Variability of Echocardiographic Discontinuity in Double Outlet Right Ventricle and Truncus Arteriosus

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SUMMARY

The widespread application of echocardiography to the field of congenital heart disease has led to the development of a concept of "echocardiographic discontinuity" for the diagnosis of some conditions. Although this is a valuable sign in differentiating such entities as tetralogy of Fallot, truncus arteriosus and double outlet right ventricle, the reported cases illustrate that the echocardiographic recognition of discontinuity may be complicated by technical factors. In addition, the differential diagnosis must include truncus arteriosus with coexistence of anterior and posterior discontinuity.

Additional Indexing Words: Ultrasound Continuity Tetralogy of Fallot Noninvasive techniques

The recent application of echocardiography to the field of congenital heart disease has generated several new "diagnostic" signs. Ultrasonic discontinuity between the aortic root and the mitral valve (posterior discontinuity) is reported to differentiate double outlet right ventricle from tetralogy of Fallot where the anterior aortic root overrides the interventricular septum (anterior discontinuity) but aortic-mitral continuity remains intact. Anterior discontinuity has also been described in truncus arteriosus. This report demonstrates that the continuity-discontinuity sign is variable and may be susceptible to technical factors. In addition, it is noted that the differential diagnosis for the coexistence of both anterior and apparent posterior discontinuity must include truncus arteriosus as well as double outlet right ventricle, a diagnostic point not previously emphasized.

Methods

The echocardiographic examination was performed with a commercially available ultrasonoscope (Smith-Kline Instruments) using a 2.25 MHz transducer of 0.25 inch diameter acoustically focused at 7.5 cm. The patients were studied in the supine position with the transducer at the left sternal border. Permanent records were made in a time-motion presentation on a strip chart recorder as the ultrasonic beam was directed smoothly along the course of the left ventricular outflow tract, as previously described by others. Multiple transducer positions at the third, fourth and fifth intercostal space on the anterior chest wall were utilized.

Results

Previous reports have not precisely defined anterior discontinuity. We have developed the following: anterior discontinuity or overriding of the aorta (rarely pulmonary artery in Taussig-Bing anomaly, partial transpositions or variants of double outlet right ventricle) requires anterior displacement, usually abrupt, of the plane of the anterior aortic wall measured on the interior surface at end diastole from the plane of the left ventricular septal wall also at end diastole, determined at the level of the mitral valve (fig. 1). Currently, we interpret anterior aortic displacement of > 5 mm as a positive sign of overriding, or anterior discontinuity. It should be emphasized that many normals have no anterior displacement with a careful sweep and in some cases the plane of the anterior aortic wall is slightly posterior to that of the left septal surface as is the case in the illustration. Anterior displacement of 5–6 mm represents a range in which visual observation without measurement may suggest continuity and in which angiography indicates minimal anterior overriding. Careful inspection for abrupt change between aortic wall and septum is important in this group. Such a case is illustrated below. Posterior discontinuity[1,5] requires an abrupt anterior displacement of the posterior wall of the great artery, usually the aorta, from that of the mitral valve. As fixed reference points the displacement should be determined between the end-diastolic position of the posterior wall of the great artery and the end-diastolic...
position (C point) of the mitral valve (fig. 1). In our experience, a displacement of several millimeters may be normal, if the transition is gradual on a smooth echo sweep. Although anatomic and angiographic experience suggest that cases of mitral-aortic (pulmonic) discontinuity should have increased displacement between mitral and aortic landmarks (fig. 1), we have not found a clear-cut echocardiographic difference from the range of normal (mitral-aortic continuity intact). Interruption of the echo signal without displacement (as in a large ventricular septal defect) is not interpreted as evidence of discontinuity.

Double Outlet Right Ventricle

Three patients with double outlet right ventricle were examined. In two, the echocardiographic findings of anterior and posterior discontinuity of the great artery were noted. Diagnosis in both cases was confirmed by catheterization to be double outlet right ventricle with great arteries in a side-by-side position. The third case was an infant who displayed anterior overriding or discontinuity on echocardiogram, but who was initially thought to have posterior continuity (fig. 2A). A second echocardiographic sweep from a position approximately one interspace higher on the anterior chest wall produced the echocardiogram in figure 2B, which was interpreted as showing discontinuity between the pulmonary artery and the ventricular septum anteriorly and the pulmonary artery and the mitral valve posteriorly. The impression of double outlet right ventricle with anterior aorta and overriding pulmonary artery was confirmed by catheterization and angiography. In addition, coarcta-

tion of the aorta was diagnosed. This patient has subsequently died, and the pathologic diagnosis was double outlet right ventricle without continuity between the mitral valve and the overriding great artery, which was the pulmonary artery. The aorta was in a position anterior to the pulmonary artery.

