Ventricular septal defect — autopsy study of 46 cases

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THERE WERE 46 autopsy cases of ventricular septal defect collected from the Paediatric Department, University of Singapore, over a period of three-and-a-half years, from January 1964 to June 1967.

Mortality Rate:

There is a high mortality in infants with large ventricular septal defect and the age distribution of death is as shown in Figure 1.

There are 22 males and 24 females showing no sex bias. The number of cases that died before the age of six months is 33 (72.0%) and before the age of one year, 40 (87.0%). Therefore, the mortality in ventricular septal defect is highest during infancy.

Muir's series of 95 cases (1960)[1] showed that 54 were males and 40 females, one with no sex stated. In this series, 23% died under the age of one month, compared with the present series of 22%; and 62% between one month and one year, compared with the present series of 65% as shown in Table IV.

There is a remarkable uniformity in these two series collected in the same hospital although the

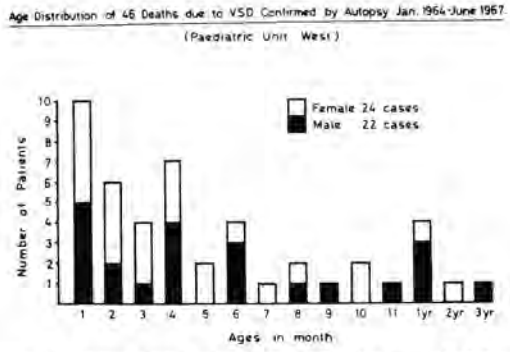

Figure 1 shows death due to ventricular septal defect at various ages. The black area denotes male and the blank female.

present series was collected ten years later. This reflects the fact that there has not been any change in the method of management as the incidence of death has remained the same for the last 20 years.

The ages of four cases listed at one year old were one year six weeks, one year two months, one year, and one year two months respectively, giving the

	Muir. 1948-57	Dept. of Paediatrics, University of S'pore (1964 – mid 1967)
V.S.D.	95	46
Male	54	22
Female	40	24
Death under 1/12	22 (23%)	10 (22%)
Death between 1 month & 1 year	59 (62%)	30 (65%)

Table IV shows the comparison of the two series of ventricular septal defect confirmed by autopsy.

total of 40 cases dying before the age of 14 months. 87% of the total mortality occurs before the age of 14 months. It would appear that the first 14 months of life are the most critical period for patients with ventricular septal defect. Engle (1954)[2] and Edward (1954)[3] have produced evidence that the most critical period of life in ventricular septal defect is the first 18 months. Keith[4], in reviewing 92 cases from the literature plus 19 of his own series, found that 34% died before the age of one year. Nagayama (1965)[11] collected reliable statistical data from the Japanese Pathological Association, showing a mortality rate of 59% in infants with V.S.D. The higher mortality rate in Singapore may be a reflection on the standard of care and availability of surgical treatment.

Size of Defect

In post-mortem patients, the pathologist measured all defects, the distribution of which is as shown in Table V.

More than 1.0 cm in diameter	= 13 cases
Between 0.5 cm to 1.0 cm in diameter	= 26 cases
Less than 0.5 cm in diameter	= 7 cases

Table V shows the size of ventricular septal defect measured by the pathologist.

Selzer (1949)[5] reviewed the literature in ventricular septal defect and emphasised that size of the defect rather than the site is the major determinant of haemodynamic status. Becu (1956)[6], from a similar survey of 50 patients, shared Selzer's opinion on the relative importance of the size of the defect. If the defect is less than 1.0 cm in diameter (or less than half the diameter of its aorta), the magnitude of left-to-right shunt is from small to moderate. If the defect is larger than 1.0 cm in diameter (or greater than half the diameter of its aorta), pressures in the two ventricles are virtually equal (Gorlin 1952)[7].

The size of the shunt is determined by the relative peripheral resistance in the pulmonary and systemic circuits. Left-to-right shunt predominates if the pulmonary vascular resistance is low. Bidirectional shunt or even right-to-left shunt as found in Eisenmenger – V.S.D. occurs if the pulmonary vascular resistance is equal or greater than systemic. It should be emphasised that the size of the defect should not be considered alone; it should have relation to the age of the patient and the size of the heart. For example, a defect of 0.5 cm diameter, which is considered small for an adult patient, is tremendously large for a small infant. Selzer (1954)[8] suggested that the size of the defect should be compared with the diameter of its aorta to be of significance. This has been generally accepted, especially in measuring the defect in open heart repair.

