The Criss-Cross Heart

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Age: 33 years  
Gender: Male  
Occupation: Salesman  
Working diagnosis: Large VSD

HISTORY
During infancy, the patient was diagnosed with a large VSD and underwent PA banding through a left thoracotomy at the age of 5 months.

At the time of PA banding for this patient, two-dimensional echocardiography and MRI were not yet available, and the complex anatomy could not be detected as easily as it can be today.

When he was 4 years old, he underwent a sternotomy to close the VSD. However, after direct inspection of the anatomic setting, the VSD appeared unsuitable for closure. The chest was, thus, closed without any intracardiac repair.

The possibility of a Fontan circulation was discussed at the age of 16 years, but it was felt better to be postponed at the time.

Over the ensuing years the patient remained well without any particular shortness of breath on exertion and without any report of cyanosis. He returned for routine follow-up.

Comments: A large VSD will allow for a pronounced intraventricular left-to-right shunt as the pulmonary vascular resistance falls soon after birth. The consequent volume overload of the LV in infancy may then give rise to heart failure. PA banding is performed to decrease pulmonary blood flow, thereby preventing or alleviating ventricular failure and—more importantly—preventing the development of pulmonary arterial occlusive disease.

A Fontan circulation would be an option in which systemic venous return could be directed to the PA without passing through an RV. This would normally involve proximal PA ligation as well. Due to the large interventricular communication, both ventricles would then have acted as a “single” systemic ventricle.

Lack of notable cyanosis in this situation suggests an optimally balanced VSD shunt, namely adequate PA banding without excessive pulmonary flow and without substantial right-to-left shunting, at least at rest. The degree of secondary erythrocytosis in such a patient, when present—provided there is no iron deficiency—indicates indirectly the magnitude of right-to-left shunting at rest and/or during exercise.

CURRENT SYMPTOMS
The patient remains essentially asymptomatic without any history of exercise-induced dyspnea, cyanosis, or angina. On average he has one or two episodes of palpitations per year, always short lived, with no associated syncope or any other symptoms. He is fit, can climb several flights of stairs, and exercises several times a week.

NYHA class: I

Comments: The history suggests the VSD shunt to be appropriately balanced by the PA banding also during exercise, with no symptoms indicating inadequate pulmonary blood flow, significant hypoxemia, or LV dysfunction precipitated by exertion. The episodes of palpitation might represent transient atrial tachyarrhythmia although they also might be explained by ectopic beats.

CURRENT MEDICATIONS
Digoxin 125 µg orally once daily

Comments: The value of the digoxin therapy is doubtful.

PHYSICAL EXAMINATION
BP 130/80 mm Hg, HR 67 bpm, oxygen saturation 93% at rest on room air

Height 165 cm, weight 70 kg, BSA 1.79 m²

Surgical scars: There was a left thoracotomy and a median sternotomy scar

Neck veins: Jugular venous pressure was not elevated

Lungs/Chest: Chest was clear

Heart: There was a right ventricular lift and a faint precordial thrill. Moreover, there was a normal first and a split second heart sound with a soft pulmonary component to it, and a grade 4/6 systolic ejection murmur at the upper left sternal edge, which radiated well to the back.

Abdomen: Soft and normal to palpation

Extremities: There was no evidence of clubbing or peripheral edema

Comments: The scars originated from the PA banding during infancy and the intended VSD closure in childhood, respectively.

The right ventricular heave suggests marked hypertrophy, presumably due to increased intraventricular pressure. Since the large VSD is not restrictive, the pronounced systolic murmur...
is therefore attributed to the impeded flow through the PA band rather than to the VSD, as is also indicated by its ejection character. This is supported by the radiation to the back, a finding more common in murmurs due to obstruction of intrathoracic vessels than in those of intracardiac origin.

These findings add to the other evidence suggesting no significant interventricular right-to-left shunting or any sign of heart failure.

