Cardiomyopathy complicated by left ventricular aneurysms in children

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Ventricular aneurysms in children are unusual. Three patients with cardiomyopathy associated with angiographically proved left ventricular aneurysms in this age group are reported. Two of them were girls. The ages were 20 months, 7 years, and 14 years. Heart failure was present in all patients. There was radiological evidence of cardiomegaly in all three, and the electrocardiogram showed signs of necrosis in two of them. Selective left ventricular angiography disclosed generalized hypokinesis in all patients. One child had an aneurysm of the diaphragmatic wall. In another the aneurysm was localized in the muscular ventricular septum, causing severe subpulmonary stenosis by encroaching in the right ventricular outflow tract during systole. The third patient had an aneurysm of the left ventricular free wall partly encircling the left ventricle. The coronary arteries appeared normal in all cases. The clinical features of the underlying disease were not altered by the presence of the aneurysm except in the patient with the septal aneurysm and subpulmonary stenosis. In this patient the aneurysm was successfully resected.

The commonest cause of left ventricular aneurysm in adults is ischaemic heart disease (Cheng, 1971). Unusual causes for this lesion include syphilis (Braunstein, Bass, and Thomas, 1940), tuberculous granuloma (Beheydt and VandenPutte, 1958), mycotic embolism (Layman and January, 1967), Chagas's disease (Moia, Rosenbaum, and Hojmen, 1955), fungal endocarditis (Mershon, Samuelson, and Layman, 1968), nonspecific myocarditis (Moreyra *et al.*, 1972), and cardiac sarcoidosis (Lull *et al.*, 1972). They may also be secondary to chest trauma (Killen *et al.*, 1969), or surgery (Wychulis *et al.*, 1971).

Left ventricular aneurysms in children are extremely unusual. They may be congenital (Paronetto and Strauss, 1963; Ruttenberg *et al.*, 1964; Treisman *et al.*, 1973), secondary to anomalous origin of the left coronary artery from the pulmonary trunk (Bland, White, and Garland, 1933), or result from rubella myocarditis (Van der Horst and Gotsman, 1970). Calcific apical

aneurysms occurring almost exclusively in Africans (Pocock *et al.*, 1965) have also been described in this age group.

It is the purpose of this communication to present three children with cardiomyopathies and angiographically proven left ventricular aneurysms.

Case reports

Case 1

A 14-year-old mentally retarded girl with no relevant past and family history was admitted to the hospital in severe congestive heart failure. She was put on bed rest, given diuretics and digitalis, but showed obvious intolerance to the latter, experiencing frequent arrhythmias. She had cardiomegaly and gallop rhythm. There were no murmurs. Laboratory studies were normal. A chest x-ray obtained at the time of the cardiac catheterization disclosed an enlarged heart with a cardiothoracic ratio of 0.54. The aortic knuckle was small and the pulmonary vasculature appeared normal. The electrocardiogram showed sinus rhythm alternating with episodes of atrial fibrillation. There was left atrial enlargement, left axis deviation ($\Delta QRS -30^\circ$), and diffuse ST-T abnormalities. Bradycardia, sinus arrests, second degree atrioventricular block, and frequent ventricular extrasystoles were seen while the girl was under treatment with digitalis.

A combined cardiac catheterization showed normal

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FIG. 1 Left ventricular cineangiography in the right anterior oblique projection, (a) in systole and (b) in diastole in Case 1. There is mitral regurgitation and a moderate-size aneurysm of the diaphragmatic wall.

intracardiac pressures except for the right ventricular end-diastolic and the right atrial mean pressures, which were raised (Table). The cardiac index was low at 2.4 l per min per m^2 b.s.a. Selective left ventricular cineangiography in the right anterior oblique projection disclosed extensive impairment of the left ventricular contractility, with an ejection fraction of 49 per cent. A medium size subvalvular aneurysm was present in the diaphragmatic wall as well as grade 2 mitral regurgitation (Fig. 1). The aorta and coronary arteries appeared normal in the retrograde aortogram.

The patient was discharged with the diagnosis of cardiomyopathy and maintained on medical treatment. Three months later she was readmitted in severe congestive heart failure unresponsive to treatment, dying shortly after. Necropsy was not permitted.

