

Post-infarction ventricular septal defect and aneurysm formation

A case presentation with successful surgical correction and review of the literature

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Summary

A fairly young man suddenly developed a large ventricular septal defect (VSD) after an extensive anterolateral and antero-septal myocardial infarction. Because of congestive cardiac failure refractory to medical treatment he underwent simultaneous closure of the VSD and left ventricular aneurysmectomy 6 weeks after the myocardial infarction. Some 14 months after operation the patient is completely asymptomatic on the minimum of medical therapy and is able to live normally. The literature on these two combined lesions complicating acute myocardial infarction and their surgical correction is reviewed. It is postulated that this patient's infarction was caused by coronary spasm of the Prinzmetal variety with underlying normal coronary arteries.

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Ventricular septal rupture after acute myocardial infarction was first described by Latham in 1845.¹ It occurs in only 1% of cases, more than two-thirds accompanying anterior wall infarctions and the remaining one-third inferior (diaphragmatic) wall infarctions.^{2,3} To date some 100 cases have been documented,⁴⁻⁷ the first successful surgical repair having been carried out by Cooley and co-workers in 1957.⁸

Left ventricular aneurysm formation after myocardial infarction has been reported in some 4-20% of post-mortem studies.^{9,10} In 1944 Beck¹¹ was the first to attempt its surgical correction and there have since been several publications documenting successful repair both in the acute and long-term phases.^{12,13}

Reports of combined repair of ventricular septal defect (VSD) and aneurysmectomy after myocardial infarction are most uncommon, 18 cases having been found on reviewing the literature. The first operation was carried out by Collis and co-workers in 1962.¹⁴ Subsequently there have been further documented cases^{3,7,15-24} with varying postoperative success. We now report the 19th case; the patient is asymptomatic 14 months after surgery.

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Case report

A 40-year-old White man was admitted to the Cardiac Unit, Tygerberg Hospital in December 1978. He had been completely healthy up until some 3 days before admission. His history was that of a typical crescendo angina which culminated in severe precordial pain associated with nausea and vomiting; pain was partly relieved by intravenous morphine given by his general practitioner.

The only risk factor for ischaemic heart disease was smoking; he had smoked 20 cigarettes per day up to 2 years previously.

Physical examination

He was generally well and there were no features of hyperlipidaemia. The radial pulse was regular and rapid at 100/min with a somewhat reduced volume. All the peripheral pulses were easily palpable and equal without any bruits. The jugular venous pressure was not elevated and the blood pressure was 100/60 mmHg. Some dyskinesia was felt on palpating the apex beat which was not displaced and there was no obvious ventricular hypertrophy. A systolic thrill was present over the left sternal edge. On auscultation of the heart the first sound was soft and there were no features of pulmonary hypertension, but a prominent left ventricular third sound could be heard. A grade 4/6 rough pansystolic murmur was present at the left lower sternal edge with fairly good radiation to the axilla and towards the base of the heart. This murmur did not vary with respiration. There were a few fine bilateral basal crepitations in the lungs. The rest of the clinical examination was negative.

Clinical course

A full blood count was normal apart from an elevated erythrocyte sedimentation rate of 65 mm/h (Westergren). The urine was normal. On admission the resting ECG (Fig. 1) showed sinus tachycardia of 100/min, a PR interval of 0.12 second and a mean QRS axis of plus 110°. There was borderline left atrial hypertrophy but no left ventricular hypertrophy. The tracing showed features of an acute transmural anterolateral and antero-septal myocardial infarction. A possible old transmural inferior myocardial infarction could not be excluded.

A chest radiograph (Fig. 2a) was negative. Levels of serum electrolytes, creatinine, urea, uric acid, glucose, total bilirubin, direct bilirubin, total protein, albumin and alkaline phosphatase were all normal. The serum cardiac enzyme levels were markedly elevated and evolved in the typical acute myocardial infarction pattern.

The patient was maintained on heparin 5 000 units every 4 hours intravenously, isosorbide dinitrate 5 mg

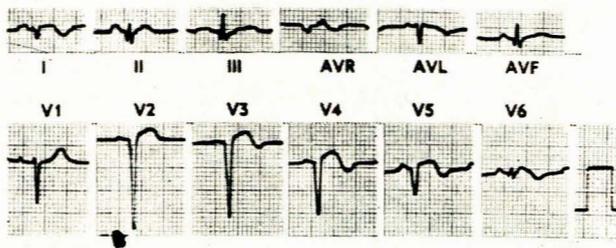


Fig. 1. ECG showing features of acute transmural antero-septal and anterolateral myocardial infarction. A possible old transmural inferior infarction can also be seen.

