Subvalvular left ventricular aneurysms
A report of 5 Ugandan cases

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The pathological features of 5 cases of subvalvular cardiac aneurysms occurring in Uganda over a 13-year period are described; 3 were subaortic and 2 were submitral. One of the latter was atypical in that the aneurysm arose at some distance below the fibrous mitral ring and was associated with endomyocardial fibrosis. In 2 cases where the coronary sinus and its ostium was examined, no abnormality was detected.

Previous descriptions of the pathological aspects of subvalvular aneurysms are briefly reviewed.

Coronary artery disease and post-infarction cardiac aneurysms are rare in Africans. However, in recent years several reports have drawn attention to the pathological features of a distinctive subvalvular left ventricular aneurysm in black Africans. Such cases have been reported from Nigeria (Robertson and Jackson, 1960; Abrahams et al., 1962; Edington and Williams, 1968), South Africa (Lurie, 1960; Chelser et al., 1965; Chelser, Tucker, and Barlow, 1967), Rhodesia (Pellatt, 1972), and the Belgian Congo (Zaire) (Yarom and Griffel, 1964). It is likely that the three cases reported by Beheyt and Vandeputte (1958) from the Belgian Congo, though attributed to tuberculosis, also fall into this category. Thus, the total cases coming to necropsy, reported since 1958, is 29. Awareness of this condition has enabled it to be diagnosed clinically (Abrahams et al., 1962; Beheyt and Joris, 1963) and the radiological features of the Nigerian cases have been fully reported by Cockshott et al. (1967). The aetiology of the aneurysms remains uncertain though an unspecified infective basis, tuberculosis, a congenital weakness of the valve rings, and ischaemia have all been suggested.

This paper describes the pathological features of 5 cases of subvalvular aneurysms seen in Uganda over a 13-year period and the published reports are briefly reviewed.

Subjects and methods
The pathological features of all cases of cardiac aneurysms in the post-mortem records of the Department of Pathology, Makerere University, Kampala, in the 13-year period 1960 to 1972, were examined. The clinical details, the gross pathology, and histology were reviewed. Sections of the heart and aneurysms were examined using a variety of stains which included haematoxylin and eosin, Weigert's elastic, elastic Van Gieson, Masson's trichrome, periodic acid Schiff, and Gram's stain. Macrosections of the aortic valve region were examined in Case 5.

Results
In the 13-year period 11,098 necropsies were carried out, 71·3 per cent of which were men and 28·7 per cent women; 67 per cent (7435 cases) were 15 years old or more.

Eight cardiac aneurysms were found, of which 5 were of the subvalvular type, and the salient features of these are shown in Table I. The aetiology of the other three aneurysms was considered to be post-infarctive, syphilitic, and indeterminate in one example of an aneurysm of the left ventricular apex.

All 5 subvalvular aneurysms arose from the left ventricle: 3 were subaortic and 2 were submitral. The ages ranged from 21 to 45 years; 3 were male and 2 were female. All were black Africans, 3 were Baganda (the tribal group found in Southern Uganda and around Kampala), 1 was a Samya (a Bantu speaking tribe from Eastern Uganda), and the tribe of the other was not known.

The incidence of subvalvular aneurysms in the adult necropsy population (over 15 years old) was 0·07 per cent.

Case reports
Case I
A 21-year-old Muganda man was admitted to Mulago Hospital in gross cardiac failure four days before death.
He had attended the cardiac clinic for the previous two years as a presumptive case of rheumatic heart disease with mitral insufficiency and stenosis. On admission, his blood pressure was 100/70 mmHg, with a pulse of 100/min of small volume and with an increased jugular vein pressure. He died suddenly while being given digitalis.

Necropsy This was a slender, but well-nourished young African man. There was no free fluid in the pericardial sacs. The heart showed a smooth pericardium and the intracardiac cavities were grossly dilated. The mitral valve orifice was deformed because the posterior mitral leaflet was bound down to a large plaque of endomyocardial fibrosis, 3 cm wide and which extended for 4 cm over the posterior left ventricular wall. The posterior leaflet was wrinkled on its atrial surface because of slight fibrosis; this extended over the endocardium of the left atrium for a small distance. The anterior papillary muscle was covered by thickened fibrous plaques. 3 cm below the mitral valve there was an ostium, 2 cm in diameter, in the wall of the left ventricle leading into an aneurysm which extended obliquely and downwards for 4 cm within the thickened ventricular wall, maintaining the same diameter as the opening and not distorting the epicardial surface (Fig. 1). It was lined by fibrous tissue and filled with thrombus. The aneurysmal opening was separated from the endomyocardial fibrosis plaque by a narrow strip of normal endocardium. The apex of the ventricle and the outflow tract were not involved by endomyocardial fibrosis. The aortic valve was normal. There was increased thickness of the walls of the left atrium (6 mm) and the left ventricle (16 mm). The right atrium and right ventricle were dilated; the tricuspid valve was normal and the pulmonary valve showed fenestration of the right cusp. The myocardium was firm and showed no scarring. The coronary arteries including their ostia were patent; the coronary sinuses and ostium were patent. The aorta showed mild atheroma.

