VENTRICULAR SEPTAL DEFECT WITH AORTIC INCOMPETENCE

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We are describing the clinical, cardiac catheterization and necropsy findings in a boy aged 15, with ventricular septal defect and aortic incompetence. The clinical findings are typical of this rare combination of defects. We also review the previously reported cases.

CASE REPORT

P.G., a European male aged 15, was first seen in September 1956 with the following history. During his mother's pregnancy no illness of note had occurred. At the age of 2 months attention was first drawn to his heart by the discovery of a cardiac murmur. Development during infancy and early childhood had been normal, though he was never robust physically and always complained of easy fatigue and dyspnoea, with palpitations on effort. Cyanosis and squatting were never present. At the age of 6 he was taken to Boston, where he was examined by Dr. Robert E. Gross, who found a loud blowing systolic murmur, most intense over the central part of the chest and considered he had a ventricular septal defect.

For 2 years before examination he had been complaining of a sharp lower chest pain on the right side, stabbing in nature, occurring with effort and relieved by rest. His general condition had deteriorated steadily and he had become noticeably more breathless on effort.

He was now a thin, normally developed youth. Cyanosis and clubbing of the fingers was absent. There was no abnormality of his palate, fingers or toes. The jugular venous pulsations and pressure appeared normal. His peripheral pulses were collapsing in type with a Corrigan pulse in the neck and bounding femorals. The blood pressure was 140/60-40 mm. Hg.

There was a slight left-sided praecordial bulge with apical pulsations in the 6th left intercostal space 4 inches from the mid-sternal line. The apex was formed by an overfilled, hyperdynamic left ventricle. A rough systolic thrill was present, felt maximally at the 3rd left intercostal space parasternally. A harsh pan-systolic murmur (grade 5/6) was present, of maximal intensity in the 3rd and 4th left intercostal spaces, close to the sternum. This murmur was suggestive of a ventricular septal defect with a left-to-right shunt. At the base the murmur was different, being shorter, with a mid-systolic crescendo and ending before the aortic second sound. The characteristics were those of an aortic ejection systolic murmur.[32] A long, loud early diastolic murmur was present, best heard parasternally at the 2nd and 3rd left intercostal spaces. Splitting of the second sound could not be detected. The above findings were confirmed on phonocardiography (Fig. 1). At no time were the murmurs thought to be fistulous. In the pulmonary area the findings were clearly those of a to-and-fro murmur. However, at the 4th left space, although the murmurs were 'continuous' in the sense that systolic and diastolic murmurs fused at the second sound, they could be distinguished as murmurs of separate origin.

The electrocardiogram (Fig. 2) showed the pattern of left ventricular hypertrophy. The chest film (Fig. 3) showed considerable enlargement of the heart affecting chiefly the left ventricle, with marked enlargement of the pulmonary arteries and pulmonary plethora. Screening confirmed the general configuration of the heart and a left-to-right shunt was suspected in view of the dilated, vigorously pulsating pulmonary arteries with pulmonary plethora.

The diagnosis of ventricular septal defect with aortic incompetence was made and, as the question of cardiac surgery was seriously considered, cardiac catheterization was performed and the following data obtained: The mean pressure in the right auricle was + 2 mm. Hg and the venous pulse waves were normal. The pressures in the right ventricle varied from 55/0 low down to 35/0 high up in this chamber (Fig. 4). The pulmonary-artery pressure was 38/18 and the mean 'wedge' pressure 13. The systemic pressure at this time was 140/40. Samples from the chambers of the heart showed an oxygen saturation in the right atrium and venae cavae of 68% (Haldane), a right ventricular and pulmonary-artery saturation of 86%, with a brachial-artery saturation of 97%. The systemic flow (calculated on the Fick principle) was 4·8 litres per minute, with a pulmonary flow of 13·2 litres per minute and a pulmonary resistance of 1 unit or 80 dynes sec/cm^5. The catheter findings were regarded as confirming a large ventricular septal defect, with a considerable left-to-right shunt at the ventricular level. The reason for the difference in pressure between the upper and lower parts of the right ventricle was not immediately apparent and is discussed later.

