Echocardiographic Appearance of the Chiari Network: Differentiation from Right-heart Pathology

JEFFREY A. WERNER, M.D., MELVIN D. CHEITLIN, M.D., BRIAN W. GROSS, M.D., SARAH M. SPECK, M.D., AND TOM D. IVEY, M.D.

with the technical assistance of Carolyn Janko, Max Hedgecock, Sharon Palella and Terryl Dooley

SUMMARY. As echocardiography is being used more often, its value and accuracy are becoming more fully appreciated. Coincident with wider application of this imaging technique is the potential for identifying normal anatomic variants and their possible erroneous interpretation as pathologic states. In this report we describe the M-mode and two-dimensional echocardiographic features of a congenital remnant known as the Chiari network. This structure can present as a highly mobile, highly reflectant echo target that can be seen in several locations in the right atrium. We report here an index case that could be well examined echocardiographically and that was a cause of considerable concern due to the presence of congestive heart failure and a history of staphylococcal endocarditis. The presence of the Chiari network was confirmed pathologically. Subsequently, we found similar echocardiographic findings in 19 of 1248 patients (1.5%) studied in our laboratory. This congenital remnant, which is found pathologically in 2–3% of normal hearts, could be confused with valve disruption, vegetation or other mass lesion, particularly when associated with a suggestive clinical situation.

M-MODE and two-dimensional echocardiography have gained wide acceptance for providing safe, repeatable and accurate diagnostic information in a variety of complicated clinical situations. Susicion of valvular heart disease, particularly infection of the valves, represents an important indication for echocardiographic examination. Increasing experience with these techniques has allowed a better, and in some cases new, appreciation of normal and abnormal cardiac anatomy, motion and structural relationships. These include structures often described at cardiac surgery or at postmortem examination, but not previously demonstrable in ambulatory patients. We describe the echocardiographic appearance of the Chiari network, a not uncommon anatomic finding that might be considered a "normal variant." This structure, particularly in the setting of fever, congestive heart failure, pulmonary infiltrates and a history of i.v. drug abuse, could be mistaken for evidence of active infection or disruption of normal right-sided structures, possibly requiring urgent cardiac surgery.

Materials and Methods

Patient 1 (the index case) was initially studied in August 1978 using a Picker System 80CI ultrasonograph. Follow-up studies in this patient in August 1979 and all other patient studies were performed using an Advanced Technology Laboratories Mark III mechanical sector scanner with a 3.0-MHz crystal. Patients were studied in both the supine and left lateral decubitus position using the three standard acoustic windows: parasternal, apical, and subcostal. Orthogonal views from each window were obtained in the conventional fashion. All M-mode records were obtained on a Honeywell 1850 strip-chart recorder and real-time studies were recorded and preserved on a Sanyo videocassette recorder using ½-inch videotape. After documentation of the index case, records were reviewed retrospectively and prospectively between April 1979 and July 1980. Studies from 1248 patients were considered and the echocardiographic features described were found in 19 patients. Three observers reviewed the M-mode and two-dimensional real-time records of the 19 patients, and the presence and location of the Chiari network were agreed upon by consensus. One patient initially thought to have this finding was rejected because review of the real-time images did not show sufficiently convincing typical features. Patient charts were reviewed for clinical information after the identification of and agreement on the presence of the echocardiographic findings. Patients were unselected and included the general population of patients referred to the echocardiographic laboratories at Harborview Medical Center. Outpatients and inpatients alike were included. Echocardiograms were excluded only if they were technically of nondiagnostic quality.

