Ventricular Septal Defect

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Now that open-heart surgery with extracorporeal circulation is being successfully used in babies weighing as little as 4 kg., it might be asked why all patients with an isolated ventricular septal defect do not have the defect surgically closed during the first year or two of life. In trying to answer this question one is immediately confronted with the tremendously wide clinical spectrum of this anomaly, which ranges from babies dying in heart failure to elderly persons free of symptoms, and which includes those who become cyanosed from pulmonary hypertension at an early age. The only feature that these patients have in common is the presence of a hole in the ventricular septum; they otherwise appear to be suffering from different diseases. It is now clear that the clinical course and management of ventricular septal defect depends largely on the size of the defect and on the presence or absence of pulmonary vascular disease. Other important factors are the spontaneous closure of some defects, and the development in some patients of progressive infundibular stenosis (Gasul et al., 1957).

The management is therefore best discussed from the point of view of the different clinical syndromes to which ventricular septal defect gives rise.

In infants (defined as a child under the age of 1 year) it is heart failure which is the chief indication for treatment, and if medical management fails then an operation is essential for survival. The Muller–Dammann operation (1952) of constricting or banding the pulmonary trunk in order to reduce the pulmonary hypertension and excessive pulmonary blood flow has become accepted as the standard palliative procedure, and in patients without other cardiac anomalies the operative mortality is less than 5 per cent (E. Aberdeen and D. J. Waterston, 1967, personal communication). This procedure not only relieves heart failure, but it permits normal development of the child so that closure of the defect and removal of the constriction can be done later. Therefore, it is especially in these infants with persistent heart failure that the question of surgical closure of the defect in infancy arises, in order to reduce the number of operations required. The difficulties and objections to such a course may be summarized as follows. (1) Extracorporeal circulation is undoubtedly a much more skilled procedure in the infant than in older children and therefore potentially more hazardous. (2) There are surgical technical problems related to the small size of the heart. The limited size of the ventriculotomy makes access to the defect difficult, and the bundle of His is much closer to the septal defect, thus increasing the risk of heart block. (3) Post-operative management, especially with regard to fluid balance and the treatment of respiratory complications, is more difficult. However, in spite of these problems several centres have been closing ventricular septal defects in infancy, and preliminary unpublished results seem to indicate that it is a reasonably safe procedure over the age of 6 months. Under 6 months the mortality for direct closure is high, and since the majority of patients who get into trouble with ventricular septal defect do so before 6 months, banding is probably the treatment of choice for the infant with heart failure. Over 6 months, direct closure of the troublesome ventricular septal defect should be considered but only at a centre with the highest possible degree of expertise, performing a large number of open and closed heart operations each week. It may be asked why patients over 6 months of age are having their defects closed when it has not been found necessary to perform a banding operation in this age-group to treat persistent heart failure. This brings us on to a slightly different criterion for surgical treatment in infancy. Certain babies, while not in persistent overt failure, are nevertheless in a considerable degree of haemodynamic trouble because of severe pulmonary hypertension with a left-to-right shunt of variable magnitude. It is probable that most of these
patients will develop progressive pulmonary vascular disease in the first few years of life, which will eventually become irreversible and render them inoperable. It is to prevent this lethal complication that surgery is being applied at a young age. If subsequent experience confirms that primary closure of the defect may be accomplished at a low mortality, then this is clearly the treatment of choice. Banding the pulmonary artery will accomplish the same end, but as already noted a second operation will always be required. Although it is traditionally taught that some patients with a right-to-left shunt due to pulmonary vascular disease have this condition from birth, it is now being doubted whether this is true and it may be that early operation will always be feasible in this group.

In a different category are those patients who are reasonably well in infancy and who do not require urgent treatment early in life. Factors that will influence a decision whether or not to operate at say 2 to 4 years of age include (i) the possibility of spontaneous closure of the defect, (ii) the possibility that progressive pulmonary vascular disease will occur and, (iii) one's opinion on the long-term prognosis of unoperated ventricular septal defect. Again the size of the defect and the presence or absence of pulmonary hypertension are the important factors. Further, it is unfortunately true that some large cardiac units have such a long waiting list for operations that they have to be deferred.

