The Management of Neonates and Infants with Congenital Heart Disease

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Donal M. Billig, M.D.
Associate Professor of Surgery
Director, Division of Cardio-Thoracic Surgery
Hahnemann Medical College and Hospital
Philadelphia, Pennsylvania

Marshall B. Kreidberg, M.D.
Professor of Pediatrics
Tufts University School of Medicine
Chief, Division of Pediatric Cardiology
Boston Floating Hospital for Infants and Children
Boston, Massachusetts

with

Harvey L. Chernoff, M.D.

Robert N. Reynolds, M.D.

Janet B. Wunderlich, M.D.

Foreword by Denton A. Cooley, M.D.

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Dedicated to

Dr. Michael Ellis DeBakey

Surgeon, Educator, Humanist

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Foreword

While the era of surgical treatment of congenital heart disease dawned some 30 years ago with operations for simple anomalies such as patent ductus arteriosus, the tetralogy of Fallot, and coarctation of the aorta, these and other subsequently developed techniques generally were not applied to neonates and infants until 10 or 15 years later. This delay undoubtedly was caused by such factors as difficulty in obtaining an accurate diagnosis in small infants, concern regarding the increased risk of operation in this age group, and perhaps a lack of appreciation of the high mortality associated with congenital cardiac anomalies during the first year of life. Thus, while a definitive medical and surgical attack upon cardiovascular anomalies was expanding, the newborn infant was being neglected. We, along with a few others, recognized the need for and feasibility of emergency surgery in infants when intensive medical treatment failed, and began, along with Dr. Dan G. McNamara, Chief of the Cardiac Clinic at Texas Children's Hospital, an active program in early diagnosis including cardiac catheterization and angiography followed by surgery. Results of this program in some 1200 infants less than 1 year of age have been most gratifying, permitting salvage of many babies otherwise doomed to death from anoxemia or congestive heart failure.

Surgical treatment for infants may be either palliative or curative and can be applied as a single operation or in two stages. The recent trend has turned away from staged procedures and many surgeons now prefer definitive one-stage operations whenever possible. This trend, of course, is highly appealing since it eliminates a second procedure with the attendant risk and morbidity. Enthusiasm for one-stage definitive treatment must be tempered, however, when the risk of open-heart surgery is prohibitive. Such situations call for careful, considered judgment. One example of the dilemma the cardiologist and surgeon face involves complete transposition of the great vessels, an anomaly with exceedingly high mortality during the first few weeks of life. Open-heart repair in the 7-lb, few-day-old infant is technically difficult, and respiratory complications after the operation may be serious. Until recently the Blalock-Hanlon

atrial septectomy was used for palliation, but this involved an open thoracotomy. The introduction of balloon atrioseptostomy by Rashkind has changed the situation, and the cardiologist now combines diagnosis and palliative treatment at the same cardiac catheterization. Septostomy allows many of the infants to survive the critical months of early development so that definitive repair can be accomplished on an elective basis when the infant is larger and respiratory function is easier to maintain. Balloon septostomy is also applicable to such defects as total anomalous pulmonary venous return, mitral atresia, and underdeveloped right and left heart syndromes.

Management of congenital cardiovascular anomalies is an ever-changing endeavor, and periodically a book summarizing the current status is extremely useful. The authors of this book have provided us with an excellent compilation based upon personal and reported experiences. The extensive references cited, which include the most significant publications regarding the particular anomalies further increase the book's value and usefulness. Dr. Billig and Dr. Kreidberg have made a notable contribution. Dr. Billig's development as an outstanding cardiac surgeon is a source of pride and personal gratification to me since he learned many of the techniques described in the book while a resident at Texas Children's Hospital in Houston. His high reputation as a pediatric heart surgeon is well deserved: He combines an aggressive spirit, an informed mind, surgical skill, and extensive personal experience. The team approach by Dr. Billig and Dr. Kreidberg, the surgeon and the cardiologist, is apparent in this book, which I predict will become a widely accepted reference and manual for treatment of neonates and infants with cardiovascular anomalies.

Denton A. Cooley, M.D.
Surgeon-in-Chief
Texas Heart Institute
Houston, Texas

Preface

The reader may be struck by the absence of many of the usual ingredients of a text on cardiac disease. He will find little discussion about murmurs, electrocardiograms, chest roentgenograms, surgical technique, or other subjects usually included in books on pediatric cardiology or cardiac surgery. To the ample number of existing texts containing all this material, we could add little, if anything, of value. These works are referred to liberally in the reference sections, for they obviously have great importance.