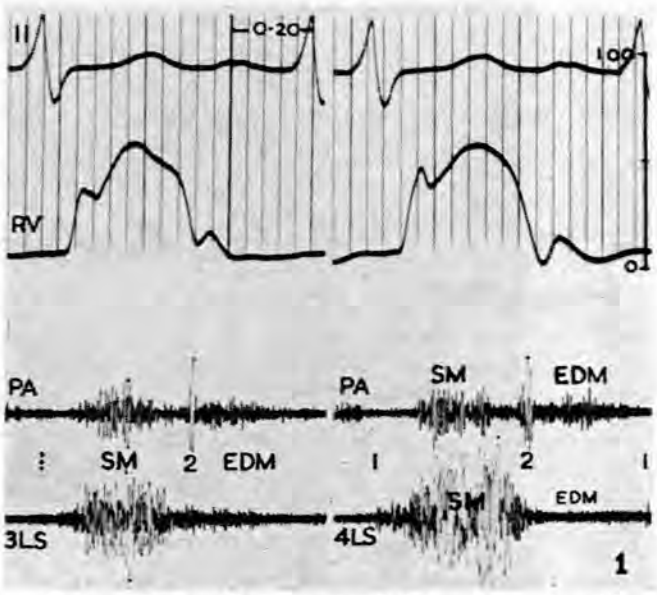

Fig. 1. Phonocardiograms recorded simultaneously with ECG and right ventricular pressure tracing (RV). In the pulmonary area (PA) there is an ejection systolic murmur (SM) with a mid-systolic crescendo, and an early diastolic murmur (EDM) with a crescendo-decrescendo configuration during diastole. The features are those of a to-and-fro murmur and not a fistulous murmur. In the 3rd left interspace (3LS), where the second heart sound is soft, the systolic murmur appears continuous with the diastolic murmur, but the mid-systolic crescendo distinguishes the systolic murmur from the late crescendo found in patent ductus arteriosus. In the 4th left interspace (4LS) a different systolic murmur is recorded. It is much louder, has a crescendo later in systole, and extends into the second sound. It is the murmur produced by a left-to-right shunt through a ventricular septal defect. The early diastolic murmur at this site is much softer than the systolic murmur and although continuous with the systolic murmur did not give the auscultatory impression of a fistulous murmur.

He was discharged until arrangements for surgery could be made. However, urgent readmission was necessary in February 1957 because of severe deterioration in his condition. Breathlessness had rapidly increased until he was orthopnoeic. Palpitations were very troublesome and severe heaving of the chest became noticeable to the patient. Abdominal discomfort, loss of appetite and vomiting followed. For 1 week before admission swelling of the legs and face occurred.

The boy was now found to be extremely ill, semi-stuporose,

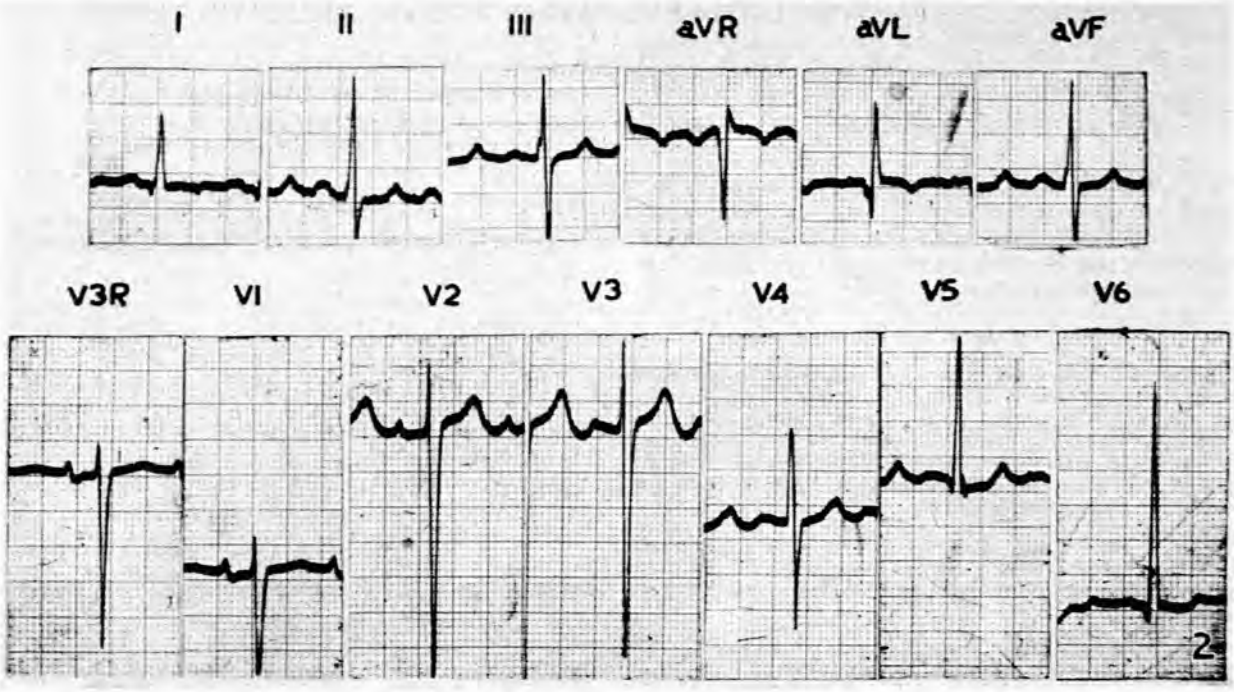

Fig. 2. Electrocardiogram, showing left ventricular hypertrophy.

