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SICKLE CELL DISEASE, PULMONARY THROMBOEN-
DARTERECTOMY, AND DEEP HYPOTHERMIC CIRC-
LATORY ARREST

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Patients with sickle cell disorders (sickle cell anemia [HbSS], sickle hemoglobin C disease [HbSC], and the sickle β thalassemias) may suffer from chronic pulmonary thromboembolic disease, resulting in pulmonary hypertension and right heart strain, eventually leading to cardiac failure and death. For patients with surgically accessible disease, pulmonary thromboendarterectomy (PTE) requiring deep hypothermic circulatory arrest represents the only possible curative intervention. We present a patient with sickle cell disease who presented for PTE.

Case Report: The patient was a 53-year-old African-American female with sickle cell anemia, chronic pulmonary thromboembolic disease, and pulmonary hypertension. On admission, the patient's pulmonary artery pressure was 73/24, mean 42mmHg, pulmonary vascular resistance (PVR) 554 dynes/sec/cm⁵, and hematocrit 30%. Hemoglobin electrophoresis confirmed her hemoglobinopathy with 64% HbS and 34% HbF. After a complete (five-unit) exchange transfusion the day prior to surgery and a partial (2-unit) exchange transfusion at the initiation of cardiopulmonary bypass, the patient underwent uneventful bilateral pulmonary thromboendarterectomies

requiring 46 minutes of circulatory arrest at 18°C. Intraoperative hemoglobin electrophoresis demonstrated 80.5% HbA, 11.5% HbS, and 5.1% HbF. During periods of cardiopulmonary bypass, hemoglobin levels were maintained between 6.6 and 9.3 g/dl, and the base excess peaked at -5.9 before correction with sodium bicarbonate. There were no complications from the procedure, which resulted in a modest decrease in PVR (491 dynes/sec/cm⁵). The patient was extubated on the second postoperative day, and discharged on postoperative day seven.

Discussion: The medical literature concerning the management of sickle cell patients who require profound hypothermic circulatory arrest is limited to one report of two cases¹. Goals in management include maintenance of low HbS concentration (<40%), adequate tissue oxygenation and perfusion, normal pH, and normothermia during non-bypass periods. PTE with deep hypothermic circulatory arrest can be performed safely in sickle cell disease patients if these goals are met. The pre-operative preparation, induction and maintenance of anesthesia, as well as significance of peri-operative exchange transfusion(s) will be presented.

References:

1. Yung GL, Channick RN, Fedullo PF, Auger WR, Kerr KM, Jamieson SW, Kapelanski DP, Moser KM Successful Pulmonary Thromboendarterectomy in Two Patients with Sickle Cell Disease. *Am J Resp Crit Care Med*1998;157:1690-1693