CASE CONFERENCE

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CASE 5—2016
Complex Congenital Cardiac Surgery in an Adult Patient With Hereditary Spherocytosis: Avoidance of Massive Hemolysis Associated With Extracorporeal Circulation in the Presence of Red Blood Cell Fragility

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EXTRACORPOREAL CIRCULATORY support among patients with genetic disorders producing erythrocyte fragility requires specific considerations involving decreasing shear stresses on red blood cells (RBCs) and vigilant intraoperative and postoperative monitoring for catastrophic hemolytic anemia. Hereditary spherocytosis (HS) is an autosomal dominant hemolytic anemia characterized by spheroid-shaped erythrocytes with increased osmolality and rigidity. Clinical presentation of HS varies depending on genetic penetrance. Decreased flexibility within the RBC membrane limits deformation and increases the possibility of hemolysis. Specifically, the mechanical stress of cardiopulmonary bypass (CPB) on HS erythrocytes presents a challenge during cardiac surgery. Although previous case reports of successful use of CPB during cardiac surgery in HS patients have been published,[1-13] significant perioperative hemolysis also has been reported.[6] The authors report the successful use of CPB in an adult HS patient undergoing multiple complex cardiac congenital repairs and review perioperative concerns and management.

CASE REPORT

A 28-year-old female with HS presented for closure of a ventricular septal defect (VSD), resection of a subaortic membrane, septal myectomy, aortic valve (AV) replacement, and repair of her dilated ascending aorta. Her medical history included a mild presentation of HS, requiring blood transfusions on 2 occasions. She had not undergone a previous splenectomy. Her preoperative echocardiogram revealed the following: severe asymmetric basal septal ventricular hypertrophy with an associated subaortic membrane (peak left ventricular outflow tract gradient of 112 mmHg with amyl nitrite provocation), a stenotic bicuspid AV (peak/mean gradients of 83/44 mmHg), a dilated ascending aorta (4.7 cm), and a perimembranous VSD (Figs 1–3). Additional congenital abnormalities identified included a left persistent superior vena cava and an anomalous origin of the circumflex coronary artery originating from the right coronary artery. Preoperatively, she was mildly anemic with a baseline hematocrit (Hct) value of 27% and a reticulocyte count of 12.8%. Her total bilirubin count was 3.9 mg/dL. All other preoperative laboratory values, including an electrolyte panel and coagulation studies, were within normal limits.

Intraoperative management included a hemodynamically stable, titrated anesthetic induction with midazolam (2 mg), fentanyl (250 μg), etomidate (12 mg), and rocuronium (50 mg). An anesthetized state was maintained with the combination of a continuous sufentanil infusion (0.3 μg/kg/h) and inhaled isoflurane (1.0%). Aminocaproic acid was administered as a bolus of 50 mg/kg and infused at 25 mg/kg/h. Intraoperative monitors included standard American Society of Anesthesiologists monitors, a left brachial arterial catheter, a right internal jugular pulmonary artery catheter, and intraoperative transesophageal echocardiography. After systemic anticoagulation with 400 U/kg of heparin, the aorta was cannulated with a 21-Fr straight-tipped arterial cannula. Bicaval venous cannulation was achieved with two 29-Fr venous cannulae. Retrograde autologous priming of the CPB pump was used to limit hemodilution. After systemic anticoagulation was verified by an activated clotting time of greater than 480 seconds, CPB was initiated with a centrifugal pump. Warm blood cardioplegia was administered to arrest cardiac electrical activity and to promote systemic normothermia.

During CPB, cardiac indices ranged from 3.2 to 4.2 L/min/m^2. Venous blood temperature variation was kept to a minimum (34.8°-36.6°C). The aortic cross-clamp time was 50 minutes, and separation from CPB was uneventful, occurring at 60 minutes without pharmacologic support. The patient

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1053-0770/2602-0033$36.00/0
http://dx.doi.org/10.1053/j.jvca.2015.11.016
Key words: hereditary spherocytosis, extracorporeal circulation, cardiopulmonary bypass, red blood cell fragility, hemolysis, congenital cardiac surgery

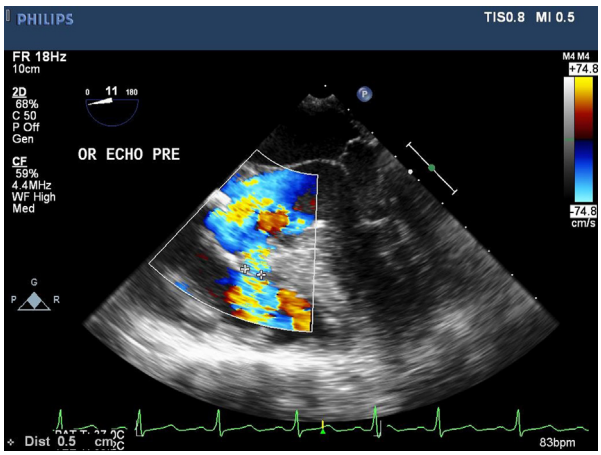

Fig 1. Transesophageal echocardiography, midesophageal 4-chamber still image displaying color-flow across the interventricular septum, consistent with a perimembranous ventricular septal defect measuring 0.5 cm.

