Chiari's Network Incidentally Detected in Athletes Undergoing Echocardiography Screening

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Abstract
There is a growing desire for sport physicians to screen athletes with echocardiography. Echocardiography is a fundamental diagnostic tool that is increasingly being used today as a screening test in the context of sports medicine. The applications of this noninvasive cardiovascular imaging are in continuous development; so the potential to identify normal anatomic variants of the heart increases. Here the author describes some typical cases of Chiari's network that were detected during the course of sports preparticipation screening. Chiari's network is a congenital remnant of the right valve of the sinus venosus. Chiari's network is often found as an incidental finding on diagnostic cardiovascular imaging, but its clinical significance should be well understood and evaluated.

Keywords — Chiari's network, echocardiography, sports preparticipation screening, athletes.

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I. INTRODUCTION

In the cardiovascular diagnostic imaging with echocardiography it is common to find the presence of benign abnormalities such as the normal anatomic variants called Chiari's Network. Chiari's network is a congenital remnant of the right valve of the sinus venosus, and it is present in approximately 2% of the population. It has been found in 1.3% to 4% of autopsy studies and is generally of no clinical significance. In studies using conventional Two-dimensional transthoracic echocardiography (2D TTE) the reported prevalence of Chiari's network varies considerably, ranging from 0.03% to 9.5%. The use of echocardiography is increasingly common in the setting of sport pre-participation screening and has brought significant benefits to the discovery of undetected heart diseases, but sometimes the anomalies detected can generate clinical doubts. This is the case of Chiari's network which enters into the differential diagnosis of some pathologies of the right atrium. It has been reported that Chiari's network is involved in the pathogenesis of thromboembolic disease, infective endocarditis, arrhythmias, and entrapment of catheters upon percutaneous intervention. For this reason it may be mistaken for a thrombus, or other cardiac masses, or valvular vegetation.

The association with Chiari's network, which theoretically by itself may be a source of thrombi, is thus clinically important in relation to its association with other two risk factors, namely PFO in combination with atrial septal aneurysm.

In addition to congenital associations, several publications have highlighted the clinical significance of Chiari's network in cardiac flow obstructions, heart murmurs, infective endocarditis, and cardiac tumors. The early literature regarding this cardiac anomaly suggested it had minimal clinical significance. However the hypothesis has been raised that the network could play a role in thrombus formation as well as the entrapment of thromboemboli.

In literature the obstruction of cardiac blood flow for example was reported, as a possible complication of a large Chiari’s network.

In his report Latifi describes a case of infective endocarditis on a papillary fibroelastoma attached to Chiari’s network in a 41-year old woman who presented with fever and chills without evidence of structural heart disease and no vegetation on any cardiac valve. Another case of an older male who presented with infective endocarditis of the tricuspid valve was reported. The transthoracic echocardiography revealed persistent vegetations adherent to the tricuspid valve, extending onto Chiari’s network. Neurocutaneous syndromes, such as neurofibromatosis are associated with cardiac and extracardiac tumors, as well as other cardiac anomalies. For example in a report by Koz et al. a case of a giant network of Chiari’s mimicking a right atrial tumor originating from interatrial septum was illustrated. Therefore it is possible that a network of Chiari’s can be accidentally mistaken for a tumor of the heart.

In current clinical practice, the only clear link between paradoxical embolism and PFO, is when there is an atrial septal aneurysm. In the guidelines of congenital heart disease, it is
strongly advised to close a PFO in the presence of an atrial septal aneurysm, after a cerebrovascular ischemic attack, whereas in the absence of an atrial septal aneurysm closure of a PFO has so far not shown to be beneficial in the prevention of recurrence of paradoxical embolism.

Here the author describe the cases of Chiari’s network incidentally detected during the course of sports preparticipation screening last year between January and December, in which 400 athletes underwent two-dimensional transthoracic (2D TT) echocardiography.

II. OUR EXPERIENCE

Last year 400 athletes underwent echocardiographic examination in our Sports Cardiology Medicine Center for sports preparticipation screening and fitness physical evaluation.

Written informed consent for publication of this study and the accompanying videoclips was obtained from all athletes.

A two-dimensional transthoracic echocardiography was performed in all routine transducer positions with Esaote MyLab 30 Gold ultrasonograph and a 2.5-MHz transducer probe. All images were recorded on S-VHS videotape by sonographer and independent review was done by two experienced echocardiographers in a frame-by-frame analysis for the presence of Chiari’s network in the right atrium. Chiari’s network was diagnosed if coarse or fine fibers were observed in the right atrium, originating from an eustachian or thebesian valve at the orifice of the inferior vena cava or the coronary sinus and demonstrating attachments to the upper wall of the right atrium or the interatrial septum. Care was taken to differentiate a network of Chiari’s from a large eustachian valve by looking carefully for attachments to other parts of the right atrium. Chiari’s network was found in 12 of 400 consecutive athletes undergoing 2D TTE (prevalence 3%; 4 women - 8 men; mean (+SD) age 32 ± 10 years).

Chiari’s network was clearly visualized in all echocardiographic windows: on apical 4 chamber (see video 1), on parasternal long axis view, right ventricular inflow (see video 2), on long axis subcostal view (see video 3), and on parasternal short axis view (see video 4), and modified apical 4 chamber view with focus on the right heart side if the heart (see video 5), where Chiari’s network seems a thrombus.

