Subxiphoid Two-dimensional Echocardiographic Diagnosis of Double-chambered Right Ventricle

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SUMMARY
The echocardiographic features of 14 patients with double-chambered right ventricle are presented. Diagnosis was confirmed in 13 patients at cardiac catheterization and in one at necropsy. The ultrasonic diagnosis was based on the visualization in subxiphoid short-axis view of an anomalous muscle bundle at the lower margin of the right ventricular infundibulum; this structure was present in diastole and systole and the distal portion of infundibulum was wide and free of obstruction. Associated lesions were very frequent (especially ventricular septal defects, present in 13 patients). Subxiphoid two-dimensional echocardiography is an easy, reliable, noninvasive means of assessing this disease in infants and children.

DOUBLE-CHAMBERED right ventricle (DCRV) is a division of the right ventricle into two chambers by a hypertrophied muscular bundle, usually located at the sinus-infundibulum junction, which may eventually act as a subpulmonary obstructive element.

DCRV has been considered a rare anatomic entity. In fact, it is probably misdiagnosed because of a variable clinical picture and unreliable data from ECG and chest x-ray. Fermont et al. stated that diagnosis of DCRV could be suspected from the association of the following M-mode echo features: easy visualization of the pulmonary valve, premature closure and systolic flutter of the aortic valve and false septo-aortic discontinuity. However, these features are only suggestive; M-mode echocardiography does not allow direct visualization. Cardiac catheterization has been considered essential to establish the diagnosis. Since the right ventricular outflow tract (RVOT) can be well visualized by subxiphoid two-dimensional echocardiography (S2DE), we investigated the reliability of this technique as a means of diagnosing DCRV non-invasively.

Material and Methods
From October 1980 to April 1981, 14 children with DCRV were studied by S2DE. Thirteen of these children underwent cardiac catheterization and angiography; the diagnosis was confirmed at necropsy in the other patient.

The age at S2DE studies varied from 2 days to 4 years (median 7 months). Five patients were younger than 6 months (median 1 month). Mean weight was 3.9 kg.

We also reviewed S2DE studies of 20 normal infants and nine infants with isolated ventricular septal defect (VSD) proved at cardiac catheterization performed during the same period. The age of the normal subjects varied from 1 day to 5 years (median 8 months). The nine control patients with VSD were 21 days to 5 years old (median 5 months). None had a pressure gradient between the right ventricle and the pulmonary artery or had angiographic evidence of DCRV.

Two-dimensional echocardiograms were performed with an ATL Mark IV echocardiograph using a 3.0-MHz short-focused transducer, providing a 90° sector angle. Studies were recorded on videotape for later analysis. Cardiac structures were displayed by positioning the transducer in the subxiphoid area and performing the examination as described previously. We used the left ventricular short-axis view at the level of the mitral valve apparatus. In this view, the RVOT could be seen in its entirety. It was visualized as a widely opened cavity located between the pulmonary valve superiorly and the distal portion of the right ventricular sinus (tricuspid valve apparatus) inferiorly, and limited anteriorly by the right ventricular free wall and posteriorly by the interventricular septum (fig. 1). Any muscular component contiguous to the interventricular septum and extending obliquely toward the inferior right ventricular free wall was defined as a moderator band (MB).

Cardiac catheterization and biplane right ventricular cineangiography using anteroposterior and lateral projections were performed in 13 patients. Diagnosis was based on the angiographic demonstration of a filling defect in the midportion of the right ventricle during diastole in the anteroposterior view remaining unchanged (or getting larger) during systole. In the lateral view (fig. 2), the filling defect is seen in the anterior wall between the inflow and outflow portions of the right ventricle. The parietal band is normally positioned, although it may be hypertrophied. The diagnosis was also based on the recording of a gradient within the right ventricle. Pressures were recorded with a NIH sidehole catheter. A withdrawal tracing demonstrative of an intraventricular gradient was obtained in six patients. Rapid movement of the catheter from the pulmonary artery to the right ventricular sinus frequently results in missing the gradient. However, all catheterized patients had increased systolic pressure in the right ventricular sinus (range 53–146 mm Hg) and typical angiographic features.

Results
Control Subjects
In normal hearts, the RVOT was easily identified in
the short-axis view (fig. 1). Usually, the MB was not apparent; in four cases, however, it was visualized at the junction of the infundibulum with the right ventricular sinus as a thin muscle bundle present in systole but no longer seen in diastole.

In patients with VSD, the RVOT appeared normal. The MB could not be identified in any patient with VSD.

Patients with DCRV

In all 14 patients an anomalous bundle, identified as a modified MB, was visualized just below the right ventricular infundibulum in the short-axis view. This muscular structure was present in diastole and remained identical or thickened in systole (fig. 3). The distal portion of the infundibulum located under the pulmonary valve was wide and free of obstruction.

In two cases (both with associated pulmonary stenosis [PS]), the MB was only hypertrophied. In nine other patients, the MB was hypertrophied with one or more of the following characteristics: modifications in shape (stretched upwards, fan-shaped, with broad insertions), location (high insertion), or direction (horizontal) (fig. 4). In the remaining three cases, the MB was of normal size but had abnormal location and direction.

Two patients in our series with a known diagnosis of isolated VSD made at a first cardiac catheterization had two-dimensional echocardiographic findings suggestive of DCRV. A second catheterization was performed some months later and showed a gradient between the right ventricle and the pulmonary artery and typical angiographic features of DCRV.

Associated lesions were very frequent. A VSD was present in 13 cases. Usually, the VSD was large and located just below the septal insertion of the anomalous bundle (fig. 5). It usually involved the membranous septum with posterior and inferior extension toward the sinus septum (nine patients) or superior extension.