Unfortunately, in the present series of 46 cases, no measurement of the size of the aorta was made at autopsy. The criticism in post-mortem measurement in general is the sliqht underestimation of the size of the defect compared with that in vivo because of the contracted state of the specimen after death. Be this as it may, a defect more than 0.5 cm in diameter is considered large in the present study because 37 (80%) of them were below the age of one year. Therefore, 39 cases (85%) had defects measuring more than 0.5 cm in diameter.

All the defects were of the membranous type situated posteroinferior to the crista supraventricularis involving the outflow portion of the ventricular septum. This is the commonest site of involvement and it occurred in 80% of Becu's series (1965).[6] Ward (1957)[9] reported the incidence of 85% in their 84 operated cases. However, the site of the defect is the least important compared with other factors, such as the size of the defect, the pulmonary vascular resistance and the associated lesions. (Veasy) 1960).[10]

Chest Roentgenology

The importance of chest X-ray investigation in small infants with ventricular septal defect cannot be over-emphasised. The cardiac lesion in more than half (26 cases) of this autopsy series was only suspected after chest X-ray was done because cardiac murmur was not apparent on admission. This could probably be due to tachycardia and cardiac failure with balanced pressures in both ventricles, as in most of them, cardiac murmur appeared a few days after treatment.

It is almost impossible to assess cardiac enlargement clinically in small infants and one has to rely on chest X-ray appearance. Unfortunately, not all chest X-rays were done on full inspiration which requires full co-operation of the patient. Therefore, the apparent size on measurement may not reflect the true heart size.

The cardio-thoracic ratio from a plain chest X-ray is used as an index of heart size the distribution of which is as shown in Table VI.

Cardio-thoracic Ratio	Heart Enlargement	No. of cases
More than 0.60	Severe	20
0.55 – 0.60	Moderate	18
Less than 0.55	Slight	5
No chest X-ray	–	3

Table VI shows the assessment of cardiac enlargement from the cardio-thoracic ratio in a plain chest X-ray

Thirty-eight cases (83%) had roentgenographic cardiac enlargement ranging from moderate to severe. The degree of plethora was difficult to assess because of added pulmonary consolidation as all the patients were admitted with chest infections. It was equally difficult to differentiate which chamber was enlarged. The roots of the great vessels were not well shown in the majority of chest X-rays because of prominent thymic shadow.

Electrocardiography

Not all the patients had electrocardiograms done because cardiac lesions were not suspected in some of them. Electrocardiograms were done in 31 patients and analysis shows that 17 patients (55%) had a mean QRS axis of more than +90°as illustrated in Figure 2.

The diagnosis of ventricular hypertrophy is based on the criteria laid down by Sodi-Pallares and associates (1958)[12] and Guntheroth (1965).[13] The distribution of various ventricular hypertrophies is as shown in Table VII.

All the electrocardiograms were abnormal for the respective ages except one. Isolated right ventricular

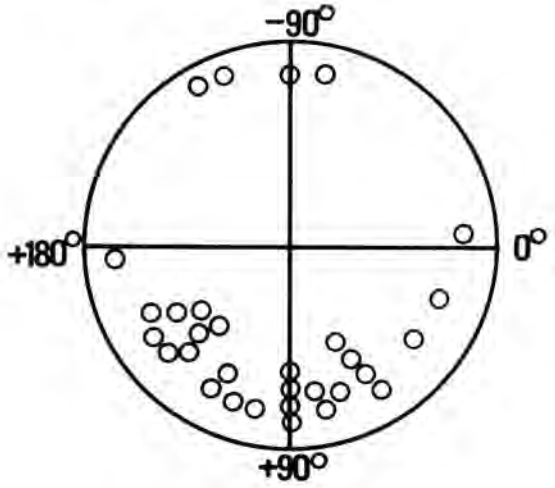

Figure 2 shows the distribution of the mean electrical QRS axis in electrocardiograms done on 31 patients who died of ventricular septal defect.

