transmural ulceration and perforation of the oesophagus we have seen. The clinical and histological evidence suggests that the leak occurred during the second week after sclerotherapy, by which time most patients will have been discharged from hospital. Minor side-effects of sclerotherapy are seen regularly but usually disappear within 48 hours. These include retrosternal discomfort, dysphagia and odynophagia, fever, tachycardia and arrhythmias, changes in the prothrombin index and partial thromboplastin time, and small pleural effusions. They are presumably related to the sclerosant entering the general circulation and to local effects.7-16

Sclerotherapy does therefore seem to be associated with certain problems, but if we are prepared to accept these, as well as the high incidence of recurrent bleeding and the need for continuous follow-up and repeat procedures, it may still be the treatment of choice when major surgical procedures pose a high risk because of severely compromised liver function.

REFERENCES

Primary cardiac hydatid disease
A case report
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Summary
A young Coloured man whose only symptom was that of minimal dyspnoea on exertion, on examination had signs of infundibular stenosis which were confirmed by cardiac catheterization. Additional features were left anterior hemiblock and cardiomegaly as delineated radiologically. Cross-sectional echocardiography revealed a very large cystic mass located within the interventricular septum which encroached upon the right ventricular outflow tract. This cystic mass was further delineated by cardiac cine angiography. It is postulated that this mass was a primary cardiac echinococcal cyst and was directly responsible for the complications of left anterior hemiblock and the infundibular obstruction. The patient declined surgery and a definitive pathological diagnosis could therefore not be made. If this was a hydatid cyst then it is the second case reported in the literature diagnosed by two-dimensional echocardiography and in which left anterior hemiblock has been recorded. The clinical features, complications and surgical correction are briefly outlined.

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Case presentation
The patient was a 27-year-old Coloured man from South West Africa/Namibia whose only complaint, mild dyspnoea on exertion, he attributed to being overweight and unfit. In June 1982 he went to his general practitioner with influenza, at which time a cardiac murmur was first detected. He worked for a building construction concern and had not been in a sheep-rearing area. The physician who examined him in SWA could find no evidence of cardiac failure. Further findings reported were a blood pressure of 125/85 mmHg, a 'significant systolic murmur' heard maximally just below the pulmonary area and a widely split second heart sound with reduced intensity of the pulmonary component. A 'right ventricular lift' was noted as well as 'pulsation in the left second and third interspace'. Chest radiography documented a 'rather globular heart' as well as 'prominence in the area of the right atrium and right ventricle'.
The possibility of pulmonary oligaeemia was also raised. A resting ECG was described as 'suggesting right ventricular enlargement' on account of S waves in leads I, II and III and the unipolar leads as well as a prominent R wave in lead aVR. The consulting physician made a diagnosis of 'acyanotic congenital heart disease', more specifically isolated pulmonary valve stenosis. Acyanotic Fallot's tetralogy was considered unlikely since exercise failed to precipitate central cyanosis.

In view of the above findings the patient was referred to the Cardiac Clinic of Tygerberg Hospital in August 1982 for further investigation and management. On clinical examination he was mesomorphic, being 1.56 m tall and weighing 67 kg. There was, however, no evidence of any congenital malformation or of infective endocarditis. Cardiovascular examination revealed normal peripheral pulses, no bruits, a blood pressure of 120/80 mmHg and an elevated jugular venous pressure of 6 cm H₂O displaying a prominent 'a' wave. Visible left parasternal pulsation was associated with an obvious parasternal lift, but the apex beat was not displaced or abnormal in character. A systolic thrill was detected over the second left intercostal space some 3 cm from the left sternal edge. The heart sounds were normal and there were no additional sounds. Auscultation revealed a grade 5/6 ejection systolic murmur heard maximally at the second left intercostal space and increasing in intensity on expiration. This murmur radiated across most of the precordium. The rest of the physical examination was unremarkable.

Side-room investigations, serum biochemical findings and hepatic function tests revealed no abnormality. Blood VDRL and FTA tests as well as tests for C-reactive protein were all negative. An Echinococcus complement fixation test after cardiac catheterization was negative. A chest radiograph (Fig. 1) delineated a globular-shaped cardiac shadow with the suggestion of some right ventricular enlargement. The pulmonary vascular markings were assessed as being normal. A resting ECG (Fig. 2) demonstrated sinus rhythm of 60 beats/min, a P-R interval of 0.19 second, left anterior hemiblock and asymmetrical T-wave inversion in leads aVL, V1 and V2. A two-dimensional echocardiogram demonstrated a large echo-free mass in the interventricular septum (IVS) which encroached anteriorly on the right ventricular outflow tract but did not appear to cause any obstruction to left ventricular outflow or have any interference with the mitral valve apparatus (Fig. 3).

