Subxiphoid Cross-sectional Echocardiographic Imaging of the “Goose-neck” Deformity in Endocardial Cushion Defect

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SUMMARY The cross-sectional echocardiographic demonstration of the “goose-neck” deformity is described in four patients with endocardial cushion defect. The diagnosis was confirmed in each patient by left ventricular angiography. The subxiphoid approach of cross-sectional echocardiography in diastole allowed visualization of an elongated, narrowed, and somewhat horizontally inclined configuration of the left ventricular outflow tract, which appeared almost identical to that obtained by angiography. In systole, the right border of the left ventricle was composed of the cleft anterior mitral leaflet, the left-sided line of which was convex toward the left ventricular cavity. The mitral valvular echoes were thickened, jagged and irregular, which seemed to correspond to the scalloped appearance of the right border of the left ventricular silhouette in a systolic phase of the left ventricular angiogram. There were no obvious differences between the goose-neck configuration of complete-type endocardial cushion defect and that of ostium primum atrial septal defect.

FROM THE DIAGNOSTIC standpoint of endocardial cushion defect (ECD), the “goose-neck” deformity as seen on the frontal projection of the left ventricular angiogram is a most important feature.1-7 M-mode echocardiography has been widely used as a diagnostic tool for various cardiac malformations. The narrowing of the left ventricular outflow tract, as judged from the distance between the left side of the interventricular septum and the mitral valve on echocardiogram at the onset of systole, and the prolonged mitral–septal apposition in diastole, corresponded to the angiographic feature of the goose-neck deformity.8-14 Recently, cross-sectional echocardiography has been used to image a more precise delineation of anatomic details of ECD.16-19 However, imaging of the goose-neck deformity in ECD by cross-sectional echocardiography has not been reported. In this report we demonstrate the ability of cross-sectional echocardiography to show the goose-neck deformity and compare it in detail with that obtained by angiographic study.

Methods

Cross-sectional echocardiographic examination was performed in four patients, ages 1–3 months, with ECD. Three had complete-type ECD and one had ostium primum atrial septal defect with a left mitral leaflet. The diagnosis was confirmed in each patient by biplane left ventricular angiography. To observe the normal configuration of the left ventricular cavity, 15 normal infants, 1–3 months of age, served as controls. The patients and control subjects were examined in the supine position using the subxiphoid technique.20 The scanner probe was placed at the subxiphoid region, tilting about 20° downward from the plane of the anterior chest wall. The sector beam was positioned through the heart in a plane parallel to a line between the patient’s shoulders to allow simultaneous visualization of the left ventricular cavity and the aortic root. As this subxiphoid sector view is nearly identical to the frontal projection of the left ventricular angiogram, it is suitable for evaluating the configuration of the left ventricular outflow tract of ECD.

We performed this study using a real-time mechanical sector instrument (Aloka SSD 1000 with ASU 25 Hand Scanner and USM 6B Amplifier). The scanner probe, which contained a 3-MHz transducer focused at 7.5 cm, was mechanically driven through a variable angle (from 30–80°) at a variable rate of 10–30 frames/sec. We usually operated this system at a rate of 30 frames/sec, which yields a line density of approximately 110 lines/frame. Cross-sectional echocardiographic images were observed in real-time during the examinations. Next, these images, which were displayed at phase-selected single frames, were photographed directly from the oscilloscope screen by a Polaroid camera, and the real-time images were recorded by a 16-mm cinecamera and analyzed subsequently in real-time, slow motion and single format.

To correspond with the frontal projection of the left ventricular angiogram, selected still-frame photographs were reversed and inverted. Thus, the echocardiographic images were shown with the liver in the apex of the fan at the bottom of the screen and the aortic root at the superior portion of it. The left-sided structures were on the right and the right-sided structures were on the left of the observer. The frontal view of the heart, therefore, is observed on echocardiogram.

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Results

In the subxiphoid view of the control subjects, the configuration of the left ventricle (figs. 1A and 1B) was somewhat elliptic, with the long axis running from the cardiac apex to the aortic root. The cardiac apex, left ventricular outflow tract and aortic root were demonstrated simultaneously in one cross-sectional plane. The size of the ellipse altered with the cardiac phase. The outflow tract of the left ventricle was widely open throughout cardiac cycle, giving neither narrowed nor elongated configuration. The long axis of the left ventricle tended to lie at about 45° horizontally. The right border of the left ventricle was composed of the interventricular septum, the left-sided line of which was smooth and convex toward the right of the heart throughout cardiac cycle. In this sector view, the echo of the anterior mitral leaflet was not seen because the mitral valve orifice was situated posteriorly apart from this section.

