Diagnosis of Cor Triatriatum by Left Ventricular Angiocardiography*

Report of a Case

DONALD A. GIROD, M.D. AND GERALD J. KURLANDER, M.D.

Indianapolis, Indiana

Cor triatriatum is a rare congenital malformation characterized by an anomalous membrane which divides the left atrium into two compartments. This membrane contains one or more openings which vary in size. If the opening is large it may render the anomaly hemodynamically insignificant and if it is small death usually occurs in infancy. A reasonable embryologic explanation for the occurrence of the membrane is failure of incorporation of the common pulmonary vein into the wall of the left atrium, associated with narrowing or lack of widening of the orifice between the two chambers. As a result, all four pulmonary veins drain into the postero-superior chamber. The left atrial appendage and mitral valve are related to the antero-inferior compartment and consistent with the development hypothesis mentioned above, the foramen ovale is usually in the atrial septum that is contiguous with this compartment. The purpose of this brief communication is to emphasize the value of left ventricular angiocardiography in the diagnosis of this anomaly.

*From the Departments of Pediatrics, Radiology and the Heart Research Center, Indiana University Medical Center. Aided by grants from the James Whitcomb Riley Memorial Association, the Marion County Heart Association of the Indiana Heart Association, and with facilities provided by Cardiovascular Clinical Research Center Grant H-6308, National Heart Institute, National Institutes of Health.

Figure 1: Frontal chest roentgenogram demonstrates cardiac enlargement with right ventricular contour. The pulmonary veins particularly in the upper lobes are enlarged. The opacity of the central third of the right lung represents fluid in the alveoli and interstitium.
CASE REPORT

The patient was an eleven and one-half month old white boy who was admitted to the Indiana University Medical Center because of congestive cardiac failure. He had been asymptomatic and had grown and developed normally until approximately three months prior to admission. At that time, it was noted that he had become somewhat listless and ate poorly. Approximately one week prior to admission he became dyspneic, pale, and more listless, and was admitted to a local hospital. A diagnosis of congestive cardiac failure probably on the basis of endocardial fibroelastosis or acute viral myocarditis was made. He was treated for congestive cardiac failure and then referred to the Indiana University Medical Center.

Physical examination revealed a respiratory rate of 60 per minute, heart rate of 180 per minute, normal blood pressure and temperature. He weighed 18 pounds. The child appeared acutely ill with rapid labored breathing associated with an audible grunt. Rales were heard in the lungs bilaterally. A faint systolic murmur was audible at the lower left sternal border. The second sound was split and the pulmonic valve closure component was accentuated. The liver was enlarged, the edge being palpable about 5 cm. below the right costal margin.

The electrocardiogram revealed an axis of 100° and showed marked right ventricular hypertrophy. Chest roentgenograms showed cardiomegaly. There was enlargement of the right ventricle and marked enlargement of the "left atrium." The pulmonary veins were large and there was evidence of fluid in the alveoli and interstitium of the lungs particularly on the right (Fig. 1).

Cardiac catheterization was performed ten days after admission. The pertinent findings included elevated right ventricular, pulmonary artery, and pulmonary arterial wedge pressures and normal left ventricular end-diastolic and aortic pressures. The catheterization did not indicate an intracardiac shunt or semilunar valvar obstruction.

Cineangiogram performed in the frontal projection demonstrated the membrane dividing the "left atrium." The membrane appeared as a radiolucent line between the contrast filled compartments of the "left atrium" and moved with atrial systole and diastole. A second cineangiographic sequence in the right anterior oblique projection with an injection in the left ventricle demonstrated filling of the lower of the two atrial compartments by regurgitation through

\begin{figure}
\centering
\includegraphics[width=0.5\textwidth]{figure2.png}
\caption{Right anterior oblique roentgenogram of the chest with barium in the esophagus demonstrates posterior esophageal displacement by a large "left atrium."}
\end{figure}
the mitral valve. Mitral regurgitation occurred during a period of cardiac arrhythmia induced by the left ventricular injection of contrast media (Fig. 3a).

Plans were then made to proceed with surgery for removal of the membrane, but the patient deteriorated on the third day following cardiac catheterization and expired.

Necropsy studies confirmed the diagnosis of cor triatriatum. A membrane extended across the left atrium so that the pulmonary veins and approximately two-thirds of the left atrial volume was included above the membrane. The left atrial appendage was part of the chamber adjacent to the mitral valve. Two communications were present in the anomalous septum between the two chambers, measuring 5 mm. and 2 mm. in diameter respectively. The mitral valve was normally formed except for the presence of several small nodular thickenings along the atrial side of the free edge of the anterior leaflet.

**DISCUSSION**

The angiocardiographic diagnosis of cor triatriatum has been established previously. This has been achieved by the use of pulmonary arteriography except in one instance. The septum has then been demonstrated in the frontal projection, contrast media having passed through the pulmonary circulation filling the chambers on both sides of the membrane. The use of left ventricular angiocardiography in the right anterior oblique projection as an aid in the diagnosis of cor triatriatum has not been previously reported. In our case, regurgitation of contrast media occurred through what appeared to be a normal mitral valve. It is probable that the regurgitation occurred secondary to arrhythmia induced by the injection of contrast media. The discrepancy between the size of the left atrium as indicated by the regurgitant contrast media and the size of the left atrium as seen on the barium esophagram in the

---

**Figure 3A**
Gineroentgenogram made in the right anterior oblique projection after injection of contrast media into the left ventricle. The lower of the two left atrial chambers is opacified by regurgitation of contrast media and is bounded superiorly by a "straight margin." The unopacified upper chamber produces the esophageal impression seen in Fig. 2. **Figure 3B**: Diagramatic representation of Fig. 3A. The broken line defines the "straight margin" which represents the under surface of the anomalous membrane.
FIGURE 4: Necropsy specimen. The posterior wall of the left atrium and left ventricle are opened revealing the anterior leaflet of the mitral valve (MV). A membrane (M) divides the left atrium into a posterior superior compartment (PS) above and antero-inferior compartment (AI) below. A probe traverses the larger communication between the compartments. An additional smaller communication is seen in the left upper margin of the membrane.

right anterior oblique projection was highly suggestive of the diagnosis of cor triatriatum (Fig. 2). In this case, further confirmation would have been obtained if a pulmonary artery injection had been made in the right anterior oblique projection for purposes of comparison with the left ventricular injection made in the same projection.

The pulmonary venous hypertension associated with this lesion resulted in the sudden onset of pulmonary edema and death. It is suggested on the basis of this experience that when the diagnosis of cor triatriatum is made, surgical treatment should be planned and executed as quickly as possible.

REFERENCES

For reprints, please write: Dr. Girod, 1100 West Michigan, Indianapolis.