Criss-cross heart

Author: Doctor Stephen Sanders[1]
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[1]Division of Pediatric Cardiology, Duke university medical center, 3000 Erwin Road, NC 27710 Durham Box 3090, United States. sande021@mc.duke.edu

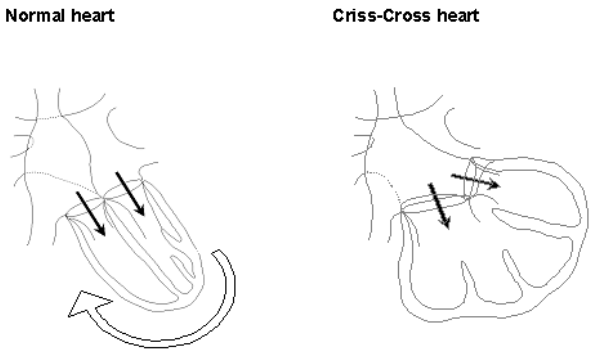
Abstract
Criss-cross heart is a rare congenital cardiac anomaly characterized by crossing of the inflow streams of the two ventricles due to an apparent twisting of the heart about its long axis. Hypoplasia of the tricuspid valve and right ventricle is a common feature, like pulmonary stenosis. The genetic or other causes and the developmental mechanisms remain unknown. The frequency of criss-cross heart is no more than 8 per 1,000,000. Most patients present as neonates with cyanosis and a systolic murmur. The diagnosis is readily made using 2-dimensional echocardiography. The diagnostic feature is crossing of the long axes of the atrioventricular valves as seen in a subxiphoid long-axis or coronal plane sweep. Treatment may include initial palliation with a systemic-to-pulmonary shunt to increase pulmonary blood flow and improve cyanosis. This is followed by staged progression toward completion of a Fontan-type operation in the majority. A few patients are candidates for a 2-ventricle repair. Antenatal diagnosis can be made using fetal echocardiography.

Keywords
Congenital heart defect; criss-cross heart; supero-inferior ventricles; straddling mitral valve; straddling tricuspid valve; juxtaposition of the atrial appendages.

Disease name and synonyms
Criss-cross heart
Twisted atrioventricular (AV) alignments (or connections)

Excluded diseases
Although supero-inferior ventricles often accompany criss-cross heart, the two are not synonyms.

Diagnostic criteria
The diagnosis of criss-cross heart is based on crossing of the long axes of the atrioventricular valves. In the normal heart, the inflow streams through the two atrioventricular valves are approximately parallel, with each aimed at the apex of its respective ventricle. In criss-cross hearts, the ventricles appear to have been twisted about their long axes while the base of the heart remains fixed. This causes the flow streams through the valves to cross each other resulting in the characteristic appearance of the anomaly. This gives the appearance of each atrium emptying into the contralateral ventricle.



INFLOW STREAMS THROUGH THE TWO ATRIOVENTRICULAR VALVES

Differential diagnosis

Other anomalies that might be confused with criss-cross heart include:
a) some forms of straddling atrioventricular valves,
b) double outlet atrium in which one orifice of the atrioventricular valve from the double outlet atrium appears to cross the other valve,
c) severe forms of Ebstein's anomaly of the tricuspid valve in which the tricuspid valve opens into the infundibulum, giving the appearance of crossing valves.

Frequency

Criss-cross heart is an extremely rare anomaly accounting for less than 0.1% of congenital heart defects (1). Congenital heart defects occur at the rate of about 8 per 1,000 live births, so that the frequency of criss-cross heart is no more than 8 per 1,000,000.

History

Criss-cross heart was first described in 1961 by Lev and Rowlatt (2). The term "criss-cross" heart appears to have been introduced by Anderson and colleagues in 1974 (3). Criss-cross heart is an especially important defect because it illustrates a key concept essential to understanding congenital heart defects: the difference between situs concordance and alignment concordance, and both concepts are important for understanding heart defects (4,5).

