

Title:

A Rare Case of Noonan Syndrome Presenting with Cor Triatriatum Dextrum

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Background:

Cor triatriatum dextrum is a rare congenital abnormality that creates a physical barrier for catheter insertion into the coronary sinus and right ventricle. We report a rare case of cor triatriatum dextrum presenting in a patient with Noonan Syndrome undergoing pulmonary vein isolation (PVI).

Case Report:

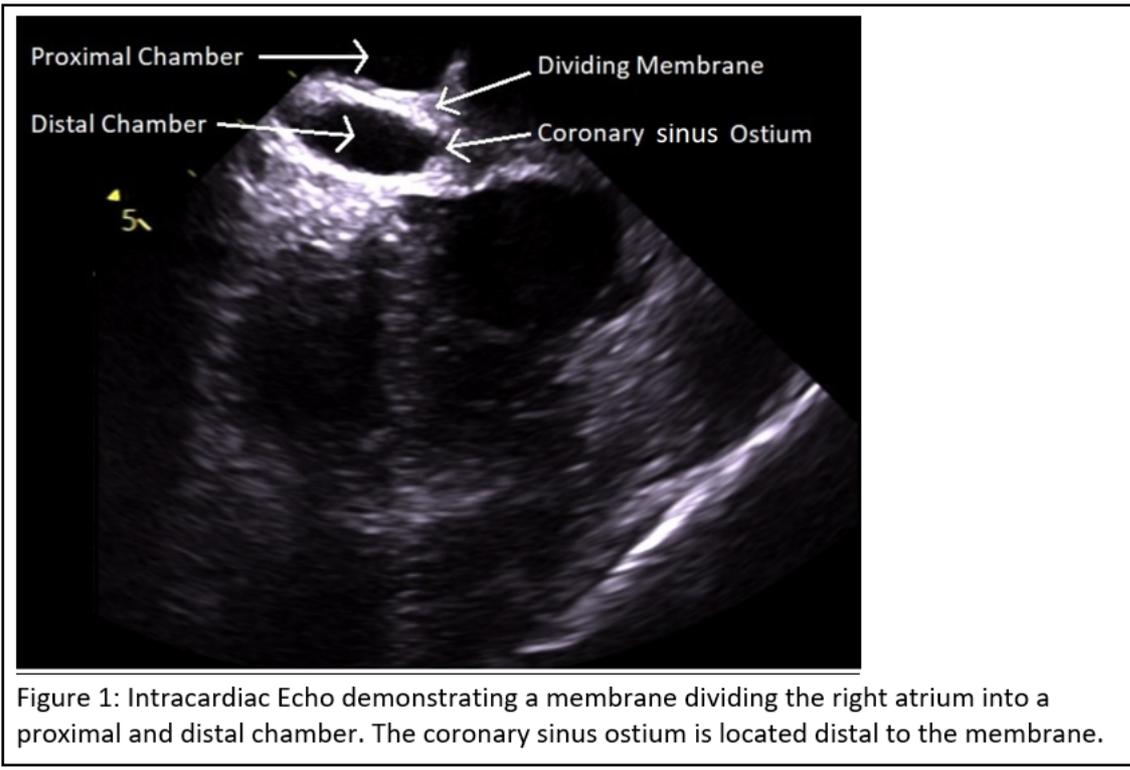
A 24-year-old woman with history of Noonan Syndrome and symptomatic paroxysmal atrial fibrillation was scheduled for PVI. Insertion of the deflectable decapolar CS catheter into the coronary sinus under fluoroscopic guidance was attempted but the coronary sinus ostium could not be located. The ICE catheter was then advanced into the right atrium where it revealed evidence of cor triatriatum dextrum with a thick membrane separating both vena cava from the rest of the right atrium with inability to advance catheters into the coronary sinus or right ventricle. The EPS was aborted due to this anatomical anomaly.

Discussion:

Cor triatriatum is a rare congenital anomaly in which the heart has three atria. Cor triatriatum is divided into two subsets; Cor triatriatum sinistrum, in which the left atrium is divided into two separate structures and Cor triatriatum dextrum in which the right atrium is divided into two separate structures¹. Cor triatriatum dextrum is extremely rare with an incidence of approximately 0.1% of all congenital heart malformations. The congenital heart diseases most commonly associated with Noonan Syndrome are hypertrophic cardiomyopathy and pulmonary stenosis of the valve, artery and vein².

Conclusion

The inability to locate the coronary sinus ostium with catheters should lead to suspicion for cor triatriatum dextrum. The use of ICE can confirm this diagnosis intraoperatively³.



References

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2. Kruszka, P., Porras, A., & Addissie, Y. e. (2017). Noonan Syndrome in Diverse Populations. *American Journal of Medical Genetics*, 2323-2334.
3. Bhatia, N., Humphries, J., Chandrasekaran, K., & Srivathsan, K. (2010). Atrial fibrillation ablation in cor triatriatum: value of intracardiac echocardiography. *Journal of Interventional Cardiac Electrophysiology*, 153-155.