Fig. 2). Ultrasonocardiogram demonstrated it more clearly and the posterior situation of the interatrial septum as well (Fig. 3).

2) Complete Form

Atrioventricular valves and interventricular septum;

There were the findings consistent with the common anterior leaflet, the high interventricular septal defect and the absence of the paradoxical motion of the interventricular septum which were not observed in cases with partial form. When the transducer was located in the mitral valve position, the interventricular septum moved in a direction opposite to the posterior left ventricular wall during ventricular ejection and the valve locating in the left

![Fig. 3. Ultrasonocardiogram of a case with partial form of endocardial cushion defects. Note the discontinuity between the mitral anterior leaflet (MV) and the interatrial septum (IAS). This could show the ostium primum defect of the interatrial septum. The ultrasonocardiogram was taken at mid-diastolic phase.](image-url)
ventricle appeared in abnormal multiple diastolic echoes (Fig. 4). The latter finding was easily detected in contrast with partial form. Abnormal anterior leaflet proximity to the interventricular septum was noticed as well. When the transducer was directed medially, the systolic disappearance of the interventricular septal echo was recorded by echocardiogram (Fig. 5). The anterior leaflet echo locating in the left ventricle had the direct connection to the
Fig. 6. Echocardiographic M-mode scan of a case with complete form of endocardial cushion defects. Note the straight connection between the anterior leaflet echo locating in the left ventricle and that in the right ventricle.

Fig. 7. Echocardiographic M-mode scan of a case with complete form of endocardial cushion defects with the beam moving medially from the mitral valve position (left panel) and adjusted slightly to detect the clear echo of the inferior segment of the common anterior leaflet in the right ventricle (left panel). Two anterior leaflet echoes are observed in the right ventricle.

The interatrial septum was not detected in all cases.
3) Special Form (VSD of ECD type)
Atrioventricular valves and interventricular septum;
The same abnormalities as in partial form were observed, but we were

Fig. 8. Ultrasonocardiotomogram (left panel) and ultrasonocardiokymogram (right panel) of a case with complete form of endocardial cushion defects. Ultrasonocardiotomogram demonstrates a single anterior leaflet (common anterior leaflet; CAL) and a large interventricular septal defect (white arrows). The same findings are obtained by ultrasonocardiokymogram. It is easy to recognize the echo sources in ultrasonocardiotomogram by ultrasonocardiokymogram. The ultrasonocardiotomogram was taken at systole. LV=left ventricle, RV=right ventricle, IVS=interventricular septum, LA=left atrium.

Fig. 9. Echocardiogram (left panel) and ultrasonocardiotomogram (right panel) of a case with interventricular septal defect of atrioventricular canal type. The interatrial septum (IAS) is detected and continuous with the anterior mitral leaflet (MV). The anterior mitral leaflet proximity to the interventricular septum (IVS) is observed. Both fail to demonstrate a definitive interventricular septal defect. The ultrasonocardiotomogram was taken at systole. TV=tricuspid valve, LV=left ventricle, LA=left atrium.
unsuccessful to demonstrate the definitive interventricular septal defect.
Continuity between the anterior mitral leaflet and the interatrial septum;
The interatrial septum was detected clearly. It was continuous with
the anterior mitral leaflet and located between the anterior tricuspid leaflet
and the posterior margin of the left atrium (Fig. 9).

**Discussion**

ECD is primarily due to a developmental defect of the atroioventricular
docardial cushions. If fusion of the cushions fails completely, the atrioventri-
cular ostia form a large and single ostium, and there is a large, central septal
defect which allows free communication between all 4 chambers (complete
form). If the cushions fuse only centrally, there is a division of the atroioventri-
cular canal into right and left atroioventricular ostia, though the mitral valve,
and often medial tricuspid leaflet, is cleft (partial form). The interatrial
septal defect varies in size from complete absence of the septum to the absence
of inferior part of the septum immediately adjoining the atroioventricular valve.
Considering the above mentioned intracardiac abnormalities, ultrasonic diag-
osis of ECD is to detect 1) the ostium primum defect of the interatrial septum,
2) the atroioventricular valve abnormalities and the left ventricular outflow
narrowing resulting from the abnormality in the mitral valve and its attach-
ment, 3) the high interventricular septal defect.

