Cardiol Young 2001; 11: 210–213

ISSN 1047-9511

Original Article

Constrictive chronic pericarditis in children

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Abstract Constrictive pericarditis is a uncommom disease in children. We have now encountered pericardial thickening as the cause of severe constrictive physiology in two patients, one also having haemodynamic features of restrictive cardiomyopathy. Both patients, who had refractory ascites and evidence of increased systemic venous pressure, underwent Doppler echocardiography, cardiac catheterisation, and magnetic resonance imaging. Resonance imaging failed to show any thickning of the pericardium, but cardiac catheterisation revealed diastolic equalisation of pressures in all four chambers, with only mild elevation of pulmonary pressure in the first patient, but nearly equalisation of diastolic pressure, and a very high pulmonary arterial pressure with a difference of 7 mm Hg between the end diastolic pressures in the two ventricles in the second patient. Doppler revealed a restrictive pattern of mitral inflow, with high E and small A velocities and a short deceleration time. The clinical background did not suggest pericardial disease in either of the patients. We conclude that a careful search is needed to uncover constrictive pericarditis when there is no previous disease which may suggest late pericardial constriction. The haemodynamic features of restrictive cardiomyopathy can co-exist with pericardial restriction, and differentiation between the two entities is critical in view of the diverse management and prognosis of the two conditions.

Keywords: restrictive pericarditis, pericardial disease.

PERICARDIAL CONSTRICTION IS USUALLY THE result of a chronic inflammatory process involving the parietal and visceral pericardial layers. The disease has a polymorphic clinical presentation, and has been described mimicking extrinsic compression of the heart and great vessels,[1,2,3] mitral,[4] and tricuspid[5] stenosis. Recently, Henein et al[6] have described two cases having haemodynamic features of restriction producing constriction with the potential presence of restrictive cardiomyopathy. Pericardial constriction is an insidious and debilitating disease, seen more frequently in adults. Its cause is usually idiopathic.[7] During childhood, pericardial diseases are typically of as acute nature, with pericardial effusion occurring due to an infective process[8] or connective tissue disorders.[9,10]

Here, we report two children with idiopathic constrictive pericarditis, in one of whom there was evidence of restrictive physiology on cardiac catheterisation that required an exploratory thoracotomy to ensure that the cause of restriction was in the pericardium itself.

Case 1

A 12 year old girl with severe ascites and cachexia was admitted to a large public hospital in Belém, in January 1998. She was treated with corticosteroids and diuretics, presumably for hepatic disease, but was discharged with no clinical improvement. In October 1998, she was admitted to our service, when we made a diagnosis of constrictive pericarditis based on the chest radiography, which

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Accepted for publication 28 September 2000

showed calcification of the pericardium. Clinical examination revealed blood pressure of 90/70 mm Hg, cardiac rate of 80 beats per minute, weight of 23.900 Kg, increased jugular venous pressure, a pronounced impulse at the left parasternal border, normal cardiac sounds, huge ascites, but neither leg nor ankle oedema.

The electrocardiogram showed sinus rhythm, normal QRS axis, and inversion of the anteroseptal T waves. Chest radiography showed an abnormal cardiothoracic ratio, with pericardial calcification seen in the posteroanterior and lateral views (Fig. 1). An echocardiogram revealed cavities of normal size, mild tricuspid and moderate mitral regurgitation, but no collection of any pericardial fluid. Continuous wave Doppler examination of the mitral flow showed a peak transmitral velocity of 0.7 m/s with deceleration time of 110 ms with no A wave (Fig. 2). Cardiac catheterisation showed diastolic equalisation of pressure in all four chambers. Resonance imaging failed to show any thickening of the pericardium.

A clinical diagnosis of significant restrictive pericardial disease was made, and the patient was referred for pericardiectomy. The pericardium was found to be thickened and so adherent to the heart in the inferior area that marked haemorrhage followed attempted resection. Because of this, it was not possible to excise all the pericardium. The postoperative course was complicated by hypotension, although the congestive symptoms regressed soon after surgery. She was discharged nine days later and remained on diuretics, digoxin and captopril. Five months subsequently, all ascites and hepatomegaly had resolved, she had gained weight, and it proved possible to discontinue the anticongestive drugs.

