

# The Chiari network: an anatomical variation or a risk factor?

## Case report

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### SUMMARY

We report an association between a Chiari network and an abnormally long coronary sinus. The network, at the Eustachian valve and a morphologically similar network in the Thebesian valve, was also associated with a permeable foramen ovale. We review the embryological basis, associated anomalies, pathological conditions and clinical relevance.

**Key words:** Chiari network – Embryological heart development – Vena cava valve – Coronary sinus valve

### INTRODUCTION

In human anatomy the concept of normality includes a range of common morphologies, while less frequent morphologies considered normal are described as variations.

The Chiari network is a fenestrated structure consisting of strands originating from the cava inferior valve in the right atrium of the heart. It is considered to be an anatomical variation, with an incidence of about 1-3% in the general population and was

described in the 19th century, becoming important after the introduction of Echocardiography. Its morphology has been studied by echography and it plays a role in differential diagnosis with some right heart pathologies (Goedde et al., 1990; Patane et al., 2009).

The variation is usually discovered during diagnostic imaging studies, necropsies, and surgical procedures (Loukas et al., 2010).

Although it has often been considered clinically insignificant, it has been associated with some pathologies (Loukas et al., 2010), such as a patent foramen ovale, atrial septal aneurysms, and paradoxical embolisms. It has also been described as associated with papillary fibroelastomas (Wasdahl et al., 1992), arrhythmias, and the development of tumours (Stanley, 2001).

Some of these associations may be a consequence of the behavior of the net itself, like the net prolapsed in the tricuspid valve orifice during the diastole period. Other associations can be explained through the study of embryological heart development.

In this report, we describe the association of a Chiari net associated with other embryological heart anomalies.

## CASE REPORT

We observed the presence of a Chiari network in a heart extracted from a male cadaver in the dissection room of our Medical School.

The heart was 165 mm in length and 287 g in weight. Its cavities displayed the following features:

The interatrial septum showed a permeable foramen ovale (Fig. 1).

The inferior vena cava opening into the atrium was 26.5 mm in diameter and 80 mm in length.

The network in the inferior cava opening, exhibited an almost semicircular shape with a base of 60 mm, representing about 75% of the total length of the opening, and it has a maximum width of 35 mm from its adherent border to the free border. A festooned veil spread out from the adherent border. This veil was formed by a superposition by several arches up to the free border (Fig. 2).

The coronary sinus was 80 mm in length between its orifice in the right atrium and the Vieussens valve, which separate the coronary sinus from the great cardiac vein. The left part of the coronary sinus was 15 mm wide and the right part 20 mm wide. The coronary sinus atrial opening was 14.7 mm in diameter and 46 mm in length (Fig. 3).

The net in the coronary sinus valve was inserted in the more external part of the orifice and covered 50% of the sinus orifice. It was 23 mm in length at its adherent border and 15 mm at its maximum width (Fig. 4). The network's morphology was similar to that of the inferior vena cava.

The histological study revealed that the network was mainly composed of fibrous tissue and some muscle cells (Fig. 5).

## DISCUSSION

The Chiari network is a congenital remnant of the right valve of the venous sinus and it is considered to be an anatomical variation, which overall presents few clinical manifestations. However, this network has been associated with other defects in embryological heart development (Schneider et al., 1995), with alterations in cardiac rhythm (Hajime et al., 1992; Doig et al., 1995), and with other heart pathologies (Benbow et al., 1987; Wasdahl et al., 1992; Loukas et al., 2010).

Many of the pathological associations with the Chiari network may be consequence of the behaviour of the network itself. The network could act as a natural filter at the inferior vena cava for the formation or trapping of thrombi (Benbow et al., 1987) and it may be a place where certain processes such as endocarditis and tumours can develop. In some cases, networks of large dimensions may occupy other heart cavities (Cooke et al., 1999), or may be a risk factor in radiofrequency ablation procedures in patients with arrhythmias. As a consequence, the Chiari network may predispose to wire entrapment (Shimoike et al., 2001).

Other associations of the Chiari network can be explained in terms of the processes involved in the embryological heart development and the relationship between the venous sinus and the development of the cardiac con-



Fig. 1. In the opened right atrium a patent foramen ovale is shown on the atrial septum.



Fig. 2. Diaphragmatic surface of the heart and opened right atrium. The veil structure of the Chari network is shown.

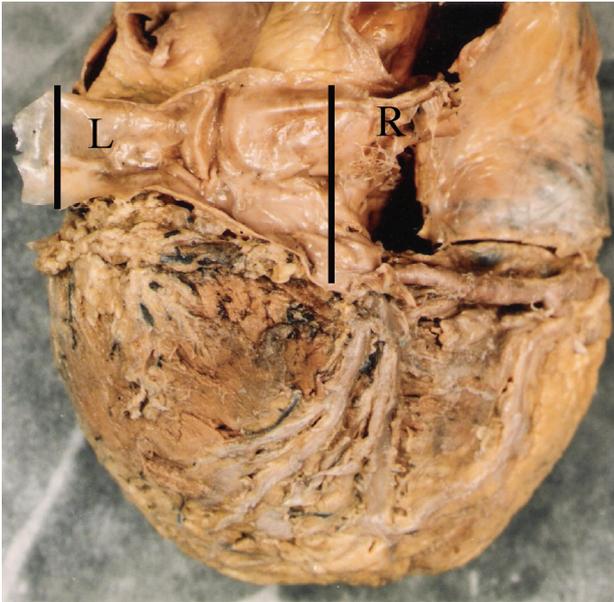


Fig. 3. Diaphragmatic surface of the heart. The coronary sinus is opened. It is possible to observe the differences between the right and the left part of the coronary sinus.



