Multiple and single ventricular septal defect

*A clinical and haemodynamic comparison*¹

K. M. FOX², R. G. PATEL³, G. R. GRAHAM, J. F. N. TAYLOR, J. STARK,
M. R. DE LEVAL, AND F. J. MACARTNEY

From the Thoracic Unit and Department of Paediatric Cardiology, The Hospital for Sick Children, Great Ormond Street, London

SUMMARY The clinical, haemodynamic, and therapeutic data were compared in 52 patients with isolated single ventricular septal defect, and 40 patients with multiple ventricular septal defects as the sole cardiac anomaly. Cardiac catheterisation and angiocardiology were carried out between 1966 and 1975 in the multiple ventricular septal defects group, and in 1973 in the single ventricular septal defect group.

The presenting symptoms, signs, and chest x-rays of the two groups were indistinguishable. However, left axis deviation was found on the electrocardiogram in 40 per cent of the patients with multiple defects as opposed to 10 per cent with single ventricular septal defects ($P < 0.01$). In these patients the frontal QRS loop was superiorly orientated and counterclockwise, but in contrast to patients with atrioventricular defects, this feature was usually seen in the absence of complete right bundle-branch block or prolongation of the PR interval. A pulmonary vascular resistance exceeding 5 units/m² was seen equally frequently in both groups, but not in those under 2½ years of age.

The total mortality from the time of presentation, with and without operation, for patients with multiple ventricular septal defects was 20 per cent as opposed to 4 per cent for patients with single ventricular septal defect. Infants with intractable heart failure from multiple ventricular septal defects were treated by banding (mortality 8.3%) and subsequently debanding and multiple ventricular septal defect closure (mortality 27%). Where operation for multiple ventricular septal defects was delayed beyond the age of 1 year, primary total correction was performed, with a mortality of 18 per cent.

Several extensive reports (Witham and McDaniel, 1970; Campbell, 1971; Collins *et al.*, 1972) on ventricular septal defect have been published. Nevertheless, there are relatively few studies concentrating on patients with multiple ventricular septal defects. This is probably both because multiple ventricular septal defect is relatively uncommon and because, apart from direct inspection at operation or necropsy, the diagnosis can only be made by angiocardiology (Taylor and Chrispin, 1971). In the 10-year period between

1966 and 1975 508 cases of isolated ventricular septal defect were catheterised at this institution; of these 40 (8%) had multiple defects. The purpose of the present report is to summarise the clinical, haemodynamic, and therapeutic data of these 40 patients. In order to establish the distinctive clinical features of multiple ventricular septal defects, these subjects have been compared with 52 patients with single ventricular septal defect catheterised in a single year.

Subjects and methods

Forty patients were admitted to The Hospital for Sick Children, Great Ormond Street, London, between 1966 and 1975 and shown to have multiple ventricular septal defects. They were compared with 52 patients admitted during 1973 who were shown to have a single high ventricular septal defect. The diagnosis in all cases was made by selective left

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Present addresses: ²Hammersmith Hospital, Du Cane Road, London W12

³The Hospital for Sick Children, 555 University Avenue, Toronto, Ontario, Canada

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ventricular cine-angiography (Taylor and Chrispin, 1971) in the left anterior oblique projection. A patient was considered to have multiple ventricular septal defects if two or more defects were seen on angiography. Ventricular septal defects were classified as being high (supracristal, infracristal) or low (muscular). Patients with any associated lesion other than a patent foramen ovale have been excluded, so as not to confuse the intended comparison of single with multiple ventricular septal defect. The patients with multiple ventricular septal defects were first seen at this hospital between 1959 and 1975, with a follow-up from the time of first presentation ranging from 1 week to 16 years (mean 8.2 years); the patients with single ventricular septal defect were first seen between 1966 and 1973, with a follow-up ranging from 1.5 years to 9 years (mean 5.7 years).

Details of the presenting symptoms, clinical signs, chest x-ray, and electrocardiogram were noted. Postoperative electrocardiograms on those patients who had undergone total correction were also examined.

Cardiac catheterisation and angiography was performed at least once in all patients. Those who subsequently underwent banding of the pulmonary artery were recatheterised before repair. Pulmonary flow and the pulmonary vascular resistance were calculated on each occasion. Oxygen consumption was not measured routinely in the laboratory during the period of this study and hence has been assumed in these calculations.

