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Case Report

The Successful Use of an Exchange Transfusion in a Child with Hereditary Spherocytosis Undergoing Congenital Cardiac Surgery

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Abstract: Little is reported in the literature regarding hereditary spherocytosis (HS) and cardiopulmonary bypass (CPB). We present a case of a 19-month-old girl child who was referred for an atrioventricular septal defect (AVSD) and HS. The patient underwent surgical repair, and an exchange transfusion was performed at the initiation of CPB. No significant hemolysis or events attributed to HS were observed during or after CPB. The surgical repair of an AVSD in a pediatric patient with HS and total volume exchange transfusion is herein reported. **Keywords:** hereditary spherocytosis, cardiopulmonary bypass (CPB), open-heart surgery, hemolysis, exchange transfusion. *J Extra Corpor Technol. 2021;53:137–9*

Hereditary spherocytosis (HS) is an inherited disease that affects the red blood cells (RBCs). Characteristic symptoms of HS are hemolytic anemia, jaundice, and splenomegaly. HS affects about 1 in 2,000 individuals in North America (1). Dysfunctional structural proteins of RBCs lead to cells that are poorly flexible, osmotically fragile, and spherically shaped. Cardiopulmonary bypass (CPB) in patients with hematologic diseases, although infrequent, introduces the potential for perioperative complications. CPB results in unavoidable hemolysis, destruction of platelets, and protein denaturation. Thus, CPB in patients with hematologic disease such as HS can result in catastrophic hematologic complications.

Description

A 19-month-old girl child with HS and an atrioventricular septal defect (AVSD) was admitted before surgery. On admission, preoperative laboratory values of significance are found in Table 1. Transesophageal echocardiogram demonstrated a transitional AVSD with moderate-to-severe valve regurgitation, primum atrial septal defect (ASD), inlet ventricular septal defect (VSD), mild pulmonary stenosis, and moderate dilation of the right heart. An arterial blood gas was recorded to assess the baseline status of the hematocrit, lactate, and coagulation before the initiation of bypass and the exchange transfusion (see in Table 1).

After median sternotomy, 400 units/kg of heparin was administered; aorta and both cavae were cannulated. Before initiating CPB, an exchange transfusion was performed using a blood-primed CPB circuit consisting of 450 mL of RBCs and 220 mL of fresh frozen plasma. This was performed by placing a ¼-inch wye connector in the venous line proximal to the reservoir and then connecting a ¼-inch line to the wye connector with a spike adapter into a blood bag. While keeping the venous line directly proximal to the reservoir clamped and opening the ¼-inch line to the blood bag, we were able to let the patient's entire blood volume drain into the blood bag while transfusing the blood prime via the aortic cannula. In total, 85 mL/kg of patient blood was removed and replaced as described earlier. An oxygenator with an integrated filter and roller pump were used to initiate CPB in a normal fashion. A perfusion index of 1.8 L/m^2/min to 2.4 L/m^2/min was established and maintained throughout CPB. The patient was cooled using moderate hypothermia (32°C). Cardioplegia was administered in antegrade fashion, using single dose of 20 mL/kg

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Table 1. Patient laboratories during course of stay.

	Preoperative	Pre-CPB (Intraoperative)	Post-CPB (Intraoperative)	Postoperative
Hemoglobin (g/dL)	10.0	6.0	9.6	12.7
Hematocrit (%)	30.6	18.0	29	37.7
Platelet (k/mm^3)	217	-	-	160
Bun (mg/dL)	11.0	-	-	12.0
Creatinine (mg/dL)	0.25	-	-	0.23
Bilirubin (mg/dL)	2.1	-	-	2.1
Plasma-free hemoglobin (mg/dL)	10.0	-	-	70.0
Haptoglobin (mg/dL)	<1.0	-	-	27.0
Lactate (mmol/L)	1.03	0.82	1.33	0.93
Activated clotting time (seconds)	-	141	104	-

BUN, blood urea nitrogen.

in a 1:4 ratio using del Nido cardioplegia. After complete repair (single-patch closure of AVSD and closure of the left atrioventricular zone of apposition), the patient was rewarmed to 36°C. Total CPB and cross-clamp times were 57 and 35 minutes, respectively. The patient had adequate urine output while on CPB and had no gross evidence of hemolysis. After separation from CPB, an arterial blood gas was again drawn to assess hematocrit, lactate, and coagulation for comparison (see Table 1). In addition to the blood prime, the patient was transfused 140 mL PRBCs, 105 mL platelets, and 30 mL cryoprecipitate before leaving the operating room. The patient was transported to the CVICU in a hemodynamically stable condition without significant bleeding.

The patient had an uneventful postoperative course, no postoperative transfusions, and was discharged on postoperative day three. At the 1-year follow-up appointment, the patient was doing very well, no residual defects or symptoms of shortness of breath or cyanosis were noted, and good cardiac function on echocardiogram was observed.

Discussion

HS is most commonly found in Northern European and Japanese families, affecting one in 2,000 individuals (2). De novo ANK1 is the most common gene mutation, resulting in more than 66% of HS cases (3). A review of the literature produced only a few case reports of children with HS undergoing open-heart surgery for congenital heart disease (4–7). There were no reports of a patient with HS and an AVSD. There have been many approaches proposed to manage hemolysis during surgery in HS patients including preemptive splenectomy, the use of a non-ionic antihemolytic detergent poloxamer 188, cardiac surgery without CPB, and haptoglobin administration to reduce plasma-free hemoglobin (4–6). Poloxamer 188 protects the RBC membranes without increasing the serum-free hemoglobin concentration during CPB (5). Hara et al. (6) reported a 10-year-old girl child with HS who underwent successful ASD closure on CPB and used poloxamer 188 and haptoglobin. They concluded that this therapy was effective in preventing intraoperative hemolysis. Aoyagi et al. reported a 9-year-old girl child with an ASD and HS who stated that CPB did not cause a significant hemolysis (7). Dal and Kumar (8) concluded that short CPB times were safe for patients with HS. With limited data, a transfusion-dependent patient, high bilirubin, and a low pre-CPB hematocrit, we believed additional precautions were necessary to minimize the risk of organ failure secondary to severe hemolysis.

Splenectomy has been proposed as an effective means of preventing hemolysis in severe cases of HS before open-heart surgery (9) and has been proposed before open-heart surgery to prevent hemolysis (10). However, spherocytosis and an increased osmotic and mechanical fragility of erythrocytes continue to persist (7), which may minimize the efficacy of this recommendation. Advocates for splenectomy caution against its use in the absence of severe HS (i.e., splenomegaly and significant anemia), as there is an increased rate of infection following this procedure (5).

In the present case, a single-volume exchange transfusion and routine hemoconcentration while on CPB were used and were effective at preventing severe hemolysis. In the absence of severe anemia and splenomegaly at baseline, splenectomy was not considered necessary. Exchange transfusions have also been efficient in cardiac operations for ABO incompatible heart transplants and patients with sickle cell disease (11,12).

Conclusion

In conclusion, HS is an inherited hematologic disease of RBCs that may be encountered in patients presenting for open-heart surgery and CPB. Minimizing CPB time, particular surgical techniques and even splenectomy have been advocated as means by which severe hemolysis may be mitigated. We report the repair of an AVSD in a child with HS, using CPB and a single-volume exchange

transfusion resulting in no significant hemolysis or anemia requiring transfusion.

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