Open Heart Operation in a Child With Congenital Heart Disease and Hereditary Spherocytosis

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An 18-month-old girl with hereditary spherocytosis underwent closure of the ventricular septal defect, commissurotomy of the pulmonary valve, and patch angioplasty of the pulmonary trunk without previous splenectomy. No serious complications as a result of hemolysis occurred in the perioperative period. Open heart surgery can therefore be safely performed in young children with congenital heart disease and hereditary spherocytosis who have not previously undergone splenectomy. (Circ J 2006; 70: 1655–1657)

Key Words: Congenital heart disease; Hereditary spherocytosis; Open heart surgery

Hereditary spherocytosis (HS) is a genetically determined red blood cell membrane disorder that results in hemolytic anemia. There are only a few case reports of patients with HS who have undergone open heart surgery. Theoretically, these patients have a high risk of perioperative hemolysis and secondary renal dysfunction attributable to the deleterious effects of cardiopulmonary bypass (CPB). Because limited information is available concerning such operations for patients with HS, we report the findings of an 18-month-old child with congenital heart disease and HS.

Case Report

A 5-month-old girl was referred to hospital after a diagnosis of ventricular septal defect (VSD) and pulmonary stenosis (PS). She was suspected to have HS because her father and grandmother were known to have suffered from HS. Her red blood cells showed increased osmotic fragility, and a diagnosis of HS was made. At the ages of 7 and 17 months, 10 ml/kg of packed red blood cells were transfused in order to treat the progressive anemia. Gradually, the pressure gradient between the right ventricle and pulmonary artery decreased, and left ventricular volume increased. Echocardiography also showed non-compaction of the left ventricular muscle. Cardiac catheterization confirmed the left-to-right shunt, with a ratio of pulmonary blood to systemic blood flow (Qp/Qs) of 1.78:1. The left ventricle was dilated to 195% of the normal volume, but overall left ventricular function was normal. Central venous pressure and pulmonary capillary wedge pressure was 9 mmHg and 13 mmHg, respectively. The systolic pressure gradient between the right ventricle and distant main pulmonary trunk was 49 mmHg. Although open heart surgery using CPB has the risk of hemolytic crisis, the most recent reports suggest that the results for patients with HS without splenectomy are acceptable. Therefore, the patient was scheduled for open heart surgery at the age of 18 months. Her parents were informed of the purpose and risk of the surgical treatment, and they gave written informed consent.

She underwent closure of the VSD, commissurotomy of the pulmonary valve, and patch angioplasty of the pulmonary trunk using autologous pericardium. The CPB circuit consisted of a roller pump and a membrane oxygenator, and the CPB time was 150 min. In order to maintain the hematocrit level and osmotic pressure of the priming solution, 150 ml of packed red blood cells and 20 ml of 20% albumin were included in the total priming fluid of 245 ml. Aggressive dilutional ultrafiltration was performed during CPB. Two thousand units of haptoglobin were administered to prevent hemolytic renal dysfunction. No serious complications as a result of hemolysis occurred in the perioperative period. The postoperative peak level of total bilirubin and of lactate dehydrogenase was 4.1 mg/dl and 678 IU/L, respectively. The hemoglobin level increased to 14.4 g/dl after transfusion of packed red blood cells (Table 2). The patient recovered uneventfully and was discharged on the 15th postoperative day. At the 6-month follow-up, she was doing well. Recent echocardiography showed no residual VSD, no pressure gradient between the right ventricle and distant main pulmonary trunk, and good left ventricular function. Hematologic investigations showed the hemoglobin level to be 11.0 g/dl and reticulocyte count of 28%. The total bilirubin and lactate dehydrogenase levels were 3.8 mg/dl, and 350 IU/L, respectively (Table 2).
HS. Despite the fact that there are no previous reports of concluded that a short CPB time was safe for patients with globin concentration and prevents secondary organ failure. Haptoglobin decreases the serum free hemoglobin concentration during CPB. Administeration of haptoglobin decreases the serum free hemoglobin concentration and prevents secondary organ failure caused by hemolysis. Previous reports recommend the use of a non-ionic antihemolytic detergent, poloxamer 188, or haptoglobin. Poloxamer 188 protects the red blood cell membranes while not increasing the serum free hemoglobin concentration during CPB. Administration of haptoglobin decreases the serum free hemoglobin concentration and prevents secondary organ failure? Hara et al report a 10-year-old girl with HS who underwent successful atrial septal defect closure, using poloxamer 188, haptoglobin, and a centrifugal pump during CPB, which they concluded was effective for preventing intraoperative hemolysis5.

Because splenectomy is a very effective treatment for reducing hemolysis, thus leading to a significant prolongation of the red cell lifespan,4 it has been advocated before open heart surgery in patients with HS to prevent hemolysis during and after surgery. Gayyed et al reported that an adult patient who underwent aortic valve replacement without splenectomy had continual postoperative hemolysis and needed splenectomy 4 months later. Kaminski et al report a 3-year-old boy who underwent splenectomy prior to undergoing anatomical correction for double-outlet right ventricle to prevent serious perioperative hemolysis. How ever, splenectomy has a high risk of infection in children under 2 years of age. Guidelines for the management of HS state that splenectomy should be performed in children with severe HS (hemoglobin level: 6–8 g/dl, reticulocyte count >10%), but such surgery should be delayed until the patient is over 6 years of age. Kawahira et al report a 15-month-old child who underwent open heart surgery without previous splenectomy and they conclude that splenectomy before cardiac operations in children with HS may not therefore always be necessary.

The present case had severe HS (hemoglobin level: 7.2 g/dl, reticulocyte count: 36.8%), and she is indicated for splenectomy in the future. However, we performed open heart surgery when she was 18 months of age without previous splenectomy because congestive heart failure and cardiomegaly had developed. In this case, haptoglobin was administered and aggressive dilutional ultrafiltration was performed during CPB. No macrohematuria occurred in the perioperative period. Two units of packed red blood cells were transfused during and after the operation. The patient had a good recovery without progression of hemolytic anemia, because the transfused red blood cells did not have structural abnormalities.

Non-compaction of the ventricular myocardium is a rare cardiomyopathy with numerous trabeculations and deep intertrabecular recesses directly connected with the ventricular cavity. It is often difficult to accurately assess the significance of ventricular dysfunction caused by non-compaction of the left ventricular myocardium. Van Heerde et al performed open heart surgery in a patient with severe mitral stenosis and non-compaction of the left ventricular myocardium, with good results. Preoperative left ventricular function was normal in the present patient, and left ventricular dysfunction did not develop during the perioperative period.

In conclusion, open heart surgery can be safely performed in young children with congenital heart disease and HS who have not previously undergone splenectomy.

### Discussion

A review of the literature produced only a few case reports of children with HS undergoing open heart surgery for congenital heart disease. One of the major concerns for patients with HS is accentuation of the risk of perioperative hemolysis, because of the fragility of the erythrocytes. Ayagi et al report a 9-year-old girl with atrial septal defect and HS, and they stated that a short duration of CPB did not cause a significant degree of hemolysis. Dal et al also concluded that a short CPB time was safe for patients with HS. Despite the fact that there are no previous reports of massive or increased intraoperative hemolysis in children, we should nevertheless take precautions to prevent secondary organ failure caused by hemolysis. Previous reports recommend the use of a non-ionic antihemolytic detergent, poloxamer 188, or haptoglobin. Poloxamer 188 protects the red blood cell membranes while not increasing the serum free hemoglobin concentration during CPB. Administration of haptoglobin decreases the serum free hemoglobin concentration and prevents secondary organ failure.

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### References


