Chronic pericardial constriction with effusion in childhood

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SUMMARY Five cases of children with nontuberculosis pericardial constriction with effusion are reported. The disease is not as rare as had been thought, and the aetiology in these cases was probably a previous virus infection. A previous diagnosis of hepatitis, nephrotic syndrome, or protein-losing enteropathy had been made and the correct diagnosis was delayed for months or even years. Pericardiectomy produced immediate relief from symptoms in 2 patients but in 2 others there was evidence of poor myocardial function postoperatively.

Constrictive pericarditis is rare, particularly in children. Watson (1968) estimated that: 'The disease represents not more than one in 10 000 cases of cardiac disease presenting in childhood in North America at present, but it may play a larger role in other countries particularly where tuberculosis is more prevalent'. The true incidence of the disease cannot accurately be given and it may be a good deal more common than Watson thought, as it is likely that some cases are asymptomatic and others are misdiagnosed. Five cases were seen in children during a 6-year period at the Bristol Royal Hospital, where about 300 new children with cardiac disease are seen annually, an incidence of 1: 360 such children.

Case reports

Case 1.

History

A girl, born at 38 weeks' gestation birthweight 1820 g, was described as 'always a poor feeder from birth'. At 18 months mild lactose intolerance was diagnosed. She was referred here at 7 years with weight loss and increasing abdominal distension.

Examination

She was small and wasted (height 111 cm, weight 18.1 kg) with conspicuous abdominal distension due to ascites and hepatomegaly. The peripheral circulation was poor, the pulse small volume with

obvious reduction in volume in inspiration, 'pulsus paradoxus'. The venous pressure was raised beyond the angle of the jaw. The cardiac impulse was faint and a very loud early diastolic sound was present.

Investigations

Chest x-ray showed an increased cardiothoracic ratio, the heart being visible as a separate shadow within the pericardium on one overpenetrated film. The presence of an effusion was confirmed by ultrasound examination. Mantoux test negative. Viral studies, including culture of ascitic fluid, negative.

Treatment

At operation (Mr R. H. R. Belsey), both parietal and visceral pericardium were considerably thickened in a uniform fashion and a sizeable effusion was found. The parietal pericardium was removed from the major part of both ventricles, as well as most of the visceral pericardium. Histology showed thickening and fibrosis, with moderate round cell infiltration.

Progress

Although initially she required antifailure treatment, 8 months later our patient was well and the heart was clinically and radiologically normal.

Case 2.

History

A girl was transferred to this hospital for investigation of hepatomegaly and ascites. She had been born by normal delivery at term. At 6 months she was noted to have bilateral interstitial keratitis. At age

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10 months she was admitted for investigation of abdominal swelling.

Examination

Obvious ascites, enlargement of the liver and spleen. Loud early diastolic sound.

Investigations

Chest x-ray showed considerable cardiomegaly and the heart shape was consistent with the presence of a pericardial effusion (Fig. 1). Tuberculin test consistently negative. Viral studies negative. Abdominal paracentesis on two occasions showed straw-coloured fluid. Transpleural liver biopsy was normal histologically. Her condition deteriorated after this procedure, and she developed pulsus paradoxus; cardiac tamponade was diagnosed. Pericardiocentesis was therefore performed, 150 ml greenish fluid being obtained, but the signs of tamponade recurred during the next 2 weeks.

Operation (Mr R. H. R. Belsey)

The heart was found to be surrounded by a densely adherent, grossly thickened visceral pericardium which was severely impeding cardiac filling. The parietal pericardium was thickened also, and was removed in its entirety, large portions of the visceral pericardium being dissected off the ventricles and

removed also. Histologically the excised tissue showed dense fibrosis with scanty chronic inflammatory cells.

Progress

During the next 7 days large volumes of fluid drained from the left chest drain, but this gradually tailed off and thereafter her postoperative recovery was rapid and uneventful with prompt disappearance of all signs of impaired right heart filling. Five years later she remains perfectly well and the cardiovascular system appears normal.

Case 3.

History

A girl was noted at age 2 years to be breathless and cyanosed on exertion. Medical advice was not sought at that time and she presented at age 8 years with effort dyspnoea and occasional right-sided chest pain on exertion.

Examination

Slight central cyanosis. Small volume pulses. Jugular venous pressure greatly raised, with prominent A- and V-waves. No abnormalities were detected on auscultation. Liver palpable 3 cm below the right costal margin.

Investigations

Chest x-ray showed gross cardiomegaly with prominence of the superior vena cava and of the veins of the upper lobes of the lungs. Multiple opacities and septal lines were present at the bases, with a small right-sided pleural effusion. Tuberculin test negative. Antibodies to Q fever, psittacosis, *Mycoplasma*, mumps, and *Toxoplasma gondii* negative. Viral culture in HeLa and monkey kidney cells negative. Cardiac catheterisation (Table).

Operation

A pericardiectomy was performed (Mr G. Keen). The pericardium looked thickened and there was a moderate accumulation of pericardial fluid. Histologically the pericardium was moderately thickened but it was otherwise normal.

