A Criss-Cross Heart with Concordant Atrioventriculo-Arterial Connections

Report of a Case

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SUMMARY A distinctive angiographic appearance is described in a case of "criss-cross" heart with concordant atrioventriculo-arterial connections.

The atrial situs was normal, and the morphological right ventricle was superior to the left ventricle, and the ventricular septum was horizontal in position. Both ventricles were connected by a small ventricular septal defect. The heart was situated anteriorly and to the right of the pulmonary artery.

THE CRISS-CROSS HEART, a term first used by Anderson and Ando, is a cardiac anomaly producing paradoxical atrioventricular concordance or discordance.

To date, 16 cases of criss-cross heart have been reported in the literature. In all cases except two, discordance was present either in atrioventricular or ventriculo-arterial connection. In the two cases double outlet right ventricle (DORV) was diagnosed.

We have recently performed angiographic studies in a case of criss-cross heart with situs solitus and with concordant atrioventriculo-arterial connections. In this presentation we describe the anatomic features of this heart and discuss its possible embryogenesis and the direction of the ventricular rotation.

Case Report

An 8-month-old girl was admitted to Osaka Prefectural Hospital in December 1974. Heart disease was first noted when she was affected by respiratory infection at the age of three months. There was no family history of congenital heart disease.

On examination, the girl weighed 5.8 kg and 65 cm long. There was no cyanosis. A bulge of the anterior wall of the chest was present. A grade 5/6 systolic murmur was heard in the third left intercostal space, and systolic thrill was palpated.

A roentgenogram of the chest showed an enlarged heart (cardiothoracic ratio 0.58). The pulmonary vasculature was increased on the right and decreased on the left. Electrocardiograms revealed conduction alternating between normal atrioventricular conduction and characteristic features of Wolff-Parkinson-White syndrome (a short P-R interval with a wide QRS complex due to a delta wave). In ECG tracing which showed normal conduction, the mean electrical QRS axis was +118° and there was biventricular hypertrophy.

A two-dimensional echocardiogram (kymogram) was obtained as the ultrasonic beam swept in a sector from the aortic root through the left venous outflow tract to the apex of the heart. Both anterior and posterior aortic walls were continuous with the interventricular septum and the anterior mitral valve leaflet, respectively. When performing an M-mode sector scan between the anterior tricuspid valve leaflet and the anterior mitral valve leaflet, the mitral valve was recorded in a position almost inferior to and posterolateral to the tricuspid valve. With the transducer placed along the left sternal border in the third or fourth intercostal space, the mitral valve was recorded behind the tricuspid valve, with echo-free space between these echoes. This finding suggested the presence of a defect of the interventricular septum. The aortic valve was in a position anterosuperior to the pulmonary valve. This anatomic relationship between the aortic valve and pulmonary valve is opposite to that expected in a normal subject.

Using cardiac catheterization techniques, we calculated total pulmonary flow at 5.3 L/min/m²; the systemic flow was 4.0 L/min/m². There was a left-to-right shunt of 1.3 L/min/m². Pulmonary vascular resistance was 3.4 units, with a pulmonary to systemic vascular resistance ratio of 0.16:1.

Selective angiograms were performed in the right atrium (fig. 1), right ventricle (figs. 2, 3), left ventricle (fig. 4), and aorta (fig. 5). The catheter was passed via the inferior vena cava into the right atrium, which was situated in a solitus position, and then into the anatomic right ventricle located above the anatomic left ventricle. The right atrial injection (fig. 1) showed the right atrium to be in continuity with the inflow portion of the right ventricle; the systemic venous flow was from the right atrium to the right ventricle through a right atrioventricular valve. The catheter was advanced through the tricuspid valve into the anatomic right ventricle which consisted of a sinus and infundibular portion.
The pulmonary artery arose from the right ventricle. The pulmonary artery trunk was moderately dilated and situated posteriorly and to the left of the aorta. The pulmonary valve was below the aortic valve. The right pulmonary artery branches were moderately dilated but not tortuous at the periphery. The left branches were hypoplastic. During the levophase (fig. 3), continuity between left atrium and anatomic left ventricle was established. The catheter was then introduced, via the femoral artery, into the anatomic left ventricle lying inferiorly (fig. 4). The ventricles were separated by a septum positioned in a nearly horizontal plane. Left-to-right shunt through the ventricular septal defect was in a vertical direction between the inferiorly positioned left ventricle and the superiorly placed right ventricle. The left atrioventricular valve was demonstrated on the left ventricular injection. The aorta arose from the left ventricle. The aortic valve was adjacent to the mitral valve. The coronary arterial pattern (fig. 5) was characteristic of ventricular rotation with the left anterior descending branch from the posteriorly shifted left coronary artery. In addition, the course of the left anterior descending branch was oriented posteriorly and then horizontally like the interventricular septum. Patent ductus arteriosus (PDA) with a small shunt was present.

From these findings, the case described was interpreted as representing a criss-cross heart with solitus atria and with concordant atrioventricular-arterial connections, in which the right ventricle was situated superiorly to the left ventricle, and the aortic valve was located anteriorly, superiorly, and to the right of the pulmonary valve. It was associated

**FIGURE 1.** Antero-posterior view of the right atrial injection showing the right atrium in situs solitus. Contrast opacification shows the right atrium to be in continuity with the right ventricular inflow tract.

**FIGURE 2.** Right ventriculogram in frontal (left) and lateral views (right). The pulmonary artery arises from the posteriorly rotated infundibular portion of the right ventricle. The left pulmonary artery branches are hypoplastic.
with ventricular septal defect, hypoplastic left pulmonary artery, and PDA.

