American Heart Association

Congenital Cardiac Defects: A Physician's Guide for Evaluation and Management

Report of the Committee on Congenital Heart Disease

RUTH WHITTEMORE, M.D., Chairman

This report was designed primarily for the physician who is not a cardiologist but who encounters, in the course of his practice, patients with congenital malformations of the heart. It will help him decide whether and when such patients should have special studies done in a cardiac center or by a cardiologist familiar with these problems. It is divided into three parts: one on infants, one on children and adults, and the last on management. The first two parts are designed to assist the physician in the selection of patients for special studies, the last to aid him in the management of patients under his care. A special section in part III is devoted to care of the pregnant woman with a congenital defect. The decision as to which studies are needed and when they should be performed should be left to the cardiologist or center to which the patient is referred.

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PART I—SIGNS AND SYMPTOMS IN INFANTS

General Considerations

EVEN though a malformation of the heart is present at birth, such a condition does not necessarily remain static. Many malformations increase the work of the heart, and some cause a progressive increase which results in progressive cardiac enlargement, cardiac failure, and even death.

Congenital malformations of the heart are a major cause of death during infancy. Cardiac surgery can be—and has been—performed successfully on infants, even in the first weeks of life. However, the mortality rate at this early age is high. It is better to defer surgery, if possible, until childhood; but the risk of waiting must be weighed against the operative mortality. The decision to operate or not is a difficult one and calls for cardiac evaluation early in life by specially trained physicians.

An infant with a congenital defect requires more frequent observations during the first two years of life than he does in later childhood. Growth is very rapid during this period: by the time an infant is two months old he has doubled his age of a month before, and normally by the time he is four or five months old he has doubled his birth weight.

For this reason, a baby with a heart murmur should be examined at monthly intervals and special attention should be paid to weight gain, heart size, changing murmurs and development of cyanosis.

In general, an infant with a heart of normal size, whose activity is normal, who has normal color, and who gains weight normally—even though a cardiac defect is diagnosed—does not need special studies such as cardiac catheterization or angiography. Nevertheless, since his condition may change, he should be kept under observation.

Indications for Prompt Evaluation

Any of the following manifestations indicates that further study should be done promptly. Listed in order of frequency, they are:

- Dyspnea in a previously asymptomatic infant
- Increasing size of the heart
- Changing murmurs
- Easy fatigability

This is a report of the Committee on Congenital Heart Disease of the Council on Rheumatic Fever and Congenital Heart Disease of the American Heart Association and reflects that committee's findings in the matter under study.
CONGENITAL CARDIAC DEFECTS

Failure to gain weight
Increasing cyanosis
Increasing polycythemia

Indications for Immediate Evaluation

Any of the following manifestations calls for immediate diagnosis and treatment:
Attacks of paroxysmal dyspnea which progress to syncope
Progressive cyanosis with dyspnea (even in the absence of a murmur or of cardiac enlargement)
Stridor or choking spells
A ventricular rate of over 200 per minute, lasting for several hours
Cerebral vascular accidents—manifested by:
   - hemiplegia
   - convulsions
   - other neurologic signs
Marked cardiac enlargement
Cardiac failure

Signs of Cardiac Failure in Infants

Cardiac failure in infants is manifested by:
   - Rapid respirations
   - Enlargement of the heart
   - Gallop rhythm
   - Engorgement of the liver
   - Rales in the lungs—usually a late manifestation
   - Pitting edema—a very late and often an ominous manifestation

Important Clues

The first sign to suggest a cardiac malformation is usually a murmur. It is not, however, a reliable sign. A murmur in the newborn may disappear and not represent a congenital defect. On the other hand, a murmur resulting from a malformation may not be heard until later in infancy.

