



LETTER TO THE EDITOR

Newly revised category for complex congenital cardiac segmental connections

In the last article [1], we grouped complex congenital cardiac defects into four subgroups with the rules of cardiac segmental coding [2]. Recently, we found some incongruities presented in these categories, so a newly revised edition (Table 1) to correct the concept is mandatory for the many readers.

There are no changes in the group of atrioventricular (A-V) and ventricular-arterial (V-A) concordance. In the group of A-V, discordance with V-A concordance, we found the type of {S.L.D} anatomically corrected malposition (ACM) should be renamed as {S.L.D} isolated atrial inversion with malposition (IAI-M). In the group of A-V concordance with V-A discordance, another two modes of {S.D.S} and {S.D.I}-posterior transposition of the great arteries (TGA) can be added. In the final group of double discordance, we consider an extra mode of {S.L.S} congenital-corrected transposition of the great arteries (ccTGA) that can be included.

The hemodynamics of {S.L.D} in group of A-V discordance with V-A concordance possesses the characteristics

of systemic venous flow returns to the right-sided left ventricle with pumping into the aorta, and the pulmonary venous flow returns to the left-sided right ventricle with pumping into the pulmonary artery. Because the hemodynamics are not the ACM's double concordance nor the TGA's V-A concordance, it should not be categorized into the ACM and should be recoded as isolated atrial concordance (IAC) or IAI-M. We favor the term of {S.L.D}-IAI-M (Fig. 1A).

For the {S.D.S}-posterior TGA (Fig. 1B), the conal septum is reversed and is lacking the spiral rotation between the conal and truncal septum. In this variant, the aortic valve is located right-posterior-inferior to the pulmonary valve and the aortic-mitral fibrous continuity may persist through an outlet type VSD. The greater part of the aortic orifice is above the right ventricle and the pulmonary artery arises to the left of the conal septum and is completely above the left ventricle [3,4]. In {S.D.I}-posterior TGA (Fig. 1C), the aortic conus is located on the right-posterior aspect; however, the spiral rotation of the truncal septum results in the pulmonary

Table 1 The revised edition of cardiac segmental connections.

A–V status	V–A status	
	Ventricular–arterial concordance	Ventricular–arterial discordance
Atrioventricular concordance	{S.D.S}/ {I.L.I}	{S.D.D}-cTGA/ {I.L.L}
	{S.D.L} – ACM/ {I.L.D}	{S.D.L}-cTGA/ {I.L.D}
	{S.D.I} – IIAI/ {I.L.S}	{S.D.S}-posterior TGA
Atrioventricular discordance	{S.L.S}–IVI/ {I.D.I}	{S.D.I}-posterior TGA
	{S.L.I} – IAI/ {I.D.S}	{S.L.L}-ccTGA/ {I.D.D}
	{S.L.D}-ACM/ {I.D.L}	{S.L.D}-ccTGA/ {I.D.L}
		{S.L.S}-ccTGA/ {I.D.I}
		{S.L.D}-IAI-M/ {I.D.L}

ACM = anatomically corrected malposition; ccTGA = congenital corrected transposition of the great arteries; cTGA = complete TGA; IAI = isolated atrial inversion; IAI-M = isolated atrial inversion with malposition; IIAI = isolated infundibulo-arterial inversion; IVI = isolated ventricular inversion; TGA = transposition of the great arteries.

