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Variability in the Clinical Manifestations of Constrictive Pericarditis

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SUMMARY

Jaiyesimi F, Aluko O, Omokhodion S and Adebo OA. Variability in the Clinical Manifestations of Constrictive Pericarditis. *Nigerian Journal of Paediatrics,* **1989; 15:0.** The variability in the clinical manifestations of constrictive pericarditis is described with the aid of 5 case illustrations. The first case, a boy who progressed from acute staphylococcal pericarditis to constrictive pericarditis within the space of 3 months, illustrates the difficulties in distinguishing between infective myocarditis and acute pericardial constriction. The second case simulated mitral stenosis while the third manifested physical and radiological signs that were suggestive of subvalvar left ventricular aneurysm. The fourth case mimicked right ventricular endomyocardial fibrosis in terms of clinical, radiologic and angiocardiologic features, and the correct diagnosis was made only at autopsy. The fifth patient presented with massive ascites and hepatomegaly, and was initially suspected to have liver cirrhosis. Owing partly to the variability in its manifestations, there is often a delay in the diagnosis of constrictive pericarditis; and in the present series, the time lapse between onset of illness and diagnosis, ranged from 3 months to 8 years. We have therefore, suggested clues to the early recognition of the disease.

Introduction

CONSTRICTIVE pericarditis is relatively uncommon in children. A previous report from our centre[1] included only 7 cases of pericardial constriction in a series of 53 children with pericardial disease. All but one of the 10 cases studied by Adebo *et al*,[2] were aged above 15 years; and less than a third of the 27 pateints reported by Mabogunje *et al*,[3] were aged below 20 years. The prevalence of the disease in technically developed countries is even much lower.[4] [5] Experience with childhood constrictive pericarditis is therefore, limited in most centres. Furthermore, considerable variability exists in the clinical manifestations of the disease[6], and this further militates against early diagnosis and treatment. The present commu-

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nication aims to facilitate timely diagnosis of constrictive pericarditis by drawing attention to its protean manifestations.

Materials and Methods

In the 5 – year period, 1982 – 86, six children with constrictive pericarditis received care in the Paediatric Cardiology Unit, University College Hospital (UCH), Ibadan. Five of the 6 patients illustrated the variability in the clinical manifestations of pericardial constriction, and have therefore, been selected for reporting. In addition to detailed clinical evaluation and general investigations, the nature and scope of which were determined by the associated diseases, each of the patients had standard chest radiographs and conventional 12-lead electrocardiograph (ECG). Four of them had cardiac catheterization and angiocardiography.

Case Illustrations

The main features in the patients are summarized in Table I and described below.

TABLE I

Clinical Data on 5 Children with Constrictive Pericarditis

	Case 1	*Case 2*	*Case 3*	*Case 4*	*Case 5*
Age (Year)/Sex	12; M	11; M	15; M	11; M	10; M
Main symptoms and signs	Antecedent purulent pericarditis; jugular venous distension; persistent heart failure	Orthopnoea; paroxysmal nocturnal dyspnoea; jugular venous distension; pericardial knock; mitral diastolic murmur; heart failure	Abdominal swelling; cough; dyspnoea; Proptosis, jugular venous distension; apical heave; Normal pulses, mitral incompetence; pericardial knock; Heart failure.	Hepatomegaly initially, followed later by heart failure and tricuspid incompetence	Proptosis; Jugular venous distension; Pericardial knock, Hepatomegaly; massive ascites; Heart failure.
Radiological findings	Cardiac enlargement; pulmonary venous congestion	Small heart; pericardial calcification	Cardiac enlargement; bulging left heart border; pericardial calcification	Mild cardiac enlargement	Normal-sized heart; irregular left heart boarder; pericardial calcification
ECG	Low amplitude QRS, Inverted T-waves	Same as in Case 1	Biphasic P-waves in VI, high-normal QRS complexes, flat T-waves in V6.	Small amplitude QRS, flat T-waves, Tall and broad P-waves	Broad P-waves, flat T-waves, flat T-waves.
Time lapse before accurate diagnosis	3 months	uncertain	8 years	4 years	2 years
Other diagnosis made/ considered initially	Bacterial myocarditis	Mitral stenosis	Left ventricular subvalvar aneuryms	Right ventricular endomyocardial fibrosis	Liver cirrhosis

Case 1. A S, a 12-year old boy, presented with malaise, fever and cough of 2 weeks' duration, followed one week later, by dyspnoea and swelling of both feet. Further questioning revealed that he sustained an abrasion on his right foot a week prior to the onset of symptoms.