Color. A cardiac defect is the most common cause of persistent cyanosis. Cyanosis may appear at or shortly after birth, or perhaps not be visible for several months or even years. Persistent cyanosis usually leads to polycythemia, although this may be absent in early infancy. Rapidly increasing polycythemia is always cause for concern. For an estimate of the severity of polycythemia, the hemoglobin and hematocrit determinations are quite as important as the red blood cell count. A hemoglobin content over 20 Gm., a hematocrit reading over 70 per cent, or a red blood cell count of 7,500,000 per mm.3 is clear indication for immediate evaluation.

Pallor in an infant suggests anemia. Anemia is known to increase the work of the heart and may therefore require treatment.

Weight. Normal weight gain usually indicates satisfactory progress. The outstanding exception is the infant with valvular pulmonary stenosis and intact ventricular septum, in whom a normal weight gain is the rule.

Failure to gain nearly always points to serious cardiac trouble. In the cyanotic infant it is of ominous significance.

Increase of weight due to edema is a late indication of serious cardiac failure.

Fatigue. One of the earliest manifestations of fatigue in infancy is an excessive length of time required to take even small feedings. Shortness of breath causes the multiple interruptions in the feeding. Labored breathing may also accompany such acts as defecation.

Later, an early manifestation of fatigue may be a shorter duration of effort, as in creeping or walking, in comparison with other young children of similar age.

Respiration. Changes in the rate or character of the respirations are often the earliest manifestations of distress.

The development of tachypnea in an infant with a murmur suggests the onset of early heart failure.

Attacks of paroxysmal dyspnea are common in infants with cyanotic malformations, particularly infants in whom there is decreased pulmonary flow. In most cases, such attacks do not represent cardiac failure. Episodes of paroxysmal dyspnea are characterized by sudden onset of labored breathing and increasing cyanosis, often with an expiratory grunt. Attacks may be precipitated by exertion, excitement, febrile illness; or they may occur without any apparent cause. They do not usually last long but may be prolonged or
severe and end in syncope. Such attacks are occasionally fatal.

Attacks of stridor may be due to a vascular ring which compresses the trachea and the esophagus. Such stridor is often accompanied by a brassy cough or choking spells.

_Pulse._ Although the heart rate of a normal infant is considerably higher than that of a child or adult, a rate of 200 or more is always of concern. Even in an infant with a normal heart, _paroxysmal tachycardia_ may result in cardiac failure. Any attack which persists 4 to 6 hours requires specific treatment.

A readily palpable radial pulse combined with a weak or absent femoral pulse usually indicates coarctation of the aorta. For this reason, palpation of femoral pulses should be part of any routine examination.

_Heart Size._ The size of the heart in an infant is difficult to determine by physical examination alone. Progressive increase in the heart size out of proportion to the growth of the chest is serious; on the other hand, a relative decrease of cardiac size is a good sign. X-ray and fluoroscopic examinations allow an accurate determination of the size and configuration of the heart and of the vascularity of the lung fields. It is usually possible to determine whether the pulmonary blood flow is approximately normal, increased, or decreased. (In infants, the shadow of the thymus superimposed on the shadow of the heart may give a false impression of cardiac enlargement.)

_Caution._ The physician must remember the dangers of excessive exposure to roentgen rays and therefore control the length and number of fluoroscopic and x-ray examinations.

_Electrocardiogram._ The electrocardiogram is seldom useful in the selection of patients to be sent to a cardiac center. It is valuable, however, in the differential diagnosis of cardiac malformations and in the clarification of arrhythmias. It is often helpful, too, in the detection of digitalis intoxication. Serial electrocardiograms may aid in the demonstration of progressive changes.

The age of the infant should always be taken into consideration. The rate, conduction intervals, axis deviation and form of the complexes vary with age. There are many electrocardiographic patterns in infants within the variations of normal.

**PART II—SIGNS AND SYMPTOMS IN CHILDREN AND ADULTS**

**General Considerations**

All children and adults suspected of heart disease should be given the benefit of detailed cardiac study. Early evaluation enables the cardiologist to assess subsequent changes.

Every child with a malformation of the heart should have a yearly cardiac examination by his physician. This should include an x-ray of the chest. The patient should be seen by a physician experienced in pediatric cardiology at regular intervals during childhood and more frequently during puberty. How frequent these check-ups should be depends on the type of malformation.