Spontaneous closure of a ventricular septal defect is now a well-documented event. It is commoner in patients with initially small defects, but occasionally occurs in those with large defects and has even been found in a patient submitted to a banding operation in infancy. Over the age of 5 years spontaneous closure is less common. Partial closure of a defect with a reduction in pulmonary artery pressure and blood flow is also clearly established and may be more common than spontaneous complete closure. It has been attributed to the defect failing to increase in size as the heart grows (Gasul, Arcilla, and Lev, 1966). Unfortunately not all defects behave in this way, as is made clear by the finding in some adult patients of defects that are larger than was the size of the entire heart at birth. However, even if less than 50 per cent of patients get a spontaneous reduction in the size of the defect, it is clearly an event worth waiting for, at any rate up to the age of 5 years.

If, however, there is the possibility that severe and irreversible changes in the pulmonary vascular bed will develop during this waiting period, then it is wrong to deny early operation. In young patients with a considerably raised pulmonary vascular resistance—as judged by the clinical and hemodynamic findings—it is unrealistic to expect many to achieve a spontaneous and worthwhile reduction in the severity of their pulmonary hypertension, and they should have the defect closed before the age of 5; the earlier the better providing the surgical team is expert at handling operative and post-operative problems in small children. In those with only a moderately raised resistance the chances of progressive changes developing in the pulmonary vessels at this age are less though they do exist. Such patients should be carefully followed and if there is doubt from the clinical and electrocardiographic findings as to the state of the pulmonary circulation they may require serial right heart catheterization.

One might mention here the problem of resolution of pulmonary hypertension following operation. The pulmonary artery pressure falls immediately after operation because the pulmonary blood flow is reduced by removal of the left-to-right shunt. However, one is more interested in the question as to whether pre-existing changes in the pulmonary vasculature regress later on. Hollman (1963) found evidence for this in only a minority of patients, who were re-catheterized after operation, and noted that acetylcholine rarely produced a selective fall in pulmonary artery pressure. Lillehei et al. (1964) were more optimistic about the late hemodynamic results in patients with severe pre-operative pulmonary hypertension, and made the important comment that an improvement in “pulmonary vascular status” was perhaps more pronounced in younger patients. This evidence indicates that any patient with ventricular septal defect with a pulmonary artery pressure of, for example, 60 mm Hg or over, may be best managed by operation within the first two years of life.

We are left then with a consideration of what action to take in the older child, over the age of 5 years, with a small or moderate sized defect, and either a slight or no increase in the pulmonary vascular resistance. It is known that some of these patients do have a spontaneous decrease in the size of the defect as late as early adult life, and it is generally agreed that those with a small defect have little or no risk of developing pulmonary hypertension. The risk of developing bacterial endocarditis has been cited as a reason for closing septal defects, but Shah et al. (1966) dispute this, quoting the incidence as only 1 in 500 patient years. They also point out that endocarditis may develop as a complication of surgical closure of the defect. Furthermore, Simmons, Moller, and Edwards (1966) have suggested that endocarditis may lead to spontaneous closure by adherence of a scarred septal leaflet of the tricuspid valve to the defect. However, endocarditis can be a difficult infection to eradicate, and it carries the hazard of late heart valve
perforation even if treatment is promptly instituted. In other words, we are left with the problem of whether to perform an operation of slight but definite risk to protect the patient against unpredictable hazards. One might even say unknown hazards, because the really long-term prognosis of patients with a small or moderate sized septal defect is still uncertain. It is not entirely clear whether the apparent rarity of ventricular septal defect in middle age is due to spontaneous closure of the defect, or death due to complications. A further "complication" of ventricular septal defect, well known outside the pediatric age-group, is the great difficulty that a patient with such a defect has of obtaining employment. Many large commercial firms refuse to employ applicants with a heart lesion, even if the cardiologist certifies that they probably have a normal expectation of life. This has led some very experienced cardiac units to advise operations for men with small defects and to advise against it for women. In women they are also partly influenced by the possibility of a large and unsightly chest scar. Clearly no hard and fast dividing line can be laid down, but in general I believe that a patient with a ventricular septal defect which is giving rise to a moderate increase in pulmonary artery pressure, with a pulmonary blood flow twice, or more, that of the systemic flow, should be submitted to operation providing it can be carried out in a unit where the operative mortality for such patients is under 2 per cent. Those patients with a genuinely small defect—no pulmonary hypertension and only a small increase in pulmonary blood flow—should be left alone. In this connexion it is worth remembering that the clinical, cardiographic, and radiological evidence of the size of the defect are at least as valuable as the catheterization data.

This review has been concerned only with those patients who have an isolated ventricular septal defect. When associated lesions such as patent ductus arteriosus, coarctation of the aorta, atrial septal defect, or mitral valve disease are considered, then the question of an operation has to take into account the relative severity of the two or more lesions that may be present.

REFERENCES