Horizontal Ventricular Septum with Dextroversion: Hearts With and Without Aortic Atresia

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SUMMARY. Two hearts with horizontal ventricular septum, dextroversion (situs solitus), ventricular septal defects, and malaligned great vessels are reported. One of the hearts had aortic atresia and the infant died; the other patient had a Fontan-type physiologic correction.

Reviewing the literature, the following conclusions are drawn: (a) Hearts with horizontal ventricular septum and those with criss-cross atrioventricular connections may be the result of different degrees of rotation of the ventricular muscle mass. This rotation is not likely to be postseptational but preseptational. (b) Only those hearts with a complete 180° rotation should be called criss-cross hearts. (c) Partial rotation results in a horizontal septum such that the right ventricle is invariably superior, regardless of atrioventricular concordance or discordance, situs solitus or inversus, or dextroversion. (d) Physiologic surgical correction is often possible but has to be tailored to the details of each heart.

KEY WORDS: Criss-cross heart — Horizontal septum — Upstairs-downstairs heart — Straddling atrioventricular valves — Dextroversion — Aortic atresia
Fig. 1. Patient 1, left ventricular (LV) injection, anteroposterior view. Note: apex points to the right; the ventricular septum is horizontal; the right ventricle (RV) is superior and is hypoplastic; and the large pulmonary artery (PA) relates primarily to the LV.

cule has a hypoplastic sinus portion, associated with tricuspid stenosis or atresia. Pulmonary stenosis or atresia has been observed.

In the group of hearts with horizontal septum, five cases were seen in situs inversus [5, 11, 21, 41]. Among those with situs solitus, levocardia was almost always present. Three hearts with dextroversion [15, 37, 41] and two with mesoversion [3, 13] were observed. We are reporting two additional hearts with dextroversion, and present a unifying concept for the different types of criss-cross hearts.

Definitions

Following Kirklin et al. [18], the term atrioventricular (AV) concordance implies that the right atrium is connected to the anatomic right ventricle, the left atrium to the left ventricle; this term does not indicate whether the right ventricle is anterior, posterior, or superior. Similarly, AV discordance implies that the right atrium is connected to the anatomic left ventricle (via a mitral valve) regardless of anterior or posterior position of the left ventricle. Ventriculoarterial (VA) concordance/discordance implies that the pulmonary valve, whether patent or atretic, relates to the right ventricle (concordance) or left ventricle (discordance). Conversely, the aorta relates to the left ventricle in VA concordance, to the right ventricle in discordance. Transposition of the great vessels (TGV) is synonymous with VA discordance.

In this article, the term dextrocardia is used in a restrictive sense, referring to mirror image arrangement of the cardiac chambers (situs inversus). The term dextroversion is used in situs solitus and implies counterclockwise rotation (viewed from apex) of the heart around the right-sided SVC-IVC axis from the left into the right chest, reversing the normal anteroposterior relationships of the cardiac chambers.

Materials

Among five hearts with horizontal ventricular septum seen at the University of Chicago, two had dextroversion; these two rare patients will be described.

Case 1

This Black infant girl was born after a full term uncomplicated pregnancy. Apgar scores were 8/9. Three cord vessels were present. Other than a skin ear tag, no anomalies were observed in addition to the cardiac defects. The infant had mild tachypnea (50/min) but was acyanotic on 40% oxygen by hood. There was