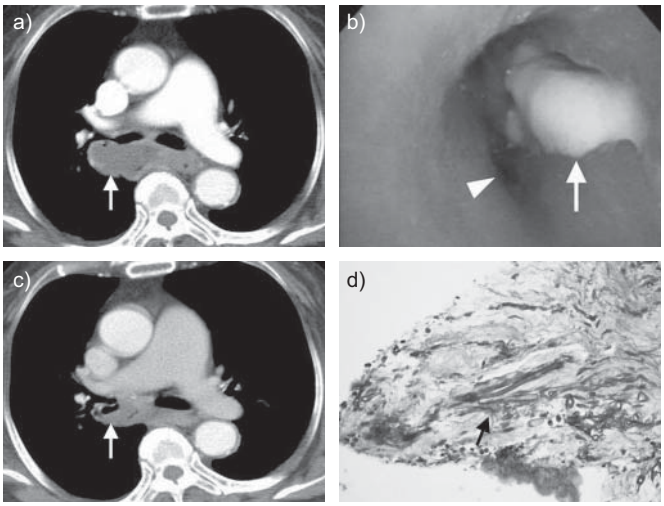

FIGURE 1. Mediastinal zygomycosis in a 76-yr-old diabetic female. a) The initial chest computed tomography (CT) scan showed a 5.7 × 5.5 × 3.7-cm mass lesion with air density on the right hilum (arrow). b) Fibreoptic bronchoscopy revealed a wedge-shaped perforation on the membranous portion of the right main bronchus and whitish caseous tissue in the mediastinum (arrow). It was also noted that the narrowed intermediate bronchus was compressed by the mediastinal lesion (arrowhead). c) After antifungal treatment for 2 months, repeated CT revealed a decrease in the size of the mediastinal mass (arrow). d) Periodic acid–Schiff-stained mediastinal tissue section showed broad, pauciseptate hyphae with branches at right angles (arrow) within necrotic tissue. Magnification: 200x.

because macrophages in diabetic hosts are impaired in the suppression of spore germination [9]. Because zygomycosis has the characteristic features of angioinvasion, vascular thrombosis and tissue necrosis [10], mediastinal zygomycosis may erode into the adjacent bronchus and produce a bronchial perforation. Though the second mechanism is a more reasonable explanation for the presentation in this case, further studies are still needed for confirmation.

Based on an early diagnosis and reversal of the underlying predisposing condition, this patient with subacute presentation of mediastinal zygomycosis was treated successfully with AMB and L-AMB. Along with histoplasmosis and tuberculosis, zygomycosis should be considered in patients with clinical manifestations of chronic mediastinitis and bronchial perforations.

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Pulmonary endarterectomy in sickle cell haemoglobin C disease

To the Editors:

Pulmonary hypertension (PH) is a common complication of sickle cell disease (SCD), with a prevalence of 10–20%, and is often a cause of death in this patient population [1]. PH can be related to two different mechanisms: 1) chronic haemolysis with endothelial dysfunction, reduction in nitric oxide bioavailability and subsequent proliferative vasculopathy; or 2) vaso-occlusive complications resulting from erythrocyte sickling and hyperviscosity [2]. According to a recent report, up to 23% of SCD patients with PH have evidence of perfusion

mismatch on ventilation/perfusion scan and 11.5% have evidence of chronic thromboembolic disease on computed tomography (CT) pulmonary angiograms [3]. Although pulmonary endarterectomy (PEA) is the best option for patients with PH related to chronic thromboembolic disease, chronic haemolysis with subsequent proliferative vasculopathy in the distal vessels put patients with SCD at increased risk of residual PH after PEA. In addition, SCD patients may be at increased risk of developing *in situ* thrombosis in the proximal vessels which can be difficult to differentiate from true thromboembolic events on radiological imaging.

Herein, we present the case of a patient with SCD due to sickle cell haemoglobin (Hb) C disease (HbSC) and chronic thromboembolic pulmonary hypertension (CTEPH) who underwent successful PEA with complete normalisation of the pulmonary haemodynamics after surgery. This patient presented with several complications of erythrocyte sickling and hyperviscosity, but had no evidence of haemolytic anaemia, supporting the possibility that PEA may be particularly helpful in SCD patients developing PH as a consequence of erythrocyte sickling and hyperviscosity rather than chronic haemolysis.