Case 2

A 20-month-old boy presented at the age of 20 days in congestive heart failure. The diagnosis of congenital heart disease was made, and thereafter he was maintained on digoxin, with a good clinical response. On physical examination he was well developed for his age. There was clinical evidence of cardiomegaly. Auscultation of the heart revealed a 4/6 ejection systolic murmur best heard along the left sternal border. There was a loud ejection sound present only on inspiration. The second heart sound was normally split, with a soft and delayed pulmonary component. Laboratory studies were normal. The chest x-ray showed cardiomegaly with a cardiothoracic ratio of 0.60. The apex of the heart was uptilted, and the pulmonary vasculature was within normal limits. The electrocardiogram (Fig. 2) had an $\bar{A}QRS$ axis of $+15^\circ$ in the frontal plane. The QRS width was 0.10 s. There were deep Q waves from V1 to V4. Tall and slurred R waves were present in the right precordial leads. Leads V5 and V6 showed increased voltage of the R waves and slurred S waves. In summary, the tracing

was compatible with incomplete right bundle-branch block, biventricular hypertrophy, and anteroseptal wall necrosis.

A combined cardiac catheterization was performed showing a 57 mmHg (7.6 kPa) gradient between the right ventricular inflow and outflow tracts (Table). The systolic pressure of the right ventricular sinus was close to systemic level. The cardiac index was normal at 5.3 l per min per m^2 b.s.a. No shunts were demonstrated by oximetry or cineangiography. Right ventricular cineangiography in the lateral view showed a round filling defect in this cavity encroaching on the outflow tract. The return circulation to the left heart showed filling of a large septal aneurysm of the left ventricle which corresponded to the filling defect visualized in the right ventricle (Fig. 3). The left ventricular cineangiogram in the right anterior oblique projection showed poor contractility (35% ejection fraction). A retrograde aortogram disclosed normal coronary arteries.

Two years after the initial study the child was re-examined. He complained of moderate fatigue and dyspnoea on effort. The auscultatory findings, electrocardiogram, and chest x-rays were essentially unchanged. A repeat cardiac catheterization showed a more severe obstruction to right ventricular outflow (80 mmHg (10.6 kPa) gradient) (Table). Left ventriculograms in both oblique projections showed again impaired left ventricular contractility (40% ejection fraction), and a large multiloculated aneurysm of the ventricular septum impinging in the right ventricular outflow tract.

Open heart surgery was performed shortly after the second haemodynamic study. The right ventricular cavity was partially occupied by a fibrous saccular aneurysm arising from the muscular septum which was 4 cm long and 2 cm wide. A membranous ventricular septal defect measuring 7 mm in diameter and hidden by the aneurysm was found. The defect was infracristal and was separated by 15 mm from the base of implanta-

FIG. 2 *Electrocardiogram of Case 2. There is evidence of left ventricular hypertrophy, incomplete right bundle branch block, and anteroseptal wall necrosis.*

tion of the aneurysm. The aneurysm was resected and the ventricular septal defect was closed with separate stitches. The patient made an uneventful recovery.

Pathological findings

The aneurysm was whitish in colour and dome shaped. The maximal length was 4 cm, and the thickness varied

between 2 and 3 mm. The surface was smooth on the convex side and slightly rugose on the opposite side. Microscopy showed that the muscular cardiac fibres were replaced by thick fibrous tissue with zones of hyalinization. There were scattered islets of myocardial fibres surrounded by the fibrous tissue. Inflammatory cells were not found.

Case 3

A 7-year-old well-developed girl with no relevant past and family history was asymptomatic up to the age of 6 years, when dyspnoea and fatigue were noted. Physical examination disclosed a heart murmur, and she was referred for investigation. There was clinical evidence of cardiac enlargement. The first heart sound was soft, and the second sound was slightly accentuated and normally split. A 2/6 ejection systolic murmur was heard along the left sternal border. Chest x-rays showed an enlarged heart with a cardiothoracic ratio of 0.56 and normal pulmonary vasculature. The electrocardiogram showed left ventricular hypertrophy and primary T wave abnormalities. There were abnormal Q waves in leads II, III, and aVF.

Cardiac catheterization showed normal pressures in both sides of the heart and no shunts (Table). The cardiac index was 4.8 l per min per m² b.s.a., and the maximal dP/dt was moderately decreased to 1260 mmHg/s (167.6 kPa/s). Selective left ventriculography in both oblique projections showed an enlarged left ventricle with diffuse hypokinesis and delayed emptying. A large, bilobed, crescent-shaped aneurysm arising from the left ventricular free wall, surrounded most of the ventricle (Fig. 4).