Results

Description of Index Case

A 20-year-old black female was admitted to Har- borview Medical Center in July 1978 with a 2-day history of severe shortness of breath. In 1976, she had presented to another hospital with acute bacterial
endocarditis due to staphylococcus aureus and mild-to-moderate aortic regurgitation. Left ventricular size by both ultrasound and x-ray was at the upper limit of normal. She was treated routinely with antibiotics for 1 month and stabilized with no symptoms of congestive heart failure. During the ensuing months, she was seen in the outpatient clinic. She continued to have moderate aortic regurgitation and was asymptomatic, but continued to use intravenous drugs intermittently. Six weeks before presentation to our emergency room, she had completed an uneventful pregnancy and uncomplicated vaginal delivery. At the time of this admission, she was severely short of breath and diaphoretic. The jugular venous pressure was elevated, but no definite "V" waves were present. She had a low-grade fever. Her blood pressure was 158/30 mm Hg. There was a grade 2/6 decrescendo murmur that lasted approximately three-fourths of diastole. A second heart sound was present, but decreased. There was a grade 2/6 systolic regurgitant-type murmur radiating across the anterior precordium that did not clearly change with respiration. Chest x-ray and M-mode echocardiography performed within hours of admission showed an increase in heart size compared with the studies performed during her previous hospital admission. The echocardiogram also showed coarse diastolic fluttering of the mitral valve without early closure. The anterior leaflet of the tricuspid valve was normal. An unusual, sinuous echo target (fig. 1) in the right atrium was seen as a curvilinear oscillating echo located relatively posteriorly toward the right atrium and at times moved toward but not through the anterior tricuspid valve leaflet in mid-diastole. No continuous echoes were seen in the right ventricular inflow tract. The largest excursions toward the tricuspid leaflet occurred recurrently with atrial contraction in late diastole, followed again by rapid posterior motion toward the posterior right atrial wall at the onset of systole. This pattern of systolic and diastolic movement seemed to recur cyclically. The finding was reproducible and was best imaged with the transducer aimed slightly inferiorly and medially in the normal fashion to visualize the tricuspid valve apparatus and right heart structures. A retrospective examination of similar right-heart sweeps from the echocardiogram recorded in 1976 showed an identical finding that was somewhat obscured by right atrial wall echoes and high gain settings. The current two-dimensional study (fig. 2) showed a long, thin, highly mobile echo target moving in a lateral-to-medial direction across the right atrium, as well as rapid, whip-like motions toward and away from the tricuspid valve ring. The most prominent apparent motion included movement toward the tricuspid valve in early diastole, movement back posteriorly and at times out of the scan plane and then the most rapid and largest excursion toward the reopening tricuspid valve leaflets with atrial systole. The combined lateral-to-medial sweeping motion and the rapid superior-to-inferior diastolic excursions in and out of the scan plane gave the impression of an overall rotatory motion of this structure about a fixed point at the junction of the mouth of the inferior vena cava as it entered the right atrium. Multiple sweeps of the right heart using parasternal, apical and subcostal approaches failed to demonstrate a clear-cut medial or inferior attachment of the structure, which otherwise appeared to be anchored only at one point and to be moving freely in the right atrial cavity.

Figure 1. M-mode sweep of the right heart showing a curvilinear, hypermobile echo located posteriorly in the right atrium and not obviously associated with the tricuspid valve apparatus. RV = right ventricle; ALTV = anterior leaflet of the tricuspid valve; RA = right atrium; CN = Chiari network.

Figure 2. Short-axis parasternal two-dimensional view at the level of the aorta. Three stop frames are shown to demonstrate the whip-like mobility of the Chiari network as well as the fairly clear-cut separation from the tricuspid valve and interatrial septum. TV = tricuspid valve; CN = Chiari network; IAS = interatrial septum; LA = left atrium; AO = aorta.
Because of the onset of congestive heart failure and the clinical worsening of aortic valvular regurgitation, catheterization of the left and right heart was performed. A supravalvular aortic angiogram showed 4+ aortic regurgitation and no obvious left-sided vegetation. The right-heart catheterization showed normal right atrial pressures and no systolic regurgitant wave. Because of the prominent unusual right atrial echo, a right atrial angiogram was performed, but no evidence of a right atrial filling defect, flail tricuspid valve leaflet or vegetation could be identified.

The patient was initially stabilized on digoxin, diuretics and vasodilators, but continued to have intermittent paroxysmal nocturnal dyspnea and profound exercise intolerance for the next 3 weeks. Therefore, aortic valve replacement was undertaken. At the time of operation, there was no active vegetation on the aortic valve. Two of the three aortic cusps, however, were thinned and retracted, accounting for the significant valvular leak. The left ventricle was considerably dilated. The right atrium was incised and contained a large, fluttering, "honeycombed" structure associated with the orifice of the inferior vena cava. A large portion of this structure was excised (fig. 3) for culture and pathologic examination. Small remnants of attachments to the inferior vena cava were left in place. A #23 porcine heterograft prosthetic aortic valve was placed in the aortic position. The patient had an uneventful postoperative course and was discharged. Although initially lost to early follow-up, she returned to the clinic approximately 1 year later feeling well and asymptomatic. A postoperative follow-up echocardiogram was performed (fig. 4). A careful examination of the right heart using multiple transducer sweeps and gain settings and several transducer locations on the chest showed an essentially normal right atrium and tricuspid valve apparatus. A few cardiac cycles showed small remnants of the network's attachments that were left in place at operation.

Additional Patients

The 19 patients who had the echocardiographic features compatible with the Chiari network are described in table 1. There were seven females and 12 males, mean age 56 years (range 21–94 years). Three patients had a history of intravenous drug abuse and six of the 19 patients had known or suspected valvular heart disease. Two patients had suspected left-to-right shunt. Three patients were having routine echocardiographic studies after resuscitation from out-of-hospital ventricular fibrillation. Four patients were specifically referred for echocardiographic study to investigate a possible intracardiac source for pulmonary or systemic thromboembolic phenomenon. Six patients were referred for evaluation of left ventricular function. Two patients had clinically active infective endocarditis at the time of the study, and two additional patients were referred for echocardiographic study to exclude possible endocarditis. The echocardiographic finding was confirmed surgically in one patient.

