Case Report

Open Heart Operation in a Patient with Hereditary Spherocytosis: A Case Report

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A 9-year-old girl who had ostium secundum atrial septal defect (ASD) and hereditary spherocytosis (HS) is described. The patient had a history of splenectomy for HS and underwent repair of the ASD under cardiopulmonary bypass (CPB), however, no significant or persistent hemolysis was observed during and after CPB. Only 10 patients with HS who underwent cardiac operations using CPB have been reported. The case is presented due to its rarity. (Ann Thorac Cardiovasc Surg 2001; 7: 375–7)

Key words: hereditary spherocytosis, hemolysis, splenectomy, open heart operation

Introduction

Open-heart surgery for congenital or acquired cardiac lesions in patients with hematologic diseases such as inherited hemoglobinopathies, red cell dyscrasias, and coagulopathies, although infrequent, presents potential management problems during the perioperative period. Among these disorders, hereditary spherocytosis (HS) is an intrinsic, red blood cell defect resulting in hemolytic anemia. In HS, the red blood cells are spheroidal in shape and have an increased osmotic and mechanical fragility. Meanwhile, use of cardiopulmonary bypass pump during a cardiac operation causes some unavoidable hemolysis, platelet destruction, and protein denaturation. Thus, application of cardiopulmonary bypass (CPB) for patients with HS can occasionally result in fatal hematologic complications. Only 10 patients with HS who underwent open-heart surgery under CPB have been described,[1-8] to our knowledge.

In this paper, we report a patient with HS who underwent repair of a congenital cardiac defect under CPB with successful results.

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Case Report

A 9-year-old girl was admitted to our hospital for treatment of anemia. At 3 months after birth, the patient was found to have a heart murmur by her family physician. Based on the findings of cardiac catheterization and hematologic examination, the diagnoses of ostium secundum atrial septal defect (ASD) and HS were made at the age of one year. At that time, the peripheral blood smear revealed spherocytosis, and the red blood cells showed increased osmotic fragility. Family history revealed that her father also suffered from HS, and that the grandfather was known to have gallstones. On admission, the patient was neither icteric nor anemic. Physical examination showed a systolic ejection murmur and fixed splitting of the 2nd heart sound at the left 2nd intercostal space and enlargement of the spleen, which was palpable on the left subcostal region. Computed tomographic scans and abdominal ultrasonography confirmed moderate splenomegaly, but gallstones were not detected in the gallbladder or the bile duct. Hematologic investigation revealed the hemoglobin level to be 12.5 g/dl, reticulocytes, 86.3%, and platelets, 24.6×104. The total bilirubin level was 3.68 mg/dl (direct bilirubin = 1.26 mg/dl). On February 4, 1999, laparascopic splenectomy was performed without any difficulties. During the next 2 months, 800 ml of autologous blood was donated in preparation for open-heart surgery.

Five months later, the patient was readmitted for repair of the cardiac defect. Hematologic investigation at

Table 1. Perioperative plasma hemoglobin concentation

Parameters	Measured plasma hemoglobin (mg/dl)
Just before CPB	6.8
At termination of CPB	13.1
Postoperative 3 hours	44.9
Postoperative 6 hours	14.7
Postoperative 1 day	12.9

CPB: cardiopulmonary bypass.

this time showed the hemoglobin level to be 14.2 g/dl, reticulocytes 10.1%, and platelets, 34.7×104. The total bilirubin level was 0.77 mg/dl. She was operated upon on July 28, 1999. Homologous blood was not used for priming the CPB circuit, and lactate-Ringer solution, hydroxyethylated starch, 25% albumin, and Mannitol solution were used. Right thoracotomy was employed and normothermic CPB was initiated via the right femoral artery and bicaval cannulae. An ostium secundum ASD of 2.0 × 1.5 cm in size was closed under total CPB without aortic cross clamping. CPB time was 30 minutes. After termination of CPB, donated autologous blood was transfused.

