Yearly checks on radiation output of equipment should be carried out. When isotopes are used for indicator dilution studies to determine shunts and cardiac output, rapidly excreted ones should be employed with low activity and short half-life.

Recent developments in the recording of the electrical events of the heart beat include a battery-operated miniaturized radio broadcasting system which permits the recording of the electrocardiogram signal without the use of wires. The sender is attached to the patient and the receiver can be located as far away as 1,500 feet. The signal can be recorded on any appropriate device including magnetic tape. The technique permits the taking of the electrocardiogram during exercise and other procedures where wires would interfere. Currently, normal children and those with heart disease are being studied under conditions of play, work, and other physical activity. Vectorcardiography may give additional information to that obtained in the electrocardiogram by way of identifying chamber hypertrophy, conduction disturbances, and myocardial infarction. The three-dimensional presentation of the electrical events of the heart is mediated through an oscilloscopic recording and then photographed. Time and voltage relationships of the cardiac forces are thereby depicted as a loop in contrast to the scalar representation on the conventional electrocardiogram.

For over 20 years the functional status of the heart and great vessels has been investigated by cardiac catheterization techniques. Through catheters it has been possible to record the intracardiac pressures and remove blood samples for oxygen analysis. More recently indicator dilution techniques have been utilized during cardiac catheterization and as mentioned above employ isotopes, dyes, chemicals with electrical charges, and warm or cold solutions. These safe methods for the detection of shunts are superior to conventional blood gas studies both quantitatively and qualitatively. The disadvantages of the techniques are that they are rather complicated and sensitive equipment is required as well as considerable technical skill. Interpretation of the data obtained requires careful study and experience. In addition to the usual right heart catheterization techniques in cardiovascular diagnostic studies it is now possible to introduce catheters into the left heart by one of three routes: (1) insertion retrograde through an arterial channel such as the brachial artery; (2) trans-septal puncture of the wall between the atria; (3) direct trans-thoracic puncture to the left ventricle. Information gained by these approaches permit better understanding of the functional status of the left heart particularly in mitral and aortic valve disease.

One of the most exciting advances in diagnostic methodology is the application of both analog and digital computers to the solution of problems in pediatric cardiology. Only recently two members of the American Academy of Pediatrics, Dr. George Veasy of Salt Lake City and Dr. John Gustafson of Des Moines, Iowa, working independently have used computers as an aid in the diagnosis of heart disease in children. Besides predicting the probability of various diagnoses including the rarest of conditions from critical information, computers can also be used to evaluate such tests as electrocardiology, vectorcardiography, phonocardiography and pulmonary function.

THE INFANT WITH HEART DISEASE

Abraham M. Rudolph, M.D.

Department of Pediatrics, Albert Einstein College of Medicine, New York

In spite of great surgical advances, congenital heart disease is still a major cause of death in children, and this is particularly true in the infant age group. The magnitude of the problem may be appreciated by reviewing some data on mortality from con-
genital heart disease collected by Carlgren in Gotenberg, Sweden. He reviewed the history of all individuals born with congenital heart disease in a 10-year period in that city. Within 15 years after birth, 40% of these individuals had succumbed. The really important factor which emerged, however, is that 30% of these patients died in the first 6 months after birth. In reviewing the causes of death in this group, I have estimated that at least 50% of these infants died of lesions which can be cured or relieved by procedures presently available. It is thus obvious that it is incumbent on us, as pediatricians, to recognize and treat the child with heart disease at the earliest possible age.

Recognition of heart disease, and particularly of specific congenital lesions, is considerably more difficult in the infant than in the older child. One problem is that the lesions encountered in infancy may be much more complicated than in older children, because after the first year many of the infants with complicated lesions may have already succumbed. More important though, is the fact that many congenital heart lesions do not present the classical clinical features we have come to recognize with specific lesions in the older child. For example, the presence of only a systolic murmur or even an absence of murmur may occur in an infant with patent ductus arteriosus, rather than the typical continuous machinery murmur seen in the older youngster. Murmurs may also be occasionally absent in infants with critically severe aortic or pulmonic stenosis and occasionally in the neonate with ventricular septal defect. Furthermore, the x-ray and electrocardiogram may not be too helpful in infancy if we consider them in relation to the traditional changes recognized in older children with congenital heart disease. The normal infant shows right ventricular preponderance in the electrocardiogram and right ventricular enlargement on the x-ray as a holdover of the fetal circulation. No departure from the normal may thus be observed in pulmonic stenosis or pulmonary arterial hypertension which classically produce a right ventricular hypertrophy pattern on the electrocardiogram in older children. Similarly, specific chamber enlargement is difficult to interpret on the x-ray of the young infant and the pulmonary vascular pattern may be deceptive in the early weeks after birth.