A 52-yr-old male with HbSC and protein S deficiency in New York Heart Association (NYHA) class III was diagnosed with CTEPH. His history revealed episodes of acute chest syndrome, bone pain crisis and retinopathy, but no history of priapism, leg ulcers or stroke. Blood electrophoresis revealed Hb of 118 $g \cdot L^{-1}$, HbS 51%, HbC 46%, HbA_2 3% and normal HbF. Abdominal CT identified splenic atrophy and renal wedge infarcts with an elevated serum creatinine of 144 $\mu mol \cdot L^{-1}$. His blood analysis showed no evidence of haemolytic activity with normal lactate dehydrogenase, reticulocyte count and bilirubin.

TABLE 1 Pre- and post-operative investigations

	Before surgery	After surgery
Right heart catheterisation		
$P_{pa,sys}$ mmHg	61	25
$P_{pa,dias}$ mmHg	34	8
$\bar{P}_{pa}$ mmHg	41	16
Cardiac index $mL \cdot min^{-1} \cdot m^{-2}$	1.9	4.6
Total PVR $dyn \cdot s \cdot cm^{-5}$	924	136
6-min walk test		
Distance m	465	578
Borg dyspnoea score	10	0
Lowest saturation %	69	100
Echocardiogram		
RV dysfunction	Severe	Mild
RV size cm	6.2	3.8
$P_{pa,sys}$ mmHg	81	32
Tricuspid regurgitation	Moderate	Trace
Brain natriuretic peptide		
Level $pg \cdot mL^{-1}$	426	13

$P_{pa,sys}$: systolic pulmonary artery pressure; $P_{pa,dias}$: diastolic pulmonary artery pressure; $\bar{P}$pa: mean pulmonary artery pressure; PVR: pulmonary vascular resistance; RV: right ventricle.

His pre-operative investigations are summarised in table 1 and figure 1.

After a multidisciplinary discussion, the decision was made to proceed with PEA. A 20-U exchange transfusion was performed 4 days pre-operatively. After the exchange transfusion, blood electrophoresis showed HbS 14%, HbC 13% and

