PEDIATRIC CARDIOLOGY

Summary of a Round Table Discussion

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The discussion of cardiac problems included: a consideration of functional murmurs; rheumatic fever; heart failure in infancy; and congenital cardiac malformations, with respect to indications for surgery and results.

FUNCTIONAL MURMURS

Functional or innocent murmurs are always systolic in time. Occasionally a functional murmur may be heard in the pulmonic area, and must be differentiated from an organic murmur due to a small atrial secundum defect. However, the most common functional murmur is that heard between the apex and lower left sternal border. The quality best suggests the adjective “vibratory,” but other terms frequently used are “groaning,” “musical” or “twanging string.” This murmur, which is of moderate duration, can be extremely loud and frequently is intensified by exercise.

A venous hum, which is a continuous bruit throughout systole and diastole, is a common finding that has no pathologic significance. With the patient in the erect position it is audible below the clavicles and is best heard in the neck. The diastolic component may be transmitted downwards and occasionally suggests the murmur of aortic insufficiency. However, in the case of a venous hum, the diastolic bruit can usually be obliterated by compressing the neck vessels, which cannot be done when the murmur is due to aortic insufficiency. The continuous murmur of a venous hum must on occasion be differentiated from that due to a patent ductus arteriosus, but it should always be noted that the murmur of a patent ductus is loudest when the patient is in the supine position.

RHEUMATIC FEVER

Of the major manifestations of acute rheumatic fever, carditis deserves special attention. A diagnosis of carditis is based primarily on auscultatory findings. The heart sounds are frequently of poor quality, or muffled, and are associated with conspicuous apical murmurs. The usual systolic murmur in this location is long, blowing, high-pitched and transmitted into the left axilla. In addition, a short, soft, low-pitched mid-diastolic murmur is usually heard at the apex. More severe cardiac involvement is characterized by an aortic diastolic murmur (short, high-pitched), pericarditis or congestive heart failure. In the presence of cardiac decompensation, organic murmurs due to rheumatic valvulitis are always audible. A conduction defect, manifested by an increased P-R interval on the electrocardiogram, is frequently but not necessarily present in acute rheumatic fever with significant carditis.

In the management of a patient with acute rheumatic fever, the assumption should be made that carditis and beta hemolytic streptococcal infection are present. Bed rest is recommended, with a 4-week period being an arbitrary minimum. The interpretation of bed rest is liberal, and for the average case the child may be permitted to feed himself and sit up and play when he desires. However, for the patient with heart failure, heavy sedation with morphine is indicated for several days. Ambulation is begun after clinical and laboratory signs (temp-
The majority of infants who develop cardiac decompensation have a congenital anomaly of the heart associated with an increased blood flow to the lungs. The most common lesions producing large left-to-right shunts associated with heart failure in infancy are ventricular septal defect and patent ductus arteriosus. Coarctation of the aorta should also be considered as a possible cause of congestive failure, in which case it is usually accompanied by other defects resulting in an increased pulmonary blood flow. In the cyanotic group of infants, the most frequent malformation producing heart failure is transposition of the great vessels.

The most conspicuous signs of congestive failure are tachycardia, tachypnea and hepatomegaly. Infants frequently have evidence of pneumonia by the time they develop cardiac decompensation. Peripheral edema is uncommon.

In the management of infants with heart failure, digitalis, oxygen and antibiotics are indicated. In addition, morphine (1 mg/5 kg) may be employed for restlessness, and diuretics may be helpful. Low-salt milk may be used, but caution should be exercised to avoid hyponatremia if it is administered over a long period of time.

As regards the choice of digitalis preparation, familiarity with one product is strongly recommended. A suitable product that has the desirable qualities of fairly rapid utilization and excretion is digoxin (Lanoxin\textsuperscript{\textregistered}). Usually, digitalization is accomplished by the intramuscular route, and then for daily maintenance the oral preparation is indicated when the infant's condition has become stabilized.

There are tremendous individual variations in the digitalis requirement, so that the dosage schedule may need to be adjusted for a particular patient. An estimated digitalizing dose of Lanoxin\textsuperscript{\textregistered} for infants in the first year of life is 0.075 mg/kg, usually divided into three or four doses and given in a 24- to 36-hour period. An electrocardiogram should be obtained before the final dose is administered to ascertain whether any conduction defect or arrhythmia has
CONGENITAL CARDIAC MALFORMATIONS—INDICATIONS FOR SURGERY AND RESULTS

Patent Ductus Arteriosus

Surgery for uncomplicated cases of patent ductus arteriosus (normal heart size without pulmonary hypertension) is recommended at approximately 4 years of age. However, if an infant develops cardiac failure, and cardiac catheterization demonstrates the presence of a patent ductus, surgery should be undertaken immediately. Despite a higher surgical risk, those patients with pulmonary hypertension, but who have a predominantly left-to-right shunt from aorta to pulmonary artery, should have operative closure of the ductus. Patients usually considered inoperable are those with marked pulmonary hypertension resulting in a significant right-to-left shunt through the patent ductus.

This lesion is successfully cured by surgical ligation and division, with a mortality of approximately 1%. Those cases with patent ductus who need surgery in infancy, or who have pulmonary hypertension noted at later age, represent an increased operative risk, with a mortality of about 10%.