Clinical description

Criss-cross heart is almost always associated with other severe cardiac anomalies and therefore presents in infancy. The majority of patients with criss-cross heart have hypoplasia of the tricuspid valve and right ventricle, a ventricular septal defect, abnormal ventriculo-arterial alignments (either transposition of the great arteries or double outlet right ventricle) and pulmonary stenosis (6). Consequently, neonates typically present with cyanosis and a systolic murmur.
Rare patients with criss-cross heart have only a ventricular septal defect with normally related great arteries (or nearly so) and present with symptoms of heart failure from the left-to-right shunt (7,8).
Some patients with criss-cross heart have juxtaposed atrial appendages (4,9-12).
In most patients with criss-cross heart the AV segmental situs is consistent with the AV alignments. That is, in patients with situs solitus of the atria and d-looped or right-handed ventricles (concordant AV segmental situs), the right atrium is aligned with (opens into) the right ventricle and the left atrium with the left ventricle (concordant AV alignments). Conversely, in patients with situs solitus of the atria but l-looped or left-handed ventricles (discordant AV segmental situs), the right atrium is aligned with the left ventricle and the left atrium with the right ventricle (discordant AV alignments).
In some, however, there is disharmony between the segmental situs and the AV alignments (4,9-13). For example, rare patients with situs solitus of the atria but l-looped or left-handed ventricles (discordant AV segmental situs) have AV alignment concordance. That is, despite segmental situs discordance (solitus atria but l-looped ventricles), the right atrium is aligned with the right ventricle and the left atrium with the left ventricle. Similarly, rare patients with situs solitus of the atria and d-looped or right-handed ventricles (concordant AV segmental situs) have AV alignment discordance. That is, the right atrium is aligned with the left ventricle and the left atrium with the right ventricle. Criss-cross heart illustrates the important concepts that AV segmental situs is not always predictive of AV alignments, that both are important, and that both must be elucidated and described independently (5).
In about 80% of patients with criss-cross heart there is discordant ventriculo-arterial (VA) segmental situs (inverse loop rule of Van Praagh) (14). That is, if the ventricles are d-looped or right-handed, the great arteries are l-malposed (aorta anterior and leftward of the pulmonary artery) and if the ventricles are l-looped or left-handed, the great arteries are d-malposed (aorta anterior and rightward of the pulmonary artery).
Associated defects, in addition to those noted above, that have been reported with criss-cross heart include: straddling mitral valve (15-19) and tricuspid valve (17,19); subaortic stenosis and aortic arch obstruction (15); mitral stenosis (17). Rarely criss-cross heart has an intact ventricular septum, associated with transposition of the great arteries (3,20-22).

Management

Initial management is usually determined by the severity of pulmonary stenosis. If pulmonary blood flow is inadequate, short-term palliation with prostaglandin E1 is indicated to maintain patency of the ductus arteriosus. Then a systemic-to-pulmonary shunt is often created to

provide adequate pulmonary blood flow until more definitive surgical management can be undertaken.
Surgical management is determined by the potential for complete repair using the ventricles independently. Only the small minority of these patients are suitable for a two-ventricle repair because of hypoplasia of the tricuspid valve and right ventricle (8,23). If such a repair is possible, the ventricular septal defect is closed so that the left ventricle is aligned with one great artery (usually the pulmonary artery) and the right ventricle with the other (usually the aorta). An arterial switch operation is then performed. This approach is only possible in the absence of pulmonary stenosis.
In rare patients with normally related great arteries (or double outlet right ventricle with subaortic ventricular septal defect) and only a ventricular septal defect, closure of the septal defect can be corrective if the right ventricle and tricuspid valve are of adequate size (24).
In the majority of patients with criss-cross heart, a two-ventricle repair is not possible. These patients are staged toward a Fontan-type operation (25,26).

Etiology
The etiology of criss-cross heart has been not understood yet. The anomaly seems to be due to abnormal twisting of the apex of the heart while the base remains relatively fixed (3). This accounts for the crossing of the AV valves, the abnormal position of the ventricles and the VA segmental situs discordance. The cause of and mechanism for the twisting of the ventricles remains unclear. Although it is clear that the degree of twisting and the size of the angle between the inflow streams are related to the size of the tricuspid valve and right ventricular sinus (the greater the twist and the angle, the smaller are the tricuspid valve and right ventricle), it is not clear if the right heart hypoplasia is primary or secondary (15). Even less is understood about the development of hearts with disharmony between segmental situs and alignments.

Diagnostic methods
Echocardiography is the primary diagnostic tool, like for all forms of congenital heart disease in the neonate. The diagnosis is made easily in a subxiphoid long-axis scan of the heart by showing the crossing axes of the two atrioventricular valves in adjacent cuts (15, 17, 27). Similarly, a scan from posterior to anterior in the apical four-chamber view shows the crossing axes of the atrioventricular valves. Doppler color flow mapping has been reported to facilitate detection of crossing of the inflow streams (28).
Angiography can also show the crossing of atrioventricular valves by injecting in the left ventricle in a hepato-clavicular view or by superimposing right and left atrial angiograms that show the inflow streams into the ventricles (7,14,27,29).
More recently, magnetic resonance imaging has been shown capable of detecting the crossing atrioventricular valves and many of the associated defects (28,30-32).

Genetic counseling
Little is known regarding possible genetic causes of criss-cross heart. At present only very general counseling regarding recurrence risks can be provided.

Antenatal diagnosis
Prenatal diagnosis can be carried out with fetal echocardiography. The optimal time for imaging the fetal heart is 18-24 weeks of gestation. The same diagnostic principles are applied to the fetus as to the neonate.

Unresolved questions
The cause of criss-cross heart remains unknown.

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