

1959

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Recommended Citation

Dzmura, A. R.; Miroyiannis, S. D.; and Enzmann, E. V. (1959) "An Interesting Cardiac Anomaly: Two Cases of Chiari's Network," *Proceedings of the Iowa Academy of Science*, 66(1), 558-563.

Available at: <https://scholarworks.uni.edu/pias/vol66/iss1/75>

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An Interesting Cardiac Anomaly: Two Cases of Chiari's Network

By A. R. DZMURA, S. D. MIROYIANNIS, and E. V. ENZMANN

Abstract. Two hearts were found with anomalous reticula of the venous valves of the right atrium. Fibrous cords extending from the eustachian and thebesian valves to the atrial wall in the area of the crista terminalis corresponded to the original descriptions of Chiari's network. No adequate clinical history was available, and no attempt was made to evaluate the clinical significance of these anatomical findings. The literature is summarized and tabulated with respect to the possible relationship of the presence of Chiari's network to pathological conditions.

In 1936, Wallace Yater wrote as follows in "The Paradox of Chiari's Network": "It is hoped that the title of this paper will stimulate interest in a cardiac anomaly which is not exactly rare but concerning which there is little general knowledge."

Since that time, some interest has been exhibited, but little more information has been brought to light concerning these delicate webs in the human heart which extend from the eustachian and thebesian valves to the crista terminalis or the interatrial septum. Various authors estimate the frequency of this anomaly at about two to three percent, although it is probably somewhat higher, since the right atrium is generally opened in dissection by cutting through the orifices of the vena cavae, a procedure which would destroy these fine strands.

The two cases presented here are from our dissection laboratories and it is unfortunate that no accurate histories were available. The description, however, may be of value to future investigators.

Case No. 1. The weight of the heart was 296 grams. Superficially, the right atrium had chorda tendinae-like structures which extended from the eustachian and thebesian valves to the lower portion of the crista terminalis. More specifically, the network began as two thin, fibrous cords attached to the atrial wall above the orifice of the inferior vena cava, between the right portion of the limbus of the fossa ovalis and the crista terminalis. A third cord, originating on the atrial wall inferiorly and laterally to the first two cords, united at a point into a single and thicker band which bifurcated immediately. The more lateral branch divided into three thin fibers which were continuous with the dentate processes on the eustachian valve. The medial branch of the bifurcation continued for 1.5 cm to the largest portion of the eustachian valve, medial to the part just described. It separated into three branches, two of which formed an

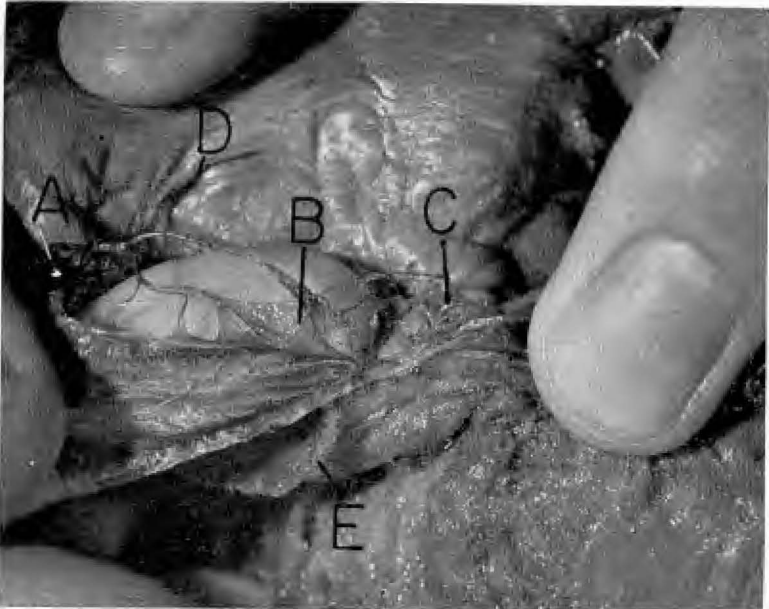


Figure 1. Case No. 1. A, attachment of network to area of crista terminalis; B, eustachian valve; C, network over the coronary sinus; D, limbus fossa ovalis; E, cut rim of inferior vena cava.



Figure 2. Case No. 1. Superior view, looking into orifices of inferior vena cava and coronary vessels.

irregularly branching and anastomosing network attaching itself to the medial extremity of the eustachian valve, and the third branch became a small network which attached itself to the thebesian valve over the coronary sinus. The eustachian valve was 7 mm. wide at its widest point, and 3.4 cm. long. It was situated on the anterior rim of the inferior vena cava. The thebesian valve was situated anteriorly and laterally on the rim of the coronary sinus (Figures 1 and 2).

Case No. 2. The weight of this heart was 181 grams. Here the network originated as a flattened, fibrous band arising on the medial extremity of the crista terminalis. This band then extended and became slightly narrowed. Immediately over the orifice of the inferior vena cava a single strand separated itself and became attached to a dentate process on the eustachian valve; the main portion of the band then formed into a large fenestrated membrane which twisted upon itself assuming the appearance of a pedicle suspended freely over the orifice of the inferior vena cava. At the base of this structure, the main band became separated into a branched network which was attached to the remaining part of the eustachian valve. A few small strands extended to the thebesian valve (Figure 3).

DISCUSSION

Most authors agree that Chiari's network is an insignificant anomaly, compatible with life, and probably produces no symptoms.