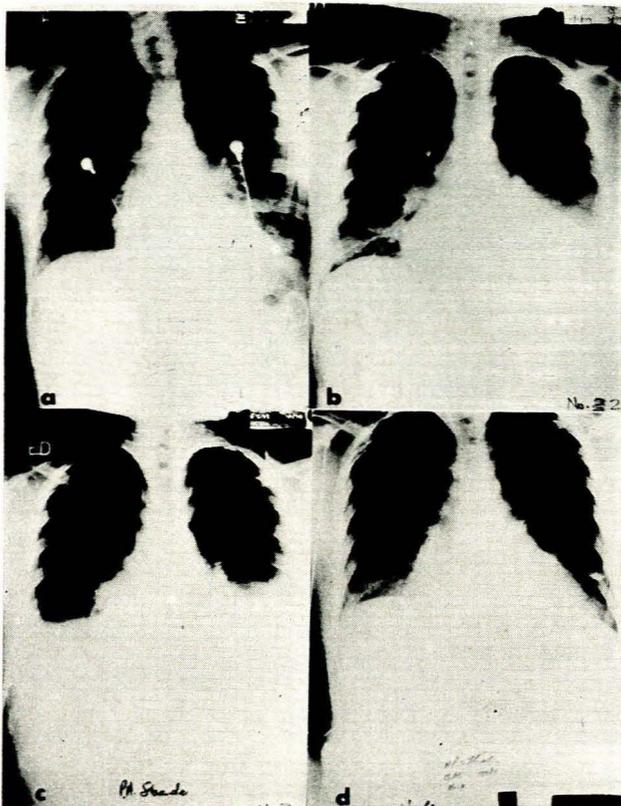


Fig. 2 (a - d). Sequential chest radiographs showing evolution of cardiac failure and pulmonary plethora pre- and postoperatively.

sublingually every 2 hours, intravenous furosemide and sedation. Initially the pansystolic murmur was diagnosed as being due to papillary muscle dysfunction (either the anterolateral or posteromedial one), but the possibility of an acute VSD was also considered although there were features against this diagnosis. On the day following admission the murmur appeared to have changed its character somewhat; it was a little more harsh and did not radiate so strikingly to the axilla. It was decided to exclude a possible VSD, and a double-lumen Swan-Ganz catheter was inserted via an antecubital vein. Three successive determinations of oxygen saturation (%) were documented as follows:

	1st	2nd	3rd
Right atrium (mid)	64	62	64
Right ventricle (high)	87	94	90
Right ventricle (mid)	91	85	91
Right ventricle (low)	67	71	81
Pulmonary artery	77	75	75

The simple bedside investigation thus established the presence of a large VSD high up in the interventricular septum, as evidenced by a step-up in oxygen saturation due to left-to-right shunting. It was then decided to begin therapy with sodium nitroprusside infusion ($1 \mu\text{g}/\text{kg}/\text{min}$) which was titrated against the pulmonary capillary wedge pressure (PCWP) and systolic blood pressure (SBP).

During the subsequent week of the patient's sojourn in the intensive coronary care unit (ICCU) he maintained an adequate blood pressure as well as a mean PCWP which varied between 14 - 20 mmHg. He did not complain of any further angina and no arrhythmias were seen. The sodium nitroprusside was gradually reduced and discontinued, to be replaced with oral and sublingual isosorbide dinitrate. His intravenous line was stopped following a change to oral warfarin and he was maintained on oral diuretic agents.

Some 3 weeks after admission he was gradually mobilized in the intermediate coronary care unit and appeared to be recovering very satisfactorily. However, he suddenly began to complain of dyspnoea as well as a cough. A chest radiograph (Fig. 2b) showed some cardiomegaly as well as a left-sided pleural effusion and other features of pulmonary congestion. A pleural aspiration showed this to be a transudate. Ventilation-perfusion studies were carried out and these tended to exclude pulmonary embolism but suggested possible pneumonia. The patient was therefore given large doses of intravenous ampicillin and cloxacillin, as well as digitalis. Clinically, his condition was fairly satisfactory although a repeat radiograph (Fig. 2c) a few days later showed bilateral pleural effusions and cardiac failure with the possibility of superadded pulmonary plethora caused by the VSD.

Some 5 weeks after admission it was decided to perform a full cardiac catheterization with a view to possible surgery.

Cardiac catheterization

This procedure was performed employing the Seldinger technique from the right groin. The oximetry results, intracardiac pressures and haemodynamic calculations are indicated in Tables I - III respectively. There was evidence of left ventricular dysfunction in that the dp/dt was markedly reduced, the left ventricular end-diastolic pressure (LVEDP) was quite elevated, the mean PCWP was raised, and there was moderately severe pulmonary hypertension.

The following cine angiograms were done: left ventricular (right anterior oblique and left anterior oblique projections); ascending aorta (left anterior oblique projection); selective coronary injections in multiple projections.

As can be seen from Figs 3 and 4, there is a very large aneurysm involving the free anterior wall of the left ventricle as well as the interventricular septum. The remainder of the left ventricle contracted normally. Fig. 4 indicates the presence of a VSD placed high in the interventricular septum. No mitral insufficiency could be seen. Total occlusion of the left anterior descending (LAD) coronary artery, distal to the first septal perfora-

TABLE I. OXIMETRY RESULTS

Catheter position	Oxygen saturation	Comment
Superior vena cava	54	No left-to-right shunting at atrial level
Right atrium (high)	53	
Right atrium (mid)	56	
Right atrium (low)	59	
Inferior vena cava (at diaphragm)	64	
Inferior vena cava (below diaphragm)	71	
Main pulmonary artery	80	No right-to-left shunting
After oxygen	84	
Central aorta	93	Step-up in pulmonary artery saturation
After oxygen	95	
Main pulmonary artery	80	Left-to-right shunting via the ventricular septal defect with 'streaming' to pulmonary artery
Right ventricle (high)	79	
Right ventricle (low)	77	
Right atrium (mid)	63	
Right atrium (low)	61	
Inferior vena cava (at diaphragm)	65	

TABLE III. HAEMODYNAMIC CALCULATIONS

Parameter	Result
Oxygen consumption (l/min)	349
Pulmonary blood flow (l/min)	17,4
Index (Fick) (l/min/m ²)	9,8
Systemic blood flow (l/min)	7,1
Index (Fick) (l/min/m ²)	4,0
Pulmonary vascular resistance (units)	0,9
Index (units/m ²)	1,5
Systemic vascular resistance (units)	11,3
Index (units/m ²)	20,0
Resistance ratio (pulmonary/systemic) (%)	8
Left-to-right shunt (oximetry) (%)	59
Left-to-right shunt (flows) (%)	59
Right-to-left shunt	Zero

tor, is demonstrated in Fig. 5. The left circumflex coronary (a dominant vessel in this patient) and right coronary artery appeared completely normal angiographically (Fig. 6).