Details of treatment, medical and surgical (pulmonary artery banding, repair), and the subsequent outcome were noted in all cases. The surgical procedures used for the repair of both single and multiple ventricular septal defects have been previously described (Breckenridge *et al.*, 1972; Aaron and Lower, 1975; Barratt-Boyes *et al.*, 1976; Singh *et al.*, 1977).

Electrocardiographic analysis

Ventricular hypertrophy (right/left/biventricular) was

defined using conventional criteria (Pagnoni and Goodwin, 1952; Hollman, 1958). Left axis deviation was considered to be present when the mean frontal plane axis lay between -30° and -120° inclusive. When the axis lay between -90° and -120° the loop was invariably counterclockwise.

Chest x-ray analysis

The heart was considered to be enlarged if the maximum transverse cardiac diameter exceeded 60 per cent of the total chest diameter.

Student's *t* test and the χ^2 test were used for statistical analysis.

Results

Thirty-six (90%) of the 40 patients with multiple ventricular septal defects and 44 (85%) of the 52 patients with single ventricular septal defect presented in infancy, that is under the age of 1 year.

Presenting symptoms and signs of both groups are shown in Table 1. The clinical signs, in particular the site and radiation of the pansystolic murmur and the presence of a mid-diastolic murmur, were the same in patients with multiple ventricular septal defects as in patients with single ventricular septal defect.

The chest x-ray in all cases (both single and multiple ventricular septal defects) showed an enlarged heart, prominent pulmonary arteries, and pulmonary plethora.

The electrocardiogram was abnormal in 37 (93%) cases with multiple ventricular septal defects and 47 (90%) of those with single ventricular septal defect. Details of the abnormalities found at the time of presentation are shown in Table 2; both groups are divided according to whether the patient first presented in infancy or childhood. Left axis deviation was seen in 16 (40%) patients with multiple ventricular septal defects at the time of presentation but in only 5 (10%) patients with single ventricular septal defect ($P < 0.01$). Table 3 lists associated electrocardiographic abnormalities

Table 1 *Presenting symptoms in patients with single and multiple ventricular septal defects*

<i>Age at presentation</i>	<i>Multiple ventricular septal defects</i>			<i>Single ventricular septal defect</i>		
	<i>Number</i>	<i>Incidental murmur</i>	<i>Failure to thrive/dyspnoea</i>	<i>Number</i>	<i>Incidental murmur</i>	<i>Failure to thrive/dyspnoea</i>
Neonates (less than 1 month)	10	4 (40%)	6 (60%)	14	5 (36%)	9 (64%)
Infants (1 month to 1 year)	26	9 (35%)	17 (65%)	30	9 (30%)	21 (70%)
Children (over 1 year)	4	2 (50%)	2 (50%)	8	5 (63%)	3 (37%)
Total	40	15 (38%)	25 (62%)	52	19 (37%)	33 (63%)

Table 2 *Electrocardiographic features of patients with single and multiple ventricular septal defects*

Age at presentation		Normal	Left ventricular hypertrophy	Right ventricular hypertrophy	Biventricular hypertrophy	Left axis deviation	Complete right bundle-branch block	First degree heart block	Total
Multiple ventricular septal defects	< 1 year	2 (6%)	8 (22%)	10 (28%)	8 (22%)	15 (42%)	9 (25%)	2 (6%)	36
	> 1 year	1 (25%)	1 (25%)	1 (25%)	—	1 (25%)	—	—	4
Single ventricular septal defect	< 1 year	4 (9%)	10 (23%)	14 (32%)	12 (27%)	5 (11%)	2 (5%)	—	44
	> 1 year	1 (13%)	1 (13%)	3 (38%)	3 (38%)	1 (13%)	—	—	8

Table 3 *Associated electrocardiographic abnormalities in 16 patients with multiple ventricular septal defects who had left axis deviation at time presentation*

Age at presentation	No.	Left ventricular hypertrophy	Right ventricular hypertrophy	Biventricular hypertrophy	Complete right bundle-branch block	Prolonged PR interval
> 1 year	15	1 (6%)	4 (27%)	2 (13%)	2 (13%)	—
< 1 year	1	—	—	—	—	—

in the patients with multiple ventricular septal defects who had left axis deviation. Left ventricular hypertrophy, complete right bundle-branch block, and first degree heart block occurred infrequently.