A review of the changes in the circulation after birth is important to the understanding of some of the hemodynamic derangements and complications seen in infants with a number of congenital heart lesions. The circulation in the neonatal period is in a transitional state from that of the fetus to that of the adult. Two major transformations occur in the circulation at birth; one is the elimination of the placental circulation by either clamping of the cord or constriction of the umbilical arteries, and the second is the opening up of the pulmonary circulation associated with expansion of the lungs with air. The relaxation of the pulmonary vasculature may be partly due to a mechanical factor but is related predominantly to the effect of oxygen in the inspired air. The normal adult circulation presents a continuously sequential flow of blood from pulmonary to systemic to pulmonary circulation. The pulmonary circulation is a low-pressure, low-resistance one, and the systemic circulation is a high-pressure, high-resistance circuit. The fetal circulation is, however, characterized among other features by the presence of a functionally single ejection system. The very large communication afforded by the ductus arteriosus results in equalization of pressures on the left and right sides of the circulation. In this type of circulation, distribution of blood flow is determined by the relative resistances of the various components. Thus, in the fetus with a relatively low resistance placental circulation, major flow occurs via the aorta to this organ. A high vascular resistance in the pulmonary circulation allows for only a small pulmonary blood flow and blood is therefore deflected through the ductus arteriosus as a right-to-left shunt. Soon after birth the elimination of the placental circulation removes the low-resistance circu-
ulation from the aorta and systemic circuit, whereas ventilation of the lung results in a significant decrease in resistance in the pulmonary circulation. For a short period of time a balanced situation exists between the pulmonary and systemic circulation; the pulmonary vascular resistance falls further and if the ductus arteriosus remains open, which in the normal infant it does for 10–15 hours, a left-to-right shunt occurs. If a large communication between the aorta and pulmonary artery, or between the two ventricles is present after birth, the pulmonary vascular resistance is a major determinant of the hemodynamic clinical manifestations of the lesion. This is well demonstrated experimentally by the results of opening and closing an artificial ductus in a newborn calf. When the animal breathes 13% oxygen which constricts the pulmonary vessels no change is detected whereas when the animal breathes air the pulmonary vessels are relaxed, decreasing the pulmonary vascular resistance, and a left-to-right shunt occurs. The particular feature of the hemodynamic changes on opening a shunt that should be stressed is that when the pulmonary vascular resistance is decreased a left-to-right shunt occurs, which results in an increase of blood returning to the left atrium and left ventricle with an increase of work load on the left side of the heart and elevation of left atrial pressure. If the pulmonary vascular resistance is markedly reduced, then an enormous left-to-right shunt would occur, resulting in a tremendous overload of the left ventricle with left ventricular failure.

In infants with large ventricular septal defect the pulmonary vascular resistance is high before birth as in normal infants and a right-to-left shunt with small systemic resistance occurs. In the period soon after birth a balanced situation exists with systemic resistance equaling pulmonary resistance and no significant shunt in either direction. No murmurs, no symptoms, and no abnormality of electrocardiogram or x-ray may be noted. As pulmonary vascular resistance drops, a left-to-right shunt occurs placing an extreme load on the left ventricle. The systolic murmur with cardiomegaly and an increase in pulmonary vasculature appears, namely, the typical criteria of ventricular septal defect. The time sequence of these clinical events is related to the rapidity with which pulmonary vascular resistance falls. The pulmonary vascular resistance continues to fall after birth due to degeneration of the muscular layer of the pulmonary arterioles. In animals, the pulmonary vascular resistance falls to adult levels within 7 to 14 days associated with a fall of right ventricular and pulmonary arterial pressure.