In view of the rather surprising coronary angiographic findings, investigations were carried out to exclude possible systemic disease. These were all negative, including serological tests. The patient continued to show evidence of cardiac failure despite maximal medical therapy, and it was decided to operate a week later.

Surgical correction

A median sternotomy was performed and the patient was placed on cardiac bypass with body cooling. Significant oedema and adhesions were noted between the

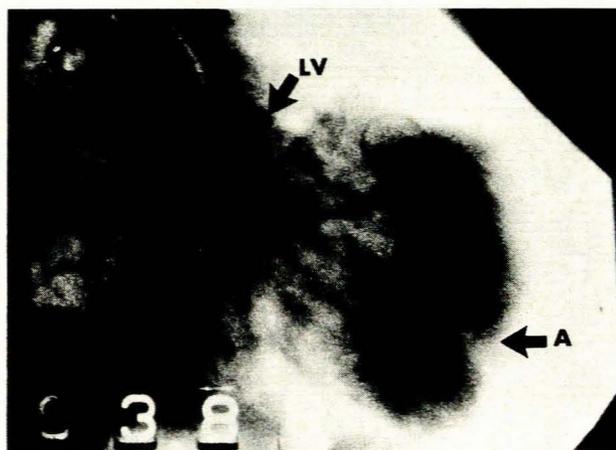


Fig. 3. Left ventricular cine angiogram in systole (right anterior oblique) showing large aneurysm (A), but no mitral regurgitation.

visceral pericardium and epicardium. A large 15 cm diameter aneurysmal sac was present with a fairly well-demarcated fibrotic edge. This aneurysm involved the anterolateral aspect of the left ventricle as well as the apical portion of the interventricular septum. A vent was inserted into the left ventricular cavity via the aneurysm,

TABLE II. INTRACARDIAC PRESSURES

Catheter position	Pressure (mmHg)	Comment
Right atrium	'a' wave 9 'v' wave 5 (mean 4)	Large 'a' waves
Right ventricle	59/0 - 9	Moderately elevated
Main pulmonary artery	59/36 (mean 46)	Moderately severe pulmonary hypertension
Ascending aorta	105/67 (mean 84)	Normal pressure
Left ventricle	105/2 - 27	Very raised end-diastolic pressure
Dp/dt	(mm/s) 804	Markedly reduced
Pulmonary capillary wedge	'a' wave 30 'v' wave 37 (mean 31)	Markedly elevated pressures

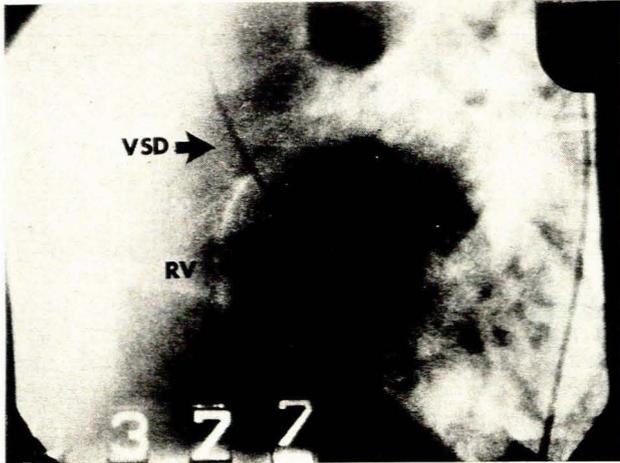


Fig. 4. Left ventricular cine angiogram in systole (left anterior oblique). High VSD is clearly seen (arrowed) between left ventricle (LV) and right ventricle (RV).

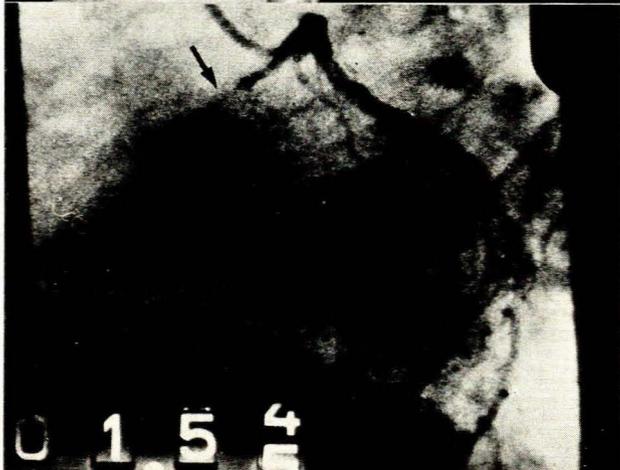
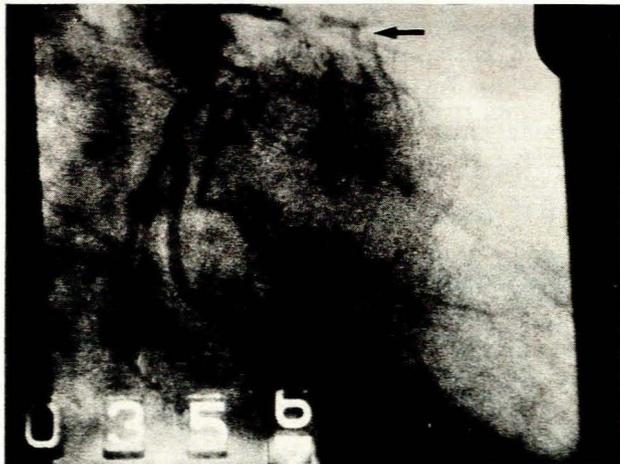


Fig. 5. Coronary cine angiogram (right and left anterior oblique) showing total occlusion (arrowed) of left anterior descending coronary distal to first septal perforator. A normal dominant left circumflex coronary can be seen.