and with Cheyne-Stokes breathing. Marked tachycardia was present, but no cyanosis. There was marked oedema of the legs, thighs and sacrum, with ascites. The jugular veins were so distended that the pressure could not be assessed clinically, and the liver was 3 fingers below the costal margin, firm and tender. The murmurs were essentially unchanged. The blood pressure was 150/40-0. The electrocardiogram remained unchanged. The X-ray now showed considerable increase in the cardiac size, especially involving the ventricles and pulmonary arteries (Fig. 5). Congestion of the lungs was marked.

Treatment was commenced with digitalis, mercurials and aminophyllin. Penicillin and streptomycin was administered for suspected subacute bacterial endocarditis, despite negative blood cultures. His general condition deteriorated, however, without relief of his acute dyspnoea and he died 4 days after admission.

Necropsy Findings: The findings were those of ventricular septal defect with aortic incompetence. The heart was grossly enlarged (weight 790 g.). The left ventricular wall was 1·5 cm. thick and measured 12·5 cm. in length from the aortic region to the apex. Beneath the medial aortic cusp a ventricular septal defect was present, measuring 2·5 cm. by 1 cm. The lower edge was semilunar in shape and a thickened ridge of endocardium demarcated the boundary (Fig. 6).

The upper limit of the defect on its left ventricular aspect consisted of two of the aortic valve cusps. These cusps were freely mobile, the surfaces being irregularly thickened. The third cusp was less severely thickened. The medial aortic cusp overhung the septal defect and was markedly thickened and displaced medially into the defect; thus the aortic cusps could not be approximated in diastole, resulting in the clinical aortic incompetence.

The posterior papillary muscle of the left ventricle was considerably hypertrophied. A few small linear areas of endocardial thickening were present, presumably the result of aortic incompetence. The sinuses of Valsalva were normal. The right ventricle and auricle were dilated, the former measuring 0·5 cm. thick, with papillary muscles as large as those normally found on the left side. Viewed from the right ventricular cavity (Fig. 7), the defect was directly beneath the medial leaflet of the tricuspid valve. The ventricular septum below the defect bulged convexly towards the right ventricular chamber, hiding the defect from view when seen from below. From the site of the defect and its relation to the tricuspid valve the jet of blood from the left ventricle to the right ventricle may well have impinged on the under surface of the medial leaflet of this valve.

DISCUSSION

The diagnosis of ventricular septal defect was made in this case on the basis of a loud pan-systolic parasternal murmur associated with left ventricular enlargement and a left-to-right intracardiac shunt on screening. The presence of bounding carotids and a collapsing pulse in association with an ejection systolic and early diastolic murmur at the base suggested aortic incompetence. The combination of a ventricular septal defect with aortic incompetence would account for the marked clinical electrocardiographic and radiological evidence of left ventricular enlargement.

The same haemodynamic findings are encountered in a large patent ductus or aortico-pulmonary septal defect. However, in these conditions the murmurs are truly continuous with a late systolic crescendo[33] and decrescendo diastolic murmur and are best heard in the pulmonary area. In our case, the murmurs at the pulmonary area were clearly not continuous, being to-and-fro (Fig. 1.) However, at the 4th space, although in a sense 'continuous', the crescendo of the pan-systolic murmur was in late systole and the diastolic murmur had a crescendo-decrescendo configuration as found in aortic incompetence[42] (Fig. 1). In a ruptured sinus of Valsalva with an aortic-right ventricular shunt, the murmurs are sometimes loudest at the 4th space, simulating very closely ventricular septal defect with aortic incompetence. However, in our case, the history of a loud, low parasternal systolic murmur without any diastolic murmur during childhood strongly favoured the presence of a ventricular septal defect rather than an unruptured aneurysm.