On admission, he was toxic, pale, pyrexic (temperature, $39°C$) and had a septic ulcer on the lateral aspect of his right foot. His respiratory rate was 42/minute and breath sounds over both lung bases were reduced. The cardiovascular system was normal except for a sinus tachycardia (rate, 136/minute). Chest radiograph revealed a small right pleural collection, which later turned out to be an empyema; his heart size was however, normal. Staphylococcal septicaemia was suspected and appropriate management measures were instituted.

Three days later, he was visibly worse; his heart rate had risen to 152/minute, the jugular veins were distended, and the liver was 3 cm enlarged. His blood pressure (110/75 mmHg) and heart sounds were however, normal. Repeat chest radiograph showed an enlarged, globular cardiac silhouette, and diagnostic pericardiocentesis confirmed the diagnosis of purulent pericarditis. *Staph. aureus* was subsequently cultured from the pericardial aspirate.

Antifailure medications were administered, and pericardial drainage was commenced via a pericardiostomy tube. Pus ceased to drain from the pericardial space after a week, and signs of speticaemia subsided after 3 weeks of treatment with intravenous cloxacillin; but the signs of cardiac failure persisted.

Two months later, he was still in heart failure, repeat chest radiographs showed persistent cardiomegaly and ECG showed diminution of QRS complexes and inversion of T-waves. At that stage, it was reasoned that he had either acute pericardial constrictioin or severe myocardial damage from the antecedent septicaemia. Echocardiography could not be performed, but a limited right atrial angiocardiography revealed features suggestive of constrictive pericarditis, namely, a thickened right atrial wall with concave endocardium. Subtotal pericardiectomy was performed shortly afterwards. He subsequently improved appreciably, but still needed antifailure drugs 6 months after the operation.

Case 2. AO, an 11-year old boy, was referred to UCH from a specialist hospital where he had received 3 weeks' treatment for congestive heart failure, but without any discernible improvement. He had presented there with cough, effort dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea and abdominal swelling.

Physical examination revealed a very sick boy with puffy face, distended jugular veins, weak peripheral pulses, and an unremarkable apex beat. His blood pressure was 105/80 mmHg. Abnormalities detected on cardiac auscultation comprised a 3rd heart sound located near the base of the heart this was initially thought to be either an opening snap or pericardial knock) and a grade 2/6 diastolic murmur which was loudest at the mitral area. His liver was 11cm enlarged, and there was moderate ascites. He was in severe heart failure.

Initially, it was unclear whether the congestive cardiac failure was secondary to constrictive pericarditis or to mitral valve stenosis. However, results of investigations re-

solved the uncertainty: the ECG findings comprised low-voltage QRS complexes and inverted T-waves in the precordial leads while the chest radiograph showed pulmonary venous congestion, a small box-like heart and, most importantly, flecks of pericardial calcification. Further confirmation of the diagnosis of chronic calcific pericardial constriction was obtained at angiocardiography. He subsequently had partial pericardiectomy, and made a speedy and complete recovery. At discharge, he had no 3rd heart sound or murmur.

Case 3. OJ, a 15-year old boy, presented with a history of abdominal swelling of 8 years duration, intermittent swelling of the face and feet for 5 years, cough for 3 years, and extertional dyspnoea for one year. He had received antifailure treatment from various medical institutions, with short-lived relief on each occasion. There was no history of sore throat or joint pains.

