The Chiari Network in an Echocardiography Student

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Case Report

A 27-year-old male cardiovascular technology student underwent an echocardiogram by fellow students as a laboratory exercise. A serpentine highly mobile reflective structure was noted in the right atrium (RA). By parasternal long- and short-axis views it appeared to whip toward the posterior aspect of the right atrium during systole, and then prolapse across the tricuspid annulus during diastole (Figs. 1 and 2). From these views as well as apical (Fig. 3) and subcostal (Fig. 4) views, the mass moved in and out of the scan plane. The structure was constrained near the orifice of the inferior vena cava.

Figure 1. Parasternal short-axis systolic frame at the level of the aortic valve. The Chiari network is located in the posterior right atrium. Although precise attachment sites could not be identified, real-time imaging suggested constrained motion near the interatrial septum. RA = right atrium; RV = right ventricle.

Figure 2. Parasternal short-axis diastolic frame at the level of the aortic valve. The Chiari network is noted to prolapse across the tricuspid annulus. RA = right atrium; RV = right ventricle.
The Chiari network (arrow) in this apical four-chamber view appeared serpentine. By real-time imaging it displayed an undulating motion. RA = right atrium; RV = right ventricle.

cava as well as the interatrial septum (IAS). These findings were consistent with a diagnosis of Chiari network.

Discussion

In the developing embryo, the right horn of the sinus venosus becomes incorporated into the wall of the right atrium. The valve of the right sinus venosus will then regress. The cranial portion of the valve forms the crista terminalis of the adult heart, whereas the caudal aspect of the valve forms the valves of the inferior vena cava (Eustachian valve) and coronary sinus (Thebesian valve). First described by Hans Chiari in 1897, the right sinus venosus valve may fail to resorb and will remain as a large fenestrated network of tissue, with additional attachment sites in the wall of the right atrium or the interatrial septum. Whereas the Chiari network will originate from either the Eustachian or Thebesian valve and attach to the upper wall of the RA or the IAS, the Eustachian valve does not, and should not be termed a Chiari network, even if it is mobile and fenestrated.

Found in about 2% to 3% of hearts at autopsy, the incidence detected by transthoracic echocardiography is very low (<0.6%), but by transesophageal echocardiography (TEE) is comparable to that at autopsy. Of patients found by TEE to have a Chiari network, most will have a patent foramen ovale (PFO). These PFOs tend to be anatomically large, and display significant shunting with peripheral vein injection of agitated saline contrast. An atrial septal aneurysm (ASA) also appears to be associated with the Chiari network. In a study evaluating patients with embolic stroke, of those with a PFO and ASA, a significant number (6/32) had a coexistent Chiari network.

The Chiari network is often considered clinically insignificant. However, it has been associated with catheter entrapment, right atrial thrombus entanglement, fetal arrhythmias, and patients with embolic stroke. Its association with embolic stroke probably is related to the relatively high association with PFO and ASA. The primary role of the echocardiographer is to recognize this as the Chiari network, and to distinguish it from other normal structures (Eustachian valve, Thebesian valve) or abnormalities such as thrombus, vegetation, tumor, or tricuspid chordal rupture.
References