A New Roentgenographic Sign of Hypoplastic Left Heart*

Gordon M. Folger, Jr., M.D.** and Abraham Saied, M.D.†

Thirteen infants with autopsy-proved hypoplastic left heart syndrome were studied, and their plain chest roentgenograms were reviewed for the presence of features specific for this anomaly. The infants were divided into two groups determined by their hemodynamic findings, which further appeared to determine longevity. Seven of these infants had similar roentgenographic findings, considered due principally to the absence of the ascending aortic shadow. This finding was present in all infants surviving longer than one week. When not found, its absence was best related to obscuration by the large thymus seen in the immediate postnatal period. The sign is considered an important diagnostic aid in determining the presence of this difficult neonatal cardiovascular problem.

Isolated roentgenographic findings are frequently of value in the recognition of malformations of the heart. Thus, it is well recognized that certain radiographic features frequently associated with total anomalous pulmonary venous return to the superior vena cava, transposition of the great vessels, tetralogy of Fallot, and the scimitar syndrome may all suggest or confirm the diagnosis. This may assume great importance when one considers the difficult diagnostic conditions present in the neonate and small child.

The purpose of this report is to describe a roentgenographic finding, observed in infants with the hypoplastic left heart syndrome, which has been extremely helpful in the diagnosis of this particularly troublesome problem and which, in our experience, has been quite specific for this and closely related to malformations. To our knowledge, this finding has not been previously reported.

Materials and Methods

The hypoplastic left heart syndrome (HLHS) as described in this communication, refers to patients with aortic or mitral valvular atresia or both, with normal alignment of the great arteries and intact interventricular septum (Fig 1a).

All the infants had aortic valvular atresia and three had mitral valvular atresia in addition. Ten were males. The age at time of death was between 24 hours and 10 weeks. There were no survivors. Chest roentgenograms were reviewed and postmortem drawings of the same heart superimposed. Characteristic anatomic findings in typical heart with HLHS are seen. Note size and position of ascending aorta (arrow), which preclude its contributing to right superior aspect of cardiovascular silhouette. Ao Ar = aortic arch; PDA = patent ductus arteriosus; MPA = main pulmonary artery; LV = left ventricle; RV = right ventricle; and RA = right atrium.

Figure 1a. Composite of frontal chest roentgenogram and postmortem drawing of same heart superimposed. Characteristic anatomic findings in typical heart with HLHS are seen. Note size and position of ascending aorta (arrow), which preclude its contributing to right superior aspect of cardiovascular silhouette. Ao Ar = aortic arch; PDA = patent ductus arteriosus; MPA = main pulmonary artery; LV = left ventricle; RV = right ventricle; and RA = right atrium.

*From the Division of Pediatric Cardiology, Henry Ford Hospital, Detroit, Mich., and The Department of Pediatrics, Division of Pediatric Cardiology, Medical College of Georgia, Augusta, Ga.
**Director, Division of Pediatric Cardiology, Henry Ford Hospital, Detroit, Mich.
†Formerly Fellow in Pediatric Cardiology, Department of Pediatrics, Medical College of Georgia, Augusta.

Manuscript received February 23; revision accepted March 28, 1973.

Reprint requests: Dr. Folger, 2799 West Grand, Detroit 48202

CHEST, VOL. 64, NO. 3, SEPTEMBER, 1973
NEW ROENTGENOGRAPHIC SIGN OF HYPOPLASTIC LEFT HEART

were analyzed, with special attention to heart size and contour, chamber enlargement and pulmonary vascular appearance.

Cardiac catheterization was performed in six of the patients, with analysis of pressures, oxygen saturation and pulmonary and systemic blood flow. Postmortem examination was performed in all 13 patients and the anatomy of the cardiac chambers, interatrial and interventricular septa, great vessels and coronary arteries was confirmed.

RESULTS

The detailed clinical and hemodynamic appearance of these infants has been the subject of a previous report. Summarizing these findings, the infants could be grouped according to longevity: Group 1 consisted of nine patients, all of whom died under one week of age; and Group 2 (four patients), who survived beyond this period. Hemodynamic correlation with the clinical findings was possible when the two groups were compared. The principal findings indicated preservation of systemic blood flow by the maintenance of high pulmonary vascular resistance, and thus potentiation of the necessary flow from the pulmonary to the systemic circuit via the ductus arteriosus in the longer survivors. In contrast, the patients in Group 1 had greatly elevated pulmonary blood flow and died early in a peripheral shock-like state.