Initial examination revealed a severely wasted boy (weight, 19kg) with bilateral proptosis, circumoral and periorbital hyperpigmentation. His digits were mildly clubbed, and small discrete lymph nodes were palpable in his neck, axillae and groins. He was dyspnoeic (respiratory rate, 36/min) and fine crepitations were audible over both lung bases. The cardiovascular findings were very striking: the neck veins were distended, his peripheral pulse was fast (126/min) but of good volume (BP, 110/70 mmHg), the apex beat was displaced to the 6th left intercostal space, and was diffuse and vigorously heaving, indicating excesive volume-loading

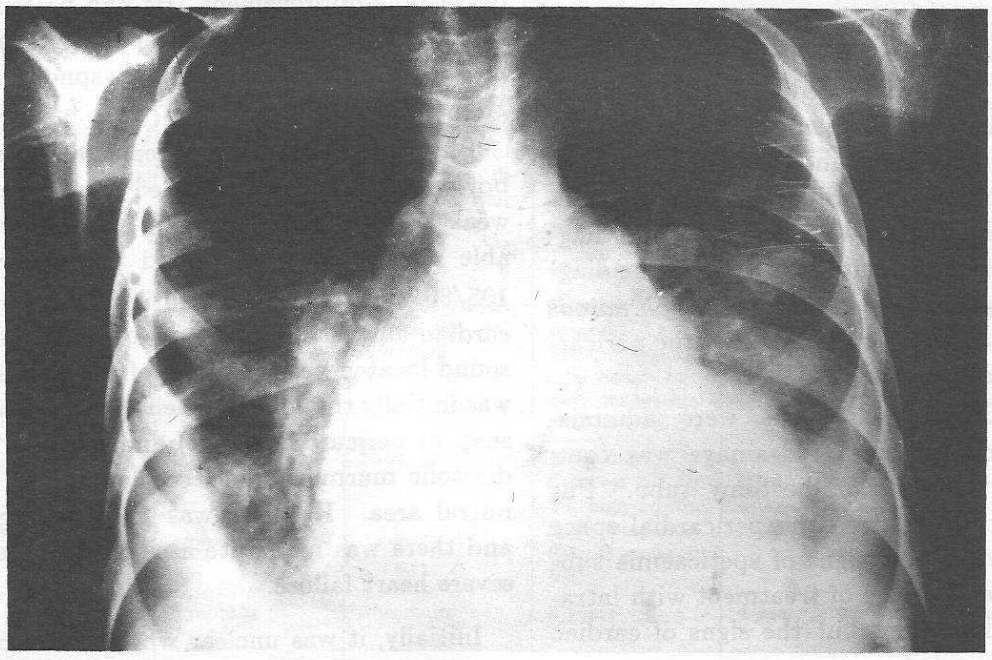

Fig 1 Chest radiograph showing a 'bumpy' left heart border, reminiscent of left ventricular subvalvar aneurysm, and bilateral pleural effusion (Case 4).

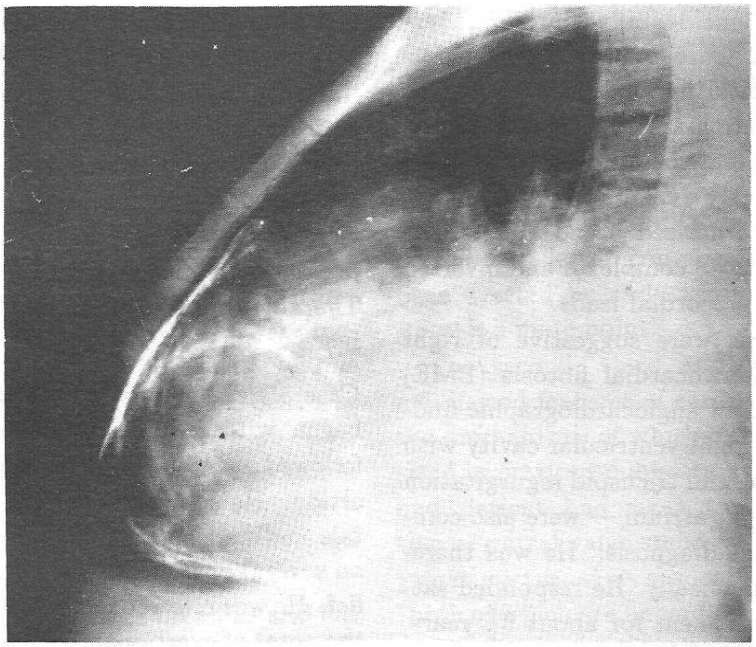

Fig 2 Chest radio graph (lateral projection) of same patient as in Fig 1, showing pericardial calcification.

of the left ventricle. The ausculatory signs consisted of an early 3rd heart sound and a grade 2-3/6 mitral regurgitation murmur. His liver was 10 cm enlarged, spleen was 5 cm; he was clearly in severe heart failure.