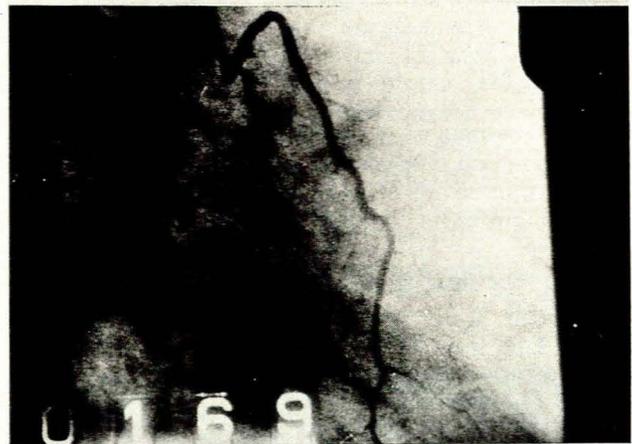
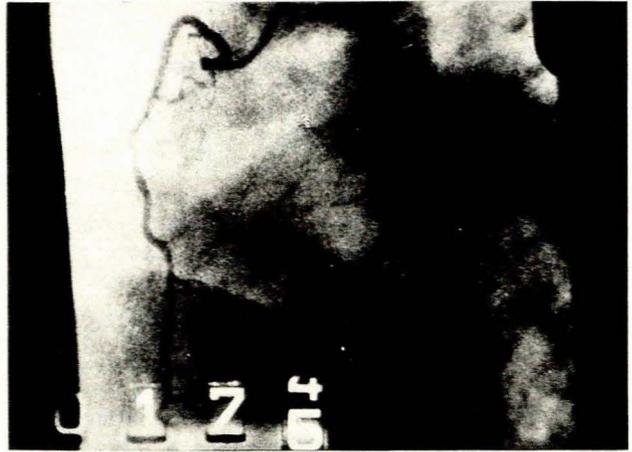


Fig. 6. Coronary cine angiogram (right and left anterior oblique) showing normal, non-dominant right coronary artery.

with resultant collapse of this structure. The largest part of the aneurysmal sac extending to the base of the papillary muscle and the apical portion of the interventricular septum was excised. High up in the muscular interventricular septum a 2 × 1 cm VSD was seen with a well-circumscribed fibrous margin. The VSD was

closed by 3 4-0 Tycron interrupted sutures secured with Teflon pledgets. After removal of the aneurysm an approximately 12 cm cardiac defect remained; this was repaired by interrupted sutures reinforced with Teflon strips. The anastomosis was further secured with a continuous suture of the same material.

Histological examination of the excised specimen verified marked fibrosis within the cardiac muscle. An artery in the myocardium (LAD) exhibited marked, nonspecific arteritis, but no features of atherosclerosis were seen. This picture was in keeping with a myocardial infarction about 6 weeks previously.

Postoperative course

Postoperatively the patient had no complications. A repeat chest radiograph (Fig. 2d) showed no further pulmonary plethora or pleural effusions, but there was some probable linear fibrosis in the left lower lobe. At no time was a systolic murmur heard to suggest a residual VSD. After 2 weeks the patient was discharged on digoxin and a small dose of diuretic. He is being seen regularly at the Cardiac Clinic and continues to deny any symptoms some 14 months after successful corrective surgery.

Anatomical considerations

It has been known for a long time that the interventricular

septum (IVS) has a dual blood supply. The most important contribution is from the left anterior descending coronary artery by way of several long 'septal perforators' supplying more than 60% of the IVS (mainly the superior portion). The remainder of the IVS is supplied by septal perforators which originate from the posterior descending branch of the right coronary when this vessel is 'dominant', or otherwise from the left circumflex coronary artery. Thus there are instances in which the IVS receives all its blood supply from the left coronary artery. However, the IVS is not always completely protected by this dual arterial supply since after infarction VSD can follow occlusion of only one or other descending artery. The presence of functioning 'collaterals' appears to be more important than the dominant arterial source, which would explain the relative infrequency of this complication following acute myocardial infarction.

In contrast to the congenital VSD, which occurs in the upper IVS (membranous portion), post-infarction VSDs are confined to the lower muscular part and may be multiple. The association of a VSD with left ventricular aneurysm formation should be suspected, as a single significant coronary artery obstruction can give rise to both these lesions simultaneously. A single series¹⁷ reported an aneurysm incidence of more than 30% in patients undergoing surgical correction for post-infarction VSD.

