

Prevalence and Clinical Profile of Chiari network in Bangladeshi subjects: Analysis of 1832 subjects by Echocardiography

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Abstract:

Keywords:
Chiari network,
Embryonic
Structures,
Right Atrium,
Bangladesh.

Background: The Chiari network is mobile, serpentine, filament-like structures occasionally seen in right atrium near the opening of inferior vena cava and coronary sinus. Sometimes it may cause diagnostic confusion with right atrial pathologies.

Methods: This was a cross sectional study conducted from September 2017 to August 2018 in a private consultation center in Dhaka. All patients who underwent transthoracic echocardiography were included. Chiari network was diagnosed and any relevant clinical condition was recorded.

Results: Out of 1832 echo studies, Chiari network was found in 90 subjects giving a prevalence of 4.91%. More than half (55.6%) had otherwise normal heart, 20% had ischaemic heart disease, 7.8% had cardiomyopathy, 6.7% congenital heart disease, while 3.3% had valvular heart disease.

Conclusion: The Chiari network is often an incidental diagnosis, though it may not be so rare. Clinicians should be aware of it to avoid diagnostic dilemma and to overcome difficulties while performing cardiovascular interventions.

(*Cardiovasc. j.* 2019; 11(2): 93-97)

Introduction:

The Chiari network is a mobile structure occasionally seen in the right atrium, most commonly encountered during echocardiography. It may be characterized as reticular network of fine strands attached to right atrium¹ or, as membranous fenestration involving valve of inferior vena cava or coronary sinus, extending to crista terminalis and interatrial septum.² Embryologically, it results from incomplete resorption of right valve of sinus venosus. Its prevalence has variably been reported to be 2–13.6%.³⁻⁵

Traditionally, Chiari network has been considered as a benign anatomical entity having no clinical significance. Subsequently, the outlook

has been challenged, and it has been reported to cause diagnostic confusion,⁶ be associated with diverse clinical conditions,⁷⁻⁹ or act as a physical barrier to invasive procedures.¹⁰ The prevalence of Chiari network in Bangladeshi subjects is not known. Also, data regarding its association with other conditions are scanty. Here, a study was carried out to find out the prevalence of Chiari network by trans-thoracic echocardiography in an outpatient population, and associated conditions have also been studied.

Methods:

The study was carried out in private consultation centres of Dhaka City during September 2017 to August 2018. All patients sent for

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transthoracic echocardiography were included. Echocardiographic examination was carried out by Vivid E95 (GE Healthcare, 9900 Innovation Drive, Wauwatosa, WI 53226, USA). The examination was carried out as per the recommendation of American Society of Echocardiography. Chiari network was sought for in all the standard views and also the modified views as needed. The network structures were arbitrarily classified into well-, moderately- and poorly-visualized on the basis of how organized they appear in 2D echocardiography.

Results:

In this study, 1832 subjects were examined. Majority of the patients with Chiari network belonged to middle-age group; 38.9% were <40 years of age while 34.4% were of 41-60 years. Almost 2/3rds of the study subjects (63.3%) were male. Out of 1832 echo studies, Chiari network

was found in 90 subjects giving a prevalence of 4.91%. Out of 90, well-, moderately- and poorly-visualized Chiari networks were 41 (45.6%), 28 (31.1%) and 21 (23.3%), respectively. More than half (55.6%) had otherwise normal heart, 20% had ischaemic heart disease, 7.8% had cardiomyopathy, 6.7% congenital heart disease, while 3.3% had valvular heart disease (Table I). In either sex, majority had otherwise normal heart. In male patients with Chiari network, IHD was the commonest diagnosis, whereas in female patients with Chiari network, cardiomyopathy was found most commonly, among those who have some sort of echo diagnosis. In younger age groups, Chiari network was more commonly associated with otherwise normal heart in echocardiography. As the age increases, probability of associated diseases increased, and beyond 80 years, all were associated with cardiac disease (Table II).

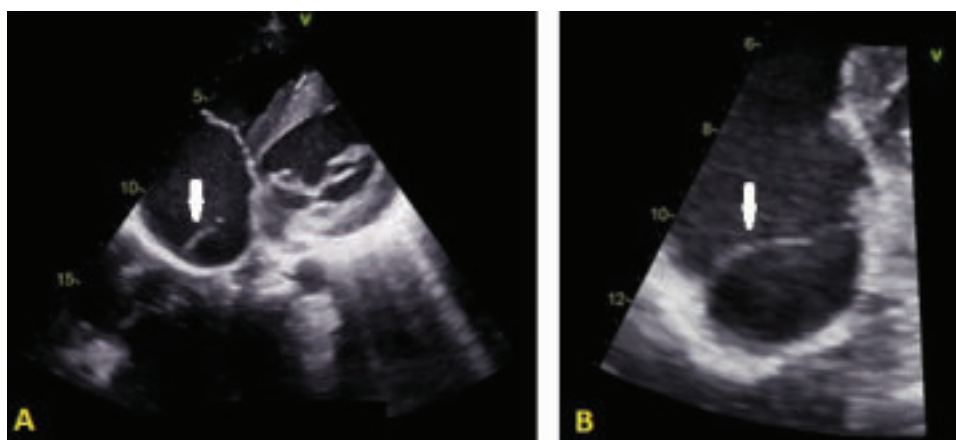


Fig 1: 2D echocardiography showing Chiari network in right atrium in off-axis view (Panel A), and zoomed up view (Panel B), marked by white arrows.