Cardiac catheterization was carried out on 26 August 1982. The Seldinger technique via the right femoral artery and vein was employed using 7F Goodale-Lubin and pigtail catheters. No intracardiac shunts could be demonstrated. All the left-sided pressures were within normal limits, as were indices of left ventricular contractility. Specifically, there was no gradient detectable within the left ventricular cavity or via the aortic valve, but this was not the case with the right-sided intracardiac pressures. Withdrawal of the catheter from the main pulmonary artery (pressure 26/17 mmHg) to the right ventricle (pressure 26/3-11 mmHg) failed to show any pulmonary valvular gradient or obstruction (Fig. 4a), but further withdrawal into the low right ventricle (pressure 72/8-13 mmHg) delineated a peak systolic infundibular gradient or obstruction measuring 46 mmHg (Fig. 4b). A left ventricular cine angiogram in the right anterior oblique (RAO) projection (Fig. 5) and left anterior oblique (LAO) view (Fig. 6a) demonstrated the presence of a
Fig. 3. a — two-dimensional echocardiogram (long-axis view) showing a large, echo-free mass within the IVS, representing the hydatid cyst. b — diagrammatic representation of the two-dimensional echocardiogram (ACW = anterior chest wall; RV = right ventricle; LV = left ventricle; LA = left atrium; MVO = mitral valve orifice; HC = hydatid cyst; LVPW = left ventricular posterior wall; PC = pericardium; AO = aorta; AV = aortic valve).

Fig. 4. a — pressure withdrawal from pulmonary artery (PA) to high right ventricle (RV) demonstrating absence of pulmonary valvular stenosis. b — pressure withdrawal from high right ventricle (RVH) to low right ventricle (RVL). Moderately severe infundibular stenosis (gradient of 46 mmHg) is visualized.

Fig. 5. Left ventricular cine angiograms in the RAO projection demonstrating the large hydatid cyst (arrowed) within the IVS (LVED = left ventricle in end-diastole; LVES = left ventricle in end-systole).
large mass in the IVS which caused marked anterior displacement of the coronary arteries. Right ventricular cine angiography in the LAO projection (Fig. 6b) also showed undoubted severe infundibular stenosis. The non-calcified mass also appeared to encroach on the outflow tract of the left ventricle (Fig. 5) despite no intracavitary pressure gradient having been recorded. Cardiac catheterization was completed without any complications.

Since there seemed to be overwhelming evidence in favour of a hydatid (echinococcal) cyst situated within the IVS and of such large dimensions that it had given rise to conduction abnormality in the form of left anterior hemifascicular block, it was decided to offer the patient emergency surgery for fear of the dreaded complications of this condition. Despite exhaustive discussions with the patient he refused any form of cardiac surgery; he was given no medication and was discharged home to SWA to be followed up by his physician. To date I have contacted this physician on several occasions only to be reassured that the patient is still alive and totally asymptomatic.

Discussion

Infestation of humans by Echinococcus granulosus is usually associated with involvement of the liver or lungs or both of these organs. Primary cardiac echinococcosis ('hydatid heart disease') is exceptionally uncommon, the incidence being quoted as between 0.02% and 2.0% of all cases of human hydatid infestation.7 The great majority of cases reported in the literature were unsuspected during life and only diagnosed at postmortem examination. Most hydatid cysts found in the heart are primary and unilocular and are thought to reach this organ via the coronary arteries, a feature which prompted some authors to claim that these cysts were commoner in the left side of the heart.8,9 The cyst is thought to become multilocular only after spontaneous rupture into the cardiac chambers or pericardial space and then to give rise to the formation of daughter cysts which may be dispersed within the systemic or pulmonary circulation, only to manifest much later with complications such as chronic cor pulmonale. Another possible outcome is death of the echinococcal cyst with subsequent calcification of its constituent surrounding wall, a stage commonly associated with negative Casoni and Weinberg tests.

The first and only occasion that a cardiac echinococcal cyst was documented in the SAMJ was in 1898 when Guillemand made the following contribution: 'A boy of fourteen died suddenly, having shown no previous symptoms, except that he complained of being tired after his work. Post mortem, when the pericardium was opened, a glistening white swelling was seen projecting to the front at the base of the heart, just below the auricles. This was found to be a hydatid cyst in the septum between the ventricles, so that the cyst projected on either side into the ventricles. The muscular tissue had been displaced and there was merely the thin cyst wall between the cavities of the ventricles and the cyst.'