Surgical treatment was performed when the patient was three years old. The angiographic findings were confirmed except for aortico-mitral continuity. The ventricular septal defect was successfully closed under right ventriculotomy.

Discussion

This heart exhibits abnormal atrioventricular (A-V) relations in that the atria are in solitus position and the right ventricle is superior to the left ventricle. However, this right ventricle receives right atrial blood and the left ventricle receives left atrial blood so that concordant connections are present. Both the heart with abnormal A-V relations of a concordant A-V connections and the heart with normal A-V relations of a discordant A-V connections represent crossing of systemic and pulmonary venous blood streams, without mixing, at atrioventricular level. Such hearts were designated as criss-cross hearts by Anderson who reported a case with complete transposition of great vessels (TGV) and one with corrected transposition of great vessels (CTGV).

In Japan, Ando et al. reported four cases designated as criss-cross heart at about the time of publication of Anderson's report. Four additional cases were reported later: five cases of TGV, two cases of CTGV and one case of DORV. Two cases of Lev and Rowlatt (TGV and DORV) and a
case of Van Praagh \cite{vanpraagh} (TGV) were designated as criss-cross heart by Anderson. Case 6 of Wagner et al.\cite{wagner} with anatomic corrected malposition and case 2 of Kinsley,\cite{kinsley} (CTGV) are similar to these but did not show a complete criss-cross relationship. Recently, two cases of TGV by Guthaner et al.\cite{guthaner} and a case of TGV by T. Sato\cite{sato} were reported.

Altogether, 16 cases of criss-cross heart have been reported in the literature, to the best of our knowledge: ten cases of TGV, four cases of CTGV and two cases of DORV. In all of these cases except two cases of DORV, either discordant A-V connections or discordant arterio-ventricular connections were present. The case presently described is the only criss-cross heart reported with concordant atrioventricular and arterio-ventricular connections.

In criss-cross heart, the apparent position of the cardiac segments does not indicate the real “connection” of the segments, and this contradicts the loop-rule of Van Praagh.\cite{vanpraagh} The terminology by Kirklin et al.\cite{kirklin} represents a refinement of the segmental descriptions which included the terms such as concordant and discordant A-V relation and position of the great arteries. However, such terminology would still be confusing since apparent position of cardiac segments are different in criss-cross hearts. Thus Anderson\cite{anderson} proposed the new nomenclature describing the bulboventricular loop and adopting the recent definition of Van Praagh\cite{vanpraagh} pertinent to transposition. Using this nomenclature in the case presently studied, the heart can be described as solitus-concordant (l-rotated)-normal.

The morphogenesis of criss-cross heart is considered to be caused by abnormal rotation of the ventricular loop. Anderson\cite{anderson} interpreted his case 1, d-loop criss-cross heart, as the result of counterclockwise rotation around the base-apex axis following septation. We interpret our case as clockwise rotation of 90° around the long axis. T. Sato et al.\cite{sato} also interpreted rotation in d-loop criss-cross to be clockwise. In our opinion, the relative positions of both atrioventricular valves (tricuspid valve situated anterior to mitral valve) and the course of the left coronary artery prove this interpretation to be valid. In addition, in our case, the right ventricle

\begin{figure}
\centering
\includegraphics[width=0.8\textwidth]{figure5}
\caption{The coronary arterial pattern in frontal (left) and lateral views (right) is characteristic of ventricular rotation with the left anterior descending branch from the posteriorly shifted left coronary artery, and the course of the left anterior descending branch runs horizontally as in the interventricular septum.}
\end{figure}

\begin{figure}
\centering
\includegraphics[width=0.8\textwidth]{figure6}
\caption{Diagrammatic representation of the relations and the connections between the atria, ventricles and great arteries. The atrial septum is in the almost usual plane with solitus atria. The ventricular septum is in the horizontal plane. The right ventricle is superior to the left ventricle. Blood flow is from right atrium to right ventricle and left atrium to left ventricle with crossing of the two circuits without mixing at the atrioventricular level. The aorta arises from the left ventricle and the pulmonary artery from the infundibular portion of the right ventricle, and the relation between the great arteries is abnormal.}
\end{figure}
was superior to the left ventricle and the ventricular septum lay horizontally. This superior-inferior arrangement of both ventricles in criss-cross heart was seen in two cases of Guthaner and a case of T. Sato. One case of CTGV reported by Kinsley and two by Momma with horizontal ventricular septum were interpreted to be the result of ventricular rotation about the anteroposterior axis. These were not criss-cross hearts however.

In summary, our case represents ventricular clockwise (viewed from postero-anterior view) rotation not only around the long axis but also around the anteroposterior axis. In our case, the aortic valve lies anteriorly, superiorly and to the right of the pulmonary valve, and the angiographic and echographic evidence indicates aortico-mitral continuity. Complex ventricular rotation would turn the originally normal great arteries into their demonstrated abnormal position.

The definition of "malposition" proposed by Van Praagh and the presence of anatomically corrected malposition (ACM) with aortico-mitral continuity reported by Anderson, would lead our case to be classified as ACM. However, our case had a right-sided aorta and does not belong to their category of ACM.

In a heart with A-V concordance, as our present case, whether it is the result of abnormal ventricular rotation of normal heart or of ACM, the conduction tissue is arranged normally. The surgeon can depend on the normal guidelines for locating bundle of His in repair of associated ventricular septal defect. The conduction pattern is not the same in A-V discordance.

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