Coarctation of the Aorta

All patients with this malformation should have surgical repair, preferably between the ages of 8 and 12 years. For the infant with coarctation who develops congestive heart failure, surgery is indicated if medical management fails to maintain an adequate state of compensation.

The operative mortality varies between 2 and 9%. It is to be noted that in 5 to 15% of cases with coarctation, a patent ductus coexists. The latter is usually repaired at the time of surgery for coarctation.

Aortic Stenosis

At the present time, the most important indication for surgery for aortic stenosis is the development of ST- and T-wave changes in the left precardial leads of the electrocardiogram, which reflect marked left ventricular hypertrophy or strain. Before surgery is performed, left ventricular catheterization should be undertaken to establish the gradient across the aortic valve. Concomitant aortic insufficiency does not contraindicate surgery.

Surgical intervention in this lesion has recently produced good results by a direct approach to the valve, with the aid of means for extracorporeal circulation.

Atrial Septal Defect

Most patients with atrial (secundum) defects do well in infancy and childhood, but may get into difficulty during adolescence or adult life. Indications for surgery include any or all of the following: 1) roentgenographic evidence of cardiomegaly; 2) symptoms of fatigue, diminished exercise tolerance, or failure to thrive; or 3) clinical evidence of pulmonary hypertension, manifested by a loud, snapping, split, pulmonic second sound.

Excellent surgical repair of this lesion is achieved either by closure under direct vision (using extracorporeal circulation or hypothermia) or by a closed atrial-well technique. The surgical mortality averages between 2 and 9%.

Endocardial Cushion Defect

This entity comprises a combination of defects sometimes referred to as atrioventricular canal, or ostium primum syndrome. In addition to the persistence of the ostium primum (low atrial defect) there are frequently associated abnormalities of the atrioventricular valves (a cleft in a leaflet of either the mitral or tricuspid), and a ventricular septal defect. The majority of patients with this malformation develop cardiomegaly and pulmonary hypertension during childhood, hence surgical repair would be
desirable. The indication for operative intervention will depend upon the changing surgical results.

Surgery for endocardial cushion defects must be undertaken by direct vision, using extracorporeal circulation. The mortality for the "partial" form (ostium primum plus cleft mitral valve) is estimated to be between 10 and 12%. However, if there is "complete" involvement of the atrial and ventricular septa and atrioventricular valves, the mortality varies between 40 and 60%.

**Ventricular Septal Defect**

The optimal candidate for surgical correction of a defect of the ventricular septum (utilizing extracorporeal circulation) is the child or young adult with cardiomegaly, in whom the systolic pressure in the pulmonary artery is in the moderate range, i.e., 40-50 mm. The asymptomatic patient without cardiac enlargement should probably not be considered for surgery at this time.

The present open-heart repair of ventricular septal defects has a mortality of about 10% in patients who do not have severe pulmonary hypertension. The risk increases roughly proportionate to the increased pulmonary vascular resistance and elevation of pulmonary artery pressure. Postoperative complications, such as complete heart block, may occasionally ensue; this is dangerous and difficult to manage.

**Tetralogy of Fallot**

The present recommendation for surgery in the tetralogy of Fallot is that a palliative anastomotic procedure be carried out in the infant who is suffering from paroxysmal dyspnea or syncope. It is the opinion of many surgeons that the procedure of Blalock (subclavian artery to pulmonary artery) should be attempted rather than that of Potts (aorta to pulmonary artery), because it may be easier to undo the former when a definitive open-heart repair can be carried out at a later date. If the infant with a tetralogy of Fallot is doing relatively well, he should be observed with the expectation of undergoing an elective open-heart repair when he is approximately 5 years of age or older.

The results of extensive surgical experience indicate that with either a Blalock or Potts anastomosis, the mortality in the infant under 18 months of age is about 30%, whereas in the patients above this age the mortality is approximately 5%. In the more recent, yet limited, experience of clinics utilizing extracorporeal circulation to carry out an anatomically corrective procedure, the anticipated risk in the childhood age range is 20%.

As regards the follow-up of patients who underwent Blalock anastomoses, Dr. Helen Taussig has reported that in a series studied 5 to 8 years after surgery, 70% of the group had maintained good post-operative results. However, some of the patients had required a second Blalock procedure. It is not known what the long-term results will be in those patients with tetralogy of Fallot who have recently undergone open-heart surgery, in most of whom prosthetic materials were incorporated into the corrective procedure.

**Pulmonic Stenosis**

Candidates for surgical repair of pulmonic stenosis, who have an intact ventricular septum, are arbitrarily selected on the basis of the systolic pressure in the right ventricle, determined by cardiac catheterization. Those patients in whom the pressure in the right ventricle is elevated to 100 mm or more should have immediate surgery. The patients with pressure in the right ventricle below 80 mm may be observed annually; and, if they are asymptomatic and do not have significant roentgenographic or electrocardiographic evidence of progression, surgery then may be carried out electively.

Utilizing the Brock procedure for closed valvulotomy, the anticipated surgical risk is approximately 1 to 2%. The patient with valvular pulmonic stenosis and, in addition, evidence of an atrial septal defect, should have surgical repair of both lesions by open-heart techniques.
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