Haematological investigations showed no diagnostic features, and tuberculin test (Mantoux) was negative. However, chest radiographs revealed remarkable findings: the anterior-posterior film (Fig. 1) showed a bulge on the left heart border, reminiscent of left ventricular subvalvar aneurysm,[7 8] while the lateral films (Fig 2) showed pericardial calcification, the type usually seen in chronic calcific pericarditis.

On the basis of the clinical features, the patient's major problems were considered to be (i) heart failure secondary to (ii) either left ventricular subvalvar aneurysm or constrictive pericarditis, and (iii) probable disseminated tuberculosis. Cardiac catheterization and echocardiography, two investigations that could unravel the correct cardiac diagnosis, could not be performed because of technical constraints but surgery subsequently confirmed the diagnosis of constrictive pericarditis. His postoperative course was marked by intermittent segmental pulmonary collapse, recurrent empyema thoracis, relentless heart failure and, terminally, respiratory failure. He died 7 weeks after surgery. Autopsy was not allowed, hence the detailed intracardiac anatomy was not ascertained.

Case 4. This has already been fully reported elsewhere[9] and will only be summarized here. It is the case of an 11-year old boy in whom massive hepatomegaly (10cm)

was discovered incidentally during an attack of malaria. Three months later, he developed periorbital hyperpigmentation, tricuspid regurgitation and right heart failure. Chest radiograph at that stage showed moderate cardiomegaly (CT ratio, 64%) while ECG findings comprised broad and tall P-waves, dwarfed QRS complexes and inverted T-waves in the precordial leads.

These features were suggestive of right ventricular endomyocardial fibrosis (EMF) and the subsequent angiocardiographic findings — reduced right ventricular cavity with obliterated apex, and tricuspid regurgitation into a dilated right atrium — were also compatible with that diagnosis. He was therefore, managed medically. He responded satisfactorily to treatment for about $3\frac{1}{2}$ years, and then defaulted from follow-up. When he resurfaced after a lapse of 6 months, he was *in extremis;* he died shortly after readmission.

Autopsy revealed constrictive pericarditis involving principally, the right ventricle, which was severely shrunken. The tricuspid ring was dilated, so also were both atria. There was no sign of endomyocardial fibrosis.

Case 5. The illness in this 10-year old boy began with painless abdominal distension, followed a year later by breathlessness on exertion. He sought care from various hospitals and had abdominal paracentensis done on several occasions, but without lasting relief. He was referred to UCH 10 months after the onset of exertional dyspnoea, and almost