Pathophysiological aspects

It is widely accepted that rupture of the IVS can occur at any time from a few hours to several weeks after an acute myocardial infarction. The acutely damaged left ventricle is subjected to further stress on account of the increased volume of blood returning to this chamber having crossed the VSD into the right ventricle and through the pulmonary circulation. In the presence of an additional right ventricular infarction the right ventricle is acutely stressed due to an augmented volume load, including the transmission of the higher left ventricular pressures.²⁵ However, the pressures in the ventricles do not equalize provided there is no significant rise in pulmonary vascular resistance. Equalization of pressures after VSD formation is further counteracted by the fact that the right ventricle is more distensible (being mostly a 'volume-regulated' pump) than its left counterpart (mainly a 'pressure-regulated' pump). It must be remembered that left-to-right shunting also occurs across the VSD in diastole because of the higher pressures in the infarcted left ventricle. Over a longer period of time, when the infarcted ventricular myocardium becomes a true left ventricular aneurysm, elevated pressures on the left side remain, as does the left-to-right shunting. Klein and co-workers²⁶ have demonstrated that when the aneurysm constitutes less than 20% of the left ventricular free wall any increased shortening of normal remaining myocardium can often adequately maintain the stroke volume, avoiding a detrimental increase in cardiac size. With larger aneurysms the stroke volume can only really be augmented with an increase in the left ventricular end-diastolic volume. The compensation then increases intramyocardial wall tension, which in turn increases the oxygen requirements of the remaining viable myocardium. Unfortunately, with a superadded VSD there is a further volume overloading of the compromised left ventricle and a vicious cycle is created. It is thus understandable why these patients develop cardiogenic shock.

Classically a diagnosis of a VSD is made by demonstrating an increase in oxygen saturation at ventricular

level as in our patient, but one must guard against making this diagnosis simply because oxygen saturation has risen in the pulmonary artery alone, because this may well be caused by a ruptured papillary muscle giving rise to secondary mitral insufficiency with reflux of oxygenated blood into the pulmonary arterial circulation by way of the pulmonary veins and capillaries.²⁷ Furthermore, an increase in oxygen saturation in the right atrium has been described in association with a patent foramen ovale being stretched by mitral insufficiency secondary to papillary muscle rupture.²⁸

It is generally accepted that the holosystolic murmur of mitral regurgitation due to papillary muscle rupture can be differentiated from that due to a VSD on account of excessively large 'V' waves in the pulmonary capillary wedge pressure (PCWP). Nevertheless, Drobac and associates²⁹ demonstrated the presence of giant 'V' waves in the presence of post-myocardial infarction VSD without concomitant mitral regurgitation. This feature has been thought to be due to the sudden volume overloading of an unprepared, non-compliant, left atrium after an acute VSD, the 'V' waves decreasing in amplitude over time, analogous to the effect on mean left atrial pressure in chronic severe mitral insufficiency. Thus, all the preceding comments have to be considered in analysis of a Swan-Ganz catheterization in attempting to arrive at a bedside diagnosis.

Clinical features

Auscultory findings

In the 19 cases (inclusive of our present case) in the literature a few features are quite outstanding. Every single patient passed through a phase of congestive cardiac failure and many actually developed cardiogenic shock. The systolic murmur was always rough in quality and accompanied by a thrill. However, contrary to what is generally believed, only some 25% of patients demonstrated the typical site of the ventricular septal rupture murmur at the lower left sternal edge; others presented with a murmur which radiated significantly to the axilla as well as up the left sternal edge. Thus, auscultation is not very reliable in distinguishing the murmur secondary to an acute VSD from that due to mitral regurgitation caused by papillary muscle rupture or dysfunction. This fact underlines the importance of attempting to come to a more definitive diagnosis by employing bedside right heart catheterization by means of a Swan-Ganz catheter.

Sex and age incidence

Out of a total of 19 patients with combined post-infarction VSD repair and aneurysmectomy only 4 were female (Table IV). This fact is not that surprising when one considers that symptomatic ischaemic heart disease, especially in the young, is far more prevalent in males. The age of the patients ranged between 40 years and 76 years, with a mean age of 59 years. Our male patient was the youngest at 40 years of age; some 20% of patients were 70 years or older.

Interval between infarction and VSD

The approximate time interval between acute myocardial infarction and ventricular septal rupture varied between an almost simultaneous occurrence in 4 cases and as late an onset as 7 years. However, some 63%

TABLE IV. POST-MYOCARDIAL INFARCTION VENTRICULAR SEPTAL RUPTURE AND VENTRICULAR ANEURYSM

Author	Age (yrs)	Sex	Time intervals		Cardiac catheterization			VSD		Aneurysm			Result	
			Infarct to rupture (days)	Rupture to operation (days)	Qs (l/min)	Op/Qs	PA (mmHg)	Size (cm)	Closure	Location	Size (cm)	Status and survival (mo.)	Residual VSD	
Collis <i>et al.</i> ¹⁴	49	M	17	91	3.4	2.8	81/31	3 X 1.5	S	LV	8 X 5	7	Alive	-
Taylor <i>et al.</i> ¹⁵	47	M	11	26	-	-	-	2,2	S	LV	4 X 3	8 1/2	Alive	-
Heimbecker <i>et al.</i> ¹⁶	56	M	12	<1	-	~2.5	-	-	P	RV and LV	-	1	Dead	-
Daicoff <i>et al.</i> ¹⁷	76	M	6	37	2.5	2.0	75/20	1.5 X 1.3	S	LV	8	13	Alive	+
Stinson <i>et al.</i> ¹⁸	57	M	8	8	-	-	-	2.0	P	RV and LV	-	6	Alive	+
Selzer <i>et al.</i> ¹⁹	52	M	3	38	5.6	3.1	60/24	2 X 1.5	P	LV	-	<1	Dead	-
Limsuwan <i>et al.</i> ²⁰	55	F	1	55	4.1	4.1	59/18	2 X 2	S	LV	2 X 2.5	16	Alive	-
Daggatt <i>et al.</i> ²¹	69	F	<1	12	-	-	-	-	S	LV	8	9	Alive	-
Freeny <i>et al.</i> ²²	47	M	2	68	-	-	-	2 X 1	?	LV	10 X 6	13	Alive	-
	46	M	<1	83	-	3.6	78/20	4 X 2	S	LV	4	24	Alive	-
	60	M	21	119	-	3.7	79/20	1	?	LV	7 X 4	8	Dead	-
	70	M	7 (yrs)	364	-	1.5	65/36	1	?	LV	4.5 X 4	7	Dead	-
Schlesinger <i>et al.</i> ²³	70	F	23	120	1.0	2.7	50/18	0.9	S	LV	8	9	Alive	-
Longo <i>et al.</i> ⁵	61	M	6	18	-	-	35/16	-	S	LV	-	20	Alive	-
Windsor <i>et al.</i> ⁷	76	M	5	140	-	2.0	50/20	-	P	LV	-	?	Alive	-
	72	M	5	42	-	2.4	90/27	-	S	LV	-	?	Alive	-
	59	M	4	168	-	2.0	-	-	P	LV	-	?	Alive	-
Egeblad <i>et al.</i> ²⁴	48	F	<1	150	1.0	3.0	58/26	0.8	S	LV	8	14	Alive	-
Przybojewski <i>et al.</i>	40	M	<1	42	7.1	2.4	59/36	2 X 1	S	LV	15	-	Alive	-