TABLE I: 46 CASES OF VENTRICULAR SEPTAL DEFECT WITH AORTIC INCOMPETENCE, WITH AVAILABLE CLINICAL AND PATHOLOGICAL DATA

(a)	X-ray (b)	ECG	Blood Pres.	Murmur	Age (years)		Pathology	Clin. Diagnosis
					First seen	Death		
2	H III	abnormal	130/20	to-and-fro	7	15	VSD 1·5 × 1·2 cm. R aort. cusp protruded into RV occluding VSD. Fused with ant. pulm. cusp. Wt 575 g.	PDA or AI
4							VSD + AI	
3	LA II LV II H IV + PA II	LV +	110/40	S and D 'continuous'	2/12	3	VSD + R aort. cusp herniation through VSD and adherent to pulm. valve. Wt 180 g.	PDA + VSD
5	H IV PA I AoI	LV +	110/30	At age 6 only syst. At age 9 S and D and thrills.	8/12	9	Ballooned ant. aort. cusp completely hiding the high VSD. Defect 2 × 1·2 cm. LV + + +. Aorta overrides septal defect. Wt 500 g.	? aort.-pulm. septal defect
	RV I LV III	rs-RS in VI, RV +	100/60	left sternal edge S and D		8	RV wall 9 mm. thick. VSD 20 mm. across. LV 11 mm. thick. Aort. valves override VSD. Wt 370 g. Normal for age 115 g.	VSD + pulm. hypertension
8							VSD + AI	
12	H IV + heart					15	VSD + AI	VSD + AI
	H IV +							VSD + AI
15	PA II LV II RV II	LV + + RV + +	130/40-0	to-and-fro S and D	1/12	6 (died at op.)	'Hypertrophy and inf. displacement of the non-coronary cusp of the aort. valve.' VSD in membranous part, 1 cm. across. Endocarditis due to *Str. viridans*. Wt 310 g.	PDA
16			84/0	S and D		14	VSD 1 cm. diam. Almost occluded by pouch-like dilatation of ant. aort. cusp protruding through VSD. Wt 150 g.	PDA
20	H IV LV I RV I PA I	LV +	130/0	S and D	20	21 died suddenly	RV 11 mm. thick, LV 17 mm. thick. High VSD 23 mm. long. Aort. cusp over the defect was as large as the other 2 together and was fenestrated, leading to insufficiency. Death from heart failure	PDA
23	(12 cases)			Murmurs not continuous more properly 'to-and-fro'. Diastolic murmurs may change from time to time. Low diastolic pressure and marked aortic pulsations				
26	H IV	LV + + RV + +	145/80-50	S. Aort. incompetent murmur first heard age 20			VSD + roughening and calcification of R aort. cusp and sac-like dilatation of cusp. Slightly dextroposed aorta. Wt 675 g.	PDA
30							High VSD. Aort. cusp drawn down by fibrous band towards defect, rendering valve incompetent.	
31	(5 cases) H large		'high pulse pressures'	Loud aort. D		18	2 cases died and both had high VSD to which a cusp of the aort. valve was tethered by a fibrous band. L case cyanosed before death	
33	LV I PA II	LV +	180/60	'continuous' holosystolic	34			PDA + VSD
37	H III PA III LV II RV II	RV + LV +		loud S and D	13	13 (died at op.)	Septal defect 2·5 cm. long. Some dextroposition of aorta. LV + + and RV + +. Aort. cusps deformed. Wt 670 g.	PDA
39	H II		112/0	to-and-fro. Aort. D.	7	7	Died from staph. endocarditis. Aorta partially overlay the VSD. Right aort. cusp thickened; its line of closure markedly retracted and lay at a lower level than the other cusps leading to AI. Defect 1·75 cm. in diam.	? VSD ? 1. subaort. stenosis ? 2 bicuspid aort. valves ? 3 congen. aneurysmal dilatation of sinus of Valsalva
43 (c)	H III LV I RV I PA I	RBBB	160/0	to-and-fro	37	56	VSD 1·5 × 5 cm., high in membranous septum, immediately beneath right cusp of aort. valves. Also aneurysm of sinus of Valsalva, opening into RV beneath left cusp of pulm. valve. R. aort. cusp thickened. AI. Wt 960 g.	PDA
	LV III RV III PA III	LV +						
	LV III RV III PA III	Auric. fibril.	160/60	to-and-fro	13	33	Most enlargement in RV. Staph. endocarditis in mitr. and aort. valves. High membranous VSD, 2·5 cm. diam. R aort. cusp immediately above was medially displaced and inferiorly thickened, preventing closure of aort. valve	
45	LV II RV I	LV + +	110/50	S and D	1	21	VSD 2 cm. in diam. Ant. aort. cusp partly prolapsed into defect and much thickened at its edge. Extensive subendocardial fibrosis at base of valve	VSD + AI
								VSD + AI
46	LV II PA II	LV +	140/50	S and D	17			PDA
	LV II PA II	LV +	125/45	S and D	24			PDA
	RV II LV II PA II	LV +	180/60	S and D	Infancy			VSD + AI
48								VSD
47								VSD + AI

Abbreviations: H = heart. RV = right ventricle. LV = left ventricle. PA = pulmonary artery. Ao = aorta. VSD = ventricular septal defect. PDA = patent ductus arteriosus AI = aortic incompetence. RBBB = right bundle-branch block. S = systolic. D = diastolic. Grade of enlargement: mild = I, moderate = II, severe = III, gross = IV.