ROENTGENOGRAPHIC FINDINGS

The chest roentgenograms (Fig 1, 2) revealed marked cardiomegaly, with the cardiothoracic ratio averaging 0.60 (range 0.55 to 0.68). All patients were considered to have radiographic evidence of right ventricular enlargement, with eight exhibiting distinct right atrial enlargement. All had increased pulmonary vascular markings, which were more prominent in those infants in acute distress at the time the films were taken. These findings are quite routine for this anomaly, but may be found with other conditions as well.

The most prominent and important radiographic feature was the absence of the shadow of the ascending aorta, which produced in the right aspect of the cardiac silhouette an angulation at the junction of the superior vena cava and the right atrial shadow, giving to this segment the appearance of the

Figure 1(b). Representative frontal chest roentgenogram with superimposition of reversed "5" sign. See text for description.

Figure 2 (a, b, left and right). Absence of aortic shadow. Note striking similarity. Panels a and b also show prominent pulmonary venous congestion.

CHEST, VOL. 64, NO. 3, SEPTEMBER, 1973
numeral "5" in reverse (Fig 1b). This feature was present in seven patients and was most evident when there was marked right atrial enlargement.

Although not seen in all patients, it was universally present in those who survived longer. Three patients from Group 2 failed to show this finding in the films.

![Figure 2](image1)
**Figure 2** (c, d, left and right). C. Absence of aortic shadow. D. Full right heart border due to presence of thymic shadow in patient from Group 1.

![Figure 3](image2)
**Figure 3.** Frontal chest roentgenogram of same patient; (a, left) three hours after birth; (b, right) 96 hours after birth. Although there is obvious difference in radiographic technique and patient positioning, respiratory effort appears comparable as does relationship of barium-filled esophagus to tracheal air shadow, indicating no difference in rotation. Width of right cardiothymic shadow at age 3 hours contrasts with that at 96 hours, suggesting diminution of thymic size resulting in visualization of reversed "5" sign.
NEW ROENTGENOGRAPHIC SIGN OF HYPOPLASTIC LEFT HEART

FIGURE 4. Single frame from retrograde cineaortogram in left anterior oblique projection. Injection was performed in descending aorta with catheter through ductus arteriosus in left anterior oblique projection. Note retrograde opacification of diminutive ascending aorta (arrow) and coronary arteries. RCA = right coronary artery; LCA = left coronary artery; MPA = main pulmonary artery; PDA = patent ductus arteriosus; and Ao Ar = aortic arch.

obtained in the first 48 hours, but it was apparent in films obtained several days later (Fig 3).

The extreme reduction in the size of the ascending aorta, which is the principal cause of the alteration of the cardiac silhouette is readily seen at the time of angiographic study, with contrast material injected either directly into the ascending aorta, or as we have preferred, at the ductal level (Fig 4). Although the illustrated angiogram has been performed in the left anterior oblique projection, the failure of the small ascending aorta to project anteriorly and to the right can be appreciated.

The other angiographic, as well as the hemodynamic, features of this condition are reported elsewhere. Briefly, they confirm the presence of function of only the right ventricle, which therefore is the sole means of perfusion of both the pulmonary as well as the systemic beds. Systemic pressure sufficient to maintain life thus depends on the integrity of the ductus arteriosus and, equally important, the persistence of significantly elevated pulmonary vascular resistance.

DISCUSSION

In the last decade, there has been an increased interest in this relatively common anomaly. A diagnosis of most of the cases, however, has been made at autopsy. Patients with HLHS rarely survive past the first month of life and, as with our patients, survival past one week is unusual. This, in addition to the distinct possibility that, as recently indicated, a surgical procedure may now be a reality for these extremely ill infants, makes early recognition of their condition, before its predictable and probably irreversible deterioration, of extreme importance.

The interpretation of the chest roentgenograms in newborn infants is difficult, and those with congenital heart malformations may present with a wide variety of roentgenographic appearances of the heart. However, the clinical diagnosis of certain cardiovascular malformations, such as transposition of the great vessels and, as well, tetralogy of Fallot may be established with significant accuracy on the basis of the appearance of the heart shadow alone; the characteristic radiographic patterns of various other defects may often not be so evident in early life.

