# An unusual Chiari Network: Potential Risk in Cardiac Interventional Procedures

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# **ABSTRACT**

The Chiari network, seen in the right atrium, is a remnant of the right valve of the sinus venosus, resulting due to the incomplete regression of the valve during development. It is found in approximately in 1.5 to 3% individuals. During routine dissection for the first year M.B.B.S. students, we came across a cadaveric heart that exhibited an unusual Chiari network. The network observed was a combination of thread like strands and mesh with fenestrations. It was attached to the right margin of the opening of the coronary sinus. The shape of the coronary sinus opening was oval, with a transverse diameter of 15.2 mm and a maximum width of 6.5 mm. No other anomaly of the right atrium was found. The Chiari network is commonly related with patent foramen ovale and with thromboembolic phenomenon. Though found incidentally, its importance cannot be overlooked as the network is possibly associated with complications during certain intracardiac interventional procedures. The authors have presented this case with a view to add to both, the morphological and pictorial data of the said developmental variation.

**KEYWORDS:** Chiari network, coronary sinus opening, fenestrated, mesh.

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# **INTRODUCTION**

The Chiari network, seen in the right atrium, is a remnant of the right valve of the sinus venosus resulting due to incomplete regression of the valve during development. It is found in approximately in 1.5 to 3% individuals. It was first elaborated upon by Hans Chiari in 1897 [1]. Chiari network presents as an abnormal network of threads and fibres, lace like in nature, found in the right atrium at the orifice of the inferior vena

cava and the coronary sinus, showing attachments to the upper region of the right atrium near the crista terminalis, to the interatrial septum or to other parts of right atrium.

Embryologically the right horn of the sinus venosus gets incorporated into the wall of the primitive right atrium during development. The right valve of the sinus venosus then contributes to the formation of the Crista terminalis, valve of Inferior Vena Cava and valve of Coronary Sinus [2].

The coronary sinus, that drains most of the venous blood from the heart opens into the right atrium, posteromedially between the inferior vena cava and the right atrioventricular orifice. The coronary sinus opening is guarded by the Thebesian valve, which has been found to have a variable morphologytype I(remnant), type II(semilunar), type III(fold), typeIV(cord)and type V(mesh and fenestrated) [3]. The Chiari network appears when there is failure of resorption of certain parts of the right valve of the sinus venosus.

With the use of echocardiography it has become apparent that the network is possibly associated with complications during certain intracardiac interventional procedures [4]. Studies by various authors have shown concurrence of clinically important cardiac abnormalities such as Patent Foramen Ovale, Atrial Septal Defect and Interatrial Septal Aneurysms with this anomaly [5,6]. It may sometimes produce diagnostic confusion in echocardiography of tumors and thrombii of right atrium. The network may also act as a site for thromboembolism and infective endocarditis [7]. Although found incidentally its importance cannot be overlooked. Since there is paucity of anatomical literature of this anomaly the authors have presented this case with a view to add to both the morphological and pictorial data of the same.

## **CASE REPORT**

During the routine teaching learning session of dissection for the first year undergraduate M.B.B.S. students, we came across a cadaveric heart that exhibited an unusual Chiari network. The formalin embalmed cadaver was of a male, aged 62 years whose cause of death was unknown. The dissection of the heart was done, as per the standard steps outlined in the Cunningham's Manual of Practical Anatomy, Volume 2. The Right atrium was opened by a vertical incision extending from the orifice of the superior vena cava to the orifice of the inferior vena cava. Care was taken to keep the valve of the Inferior Vena Cava (IVC) intact. On examining the interior of the heart, the valve of the IVC and valve of the coronary sinus, we found a large fenestrated structure over the opening of the coronary sinus. It was attached to the right margin of the opening of the coronary sinus, extending to the anterior and posterior margin of the coronary opening on the right side, covering more than two thirds of the opening. There were multiple thread-like strands to the left and a sieve-like fenestrated structure towards the right. No other anomaly of the right atrium was found. The structure observed was ascertained to be a Chiari network that was a combination of thread like strands and mesh like fenestrations unlike any one of the enumerated types I, II, III, IV or V. The coronary sinus opening was observed and the shape of the opening was found to be oval. Using a vernier caliper measurement of the dimensions of the coronary opening was taken. A transverse diameter of 15.2 mm and a maximum width of 6.5 mm was observed. All descriptions and measurements were taken keeping the heart in anatomical position.

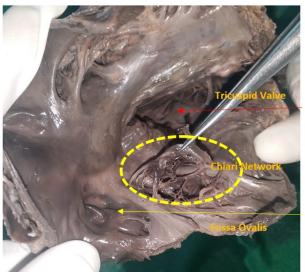


Fig. 1: Superior view of Right atrium showing the Chiari network covering the opening of Coronary sinus Red arrow showing Tricuspid valve, Yellow arrow showing Fossa Ovalis and Yellow dotted oval with star showing Chiari network

#### DISCUSSION

Chiari's network has been considered as a normal anatomic variant that is rarely of clinical importance. But certain associations are noteworthy like its occurrence with other cardiac anomalies like patent foramen ovale, atrial septal defects and interatrial septal aneurysms. <sup>[5,6]</sup> This can be explained with the function of the coronary sinus valve during

embryonic life. The Thebasian Valve helps in directing the blood flow from the inferior vena cava preferentially toward the region of the fossa ovalis. Hence, it may favor the persistence of a patent foramen ovale.

With use of transesophageal echocardiography nowadays, identifying the fine threads and fibers seen attached to the upper region of the right atrium and to the interatrial septum has become easier. [5] Thus helping in differentiating the Chiari network from atrial septal tumors or atrial septal aneurysms that may cause an error in diagnosis.

This anomaly has also been suspected to be a cardiac source of arterial embolism [5].

Amongst the newer cardiac techniques for resynchronization therapy the Coronary Sinus is used for left ventricular epicardial lead placement. Presence of network around the opening of the sinus may cause entrapment of right heart catheters, guidewires and pacemaker leads used during invasive procedures leading to procedural difficulties [4].

Even though the Chiari network is found incidentally we authors have seen a change in opinion regarding the importance of this structure in various studies and case reports published since 1995 onwards [3-8].

We see an increasing trend in the viewpoint that this structural anomaly may have a clinical significance in the field of cardiology and interventional cardiology. The authors have presented this case with a view to add to both the morphological and pictorial data of the said anomaly towards the same opinion.

### **ABBREVIATIONS**

**M.B.B.S:** Bachelor of Medicine and Bachelor of Surgery;

IVC: Inferior Vena Cava.

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# **Author Contributions**

**PB** drafted the manuscript.

**NK** provided the technical support.

**PVS** commented on the draft. All authors read and approved the final manuscript

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**Conflicts of Interests:** The authors declare that there are no conflicts of interests.

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