M = male; F = female; Qs = systemic blood flow; Op = pulmonary blood flow; PA = pulmonary artery pressure; VSD = ventricular septal defect; S = suture; P = patch; LV = left ventricle; RV = right ventricle.

developed the VSD within the first week after infarction, which is in line with experience in the literature relating to ventricular septal rupture alone. This time interval had no influence on the success of the operation.

Electrocardiographic findings

The most prevalent site of the acute infarction electrocardiographically, in the 19 cases, was anterior (15 cases); 3 infarcts were inferior (15%), and 1 was inferolateral (6%). Unfortunately, an accurate correlation with selective coronary arteriographic data is not possible as the majority of patients did not undergo this investigation. These findings are in general agreement with the belief that most ventricular septal ruptures as well as anterior left ventricular aneurysms are related to anterior infarctions and predominantly left coronary artery obstructive lesions.

Interval between VSD and surgery

The time lapse between diagnosis of ventricular septal rupture and corrective surgery ranged from a few hours to 1 year. Some 63% reached a minimum period of 6 weeks before operation, and a further 31% were not operated upon for at least 3 months. This time interval had no significant bearing on the postoperative survival period, although widespread opinion has emphasized the crucial role of timing of operation. Generally, the patient is treated medically for as long as possible to allow time for formation of fibrous tissue around the periphery of the VSD as well as the periphery of the left ventricular aneurysm. This makes technical repair by the surgeon much more simple and should theoretically reduce the possibility of postoperative VSD due to detachment of patches or stitches. Supportive medical therapy has consisted of vasodilator drugs, digoxin, diuretics, and intra-aortic balloon pumping (IABP). IABP is the most appropriate therapy theoretically because during diastole the increase in intra-aortic pressure increases coronary perfusion pressure as well as flow, and thus can augment the contractility of the ischaemic myocardium. During systole the intra-aortic balloon unloads the pressure against which the ischaemic ventricle must contract and thus it increases the forward flow from left ventricle to aorta, decreasing left-to-right shunting. However, it must be realized that IABP is only temporary treatment; over a longer term it becomes less effective in cardiogenic shock and many technical difficulties appear.

Diagnostic techniques

Swan-Ganz catheterization

This bedside procedure was first suggested for diagnostic purposes in the acutely ill patients with post-myocardial infarction ventricular septal rupture by Lufschanowski *et al.*³⁰ and Kaplan *et al.*⁹ Their argument was that this was quicker to perform and less hazardous than left ventricular cine angiography. Longo *et al.*⁵ (Table IV) were the first to use this procedure in post-infarction VSD associated with left ventricular aneurysm, but then went on to perform complete cardiac catheterization and selective coronary arteriography. They were also the only workers in this series to use IABP pre-operatively. Apart from Longo's patient we were the only workers to apply the bedside Swan-Ganz catheterization in this group of patients, as well as to follow up this procedure with definitive right and left heart catheterization, left ventri-

cular cine angiography, and selective coronary arteriography.

Complete cardiac catheterization

Most of the earlier patients¹⁴⁻²³ had a simple right heart catheterization (under fluoroscopic vision), including measurement of pressures and oxygen saturations taken from either the brachial or radial artery. Because of improvements in cardiac catheterization techniques the subsequent workers^{27,28} obtained left ventricular pressures as well.

M-mode echocardiography

This non-invasive procedure has delineated several features in acute ventricular septal rupture. Dilatation of the right ventricle, due to volume overload from the shunt, was the most striking characteristic.³¹ Paradoxical septal motion (PSM) has also been described,^{32,33} as has a reversed or reduced EF-slope of the tricuspid valve.³² Differentiation from acute papillary muscle rupture of the mitral valve is possible in that in this situation septal motion amplitude is increased and dilatation of the left atrium and ventricle occurs.²⁴ Egeblad and Haunsø²⁴ were the first to diagnose post-infarction acute VSD with an associated left ventricular aneurysm in a patient who later had a VSD closure and aneurysmectomy. His case was the only one studied by echocardiography in the series of 19 cases. Prognostic assessment following aneurysmectomy in these cases can be made. Feigenbaum *et al.*³⁵ indicated that prognosis was good if the base of the left ventricle was normal and if there was normal contractility of the non-infarcted myocardium.