The gastrointestinal tract and the parenchymal organs showed chronic venous congestion. Pyelonephritic scarring was noted on both kidneys and the left one showed one small infarct at the upper pole.

**Case 2**

This 45-year-old Muganda woman was admitted in cardiac failure 8 days before death. She had had a similar illness 3 years previously which lasted for 3 weeks. On examination she was oedematous, dyspnoeic, with a rapid irregular pulse of 180/min and a blood pressure of 90/85 mmHg. A chest x-ray revealed an abnormal enlarged cardiac shadow which was interpreted as a left ventricular aneurysm. The cardiac arrhythmia persisted in spite of quinidine treatment.

Necropsy This elderly African woman was slightly obese with pitting oedema of the lower legs, and free yellow fluid in the pleural and peritoneal cavities. The pericardium was smooth and the heart considerably enlarged. Projecting from the anterolateral aspect of the upper part of the left ventricle was a tumour-like mass measuring 7 x 5 x 4 cm which on section was found to be a thrombus-filled aneurysm. It communicated with the left ventricular cavity by a narrow ostium situated below the annulus fibrosus adjacent to the anterior mitral leaflet. The aneurysm wall was 3 mm thick. There was mitral incompetence, the orifice admitting three finger tips. There was a small patch of endocardial thickening in the left ventricle. All cardiac chambers were dilated and hypertrophied. No data concerning the coronary sinuses are available. The myocardium showed a fibrous area (2 x 3 cm) in the middle of the interventricular septum. No obvious pathology was found in the valves or the coronary arteries. There was pronounced venous congestion of the parenchymal organs.

**Case 3**

This adult African man was referred from a district
hospital with a two-month history of cough, general malaise, and night sweats. The sputum contained acid-fast bacilli. He died in the casualty department of Mulago Hospital with a diagnosis of pulmonary tuberculosis.

Necropsy This showed a well-built and well-nourished African. A small amount of straw-coloured ascites was found. Both lungs were consolidated due to extensive bilateral tuberculous bronchopneumonia. There were severe pleural adhesions. The mediastinal lymph nodes showed caseous tuberculosis. The heart was greatly hypertrophied and dilated. There was a circular hole in the membranous part of the interventricular septum, on the left ventricular side, below the aortic valve, which communicated with a thick-walled aneurysm extending behind the aortic valve ring. The aneurysm did not communicate with or deform the right ventricle. The coronary sinus was not specifically examined. The aortic valves showed fenestration of the cusps. The liver and spleen showed pronounced venous congestion.

Case 4
A 37-year-old Muganda man was admitted to hospital 2 weeks before death with a history of intermittent fever and cough. On examination he was found to have aortic and mitral incompetence. A possible diagnosis of subacute endocarditis was made.

Necropsy This showed a well-nourished and well-built African with no external abnormalities and no free fluid in the abdomen or pleural cavity. There was an obliterative fibrino-haemorrhagic pericarditis with a small haemorrhagic effusion. The heart was enlarged and showed bilateral ventricular hypertrophy and dilatation, more pronounced on the left side. Below the aortic valve, there was a small thrombus-filled aneurysm which extended towards the ventricular surface of the mitral valve. There was no evidence of rheumatic carditis. The coronary sinus was not examined. The lungs were extensively oedematous, and the liver and spleen were moderately congested.

Case 5
This was a 25-year-old Samya woman who died on the day of admission to hospital. She had complained of chest pain, dry cough, headache, and fever for one week; and palpitations, dyspnoea, and vomiting for one day. On examination, she was anaemic and dyspnoeic; she also had a rapid, paradoxical pulse (140/min) and the blood pressure was 90/60 mmHg. The haemoglobin was 11.2 g/100 ml and the white cell blood count was 20,000 mm$^3$. A diagnosis of anaemia and pericarditis was made.