The frontal plane QRS vector loop was plotted from the 12-lead electrocardiogram and was counter-clockwise in all patients with multiple ventricular septal defects and left axis deviation. The initial QRS deflection was found to be downwards and to the right in 9 patients and downwards and to the left in 7 patients.

All 5 patients with multiple ventricular septal defects who had left axis deviation and who have subsequently undergone repair through a right ventriculotomy were found to have complete right bundle-branch block and left axis deviation on their postoperative electrocardiogram. This conduction disturbance has been persistent over the period of follow-up (mean 3 years) and no patient has developed complete heart block (either transient or permanent).

CARDIAC CATHETERISATION AND ANGIOCARDIOGRAPHY

The initial haemodynamic data together with the post-banding data in those patients who had their pulmonary arteries banded is presented for both groups in Table 4.

Seven patients with multiple ventricular septal defects and 9 with a single ventricular septal defect had a pulmonary vascular resistance above 5 units/m² at their initial cardiac catheterisation. The ages of these patients ranged from 2½ to 8 (mean 4 years) at the time they were studied. All patients who had a normal pulmonary vascular resistance before banding had a normal level at recatheterisation after banding.

The sites of the defects found in patients with multiple ventricular septal defects are shown in Table 5. There appeared to be no association between either the site or number of defects and the presence of left axis deviation on the electrocardiogram.

Table 4 *Initial haemodynamic and post-pulmonary artery banding study of patients with single and multiple ventricular septal defects*

		No. of cases	Pulmonary flow (l/min per m ²) (mean ± SD)	Pulmonary vascular resistance (units/m ²) (mean ± SD)
Multiple ventricular septal defects	Initial study	40	7.1 ± 3.1	4.3 ± 2.9
	Post pulmonary artery banding	11	2.9 ± 1.1	2.7 ± 1.6
Single ventricular septal defect	Initial study	52	7.3 ± 3.5	4.4 ± 3.4
	Post pulmonary artery banding	13	2.6 ± 1.0	2.8 ± 1.1

Table 5 Site of defects found at selective angiocardiology in 40 patients with multiple ventricular septal defects

Site High	Low	No. of cases	Left axis deviation present on electrocardiogram
1	1	10	3
1	2	5	2
1	> 2	3	2
—	2	10	4
—	> 2	12	5

THERAPY AND OUTCOME

A summary of the outcome in the 40 patients with multiple ventricular septal defects and the 52 patients with single ventricular septal defect is presented in Table 6. Eight patients (20%) with multiple ventricular septal defects died as opposed to 2 patients (4%) with single ventricular septal defect ($P < 0.05$). All the deaths in both groups were among the patients who presented in infancy. There were, however, 2 patients with single ventricular septal defect who presented in childhood and who were found to be inoperable because of pulmonary vascular disease (though in one pulmonary artery banding was attempted).

Discussion

Multiple ventricular septal defects might be assumed *a priori* to be a more serious condition than single ventricular septal defect. The results of this investigation show that this is only true to a certain extent. The patients investigated with multiple ventricular septal defects presented with the same symptoms and signs, at the same age, and with the same haemodynamic disturbance as those with single ventricular septal defect. This indicates that in symptomatic patients the clinical features are indistinguishable. Thus, if there is a difference in the population as a whole between single and multiple ventricular septal defects it must lie in the proportion of patients with the two conditions who are ill enough to require cardiac catheterisation. This information

would be impossible to obtain without performing left ventriculography in all patients with the physical signs of a ventricular septal defect regardless of apparent size, a procedure which we do not feel is justified.

