Do we really need a new classification for cor triatriatum sinister?

Alhanouf Alaloola[1], Rawan Alghamdi[1], and Amr Arafat[2]

[1]Affiliation not available
[2]Tanta University Faculty of Medicine

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Abstract

Current classifications of Cor Triatriatum Sinister (CTS) do not address the associated heart defects or single ventricle pathology. Therefore, these classifications are not prognostic classifications and only describe the anatomy and the pulmonary venous drainage. The proposed classification considered the associated congenital cardiac lesions and the single ventricle pathology, therefore, it could have prognostic value. Future multicenter studies are required to measure the performance of this classification and its prognostic value in patients with CTS.

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Alhnouf A. Alaloola, MD (1), Rawan Alghamdi, MD (2), Amr A. Arafat, Ph.D (2, 3)

1. King Fahd Medical City- Riyadh- Saudi Arabia
2. Prince Sultan Cardiac Center- Riyadh- Saudi Arabia
3. Cardiothoracic Surgery, Tanta University, Tanta, Egypt

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Corresponding author:

Amr A. Arafat, MD, Ph.D.

Correspondence Amr A. Arafat, MD, Department of Adult Cardiac Surgery, Prince Sultan Cardiac Center, Building 6, Makkah Al Mukarramah Branch Road, As Sulimaniyah, Riyadh 12233, Saudi Arabia.

Email: amr.arafat@med.tanta.edu.eg

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We have read with great interest the manuscript written by Mashadi and colleagues, "Cor Triatriatum Sinister: Long-term Surgical Outcomes in Children and a Proposal for a New Classification. (1)" Cor triatriatum sinistrum (CTS) is a rare cardiac congenital malformation and represents around 0.1-0.4% of all congenital heart defects, with a very limited number of cases reported in the literature (2, 3). CTS describes an anomaly where the left atrium is subdivided by a thin membrane resulting in two chambers. Many classifications have been used to describe CTS; the most commonly used are the Loeffler's, Lam's, and Lucas's, which are mainly anatomical.

Loffler's classification was developed in 1949, and it depends on the size and number of fenestrations communicating the two atrial chambers. In Loffler's classification, group 1 has no connection between the two chambers, and group 3 has free communication between the two chambers (4). Later in 1962, Lam's classification came to elaborate more on the morphological features of CTS, including pulmonary venous drainage and the site of the atrial septal defect (5). Lucas' classification added further anatomical details and described the association between CTS and anomalous drainage of the pulmonary veins, which the Society of Thoracic Surgeons project adapted for Congenital Heart Surgery Nomenclature and Database (6). Other uncommon classifications described the anatomical features of the accessory left atrial chamber and membrane (7) or the entry of the pulmonary veins into the accessory chamber (8).

The current classifications do not describe the complex nature and possible presentation of CTS that may affect long-term outcomes. Lam's classification takes into consideration the location of drainage of the pulmonary veins and if there is an atrial septal defect (ASD) or not. However, it does not consider the number and size of communication between the two chambers (9, 10). CTS can be associated with other cardiac anomalies in up to 80% of the cases; the most commonly associated cardiac anomalies are atrial septal defect (ASD) in around 60% of patients (11, 12) and partial or total anomalous pulmonary venous return, which has been reported in nearly one-third of the patients with CTS (13). Several studies showed the association of CTS with other congenital anomalies, such as bicuspid aortic valve, atrioventricular canal, tetralogy of Fallot and ventricular septal defect (1, 14). The early and long-term outcomes may vary according to the associated congenital cardiac defects, and in the previously mentioned classifications, a full range of associated congenital heart defects was not taken into consideration.

Additionally, Nagao and colleagues found better survival in CTS patients who had biventricular repair compared to the univentricular pathway (15). Obviously, the univentricular heart was not considered in the previous CTS classifications despite the reported worse outcomes in this category. Furthermore, the exact surgical approach is chosen primarily based on the presence of associated lesions and the size of the atria. The proposed classification mainly depends on the possible presentations and potentially addresses the long-term outcome (1). Mashadi and colleagues divided the patients into three groups: isolated without other congenital heart defects, in association with single ventricle physiology, and CTS concomitant with other congenital heart defects. This proposed classification addresses the anatomical factors previously presented in older classifications, in addition to the associated heart defects and single ventricle pathology. The outcomes could differ in each group; therefore, this proposed classification could have a prognostic value and can anticipate the possible outcomes for each group.

This classification seems beneficial in underlining the anatomical and pathophysiological presentation of CTS; however, its value is still limited, pending a multicenter study or a pooled analysis with a larger sample size to correlate the outcomes of CTS with each classification category.

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