This book attempts to relate pathologic anatomy and pathophysiology as they affect the natural history of the various diseases, and to review current thinking on management and its effects on anatomy, physiology, clinical course, and prognosis. Thus the book is really a compendium of the indications for, the risks and expected accomplishments of and the currently available treatments for those cardiac diseases causing symptoms in neonates and infants.

The management of neonates and infants with cardiac disease is one of our current "frontiers," and as such, is open to disagreements and to changes in attitudes and approaches to various problems. We have attempted to point these up, and while we have not refrained from editorializing, we hope we have given the reader enough information on either side so that he can come to his own conclusions.

The book was a year in the writing, and in such a field a year brings many changes. Those bibliographic references which read "in press" or "personal communication" constitute an attempt to stay abreast of these changes and are possible through the kindness of friends in sending manuscript copies or letters in response to specific questions.

Finally we should like to express our thanks to our contributing authors for their excellent chapters and to Miss Candi McGonagle for her tireless efforts in readying the manuscript for publication through seemingly endless drafts.

Contents

I Cardiac Failure

Cardiac failure may be defined as the inability of the heart to pump sufficient blood to satisfy the metabolic demands of the body.[8,11,12,17–22,25] Early recognition of heart failure in the neonate is often difficult because it may be confused with other disease entities. In this chapter we are chiefly concerned with cardiac failure secondary to structural congenital heart disease, although one must keep other possibilities in mind (Table 1-1).

Table 1-1. Etiology of Cardiac Failure (In order of patient's age at onset)

Hypoplastic left heart syndrome, aortic atresia, interruption of the aortic arch
Transposition of the great vessels
Coarctation of the aorta
Ventricular septal defect
Severe pulmonary stenosis with intact ventricular septum
Total anomalous venous drainage with venous obstruction
Endocardial cushion defect
Tricuspid atresia
Patent ductus arteriosus, systemic arteriovenous fistulas
Single ventricle
Persistent truncus arteriosus
Primary myocarditis
Endocardial fibroelastosis
Cardiac arrythmias, paroxysmal atrial tachycardia, complete heart block

ETIOLOGY AND PATHOPHYSIOLOGY

The causes of cardiac failure in infancy may be classified in terms of the functional disturbance imposed on the myocardium. Causes include (1) volume overloading of the left or right ventricle as in the case of a large left-to-right shunt, valvular regurgitation,[24] or a systemic arteriovenous fistula; (2) pressure overloading of the ventricles as in left or right ventricular outflow obstruction, pulmonary valvular obstruction, coarctation of the aorta, and systemic arterial hypertension; (3) obstruction to pulmonary venous return as in

anomalous pulmonary venous drainage below the diaphragm, congenital mitral stenosis, cor triatratum, stenosis of the pulmonary veins, and the hypoplastic left heart syndrome with mitral atresia; and (4) primary disorders of cardiac muscle function caused by inflammatory processes, anomalous coronary perfusion or other causes of ischemia, metabolic derangements, or fibroelastosis.

Pulmonary congestion may result from left ventricular failure secondary to severe coarctation of the aorta, aortic stenosis, other lesions which obstruct left heart outflow, aortic valve incompetence, or primary myocardial disease. Obstruction to pulmonary venous return may also cause pulmonary congestion.

Hyperdynamic engorgement with cardiac failure can also result from excessive left-to-right shunting as in large ventricular septal defect or patent ductus arteriosus. Left atrial and pulmonary venous hypertension is also present in these forms of "high-output" cardiac failure.

Pulmonary vascular resistance is elevated in the newborn as a result of medial thickening. This normally begins to regress at 4 weeks of age and is normal (20 percent of systemic vascular resistance) by 8 weeks of age. These involutional changes toward normal occur slowly or not at all in some infants with excessive left-to-right shunts or pulmonary venous hypertension from other of the above causes.