When the cyst is located in the IVS it is most commonly seen in the upper part of this structure and least frequently in the middle portion of it.3 Very rarely does the hydatid cyst occupy most of the IVS,4 when it usually extends into either the left or right ventricular wall, or even both.4 Probably because of the smaller resistance to extension, the cysts tend to expand into the surrounding pericardial space rather than into the left or right ventricular cavities. A most unusual case was reported by Kurban et al.7 in which a cyst had expanded so as to compress the left coronary artery in its interventricular groove course with subsequent angina pectoris. Despite frequent localization of echinococcal cysts in the IVS no case of rupture into both ventricles has been documented. When the cyst expands inwards towards the ventricular chambers it usually involves the right ventricle, probably because of the lower intraventricular pressure. This situation was clearly encountered in our patient and was severe enough to cause a harsh systolic murmur and infundibular stenosis. A similar case was reported by De los Arcos et al.,6 their patient also had the additional complication of bifascicular block (left anterior hemiblock plus complete right bundle-branch block), thought to have been due to compression of the bundle branches in the superior portion of the IVS. Left anterior hemiblock was also witnessed in our patient, and it would appear that this is the second such case in the world literature. Other very rare conduction abnormalities, such as
complete atrioventricular heart block with resultant Stokes-Adams attacks, have been documented previously.\(^9\)

Other complications of cardiac echinococcal cysts, usually related to acute rupture, have been described, including acute pericarditis,\(^{10,11}\) chronic pericarditis with effusion (sometimes massive),\(^{12}\) and constrictive pericarditis.\(^{13}\) Recurrent pulmonary embolism with ensuing pulmonary hypertension\(^{14,15}\) is less commonly encountered. Even more rare sequelae, such as intermittent obstruction of the tricuspid valve (atrial pseudo-myxoma)\(^{16}\) and pseudoaneurysms of the heart\(^4\) are known to occur. Systemic hydatid emboli giving rise to cerebrovascular accidents can be most debilitating.\(^11\) Allergic and anaphylactoid reactions follow most echinococcal cyst ruptures, but it is important to be aware of the fact that these ruptures can pass unnoticed by the patient. Ruptures (usually of cysts located within the IVS) into the right ventricle\(^4\) or left ventricle\(^4,16\) can prove quite catastrophic. Paroxysmal supraventricular tachycardia\(^{17}\) as well as ventricular arrhythmias\(^{18}\) have been documented.

The ECG is generally normal when the hydatid cyst is located within the right ventricular myocardium. However, if the cyst is situated within the left ventricle then the classic features of ‘coronary T waves’ (inverted T waves in the anteroseptal and anterolateral leads) are encountered. It is very rarely that pathological Q waves are documented. The ECG can thus prove to be quite useful in localizing the cyst.

Diagnosis by means of the non-invasive technique of cross-sectional (two-dimensional) echocardiography was described for the first time by Oliver et al.\(^{17}\) These authors documented the case of a 26-year-old male farmer with neurological symptoms who had a hydatid cyst localized within the apical third of the IVS as well as several cerebral cysts. This patient also displayed deeply inverted T waves in the inferolateral and apical leads, and had a harsh apical systolic murmur. Our patient is the second one in the literature in whom cross-sectional echocardiography was employed for the diagnosis. M-mode echocardiography proved to be of no diagnostic use in either of these 2 patients.

A patient is not often entirely asymptomatic in the presence of a primary cardiac echinococcal cyst, but this appeared to be the case with our patient. The fact that the patient may be asymptomatic is no indication that surgery must be postponed. There is no doubt that surgery is the only form of therapy for this fairly uncommon condition, although this might prove extremely difficult if most of the IVS is involved by the cyst, in which case permanent cardiac pacing may be required because of involvement of the conduction system. Once hydatid pulmonary hypertension is established the prognosis is uniformly poor, although the occasional survivor has been documented.\(^{14}\) If the cyst is epicardial, removal is possible by means of a simple thoracotomy,\(^{18}\) but if it is situated intramyocardially extra-corpooreal techniques must be employed.\(^{14}\)

Most authors\(^{1,2,19-22}\) recommend the instillation of concentrated saline solution after the removal of any daughter cysts with subsequent closure of the cavity formed. Unfortunately our patient refused to undergo surgery and it would seem that his prognosis is extremely poor.

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