Plasma hemoglobin was measured immediately before and after CPB, at 3 hours, 6 hours, and 1 day, postoperatively, as shown in Table 1. A moderate increase of the plasma hemoglobin concentration was temporarily found at 3 hours after the operation, however, it recovered with administration of haptoglobin by 6 hours postoperatively. Hemoglobinuria was not observed, and postoperative bleeding was minimal. The patient recovered uneventfully and was discharged on the 13th postoperative day.

Discussion

Hereditary spherocytosis (HS) is an intrinsic, red blood cell defect, characterized by increased red cell osmotic and mechanical fragility which leads to hemolytic anemia. On the other hand, some degree of hemolysis is unavoidable in open-heart surgery under CPB. Thus, one of the major concerns in open-heart surgery for patients with HS is an accentuation of a risk of perioperative hemolysis caused by fragility of the erythrocytes.

Generally, previous investigators[3,4,6] have emphasized that splenectomy is recommended before a cardiac operation to prevent significant hemolysis. As shown in Table 2, to our knowledge, only 11 patients with HS, including the present patient, underwent open-heart operations using CPB.[1-8] According to the results, all of the patients survived their cardiac operations without any mortality or serious morbidity. Of the 11 patients, six had received splenectomy before the cardiac operations. The other five patients underwent cardiac operations without previous splenectomy, and significant hemolysis was found in only one patient, who required splenectomy 4 months after aortic valve replacement because of

Table 2. Patients with hereditary spherocytosis who underwent open heart operations

Cases	Age (year)	Type of operation	Previous splenectomy	Perioperative complications	Preventive procedures	Results
1[1)]	16	MV repair	yes	-		survive
2[2)]	5	Correction of TOF	no	-		survive
3[3)]	-	MVR + TVR	yes	-		survive
4[4)]	16	ASD repair	yes	-	haptoglobin	survive
5[5)]	38	AVR + MVR + TVR	no	-		survive
6[6)]	60	MVR	no	-		survive
7[6)]*	67	AVR	no	hemolysis	blood exchange	survive
8[6)]	64	Post MI VSR repair	yes	-		survive
9[7)]	1.3	MV repair + PDA ligation	no	-	poloxamer 188 haptoglobin	survive
10[8)]	31	ASD repair	yes	-		survive
11	9	ASD repair	yes	-	haptoglobin autologous blood donation	survive

*: This patient underwent splenectomy for severe hemolytic anemia 4 months after the operation.
ASD: atrial septal defect, AVR: aortic valve replacement, MVR: mitral valve replacement, MV: mitral valve, PDA: patent ductus arteriosus, MI: myocardial infarction, TOF: tetralogy of Fallot, TVR: tricuspid valve replacement, VSR: ventricular septal rupture.

continued hemolysis.

The plasma hemoglobin concentration is generally considered to be one of the most sensitive and reliable indices of hemolysis.[9] We measured the plasma hemoglobin concentration in the perioperative period, and observed that with the short duration of CPB, the use of CPB did not cause increased or persistent hemolysis in this patient. Dal and colleagues[8] also found no increase of plasma hemoglobin concentration during and soon after CPB, and concluded that a short CPB time can be tolerated by patients with HS. In this patient, autologous blood was donated before open-heart surgery and was transfused immediately after termination of CPB. We believe that this procedure may be useful to prevent the mechanical hemolysis of the fragile red blood cells by CPB pump.

Splenectomy is indicated in every patient with HS, and it achieves clinical cure of hemolytic anemia for most patients. However, spherocytosis and the increased osmotic and mechanical fragility of the erythrocytes persist after splenectomy throughout life. Considering these facts, splenectomy before or during cardiac operations in patients with HS may not always be necessary for the imposition of CPB, and open-heart operations using CPB may be safely performed with essentially the same risks as those in patients without HS.

In conclusion, a patient with HS and ostium secundum ASD who had received a previous splenectomy was operated under CPB. The use of CPB did not cause significant or persistent hemolysis in this patient as observed in analyses of the plasma hemoglobin concentration. Open-heart operations using CPB in patients with HS may be safely performed with essentially the same risks as those in patients without HS.

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