

Incidence rate of patent foramen ovale in Chiari type 1 malformation cases and its relationship with tonsillar descent degree and cervical syringomyelia: A contrast transthoracic echocardiography study in adults and review of literature

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Aim: Hans Chiari, who first described the Chiari type 1 malformation (CMI), also reported on the Chiari network (CN)—the embryonic remnant in the right atrium of the heart. CN does not affect the clinical outcome unless accompanied by a patent foramen ovale (PFO), which results in severe cerebrovascular events leading to paradoxical embolism formation. Although 25% of the general population has PFO, in our literature review, we found no study on the incidence of PFO in CMI cases. Therefore, we aimed to investigate the incidence rate of PFO in adult CMI cases using contrast transthoracic echocardiography (cTTE) and assess its relationship with the tonsillar descent (TD) degree and cervical syringomyelia (CMI/Syrinx).

Material and Methods: The cTTEs of randomly selected 37 adult patients with CMI with TD \geq 3 mm were performed. Pearson's chi-square and Fisher–Freeman–Halton exact tests were used to compare qualitative data. Statistical significance was set at $p < 0.05$.

Results: In CMI cases, the incidence rates of PFO, CN, and atrial septal aneurysm with atrial septal defects were 48.6%, 5.4%, and 2.7%, respectively. Moreover, PFO was twice as common in men than in women. There was no statistically significant relationship between the PFO grades and the degree of TD and CMI/Syrinx

Discussion: Since male patients are twice as prone to PFO than female patients, and that coexisting PFO and inter atrial septum pathologies are observed mainly in male patients with CMI, the possibility of embolism might be higher in men than in women.

Conclusion: The results suggest that PFO in CMI cases can impact prognosis by two- fold relative to the general population, moreover its impact on prognosis can be by two- fold in male cases relative to the female cases. However, PFO may not be one of the main etiopathogenetic factors for CMI with TD \geq 3mm and cervical syringomyelia.

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