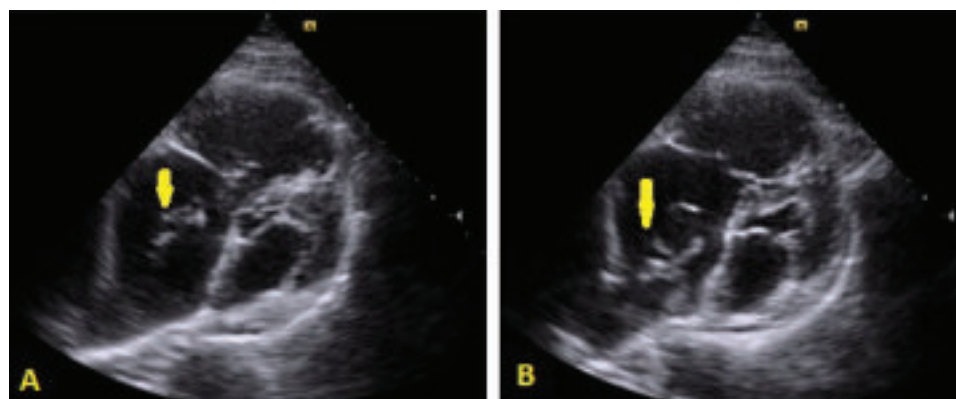


Fig 2: Panel A & B: 2D echocardiography of a case of pulmonary thromboembolism showing Chiari network in dilated right atrium in different phases of off-axis views marked by yellow arrows.

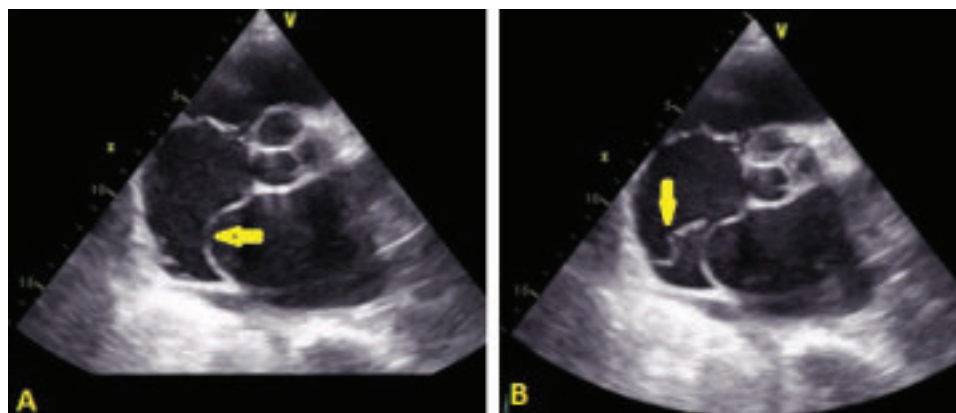


Fig-3: 2D echocardiography, parasternal short axis view. Panel A showing atrial septal aneurysm marked by arrow; Panel B showing Chiari network in right atrium marked by yellow arrow.

Table-I

Distribution of the study subjects with Chiari network by clinical diagnosis (n=90).

| Clinical diagnosis | Differentiation of Chiari network | | | Total Number (%) |
|--------------------------|-----------------------------------|------------------------|--------------------|---------------------|
| | Well Number (%) | Moderate Number (%) | Poor Number (%) | |
| Normal | 19(21.1) | 14(15.6) | 17(18.9) | 50(55.6) |
| Congenital heart disease | 5(5.6) | 1(1.1) | 0 | 6(6.7) |
| Valvular heart disease | 2 (2.2) | 0 | 1(1.1) | 3(3.3) |
| Ischemic heart disease | 6 (6.7) | 9(10) | 3(3.3) | 18(20) |
| Cardiomyopathies | 5(5.6) | 2 (2.2) | 0 | 7(7.8) |
| Others | 4(4.4) | 2 (2.2) | 0 | 6(6.7) |

Table-II

Relationship between age group and clinical diagnosis of the study subjects with Chiari network (n=90).