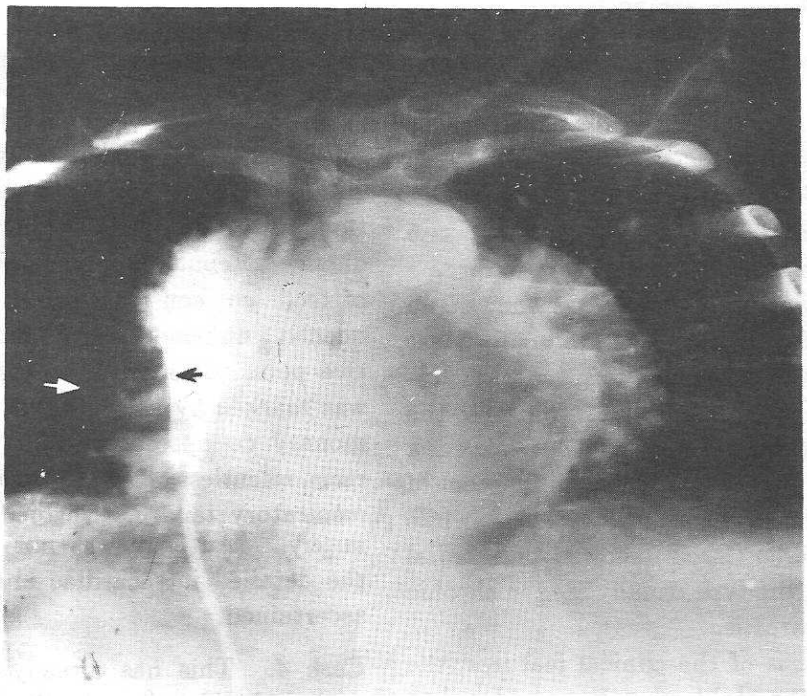

Fig 3 Right atrial angiocardiography in constrictive pericarditis (Case 5). Note the thickened right atrial wall (arrowed) and the concave endocardium.

2 years after the onset of abdominal distension, with a diagnosis of liver cirrhosis.

Examination in UCH revealed moderate wasting and proptosis. His chest was clear but there was slight tachypnoea (rate, 28/min). The clues to the diagnosis were found mainly in the cardiovascular system: his jugular venous pressure was raised, and rose even higher during inspiration (Kussmaul's sign), his peripheral pulses were regular (rate, 106/min) but of small volume (BP, 70/50 mmHg), and the apex beat was indistinct. The heart sounds were however normal, as neither a murmur nor a 'pericardial knock' was audible. Abdominal findings comprised massive ascites, hepatomegaly (12 cm) and spenomegaly (5 cm). ECG findings consisted of broad P- and flat T-waves in the precordial leads. On chest radiograph, the heart was of normal size but its left border was irregular and there was a thin rim of pericardial calcification.

The above features were interpreted to be very suggestive of constrictive pericarditis, which diagnosis was subsequently confirmed at angiocardiography (Fig 3) and surgery. He made steady progress postoperatively and at discharge, there was no ascites, his liver was only 3 cm enlarged and the spleen was no longer palpable.

Discussion

The patients described here illustrate the variability in the clinical manifestations of pericardial constriction, both in terms of temporal progression of the disease as well as in the multiplicity and complexity of its signs. The first patient, for example, typified acute constriction: he remained in cardica failure after the initial pericardial tube drainage, and proceeded to constriction within 3 months. The rapid progression in this case was, at least in part, attributable to the virulence of the infecting pathogen. The persistence of cardiac decompensation after pericardiectomy indicated the presence of significant myocardial disease. Whether or not this latter complication could have been averted by early primary pericardiectomy is a mute point.[2 3 10 11]

The second, third and fourth patients manifested features of cardiac valve defects, the mechanisms of which have been summarized in previous publications.[6 12] The second patient had signs that were suggestive of mitral valve stenosis, namely, paroxysmal nocturnal dyspnoea and orthopnoea (both of which are uncommon in constrictive pericarditis[6]), juxta-apical 3rd heart sound, and an apical diastolic murmur. Given this type of setting, an accurate interpretation of the nature of the 3rd heart sound is vital. If it is taken to be an 'opening snap' a diagnosis of organic mitral stenosis becomes invetable, whereas if it is interpreted to be a 'pericardial knock' constrictive pericarditis becomes the logical diagnosis. The crucial issue therefore is how to distinguish an 'opening snap' from a 'pericardial knock'. The task demands a diligent clinical evaluation: an 'opening snap' is commonly heard in early diastole, and is usually associated with a loud first heart sound (closing snap) and a tapping apex beat. Although a 'pericardial knock' is also heard in early diastole, it is characteristically accompanied by soft heart sounds and a feeble or impalpable apex beat. Where facilities for phonocardiography are available however, distinguishing a 'knock' from a 'snap' should not pose a problem.