(a) This column shows who reported the cases (numerals refer to list of references on page 96)

(b) The roman numerals refer to grades of enlargement.

(c) Six cases, of which details of only 4 are set out.

The findings at cardiac catheterization indicated a left-to-right shunt at the ventricular level. The absence of pulmonary hypertension was confirmed, and the increased saturation in the right ventricle could not be attributed to pulmonary incompetence. A ruptured sinus of Valsalva was not completely excluded on the catheter findings.

The natural history of this case is of great interest. At the age of 6 a loud systolic murmur only was present, presumably caused by the ventricular septal defect. With the passage of time the aortic incompetence developed and gradually became a feature of the boy's condition. In fact, the ultimate development of gross aortic incompetence must be regarded as the major contributory cause of death. The later development of aortic incompetence has also been reported by Baylis *et al.*[5] and Hurst and Schemm.[26]

About 10% of ventricular septal defects are openings due to imperfections in the formation of the muscular septum, which may be located in various parts of the septum, whereas 90% of the isolated ventricular septal defects and virtually all such defects occurring in combined malformations of the heart are located in the region of the membranous septum.[35] The term 'membranous' is inadequate, because the defect also involves the large zone of muscular tissue adjacent to the membranous septum. When the defect is large it involves the septal tissue immediately below the annulus of the aortic valve. The aortic leaflets are thus seen to overhang the defect.[27] The aortic cusp which lies above the septal defect is often abnormally large and scarred.[39]

The defect, when viewed from the right ventricle, involves the right ventricular outflow tract fairly low down below the crista supraventriculars[41, 27] and is well separated from the pulmonary valves. When viewed from the left ventricular aspect, on the other hand, the defect is seen to lie immediately beneath the aortic valve cusps[27] and it can readily be understood how the aortic valve cusp overlying the defect may be affected by a stream of blood flowing from the left to the right ventricle.

The case described here represents such a defect and, in addition, the aortic cusp overlying the defect was slightly lower, thickened and scarred, and seemed to hang as it were in mid-air, resulting in aortic incompetence. It seems that the changes found in this cusp may be slowly acquired as a result of trauma or motion. It is also possible for the cusp to be dragged downwards by the Venturi effect of a jet of blood passing below it from the left to the right ventricle through the defect.

In some cases[3, 45, 2] the right aortic cusp may actually protrude through the ventricular septal defect into the right ventricle, partially occluding the defect. In 2 cases[3, 2] the protruded valve adhered to the pulmonary valve. In a case described by Hurst and Schemm[26] the right aortic cusp was calcified. An associated aneurysm of the sinus of Valsalva opened into the right ventricle. Laubry *et al.*[31] described a case in which the right aortic cusp was tethered to the septal defect by a fibrous band. In a case described by Espino-Vela and Mata[20] the right aortic cusp was twice as large as either of the other two aortic cusps and was fenestrated, resulting in incompetence.

Ventricular septal defect with aortic incompetence is not common and, according to Collins *et al.*,[46] was first described by Breccia[47] in 1906 and then by Laubry and Pezzi[30] in 1921. Wood[44] states that the frequency of this association in one series was 2%. As far as we are aware from the literature (Table I) our case is the 47th published case and the 8th to be catheterized.[15] A study of the reported cases (Table I) reveals the remarkable uniformity both of the clinical and pathological findings. Most cases were seriously handicapped. All had a high systemic pulse pressure with evidence of aortic incompetence and marked carotid pulsations.

Although the murmurs are often described as 'continuous', it is likely from the descriptions and from the phonocardiograms published that the systolic murmur of ventricular

