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. Pericardietectomy 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
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 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 pericardectomy 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.

Table

<table>
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<th>Right ventricle (mmHg)</th>
<th>Pulmonary artery (mmHg)</th>
<th>Wedge (mmHg)</th>
<th>Left ventricle (mmHg)</th>
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Fig. 1 Case 2 x-ray. Cardiac shadow is enlarged but a faint medial opacity due to the heart itself is also shown.
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 cardogram 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 2 1/2-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 precordium.

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 moder-
ate 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 con-
striction 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 tuber-
culosiss 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 (Morten-
sen 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 pro-
minently 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 histo-
plasmosis, Coxsackie pericarditis (particularly strains
B3 and B5), traumatic haemopericardium, rheuma-
toid 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

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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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References


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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 will accept manuscripts presented in one agreed style.

The system numbers references consecutively in the order in which they are first mentioned in the text. References are identified in the text by arabic numerals. For further details see instructions to authors inside the front cover, and the references cited.¹,²

From now onwards all manuscripts submitted should accord with the new style.