The previous reports\(^3\)-\(^7\) showed the echocardiographic findings con-
sistent with 2). They are the abnormal multiple diastolic echoes of the mitral
valve, the anterior tricuspid leaflet’s crossing the interventricular septum into
the left ventricle and the anterior mitral leaflet proximity to the interventri-
cular septum. The abnormal diastolic echoes, in our experience, were com-
posed of 2 anterior and 1 posterior echoes and might show the cleft of the
anterior mitral leaflet. The anterior mitral leaflet proximity to the inter-
ventricular septum might be the echocardiographic representation of the left
ventricular outflow narrowing which is seen as “goose-neck” sign angi-
ographically.

The interatrial septum is easily detected in cases with right ventricular
cavity dilatation and continuous with the anterior mitral leaflet by transverse
M-mode scan and ultrasonocardiogram.\(^2\) Nimura et al\(^6\) demonstrated
that the discontinuity between the interatrial septum and anterior mitral
leaflet was observed in ECD and might reflect 1). This finding was observed
in all our cases detectable the interatrial septum except for VSD of ECD type.
There were the 2 additional findings in complete form. They were the
common anterior leaflet and 3) the high interventricular defect. Rastelli et
al8) classified complete form into 3 types according to the morphology of the common anterior leaflets and its attachment. In recent years, Tenckhoff et al9) revealed that complete form was mainly classified into Type I having a free-floating undivided common anterior leaflet and Type II having a divided common anterior leaflet. These authors found that the anatomy of the common anterior leaflet correlated well with amenability to surgical repair, the left ventriculographic profile and the incidence of associated cardiac abnormalities. The common anterior leaflet could be expressed as the direct connection between the anterior leaflet echo locating in the left ventricle and that in the right ventricle by echocardiogram and a continuous single anterior leaflet echo by ultrasonocardiotomogram. Further, it would be possible to know whether the common anterior leaflet is divided or not, by the way of the connection. The straight connection was seen in a case with a free-floating undivided common anterior leaflet and the step-wise connection in a case with a divided common anterior leaflet. In the latter case, 2 different anterior leaflet echoes moving the same were observed in one beam direction medially from the interventricular septum. It is supposed that this pattern would come from the anterior tricuspid leaflet and the divided common anterior leaflet.

Ultrasonocardiotomogram sometimes fails to detect the interventricular septal defect.10) It defected, however, the defect in all cases with complete form. The defect is manifested as a echo-free space between the common anterior leaflet and the intact interventricular septum. The echocardiographic finding of the high interventricular septal defect would be the interruption of the echo in systole between the intact interventricular septum and the common anterior leaflet moving into the right ventricle. This is supposed because the beam is sent into the large defect itself. The easiness of detecting the interventricular septal defect could be explained by the different situation and size of the defect from those in ordinary interventricular septal defect. The defect is large and situated just beneath the common atrioventricular valve,81 so that easily detectable in the beam direction toward the common atrioventricular valve. On the other hand, the definitive interventricular septal defect was not detected in cases with VSD of ECD type possibly due to the small size of it.

The absence of the paradoxical motion of the interventricular septum would be characteristic of complete form. This could be postulated for the presence of the pressure equalization between the right and left ventricles. Diastolic overloading of the right ventricle seen in both partial form and the secundum defect of the interatrial septum disappears in complete form. The interatrial septum was not detected in all cases with complete form. This
would reflect the large or complete defect of the interatrial septum, so that be rather acceptable than neglecting in the diagnosis of complete form.

This study indicates that the ultrasonic abnormalities correlates well with the anatomic abnormalities of ECD. Partial form could be diagnosed by the ostium primum defect of the interatrial septum, the atrioventricular abnormalities and the left ventricular outflow narrowing. Complete form could be correctly diagnosed by the additional 2 findings which are the common anterior leaflet and the high interventricular septal defect. VSD of ECD type, special form, could be strongly suspected by the intact interatrial septum continuous with the anterior mitral leaflet as well as the atrioventricular valve abnormalities and the left ventricular outflow narrowing.

REFERENCES

5. Piteroni DR, Freedom RM, Homsy E: Echocardiography in atrioventricular canal defects: A clinical spectrum. (abstr) 2nd World Congress on Ultrasonics in Medicine, June, 1973