Echocardiograms

The Chiari network could be seen with all three standard approaches. The most diagnostic view was the long-axis, parasternal view (right ventricular inflow tract) in five patients, the short-axis parasternal view in eight patients, the subcostal view in three patients, and the four-chamber apical view in three patients. The curvilinear right atrial echoes described...
were variable in location within the right atrium and could not be seen convincingly on the M-mode study in three of the 19 patients. The two-dimensional real-time appearance of these structures varied, as would be expected by pathologic description6 (table 1). In nine of the 19 patients, the Chiari network clearly originated from the inferior vena cava orifice in the posterolateral right atrium and coursed inferomedially toward the tricuspid ring. A predominantly horizontal appearance of the structure fairly low in the right atrium immediately superior to the tricuspid ring and apparently associated with the interatrial septum was seen in six of the 19 patients and an intermediate position was seen in the remaining four patients. The structure was very small, bright and far removed from the tricuspid apparatus (posterior in the right atrium) in two patients and probably would not have been confused with the tricuspid apparatus itself. In five patients the structure was highly reflectant and close to the tricuspid ring and could have been indistinguishable from a partially disrupted septal tricuspid valve leaflet or vegetation.

Discussion

The echocardiographic appearance of a Chiari network has not previously been described. While such a congenital remnant is seldom clinically important, such membranes have been reported as sites of thrombus formation, and hence potential etiologies for pulmonary emboli7 as well as a source of entrapment of a right-heart catheter. In this case, the identifica-
tion of the Chiari network was important for two reasons: These M-mode and two-dimensional echocardiographic findings had not previously been described, and the wildly mobile appearance within the right atrium could be confused with other curvilinear, highly mobile, cyclically recurring pathologic echo targets such as right-heart vegetation, flail tricuspid leaflet (particularly the septal or posterior leaflet), ruptured chordae tendineae to the tricuspid apparatus, a small right-heart thrombus or even a pedunculated right-heart tumor. This distinction, therefore, could have considerable clinical importance, particularly in febrile and acutely ill patients. While this echocardiographic finding is not uncommon, the reported pathologic incidence of such a structure is 2–3%. Indeed, since we recognized and confirmed the echocardiographic appearance of this embryologic remnant (the index case described), we have seen similar echocardiographic findings in 19 of 1248 patient studies (1.5%) (table I). Illustrations of the echocardiographic examination of an additional recent patient from this series are shown in figures 5–7. This young female patient was being examined for the presence of an asymptomatic systolic ejection murmur. Figure 5 is an M-mode echocardiogram in this patient. Although some angulation artifact is present, portions of the anterior leaflet of the tricuspid valve are clearly delineated. A somewhat more posterior, intermittent, highly reflectant curvilinear echocardiographic target was seen toward the posterior right atrial wall. Figure 6 shows this structure in multiple stop frames from the real-time, short-axis study. A prominent, curvilinear, highly mobile echocardiographic target was seen to extend from the inferolateral portion of the right atrium at the orifice of the inferior vena cava, medially, toward the junction of the interatrial septum and the tricuspid ring. A right ventricular inflow tract view from the para-sternal region in this patient (fig. 7) further delineates similar anatomy in motion from a different tomographic plane. In the majority of these patients, these structures can be identified and validated using at least two and sometimes three orthogonal tomographic views on the two-dimensional study: views of the right ventricular outflow tract from a short-axis, para-sternal approach, and from the four-chamber apical or subcostal views. The key differential points include identification of two, and ideally three, normally appearing tricuspid valve leaflets; the presence of a bright, rotatory, highly mobile echocardiographic target that does not move into the right ventricular inflow tract or right ventricle in diastole as would be typical of a tricuspid leaflet vegetation; and, in the four-chamber apical or subcostal view, the typical posterolateral orientation and anteroinferior and medial course of this structure across the right atrium.
The latter finding is more obvious when the Chiari network is associated with the orifice of the inferior vena cava. The use of intravenous bolus contrast material to outline the course of the inferior vena cava, right atrium, tricuspid valve and right ventricle can be of additional benefit in excluding the presence of right atrial mass and tricuspid leaflet disruption. There is some variability in location, however, depending on the primary origin of the network; such structures have been associated with the right atrial wall, coronary sinus and interatrial septum, in addition to the more common association with the orifice of the inferior vena cava, as in the two patients illustrated.

In conclusion, we observed and documented the echocardiographic appearance of a previously described anatomic entity and emphasized the importance of careful interpretation of new echocardiographic findings and the use of multiple acoustic windows, tomographic views and, possibly, contrast medium to separate normal from abnormal anatomy. Prominent, highly mobile, cyclical echoes are always of concern, particularly in an acutely ill or febrile patient. Such findings should be interpreted cautiously, particularly in view of the not uncommon presence of this “normal variant.”

References
Echocardiographic appearance of the Chiari network: differentiation from right-heart pathology.
J A Werner, M D Cheitlin, B W Gross, S M Speck and T D Ivey

Circulation. 1981;63:1104-1109
doi: 10.1161/01.CIR.63.5.1104

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/63/5/1104