Change in murmurs, development of new symptoms, or increase in the size of the heart are always indications for prompt re-evaluation. A physician experienced in pediatric cardiology is in the best position to determine the proper time for special studies and, together with the cardiac team, to determine which procedures, if any, will provide the most needed information.

Adolescent patients must be followed closely. The spurt of growth and the increase in strenuous physical activity common at this age may increase the strain on the heart.

Every woman with a cardiac defect should be carefully evaluated before she marries. If she has not had a cardiac evaluation prior to pregnancy, it should be done at the earliest possible date.

**Indications for Prompt Evaluation**

Any one of the following signs indicates that cardiac evaluation should be done promptly. Listed in order of frequency, they are:

- Shortness of breath
- Decreased exercise tolerance
- Cyanosis
- Abnormalities of heart rate or rhythm
- Cardiac enlargement
- Changing cardiac murmurs
Cardiac failure
Hypertension
Retardation of growth or development
Progression of any signs or symptoms calls for prompt re-evaluation.

Common Cardiac Defects Amenable to Surgery

Surgery may be advisable—and possible—for many children and adults with congenital defects. The following paragraphs list some of the signs associated with defects that are operable during childhood:

A continuous murmur over the pulmonary area in a noncyanotic child usually indicates patent ductus arteriosus. An operation to correct this defect is best done before the child enters school.

A strong pulse in the upper extremities, combined with a weak or absent pulse in the lower extremities, suggests coarctation of the aorta. It is advisable to defer surgery until between the eighth to twelfth year if the child’s heart is of normal size and his blood pressure is not excessively elevated.

Cyanotic children who squat when they are tired usually have pulmonary stenosis and decreased pulmonary blood flow as in tetralogy of Fallot. These children can frequently be helped by surgery.

Patients with a harsh systolic murmur over the pulmonary area and a weak or absent pulmonic second sound may have valvular pulmonary stenosis with an intact ventricular septum. Surgery is usually indicated if these patients show dyspnea, cyanosis, cardiac enlargement, or electrocardiographic evidence of marked right ventricular hypertrophy.

Closure of certain septal defects are being considered in a few cardiac centers.

Important Clues

Color. Pallor is an important sign and may be due to anemia. Severe anemia, regardless of etiology, may of itself cause murmurs or cardiac enlargement.

Persistent cyanosis of the mucous membranes usually indicates a venous-arterial shunt. Cyanosis may occur at any age and may appear initially only on exertion or increase with stress. Usually it leads to polycythemia.

Relative anemia may occur in a cyanotic child, and the greater the anemia the less obvious is the cyanosis. It must be remembered that in every instance as much as 5 Gm. per cent of reduced hemoglobin circulates in the blood before cyanosis becomes visible. For instance, in the presence of a total hemoglobin of 14 Gm., the cyanotic child has less than 9 Gm. per cent of oxyhemoglobin. Therefore, in the presence of cyanosis, any anemia indicates a greater deprivation of oxygen than is indicated by the hemoglobin level.

Clubbing occurs in most patients with cardiac malformation associated with long-standing cyanosis. It may, however, occur as a manifestation of chronic pulmonary disease and bacterial endocarditis, and occasionally it appears in normal individuals as a familial trait.

Growth and Development. Although many children with cardiac defects grow normally in height and weight, children who have large shunts often grow more slowly than normal children and may develop a “gracile habitus.” Retarded growth may be the first indication that cardiac output is inadequate to meet the demands of the body, as in large left-to-right shunts or severe anoxia.

A sudden gain in weight may indicate edema due to congestive failure.

Exercise Tolerance. A large majority of patients with congenital malformations are able to lead normal, active lives without undue fatigue or strain. Reduced exercise tolerance is an indication for referral. Minor changes of exercise tolerance in the cardiac child are difficult to evaluate because of considerable variations in the activities of normal young children. Comparison of the child’s play activities with those of playmates or siblings is often helpful. The cardiac child, for example, may seek quieter games, rest more frequently, or stop active play sooner than do other children of his age.