There were three extremely remarkable things about the third patient. First, it was the longest history — 8 years of niggling cardiac decompensation. Secondly, he had full peripheral pulses, a normal blood pressure, and a vigorous left ventricular impulse. All these signs are extremely unusual in constrictive pericarditis, and can only be attributed to the associated mitral regurgitation. The third remarkable thing was the chest radiograph which showed features that were compatible with both calcific pericarditis and subvalvular left entricular aneurysm. The genesis of these puzzling features became obvious at surgery where it was discovered that the main constrictive tissue extended round the ventricles like a band, sparing the apex, the atrioventricular groove and the atria. The areas above and beneath the constricting band were dilated and hyperdynamic, hence the hyperactive apex beat, the bulge seen on the left heart border on the chest radiograph, and the mitral regurgitation which presumably resulted from dilatation of the mitral annulus.

Tricuspid regurgitation, an exceedingly rare finding in constrictive pericarditis.[12] was a prominent sign in the 4th patient. As stated earlier, the case has been reported fully elsewhere[9] and will therefore not be discussed here. Suffice it to state that the case vividly illustrates the close similarities between constrictive pericarditis and endomyocardial fibrosis involving the right ventricle. The echocardiographic features of both diseases are, however, fairly distinct,[13–16] and should come in handy in clinical decision-making.

The 5th patient initially manifested abdominal symptoms, as many cases of constrictive pericarditis are wont to do[6]. During the first 12 months of his illness, all he had was abdominal distension caused by hepatomegaly and ascites; there were no symptoms that were directly referrable to the heart. Understandably, the attending physician made a diagnosis of liver cirrhosis. Signs of constrictive pericarditis were detected in UCH almost two years after the onset of symptoms.

With regard to the 3rd, 4th and 5th patients, there was an unacceptably long lapse between the onset of illness and the time the correct diagnosis was made. But they are not isolated cases as many authors have repeatedly drawn attention to the delays in diagnosing pericardial constriction[6]. Where echocardiography is available, however, such delays should not occur anymore.

All five cases described here illustrate the spectrum of the clinical manifestations of constrictive pericarditis in children, a spectrum that is so wide that it often leads to errors in diagnosis. The errors will be fewer, however, if adequate care is taken during clinical evaluation. With haemodynamically significant pericardial construction, the jugular venous pressure is invariably elevated, even when the other classical signs of heart failure are not discernible. Such jugular venous distension was present in all the 5 patients described here, and in 95% of the cases reviewed by Hirschman[6]. It is therefore a sign that should be diligently sought. Besides, the presence of heart failure in a child who has a globular 'murmurless' heart should always prompt a search for a pericardial disease or cardiomyopathy. Table II, a synopsis of the main clinical features of constrictive pericarditis and right ventricular EMF, is aimed at facilitating the clinical recognition of these two

TABLE II
Clinical Signs of Chronic Constrictive Pericarditis and Right Ventricular Endomyocardial Fibrosis (RV FMF)

	Constrictive Pericarditis	*RV EMF*
Central cyanosis	Rare	Occurs in 40-50% of patients
Arterial pulse volume/ pressure	Usually low	Invariably low
Jugular venous pattern	Inspiratory rise; rapid diastolic collapse	Giant 'V' waves
Precordial activity	Precordium usually quiet	Infundibular heave occasionally palpable
Heart sounds (H/S)	3rd H/S (pericardial knock) in approx. 40%; cardial murmur uncommon	3rd H/S (endocardial knock') common; TI murmur in 10-20% of patients
Ascites	Variable quantity	Usually massive
Chest radiograph	Cardiac enlargement in 40% of patients; pericardial calcification in approx. 50%, pulmonary venous congestion	Enlarged cardiac silhouette in practically all cases; no pericardial calcification, but infundibular calcification in 10-15% of cases; pulmonary oligaemia.
Electrocardiogram	Low amplitude QRS, flat or inverted T-waves	Tall P-waves, low-amplitude QRS, qr pattern in V_1/V_2, flat or inverted T-waves.

TI: tricuspid incompetence.

very similar conditions. Where the listed features are insufficient to make the necessary distinction additioinal clarification can be obtained at echocardiography[13 15 16] or angiocardiography.[17–20] Finally, a normal or only marginally increased CT ratio in a child who shows all the other classical signs of congestive heart failure should always prompt a thorough search for constrictive pericarditis.

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