The Significance of Asplenia in the Recognition of Inoperable Congenital Heart Disease

By William C. Roberts, M.D., William B. Berry, M.D., and Andrew G. Morrow, M.D.

SPLENIC AGENESIS in association with congenital heart disease implies the presence of multiple and complex malformations of the heart and great vessels, and consequently inoperable cardiac disease. Such was the case in an extremely cyanotic and dyspneic 3-month-old male infant with no cardiac murmur. The electrocardiogram showed right axis deviation, and suggested right atrial enlargement, right ventricular hypertrophy, and abnormal positioning of the ventricles. Chest roentgenogram (fig. 1) disclosed a normal-sized, normally located heart and a right-sided gastric air bubble. Selective angiography (fig. 2) showed transposition of the great vessels, a single ventricle, a right aortic arch, and probable pulmonic valvular and subvalvular stenosis. Necropsy following sudden death revealed additional malformations (figs. 3 and 4).

This patient demonstrates the syndrome of splenic agenesis, partial situs inversus, and multiple congenital cardiovascular anomalies. Approximately 100 patients with this entity have been reported. All of the more common malformations of the heart and great vessels associated with this syndrome are illustrated in the patient described herein. The complexity of the cardiac disorders can be determined only by intracardiac catheterization and usually by angiography. The diagnosis of asplenia, on the other hand, frequently may be made simply by study of the peripheral blood smear. Howell-Jolly bodies or siderotic granules in the erythrocytes of the peripheral blood for practical purposes indicate an absent spleen. The presence of situs inversus, which often suggests that asplenia also exists, can usually be determined by routine radiographic examination. The gastric air bubble on the right means situs inversus. The upper abdominal roentgenogram is extremely helpful in determining the presence or absence of the spleen and will frequently demonstrate the anteroinferior margin of the liver on the left, and also flattening of this margin.

Summary and Conclusion

The finding of asplenia and situs inversus in a patient with congenital heart disease virtually precludes the presence of cardiac lesions which would be benefited by corrective or even palliative surgical procedures.
Figure 2
Angiocardiograms. The contrast material is injected into the systemic ventricle. a. Anteroposterior projection showing that the aortic valve (A.V.) lies directly to the left of the pulmonic valve (P.V.), and that the two semilunar valves are on the same frontal plane. The rudimentary chamber proximal to the pulmonic valve appears to be filled in a retrograde fashion, since no contrast material is seen entering this subvalvular outflow tract from the systemic ventricle. The aorta descends (D.A.) on the right and the great arteries arising from the arch have a mirror-image reverse of normal. The patent ductus arteriosus is not clearly identified. b. Lateral view. The aorta arises anteriorly, indicating transposition of the arterial trunks. In this view the pulmonary trunk is apparent directly behind the proximal portion of the ascending aorta.

Figure 3
Diagrammatic representation of the heart and great vessels. Blood enters the right atrium (R.A.) through the superior vena cava (S.V.C.) and the hepatic vein (H.V.). The termination of the inferior vena cava was not determined at the time of the original dissection, but it is apparent that this vessel did have an abnormal course.

The coronary sinus is absent. Blood in the right atrium either enters the left atrium ("L.A.") through defects in the lowermost and midportions of the atrial septum, or enters the systemic ventricle directly through a common atrioventricular valve. No vessels are connected to the left atrium, although a small protrusion on its surface suggests a rudimentary vascular bud. The right and left pulmonary veins drain into a common pulmonary vein (shown in figure 4), which in turn terminates by dividing into two branches: the larger one connects to the "left" gastric vein; the smaller one, to the portal vein. The left-sided atrium is anatomically a right atrium in that its wall is composed entirely of pectinate muscles. The systemic ventricle, which is large and thick-
walled, functions as a single ventricle. The leaflets of the common AV valve are not continuous with those of either the aortic or pulmonic valves. An intramural opening (0.3 cm. in diameter) below the aortic valve connects the systemic ventricle to a rudimentary, smooth-walled chamber below a stenotic dome-shaped, unicuspid, unicommisural pulmonic valve. The pulmonary trunk is hypoplastic. The aortic valve is located on the same plane and directly to the left of the pulmonic valve. The aorta arises anteriorly and does not cross the pulmonary trunk in its ascent. A small patent ductus arteriosus, which is connected to the right pulmonary artery, is present. The pressure in the systemic ventricle was recorded as 70/5 mm. Hg and the peripheral arterial oxygen saturation was 58 per cent. In summary, there is total anomalous pulmonary and systemic venous drainage, persistent common atrioventricular canal, common ventricle, transposition of the great vessels, stenotic subpulmonary outflow tract with pulmonic valvular stenosis, patent ductus arteriosus, right aortic arch, absent coronary sinus, and anatomic double right atrium.

References

This drawing demonstrates partial situs inversus, symmetrically lobed lungs, and abnormal systemic and pulmonary venous connections in the patient described. The largest lobe of the liver is on the left, the stomach and tail of the pancreas, on the right. The gallbladder is in the midline, and the spleen is absent. The colon and appendix are normally located but the mesenteric attachments of the small intestine are abnormal.

Amid the racket and hurly-burly few of us have the chance to warm both hands at the fire of life.—SIR WILLIAM OSLER. Aphorisms From His Bedside Teachings and Writings. Edited by William Bennett Bean, M.D. New York, Henry Schuman, Inc., 1950, p. 81.
The Significance of Asplenia in the Recognition of Inoperable Congenital Heart Disease

WILLIAM C. ROBERTS, WILLIAM B. BERRY and ANDREW G. MORROW

Circulation. 1962;26:1251-1253
doi: 10.1161/01.CIR.26.6.1251

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1962 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/26/6/1251.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/