In all athletes 2D TT echocardiography disclosed a large, highly mobile, reticulated network of fibers originating from the Eustachian valve connecting to different parts of the right atrium. However the fine attachments to other parts of the right atrium, meeting the criteria of Chiari’s network, were identified only in half the cases. The network was highly mobile, with an oscillating or whiplike motion pattern during each cardiac cycle. In nine (75%) of twelve athletes Chiari’s network was associated with a large eustachian valve. A thebesian valve at the orifice of the coronary sinus was present in one of these patients. The sites of attachment were multiple, including the upper region of the right atrium, the interatrial septum, the lateral wall of the right atrium and the tricuspid valve area. Thrombus-like material in the network could not be detected in any athletes. All subjects examined were asymptomatic and no other anomalies were detected.

III. DISCUSSION

During early embryogenesis, two venous valves protect the right horn of the sinus venosus. The smaller left valve becomes incorporated into the septum secundum. The larger right valve becomes incorporated into the right atrium (RA). Normally, the right valve regresses between the 9th and 15th gestational weeks. The cephalad portion of the valve remains as the crista terminalis. The caudal portions divide forming the eustachian (EV) and thebesian valves (TV). Failure of regression gives rise to Chiari’s fenestrated network. The eustachian valve (EV) or the inferior vena cava (IVC) valve, is located at the junction of the IVC and right atrium (RA). Embryologically, the EV is not typically seen on echocardiography, CT, or cardiac MRI. Yet, on occasions persistent fetal remnants of the valve referred to as Chiari’s network may be seen as in this case and should not be misinterpreted for a pathologic process. On both echocardiography and real-time cinematic cardiac MRI imaging, the EV remnant or Chiari’s network appears as a thin, linear, mobile structure attached at the junction of the RA and IVC. This remnant may even extend to the fossa ovalis. This fetal remnant is usually of no any clinical significance. However right heart catheters and other central venous devices may occasionally become entangled within this structure.

Hans Chiari described for the first time in 1897 15 eleven cases in which a network of threads and fibers was found in the right atrium. These reticula were in connection with the eustachian and thebesian valves at the orifice of the inferior vena cava and the coronary sinus, and showed attachments to the upper region of the right atrium near the crista terminalis, to the interatrial septum or to the tubercle of Lower. The finding echocardiographic network imposes some clear clinical considerations: a) its associations with the presence of a patent foramen ovale in about 80% of patients, reported in some studies, with intraatrial thrombus and thromboembolic events, and with atrial aneurysm. Furthermore its presence has recently been also correlated with infective endocarditis. Cardiac arrhythmias in association with Chiari’s networks are also being reported. There are reports of Chiari’s networks as a physical barrier which interferes with the endovascular procedures inside the right heart side. Chiari’s network is often diagnosed incidentally with echocardiography as a right asymptomatic atrial mass. Both transthoracic and transthoracic echocardiographies are reliable methods to diagnose Chiari’s networks. The Differential Diagnosis is from: 1) Prominent eustachian valve; 2) Valvular disruption; 3) Valvular vegetation; 4) Atrial septal aneurysm; 5) Various cardiac masses. The most common ancillary form of investigation in sport cardiology medicine is the echocardiography. Echocardiographers can visualize Chiari’s network with almost all standard approaches of echocardiographic examination. The classic windows to
visualize Chiari’s network is the long axis parasternal view (right ventricular inflow tract), but also the short-axis parasternal, subcostal view, and the apical 4 chamber view are more efficacious. Chiari’s network appears as a thin, web-like, fenestrated membrane at the orifice of the inferior vena cava. On real-time imaging, this network appears very impressive by its chaotic, random motion and by its highly reflective appearance within the right atrium. In most of our subjects Chiari’s network clearly took origin from the hole in the lower vena cava, at posterolateral right atrium and coursed inferomedially toward the tricuspid ring. Most athletes have a predominantly rather low horizontal structure in the right atrium immediately above the tricuspid ring and apparently associated with interatrial septum, while an intermediate position is observed in a small number of patients. Only few athletes showed a very small structure, light and away from the apparatus tricuspid (near right atrium), and probably should not be confused with the tricuspid apparatus itself. In other athletes the structure was highly reflective and the close tricuspid ring was indistinguishable from a tricuspid septal, partially interrupted leaflet valve or vegetation. In these difficult cases even for trained echocardiographers the definitive diagnosis is made only by transesophageal echocardiography. However, an experienced operator can clearly discriminate the network from the pathologies of the right atrium, fortunately because the clinic comes to the rescue.

IV. CONCLUSIONS

Chiari's network is often found as an incidental finding by diagnostic cardiovascular imaging, but its clinical significance should be well understood and evaluated. However, in athletes it is reasonable to believe that the discovery of Chiari’s network should be considered a benign anatomical variant, and therefore without any complication. Its differentiation from right-heart pathology is easy for expert echocardiographers, while it can be difficult for less expert echocardiographers. It has to be recognized appropriately, otherwise it may lead to misdiagnosis. Obviously the diagnosis must be certain and free from errors of echocardiographic interpretation, with the support of the clinical data and medical history. For this reasons clinicians, sports cardiologist and echocardiographers should keep this in mind in order to avoid diagnostic dilemma.

V. SUPPLEMENTAL DATA

Supplemental videos (1-5) are available online: http://www.researchpub.org/journal/jcvd/jcvd.html

REFERENCES

Figure 1. Chiari’s network, showing reticulated structure extending from the sinus atrial node (on the left) to atrial ventricular node (on the right).