From the physiologic standpoint cardiac failure represents an inadequate cardiac output in relation to the body needs (myocardial component) and an accumulation of abnormal amounts of blood in the pulmonary and systemic venous system (congestive component). A rise of ventricular end-diastolic and filling (atrial mean) pressures is the first hemodynamic manifestation of ventricular failure. The end-diastolic pressure is usually abnormally elevated in the presence of congestive failure. It may also be elevated when marked ventricular hypertrophy is present, reflecting decreased diastolic compliance.[2,4] In this instance the elevation is not necessarily an expression of impaired ventricular contractility.[3]

As ventricular function becomes impaired, increased diastolic fiber length (cardiac dilation) may allow the heart to maintain an adequate stroke volume.[23,26,27] Tachycardia ensues, maintaining cardiac output in the face of a falling stroke volume as myocardial failure progresses.[9] When cardiac output can no longer be maintained by these mechanisms, the arteriovenous oxygen difference widens, extracting more oxygen per liter of blood flow. These are rapid adjustments. Myocardial hypertrophy is a more gradual compensatory adjustment and is most marked when there is systolic or afterloading, as in severe semilunar valve stenosis or in systemic or pulmonary arterial hypertension.

Pulmonary vascular congestion from any of the above causes results in a decreased pulmonary compliance.[5,29] This stimulates receptors at various cardiac, respiratory, and central nervous system sites and results in tachypnea, hyperventilation, and respiratory alkalosis.[1,16] The work of breathing is markedly increased. With time the severely ill infant in cardiac failure may be unable to maintain this respiratory work level. Hypoventilation, hypoxemia, hypercarbia, and respiratory acidosis ensue.[28] In the final stages transudation of edema fluid into the alveoli and tracheobronchial tree creates the picture of overt pulmonary edema. Superimposed pulmonary infection is common, and may further impair cardiorespiratory function by consolidating areas of lung parenchyma and by the increase of metabolic requirements imposed by fever and sepsis.[6,14]

Certain types of malformations of the heart predispose to the development of cardiac failure in characteristic age periods. (Table 1-1) During the first week of life the most common cause of cardiac failure is the hypoplastic left heart syndrome with aortic atresia. Transposition of the great vessels is second. Both produce signs of pulmonary or systemic congestion and severe hypoxemia. From 1 to 4 weeks of age coarctation of the aorta and transposition of the great vessels are the leading causes of congestive heart failure, the former because the left ventricle cannot maintain systemic perfusion in the presence of a severe left ventricular afterload, and the latter as a result of hypoxemia and increased pulmonary blood flow.[14] Between 1 and 3 months of age left-to-right shunts, as in ventricular septal defect, patent ductus arteriosus, and endocardial cushion defects are important causes of failure, as pulmonary vascular resistance decreases and left-to-right shunting increases, overloading the left ventricle. Frequently more than one defect is found in the infant who develops heart failure in the early weeks of life.

Certain combinations of lesions produce systolic as well as diastolic overloading of both ventricles. This is particularly true of the frequently seen combination of coarctation of the aorta, patent ductus arteriosus, and/or ventricular septal defect. A bicuspid aortic valve is present in many, and in some there is a significant transvalvular pressure gradient (aortic stenosis). These combined hemodynamic liabilities are refractory to medical treatment alone and usually require combined medical and surgical therapy.

Clinical Features

Dyspnea and tachypnea are frequently noted in cardiac failure in infancy.[7,10,14,17,19–22] The respiratory rate may be 50 breaths per minute or higher. Dyspnea may be observed at feedings, when the

process of sucking and swallowing leads to breathlessness. This observation in an infant who is not gaining weight adequately should suggest the possibility of cardiac failure.[15]

The liver usually becomes enlarged early in an infant with cardiac failure and the edge may be rounded.

Tachycardia with rates of 150 to 200 beats per minute is frequently found.

Gallop rhythm in infancy is usually indicative of cardiac failure. It is probably to be explained on the basis of distention of the ventricles in the rapid filling phase of diastole, the same explanation given for the production of the third heart sound. It should be pointed out, however, that the physiologic third heart sound is not heard well in infancy. A well-heard third heart sound is therefore considered pathologic and in most forms of heart disease is an indication of incipient cardiac failure.

In left ventricular failure rales may be audible at the bases in the late stages. The alternative possibility of pulmonary infection must be considered, especially when pulmonary vascular markings are increased. In this case it is difficult to tell whether the rales are due to infection, heart failure, or both.

