Coronary arterial pattern in superoinferior ventricular heart

Implications on significance of morphogenesis of this anomaly

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SUMMARY The coronary arterial pattern was studied in two patients with superoinferior ventricular heart and haemodynamically complete transposition. In one of them the aorta was dextro-positioned and in the other laevo-positioned. In both the right coronary artery arose from the posterior aortic sinus and the left coronary artery from the left aortic sinus. This pattern is similar to the common coronary arterial pattern in complete transposition of the great arteries. The similarity supports the theory that in the superoinferior ventricular heart the connections between the ventricles and the great arteries (concordant or discordant) are those of the basic condition – complete transposition or corrected transposition. The angiocardigraphic findings indicate that relations are not always as expected for a given connection.

Superoinferior ventricular heart is a rare anomaly in which the interventricular septum is horizontal, the morphologically left ventricle is inferior, and the morphologically right ventricle is superior. It may be associated with criss-cross heart – a rare anomaly in which the systemic and pulmonary blood streams cross at the atrioventricular level, without mixing. The systemic venous atrium is right sided (in situs solitus) and the venous ventricle is left sided. The arterial blood stream similarly courses from a left sided atrium, to a right sided ventricle.¹⁻⁹

It is the purpose of this paper to describe the coronary arterial pattern in this anomaly, and to discuss the morphogenetic implications of it.

Case reports

CASE 1

A 3-year-old girl was admitted because of cyanosis which was first found at the age of 3 months. Physical examination showed severe cyanosis with clubbing and no signs of congestive heart failure. The second heart sound was normal and a systolic murmur grade 3/6 was heard over the left sternal border. Electrocardiogram showed signs of right ventricular hypertrophy. Thoracic x-ray showed cardiac enlargement, with prominent pulmonary vasculature. Cardiac catheterisation and angiocardigraphy indicated a

normal left atrium draining the pulmonary veins. Contrast material from the left atrium filled an inferior ventricle which was similar to an anatomical left ventricle. The interventricular septum was horizontal. A superior anatomical right ventricle was filled via a ventricular septal defect. It gave rise to a wide aorta which was anterior and right sided. The poor filling of the pulmonary arteries from the left ventricle suggested pulmonary valvular or subvalvular stenosis. Injection of contrast material into the right atrium showed this chamber to be normal. The superior left-sided right ventricle was filled from the right atrium and gave rise to the pulmonary artery. Subpulmonary stenosis was demonstrated (Fig. 1).

Injection of contrast material into the ascending aorta showed the right coronary artery arising from the posterior aortic sinus. The left coronary artery arose from the left aortic sinus, and branched into the left anterior descending and left circumflex arteries (Fig. 2).

The final diagnosis was criss-cross superoinferior ventricular heart, complete transposition of the great arteries, ventricular septal defect, and pulmonary stenosis.

CASE 2

A 1-day-old boy was examined because of cyanosis and tachypnoea. The heart was hyperactive with a single second sound and a holosystolic murmur grade

Fig. 1 Angiocardiography in the lateral projection with injection of contrast material into the inferior left ventricle. The superior right ventricle (RV) is filled via a ventricular septal defect and the interventricular septum occupies a horizontal position. The aorta (A) arises anteriorly and high from the right ventricle. The left ventricle (LV) is inferior. The pulmonary artery (P) arises posterior to the aorta.

3/6 over the left sternal border. Electrocardiogram showed an S1, S2, S3 pattern and absence of Q waves in the praecordial leads. Thoracic x-ray film showed normal heart size, absent pulmonary segment, and prominent upper left cardiac border. Angiocardiography showed a normal left atrium from which a right sided inferiorly positioned morphologically left ventricle was filled. A normal right atrium was filled via an atrial septal defect.

An atric posterior outflow tract arose from the left ventricle. The morphologically right ventricle was left

sided and superior. It was filled via a ventricular septal defect. The interventricular septum occupied a horizontal position. The aorta arose from the left sided infundibulum of the right ventricle. Mitral regurgitation was also demonstrated. Aortography showed the right coronary artery arising from the posterior aortic sinus. The left coronary artery arose from the left anterior aortic sinus. Normal sized pulmonary arteries were filled by a persistent ductus arteriosus.

The final diagnosis was criss-cross superoinferior ventricular heart (in situs solitus), complete transposition with laevo-positioned aorta, pulmonary atresia, ventricular septal defect, atrial septal defect, mitral regurgitation, and persistent ductus arteriosus.

Discussion

Criss-cross and superoinferior ventricular heart represents an exception to Van Praagh's loop rule.¹⁰⁻¹⁵ The spatial relations between the cardiac chambers are disharmonious despite the presence of concordant connections.⁴ It was suggested,¹⁶ however, that this angiographic appearance of the criss-cross atrio-ventricular connection may create an illusion to the eye of the observer who believes that relations are always harmonious with connections. Despite the angiographic appearance the right atrium communicates with the morphologically right ventricle and the left atrium communicates with the morphologically right ventricle and the left atrium communicates with the morphologically left ventricle (concordant connections).

The coronary arterial pattern may help to resolve the question whether the criss-cross and superoinferior connection, represents an anatomical reality

Fig. 2 (a) Aortography in the lateral projection, showing the right coronary artery (R) arising from the posterior aortic sinus. (b) This shows the origin of the right coronary artery more clearly but does not show its bifurcation.

(as a new entity) or an angiocardigraphic illusion. The coronary arterial patterns in complex congenital heart diseases are well known. In a previous study¹⁷ we suggested that in transposition complexes the coronary arterial pattern is related to the type of ventricular loop (inverted or non-inverted ventricles). Thus, the coronary arterial pattern in complete transposition of the great arteries with laevo-position of the aorta will be the same as that found in 'classical' complete transposition of the great arteries.¹ This had also been shown by others.¹⁶

In the most common coronary arterial pattern in complete transposition of the great arteries the right coronary artery arises from the posterior aortic sinus.¹⁸ This pattern was found in our two patients. This supports the theory that the 'crossing', at least in these cases, is a variant of the usual atrioventricular connection. This variant is formed, as suggested by Anderson *et al.*,¹ by rotation along the apex-base axis.

Reports of the coronary arterial pattern in criss-cross and superoinferior ventricular heart are rare. Sato *et al.*⁸ reported a case of criss-cross heart with concordant atrioventricular and ventriculo-arterial connections. The left coronary artery arose from the left posteriorly rotated aortic sinus. The origin and course of the right coronary artery were not described. The left anterior descending artery was oriented horizontally, like the interventricular septum.

Symons *et al.*¹⁹ reported a case of criss-cross heart with corrected transposition and right sided anterior aorta. The origin of the coronary arteries was not specified. There were, however, downgoing branches from both the right and left coronary arteries, which delineated the oblique position of the interventricular septum.

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