Figure 2A is a cross-sectional echocardiogram of case 1 with complete-type ECD illustrating the left ventricular cavity and aortic root in an early systolic phase. The left ventricular outflow tract lay at about 35° to the horizontal, and its right border was shown to be composed of the cleft anterior mitral leaflet and was slightly convex toward the left ventricular cavity. In this still photograph the echo of the cleft anterior mitral leaflet may look like that of the interventricular septum, but in real-time pictures the movement of this echo corresponded to rapid valvular, rather than septal, motion. On the early diastolic image of this case (fig. 2B), the echo of the superior segment of the anterior mitral leaflet moved superiorly and reduced the width of the outflow tract, resulting in an elongated and almost horizontally inclined configuration of the outflow tract that simulated a goose neck. In actual examinations or real-time movies, this opening and closing movement of the superior segment of the anterior mitral leaflet was observed well. As the image of the figure 2A was photographed on an early but not a late systolic phase, the left ventricular size appears larger than that of the diastolic phase (fig. 2B), which was recorded in early diastole. On the angiocardiogram of a systolic phase (fig. 2C), the left ventricular outflow tract was widely open and inclined at about 35° to the horizontal, and its right border was formed by anterior mitral leaflet and had a scalloped contour. In diastole (fig. 2D), the superior segment of the anterior mitral leaflet was pushed upward, resulting in a narrowed and elongated appearance of the left ventricular outflow tract, similar to a goose neck. The echocardiographic findings of the left ventricular outflow tract coincided well with the angiographic observations.

Figure 3A is a cross-sectional echocardiogram of case 2, who had complete-type ECD, illustrating the left ventricular cavity and aortic root in early systole. The right border of the left ventricle was composed of the cleft anterior mitral leaflet, the echoes of which were thickened and slightly convex toward the left ventricular cavity. Furthermore, the sharp and smooth line of the right border of the left ventricle was replaced by a jagged and irregular line. On this echocardiogram, the left ventricular outflow tract was not narrow. Figure 3B is an echocardiogram of the same case in an early diastolic phase. The superior segment of the cleft anterior mitral leaflet moved superiorly, raising the "floor" of the left ventricular outflow tract and forming an elongated, narrowed, and almost horizontally inclined configuration. The free edge of the superior segment of the cleft anterior mitral leaflet appeared to be thickened.

![Figure 1. Subxiphoid cross-sectional echocardiograms of a normal left ventricular (LV) cavity and aortic root. The configuration of the LV cavity is somewhat elliptic, with the long axis running from the cardiac apex to the aortic root. The LV outflow tract (ot) is widely open in both a diastolic frame (A) and a systolic frame (B). The right border of the left ventricle is composed of the interventricular septum (IVS), the left-sided line (small white arrows) of which is smooth and convex toward the right of the heart throughout cardiac cycle. S = superior; I = inferior; R = right; L = left; Ao = aorta.](image-url)
An echocardiographic goose-neck configuration was also demonstrated on a case of ostium primum atrial septal defect with a cleft mitral leaflet. The right border of the left ventricle was composed of the echoes of the anterior mitral leaflet in systole (fig. 4A), which moved superiorly in diastole, forming an elongated, narrowed, and almost horizontally inclined outflow tract (fig. 4B). These echocardiographic features of ostium primum atrial septal defect appeared to be almost identical to those of complete-type ECD.

Discussion

The appearance of the left ventricle in the frontal left ventricular angiocardiogram in cases of ECD is characteristic, and is commonly called the goose-neck deformity. This angiocardiographic appearance has been recognized since the early 1950s, but not until 1964 did Baron et al. lucidly describe the anatomic basis of this sign. They said that the selective left ventricular angiocardiogram appears to be the most ac-