Case 2

A 12-year-old boy came to our cardiology service for investigation of cardiomyopathy. Five years previously, he had developed ascites and hepatomegaly. He was investigated by our hepatologists, when liver biopsy failed to show schistosomiasis or any other disease. He was treated with spironolactone and furosemide, albeit with no clinical improvements. During this period, he was hospitalized on several occasions. As he did not present with classical symptoms of cardiac disease, except for the presence of refractory ascites, he was re-admitted to investigate the possiblility of cardiac disease. On the latest admittance his weight was 29.300 Kg, blood pressure was 90/65 mm Hg, and he was mildly breathless with hepatomegaly and increased jugular venous pressure.

The electrocardiogram showed sinus rhythm, a normal QRS axis and flat T waves throughout the tracing. Previous chest radiography did not show calcification, but on this latest hospitalisation pericardial thickening was seen in the lateral view (Fig. 3). Resonance imaging failed to show any abnormalities. An echocardiogram showed normal ventricular size, and mild dilatation of the atriums with moderate dilation of inferior caval vein and the hepatic veins. In addition, there was diastolic regurgitation across the mitral valve, and a deceleration time of 80 m/s (Fig. 4). There was abnormal diastolic filling, with an abrupt stop which was suggestive of constrictive pericarditis. Cardiac catheterisation showed that the diastolic pressures were almost equalised and provided evidence of significant pulmonary hypertension (Table 1).

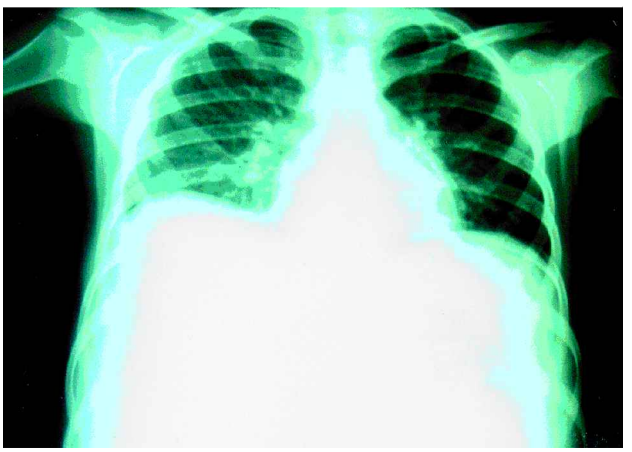

Figure 1.
Case 1 – Chest radiography (posteroanterior view) showing pericardial calcification.

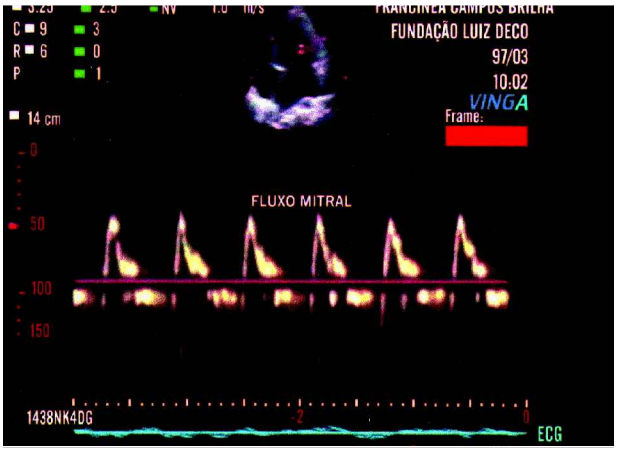

Figure 2.
Case 2 – Transmitral Doppler flow showing a typical restrictive filling pattern.

https://doi.org/10.1017/S1047951101000130 Published online by Cambridge University Press

Table 1. Haemodynamics Data (mm Hg)

	Case 1	Case 2
Right atrium (mean)	32	35
Right Ventricle	37/32	85/35
Pulmonary arteries	37/32	85/60
Wedge pressure (mean)	32	42
Left ventricle	92/32	112/42
Aorta	92/74	112/86

On the basis of the haemodynamic data, the patient was referred for surgical exploration through an anterior incision. The pericardium was found to be thick and heavily calcified. The incision was thus enlarged to permit total pericardiectomy and a myocardial biopsy was performed. The pericardium was successfully removed and, macroscopically, it had a stony consistence. The microscopic examination of the resected specimen showed dense connective tissue with extensive areas of hyalinasation with fibrous and calcific deposits (Fig. 5). The posoperative course was uneventful, so that the patient was discharged seven days after operation.