| Age Group (Years) | Clinical diagnosis | | | | | | Total Number (%) |
|----------------------|----------------------|--------------------------|------------------------|-------------------|------------------------------|----------------------|---------------------|
| | Normal Number (%) | Congenital Number (%) | Valvular Number (%) | IHD Number (%) | Cardiomyopathy Number (%) | Others Number (%) | |
| <41 | 23(25.6) | 4(4.4) | 1(1.1) | 2(2.2) | 1(1.1) | 4(4.4) | 35(38.9) |
| 41-60 | 17 (18.9) | 0 | 1(1.1) | 9 (10) | 3 (3.3) | 1(1.1) | 31(34.4) |
| 61-80 | 10 (11.1) | 2 (2.2) | 1(1.1) | 5 (5.6) | 3 (3.3) | 0 | 21(23.3) |
| >80 | 0 | 0 | 0 | 2 (2.2) | 0 | 1(1.1) | 3 (3.3) |
| Total | 50 (55.6) | 6 (6.7) | 3 (3.3) | 18 (20) | 7 (7.8) | 6 (6.7) | 90 (100) |

Discussion:

Chiari network was first described by Hans Chiari, an Austrian pathologist, in 13 human right atria in 1897.¹¹ Since then, its prevalence has been reported sporadically. Schneider et al.

conducted a prevalence study on Chiari network based on trans-oesophageal echocardiography (TOE); the prevalence was 2% (29 of 1,436 patients).⁴ In their study, Bhatnagar et al. found Chiari network in 13.6% of the 213 cadaver hearts,

and 10.5% of the 38 autopsied hearts examined.³ This apparent disparity in prevalence may be due to the way of diagnosing the Chiari network, i.e. imaging modality or cadaveric heart. However, the incidence of Chiari network in a more recent study involving 80 cadaveric hearts was 3.75%.¹² Another explanation behind the varying prevalence may be the criteria used for defining Chiari network. All structures found in the right atrium in relation to the IVC or coronary sinus is not well visualized, especially in imaging study. In fact, the prevalence of Chiari network of 4.91% found in the present study comes down to 3.77% when the poorly-visualized structures are excluded from calculation.

Chiari network is often regarded as an ‘innocuous bystander’ diagnosed incidentally. However, this structure is being increasingly recognized to have clinical importance – both diagnostic as well as therapeutic. This right atrial network may occasionally mimic thrombus, vegetation or mass.¹³⁻¹⁶ From the therapeutic point of view, Chiari network has been reported to be associated with thromboembolism,⁷ infective endocarditis,⁶ arrhythmia,⁷ and others. Contrarily, the sieve-like structure of Chiari network might play protective role against migration or propagation of thrombus formed in IVC to cause pulmonary embolism.¹⁹ During intervention including electrophysiological study and radiofrequency ablation, the catheter or guidewire may get entangled into the CN, causing procedural complications.²⁰⁻²²

In our observation, among the subjects with Chiari network, 20% had ischaemic heart disease, 7.8% had cardiomyopathy, 6.7% had congenital heart disease while 3.3% had valvular heart disease. Again, 4 patients had haemoglobinopathy, 2 patients hypertrophic cardiomyopathy, 2 patients atrial septal defect, 1 patient atrial septal aneurysm, and 1 patient had complex congenital heart disease (transposition of great arteries, ventricular septal defect and pulmonary stenosis). One of the 90 patients with Chiari network, a 45-year-old man, presented with pulmonary embolism, and he had well-developed Chiari network. Previously, Chiari network has been reported in association with other congenital heart diseases,

e.g., atrial septal defect and anomalous pulmonary venous drainage, atrial septal aneurysm, and Ebstein anomaly of tricuspid valve.⁹ Interestingly, we have found 4 cases of haemoglobinopathy having Chiari network; whether this observation is merely casual or a positive association needs further study.

The study has got some limitations. Echocardiographic evaluation was done on the referred adult patients who may not be representative of the general population. Also, only transthoracic echocardiography was utilized in this study; the prevalence of Chiari network may differ from that we have found if transoesophageal and 3-dimensional modalities would have been used. However, to the best of our knowledge, this is the first attempt to find out the prevalence of Chiari network and its association in Bangladeshi population.

Conclusion:

The Chiari network is often an incidental diagnosis, though it may not be so rare. This apparently normal anatomical variant may cause diagnostic confusion and have therapeutic implications. It may be associated with other conditions like congenital heart disease, hypertrophic cardiomyopathy, arrhythmia or even haemoglobinopathy. Whether these co-occurrences are merely casual or positive association needs further evaluation. Chiari network may predispose to thromboembolism, even pulmonary embolism. Clinicians should have appropriate preparedness while doing echocardiography to avoid diagnostic dilemma, while performing cardiovascular interventions to overcome difficulties, and while dealing with patients to make the appropriate diagnosis.

Conflict of Interest - None.

Acknowledgement:

Ms. Nusrat Zahan, Echo Lab Assistant, Popular Diagnostic Centre Ltd. (Shyamoli Branch), Dhaka.

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