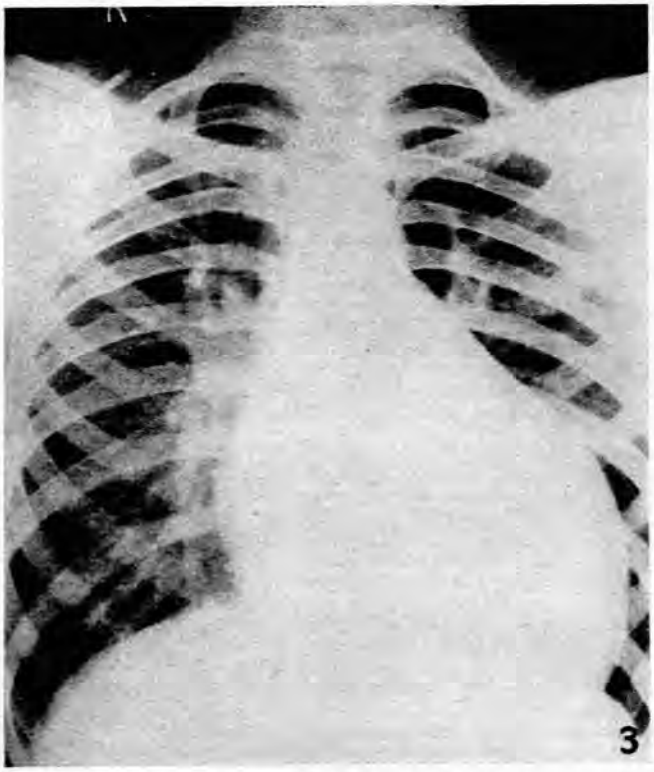

Fig. 3. X-ray of heart, 12 October 1956, showing dilated pulmonary arteries, marked pulmonary plethora, and enlargement of right and left ventricles.

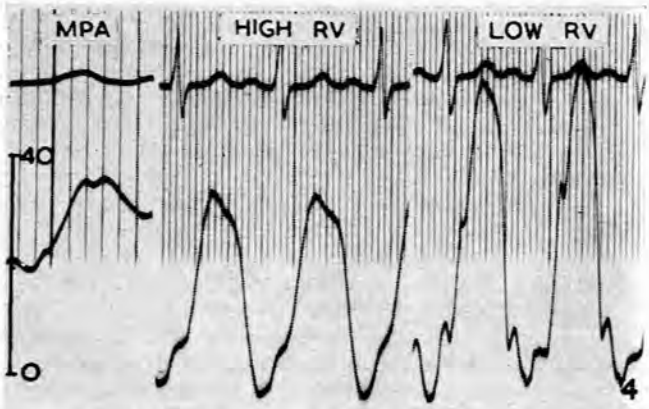

Fig. 4. Cardiac catheterization findings, showing pressure tracings in main pulmonary artery and high and low right ventricle. Note similar systolic pressures of 35 mm. Hg in main pulmonary artery and high right ventricle with a low ventricular systolic pressure of 55 mm. Hg.

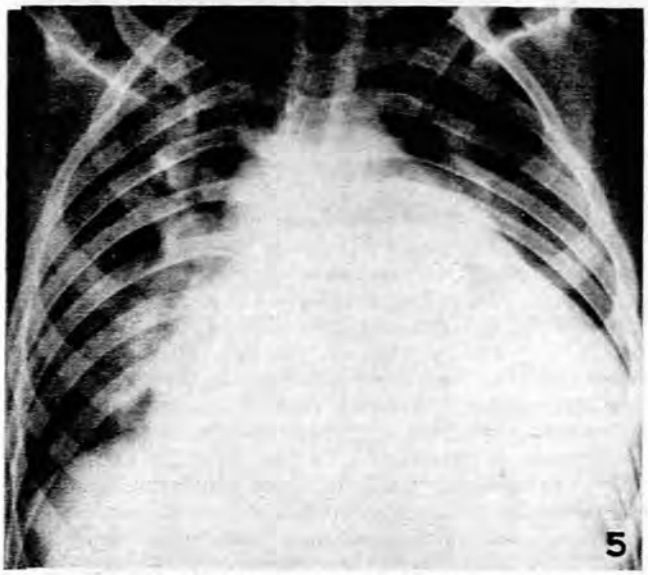

Fig. 5. X-ray of chest 16 February 1957, showing gross cardiomegaly and extreme pulmonary plethora and dilatation of all chambers of the heart.

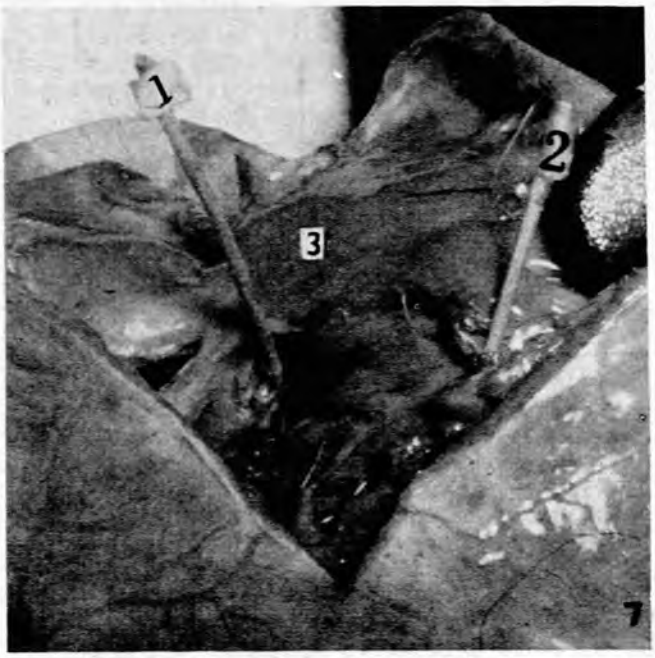

Fig. 7. The specimen shows the cavity and outflow tract of right ventricle. Probe 1 is in ventricular septal defect. Probe 2 is on muscular septum and 3 is on hypertrophied outflow tract below pulmonary valves.

septal defect and the early diastolic murmur of aortic incompetence are present, as in our case.