She was discharged on digitalis and six months later was restudied. The heart size had increased, and there were frequent ventricular premature beats. Left heart

FIG. 3 *The left heart-phase of the right ventricular angiogram in the lateral projection of Case 2 showing a large, multiloculated aneurysm of the ventricular septum.*

TABLE Haemodynamic data

	Pressures (mmHg)			
	Case 1 14 yr	Case 2 1 yr 8 mth	3 yr 8 mth	Case 3 7 yr
Right atrium	(10)	(4)	(3)	(3)
Right ventricle				
inflow	28/10	85/6	110/6	22/8
outflow		28/6	30/6	
Pulmonary artery	28/13 (20)	26/8 (15)	28/10 (15)	22/12 (9)
P A wedge	(11)	—	(5)	(5)
Pulmonary vein	—	(10)	(5)	—
Left atrium	—	(8)	(5)	—
Left ventricle	110/11	96/8	110/9	110/7
Aorta	100/80 (86)	98/68 (80)	105/60 (75)	110/65
Cardiac index (l/min per m ²)	2.37	5.35	4.91	4.8
Ejection fraction	49%	35%	40%	—

Numbers in brackets represent mean pressures. Left atrial and pulmonary vein pressures in Case 2 were obtained through a patent foramen ovale. Conversion factor from Traditional to SI units. 1 mmHg $\approx$ 0.133 kPa.

catheterization disclosed further decrease of the maximal dP/dt to 700 mmHg/s (93.1 kPa). The coronary arteries were selectively opacified and appeared completely normal.

Discussion

Left ventricular aneurysms are very frequent in adults owing to the high incidence of ischaemic heart disease. Other causes are rare. On the other hand, the occurrence of left ventricular aneurysms in children is extremely unusual.

Our first patient had the clinical features of a congestive cardiomyopathy. The obvious cardiomegaly with diffuse impairment of the left ventricular contractility favours the generalized involvement of the myocardium. The location of the aneurysm in the diaphragmatic wall indicates that this was the area of maximal weakness. We have recently seen three adults with similar aneurysms in the presence of normal coronary arteriograms (Moreyra *et al.*, 1972).

The second patient of our series is unique because the aneurysm involving the muscular ventricular septum produced severe subpulmonary stenosis. The paradoxical movement of the aneurysm during systole made it protrude into the right ventricular outflow tract. This type of obstruction to right ventricular outflow is similar to that found in some cases of aneurysms of the membranous ventricular septum (Das, Jahnke, and Walker, 1964), which often accompany small ventricular defects. In our patient there was also a membranous defect which had not been evident until surgery despite two haemodynamic studies and selective left ventricular cineangiograms previously performed. It appears that the defect was occluded by the redundant aneurysmal sac arising from the muscular septum.

This child's electrocardiogram showed antero-septal wall necrosis. Coronary arterial anomalies were ruled out by retrograde aortography. A case of myocarditis caused by neonatal rubella showing an infarct pattern in the electrocardiogram and complicated by a left ventricular aneurysm has been reported by Van der Horst and Gotsman (1970). Our patient had no rubella stigmata but other intrauterine viral infection cannot be excluded. A

FIG. 4 Left ventriculography in the right anterior oblique projection of Case 3. A crescent-shaped bilobed aneurysm encircling the ventricle is clearly seen. The left ventricular cavity is huge.

myocardial infarct with normal coronary arteries, which is being recognized with increasing frequency in young adults (Khan and Haywood, 1974), is another possibility, though unlikely. Histological studies of the resected aneurysm showed that this was a fibrous structure with scattered myocardial fibres. Inflammatory cells were not present. However, since more than three years had elapsed from birth a myocarditis causing the original lesion could not be ruled out.

The last patient of our series also had evidence of myocardial disease with impaired left ventricular function. The electrocardiogram showed inferior wall necrosis and left ventricular hypertrophy. In this case the aneurysm had a very unusual crescent shape surrounding most of the left ventricular cavity. The coronary arteries were normal. To the best of our knowledge there are only two apparently similar cases reported. Paronetto and Strauss (1963) and Ruttenberg *et al.* (1964) found loculations and septations within the left ventricle corresponding to areas of myocardial wall hypoplasia and bridges formed by hypertrophied myocardial fibres. Their cases were probably congenital in origin, as we think is ours.