SIGNIFICANCE

Congestive heart failure is sufficient reason for cardiac catheterization in infancy.[13] No patient should be considered too ill to undergo cardiac catheterization unless he or she is moribund. If the differential diagnosis includes a potentially correctable lesion, cardiac catheterization is indicated and the study is not to be considered complete until such a lesion has been found or excluded. Obviously the congestive heart failure, pneumonia, anemia, and acidosis must be remedied to the optimal degree before cardiac catheterization is undertaken. If the infant is not improving on medical management and surgical therapy is possible, the risks of study and surgery are justified, since in such infants the ultimate prognosis is poor unless aggressive medical and surgical therapy is carried out early.

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2 Cyanosis

Whether of congenital or acquired origin there are four basic pathophysiologic types of cardiovascular abnormalities:

1. Admixture of unoxygenated blood in the left side of the heart or aorta leading to arterial oxygen unsaturation
2. Volume overloading in which an excess amount of blood is pumped by either or both ventricles
3. Obstruction to flow entering or leaving either ventricle
4. Abnormal ventricular contraction and/or relaxation

In congenital cardiac disorders more than one of these abnormalities are usually present simultaneously. Right ventricular outflow obstruction and admixture of unoxygenated blood to the left heart (right-to-left shunt) are found in the tetralogy of Fallot and in pulmonary atresia with intact ventricular septum. Right-to-left shunting and inflow obstruction occur in tricuspid atresia. In transposition of the great vessels volume overloading plus admixture of unoxygenated blood are noted, and severe aortic stenosis is frequently associated with myocardial ischemia or fibroelastosis which produces altered contraction and compliance of the ventricle. In this chapter we will attempt to examine factors related to arterial oxygen unsaturation and cyanosis, but it must be kept in mind that almost every lesion is accompanied by other pathophysiologic liabilities and that the clinical features are due to hypoxemia plus the associated physiologic abnormalities imposed by the altered cardiac anatomy.

Blood in the pulmonary veins, left side of the heart, and systemic arteries normally has an oxygen saturation of 95 percent or more. At least 5 gm of reduced hemoglobin per 100 ml of blood must be present in the capillaries before clinical cyanosis is evident.[8,9] Therefore cyanosis is related not only to the degree of arterial unsaturation but to the absolute amount of circulating hemoglobin (i.e., the polycythemic patient has a greater tendency to appear cyanotic).[13]

Cyanosis may be "central," due to true arterial oxygen unsaturation, as may occur with right-to-left intracardiac shunting or severe pulmonary disease with intrapulmonary shunting or hypoventila-

tion. Cyanosis may also be "peripheral," associated with a normal arterial oxygen saturation and slow blood flow in the subpapillary venous plexus of the skin and nail beds; this cyanosis is due to peripheral vasoconstriction secondary to cold extremities, reduced cardiac output, or both. Since blood flow tends to be lowest in the extremities, peripheral cyanosis is most apparent in the nail beds and less obvious in the mucous membranes.

When arterial oxygen unsaturation is due to abnormal ventilation/perfusion relations in the lung (pulmonary shunting) or to hypoventilation, the arterial oxygen saturation may be returned to normal by having the patient breathe 100 percent oxygen. When unsaturation results from right-to-left intracardiac shunting, the saturation will remain low regardless of the oxygen content of inspired air because the shunted blood is never in contact with the alveolar-capillary interface in the lung.[6,7] In such infants breathing 100 percent oxygen may increase only the amount of dissolved oxygen in the plasma.

The pathologic mechanisms of central cyanosis are the following:

1. Congenital malformations of the heart with right-to-left shunting. In most cyanotic infants there is not only admixture of unsaturated right heart blood to the left heart chambers, but almost invariably reduced pulmonary blood flow as well. Cyanosis is minimal when pulmonary blood flow is not reduced. In such instances even though arterial oxygen unsaturation is present, so long as the saturation remains at approximately 80 percent or above polycythemia does not develop, and this further reduces the degree of clinical cyanosis by curtailing the amount of reduced hemoglobin in the capillaries.

2. Respiratory abnormalities. Infants with hypoglycemia,[1,3,11] central nervous system damage, or severe sepsis may be cyanotic because of alveolar hypoventilation. When respiratory work is increased and the infant cannot perform the work of breathing, hypoventilation, severe hypercarbia, acidosis,[5] and hypoxemia ensue. This chain of events may be due to loss of pulmonary compliance,[10] as in severe congestive heart failure, pneumonia, respiratory distress syndrome,[12] or Mikity-Wilson syndrome. Neonatal atelectasis or obstruction to the trachea or bronchi from vascular ring, foreign body, or giant pulmonary artery may similarly increase respiratory work. Congenital lobar emphysema, diaphragmatic hernia, or tension pneumothorax may reduce the lung volume enough to impair gas exchange and cause hypoxemia and often hypercarbia and acidosis.

