Crisscross Hearts in Adults: Echocardiographic Evaluation and Natural History

Piotr Hoffman, MD,* Piotr Szymański, MD, Barbara Lubiszewska, MD,
Jacek Różański, MD, Magdalena Lipczyńska, MD, Anna Klisiewicz, MD, *Warsaw, Poland*

Crisscross heart is a rare congenital, rotational abnormality of the ventricular mass, resulting in the crossing of the inflows of the 2 ventricles and each atrium emptying into the contralaterally located ventricle. Few cases of adult patients are described in the literature. We present a case series of 7 adult patients aged 18 to 53 years with crisscross hearts, followed up for 5 to 22 years (mean 14.4 years) at the Institute of Cardiology Warsaw. Only in 2 patients was the diagnosis of crisscross heart made at infancy. In the remaining 5 patients, the diagnosis was made at the age of 7 to 26 years. All patients had complex congenital heart defects, including double outlet right ventricle in 4 patients and discordant ventriculoarterial connections in 3 patients, with numerous associated lesions. In all patients, the diagnosis of crisscross heart was made by transthoracic echocardiographic examination. Five patients underwent surgical procedures, including Rastelli operation, arterial switch, Glenn anastomosis, and central shunt, according to the individual morphology and hemodynamics. The clinical outcome depended predominantly on the underlying hemodynamic abnormalities and the results of surgical management rather than on crisscross anatomy. Successful surgery allowed acceptable functional status in adulthood. (J Am Soc Echocardiogr 2009;22:134-140.)

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Crisscross heart is a rare congenital, rotational abnormality of the ventricular mass, resulting in the crossing of the inflows of the 2 ventricles and each atrium emptying into the contralaterally located ventricle.[1] The atrioventricular and ventriculoarterial connections can be concordant as well as discordant, and the relationships of the ventricular chambers cannot be anticipated from these connections.[2] The real incidence of the condition is unknown and can only be estimated as several cases per million live births. There are more than 150 cases described in the literature, a few in adult patients.[3,4]

The diagnosis of crisscross hearts is usually based on the principles of segmental analysis described by Van Praagh[2] and Anderson et al.[5,6] Atrial situs is principally identified from the arrangement of the atria and the great vessels, with the aorta to the left of the spine and inferior vena cava to the right in situs solitus. The opposite arrangement is situs inversus. The shape of atrial appendages is helpful in identifying left and right atria. The ventricular chambers are identified on the basis of several anatomic features. The right ventricle has more coarse trabeculations in the apical area, moderator band, slightly lower insertion of the tricuspid valve on the ventricular septum, and attachment of its chordae to the ventricular septum. The anatomic left ventricle has the bicuspid atrioventricular valve inserted higher on the ventricular septum, smooth walls, fine apical trabeculations, and, usually, 2 papillary muscles.

Crisscross heart results principally from the major ventricular malposition, and because of this malposition atrioventricular relations are angulated relatively to each other. At the embryologic straight tube stage (preloop), the ventricles are oriented in a superoinferior manner—right ventricle above, left ventricle below. During D-looping, a 90-degree clockwise rotation occurs, so that the ventricles wind up side by side, with the right ventricle to the right and the septum more or less vertical in orientation. If, however, looping is in the wrong direction, the ventriculoatrial relations become crisscross. From the straight tube stage, looping would proceed counterclockwise (ie, in the wrong direction) by 90 degrees or somewhat less. Now, while the right ventricle is still connected to the right-sided right atrium, it is located way over to the left. The left ventricle is now right sided and connected to the left-sided left atrium. To sum up, when a normal straight heart tube rotates in the wrong direction, a crisscross heart is the result.