Truncus Arteriosus

Seven patients with truncus arteriosus or pseudotruncus arteriosus (tetralogy of Fallot with pulmonary atresia) were examined. Three patients revealed the reported findings of anterior discontinuity or overriding of the truncal vessel wall with continuity of the posterior vessel wall and the mitral valve. On initial echocardiographic examination, three patients, including two with truncus arteriosus and one with pseudotruncus arteriosus (fig. 3A) did not appear to have anterior discontinuity although careful measurement suggested an anterior displacement. By repositioning the transducer higher on the anterior chest wall and repeating the standard echocardiographic sweep from left ventricle to great artery, anterior discontinuity was clearly demonstrated (fig. 3B). A lateral angiogram on this patient demonstrates a single great artery which is mildly overriding the interventricular septum (fig. 4). This patient has subsequently undergone repair with a Rastelli-type procedure and is currently having a satisfactory postoperative course.

Finally, a case of type II truncus arteriosus was examined in which there was both anterior and apparent posterior discontinuity — between the truncal vessel and the interventricular septum, and the interven-

Figure 1

Smooth echocardiographic sweep from mitral valve (left) to aorta. Upper set of horizontal lines define landmarks for determination of anterior continuity. Anterior aortic surface is displaced 2 mm, in this case posteriorly. Lower set of horizontal lines define landmarks for determination of posterior displacement. Posterior aortic wall is displaced 3 mm anteriorly. Note gradual transition. Ao = aorta; av = aortic valve; aw = (large vertical solid arrow) = anterior aortic wall; paw = (large vertical solid arrow) = posterior aortic wall; S = septum; sed = (small vertical solid arrow) = septum, end-diastolic position; C = (open arrow) = mitral closure point; M = mitral valve.

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A) Smooth echocardiographic sweep of patient with double outlet right ventricle. Transducer positioned at fourth intercostal space. The anterior pulmonary artery (PA) appears to be displaced anterior to the ventricular septum (VS). There is continuity between the posterior PA wall (lower vertical arrow) and the anterior mitral leaflet (AML). This record was interpreted as anterior discontinuity and posterior continuity. ecg = electrocardiogram, PV = pulmonary valve, LA = left atrium. B) Smooth echocardiographic sweep from the ventricular (left) to great artery (right) level in the same patient with double outlet right ventricle. Transducer at third ICS. Horizontal open arrows indicate anterior displacement (5 mm) of PA from AML. This record was interpreted as posterior discontinuity consistent with double outlet right ventricle.

Both tricuspid septum and anterior mitral valve, respectively (fig. 5A and B). The lateral angiogram is shown on this patient in figure 6. The truncal vessel appears to be primarily associated with the anterior ventricle and the distance between the truncal root and the anterior mitral valve ring shadow appears to be excessive and consistent with anatomic discontinuity. This patient has subsequently had bilateral pulmonary artery banding with a very satisfactory course but to date has not had a complete repair.
DISCONTINUITY IN DORV AND TA

Discussion

Although anterior discontinuity is mandatory in double outlet right ventricle and usually present in cases of truncus arteriosus and also tetralogy of Fallot, there is a documented variability in the degree of overriding of the aortic vessels.\textsuperscript{3, 8, 9} Marked overriding is easily and reliably diagnosed on echocardiogram (fig. 7). However, in patients with lesser degrees of aortic overriding, echocardiographic, angiographic and even anatomic determination of overriding is more difficult to interpret.

There is a geometric explanation which may be important in the echocardiographic variation. The echocardiographic definition of discontinuity as defined above requires abrupt displacement of the echocardiographic signal anteriorly as one smoothly sweeps the echo beam from the body of the heart to the great artery. By this definition, two structures which lie on an arc at a given length from the transducer will appear continuous and a structure not on the same arc will appear discontinuous. As illustrated in figure 8, the position of the transducer on the anterior chest wall may have a direct influence on

Figure 3
\textit{A)} Smooth echocardiographic sweep from the ventriculocardiac (left) to great artery (right) level in a patient with pseudotruncus arteriosus. Transducer at fourth ICS. Note apparent continuity between the septal echo (S) and the anterior wall of ascending aorta (AA) (4.4 mm anterior displacement). The continuous transition between the anterior mitral leaflet (AML) and the posterior wall of the ascending aorta (vertical arrow) is shown. This record was interpreted as showing posterior and probable anterior continuity. \textit{AV} = aortic valve. \textit{B)} Smooth echocardiographic sweep from the aorta (left) to the mitral valve (right) in the patient with pseudotruncus arteriosus but with transducer at third ICS. Solid horizontal arrows indicate anterior displacement of the anterior AA wall from the septum (6 mm displacement). There continues to be posterior continuity. This record was interpreted as showing clearcut overriding of the aorta or anterior discontinuity.