Cyanotic patients limit their own activity. Some cyanotic children are unable to walk more than a few feet without having to rest. They
often assume a squatting position or insist on being carried. As they grow older their exercise tolerance usually improves. If it does not, re-evaluation is indicated.

Respiration. Dyspnea on exertion, a frequent complaint, is variable and requires evaluation. It may easily be overlooked in patients with large left-to-right shunts. In a cyanotic child extreme dyspnea often precedes the need for immediate rest and in most instances is quickly relieved by the squatting position. In the young cyanotic child such respiratory distress may progress to an attack of paroxysmal dyspnea, with or without loss of consciousness. In the older child these attacks are less frequent, but if they occur they are a sign of severe anoxia.

Pulse and Blood Pressure. In any patient, child or adult, the physician should palpate the pulses of the upper and lower extremities and record the blood pressures. He should note any discrepancy between the blood pressures in the two arms or between the arms and legs and should watch for hypertension in the upper extremities. In some instances, abnormal pulsations may be seen in the neck or collateral pulsations felt over the thoracic cage.

An abnormal pulse pressure is also significant in many congenital malformations.

The pulse may reveal certain abnormal cardiac rates or rhythms, such as atrial fibrillation, paroxysmal tachycardia, ectopic beats and complete atrioventricular dissociation. The latter should be suspected in a child with a pulse lower than 60. It is wise to confirm the nature of these abnormalities by an electrocardiogram.

Heart Sounds and Murmurs. The quality of heart sounds is always important: a forceful heart sound may indicate a laboring heart; poor quality of heart sounds or gallop rhythm may indicate impaired myocardial function; a widely split pulmonic second sound, or its absence, diminution, or increase in intensity, is usually abnormal.

A question of great practical importance is whether a murmur is innocent or organic in origin.

Innocent (nonsignificant or "functional") murmurs are extremely common, occurring at one time or another in over 50 per cent of children. Such murmurs are systolic and are usually located along the left or upper right sternal border. They are vibratory or groaning, poorly localized, and may vary on change of position.

A venous hum is also innocent; it is a high-pitched continuous bruit heard lateral to the base of the heart, above or below either clavicle. This hum usually disappears when the patient lies down or when pressure is applied over the jugular vessels. It varies greatly on turning the patient's neck.

Systolic murmurs associated with a thrill are usually organic. Their quality, intensity, and location are important in the differentiation of congenital defects from acquired heart disease.

A harsh systolic murmur and thrill close to the sternum or over the base of the heart are more often indicative of congenital than acquired heart disease.

Diastolic murmurs other than those associated with a venous hum are organic. It is important to differentiate between a short diastolic murmur and a third heart sound.

Apical or basal diastolic murmurs, particularly with cardiac enlargement, occur not only in certain congenital malformations of the heart but also in acute rheumatic fever and rheumatic heart disease. (An analysis of the murmurs of rheumatic fever and rheumatic heart disease will be found in "Jones Criteria (Modified) for the Diagnosis of Rheumatic Fever.")

In most cases, a continuous murmur over the pulmonic area is due to a patent ductus arteriosus. It is usually audible in childhood but occasionally does not develop until the child is about 5 years old. This murmur is usually accentuated by exercise and is often better heard in the recumbent than in the erect position. This must be differentiated from the venous hum. Continuous murmurs may also be noted in cyanotic patients or in those with less common cardiac defects.

Cardiac Size and Configuration. The cardiac configuration, enlargement of specific chambers, and pulmonary vascularity are easier to
evaluate with x-ray in children and adults than in infants. Serial x-rays are important aids in the detection of a progressive increase in heart size or changes in vascular markings.

X-rays for size of the heart should be standard telerontgenograms taken six feet (or 2 meters) from the roentgen tube.