3. Red cell or hemoglobin abnormalities. Methemoglobinemia or other disorders of oxygen transfer due to red cell or hemoglobin abnormalities may produce cyanosis.

If the cyanosis persists after the administration of 100 percent

oxygen by mask for 10 to 15 minutes, it is unlikely that the arterial desaturation is the result of a pulmonary disorder, and cardiac evaluation is indicated. The absence of a loud murmur is not necessarily reassuring: In many serious congenital malformations of the heart, such as transposition of the great vessels, severe tetralogy of Fallot, malformations associated with pulmonary atresia, severe pulmonary valvular stenosis, and total anomalous pulmonary venous drainage, especially below the diaphragm, a murmur may be absent or insignificant.

The initial diagnostic procedures include electrocardiography and posteroanterior and lateral chest roentgenography. Interpretation of pulmonary vascularity and of cardiac size and shape is of the utmost importance in the cyanotic newborn.[2] All too frequently the observer not experienced in evaluating the chest roentgenograms of neonates fails to appreciate the slight decrease in pulmonary vascularity that is often the only abnormal finding. The cyanotic infant with a small heart, slightly decreased pulmonary vascular markings, and an insignificant or absent murmur may have a serious malformation of the heart. Blood samples should be obtained (most easily by catheterization of the umbilical artery) for determination of arterial oxygen tension (or oxygen saturation) and pH.

A prospective diagnosis may be made from the aforementioned studies, but in almost every case cardiac catheterization and angiocardiography are required for a precise and definitive anatomic diagnosis of the congenital malformation of the heart. An aggressive diagnostic approach is always justified because the majority of cases are amenable to medical or surgical therapy, more often a combination of the two.

Anoxic or cyanotic spells are characterized by paroxysms of dyspnea, cyanosis, convulsions, and loss of consciousness. They occur most frequently in the tetralogy of Fallot, where they are thought to be due to a sudden fall in pulmonary blood flow secondary to spasm of infundibular muscle in the right ventricle. They may be present in any of the other anomalies in which decreased pulmonary blood flow and right-to-left shunting are present, such as pulmonary atresia with intact ventricular septum or tricuspid atresia. In both of the last two conditions the spells may be secondary to intermittent spasm of the ductus arteriosus. In either case the convulsions and loss of consciousness indicate severe cerebral hypoxemia. They herald a grim prognosis, may result in irreversible brain damage or death, and constitute a pressing indication for cardiac catheterization, in some instances balloon atrioseptostomy, and surgical treatment.

The acidosis that accompanies severe cyanosis is secondary to tissue hypoxemia (anaerobic glycolysis). In the first hours of life a low arterial oxygen tension may be tolerated quite well. The infant

may become unable to compensate, however, and his condition may deteriorate rapidly, with progressive acidosis.[14,15] Acidosis has a deleterious effect upon myocardial function and may result in more inadequate tissue perfusion, with further increase in the acidosis. A vicious cycle may thus be set in motion, and it is of extreme importance to prevent or correct acidosis expeditiously.

If a diagnosis has been established, it is often possible to carry out some form of palliation such as balloon septostomy or a systemic-pulmonary anastomosis to increase the oxygen content in the systemic circulation. This often corrects the acidosis, but judicious administration of sodium bicarbonate is usually necessary as well. Morphine is of little use in the progressively ill cyanotic infant, but is used to relieve the acute hypoxic spells experienced in, for example, the tetralogy of Fallot. When the condition of the cyanotic infant is rapidly deteriorating, morphine plays a decreasingly effective role and may be dangerous unless ventilatory assistance is available.

As will be noted in the chapters devoted to the various malformations, untreated cyanotic infants have an extremely poor prognosis, and an aggressive diagnostic approach leading to appropriately combined medical and surgical treatment offers them the only reasonable hope for survival beyond infancy. As experience accumulates both in this country and abroad, it is becoming increasingly apparent that most infants with heart malformations producing cyanosis can be salvaged by a palliative or totally corrective operation performed in the early weeks of life. Therefore an aggressive diagnostic and therapeutic regimen is warranted in view of the poor prognosis without such treatment.[4]

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