Fig. 1 Case 2 x-ray. Cardiac shadow is enlarged but a faint medial opacity due to the heart itself is also shown.

Table Cardiac catheterisation

Case	Right atrium (mmHg)		Right ventricle (mmHg)	Pulmonary artery (mmHg)		Wedge (mmHg)	Left ventricle (mmHg)	A-V O ₂ difference	
	A-wave	Mean		S/D	Mean			%	O ₂ content
3	36	28	100/12/25	100/65	80	26	100/12/25	54	9.6
4	9	8	35/ 6/ 8	33/12	18	12	—	—	—
5	30	25	42/15/25	40/30	32	26	—	39	6.3

Progress

Jugular venous pressure fell almost to normal and she was symptomatically greatly improved, but continued to show mild cardiomegaly and abnormal T-waves in the ECG. She died suddenly 2 years later, presumably from an arrhythmia.

Case 4.**History**

A girl born by normal delivery at 38 weeks' gestation, birthweight 3200 g. She was admitted to hospital at age 2 months for investigation of convulsions. A chest x-ray showed a cardiothoracic ratio of 0.6, and an ultrasound cardiogram showed a moderate sized pericardial effusion and a small or normal sized heart.

Progress

Radiological signs of pericardial effusion remained for 2 years but then gradually subsided and our patient has since remained well.

Case 5.**History**

A 2½-year-old girl was referred to this hospital with a one-year history of lethargy, oedema, and abdominal swelling. She was seen at another hospital and found to have albuminuria, and a tentative diagnosis of nephrotic syndrome was made; later this was changed to active hepatitis and then to protein-losing enteropathy.

Examination

Oedema of legs and face and moderate ascites. Liver 10 cm below the costal margin, jugular veins engorged with no obvious pulsation. A loud, early diastolic sound was audible all over the praecordium.

Investigations

Chest x-rays showed an enlarged cardiac shadow. ECG showed right and left atrial hypertrophy, small voltage QRS complexes, and flat T-waves (Fig. 2). Cardiac catheterisation (Table) showed findings

Fig. 2 Preoperative electrocardiograms of the 5 patients.

consistent with pericardial constriction, there being a pronounced 'dip and plateau' pattern in the right ventricular trace.

Operation (Mr R. H. R. Belsey)

The parietal pericardium was mildly thickened and distended by a large straw-coloured effusion. There was no adhesion of visceral and parietal pericardium. The pericardium was excised between the phrenic nerves. It was uniformly thickened with moderate numbers of chronic inflammatory cells.

Progress

Postoperatively the patient continued to drain moderate quantities of fluid for 7 days (1.6 litres) through the left pleural drain. After removal of the drain, fluid reaccumulated in the left chest and a further 2.8 litres were removed, by repeated paracentesis, but eventually the effusion was controlled by frusemide 20 mg daily. During the next 6 weeks the dose was reduced and she remained well with a normal venous pressure and no abnormal signs. Four months after operation the venous pressure was again raised. The cardiac size on x-ray was within normal limits. Repeat cardiac catheterisation still showed equally raised right atrial and pulmonary wedge pressures. The chest was re-explored to exclude recurrent constriction but none was found. Six months later she still required a moderate dose of diuretic (10 mg frusemide daily) to keep her out of failure, and this has continued to do so to date.

Discussion

Aetiology. The 5 cases described have in common the presence of persistent pericardial effusion. This was associated with impaired heart filling in 4 and proved thickening of the pericardium in 3. The aetiology is obscure in all, although in 2 of the children there were associated features which support the suggestion of viral infection. Presumptive evidence for a virus aetiology to explain 'idiopathic' pericardial constriction was produced by Robertson and Arnold (1962) who found a cluster of about 5 such cases a year after an epidemic of virus pericarditis: 2 of their patients had had pericarditis during the epidemic (caused by Coxsackie B5 virus). In our Case 2 the coexistence of interstitial keratitis with negative serology for syphilis, and in Case 1 the low birthweight for gestational age and poor progress from birth, would be consistent with this possibility.

It was usually assumed, before the introduction of effective antituberculous chemotherapy, that tuberculosis accounted for most of the cases diagnosed as constrictive pericarditis. Simcha and Taylor (1971)

stated: 'Tuberculosis plays a major role in the aetiology of constrictive pericarditis; strong evidence of this is found in this series'. This claim seems difficult to justify on the basis of their paper since in only one of 5 cases was a diagnosis of tuberculosis well established, and the evidence adduced in favour of this disease in 2 other cases was not convincing. Indeed, some doubt existed about the importance of tuberculosis as the main cause of constrictive pericarditis even before the introduction of streptomycin; of 37 cases (all children) reviewed by Rothstein (1934) tuberculosis was given as the diagnosis in only 9, with one doubtful, the remainder being attributed to rheumatism or 'unknown' in roughly equal proportions. Other authors (Mortensen and Warburg, 1948; Kuipers, 1955; Vining, 1955; Dalton *et al.*, 1956; Shea *et al.*, 1957; Caddell *et al.*, 1960) have presented small series of children, or included the occasional child in series of adults, but in these papers tuberculosis did not feature prominently as a proved cause. Even in countries where tuberculosis is common it probably only accounts for about half the number of cases (Das *et al.*, 1973).