Fluoroscopy aids in the study of:
- Size and configuration of the heart and its chambers
- Pulsations of the cardiac border
- Vascularity of the hilar shadows
- Course of the barium-filled esophagus
- Position of the aorta and anomalous vascular structures.

Fluoroscopy should be a part of the roentgenographic study of the heart and preferably be performed by the cardiologist.

Caution. The amount of total x-ray exposure to the patient should always be carefully limited.

Electrocardiogram. An electrocardiogram is an important adjunct in the cardiac evaluation and should always include precordial leads. Cardiac arrhythmias should be confirmed by electrocardiogram.

The age of the child always should be taken into consideration in the interpretation of the electrocardiogram as the range of normal measurements varies according to age and pulse rate of the individual child and differs from the norms established for adults.

**Part III—Management**

**Attitude and General Care**

The physician should encourage parents to allow the cardiac child to lead as normal a life as possible. He should remind them not to spoil the child. Both physician and parent should maintain a cheerful outlook; this is certainly justifiable, as a malformation which cannot be helped at the present time may well become amenable to surgery in the near future.

The child with a cardiac defect should be given immunizations, particularly pertussis vaccine, at the usual time. Education is just as important as for normal children and whenever possible the cardiac child should attend a regular school.

**Physical Activity**

An infant need not be restricted in any way; he should be allowed to crawl and walk as he likes. It is not harmful to let him cry. In general, it is both unwise and unnecessary to restrict the young child with a congenital defect so long as he can rest easily and without embarrassment when he chooses. For the older child, the physician can outline a suitable program of physical activities after the child’s cardiac status and functional capacity have been evaluated.

**Protection Against Bacterial Endocarditis**

In individuals who have congenital heart disease, as in those who have rheumatic heart disease, bacteria may lodge on the heart valves or other parts of the endocardium, producing bacterial endocarditis. Transient bacteremia which may lead to bacterial endocarditis is known to occur following various surgical procedures including dental extractions and other dental manipulations which disturb the gums, the removal of tonsils and adenoids, the delivery of pregnant women, and operations on the gastrointestinal, genital, or urinary tracts.

It is good medical and dental practice to protect patients with congenital heart disease by prophylactic measures.

Penicillin is the drug of choice for administration to patients with congenital heart disease undergoing dental manipulations or surgical procedures in the oral cavity. Although the exact dosage and duration of therapy are somewhat empirical, there is some evidence that for effective prophylaxis reasonably high concentrations of penicillin must be present at the time of the dental procedure. High levels of penicillin in the blood over a period of several days are recommended to prevent organisms from lodging on the endocardium during the period of transient bacteremia.

In general, the combined oral and parenteral route of administration is preferred. All patients should be instructed to report to their physician or clinic should they develop a fever within a month following the operation.
First Choice—Intramuscular and Oral Penicillin Combined

For two days prior to surgery—200,000 to 250,000 units by mouth four times a day. On day of surgery—200,000 to 250,000 units by mouth four times a day and 600,000 units aqueous penicillin with 600,000 units procaine penicillin shortly before surgery. For two days thereafter—200,000 to 250,000 units by mouth four times a day.

Second Choice (if injection is not feasible)—Oral Penicillin

200,000 to 250,000 units four times a day beginning two days prior to the surgical procedure and continued through the day of surgery or dental procedure and two days thereafter.

Contraindications

A history of sensitivity to penicillin.

Other Antibiotics

Erythromycin or the broad spectrum antibiotics should be employed as prophylaxis in patients who are sensitive to penicillin. In those who are undergoing surgery of the genitourinary or lower gastrointestinal tract, oxytetracycline or chlorotetracycline should be administered in full dosage for five days, beginning treatment two days prior to the surgical procedure.

Protection against Respiratory Infections

Most children with cardiac defects tolerate infections (with the possible exception of pertussis) as well as other children. Patients with increased pulmonary flow or congestive failure, however, are very susceptible to respiratory infections. These patients may be greatly benefited by prolonged prophylactic use of chemotherapy or antibiotics during the younger years.