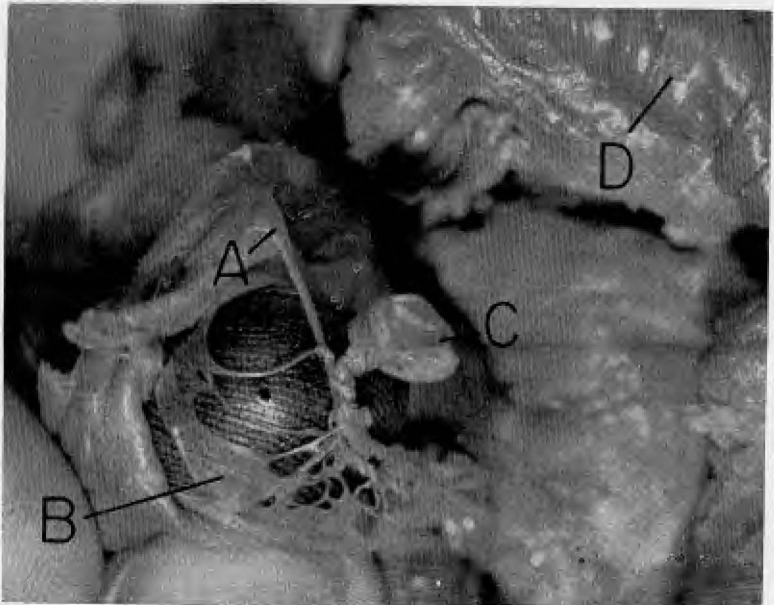


Figure 3. Case No. 2. A, attachment to crista terminalis; B, eustachian valve; C, twisted, fenestrated membrane; D, musculi pectinati.

Table 1 lists the case reports of Chiari's network. Table 2, compiled from the literature, shows most of the reports of pathological states or symptoms which have some relationship, real or postulated, to the presence of a Chiari network. In all of these, save Chiari's case 1, there was no proof that the network was the cause of fatal pathology. Chiari (1897) thought that the network was the site of formation of a thrombus which caused a pulmonary embolism. In Haas's case (1916), and one of Yater's cases, it was thought that the network trapped an embolus and prevented death; thus, "The paradox of Chiari's network".

The present authors suggest that the following points emphasize the possible pathological significance of Chiari's network:

1. Thrombi and emboli have been found in the network.

2. It can easily be imagined that a network of fibers suspended over the inferior vena cava, in direct line of flow of the blood, could be a source of trouble if the circulatory rate should become depressed, or if for any reason the blood became more coagulable.

3. Abbott (1926) pointed out that cardiovascular malformations offer a favorable base for nidation of bacteria and their multiplication.

4. A catheter passed into the right atrium might be deflected by the network, or it might break the net. Trabeculae have been reported in cardiac catheterizations. If catheterization were performed on a heart such as Yater's case 4, which contained a flat band of

Table 1
Reported Cases of Chiari's Network

Author	Number of Cases	Year
Chiari	11	1897
A. Weber	1	1898
Le Count	1	1901-03
Looser	1	1902
Ebbinghaus	1	1904
Thilo	2	1909
Haas	1	1916
F. P. Weber	2	1920
Jordan	2	1926
Yater	4	1929
Heupner and Berghoff	2	1929
Alvarez and Herrmann	1	1931
Helwig	8	1932
Yater	1	1936
Wilson	1	1938
Evangelisti	1	1940
Wright, <i>et al.</i>	1	1948
Orbison	1	1949
Total	42	

Note: Mönckeberg (1924) mentioned 6 cases which he saw in Düsseldorf; he did not describe these.

cardiac muscle continuous with either side of the atrium near the orifice of the inferior vena cava, and which was probably the path of a circus movement, instrumentation on such a heart might conceivably initiate flutter.

ACKNOWLEDGMENTS

The authors desire to express their appreciation to Mr. E. Lynn Baldwin, Director of the Department of Medical Photography of this institution, for his painstaking photographic work.

Table 2

Cases of Chiari's Net Associated With Various Types of Pathology

Case	Type of Pathology or Symptom	Relation of Net to the Pathology
Chiari Case 1	Net was the site of thrombus formation which produced pulmonary embolism.	Caused death of the patient.
Looser	2 cm. thrombus attached to the net.	Other thrombotic areas were found which were probably the true sources of emboli.
Thilo	A grayish-red "pea-sized" spherical thrombus attached to the net.	Other thrombotic areas existed.
Haas	6 cm. embolus ensnared in the net.	Prevented fatal pulmonary embolism.
Yater (1929) Case 11	Thrombus attached to the network.	Other thrombotic areas.
Yater (1936)	Network ensnared a large embolus.	Prevented fatal pulmonary embolism.
Alvarez and Hermann	Syphilitic heart disease and aortic regurgitation with congestive failure.	Network may have obstructed blood flow contributing to the failure; net produced a peculiar murmur.
Wilson	Hypertensive vascular disease with arterial nephrosclerosis and lobular pneumonia.	Net produced a murmur resembling the bruit de Roger.
Evangelisti	Acute endocarditis of medial zone of tricuspid valve with congenital defect of I. V. septum, neurinomas of the adrenals.	Apparently the network had nothing to do with any of these.
Orbison, J. L.	Thrombi on the network, and emboli in the pulmonary arteries. No other origins for emboli were found.	These thrombi may have caused the emboli in the pulmonary arteries. The author states, however, that the leg veins were not examined for the presence of thrombi.

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