Natural History of Ventricular Septal Defect

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PALLIATIVE or corrective surgery of ventricular septal defect has been almost established. But there are still many questions to be answered, particularly in deciding how to select patients for surgery. Therefore more informations about the natural history of ventricular septal defect are needed.

Materials

829 patients with isolated VSD who visited the Department of Pediatric Cardiology, Kyushu University Hospital in 1962–1970 were studied. 449 patients (54.2%) visited during infancy (under 1 year of age) and most of them have been followed up over 2 years. 405 cases were catheterized.

Spontaneous Closure and Decrease in Size of Defect

Closure of the defect is defined as the disappearance of all clinical signs of VSD. It is very difficult to work out the true incidence of spontaneous closure of VSD. So there are a variety of figures given in the literature of the last 10 years on the incidence of spontaneous closure of VSD^{1,2}. Nevertheless, several large clinical series show a high incidence of spontaneous closure such as 25% (Li *et al.*³) and 41% (Hoffman and Rudolph⁴).

In our unselected cases the incidence of spontaneous closure of the defect appeared to be as high as 22.8% in all VSD and 28.1% in small VSD. The expected rate of spontaneous closure of VSD at different age was calculated by the

Fig.1. Expected rate of spontaneous closure of ventricular septal defect at different age.

method of the life table. (Fig. 1) For instance, 10% of the patients with small VSD at 5 years of age have possibility of spontaneous closure in future. Spontaneous closure occurred chiefly in the first 5 years (86.4%) and was uncommon in the order age. Most defects that closed spontaneously were small initially (93.2%).

Phonocardiograms were recorded several times in 31 patients until the defect completely closed (Fig. 2). The harsh pansystolic murmur changed in three ways at first, then a faint band-like pansystolic murmur, finally early ejection systolic murmur and disappeared. It might show that the defect had been gradually smaller in size and finally closed.

Decrease in size of the defect can be defined as significant reduction of pulmonary to systemic flow ratio and right ventricular systolic pressure by cardiac catheterization or clinical investigation. It occurred in 20% of large VSD and in

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Fig.2. Various patterns of phonocardiogram in spontaneously closed VSD.

Fig.3. Hemodynamic changes in ventricular septal defect with acquired infundibular stenosis.

27.4% of moderate VSD, and chiefly in the first 2 years of age. So many infants with large or moderate VSD had been improved clinically and hemodynamically after 1 year of age.

Acquired Infundibular Stenosis

In 1957, Gasul *et al*⁵ first described the development of infundibular stenosis in a patient

who had previously had an isolated VSD. Keith *et al*⁶ reported that this change occurred in 6 to 11 per cent of the cases who had a pulmonary to systemic flow ratio greater than 2:1.

In our series there were 5 cases of VSD with acquired infundibular stenosis confirmed by subsequent catheterization. In all cases a left to

Fig.4. Onset of congestive heart failure and pulmonary vascular resistance in large ventricular septal defect.

right shunt and pulmonary artery pressure had been reduced, and in one case a right to left shunt had been caused (Fig. 3). The frequency and mechanism of this change are still uncertain, but operated cases had a fibrous infundibular stenosis or an anomalous muscle bundle in the right ventricle.

Pulmonary Vascular Resistance

One of the most important complication of VSD is the development of progressive increase in pulmonary vascular resistance. And it is very interesting and important problem when and how to develop the progressive pulmonary vascular disease⁷ Fig.4 shows pulmonary to systemic resistance ratio (TPR/TSR) in large VSD at different age. Infants with large VSD before 1 month of age had high pulmonary vascular resistance, but in 1 to 4 months all cases with large VSD had lower pulmonary vascular resistance. Decrease in pulmonary vascular resistance in large VSD in newborn period may be slower than physiological decrease in pulmonary vascular resistance in normal newborn.⁸ After 5 months of age many infants with large VSD had still low or slightly elevated pulmonary vascular resistance, but small number of cases had higher pulmonary vascular resistance with increasing age. Fig.6 shows the changes in pulmonary to systemic pressure ratio with increasing age by subsequent cardiac catheterisation. In many in-

fants with large VSD pulmonary to systemic pressure ratio had declined after 1 month of age. It may be explained by the decrease of pulmonary blood flow due to smaller in size of the defect or development of infundibular stenosis.