The electrocardiograms, where reported, showed the signs of severe left ventricular hypertrophy, as found in our case.

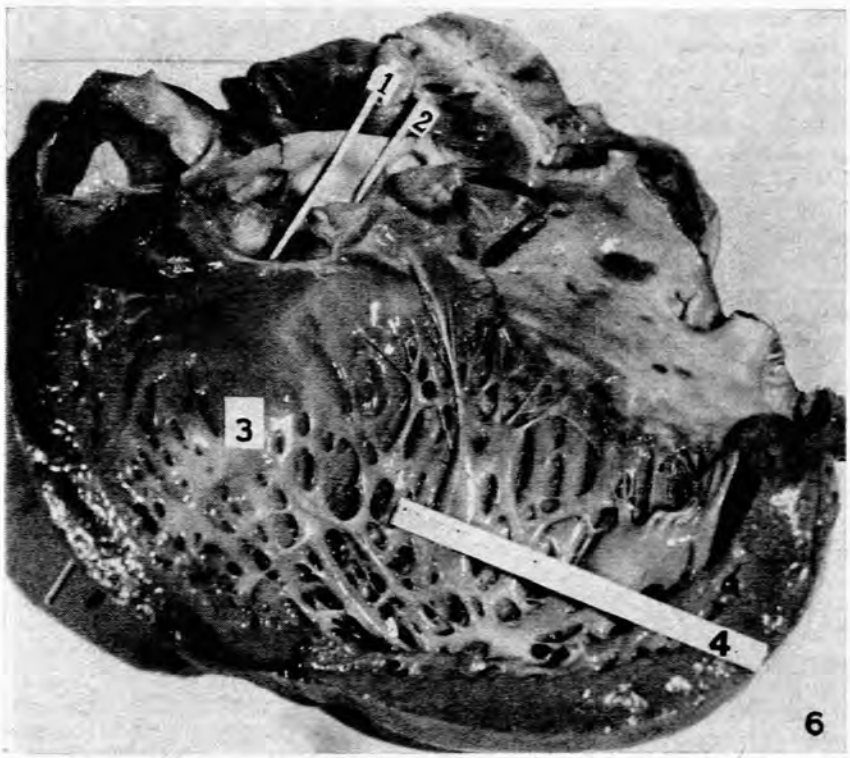

Fig. 6. Heart viewed from left ventricular cavity. Probe 1 is through the ventricular septal defect, just below the hypertrophied, thickened and prolapsed medial aortic cusp. Probe 2 is in the middle aortic cusp. No. 3 is on interventricular septal muscle. No. 4 is on rule showing thickness of left ventricular wall of 15 mm.

Radiograms of the heart in most of the reported cases have shown gross cardiomegaly. The left ventricle is the most severely involved chamber, because of the aortic incompetence, but the associated radiological signs of a ventricular septal defect such as pulmonary plethora and right ventricular enlargement also occur.

The association of the two defects results in severe strain on the left ventricle and carries with it a serious prognosis.

Death in heart failure is the rule, but 3 cases have been described where the cause of death was infective endocarditis,[43, 15, 39] 2 from staphylococcal endocarditis and 1 from *Streptococcus viridans* infection. In 2 of the cases, the endocarditis involved the deformed aortic cusp and the mitral valve, the ventricular defect escaping. None of the 41 previously reported cases were cyanosed except for one,[31] and this patient was cyanosed only while in severe congestive cardiac failure.

The pathogenesis of aortic incompetence in cases with ventricular septal defect is worthy of speculation. It would appear to depend on the presence of a large left-to-right shunt through a septal defect of some size just below the valve. In support of this the

rarity of aortic incompetence in cases of equally large ventricular septal defects where there is no left-to-right shunt, and where there are balanced ventricular pressures either from severe pulmonary hypertension (Eisenmenger's complex) or severe pulmonary stenosis (Fallot's tetralogy).