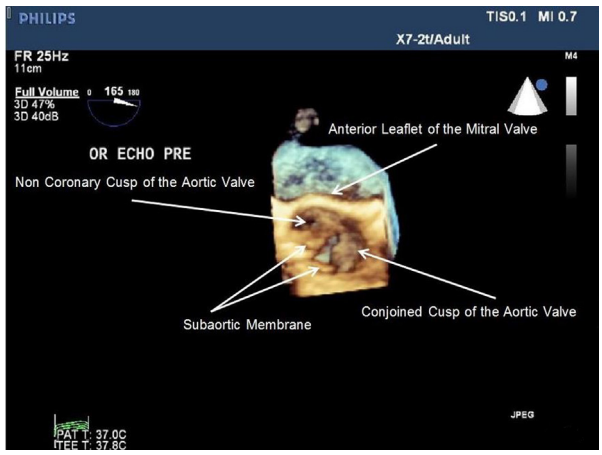

Fig 3. Transesophageal echocardiography, midesophageal, short-axis aortic valve still 3-dimensional image displaying the bicuspid composition of the aortic valve and the presence of a subaortic membrane within the left ventricular outflow track.

underwent successful closure of her VSD, subaortic membrane resection, a septal myectomy, AV replacement with a 21-mm Carpentier Edwards bioprosthesis, and ascending aortoplasty. Her Hct value varied from 21% to 18% during CPB. After a transfusion of 555 mL of autologous cell-saver-recovered blood, the final intraoperative Hct was 29%. On arrival to the ICU, the patient's cardiac index was 4.4 L/min/m^2, and her airway was extubated 2 hours later. Due to persistent anemia (Hct 20%) and mild hypotension, the patient received 1 U of packed RBCs 4 hours after surgery and 2 additional units on postoperative day 1. Evaluation at this time was consistent with hemolysis—with lactate dehydrogenase 470 U/L, haptoglobin <20 mg/dL, and reticulocyte count 22.2%. The total bilirubin on postoperative day 3 was 3.6 mg/dL and the reticulocyte count was 15.7. She was discharged on postoperative day 7.

DISCUSSION

Although recessive inheritance patterns have been reported, HS typically presents as an autosomal dominant disorder and is

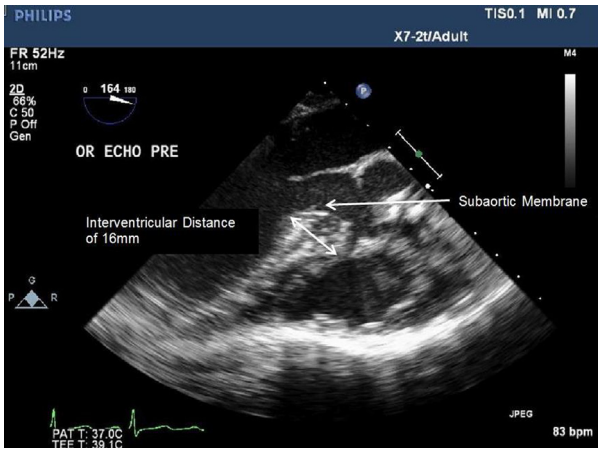

Fig 2. Transesophageal echocardiography, midesophageal long-axis still image displaying a hypertrophied interventricular septum and subaortic membrane.

the most common form of chronic hemolytic anemia among Northern Europeans and Americans.[14] Within the Caucasian population, the incidence is reported as 1 in 2,000.[14] Erythrocyte membrane protein deficiencies (spectrin, ankyrin, band 3-protein, protein 4.2, and Rh complexes) decrease the cell surface area and volume, producing rigid, spherical-shaped erythrocytes.[15-18] Spherocytes, unable to traverse the microcirculation, are retained in the splenic pulp and eventually destroyed. The clinical presentation varies from mild to severe anemia.[14,19] Formal classification of HS severity depends on the following variables: hemoglobin, bilirubin, and reticulocyte counts.[14] Physical examination may be remarkable for splenomegaly, jaundice, and scleral icterus. Hemolytic, aplastic, and megaloblastic crises may occur in severe cases.

Currently, medical therapy involves supportive management of the chronic anemia. Mild presentations may require an occasional transfusion, whereas severe presentations of persistent anemia may require splenectomy. Despite the persistence of spherocytes after splenectomy, the adverse effects of chronic hemolysis disappear; serum bilirubin levels and reticulocyte counts normalize.[14] Although hemoglobin levels increase after splenectomy, the presence of rigid RBCs unable to navigate the microvasculature remains. These erythrocytes are no longer sequestered within the spleen, which predisposes HS patients to thrombotic events.[20-22] In recent years, removal of the spleen has only been recommended for severe presentations of HS because of the increased incidence of stroke and myocardial infarction in post-splenectomy HS patients.[14]

Preoperative considerations for HS patients undergoing cardiac surgery using CPB include a detailed history and evaluation regarding the severity of the disease. Baseline hemoglobin, total bilirubin, and reticulocyte counts should be obtained and provide an indication of the disease severity. The frequency of previous transfusions provides an indication of genetic penetrance of HS and the fragility of the individual patient's erythrocytes. Although previously debated, a splenectomy before CPB (to eliminate subsequent splenic sequestering of the spherocytes damaged with extracorporeal circulation) is no longer required.[6,7] Only 9 of the 15 reported HS patients