Haemodynamic characteristics

Of the 19 patients, 14 had pulmonary to systemic blood flow ratios (Qp/Qs) calculated. These ratios varied between 1.5 and 4.1 (Table IV). The left-to-right shunting in the majority of cases was significant, that is, 10 out of 14 patients had a Qp/Qs ratio of above 2.0. As expected, the magnitude of the shunt was closely related to the systolic pulmonary artery pressure recorded, the latter ranging between 35 mmHg and 90 mmHg, with a mean value of 64 mmHg. There did not appear to be any correlation between the severity of the pulmonary hypertension, magnitude of Qp/Qs ratio and size of ventricular septal defect found at operation. Likewise, there was no relationship between these parameters and the length of survival. The other salient feature was the absence of any detectable right-to-left shunting across the VSD, which is to be expected since ventricular pressures are not equalized in these acquired defects, in contrast to a congenital membranous VSD.

In only 2 cases was a pulmonary capillary wedge pressure (PCWP) documented; it was elevated to a mean value of 22 mmHg (normal 12 mmHg) in Longo's⁵ patient, and to 31 mmHg in our patient. Neither had mitral insufficiency, although our patient had peaked 'V' waves reaching an amplitude of 37 mmHg; this feature seen on bedside Swan-Ganz catheterization is thus not pathognomonic of mitral insufficiency and cannot be absolutely relied upon in differentiation from an acute ventricular septal rupture. Furthermore, our patient had a markedly elevated left ventricular end-diastolic pressure (LVEDP) of 27 mmHg, as well as a severely reduced dp/dt of 804 mm/s (Table II). These features are entirely explicable on the magnitude of the left ventricular aneurysm

with secondary cardiac failure, also aggravated by the volume overloading from the VSD. Our patient still had a well-preserved cardiac output of 7.1 l/min (Table III), indicative of adequate remaining viable left ventricular myocardium, and thus predicting a good surgical outcome.

Angiography and selective coronary arteriography

The first few cases of post-infarction ventricular septal rupture associated with left ventricular aneurysm were not diagnosed with absolute certainty by angiography because cardiac catheterization techniques had not advanced sufficiently. Thus, the left ventricular aneurysm was suspected on the grounds of bedside examination, persistence of ST-segment elevation electrocardiographically, and the presence of congestive cardiac failure (unfortunately usually overshadowed by the features of the ventricular septal rupture). Many of the aneurysms were discovered by chance in the operating theatre. Schlesinger and co-workers²⁵ were the first to employ angiocardiology pre-operatively; however, they did not catheterize the left heart but injected contrast material into the main pulmonary artery and waited for re-circulation through the lungs to occur. This technique showed up the increased pulmonary vascularity, enlarged left atrium, left ventricular aneurysm, and remaining viable ventricle. These workers preferred this approach to that of direct left ventriculography as such complications³⁶ as acute left heart failure, acute rhythm disturbances, and embolic phenomena originating from the aneurysmal thrombi could be avoided. They also felt that this method, undertaken during right heart catheterization, saved an additional procedure. Finally, in their experience, the degree of radiological opacification of the aneurysmal sac resulting from right heart approach was satisfactory.

Longo *et al.*⁵ were the first to perform selective coronary arteriography in these patients. Their patient had a total occlusion of the posterior descending branch of the right coronary artery, insignificant lesions in the left circumflex coronary artery, and a normal left anterior descending coronary artery.

The first workers to undertake direct left ventricular cine angiography in patients with post-infarction VSD and concomitant left ventricular aneurysm were Windsor *et al.*⁷ They experienced no complications but claim that it was probably unnecessary because the surgeon could readily ascertain the presence of an aneurysm at operation. It is unfortunate that these workers did not perform coronary arteriography as there may have been an indication to bypass obstructive lesions (*vide infra*).

In the case reported by Egeblad *et al.*²¹ direct left ventricular cine angiography and selective coronary arteriography were undertaken. They found a subtotal obstruction in the left anterior descending branch 4 cm distal to its origin from the left mainstem coronary, but this vessel was not bypassed at surgery. The remaining coronary arteries and their branches were reported as normal.

In our case direct left ventricular cine angiography in 2 radiographic views enabled accurate delineation of the left ventricular aneurysm, as well as the VSD. In addition, selective coronary arteriography revealed total occlusion of the left anterior descending branch just distal to the first septal perforator.

Operative findings and surgical procedures

The VSD was located in the apical portion of the septum in 7 cases, anterior in 9 (high-anterior in 3 cases) and posterior in 3 cases. Its diameter varied between 1 cm and 4 cm, some 50% being about 2 cm in size (Table IV). Most of the aneurysms involved the anterolateral wall of the left ventricle, although 2 patients had posteriorly situated aneurysms. Two of the 19 had additional right ventricular aneurysms^{16,19} which were usually large, varying from 2 cm to 15 cm in their long axis.

Some variation in repair of both the VSD and aneurysm was noted. In 18 of the 19 patients the defect was approached by an incision into the sac as recommended by Stinson *et al.*¹⁹ as well as by Kitamura *et al.*³⁷ This approach prevents any possible damage to healthy myocardium and provides good visualization and access to the VSD. In only 1 of the cases²⁰ was a right ventriculotomy performed. Eleven of the 19 patients only required interrupted sutures, reinforced by Teflon strips, to repair the VSD; 5 patients had some form of patch inserted and in the remaining 3 patients the method of VSD closure is not documented.

