pertension, however, in ours and other people’s experience is outweighed by the advantage of operative closure of the defect at a time optimal for the surgeon.

Data available on endocardial cushion defect are scanty indeed and profiles of natural history are not available. Of the entire group, it would seem that the defect is partial (ostium primum atrial septal defect with or without a cleft in the mitral or tricuspid valve) in 75% and complete (atrioventricular canal) in 25%. Near normal life expectancy can probably only be anticipated in those with small partial defects. Congestive heart failure is the usual cause of death and may occur under 2 years in those with complete canals. Bacterial endocarditis and pulmonary vascular obstructive disease are rarely encountered in patients with endocardial cushion defects.

The natural history of aortic stenosis is also incomplete. Operation on the aortic valve or the subvalvar area is indeed safe. However, we do not have any significant information on a large number of patients with any long-term postoperative follow-up. We have to know who are the patients in whom operation may be safely postponed to a time when the effect of surgery on those with maximal aortic obstruction may be assessed from the point of view of (1) reduction of gradient, (2) persistence of reduction, (3) reversibility of electrocardiographic changes, and (4) creation of aortic regurgitation. In reviewing statistics on mortality it is apparent that the nonsurgical group with aortic stenosis is probably an unnaturally low number since most of the severe cases are operated upon and they would have been the ones who would have died without operation. This is a good example of the difficulty in assessing problems of natural history accurately.

CURRENT CONCEPTS IN PEDIATRIC CARDIOVASCULAR SURGERY

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Among the many interesting congenital cardiac malformations amenable to surgery today, four will be discussed, namely, coarctation of the aorta, ventricular septal defect, Tetralogy of Fallot, and transposition of the great vessels. Recommendations for surgical therapy are based on the experience at the Mayo Clinic.

Coarctation of the aorta continues to be most satisfactorily treated by operation and the hospital mortality risk is extremely low. In general, the optimal age for surgery is about 10 to 12 years. Unusually severe systemic hypertension or cardiac enlargement constitute an indication for somewhat earlier surgery. When infants with isolated coarctation of the aorta show severe cardiac failure, and this certainly occurs although uncommonly, operation should be advised at that age. The risk even then is very low, but a few cases may require reoperation at age 12 because of failure of adequate growth of the anastomosis.

The indications for repair of ventricular septal defect must take into account the estimated size of the lesion and its hemodynamic effects. Patients with small ventricular septal defects who have normalized hearts and normal electrocardiograms, that is, those without clinical evidence of significant hemodynamic derangement, are not advised by us to undergo repair of the defect at present. Patients with ventricular septal defect and mild or moderate pulmonary artery hypertension (that is, patients whose pulmonary artery pressure is less than 75% of systemic artery pressure) can be operated upon at essentially no hospital risk. Operation is clearly advisable in all of these patients. Patients with ventricular septal defect who have severe pulmonary artery hypertension associated with a large
pulmonary blood flow (and thus with only mild elevation of pulmonary vascular resistance) can be operated upon with a low hospital mortality rate, in the neighborhood of 5%. These patients should clearly be subjected to surgery and the long-term results are excellent. The patients with severe pulmonary artery hypertension but with moderate pulmonary vascular disease should also be advised to have operation for the risk of surgery under these circumstances is in the neighborhood of 10%. The long-term results in these cases seem acceptably good although such patients continue to have moderate elevation of pulmonary artery pressure and pulmonary vascular resistance. Patients with ventricular septal defect who have severe pulmonary artery hypertension and severe pulmonary vascular disease, indicated by marked elevation of pulmonary vascular resistance, are at the present time difficult of decision. The hospital mortality rate is now in the neighborhood of 25% but there is considerable uncertainty as to the long-term fate of these patients after repair. The data so far suggests that the pulmonary vascular resistance usually does not fall in the months after repair. There is some reason to suspect that these patients may live less long with their defect closed than with it open. In those patients with even more severe elevation of pulmonary vascular resistance in whom the ventricular shunt is predominantly right to left, repair of the ventricular septal defect is clearly contraindicated.

Though the optimal age for repair of ventricular septal defect is 5 to 6 years there are indications for repair at an earlier age. These include chronic heart failure, marked retardation of growth and progressing pulmonary vascular disease. The latter may occur in some patients between the ages of 6 months and 3 years. When operation is indicated in a small patient over 6 months of age, open-heart repair of the defect is to be advised as the risks are low and the results good. Pulmonary artery banding should be reserved for patients under 6 months of age and in actual practice is rarely necessary.

The results of open repair for Tetralogy of Fallot, viewed now 9 years after its beginning, continue to be excellent. In some ways they are the most outstandingly good results seen in intracardiac surgery. The ideal age for open operation for the Tetralogy of Fallot is about 6 years. Younger patients who are doing reasonably well may not be operated upon until this age. Infants and small children with serious symptoms and progressing polycythemia and disability are operated upon by us using an anastomotic operation; the open operation should then be done at about the age of 6 years. The risk of open intracardiac repair has been acceptably low in our institution in the last three years, being about 10% for cyanotic patients with Tetralogy of Fallot. In at least 85% of these patients in the last year an outflow patch over the right ventricle has not been required since we seem to have learned to relieve the pulmonary obstruction in most even deeply cyanotic patients without the need for this.

Corrective open-heart surgery for transposition of the great vessels should be continued utilizing the Senning procedure. It is recommended for all patients with transposition over the age of about 10 months except those whose pulmonary vascular disease is considered severe. The hospital mortality rate continues to approximate 50%. Under the age of 10 months an atrial septal defect is created as a palliative procedure by the Blalock-Hanlon technique and open operation, it is hoped, can be done approximately a year or two thereafter.
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