LABORATORY DATA

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<thead>
<tr>
<th>Test</th>
<th>Result</th>
<th>Normal Range</th>
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<tbody>
<tr>
<td>Hemoglobin</td>
<td>16.1 g/dL</td>
<td>(13.0–17.0)</td>
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<tr>
<td>Hematocrit/PCV</td>
<td>46% (41–51)</td>
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<tr>
<td>MCV</td>
<td>91 fl</td>
<td>(84–98)</td>
</tr>
<tr>
<td>Platelet count</td>
<td>$205 \times 10^9$/L</td>
<td>(136–343)</td>
</tr>
<tr>
<td>Sodium</td>
<td>138 mmol/L</td>
<td>(134–145)</td>
</tr>
<tr>
<td>Potassium</td>
<td>4.0 mmol/L</td>
<td>(3.5–5.2)</td>
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<tr>
<td>Creatinine</td>
<td>72 mg/dL</td>
<td>(60–120)</td>
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<tr>
<td>Blood urea nitrogen</td>
<td>3.8 mmol/L</td>
<td>(2.5–6.5)</td>
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<tr>
<td>Ferritin</td>
<td>237 µg/L</td>
<td>(32–284)</td>
</tr>
<tr>
<td>Transferrin</td>
<td>2.5 g/L</td>
<td>(2.0–3.2)</td>
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</table>

**Comments:** A significantly increased hemoglobin and hematocrit would reflect right-to-left shunting. The opposite is not always true: An apparently normal hemoglobin/hematocrit may be present with cyanosis if the patient has concomitant iron deficiency or other cause of anemia.2

In this case, the hemoglobin and hematocrit values are not elevated and there is no evidence of iron deficiency. The laboratory findings therefore indicate that there has been no significant secondary erythrocytosis. This supports the clinical impression of no significant right to left shunting.

ELECTROCARDIOGRAM

**Findings**

Heart rate: 67 bpm
PR interval: 220 msec
QRS axis deviation: 254°
QRS duration: 129 msec
Sinus rhythm with one ventricular ectopic beat, first-degree heart block, extreme axis deviation, right bundle branch block.

**Comments:** The marked QRS axis deviation is similar to that usually associated with AV septal defects or tricuspid atresia, and is not a typical finding in patients with VSD. The depolarization pattern might therefore suggest an unusual topographical arrangement of the ventricular myocardium.

The diagnosis of RVH is difficult in the presence of right bundle branch block.

CHEST X-RAY

**Findings**
Cardiothoracic ratio: 52%

The cardiac silhouette was mildly enlarged with evidence of right atrial dilatation. The central pulmonary arteries were dilated while the peripheral vascular markings were not definitively increased.

**Comments:** Central PA dilation is consistent with PA banding and enlargement of the vessel distal to the narrowing from poststenotic dilatation as well. RVH resulting from PA banding may impair ventricular filling thereby explaining the enlarged atrium. The lack of overtly increased vascular markings suggests there is no hemodynamically important excess pulmonary blood flow.

**Exercise Testing**

**Exercise Protocol:** Modified Bruce
**Duration (min:sec):** 11:13
**Reason for stopping:** Dyspnea
**ECG changes:** None

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<tr>
<td>Heart rate (bpm):</td>
<td>67</td>
<td>125</td>
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<tr>
<td>O₂ saturation (%):</td>
<td>93</td>
<td>87</td>
</tr>
<tr>
<td>Blood pressure (mm Hg):</td>
<td>130/80</td>
<td>170/90</td>
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<tr>
<td>Double product:</td>
<td>21,250</td>
<td></td>
</tr>
<tr>
<td>Peak VO₂ (mL/kg/min):</td>
<td>22.8</td>
<td></td>
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<tr>
<td>Percent predicted (%)</td>
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<td></td>
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<tr>
<td>VO₂/VO₂</td>
<td>68</td>
<td></td>
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<td>Metabolic equivalents:</td>
<td>4</td>
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</table>

**Findings**

Exercise tolerance was fair, with mild desaturation during exercise.