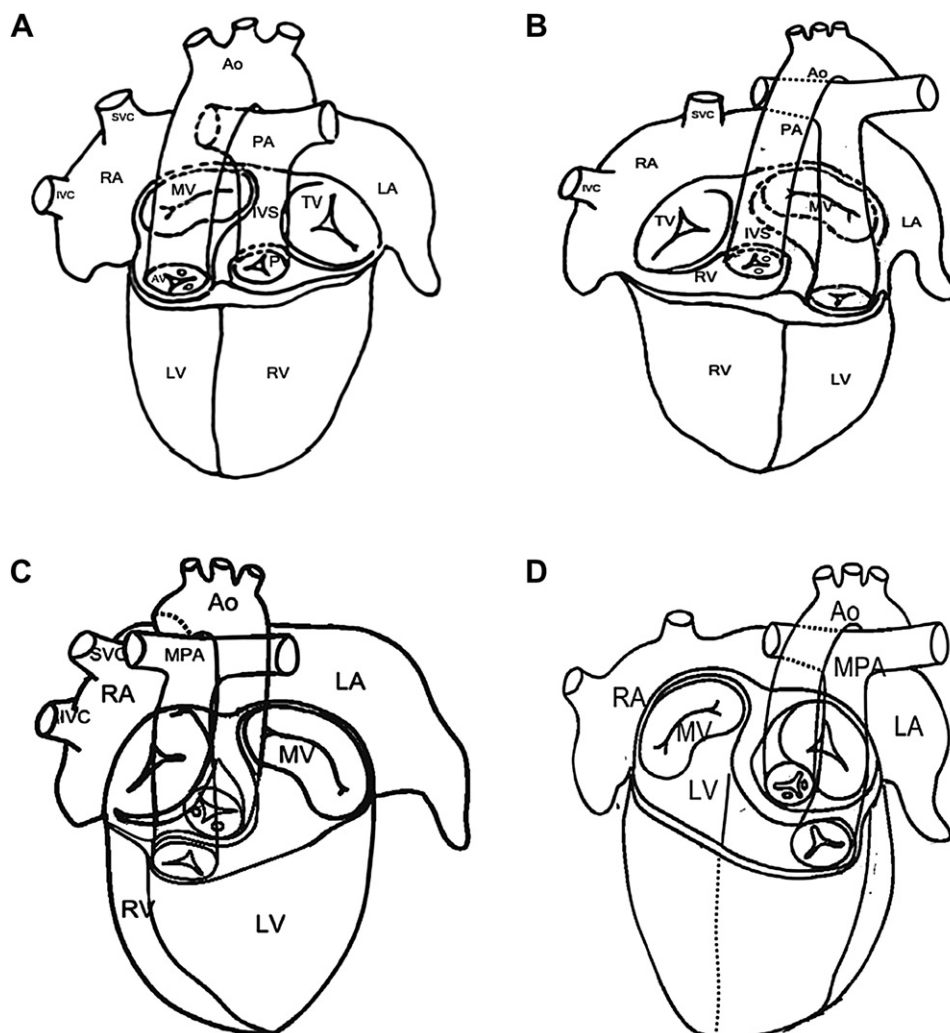


Figure 1. (A) Instead of the name of ACM malformation, the new code of {S.L.D.}-IAI-M is used because this variant is hemodynamically A-V discordant with V-A concordant rather than the ACM's A-V and V-A concordant. (B) The reversed conal septum and the lack of the spiral rotation between the conal and truncal septum make the variant of {S.D.} to be the anomaly of posterior TGA, and is newly listed in the group of A-V concordant and V-A discordant. (C) In {S.D.}-posterior TGA the aortic conus is located on the right-posterior aspect but the rightward rotation of the truncal and conal septum result in the pulmonary trunk is more right-anteriorly. The ventricular-arterial connections are mitral-pulmonary fibrous discontinuity and tricuspid-aortic fibrous continuity as the cTGA. (D) The different direction of conal septum (right-posterior to left-anterior rather than left-posterior to right-anterior) makes the {S.L.S.}-IVI to be a newly variant of {S.L.S.}-ccTGA. Though the aorta is located posteriorly to the pulmonary artery like as posterior TGA, but the hemodynamic is congenitally corrected, so we group as the {S.L.S.}-ccTGA.

trunk being more anterior and rightward, so the pulmonary valve is right-anterior to the aortic valve with the further extension of the pulmonary trunk. The V-A connections are mitral-pulmonary fibrous discontinuous and tricuspid-aortic fibrous continuous. We also found two different hemodynamics of {S.L.S.} isolated ventricular inversion and {S.L.S.}-ccTGA (Fig. 1D) can be divided according to the directions of conal and truncal septum. In {S.L.S.}-ccTGA the truncal septum extends more left and anteriorly, which causes the left ventricle to connect with the pulmonary artery with conus, and the right ventricle to connect with the aorta in fibrous continuity. The aorta is located posteriorly to the pulmonary artery as posterior TGA however, the hemodynamics are congenitally corrected, so we listed the {S.L.S.} as

ccTGA. The hemodynamics in this group are double discordances, so cyanosis is not the main symptom unless other cardiac anomalies exist [5].

We concluded that the newly revised edition ideally corrects all the errors in the previous one and makes the categories clearer and the clinical usage correct.

References

- [1] Chen H-M, Chang P-C, Lee M-S, Wu J-R, Chiu C-C. Easy category for complex congenital cardiac segmental connections. *Kaohsiung J Med Sci* 2007;23:30–3.
- [2] Van Praagh R. The segmental approach to diagnosis in congenital heart disease. *Birth Defects* 1972;3:4–22.

- [3] Miyake T, Yokoyama T, Shirotani H. Transposition of the great arteries with posterior aorta: detection by two-dimensional echocardiography. *Pediatr Cardiol* 1990;11:102–4.
- [4] Ishibashi N, Aoki M, Watanabe M, Nakajima H, Aotsuka H, Fujiwara T. Intraventricular rerouting for transposition of the great arteries with posterior aorta: ventricular septal defect creation and total resection of the infundibular septum. *J Thorac Cardiovasc Surg* 2005;130:593–4.
- [5] Wilkinson JL, Cochrane AD, Karl TR. Congenital Heart Surgery Nomenclature and Database Project: corrected (discordant) transposition of the great arteries (and related malformations). *Ann Thorac Surg* 2000;69:S236–48.

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