The presence of a large defect between the two sides of the circulation in itself appears to interfere with a normal maturation process of the pulmonary vessels and a slow decrease in pulmonary vascular resistance which thus results is a protective mechanism against the early development of severe left ventricular failure. The mechanism whereby this delay in the maturation process of the pulmonary vessels occurs is not known. It may be due to persistent pulmonary hypertension or to increased pulmonary blood flow. I would like to present a hypothesis for the persistence of a high pulmonary vascular resistance in some infants with ventricular septal defect and patent ductus arteriosus. The high flow through the pulmonary circulation produces left ventricular failure of some degree with resulting pulmonary edema. This may interfere with alveolar ventilation thus producing hypoxia of pulmonary venous blood. This hypoxia may stimulate pulmonary vascular constriction, thereby decreasing the left-to-right shunt and preventing critical cardiac failure. This persistent constriction of the pulmonary vessels due to hypoxia could result in more permanent changes in these vessels.

Cardiac failure is a major symptom arising from noncyanotic congenital heart lesions in infancy. Frequently, however, this is not recognized clinically until it has progressed to severe degree. In the common course of cardiac failure in infancy left ven-
tricular overload is the major problem and left ventricular failure occurs. Since for many years the importance of hepatomegaly in the diagnosis of cardiac failure has been stressed, a large number of infants with dyspnea, continuous or paroxysmal, are diagnosed as having bronchopneumonia or bronchiolitis when what they have is left ventricular failure. The importance of this point cannot be overstressed as the sooner pediatricians recognize that cardiac failure in infants most commonly presents as left ventricular failure with respiratory distress, the more rapidly will these infants be treated. Cardiac failure should be considered in all infants with attacks of respiratory distress or wheezing even in the absence of hepatomegaly.

Specific diagnostic studies should be carried out in any infant with noncyanotic heart disease and evidence of left ventricular failure, in addition to those babies with suspected cyanotic heart disease. Age or size of infant should in no way determine whether definitive cardiovascular diagnostic study should be attempted, since cardiac catheterization can be performed with only minimal risk in experienced hands.

THE NATURAL HISTORY OF CERTAIN CONGENITAL CARDIOVASCULAR MALFORMATIONS

Alexander S. Nadas, M.D.
Department of Pediatrics, Harvard Medical School, Boston, Massachusetts

Information on the “natural history” of congenital cardiac malformations is difficult to obtain and may have very little meaning for the statistician. One is aware that the facts obtained may yield a profile only of those patients with heart malformations who have signs and symptoms significant enough to bring them to a cardiac center. The others, either not sick enough to come to a center, or too sick and even dying before they can be brought to the specialist, or living too far from the medical center, therefore, will not be included in any survey. The only way the entire congenital heart disease population may be caught in the net of the investigator is by following up carefully a large group of newborns.

Though information gathered may not be statistically significant it is none the less important as a frame of reference in making recommendations to patients and their families. Also it is of necessary historical import that we document the course of patients with malformations as yet unaltered by the hand of the surgeons. The effect of surgery on patients with certain malformations is far reaching and changes the course of their natural history. Another reason why we should try to find out what happens without operation to certain patients is that there will be a tremendous advance in cardiovascular surgical techniques for many lesions yet in the future. Nobody questions the fact that cardiac surgery, in many, if not most, areas will be better tomorrow than it is today.

The notable exceptions to improvement of surgical techniques include repair of patent ductus arteriosus, coarctation of the aorta, secundum atrial septal defect, and pulmonic stenosis. These defects are operable at an irreducible risk and the results of surgery have proven to be good. The length of followup after repair of patent ductus extends to 25 years, of coarctation of the aorta to over 15 years, and of secundum atrial defect or pulmonic stenosis (by closed technique) to over 10 years. Since in patients with these malformations operative mortality is negligible and certainly much lower than the risk of early death or prolonged morbidity, one should not really spend time discussing their natural history without surgery.

Congenital cardiac malformations which
THE INFANT WITH HEART DISEASE
Abraham M. Rudolph
Pediatrics 1964;33:990

The online version of this article, along with updated information and services, is located on the World Wide Web at:
/content/33/6/990