Fig. 4. Diaphragmatic surface of the heart. The coronary sinus lies in the atrioventricular groove. A network is shown in its opening into the right atrium.

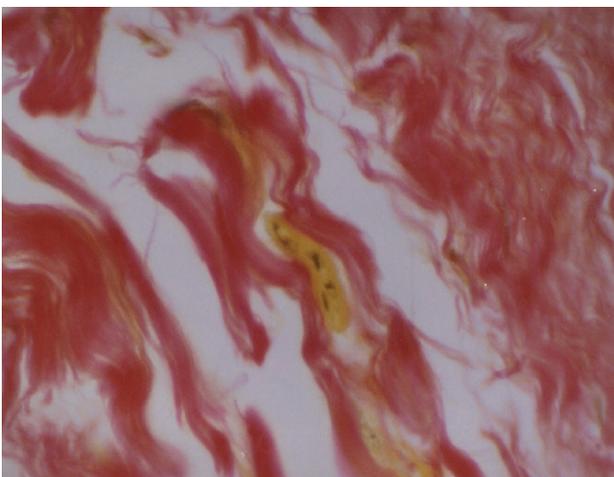


Fig. 5. Van Giensson staining of the Chiari network. Connective tissue in red, muscle cells in yellow. x 160.

duction system. During embryological development, the right horn of the venous sinus increases to the detriment of the left horn, and before its incorporation in the right atrium the venous sinus opens into the right atrium; the sinus atrial-orifice is formed and is guarded by right and left venous valves. These valves fuse rostrally as the septum spurium. At the inferior end, the two valves fuse with the dorsal endocardial cushion of the atrium. The embryological development of the right sinus valve forms the crista terminalis, the intervenous tubercle, the valves of the coronary sinus, and a large part of the inferior cava valve.

The sinus septum, whose origin is located in the region where the right and left horn meet, takes part in the subdivision of the right sinus valve into the Eustachian and Thebesian valve. The Eustachian valve is formed by the superior portion of the right sinus valve, and its left end by a small fold that spreads from the sinus septum. The Thebesian valve is formed by the inferior portion of the right sinus valve. The left sinus valve fuses with the right part of the interatrial septum, and at the same time the left horn of the sinus venosus forms the coronary sinus.

The heart studied showed at least two structural modifications, a consequence of defects in heart development; a) a patent foramen ovale and b) an abnormally long atrial coronary sinus opening.

In the reported case, the heart showed a patent foramen ovale. The recognition and treatment of a permeable foramen ovale has attracted interest due to the importance and frequency of paradoxical embolism (Price et al., 2004) and a causal relationship between a patent foramen ovale and atrial septal aneurysm and neurologic ischemic events has been considered in patients younger than 55 years old. A patent foramen ovale has also been related with decompression sickness in divers and astronauts, and with migraine headaches (Kerut et al., 2001; Holmes et al., 2004). The incidence of a patent foramen ovale in autopsy patients in the general population is 27.3% with no sex differences (Hagen et al., 1984). The association between Chiari network and a patent foramen ovale is 83% (Schneider et al., 1995).

During the embryological development the cava valve directs the blood towards the interatrial foramen. The high incidence of association between the Chiari network and a persist-

ent foramen ovale can be attributed to the defect in the regression of the right venous valve. In this case, the valve, which had not regressed, may have created abnormalities in intraatrial circulation, causing difficulties in the closing of the foramen (Schuchlenz et al., 2004). The presence of a Chiari network should alert about the presence of a patent foramen ovale.

The heart studied showed an 80 mm-length sinus coronary, which is considered normal. However, its morphology was not tubular but resembled a wind sock with an abnormally large atrial coronary sinus opening diameter. The ostium diameter was 14.7 mm as compared with control dimensions  $8.5 \pm 1.5$  mm. The ostium, in this case, was 43% larger than in the controls. This association, a long coronary sinus ostium, longer than  $12.2 \pm 2$  mm, and coronary sinus with a wind sock shape, has been shown to be related to the presence of atrioventricular junctional reentry tachycardia (Doig et al., 1995).

Alterations in cardiac rhythm often seem to be frequently associated with the Chiari network. In this sense, a relationship between the structures derived from the venous sinus and those of the development of the cardiac conduction system has been described.

The conducting system of the heart appears at 4.5-5 weeks of embryological development and is well developed by 8 weeks. The cells of the conducting system are related to structures in the venous sinus and structures derived from the right venous valve, such as the cresta terminalis and the inferior vena cava valve. Insufficient absorption may occur in conjunction with abnormalities of the cardiac conduction system and alterations in cardiac rhythm.

## CONCLUSIONS

Although the Chiari network can be considered an anatomical variation without pathological consequences, it may represent a potential risk for patients.

The presence of a Chiari network should alert physicians about three kinds of clinical consequences:

1. The network in the right atrium may be herniated in other cavities, it could be a site for guide wire entrapment in patients subjected to ablation procedures for arrhythmias and an area where infection and tumour can develop.

2. The network may be associated with a permeable foramen ovale and its complications, such as a paradoxical embolus or atrial septal defects.
3. The Chiari network can be associated with alterations in heart rhythm.

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