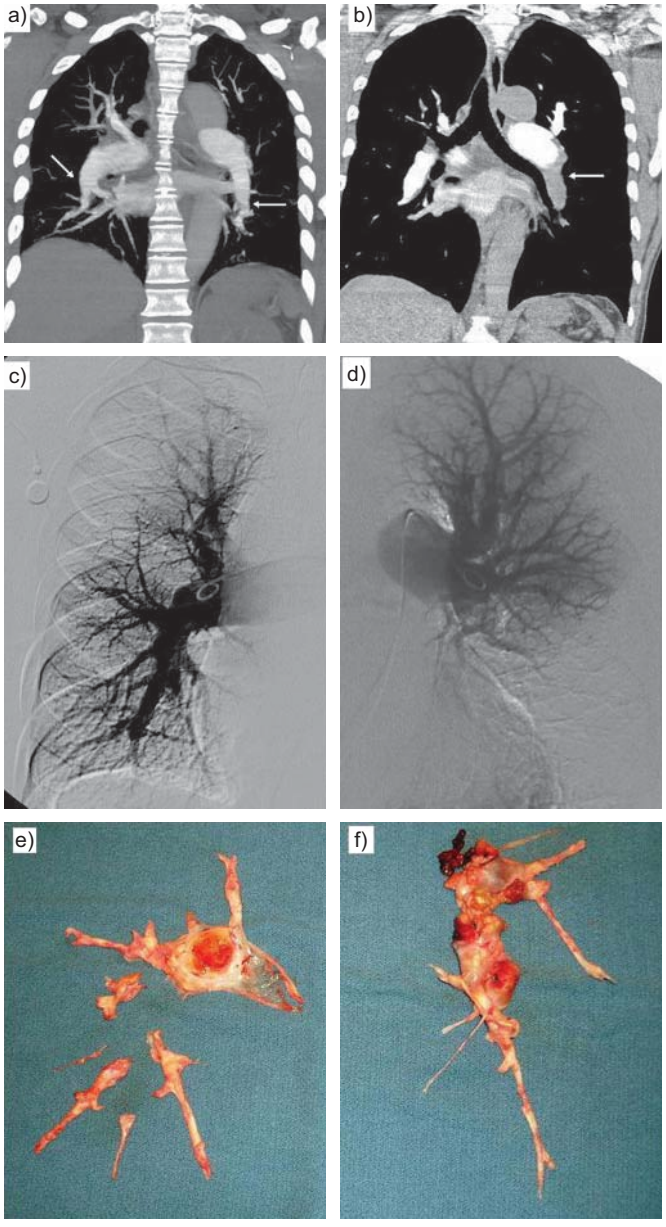

FIGURE 1. a) Computed tomography (CT) pulmonary angiogram showing evidence of webs in the descending branch of the right and left pulmonary artery (arrows). b) CT pulmonary angiogram performed 1 yr later after referral to our centre, demonstrating the presence of new large thrombi in the left pulmonary artery (arrow) that occurred despite treatment with low molecular weight heparin. c) Pulmonary angiogram of the right pulmonary artery showing evidence of segmental defects in the right upper and lower lobes. d) Pulmonary angiogram of the left pulmonary artery showing an occlusion of the descending branch to the left lower lobe. e) Pulmonary endarterectomy specimen of the right lung. f) Pulmonary endarterectomy specimen of the left lung.

HbA 71%. On cardiopulmonary bypass (CPB), an additional 1 L of blood was exchanged with 2 U of packed red cells and haematocrit was reduced to ~25%. Blood electrophoresis performed after this second exchange revealed HbS 13% and HbC 12%. The patient was cooled to 18°C. Blood gases were managed using α-stat. Successful PEA of the right and left pulmonary artery was performed with two circulatory arrests totalling 32 min (fig. 1). The patient was rewarmed to a core temperature of 37°C and weaned easily from CPB. Total CPB and cross-clamp times were 239 and 107 min, respectively. The patient was extubated on the first post-operative day and discharged from hospital on the sixth post-operative day. After PEA, the patient presented with complete normalisation of his pulmonary haemodynamics and remained in NYHA class I 15 months after surgery (table 1).

This case demonstrates that normalisation of pulmonary haemodynamics is possible after PEA in patients with SCD. Our patient had a diagnosis of HbSC and presented with typical complications of hyperviscosity (acute chest syndrome, bone pain crisis and retinopathy) and no evidence of haemolytic anaemia [2]. The higher frequency of complications from hyperviscosity in patients with HbSC supports the notion that PH may be more frequently related to local thrombosis and embolic events than to a proliferative vasculopathy related to haemolytic anaemia in this subgroup of patients with SCD disease. The presence of an atrophic spleen may also have been a potential risk factor for the development of CTEPH in our patient [4, 5]. However, the results of PEA in patients with previous splenectomy may carry increased risk of residual PH after surgery due to the presence of thromboembolic disease in the small pulmonary arteries [5, 6].

Patients with SCD are at risk of sickling crisis during deep hypothermic circulatory arrest (DHCA) because of hypothermia, hypoxia, acidosis and low-flow states. Although there is no consensus on absolute safe values of HbS in patients undergoing surgery, it is proposed that the level of HbS should be reduced to <30% for major surgical procedures and <10% for patients undergoing cardiac surgery [7]. As shown in this case and in an additional three cases reported in the literature, PEA is possible under DHCA after exchange transfusion if HbS is reduced to ~10% [8, 9]. During CPB, particular attention should be paid to maintain good flows to limit end organ ischaemia, maintain normal acid–base status and limit the duration of circulatory arrest periods. The use of a cell saver is controversial, but filtered and washed blood from the cell saver system may be more prone to sickling and was, therefore, not used in our patient.

In conclusion, the current report supports the feasibility of PEA under DHCA in patients with SCD. This case also demonstrates that normalisation of the pulmonary haemodynamics is possible after PEA in patients with SCD. Hence, a thorough evaluation for potentially curable (*i.e.* by surgery) chronic thromboembolic disease should be undertaken for all patients presenting with PH, irrespective of the underlying disease.

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