From the embryologic perspective, it is helpful to consider chirality (handedness).[7,8] A hand can be used to determine the handedness of a ventricle in space. By putting hands out in front, palms together, fingers pointing forward, this is more or less how the ventricles are normally related. The right ventricle is in the position of the right hand. The palm of the right hand represents the right ventricular septal surface, whereas the dorsum of the right hand represents the right ventricular free wall. The palm of the left hand represents the left ventricular septal surface, whereas the dorsum of the left hand represents the left ventricular free wall. When the ventricles are inverted, the left hand is the morphologic right ventricle and the right hand is the morphologic left ventricle. Thus, in normal D-loop ventricles, the right ventricle is right handed and the left ventricle is

From the Department of Adult Congenital Heart Disease (P.H., P.S., M.L., A.K.), Department of General Cardiology (B.L.), and Second Department of Cardiac Surgery (J.R.), Institute of Cardiology, Warsaw, Poland.

Reprint requests: Piotr Hoffman, MD, Head, Department of Adult Congenital Heart Diseases, Institute of Cardiology, Alpejska 42, 04–628 Warszawa, Poland (E-mail: *hoffman@ikard.pl*).

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left handed. In inverted L-loop ventricles, the right ventricle is left handed and the left ventricle is right handed.

Imaging crisscross hearts is difficult, and patients usually undergo several different diagnostic procedures, including cardiac catheterization, until the final diagnosis is reached—crisscross heart is sometimes called a "Rosetta stone" of segmental analysis.

Echocardiographic diagnosis of crisscross heart is based on the above-mentioned anatomic principles, described by Van Praagh[2] and Anderson and colleagues.[5,6] Accurate echocardiographic diagnosis requires scanning at different planes and modified, nonstandard views. Segmental approach has to be carefully followed. All cardiac segments, their arrangement, relations, and connections have to be thoroughly identified. In particular, the relative position of both atria and the ventricles (their inflow parts) has to be defined. Imaging of the typical crossing of ventricular inflows requires tilting and rotation of the transducer because, frequently, both inflows cannot be visualized in the same imaging plane. Coexisting anomalies and sequels of surgical procedures have to be identified.

We describe our case series of 7 adult patients aged 18 to 53 years with crisscross hearts. The cases come from the Grown-up Congenital Heart Disease Unit located at the Institute of Cardiology Warsaw. The results of the echocardiographic studies are summarized in Table 1.

Case 1 (Video 1) was an 18-year-old male student followed up at the institute for 15 years. The diagnosis of crisscross heart, pulmonary atresia and hypoplastic pulmonary arteries, atrial septal defect, and ventricular septal defect was made at a pediatric center elsewhere. At the age of 3 and 2/12 years, a polytetrafluoroethylene (Gore-Tex; WL Gore and Associates Inc, Flagstaff, AZ) anastomosis between the ascending aorta and right pulmonary artery was created, and a left pulmonary artery plasty was performed. At the age of 4 and 9/12 years, the shunt, atrial septal defect, and ventricular septal defect were closed, and the Rastelli procedure was performed, with a homograft placed between the pulmonary artery and the right ventricle. At the recent examination (2007), the patient was in a good clinical condition, in New York Heart Association class I/II. His electrocardiogram showed low atrial rhythm of 55 beats/min with a short PQ interval (0.1 seconds), left axis deviation, and indeterminate intraventricular conduction block. Apart from the crisscross-type connections (Figure 1), a recent echocardiographic examination demonstrated a moderately elevated gradient through the homograft and mild-to-moderate pulmonary regurgitation, a small muscular type ventricular septal defect, and a possible small residual ventricular septal defect patch leak (Table 1). The patient was treated conservatively.