**Comments:** Despite the unrepaired VSD, the patient has only mild desaturation with exercise. This is generally good and reflects the protection gained from his earlier pulmonary arterial band. Without this, the patient’s baseline and exercise oxygen saturations would be much lower.
Overall Findings
There was normal visceral situs. There was a conspicuous AV connection with abnormal ventricular relationships (see below). Left ventricular size and function were normal. The RV was hypertrophic with good systolic function. The LA was normal in size while the RA was mildly enlarged. There was no significant valve regurgitation. There was considerable PA obstruction with an estimated peak pressure gradient of 93 mm Hg.

Findings
The parasternal long-axis imaging demonstrated a large nonrestrictive VSD 36 mm in diameter with bidirectional flow. Ventriculoarterial concordance was reported.

Comments: The relationship between the atria and ventricles and great arteries must be clarified to be certain of the true anatomic diagnosis. Here, the AV and ventriculoatrial relationships seemed to be concordant (i.e., normally related), as shown by the normally related LA, LV, and aorta.

The VSD is large and nonrestrictive, so there is equalization of pressures between the LV and RV. The measured arterial systolic blood pressure of 130 mm Hg, which means the RV systolic pressure is also 130 mm Hg. Given the gradient of 93 to 100 mm Hg between RV and PA, one can estimate the systolic PA pressure to be between 30 and 37 mm Hg. Thus, pulmonary vascular disease is not likely to be present.

Findings
There was midline crossing of the systemic venous blood stream from the RA to the RV as the latter is abnormally positioned to the left (Fig. 59-4A). Moreover, the pulmonary venous blood stream is shown to cross the midline from the LA to the LV, which is abnormally positioned to the right (Fig. 59-4B).

Comments: Although the AV connection seemed to be concordant as determined by segmental analysis, it differs conspicuously from what is usually observed. As opposed to the normally located right and left atria, the RV is positioned to the left while the LV is positioned to the right. The AV connections are therefore crossing each other as demonstrated by the course of the depicted systemic and pulmonary venous blood streams. This anatomic setting has been denoted as a criss-cross heart.

The essence of the criss-cross heart is a rotational abnormality of the ventricular mass so that the relationships of the ventricular chambers are not as anticipated for the given AV connection. This might be explained by postseptational rotation of the ventricular mass along its long-axis during embryogenesis.
One indication of the abnormal relationship is the inability to obtain a traditional four-chamber view, since the atrial septum and ventricular septum do not lie in the same plane. A VSD is a mandatory part of the criss-cross relationship.

**MAGNETIC RESONANCE IMAGING**

![Figure 59-5 Axial SSFP cines.](image1)

**Findings**
AV discordance with criss-cross connections and VA discordance.

LV and RV systolic function were normal. There was RVH. Both mitral and tricuspid valves functioned well apart from mild mitral regurgitation. There was a large VSD.

**Comments:** Both atria and both ventricles were present, but it was not possible to obtain a four-chamber view. The mitral and tricuspid valves were not in the same plane. The mitral valve opened right to left from RA to LV (top) and the tricuspid valve opened posteroanteriorly, from LA to RV (bottom).

**Ventricular Volume Quantification**

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<th>LV</th>
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<th>LV</th>
<th>Normal range*</th>
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<td>EDV (mL)</td>
<td>112</td>
<td>(77–195)</td>
<td>86</td>
<td>(88–227)</td>
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<tr>
<td>ESV (mL)</td>
<td>48</td>
<td>(19–72)</td>
<td>32</td>
<td>(23–103)</td>
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<td>SV (mL)</td>
<td>64</td>
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<tr>
<td>EF (%)</td>
<td>57</td>
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<td>63</td>
<td>(50–76)</td>
</tr>
<tr>
<td>EDVi (mL/m²)</td>
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<td>(66–101)</td>
<td>48</td>
<td>(65–111)</td>
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<tr>
<td>ESVi (mL/m²)</td>
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<td>(18–39)</td>
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<td>(18–47)</td>
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<tr>
<td>SVi (mL/m²)</td>
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<td>(43–67)</td>
<td>30</td>
<td>(39–71)</td>
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<tr>
<td>Mass index (g/m²)</td>
<td>61</td>
<td>(59–92)</td>
<td>49</td>
<td>(22–49)</td>
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![Figure 59-6 Oblique coronal plane SSFP cine.](image2)

**Findings**
The LA was located superior to the RA. The right upper pulmonary vein was visible entering the LA. The tricuspid and mitral valves did not open in the same plane, their paths of flow being almost perpendicular to one another.