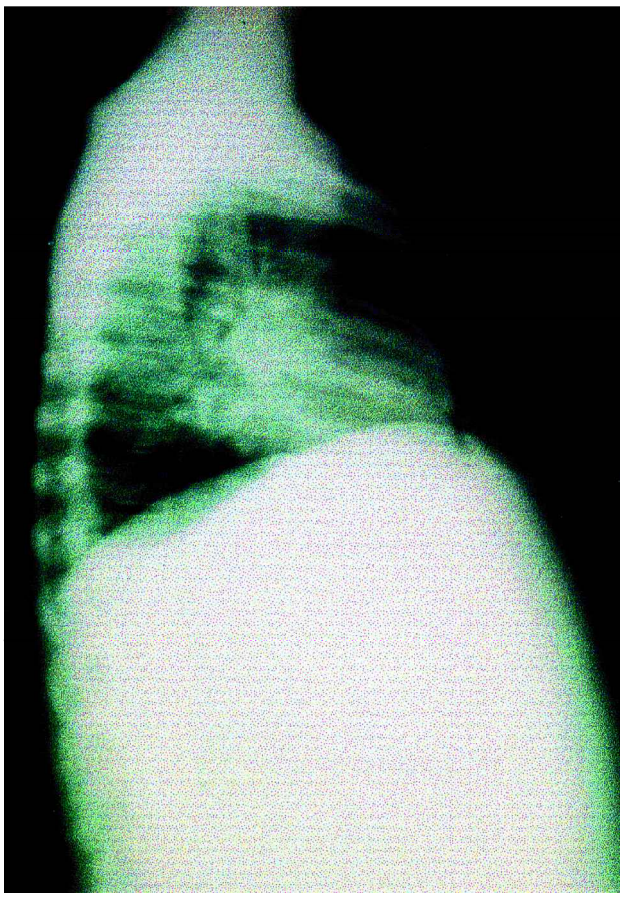

Figure 3.
Case 2 – Chest radiography (lateral view) showing pericardial calcification.

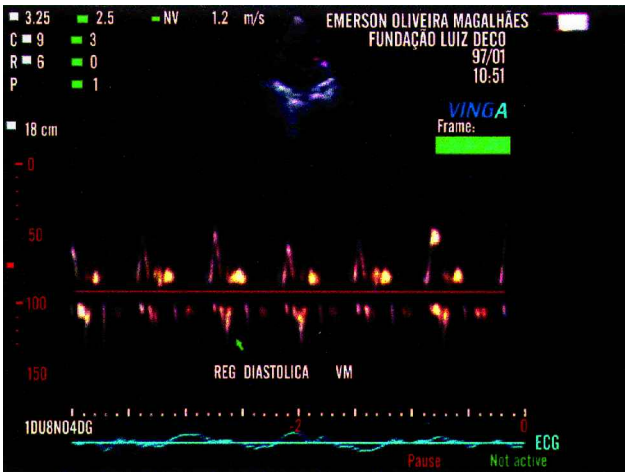

Figure 4.
Case 2 – showing a short deceleration time with minimal respiratory variation and diastolic regurgitation (arrow).

Two months later, it proved possible to discontinue anticongestive drugs and he remains symptom free.