But small number of patients with large VSD had increased pulmonary artery pressure with progressive pulmonary vascular resistance after 3 months of age. In moderate and small VSD pulmonary artery pressure had almost not changed or slightly declined. Patients with Eisenmenger's Syndrome were very rare and seen in only 4 cases. The risk of surgery in patients with pulmonary vascular disease was high as 50%. Careful attention must be paid to the fact that there are some patients with markedly elevated pulmonary vascular resistance in late infancy or 1 year of age, who may have the indication of surgical closure in infancy.

Congestive Heart Failure

The development of left-to-right shunt and increased pulmonary blood flow is directly dependent on the decrease in pulmonary vascular resistance after birth. The major decline in pulmonary resistance occurs within the 2–3 weeks after birth. (Rudolph⁸). Congestive heart failure was first noted in 3–12 weeks of age in large VSD. At this period pulmonary to systemic resistance ratio was decreased and pulmonary to systemic flow ratio was markedly increased in all

Fig. 5. Pulmonary to systemic flow ratio in large ventricular septal defect.

Fig. 6. Changes of pulmonary to systemic pressure ratio with increasing age in ventricular septal defect.

Fig. 7. Changes of pulmonary to systemic flow ratio with increasing age in ventricular septal defect.

large VSD (Fig. 4, Fig. 5). But pulmonary to systemic flow ratio in great number of large VSD had decreased after 6 months of age and improved clinically, because of smaller in size of the defect, acquired infundibular stenosis or increased pulmonary vascular resistance.

Deaths

In our 829 patients 9 cases died without

surgery. 6 cases died before 6 months of age, who had severe congestive heart failure with large left-to-right shunt and pulmonary complication such as pneumonia, lung bleeding and atelectasis. The rest of cases died of pneumonia with or without heart failure.

Association of Aortic Regurgitation

Association of aortic regurgitation appeared in

11 cases (1.3%). The mechanism or time of this development is still uncertain, but it is seldom detected in infants.⁹ The murmur of aortic regurgitation had been recognized after 5 years of age in our cases. All cases might seem to be small hemodynamically, but operated cases had a large supracristal VSD and prolapsed right coronary cusp of aortic valve.

Subacute Bacterial Endocarditis

The earlier data by Gelfman and Levine¹⁰ on the incidence of bacterial endocarditis shows high incidence of 43%. More recent papers reported that it was uncommon in VSD.^{11,12} The incidence of bacterial endocarditis in our series appeared to be low as 0.72% or 1 in 556 patient years. It occurred in moderate or small VSD after 1 year of age. All of them were survived by chemotherapy.

Medical Management and Surgical Indication of Ventricular Septal Defect

From this study on natural history of VSD we suggest that many infants with VSD should be treated conservatively in the hope that spontaneous closure or decrease in size of the defect will occur. Almost infants with congestive heart failure can be controlled medically. But a small number of infants, who have a huge left-to-right shunt and severe congestive heart failure which cannot be well controlled, has the surgical indication of the pulmonary artery banding or surgical closure of the defect in early infancy.

Patients with large VSD or congestive heart failure should be catheterized in infancy. If the infant has any clinical evidence of a rise in pulmonary vascular resistance, the recatheterization should be done before 1 year of age and the surgical closure of the defect may be needed at this age. Almost patients should be followed up with the hope of spontaneous improvement. In 3-6 years of age they should be put on the examination to decide the surgical indication. Patients with large or moderate sized VSD need the surgical closure of the defect before school age. Patients who have the VSD associated with an aortic regurgitation or an acquired infundibular stenosis have a surgical indication when the

diagnosis is established.

Subacute bacterial endocarditis is an infrequent occurrence but should be prevented in the whole life.

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