Necropsy The body was that of a well-nourished African woman showing peripheral oedema but no pleural or peritoneal effusions. There was a partly obliterative fibrino-haemorrhagic pericarditis with a small multi-loculated haemorrhagic effusion. The heart was enlarged and showed bilateral atrial dilatation. Both ventricles were hypertrophied (right 6 mm, left 16 mm) and dilated.

Inferior to the non-coronary cusp of the aortic valve was the ostium of an aneurysm, measuring $3 \times 2 \times 1.5$ cm, and containing thrombus. This aneurysm communicated
with the sinus of the non-coronary cusp after rupture of this cusp (Fig. 2). A sagittal section through the non-coronary cusp and the aneurysm (Fig. 3) showed that it had not perforated into the right atrium nor did it bulge into the left atrium. The aneurysm had also ruptured into the space between the root of the aorta and the right atrium. The blood had tracked through the connective tissue in this space and thence into the pericardial sac. The left coronary ostium and the corresponding artery were patent and normal, as was the right coronary ostium. The coronary sinus and its ostium were also normal. The aortic valve cusps were abnormal, they showed nodular, non-uniform thickening on the sinus side, the right cusp also having a nodule on the ventricular aspect. The remaining three valves, the endocardium, and myocardium were all normal. A culture of the aneurysm yielded scanty growth of contaminants. The aorta was elastic and smooth but the main pulmonary arteries showed atheromatous plaques. The lungs showed acute bronchitis and were indurated but not oedematous. There was chronic venous congestion of the liver. In the spleen a recent infarct (3 × 2 cm) was found. The parenchyma was otherwise within normal limits.

Histopathology

Heart The histopathological features of the 4 cases where material was available were broadly similar and are summarized in Table 2. (Case 3 was preserved as a museum specimen.)

In the subaortic aneurysms, the wall consisted of fibrous tissue with no myocardial fibres. In the two submitral cases, the greater part of the wall thickness consisted of fibrous tissue, but a few strands of attenuated myocardial fibres were incorporated into the wall. In Case 1, the lining of the aneurysm was similar to the adjacent endomyocardial fibrosis plaque; in Case 2 there was extensive hyalinization with occasional plaques of calcification and in one area a focus of bone trabeculae without any marrow. In 3 cases, nonspecific granulation tissue together with chronic inflammatory cells were seen in the wall of the aneurysm. No granulomata were identified.

All the aneurysms contained thrombus which was mainly of recent origin, but in 2 cases it was a mixture of fresh and organized thrombus.

In all cases there was hypertrophy of the myocardial fibres. No evidence of myocarditis was seen and neither was there any evidence of coronary artery disease. Mild scattered interstitial myocardial fibrosis was seen in 4 cases.

In the 2 cases in which the aortic valves were examined, no valvular vascularization was noted, but there was fibrous thickening of either nodular or diffuse type. In 1 case (Case 5) there was partial destruction of an aortic valve due to rupture of the aneurysm through the cusp, with resulting necrosis, haemorrhage in the neighbourhood of the cusp, and subsequent disorganization of the cusp architecture.

Other organs Histological changes in other organs were mainly a reflection of passive venous congestion. Four of the lungs showed a moderate degree of alveolar wall thickening and the same four contained haemosiderin-filled macrophages in the alveoli.

The liver showed passive centrilobular venous con-
gestion in all cases. None of the kidneys showed evidence of hypertension, but one showed mild chronic pyelonephritis (Case 1).

**Discussion**

Of the 8 cardiac aneurysms seen in Uganda over a 13-year period, 5 were considered to be of the subvalvular type. This is a rare condition in comparison with other cardiac conditions in Uganda; Hutt (1970) found that, in 3589 necropsies over a 4-year period, the cause of death was cardiac disease in 637 (17%). Hypertensive heart disease (23%), rheumatic heart disease (17%), and endomyocardial fibrosis (10%) were the three major cardiac problems.

One of the aneurysms in the present study was somewhat atypical in that its ostium was situated slightly lower than is usual; it was also associated with endomyocardial fibrosis, an association that has been noted on one previous occasion only (Robertson and Jackson, 1960). In general, the morbid anatomy and histological features were similar to the pathological descriptions of the 29 previously reported cases and are summarized in Table 3. Subvalvular aneurysms occur predominantly in a young age group; in the present series the ages range from 21 to 45 years, and this compares with an age range of 7 to 60 years in the previous cases.