The question of revascularization of the myocardium by way of aortocoronary saphenous vein bypass grafting is fairly controversial. This procedure must obviously be preceded by selective coronary arteriography, which was carried out in only 3 of the 19 cases.^{3,21} If the patient has medically refractory angina and cardiac failure, complete re-vascularization (providing the vessels are angiographically suitable) should be done at the same time as the other procedures. This would probably relieve the angina and prolong survival, as most late deaths following aneurysmectomy are secondary to new sites of infarction remote from the aneurysm. On the other hand, in patients with congestive cardiac failure without angina, who usually have complete occlusion of the left anterior descending coronary artery with anterolateral aneurysm formation (as in most of the patients reviewed), and in whom the distal part of this artery fills by way of collateralization from the remaining coronary arteries, attempts should be made to preserve the left anterior descending artery and bypass it. If there are additional significant lesions in the other coronary arteries, and these are suitable for grafting, then they should also be bypassed. Unfortunately, coronary arteriography indicated that the left anterior descending artery was unsuitable for grafting in our patient.

Results of surgery and prognosis

The outcome of surgery as judged by improvement of cardiac failure, the presence of a residual VSD clinically, and the survival rate is most satisfactory. All patients were free of congestive cardiac failure on minimum therapy. Only 3 patients had a residual VSD, but survived (Table IV). In 2 cases^{17,19} the residual VSD was small, but in the third it was large, necessitating re-operation after 2 weeks, following which no systolic murmur could be heard. This patient also required a permanent pacemaker because of complete heart block secondary to surgery. The interval between infarction and surgical correction in the 3 patients with a residual VSD was 16, 43 and 47 days respectively. Two of these residual VSDs were apical and the third posterior. None of the patients with a residual VSD was hypertensive, a factor always claimed to precipitate this complication. There was no

correlation between this time interval and the formation of a residual VSD.

There were 4 deaths among the 19 cases, at 24 days, 1, 7 and 8 months respectively (Table IV). All these patients were males, their ages ranging from 52 years to 70 years. Two died of non-cardiac causes, 1 of a probable reinfarction, and the fourth had a sudden death thought to have been arrhythmia-induced. The survival periods of the remaining 15 patients varied from 6 months to 2 years (mean 12.7 months). In assessing prognosis the underlying coronary artery disease, any additional disease, and risk factors are important. Patients also tended to be older, making prognosis less favourable. An exception is our patient who is 40 years old and whose only risk factor was smoking. The fact that most patients did not undergo coronary arteriography makes assessment of the extent of their coronary artery disease and therefore of prognosis very difficult, despite what appears to have been very adequate surgery.

We postulate that our patient's myocardial infarction was caused by spasm^{38,39} superimposed on normal coronary arteries, especially in the light of the histological findings. Workers⁴⁰ have shown that locally aggregated platelets can cause coronary spasm by release of vasoconstrictors such as the prostaglandin-related thromboxane A₂. Madias⁴¹ has shown that smoking can precipitate variant (Prinzmetal's) angina, and Maseri and co-workers⁴² found a fresh coronary artery thrombosis in a patient who shortly before death had coronary spasm on angiography. The prognosis of patients suffering from this variety of angina was previously considered to be most grave⁴³⁻⁴⁵ but more recently this point has been disputed.⁴⁶⁻⁴⁸ In conclusion, provided our patient does not have any further attacks of coronary artery spasm involving his remaining angiographically normal vessels, he may well survive many more years. This suggestion appears more realistic when one considers that resection of as much as 35% of the surface of a dilated ventricle has been shown to be tolerated.⁴⁹

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Umbilical endometriosis

A case report

N. J. BLUMENTHAL

Summary

A patient presented with a dark, painful and bleeding umbilicus, and was found to have endometriosis of the umbilicus. The details are presented, as well as a discussion of the aetiology, incidence, differential diagnosis and histological features of the condition.

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Endometriosis is a common disease in which ectopic uterine mucosa is found in widely scattered areas of the body. Various theories as to its pathogenesis have been suggested, none of which can unequivocally account for endometriosis in all the various locations in which it has been reported. The umbilicus is one of the less common sites for the localization of ectopic endometrium.

According to the literature a total of only 109 cases of umbilical endometriosis has been reported; it is particularly rare among Blacks. A case is described in a Black patient with classic symptoms and signs of umbilical endometriosis; the pathogenesis of the condition is discussed.

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Case report

A 46-year-old patient (para 10, gravida 10), was first seen in the Gynaecological Outpatient Department at Baragwanath Hospital in 1979. Her main complaint was of a lump on the umbilicus, which had been present for 2 years and had continued to grow. The lump enlarged at the time of menstruation, became dark, painful and often secreted dark blood. The menarche had occurred at the age of 14 years; thereafter normal, regular, monthly periods took place, with menses lasting 4 days. She had never used any form of contraception and denied any history of dysmenorrhoea or dyspareunia. She had no history of previous abdominal surgery or any congenital abnormalities.

Abdominal examination revealed a 4 x 4 cm papillomatous, fungating umbilical mass which was dark and not tender. No blood could be expressed. Vaginal examination revealed a 14-week fibroid uterus, normal ovaries and no suggestion of pelvic endometriosis.

A diagnosis of umbilical endometriosis was made and an excision biopsy was performed. Through a mini-laparotomy it was found that the umbilical tumour had no connections or extensions; it was therefore excised with a small cuff of skin and sheath. No evidence of pelvic endometriosis could be found.

The pathologist reported typical endometrial tissue consistent with endometriosis. The patient had an uneventful postoperative course and follow-up showed no residual or new endometriosis.