![Figure 1. Normal heart in diastole, subxiphoid lateral short-axis view. The infundibulum is wide open without any obstruction. Inf = infundibulum; IVS = interventricular septum; LV = left ventricle; PV = pulmonary valve; TV = tricuspid valve.](image)

![Figure 2. Right ventriculogram of a 2-year-old patient with a double-chambered right ventricle. This lateral view shows the filling defect (arrows) caused by the anomalous muscle bundle. Inf = infundibulum; PA = pulmonary artery; RV = right ventricular sinus.](image)

![Figure 3. Subxiphoid two-dimensional short-axis view of a 2-year-old boy with double-chambered right ventricle. The hypertrophied anomalous bundle (arrows) is horizontal, hypertrophied and located in a high position. The anomalous bundle remains identical in systole (A) and in diastole (B). MV = mitral valve; LV = left ventricle; TV = tricuspid valve; PV = pulmonary valve; Inf = infundibulum.](image)
have been reported.\textsuperscript{9,15} Although its embryonic origin is still uncertain, DCRV is clearly different from reactional infundibular stenosis, valvular PS and infundibular hypoplasia (TOF).\textsuperscript{8,13,16}

The present study shows that S2DE can demonstrate the anatomic features of DCRV in a group of patients with angiographic evidence of this anomaly. The diagnosis is based on identification of a hypertrophied, anomalous muscle bundle at the lower margin of the right ventricular infundibulum. Rowland et al.\textsuperscript{17} and Van Praagh and McNamara\textsuperscript{18} reported that the obstructive muscle bundle in DCRV is most likely a superiorly displaced MB. This is in agreement with Goor and Lillehei,\textsuperscript{13} who pointed out the important anatomic variations in the position of the normal MB. Benjamin et al.\textsuperscript{19} suggested that DCRV is due to reactional hypertrophy of muscle bundles secondary to VSD or PS.

**Discussion**

Since Lucas,\textsuperscript{14} in 1962, described the first cases of DCRV as a new anatomic entity to be separated from other forms of RVOT obstruction, more than 100 cases towards the conal septum (four patients). The aorta was above the VSD in all cases. This gave a false impression of overriding in some patients with large defects. However, in these cases, the parietal band was normally located, the distal infundibulum was nonobstructed and there was no infundibular hypoplasia as seen in tetralogy of Fallot (TOF). Associated forms of DCRV with TOF, however, do exist (one case in our series) in which an anterior, superior and leftward deviation of the parietal band causes a malalignment VSD.

Valvular PS was present in five cases. One patient had echocardiographic features of discrete subaortic stenosis, and a systolic gradient of 12 mm Hg was found at catheterization. A right-sided aortic arch was present in two cases without associated TOF.

**Figure 4.** Same view as in figure 3, in diastole, from a 1-year-old boy. The pulmonary valve is closed and appears normal. In this case, the hypertrophied anomalous muscle bundle has a normal free wall insertion and then follows an oblique upward course. AMB = anomalous muscle bundle; other abbreviations as in previous figures.

**Figure 5.** Same view as in figures 3 and 4, in diastole, from a 6-month-old child. The anomalous muscle bundle is horizontal, moderately hypertrophied and located in a high position. It has a wide base of insertion on the right ventricular wall as well as on the interventricular septum. A ventricular septal defect (VSD) is well demonstrated just under the septal insertion of the anomalous muscle. Other abbreviations are as in previous figures.
However, in isolated forms of VSD or PS, the MB usually is in a lower, nonobstructive position and does not induce subpulmonary stenosis and DCRV. Obvious cases of DCRV may well represent advanced forms due to both hypertrophied and malpositioned MBs.

A nonobstructive anomalous muscle bundle may become obstructive with time. In our series, cardiac catheterization first failed to detect two cases of high nonobstructive MB, without significant gradient or suggestive angiographic features in early infancy. A second catheterization performed some months later, after S2DE had demonstrated an anomalous bundle, confirmed the echocardiographic diagnosis.

Associated cardiac anomalies were present in 80–90% of reported cases and in 100% in our series. The most commonly associated lesion is VSD (80% or more in other published series and 93% in ours). A membranous septal aneurysm has been angiographically or preoperatively discovered in some cases previously considered as isolated forms. This fact, and the high incidence of VSD, have led some investigators to think that isolated forms do not exist, and must be regarded as DCRV in which a VSD was present before the time of exploration but closed spontaneously. Our findings support this opinion. Valvular PS, present in four of the 14 patients in our study, is the second most frequent associated lesion; its incidence is variable — 10–30% in reported series. Associated subaortic stenosis, present in one patient in our series, has also been reported elsewhere.

Subxiphoid two-dimensional echocardiography also allowed us to rule out other forms of RVOT obstruction. In valvular PS, the valve looks thickened and excursion is limited; the main pulmonary artery and infundibulum are usually dilated. In more severe forms, reactional hypertrophy develops. In those cases, proximal as well as distal portions of the infundibulum are involved, a situation not seen in DCRV. Associated forms of DCRV with severe PS may, however, be confusing and certainly constitute a challenge for the echocardiographer. TOF, clinically undistinguishable from DCRV, has well-known echocardiographic features and should be easily ruled out. DCRV associated with TOF, present in one of our patients, does exist, however, and should be suspected when features of both entities are encountered. A right-sided aortic arch may be present in both malformations and does not constitute a distinctive feature per se.

In conclusion, S2DE provides an easy, noninvasive means of assessing DCRV in neonates and infants suspected of RVOT obstruction. It can also identify associated lesions (VSD or PS) and rule out other forms of RVOT obstruction (especially TOF). We believe that in most patients who do not require immediate surgery, early hemodynamic and angiographic investigation is not necessary.

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