These aneurysms arise in two situations, either in relation to the fibrous rings of the aortic valve or in relation to the mitral valves, hence the descriptive name given to them by Abrahams et al. (1962). In this series there were 3 subaortic and 2 submitral aneurysms, though the latter are commoner in the previous series. The two types of aneurysm are also dissimilar in other respects. The submitral aneurysms grow to a larger size, and because of the less restrictive anatomy in the mitral area, are

**TABLE 2** Histopathological features of 4 hearts with subvalvular aneurysms

<table>
<thead>
<tr>
<th>Case</th>
<th>1</th>
<th>2</th>
<th>4</th>
<th>5</th>
</tr>
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<tbody>
<tr>
<td>Pericardium</td>
<td>Pericarditis</td>
<td>+</td>
<td>-</td>
<td>++</td>
</tr>
<tr>
<td></td>
<td>Myocarditis</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Myocardium</td>
<td>Focal myocardial fibrosis</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td></td>
<td>Hypertrophiy of myocardial fibres</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Valves</td>
<td>Thickening</td>
<td>o</td>
<td>o</td>
<td>-</td>
</tr>
<tr>
<td></td>
<td>Calcification</td>
<td>o</td>
<td>o</td>
<td>+</td>
</tr>
<tr>
<td></td>
<td>Perforation</td>
<td>o</td>
<td>o</td>
<td>(?)</td>
</tr>
<tr>
<td></td>
<td>Wall structure</td>
<td>EMF</td>
<td>Calcification, bone, hyaline</td>
<td>Endocardial thickening</td>
</tr>
<tr>
<td>Aneurysm</td>
<td>Thrombus</td>
<td>F</td>
<td>R</td>
<td>F, R</td>
</tr>
</tbody>
</table>

Key: o = no histology; - = negative; + = minor change; ++ = moderate change; EMF = endomyocardial fibrosis; F = fresh thrombus; and R = organized thrombus.
Table 3 Necropsy features of published subvalvular left ventricular aneurysms since 1958

<table>
<thead>
<tr>
<th></th>
<th>Subaortic aneurysm</th>
<th>Submitral aneurysm</th>
<th>Female</th>
<th>Male</th>
<th>Aortic valve cusp rupture</th>
<th>Atrial extension</th>
<th>Multiple ostia</th>
<th>Thrombus formation</th>
<th>Wall calcification and/or ossification</th>
<th>Associated with tuberculosis</th>
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<tbody>
<tr>
<td>Beheyt and Vandeputte (1958)</td>
<td>3</td>
<td>0</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>?</td>
<td>0</td>
</tr>
<tr>
<td>Lurie (1960)</td>
<td>3</td>
<td>1</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Robertson and Jackson (1960)</td>
<td>6</td>
<td>3</td>
<td>3</td>
<td>4</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>4</td>
<td>1</td>
<td>0</td>
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<tr>
<td>Abrahams et al. (1962)</td>
<td>6</td>
<td>2</td>
<td>6</td>
<td>5</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>6</td>
<td>0</td>
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<tr>
<td>Yarom and Griffel (1964)</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>0</td>
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<tr>
<td>Chesler et al. (1965)</td>
<td>6</td>
<td>1</td>
<td>5</td>
<td>2</td>
<td>3</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Edington and Williams (1968)</td>
<td>3</td>
<td>0</td>
<td>4</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>3</td>
<td>0</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>Pellatt (1972)</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Present series</td>
<td>5</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>3</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Totals</td>
<td>34</td>
<td>38</td>
<td>32†</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

* Cases with multiple aneurysms.
† Sex not reported in two cases.

able to expand in a superior, lateral, or anterior direction. This size difference allows their radiological detection in life, whereas the subaortic aneurysms rarely grow to a size that enables them to be detected by non-contrast radiology. Expansion of the submitral aneurysm in a superior direction between the left atrial endocardium and myocardium may be such that it leads to encroachment of the aneurysm on to the cavity of the left atrium. This expansion was not seen in the present series, but five such cases have previously been recorded (Robertson and Jackson, 1960; Chesler et al., 1965; Edington and Williams, 1968). Secondary communication may then take place between the aneurysmal cavity and the lumen of the left atrium, due either to ulceration after subacute bacterial endocarditis (Chesler et al., 1965) or to mechanical causes (Edington and Williams, 1968).