What has been shown, however, is that the risks of operation in single and multiple ventricular septal defects differ considerably, as has been emphasised by others (Blackstone *et al.*, 1976). As might have been expected, the presence of a high pulmonary vascular resistance depended more on total size of the defect or defects and the age of the patient at the time of the investigation than on the number of defects in the interventricular septum, since we found a raised pulmonary vascular resistance as frequently in single as in multiple ventricular septal defects. Right ventricular hypertension was not invariably present in patients with multiple defects. Pulmonary artery banding successfully prevented the development of pulmonary vascular disease in both groups provided it was undertaken early. However, a relatively small group of patients with single ventricular septal defect were studied. In a larger group recently reviewed from this hospital, pulmonary artery banding even in infancy did not invariably prevent a rise in pulmonary vascular resistance in later life (McNicholas *et al.*, 1977). The greater surgical risk of closure of multiple ventricular septal defects (compared with single ventricular septal defect) probably reflects the more complicated techniques required to repair multiple defects, and the resultant impairment of function of the ventricular septum (Breckenridge *et al.*, 1972; Blackstone *et al.*, 1976; Singh *et al.*, 1977).

We no longer recommend a two-stage procedure (pulmonary artery banding and then elective total correction) for patients with single ventricular septal defect who require operation in infancy, but prefer primary correction, because this can be carried out with a very low mortality (Barratt-Boyes *et al.*, 1976; Blackstone *et al.*, 1976). Between 1971 and July 1977 we have closed a single high ventricular septal defect in 51 infants with one hospital death

Table 6 Outcome in patients with single and multiple ventricular septal defects divided according to age at presentation

	Age at presentation	No. of cases	Banded in infancy	Childhood	Corrected in infancy	Corrected in childhood With band	Corrected in childhood Without band	No operation
Multiple ventricular septal defects	< 1 year	36	12 (1 death)	—	—	11 (3 deaths)	15 (3 deaths)	9 (1 death)
	> 1 year	4	—	—	—	—	2	2
Single ventricular septal defect	< 1 year	44	13	—	4	13 (1 death)	18 (1 death)	8
	> 1 year	8	—	1	—	—	6	2

(1.9%). The indications for early primary correction in infants with multiple ventricular septal defects are less straightforward. There are no data available to suggest that early primary correction in these patients would carry a higher or lower risk than a two-stage procedure. At the present time, we recommend early repair in most infants with multiple ventricular septal defects. For those with associated cardiac lesions such as coarctation of the aorta, we are in favour of treating the associated lesion and perhaps banding the pulmonary artery. Infants with a 'Swiss cheese' type septum may also require banding. More short and long term follow-up data are required to establish the best form of treatment for this difficult group of patients.

At present, since the risks of surgical management are considerably higher for patients with multiple ventricular septal defects, it is important to distinguish them preoperatively from patients with a single ventricular septal defect. We could discover no means by which patients with multiple ventricular septal defects might be identified clinically or by plain chest radiography. The only clue to the presence of multiple ventricular septal defects before cardiac catheterisation appears to be left axis deviation on the electrocardiogram. Left axis deviation, which in our experience and that of previous investigators (Bäckman, 1972) occurs in only 10 per cent of cases of single ventricular septal defect occurred in 40 per cent of our cases of multiple ventricular septal defects. In some ways the electrocardiogram is similar to that found in atrioventricular canal defects in that the frontal QRS vector loop is counterclockwise, and in 56 per cent the initial QRS deflection was downwards and to the right. However, in atrioventricular canal defects of either partial or complete type, the initial QRS vector was downwards and to the right in 92 per cent (Ongley *et al.*, 1976). Furthermore, whereas PR prolongation occurred in only 5 per cent of our patients with multiple ventricular septal defects, it was found in 93 per cent of patients with complete atrioventricular canal, and 70 per cent of those with the partial form (Ongley *et al.*, 1976). Finally, QRS prolongation was found in 23 per cent of our patients with multiple ventricular septal defects, as against 60 per cent in complete atrioventricular canal (Ongley *et al.*, 1976).

No clear explanation for the high incidence of left axis deviation could be found in these patients. Certainly it did not prove possible to relate it to the site or number of interventricular defects. While it seems clear that patients in whom the ventricular septal defect lies behind and below the membranous septum, with the superior edge immediately below the tricuspid ring (ventricular

septal defect of the persistent common atrioventricular canal type), have a high incidence of a superiorly orientated counterclockwise loop (Neufeld *et al.*, 1961; Ongley *et al.*, 1976), it is also clear that this type of electrocardiogram occurs occasionally in patients with a ventricular septal defect in more usual sites (Feldt *et al.*, 1966; Bäckman 1972).