Apart from tuberculosis, a variety of other disease processes have occasionally been found to cause constrictive pericarditis; these include histoplasmosis, Coxsackie pericarditis (particularly strains B3 and B5), traumatic haemopericardium, rheumatoid disease, hydatid disease, myxoedema, amyloidosis, sepsis (for example meningococcal and staphylococcal septicaemia), and malignant disease. Practically all accounts agree, however, that in any sizeable series the majority of cases occur in the absence of any identifiable primary disease, and the known causes, even collectively, constitute the minority (*British Medical Journal*, 1968; Keith *et al.*, 1968; Watson, 1968; Mullen *et al.*, 1969).

The role of acute rheumatism, if any, in the aetiology of constrictive pericarditis is not clear. Simcha and Taylor (1971) regarded it as hardly ever leading to constrictive pericarditis, while Rothstein (1934) attributed 13 of 37 cases to this cause.

Our 5 cases had moderately large, serous effusions, separating parietal and visceral pericardium. This is not generally a feature of tuberculous constrictive pericarditis, but is in keeping with the findings of Harrold (1968) in 3 adults with non-tuberculous pericardial constriction. Cayler *et al.* (1963) found moderately large pericardial effusions in their patients, but each one presented during the course of acute pericarditis.

Diagnosis. Although 2 of our patients presented with short histories and the diagnosis was suspected before referral, the other 3 were clearly undiagnosed

for periods of several months or years. The failure to diagnose the presence of pericardial constriction was partly due to failure to notice or realise the significance of the raised venous pressure. If the venous pressure is so high that there is no longer obvious venous pulsation in the neck, the difficulties of assessment are well known. Another factor in the failure to make a diagnosis was the presence of secondary effects of the high venous pressure. Hepatic enlargement with abnormal liver function tests led to a diagnosis of active hepatitis and liver biopsy was performed (a procedure not without risk when the venous pressure is high). Similarly the effect of high venous pressure in the kidneys may cause a secondary nephrotic syndrome. Our Case 5 was similar in this respect to a 58-year-old patient reported by Pastor and Cahn (1960). Case 5 was initially diagnosed as having protein-losing enteropathy and this too has been reported as a complication of pericardial constriction (Plauth *et al.*, 1964).

The auscultatory signs in pericardial constriction are not obvious. Three of our patients had a loud, early diastolic sound, but this is often so early that it is mistaken for the second sound. The ECG is not specific and patients with only a small effusion, or none, may have a remarkably normal looking cardiac size and shape on chest x-rays. Diagnostic ultrasound (Feigenbaum *et al.*, 1967) is useful when there is a pericardial effusion.

Treatment. There were clear indications in 4 of our patients for surgical treatment. In one of them treatment was required fairly urgently for rapidly increasing signs of right heart failure and reduced cardiac output. In the other 3 there were obvious indications from the secondary effects of high venous pressure on the liver, kidneys, and gut. The surgeon's aim is to remove the thickened pericardium (visceral as well as parietal if necessary) from more than half the surface of the heart so that any further contraction of the remaining pericardium does not constrict the heart. In 2 of the operated patients the pericardial effusion separated parietal and visceral pericardium. In the other 2 a variable amount of dissection of pericardium from the heart itself was necessary. The exudative nature of the pericarditis is well shown by the 2 patients (Cases 2 and 5) who continued to drain fluid for several days or weeks after pericardiectomy. This was a surprising feature in view of the almost complete lack of signs of inflammation microscopically in the excised pericardium.

Myocardial involvement. Two of our patients (Cases 3 and 5) probably had chronic myocardial damage and Case 1 also showed temporary cardiac

failure in the postoperative period. The possible reasons for myocardial involvement were reviewed by Levine (1973). Possible explanations are associated myocardial fibrosis (particularly if a viral aetiology is suspected), and the splinting effect of the constriction preventing the heart from dilating adequately.

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Change of reference style

From January 1980, the *Archives* will change its style for citing references and will follow the number system. We are making this change in order to conform with the 'Vancouver style' of uniform requirements for manuscripts submitted to biomedical journals.

Previously the *Archives* has used the Harvard reference system. Its advantages and disadvantages compared with those of the number system have seemed evenly balanced, but the editors have been aware of the burden imposed on authors and their secretaries by the different styles which different journals require. Therefore, with the unanimous agreement of the editorial committee, we have decided to support a move whereby a large—and increasing—number of major medical journals

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¹ International Steering Committee of Medical Editors. Uniform requirements for manuscripts submitted to biomedical journals. *Br Med J* 1979; **1**: 533-535.

² Lancet. The Vancouver style. Uniform requirements for manuscripts submitted to biomedical journals. *Lancet* 1979; **1**: 429-430.