Submitral aneurysms are commonly multiple, and Lurie (1960) has described a subaortic and submitral aneurysm in the same patient; they are frequently loculated and may have multiple ostia. Four cases have been reported in which more than one ostium was present (Abrahams et al., 1962; Edington and Williams, 1968), the maximum number of openings being four. Such features as loculation and multiple ostia suggest that, initially, multiple submitral aneurysms developed but that secondary fusion produced a single, multilocular aneurysm with more than one opening. In Case 5, the subaortic aneurysm had secondarily ruptured into the base of the non-coronary cusp producing considerable distortion of the aortic valve. Two previous such cases have been reported by Robertson and Jackson (1960) and Abrahams et al. (1962).

The aneurysm wall is composed of fibrous tissue, but in the submitral type there are some attenuated myocardial fibres incorporated into its wall. There may be non-specific chronic inflammatory cells, granulation tissue, and haemosiderin containing macrophages in the wall.

A high percentage of these cases have shown either a generalized fibrous pericarditis or fibrous adhesions over the aneurysms, often associated with non-specific chronic inflammation of the visceral pericardium. The cause of this is speculative, but it is of some importance as it increases the technical difficulties if surgical excision of the aneurysm is contemplated (Chesler et al., 1965). Other pericardial changes have included pericardial effusion or fibrino-œamorrhagic pericarditis which in one of the present cases (Case 5) was caused by a leak from the aneurysm.

In the present series, all the hearts were enlarged with left ventricular hypertrophy; most of the previous cases also showed this. The aneurysms interfere with normal cardiac function in two ways: both the subaortic and submitral aneurysms deform the architecture of the valves and give rise to incompetence, though in one case subaortic stenosis was reported (Yarom and Griffel, 1964); in addition, because the submitral aneurysms reach a considerable size, they accumulate a large regurgitant flow during systole. In a minority of cases, the subaortic aneurysm may distort the anatomy of the coronary arteries thus causing attenuation and
stretching of the vessel; in one case this had resulted in secondary thrombosis in the right coronary artery leading to death (Lurie, 1960).

It is possible that single cases of left ventricular aneurysms in black Africans reported before 1958 were of the subvalvular type. Abrahams et al. (1962), in an extensive search of the earlier reports, were able to find 8 cases up to 1958, the earliest being that of Corvisart (1813). The 3 cases of submitral aneurysms in young people from Zaire, reported by Beheyt and Vandeputte (1958), were attributed to tuberculosis; however, the general structure and situation of these aneurysms corresponded to the typical subvalvular type. In 2 cases there was concomitant tuberculosis: in a 7-year-old boy the large submirtal aneurysm was associated with acute miliary tuberculosis but with no lesions in the aneurysm wall; in the other case there was chronic pulmonary tuberculosis, and histological but not bacteriological evidence of tuberculosis of the heart and aneurysm wall. A further case showed no tuberculosis but there was a non-specific giant cell reaction in the aneurysm wall. Tuberculosis is often found at necropsy in black Africans, and as tuberculosis was demonstrated in 2 of the present cases and in 2 previous cases (Abrahams et al., 1962), without attributing to it a causal role, it is possible that in 2 of the cases of Beheyt and Vandeputte (1958) this relation also applies.

Other possible aetiological factors are syphilis, rheumatic disease, coronary artery disease, and connective tissue disorders; in none of the present cases were they detected, neither was there any evidence of myocarditis though Beheyt and Vandeputte (1958), in their third case, found giant cell myocarditis. Roberts and Wibin (1966) have described a case of panaortitis with giant cell myocarditis but without ventricular aneurysm in a black African from the Republic of Congo. They suggested that a granulomatous myocarditis may be the initiating factor in subvalvular aneurysms, but is not seen in the fully formed case.

Congenital weakness of the fibrous valve rings as postulated by Abrahams et al. (1962) has certain features in its favour, for the disease occurs in young patients; however, the nature of the defect has yet to be elucidated. Pellatt (1972) described a case of subtitral aneurysm associated with stenosis of the ostium of the coronary sinus and suggested that venous obstruction with consequent chronic ischaemia was the cause of the aneurysm. However, in 2 other subvalvular aneurysms examined by him no abnormality of the ostia of the coronary sinuses was found. Similarly, in the 2 hearts available for macroscopical examination by the present authors, the ostia of the coronary sinuses showed no abnormality. Consequently, it must be considered that either subvalvular aneurysms are a heterogeneous group with differing aetiologies, or the association described by Pellatt (1972) was fortuitous.

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References


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