The three patients of this series are examples of cardiomyopathy of unknown origin complicated by left ventricular aneurysms. The presence of areas of localized weakness of the affected myocardium is the possible mechanism for aneurysm formation. The clinical features of the cardiomyopathy were not altered by the presence of the aneurysm except in the patient with subpulmonary stenosis caused by the aneurysm of the ventricular septum. In this child surgical resection of the aneurysm was carried out successfully.

References

- Beheydt, P., and VandenPutte, M. (1958). L'anevrisme ventriculaire d'origine tuberculeuse chez des jeunes africains. *Acta Cardiologica*, **13**, 419.
- Bland, E. F., White, P. D., and Garland, J. (1933). Congenital anomalies of coronary arteries: report of an unusual case associated with cardiac hypertrophy. *American Heart Journal*, **8**, 787.
- Braunstein, A. L., Bass, L. B., and Thomas, S. (1940). Gummatous myocarditis and aneurysm of the left ventricle with nodal tachycardia, and subsequently bundle branch block. *American Heart Journal*, **19**, 513.
- Cheng, T. O. (1971). Incidence of ventricular aneurysm in coronary artery disease. *American Journal of Medicine*, **50**, 340.
- Das, S. K., Jahnke, E. J., and Walker, W. J. (1964). Aneurysm of the membranous septum with interventricular septal defect producing right ventricular outflow obstruction. *Circulation*, **30**, 429.
- Khan, A. H., and Haywood, L. J. (1974). Myocardial infarction in nine patients with radiologically patent coronary arteries. *New England Journal of Medicine*, **291**, 427.
- Killen, D. A., Gobbell, W. G., France, R., and Vix, V. A. (1969). Post-traumatic aneurysm of the left ventricle. *Circulation*, **39**, 101.
- Layman, T. E., and January, L. E. (1967). Mycotic left ventricular aneurysm involving the fibrous atrioventricular body. *American Journal of Cardiology*, **20**, 423.
- Lull, R. J., Dunn, B. E., Gregoratos, G., Cox, W. A., and Fisher, G. W. (1972). Ventricular aneurysm due to cardiac sarcoidosis with surgical cure of refractory ventricular tachycardia. *American Journal of Cardiology*, **30**, 282.
- Mershon, J. C., Samuelson, D. R., and Layman, T. E. (1968). Left ventricular 'fibrous body' aneurysm caused by aspergillus endocarditis. *American Journal of Cardiology*, **22**, 281.
- Moia, B., Rosenbaum, M. B., and Hojman, D. (1955). Aneurismas ventriculares en la miocarditis crónica chagásica. *Revista Argentina de Cardiología*, **22**, 113.
- Moreyra, E., Alday, L. E., Amuchástegui, L. M., Conde, L., and Quiroga Mayor, C. (1972). Infarto de miocardio y aneurismas ventriculares con coronariografía normal. *Revista Médica de Córdoba*, **60**, 198.
- Paronetto, F., and Strauss, L. (1963). Aneurysm of the left ventricle due to congenital muscle defect in an infant. *American Journal of Cardiology*, **12**, 721.
- Pocock, W. A., Cockshott, W. P., Ball, P. J. A., and Steiner, R. E. (1965). Left ventricular aneurysms of uncertain aetiology. *British Heart Journal*, **27**, 184.
- Ruttenberg, H. D., Jue, K. L., Elliott, L. P., Anderson, R. C., and Edwards, J. E. (1964). Cardiac myopathy, probably of congenital origin. A case simulating anomalous origin of the left coronary artery from the pulmonary trunk. *Circulation*, **29**, 768.
- Treisman, B., Cooley, D. A., Lufschanowski, R., and Leachman, R. D. (1973). Diverticulum or aneurysm of left ventricle. *American Journal of Cardiology*, **32**, 119.
- Van der Horst, R. L., and Gotsman, M. S. (1970). Left ventricular aneurysm in rubella heart disease. *American Journal of Diseases of Childhood*, **120**, 248.
- Wychulis, A. R., Frye, R. L., Kincaid, O. W., Ongley, P. A., and Ellis, F. H., Jr. (1971). Postventriculotomy aneurysm in patients with idiopathic hypertrophic subaortic stenosis. *American Journal of Cardiology*, **27**, 322.

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