**Comments:** The atrial septum normally lies in a plane that is nearly parallel with the long axis of the body, that is, in a superior-inferior direction. Here the atrial septum is almost horizontal.

Both inlet valves are seen in this view, and may appear to enter a common ventricular chamber. Instead, we are actually seeing the interventricular septum _en face_, in a plane almost perpendicular to that of the atrial septum.

As mentioned, one feature of criss-cross anatomy is that it can be impossible to obtain a true four-chamber view, because of the rotation of the ventricular cavities relative to the atria.
Cases in Adult Congenital Heart Disease

The Criss-Cross Heart

Findings
In the upper frame, the relative position of the four valves was visible (clockwise from upper left were the aortic, pulmonic, tricuspid valve opening to the RV, and mitral valve below opening to the LV). The aortic valve lay anterior to the pulmonic valve, whereas the tricuspid valve lay superior and posterior to the mitral valve.

In the lower frame, taken 1 cm more apically from the base, the open tricuspid (upper right) and mitral (lower left) valves were seen. The ventricular septum between them was interrupted by a large septal defect. Farther toward the apex (not seen), the ventricular septum lies in a more anterior-superior plane.

Comments: It is difficult to conceptualize the 3D configuration between the atria and ventricle from 2D images. Knowing the relative location of the valves can be helpful.
Recall that from earlier views, the tricuspid and mitral valves do not truly lie in the same plane as is suggested here, but each deviates slightly toward its respective ventricle, opening at 90° angles to each other.

Findings
The great arteries were parallel with the aorta located anteriorly. There was mild subvalvar pulmonary stenosis and severe valvar pulmonary stenosis (peak velocity > 5 m/sec). The location of the previous band was not obvious. The PA branches appeared relatively normal.

Comments: One advantage of MRI is in its ability to clearly demonstrate the relative location of the great arteries, which can be hard to see behind the sternum with echocardiography. Here they appear transposed, meaning that the echocardiogram was likely incorrect in its description of VA concordance.

The severe pulmonic stenosis (rather than a PA band) may have protected the pulmonary vasculature from excessive flow.

Findings

Comments: It is difficult to conceptualize the 3D configuration between the atria and ventricle from 2D images. Knowing the relative location of the valves can be helpful.
Recall that from earlier views, the tricuspid and mitral valves do not truly lie in the same plane as is suggested here, but each deviates slightly toward its respective ventricle, opening at 90° angles to each other.
Complex Congenital Heart Conditions / Congenitally Corrected Transposition

The LA drained through the tricuspid valve to the anterior RV, which then ejected to the anteriorly located aorta.

**Comments**: This image illustrates the AV and VA discordance. The PA is located between the RV inlet from the LA and its outlet to the aorta (see prior image demonstrating valve planes).

Out of plane, the LV, partially seen posterior to the RV in this view, fills from the RA (inferior to the plane depicted here), and empties to the PA. The superior vena cava is present here in cross section adjacent to the aorta.

**CATHETERIZATION**

Not performed.

**FOCUSED CLINICAL QUESTIONS AND DISCUSSION POINTS**

1. What is the complete anatomic diagnosis?

   The patient has atrial situs solitus, AV discordance, and a criss-cross AV relationship with a large, nonrestrictive VSD and VA discordance, which may also be known as congenitally corrected transposition of the great arteries, or L-TGA.

   The criss-cross relationship is not difficult to conceptualize once it is understood that it refers to the relative orientations of the AV valves, and hence the locations of the ventricles relative to the atria, regardless of the atrial situs or the sequence of connections, which can each be normal or abnormal. A key to recognizing the criss-cross relationship can be failure to locate a four-chamber plane, the atrial septum and the ventricular septum being almost perpendicular to each other (see Fig. 59-4), as are the two AV flow paths. Associated cardiac anomalies are common, encompassing transposition of the great arteries, double outlet RV, VSDs, straddling AV valves, and pulmonary outflow tract obstruction.