Figure 4
A lateral angiogram with injection into right ventricle of the patient with pseudotruncus arteriosus whose echocardiograms were shown in figure 3. There is absence of the right ventricular outflow tract with a large aorta mildly overriding the interventricular septum. \textit{AA} = ascending aorta; \textit{AV} = aortic valve; \textit{RV} = right ventricle; \textit{LV} = left ventricle; open arrows indicate the right and left sides of the interventricular septum (see fig. 8).
this finding. The ventricular septum, point Y, and the anterior aortic wall, point X, are equidistant from the transducer located at position A on the chest wall. Since point X is closer than point Y to the transducer at point B on the chest wall, there is an echocardiographic discontinuity (i.e., the anterior aortic wall appears abruptly anterior to the interventricular septal echoes). In our cases, moving the transducer one to two interspaces higher on the anterior chest wall accentuated the finding of anterior discontinuity. The precise degree of overriding or anterior displacement can probably only be determined by direct inspection or on a true lateral angiogram of the heart. To date, however, the use of smooth echo sweeps at two or three interspace levels has been a helpful predictor of mild degrees of anterior position of the great artery and has not falsely predicted a double outlet right ventricle, truncus arteriosus or tetralogy of Fallot.

Posterior discontinuity is an important criterion in the echocardiographic differentiation of double outlet right ventricle from tetralogy of Fallot and may be present in truncus arteriosus. However, it is less precisely defined in our experience and more difficult to interpret. The echograms presented illustrate that posterior discontinuity may be subject to significant variation. Technically, a careful, smooth sweep is required to minimize false positives. Inspection of the

Figure 5

A) Smooth echocardiographic sweep from the truncal root (left) to ventricle (right) in a patient with truncus arteriosus whose angiogram is shown in figure 6. Transducer at 4th ICS. There is discontinuity of both walls of the truncal vessel (defined by horizontal solid arrows) from the ventricular septum (S) and the mitral valve (M). The open horizontal arrows define left septal surface and mitral C point. This record was interpreted as showing both anterior and posterior discontinuity. The displacement anterior is 7.8 mm and posterior 5.6 mm. B) Second sweep on the patient with truncus arteriosus. Transducer is at the third intercostal space. Note the more complete view of the truncal valve and the apparent increase in degree of anterior overriding (anterior displacement 12 mm). The horizontal solid arrows define the end-diastolic margins of the truncal vessel. The horizontal open arrow defines the left ventricular septal surface. TV = truncal valve; Tr = truncus valve; M = mitral valve; S = ventricular septum.
normal sweep demonstrates that the plane of the posterior aortic wall at end diastole and the end-diastolic mitral valve position may be displaced (fig. 1) and a sudden transition would suggest a discontinuity. The cause of false negatives is not entirely clear, although transducer position is important. Because of the anterior placement of the aortic root, the mitral valve is not only abnormally posterior but as the transducer is placed higher on the chest wall the angle (arc length) between the two structures is reduced and they may superimpose (fig. 9). The combination of displacement and superimposition is important in producing the abrupt change we define as posterior discontinuity.

Finally, some cases of double outlet right ventricle with proximity of anterior mitral valve and aortic root have been described and presumably these cases would show posterior echocardiographic continuity and would represent false negative studies for double outlet right ventricle. Currently, we are cautious in interpreting posterior discontinuity unless it is apparent at two or three interspace levels and the diagnosis is consistent with clinical findings.

In truncus arteriosus, the truncal vessel is usually associated with both of the ventricles equally, giving anatomic continuity with the mitral valve. However, the truncus occasionally may be primarily associated with the anterior ventricle and show both anterior and apparent posterior discontinuity on echocardiogram, as illustrated above. It is not clear in cases such as this whether evidence of echocardiographic discontinuity reflects true anatomic fibrous discontinuity. It is important to emphasize that since echocardiographic posterior discontinuity is not limited to double outlet right ventricle, the differential diagnosis must include truncus arteriosus. As Chung et al. have reported, the ability to define a second semilunar valve is useful in separating truncus arteriosus from tetralogy of Fallot. Finding two semilunar valves is an important additional sign to distinguish between truncus arteriosus with posterior discontinuity and double outlet right ventricle as well. However, locating the second

Figure 6
Lateral angiogram of the patient with truncus arteriosus whose echocardiogram is illustrated in figure 5. Injection is into the truncal root. There is an excessive distance between the truncal valve (TV) and the anterior mitral leaflet (AML) consistent with posterior discontinuity. The anterior wall of the truncal vessel (TA) overrides the ventricular septum (S) (see fig. 8).
semilunar valve may represent a technical challenge if the valves are side-by-side or if the second valve and great artery are hypoplastic as they are in many cases of double outlet right ventricle.

The use of oscillating transducers or multi-transducer instruments, although not employed in these patients, would have been helpful in identifying the second great artery and such instruments have been used for diagnosis in this type of patient. However, the determination of mild degrees of arterial overriding and precise mitral-semilunar relationship continues to be a challenge even with the more sophisticated instruments.

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