In conclusion, if a patient with clinical signs suggesting ventricular septal defect has an electrocardiogram with left axis deviation and a counterclockwise loop, but no other conduction abnormalities, and in particular, a normal PR interval, multiple ventricular septal defects should be suspected. However, since this is no more than a clue, we must re-emphasize the need for left ventricular cine-angiocardiology in a suitable projection to display the interventricular septum in all patients with ventricular septal defect in whom an operation is contemplated.

References

- Aaron, B. L., and Lower, R. R. (1975). Muscular ventricular septal defect repair made easy. *Annals of Thoracic Surgery*, **19**, 568-570.
- Bäckman, H. (1972). Influence of structural and functional features of ventricular septal defect on frontal plane QRS axis of the electrocardiogram. *British Heart Journal*, **34**, 274-283.
- Barratt-Boyes, B. G., Neutze, J. M., Clarkson, P. M., Shardey, G. G., and Brandt, P. W. T. (1976). Repair of ventricular septal defect in the first two years of life using profound hypothermia-circulatory arrest techniques. *Annals of Surgery*, **184**, 376-390.
- Blackstone, E. H., Kirklin, J. W., Bradley, E. L., DuShane, J. W., and Appelbaum, A. (1976). Optimal age and results in repair of large ventricular septal defects. *Journal of Thoracic and Cardiovascular Surgery*, **72**, 661-679.
- Breckenridge, I. M., Stark, J., Waterston, D. J., and Bonham-Carter, R. E. (1972). Multiple ventricular septal defects. *Annals of Thoracic Surgery*, **13**, 128-136.
- Campbell, M. (1971). Natural history of ventricular septal defect. *British Heart Journal*, **33**, 246-257.
- Collins, G., Calder, L., Rose, V., Kidd, L., and Keith, J. (1972). Ventricular septal defect: clinical and haemodynamic changes in the first five years of life. *American Heart Journal*, **84**, 695-705.
- Feldt, R. H., DuShane, J. W., and Titus, J. L. (1966). The anatomy of the atrioventricular conduction system in ventricular septal defect and tetralogy of Fallot: correlations with electrocardiogram and vectorcardiogram. *Circulation*, **34**, 774-782.
- Hollman, A. (1958). Electrocardiographic diagnosis of right ventricular hypertrophy in infancy and childhood. *British Heart Journal*, **20**, 129-136.
- McNicholas, K., de Leval, M., Stark, J., Taylor, J. F. N., and Macartney, F. J. (1977). Surgical treatment of ventricular septal defect in infancy. Comparative study of primary closure versus banding of the pulmonary artery. In preparation.
- Neufeld, H. N., Titus, J. L., DuShane, J. W., Burchell, H. B., and Edwards, J. E. (1961). Isolated ventricular septal defect

- of the persistent common atrioventricular canal type. *Circulation*, **23**, 685-696.
- Ongley, P. A., Pongpanich, B., Spangler, J. C., and Feldt, R. H. (1976). The electrocardiogram in atrioventricular canal. In *Atrioventricular Canal Defects*. Ed. by R. H. Feldt and D. C. McGoon. W. B. Saunders, Philadelphia, London, and Toronto.
- Pagnoni, A., and Goodwin, J. F. (1952). The cardiographic diagnosis of combined ventricular hypertrophy. *British Heart Journal*, **14**, 451-461.
- Singh, A. K., de Leval, M. R., and Stark, J. (1977). Left ventriculotomy for closure of muscular ventricular septal defects. *Annals of Surgery*, **186**, 577.
- Taylor, J. F. N., and Chrispin, A. R. (1971). Interventricular septal defect shown by left ventricular cine-angiography. *British Heart Journal*, **33**, 285-289.
- Witham, A. C., and McDaniel, J. S. (1970). Electrocardiogram, vectorcardiogram and hemodynamics in ventricular septal defect. *American Heart Journal*, **79**, 335-346.

Requests for reprints to Professor F. J. Macartney, The Hospital for Sick Children, Great Ormond Street, London WC1N 3JH.