2. What is the clinical importance of the criss-cross configuration?

   Criss-cross heart is an infrequently occurring abnormality. Nevertheless, knowledge of this condition is highly important as the complex anatomy may make the other commonly associated lesions more difficult to manage surgically. This is illustrated by the present history, as the operation embarked on in childhood had to be discontinued when exposure of the heart demonstrated an anatomy considered not suitable for VSD closure.

   In this case, the MRI proved unquestionably the double discordance by demonstrating the anterior aorta and parallel great arteries, which indicates transposition. The great arteries can be difficult to visualize with echocardiography.

3. Why did the echocardiogram and MRI differ in the anatomic interpretation?

   Both modalities identified the main criss-cross relationship, and indicated the various locations of each of the four chambers. The key difference was in differentiating the morphologic RV from LV. In identifying the more anterior ventricle as the morphologic RV, the MRI changed the diagnosis from AV and VA concordance (normal relationships), to AV and VA discordance (or L-TGA).

   The key means of differentiating the morphologic RV from LV were not readily apparent by the echocardiogram. First, trabeculations usually identify the RV, but both ventricles were fairly similar in their trabeculations given the RVH. Second, the moderator band is present in the RV, but a distinct moderator band was not easily seen in the Echo. Third, the tricuspid valve, always part of the RV, is usually apically displaced, but both AV valves arose from the same plane in this complex arrangement. Fourth, the RV is typically the more anterior ventricle, as in this case, although typically this is not seen in L-TGA. Hence, the discrepancy between the two studies is easily understood.

4. What surgical options could be entertained?

   There are three surgical options. Usually, the preferred approach is biventricular repair that would involve VSD closure and take-down of the PA band. This would relieve the pressure load on the RV, opposing the risk for heart failure and would provide adequate pulmonary blood flow. However, the criss-cross configuration would still make this a difficult repair, with a high operative risk markedly exceeding that of a regular VSD repair. Risks would include heart block, incomplete closure, or outflow tract obstruction.

   A second option would be to try and do a double switch to correct the discordance and leave the patient with a systemic LV. Of uncertain benefit in even the best of circumstances, here again, however, the criss-cross relationship makes this option impossibly complex.

   The third option is a Fontan palliation. Because of the pulmonic stenosis, the pulmonary pressure and resistance may be normal. Hence, one might still consider surgical cavopulmonary connection. This is predominantly performed in functionally univentricular hearts to eliminate cyanosis and provide adequate pulmonary blood flow.

   However, as the patient is doing very well and is relatively acyanotic, one might consider him ideally “balanced” (cf. Case 60). A Fontan circulation would not offer improvement over his hemodynamic situation at the moment, and the long-term results of Fontan connections are still associated with significant mortality and morbidity caused by atrial arrhythmia, venous congestion, protein-losing enteropathy, thromboembolism, and ventricular failure. Any surgery, therefore, would not likely improve the patient’s current clinical situation.

4. What is the patient's long-term prognosis?

   Although the patient’s clinical situation is currently satisfactory, there might be a need for further intervention over time. The RV has been exposed to systemic intraventricular pressure for many years and there is pronounced RVH, though without any evidence of impaired systolic function. However, there is a long-term possibility of right-sided heart failure. There is the risk of paradoxical embolism. Tachyarrhythmias may also occur requiring intervention.

   The patient has done extremely well thus far and has demonstrated an optimal balance of pulmonary blood flow and little cyanosis or shunt. Typically, this “balanced” physiology is best left alone (see Case 60). The patient’s future prospects are uncertain, but a limited life span can be anticipated.

**FINAL DIAGNOSIS**

Criss-cross heart with large VSD and consequent PA banding established during infancy

**PLAN OF ACTION**

Yearly follow-up.

**INTERVENTION**

None.

**OUTCOME**

Two years later, at his routine follow-up, the patient noted no change in his exertional tolerance and continued to maintain his healthy, active lifestyle.
Selected References

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