Attacks of Paroxysmal Dyspnea

Attacks of paroxysmal dyspnea are likely to occur in infants with defects which cause decreased pulmonary blood flow, such as tetralogy of Fallot. The attacks are due to anoxemia and not to heart failure.

The treatment for such an attack is:

Place the infant in knee-chest position.
Give morphine sulfate by hypodermic injection (1 mg. per 10 lb. body weight).
Give oxygen if available.
Attacks of paroxysmal tachycardia are an important indication for immediate cardiac evaluation.

Polycythemia

Polycythemia may occur in infants and is common in cyanotic children. A serious complication of polycythemia is cerebral thrombosis. As a preventive measure, the physician should prescribe an adequate fluid intake to avoid dehydration. After the first month of life, such infants require approximately one quart of fluid a day and should never be more than 8 to 10 hours without fluid even though it may be necessary to wake the infant. Children require about two quarts of fluid each 24 hours and should never be more than 12 hours without fluid. This fluid intake is especially important if the patient with polycythemia has fever, vomiting, or diarrhea, or is exposed to extreme heat.

Congestive Heart Failure

Congestive heart failure in infants and children, as in adults, requires rest, oxygen, and treatment with digitalis, morphine, and diuretics. The drugs should be given in dosages directly proportional to the body weight. If edema is present, the dosages should be based on normal body weight.

Cerebral Complications

The occurrence of cerebral thrombosis calls for immediate therapy. Oxygen therapy, venesection, fluid replacement, and other procedures may aid in lessening the extent of brain damage. Brain abscess in cyanotic children with cardiac malformations is frequent and calls for prompt treatment.

Paroxysmal Tachycardia

Paroxysmal tachycardia requires prompt therapy.
Jugular or abdominal pressure or breath holding may stop an attack. If these are un-
successful, digitalis is the drug of choice for infants and children. Great care and close observation, including frequent electrocardiograms, are advisable. Since episodes of atrial tachycardia have a marked tendency to recur in infancy, digitalis should be continued over a prolonged period of time.

Stridor

Stridor may be due to a vascular ring, a relatively rare congenital defect. During acute difficulty, hyperextension of the head and neck will often help. If choking occurs, the physician should be sure that the airway is clear. A moist atmosphere may also be helpful until the acute situation is relieved. Early surgery to correct the underlying condition is usually indicated.

Pregnancy

The therapy of pregnancy for the patient with a congenital cardiac defect begins before marriage. Many patients who survive to child-bearing age have anomalies that today are either curable or greatly ameliorated by surgical intervention. The pregnant woman with a congenital defect of the heart should be referred for evaluation at the earliest possible date. With the rapid advances in cardiac surgery today, sterilization of a young woman is seldom indicated.

With congenital defects, just as with acquired heart disease, the critical periods for development of heart failure parallel the periods of maximum load on the circulation. These critical periods are the seventh and eighth months of pregnancy, the later stages of labor, and the first postpartum days. Regardless of the type of underlying heart disease, a complete understanding of the functional capacity of the patient’s heart is important, since it is the functional capacity which dictates the management of the pregnancy. Re-evaluation of the functional capacity at each visit is necessary, because the status may change at any time during pregnancy.

Close observation of the patient during her pregnancy will help the physician predict the risk at the time of delivery and during the post-partum period. If the patient has not had any manifestations of clear-cut congestive failure prior to term, the maternal mortality is between 1 and 2 per cent; if prior to term she has developed failure which responded to treatment, the risk is considerably increased (about 10 per cent); if congestive failure is present at the time of delivery, maternal mortality is high (about 40 per cent).

Unless there are obstetrical reasons for it, Cesarean section is generally contraindicated in patients with any type of heart disease. The possible exception is coarctation of the aorta, where there is risk of a vascular accident. A woman with a congenital cardiovascular defect has a slightly greater chance of having a child with a congenital abnormality. This chance is so slight, however, that it should not be considered a contraindication to pregnancy.

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