Normal	LVH	LVH + RVH	RVH
1	6 (19%)	9 (29%)	15 (48%)

Table VII shows the numbers of various ventricular hypertrophies

overload pattern occurred in 15 cases (48%), left ventricular hypertrophy alone in nine cases (19%) and combined ventricular hypertrophy in nine cases (29%). Eighteen cases had an upright T wave in right precordial leads (V_4R and V_1) which was abnormal for the age and is characteristic of severe right ventricular hypertrophy. The deep q wave of more than 4mm and tall T wave in left precordial leads (V_5 and V_6) denoting left ventricular diastolic overload pattern were only present in four cases. Therefore, electrocardiogram obtained in this series was abnormal in 96% of the cases.

Mongolism	10
Edward trisomy	4
Cleft palate	3
PDA (probed)	6
ASD (small)	3
Twins	2
Endocardial Fibroelastosis	1
Congenital Laryngeal stenosis	1
Hydroureter	1
Intersex	1
Microcephaly	1

Table VIII shows a list of various malformations associated with ventricular septal defects with autopsy.

Associated Malformations

In this series of 46 autopsy cases of ventricular septal defect, there was a high incidence of associated malformations. The different types of abnormalities are listed as shown in Table VIII.

The presence of associated abnormalities plays an important part in contributing to the high mortality rate. Chromosomal anomalies like Mongolism and Edward trisomy were associated in 14 cases (30%) and these anomalies are noted for their high mortality in infancy and early childhood.

Summary

Forty-six cases of ventricular septal defect confirmed by autopsy are analysed and discussed in some detail. The high mortality rate seems to be confined to the first 14 months of life. Most of these cases have large defects measured at autopsy. Chest roentgenology and electrocardiography further confirm the severity of the lesions. Associated malformations are common and they play an important role in contributing to the high mortality rate.

References:

1. Muir, C.S. (1960): Incidence of Congenital Heart Disease in Singapore. *Brit. Heart J. Vol. 12,* 243-254.
2. Engle, M.A: (1954): Ventricular Septal Defect in Infancy. *Paediatrics,* 14:16, 1954.
3. Edwards, J.E. (1954): Symposium on Cardiovascular Diseases; functional pathology of congenital cardiac disease. *Paediat. Clin. North Amer.* 1:13, 1954.
4. Keith, J.D., Rowe, R.D., Vlad, P. (1958): Heart Disease in Infancy and Childhood. New York 1958. The Macmillan Co.
5. Selzer, A. (1949): Defect of the Ventricular Septum. Summary of 12 cases and review of the literature. *Arch. Unt. Med.,* 84:798, 1949.
6. Becu, L.M., Fontane, R.S., DuShane, J.W., Kirklin, J.W., Burchal, H.B., Edwards, J.E. (1956): Anatomic and pathologic studies in V.S.D. *Circulation,* 14:349, 1956.
7. Gorlin, R., Gorlin, S.G. (1951): Hydraulic Formula for Calculation of Area of the Stenotic Mitral Valve. Other Cardiac Valves and Central Circulatory. Shunts. I, *Am. Heart J.,* 41:1-29, 1951.
8. Selzer, A. (1954): Defects of the cardiac symptoms. *J.A.M.A.,* 154:129, 1954.
9. Warden, H.E., DeWall, R.A., Cohen, M., Varco, R.L., and Lillehei, C.W. (1957): A surgical-pathologic classification for isolated V.S.D. and for those in Fallot's tetralogy based on observations made on 120 patients during repair under direct vision. *J. Thoracic Surg.,* 33:21-44, 1957.
10. Veasy L.G. (1960): Clinical findings in Ventricular Septal Defects. *Am. J. Cardiol.;* 5, 185-189, 1960.
11. Nagayama, T. (1965): Ventricular Septal Defect. *International Congress of Paediatrics,* Tokyo, 7-13 Nov. 1965.
12. Sodi-Pallares, D., Portillo. B., Cisheros, F., Dela Crus, M.V., and A-costa, A.R. (1958): Electrocardiography in infants and children. *Paed. Clin. North Amer.* 5:871, 1958.
13. Guntheroth, W.G. (1965): Paediatric Electrocardiography 1965 W.B. Saunders Co.