Case 2 (Video 2) was a 19-year-old female student followed up at the Institute of Cardiology for 18 years. In infancy she was diagnosed as having complete transposition, mild subvalvular pulmonary stenosis, and 2 ventricular septal defects (perimembranous and large inflow). She underwent the Rashkind procedure at the age of 6/12 years and arterial switch procedure with atrial septal defect closure and ventricular septal defects closure with polytetrafluoroethylene (Gore-Tex) patches at the age of 1 and 1/12 years. At the time of the surgery, the difficulty in visualizing the inferior edge of the inlet defect was reported, and the tricuspid valve was detached and subsequently reattached to the tricuspid annulus, following the defect closure. After the surgery, moderate and later severe pulmonary artery stenosis was observed with the systolic gradient of up to 90 mm Hg. Unsuccessful balloon angioplasty of the pulmonary artery was performed at the age of 18 years, with a significant residual gradient at Doppler measurements. At the recent examination (2007), she was in a good clinical condition, with slightly diminished exercise tolerance, in New York Heart Association class II. Her electrocardiogram showed a normal sinus rhythm of 63 beats/min, left axis deviation, indeterminate intraventricular conduction block, suspected biventricular hypertrophy, and deep Q waves in II, III, aVF. Echocardiographic examination revealed the typical crossing of the inflows of the 2 ventricles, not described during previous examinations (Figure 2). There was a significantly obstructed flow through the pulmonary artery with mild subpulmonary stenosis. A small muscular type ventricular septal defect was detected. A nonsignificant aortic regurgitation was also found (Table 1). She was qualified for surgical intervention.

Case 3 (Video 3) was a 24-year-old female student, followed up at the institute for 5 years. The diagnosis of crisscross heart with double outlet right ventricle, subvalvular and valvular pulmonary stenosis, large ventricular septal defect, atrial septal defect, overriding, and straddling tricuspid valve was made at infancy. At the age of 10 years, a Glenn anastomosis was performed. There were no further interventions. At the recent examination (2007), she was cyanotic, with an arterial blood oxygen saturation of 78% to 88%, in a relatively good clinical condition, with diminished exercise tolerance and New York Heart Association class II. She had a sinus rhythm of 83 beats/min, with left axis deviation, left atrial enlargement, incomplete right bundle branch block, and features of left ventricular hypertrophy in the electrocardiogram. Echocardiogram demonstrated superoinferior ventricles with a large ventricular septal defect and "typical" crisscross connections (Figure 3). An unusual finding was an accessory mitral tissue obstructing the right ventricular outflow tract. The flow through the right-sided Glenn anastomosis was unobstructed. She was treated conservatively.

Case 4 (Video 4) was a 25-year-old male university graduate, followed up at the institute for 18 years. The first hemodynamic study was performed at the age of 8/12 years. The diagnosis included dextrocardia, univentricular heart, ventricular septal defect, hypoplastic tricuspid valve and right ventricle, transposition of the great arteries, pulmonary stenosis, and moderate left atrioventricular valve insufficiency. The patient was initially disqualified from surgical intervention. The diagnosis of crisscross heart was made at the institute when the patients was 7 years old. Biventricular repair was considered possible. At the age of 8 years, the Rastelli procedure was performed. After the surgery, residual ventricular septal defect and mitral regurgitation persisted. Because of deteriorating heart failure and episodes of atrial fibrillation accompanying significant mitral regurgitation at the age of 14 years, a St Jude Medical (St Paul, MN) mechanical valve was implanted into the left atrioventricular orifice. A trace residual shunt through previously patched ventricular septal defect was not closed. At the recent examination (2007), the patient was in a good clinical condition, without overt heart failure, in New York Heart Association class II. Electrocardiogram revealed atrial fibrillation 70 to 80 beats/min, right axis deviation, complete right bundle branch block, features of right ventricular hypertrophy, and negative T waves in leads V3-V6. Apart from the crisscross, a relationship of the inflows of the ventricles (Figure 4), well-functioning mitral valve prosthesis, and a nonsignificant gradient through the pulmonary homograft were demonstrated. The left ventricle was mildly dilated, and a trace residual ventricular septal defect was visible. No further interventions were planned.

Case 5 (Video 5) was an unemployed female patient in whom the final diagnosis of crisscross heart, partial atrioventricular canal, D-transposition of the great arteries, and pulmonary stenosis was made at the age of 26 years. At the time of the examination at the institute (1999), she was slightly cyanotic, with a history of syncope, without overt heart failure, and in New York Heart Association class II.