In patients with HLHS, we have observed that the right border of the cardiovascular shadow consists of two parts: the upper is formed by the superior vena cava alone, which can often be traced from beneath the right clavicle to its point of entrance into the right atrium, and inferiorly, the shadow of the right atrium itself. The ascending aortic shadow is absent, and, as the right atrium is usually enlarged, patients with HLHS may show a striking angulation at the point of junction of the superior vena cava with the right atrium, and the right cardiac silhouette assumes the appearance suggestive of a reversed “5.” We have not knowingly encountered this finding in infancy in any other condition of the heart or great vessels. A similar finding in the older child may be seen with other malformations that result in marked diminution of the size of the ascending aorta, such as large atrial septal defects and total anomalous pulmonary venous connections, but these do not produce aortic underdevelopment in utero and thus do not manifest the finding in the neonatal period.

This radiographic sign was evident in seven of our patients with HLHS and in each of the affected infants who did not have a prominent thymic shadow. When present, this sign was to us as helpful for the proper clinical diagnosis as the “figure of 8” observed in total anomalous pulmonary venous connection to the superior vena cava, the “egg-shaped” silhouette of transposition of the great arteries, and the small “boot-shaped” heart seen with typical tetralogy of Fallot.

REFERENCES

Clinical Imagery Through X-rays

The early educational background of Wilhelm Konrad Roentgen (1845-1923) is of interest. He was an average student not because he lacked talent but possibly because didactic standards of his day were too slow for his exceptional capabilities. In 1864 he was expelled from school because he refused to divulge the name of one of his mischievous classmates. Subsequently he failed his reentry examination. In 1866 he matriculated in the Zurich Polytechnic Institute where he graduated as a mechanical engineer in 1868. Then he became assistant to his former teacher, A. Kundt of Zurich. He continued in this post when Kundt became Chairman of the Department of Physics at the University of Wurzburg and in 1872 at the University of Strassburg. During his tenure there, Roentgen published papers on electric discharges through conductors and insulators. At the age of 34 he became the Chairman of the Physics Department at the University of Giessen and assumed the same post at the University of Wurzburg at the age of 43 in 1888. Seven years later, after having explored the experimental studies of Crookes, Hertz, Hittorf, and Lenard, and while experimenting with electric current passing through a vacuum tube, he made his epoch-making discovery: "new kind of rays." For this achievement he became the first recipient of the Nobel Prize in Physics, in 1901. He designated the "new rays" as x-rays. The letter x was used first by René Descartes, the French philosopher and mathematician (1596-1664) to symbolize unknown qualities. The medical application of x-rays spread rapidly all over the civilized world. To refer to Roentgen's discovery as serendipity is ill-conceived and unjustified. On the contrary, it was the result of a logical quest and systematic pursuit of deciphering the unsolved code of new phenomena. The luminous flicker on the small barium platinocyanide plate, the first revelation of the existence of x-rays, was not accidental. Roentgen intended to use this screen in his pertinent investigations. He belonged in a category of men of rigid intellectual discipline, sound logic and detached skepticism. Inquisitive, research-oriented minds of men of this type are always prepared to see the light and register new knowledge where others perceive only a vacuum. In reference to the respiratory system, several valuable diagnostic x-ray methods have become available, including the Potier-Bucky technique, tomography, and bronchography. Attempts at photofluorography started shortly after Roentgen's great discovery. Manoel de Abreu is regarded as the originator of serviceable mass photofluorography. As to cardiovascular diseases, according to Zimmerman (Intravascular Catheterization, Springfield, C C Thomas, 1966), Fritz Bleichroeder in 1905 passed catheters in arteries and veins of experimental animals as well as of himself. In 1929 Werner Forssmann of Germany after dissecting his own forearm, passed a catheter up to the right atrium under fluoroscopic control in front of a mirror held by his nurse; then he walked to the x-ray department, with the catheter in place, for x-ray pictures. Subsequently André Courmand and Dickinson W. Richards of New York contributed extensively to the clinical applicability of this modality. The three of them shared the Nobel Prize in Medicine and Physiology in 1956. Satisfactory visualization of minute vascular changes has been accomplished by direct magnification of the x-ray image with the aid of very fine focal-spot x-ray tube and of the Schonander table top which can be elevated. With expert technique and meticulous supervision, coronary angiography has attained a prominent role in clinical diagnosis. In this manner it carries very low risk, as reported from one of the medical centers, namely 22 deaths out of 25,000 patients catheterized. The manifold ramifications of medical specialties would call for encyclopedic coverage of the subject but it is not within the framework of this writing.

Andrew L. Banyai, M.D.