Discussion

It has now been established[11] that there are haemodynamic criterions which discriminate pericardial constriction from restrictive myocardial disease. Thus, it is suggested that, to make the diagnosis of constrictive pericarditis, there should be a difference of greater than 5 mm Hg between the end diastolic pressure measured directly in the two ventricles, a peak right ventricular pressure of less than 50 mm Hg, and a ratio of right ventricular end diastolic peak pressure of more than 0.33. In our first case, all these criterions were fulfilled. In the second case, however, the two first criterions were not met. Furthermore, significant pulmonary hypertension is not part of the known haemodynamic features of constrictive pericarditis,[6,12] instead being suggestive for restrictive cardiomyopathy. The finding of a peak right ventricular pressure of 85 mmHg lead us to search for concomitant myocardial disease, which could not be confirmed by myocardial biopsy. The haemodynamic profile of our cases is similar to those described by Henein et al.,[6] who reported striking evidence of restrictive physiology in their two cases where the restriction was localized in the pericardium. They claimed that these cases should be considered as a distinct diagnostic entity, since the pathological basis and treatment are different from those needed for intrinsic myocardial disease.

Preoperatively, the clinical state has been shown to be predictive of operative risk, with the mortality rate increasing to 46% among patients in functional class IV[13] of the classification of the New

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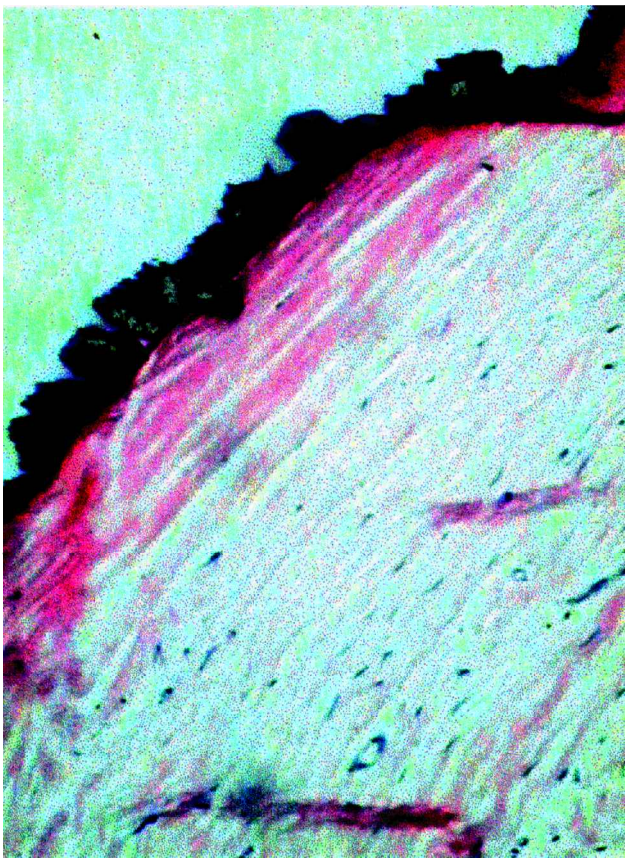

Figure 5.
Histology showing dense fibrosis and hyalinisation (red) and deposits of calcium (black).

York Heart Association. In our two cases, although neither patients had dyspnoea as a major symptom, the huge ascites and cachexia were features that permitted us to consider them in functional class IV. Nevertheless, both had rapid clinical improvement after surgery.

The aetiologic diagnosis of pericarditis is difficult, even when dealing with acute episodes characterized by pericardial effusion. A recent analysis of 38 patients has shown the limited value of pericardial biopsy as a complementary tool in establishing the aetiology of pericardial effusion.[14] In only one-tenth of cases was the biopsy able to define the aetiology. Non-specific changes were seen in the other patients. We were not able to retain the specimen from surgery in our first case, although the initial examination revealed that the pericardium took the form of a fibrous carapace similar to that seen in the second patient, as described by the surgeon who carried out both procedures. Microscopic examination in our second patient revealed a chronic and non-specific inflammatory process.

Total pericardiectomy is the appropiate surgical procedure in these cases, although the pericardium could not fully be resected in our first patient. Despite this, the patient did well after surgery.

We conclude that children with restrictive ventricular physiology must also be investigated for constrictive pericarditis in suspicious cases, even when there is no previous evidence of pericardial involvement. As we found, even when haemodynamic findings suggest myocardial restriction, successful pericardiectomy is able to achieve complete cure of the disease.

Acknowledgements

We are grateful to Clínica "Rx-Lobo" and Laboratório "Amaral Costa" for their invaluable collaboration and Superintendência do Desenvolvimento da Amazônia (SUDAM) for their support of this work.

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