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**Figure 2.** Cross-sectional echocardiograms and frontal left ventricular angiograms of case 1 with complete-type endocardial cushion defect. (A) In systole, the right border of the left ventricular outflow tract (ot) consists of the cleft anterior mitral leaflet (AML), the left-sided line (small white arrows) of which is slightly convex toward the left ventricular cavity. (B) In diastole, the superior segment of the cleft AML moves superiorly, forming an elongated and narrowed configuration of the outflow tract. (C) On the angiogram of a systolic phase, the right border of the left ventricle (LV) is formed by the cleft AML and has a scalloped contour. (D) In diastole, the superior segment of the cleft AML is pushed upward, forming a goose-neck deformity. S = superior; I = inferior; R = right; L = left; Ao = aorta; IVS = interventricular septum.
FIGURE 3. Cross-sectional echocardiograms of case 2 with complete-type endocardial cushion defect. (A) In systole, the right border of the left ventricle (LV) is composed of the cleft anterior mitral leaflet (AML). The echoes (small white arrows) are jagged and irregular. (B) In diastole, the superior segment of the cleft AML moves superiorly, forming an echocardiographic “goose-neck” deformity. $S =$ superior; $I =$ inferior; $R =$ right; $L =$ left; $Ao =$ aorta; $IVS =$ interventricular septum.

FIGURE 4. Cross-sectional echocardiograms of case 3, with ostium primum atrial septal defect. (A) In systole, the right border of the left ventricle (LV) is composed of the anterior mitral leaflet (AML). (B) In diastole, the AML moves superiorly, forming an echocardiographic “goose-neck” deformity. $S =$ superior; $I =$ inferior; $R =$ right; $L =$ left; $ot =$ outflow tract; $Ao =$ aorta.

Recently, echocardiography has been used to diagnose the anatomic details of ECD. Some reports described the echocardiographic expression of the goose-neck deformity. On M-mode echocardiography, anterior displacement of the anterior mitral leaflet was thought to be an echocardiographic representation of the goose-neck deformity in angiocardiography but this echocardiographic assessment of the narrowing of the left ventricular outflow tract was less sensitive than angiocardiography. On cross-sectional echocardiograms, Hagler et al. described the abnormal orientation of the anterior mitral leaflet to both the interventricular septum and left ven-
tricular outflow tract using a parasternal long-axis view. These findings seemed to be an echocardiographic representation of the goose-neck deformity on a parasternal long-axis view.

Lange et al. evaluated and verified the usefulness of subxiphoid cross-sectional echocardiography for examining children with various forms of congenital heart disease. In this subxiphoid approach, the plane imaging the left ventricular outflow tract and aortic root is similar to the frontal silhouette of the left ventricular angiogram. Using this subxiphoid technique, we attempted to demonstrate the details of the goose-neck deformity. It revealed that this technique could demonstrate precisely the echocardiographic goose-neck deformity in all four patients with ECD. This echocardiographic image of the goose-neck deformity was nearly identical to that of the frontal left ventricular angiogram. The right side of the left ventricular outflow tract was composed of the cleft anterior mitral leaflet during systole, and in diastole the superior segment of the cleft anterior mitral leaflet opened into the left ventricular outflow tract. This movement of the superior segment of the anterior mitral leaflet in diastole gave rise to an elongated, narrowed, and somewhat horizontally inclined configuration of the outflow tract of the left ventricle that simulated a goose neck.

In addition to the goose-neck deformity, it is also important in the diagnosis of ECD that the right border of the left ventricular silhouette in systole is composed of a scalloped and serrated contour in the frontal left ventricular angiogram. In this study we found that the images corresponding to this angiographic appearance could be demonstrated by subxiphoid cross-sectional echocardiography. The echocardiographic configuration of the right border of the left ventricle in ECD was obviously different from that of the normal left ventricle: Its right border in systole was composed of the cleft anterior mitral leaflet, the echoes of which were thickened and slightly convex toward the left ventricular cavity. Moreover, the left-sided outline of these mitral valvular echoes was jagged and irregular rather than smooth, as in a normal heart. This image seemed to correspond to the peculiar appearance of the right border of the left ventricular silhouette in a systolic phase of the frontal left ventricular angiogram.

We could find out no obvious differences between the goose-neck configuration of complete type ECD and that of ostium primum atrial septal defect. To differentiate the various subtypes of ECD, the apical four-chamber view seems to be more suitable.

In conclusion, the subxiphoid approach of cross-sectional echocardiography in ECD allowed visualization of the characteristic goose-neck deformity of the left ventricular outflow tract in diastole. In systole it demonstrated the image corresponding to the scalloped contour of the right border of the left ventricular angiogram.

References
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