Although it has been stated that aortic incompetence complicates Eisenmenger's complex,[35, 9] which is by definition 'pulmonary hypertension with reversed interventricular shunt, the cyanosis being accounted for entirely by the right-to-left shunt through the ventricular septal defect,'[36, 44, 35, 12] we have been unable to find an acceptable published case of Eisenmenger's complex with pathological proof of the co-existence of aortic incompetence. Of the 31 cases of Eisenmenger's complex collected from the literature up to 1951 with pathological proof,[36] none had evidence of aortic cusp deformity at necropsy. Although an early diastolic murmur is described in many of these cases, the clinical and pathological evidence seems to point to pulmonary incompetence as the cause. Wood[44] mentions that a Graham Steell murmur due to functional pulmonary incompetence was heard in 2/3 of all his cases of Eisenmenger's complex. No mention is made of aortic incompetence. Early diastolic murmurs were found in 6 of our 9 catheterized cases of Eisenmenger's complex and these, on good clinical grounds, were regarded as pulmonary and not aortic in origin. The differentiation between the early diastolic murmur of aortic incompetence and pulmonary incompetence is sometimes very difficult and often depends on the associated findings. Thus, the presence of a collapsing pulse in the systemic arteries indicates that the early diastolic murmur is aortic in origin, whereas in pulmonary incompetence the systemic pulse is normal or small. Furthermore, there are usually signs of severe pulmonary hypertension when the early diastolic murmur is pulmonary in origin.

In a further 83 cases of Eisenmenger's complex published since 1951 with either catheter or necropsy proof reviewed by us, we were unable to find any example of aortic incompetence.[6, 17, 13, 14, 12, 22, 25, 28, 29, 34, 44] It would therefore appear from a review of the literature that aortic incompetence is exceptional in the Eisenmenger's complex if it ever occurs.

In Fallot's tetralogy aortic incompetence is also most uncommon. We have encountered one case of trivial aortic incompetence in our series of 51 proven cases of Fallot's tetralogy and 1 case of trivial pulmonary incompetence in a mild tetralogy. In the tetralogy the differentiation is readily made because an early diastolic murmur following immediately after the second sound is aortic in origin, where as a pulmonary incompetent murmur commences well after the aortic second sound. This is due to the fact that pulmonary valve closure is always much delayed in that condition.[40]

As the combination of aortic incompetence and ventricular septal defect appears to occur most of all in cases with a large left-to-right shunt and is rare, if it occurs at all, where the ventricular pressures are balanced with a reversal of the shunt, it is not unreasonable to attribute the development of the valve lesion to the left-to-right shunt. Such a shunt may tend to displace and deform a vulnerably placed aortic cusp and in time result in its incompetence.

The pressure differences found in the right ventricle are of some interest. Thus a consistently higher pressure was recorded in the body of the right ventricle compared with that in the outflow tract (Fig. 4). This is presumably due to some obstruction to flow in the outflow tract and not to a Venturi effect, for the pressures well down in the pulmonary arteries were also significantly lower than in the body of the right ventricle. At necropsy, while no organic stricture could be demonstrated, marked hypertrophy of the outflow tract was present. This may account for the small pressure gradient in the ventricle. That hypertrophy of the outflow tract may result in subvalvular obstruction is now well recognized in both ventricles,[10,11,48] Interesting supporting evidence for this view comes from two recent reports. Gasul *et al*,[21] catheterized 2 patients with ventricular septal defects in early infancy, at which time the pressures in the right ventricle were all identical. Four years later, re-catheterization in both showed the development of a definite gradient with markedly higher pressures in the body of the right ventricle. This type of obstruction is apparently reversible, as Engle *et al.*[19] have shown in pulmonary stenosis. Three patients were observed to have a right ventricular pressure of over 100 mm. Hg after pulmonary valvotomy performed under hypothermia. The obstruction that remained was localized to the outflow tract of the ventricle. Cardiac catheterization many months later showed pressures within normal limits with only slight or no transvalvular gradients. Nine of 21 cases of ventricular septal defect operated on by Cleland *et al.*[48] had infundibular obstruction and similar intraventricular pressure gradients as described in our case.

SUMMARY

1. A case of ventricular septal defect with aortic incompetence from cusp deformity is described.

2. The clinical, electrocardiographic and radiological findings are described and related to the findings on cardiac catheterization. The unusual pressure findings in the right ventricular chamber are discussed.

3. The necropsy findings are reported.

4. The literature is reviewed.

ADDENDUM

Since this manuscript was accepted for publication 8 more cases have been reported, [49,50,51] making a total of 54 reported cases. Three of the cases were confirmed by autopsy, 2 were proved by thoracotomy, 6 were catheterized and in 2 a retrograde aortogram aided the diagnosis. The authors stress the diagnostic difficulties and mention cardiac catheterization, retrograde aortography and dye-dilution studies as aids in diagnosis.

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