Open heart surgery in an infant with hereditary spherocytosis and a review of literature

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Abstract

Hemolytic anaemia often challenges congenital heart surgery. Hereditary spherocytosis is a rare familial hemolytic anaemia. When associated with congenital heart disease, the safe performance of cardiopulmonary bypass becomes a priority. The increased risk of hemolysis during cardiopulmonary bypass could potentially lead to significant secondary organ damage. Till now, only very few reports of successful repair of a congenital heart defect in patients with hereditary spherocytosis have been reported. We report the only case of successful repair of a congenital heart defect in an infant with hereditary spherocytosis.

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Abstract : Hemolytic anaemia often challenges congenital heart surgery. Hereditary spherocytosis is a rare familial hemolytic anaemia. When associated with congenital heart disease, the safe performance of cardiopulmonary bypass becomes a priority. The increased risk of hemolysis during cardiopulmonary bypass could potentially lead to significant secondary organ damage. Till now, only very few reports of successful repair of a congenital heart defect in patients with hereditary spherocytosis have been reported. We report the only case of successful repair of a congenital heart defect in an infant with hereditary spherocytosis.

Key words : Hereditary Spherocytosis Congenital Heart Disease

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Introduction: Hereditary spherocytosis is a rare familial hemolytic disorder. This disorder involves mutations in genes responsible for membrane proteins that participate in shape changes by erythrocyte. It can be either autosomal dominant or recessive trait [1]. The red blood cells, because of the mutation, will have an abnormal spheroidal shape with resultant increased osmotic and mechanical fragility and therefore increased chances of hemolysis [1]. Cardiopulmonary bypass in these patients could potentially increase the risk of hemolysis. We describe a case of a 7 months old child with Down's syndrome diagnosed with a large Inlet ventricular septal defect (VSD), secundum atrial septal defect (ASD), moderate mitral regurgitation, and hereditary spherocytosis, for its rarity and potential associated complications in this unique situation.

Clinical summary: A 7 months old girl with Down's syndrome presented with the diagnosis of large inflow VSD, additional OSASD, moderate mitral regurgitation, and severe pulmonary hypertension. The patient had a history of treatment for neonatal jaundice. The child was diagnosed with hereditary spherocytosis at 3 months following the evaluation of fever and anemia. There was a pansystolic murmur at the left 4^{th} intercostal space with a loud pulmonary component of the second heart sound. There was a large inflow VSD, additional Secundum ASD, moderate mitral regurgitation with a cleft in the anterior mitral leaflet, and severe pulmonary hypertension on echocardiographic examination. At admission, the patient had no clinical icterus or anemia. The patient had mild compensated hemolytic anemia suggested by the presence of hyperbilirubinemia with raised serum LDH (Table -1). Peripheral smear showed mild anisocytosis with many spherocytes, constituting about 40-50% of RBCs and occasional polychromatophils (Fig 1). Preoperative hematocrit and reticulocyte count were in the normal range (Table 1)

Through a standard median sternotomy, cardiopulmonary bypass was initiated with aorto-bi caval cannulation and mild hypothermia. Anticipating the increased risk of hemolysis on cardiopulmonary bypass, a conscious decision was made to avoid hypothermia. 100 ml of blood was drained for autotransfusion and replaced with fresh frozen plasma before initiating cardiopulmonary bypass. The CPB circuit consisted of a roller pump, and in-order to maintain sufficient oncotic pressure, sufficient RBC and 20% albumin was added to the priming fluid. Ultrafiltration was used during surgery to maintain the patient's hematocrit level. The VSD was closed with a Dacron patch. The cleft in the anterior mitral leaflet was repaired with intermittent 7-0 prolene sutures. The Secundum ASD was closed with a tanned pericardial patch. CPB time was 121 min, and ACC time was 89 mins. Post repair, there was no residual VSD, and mitral regurgitation was mild. After termination of CPB, Autologous blood of 90ml was transfused. The patient was extubated the next day. The hematocrit, reticulocyte count, Bilirubin levels, and serum LDH were serially monitored during ICU stay (Table1). There was a transient rise in reticulocyte count and serum LDH levels. Postoperatively, there was no sign of macrohematuria, impaired renal, or hepatic function. The patient recovered uneventfully and was discharged on the 6^{th} postoperative day.

Discussion : Hereditary spherocytosis in patients requiring congenital heart surgery is uncommon and poses different problems for the patient and the clinician. Anaemia secondary to hemolysis can push patients with cardiac defects to the early development of heart failure if the congenital heart defect is not repaired early [2,3]. Cardiopulmonary bypass can accentuate hemolysis because of the increased fragility of the erythrocytes

and further complicate the procedure. A literature review gives only a few congenital heart disease cases with hereditary spherocytosis undergoing open-heart surgery [Table 2][2–6]. Our case is the only reported infant with hereditary spherocytosis to undergo open-heart surgery.

To decrease hemolysis risk, the use of a centrifugal pump has been described in adult patients[7]. A centrifugal pump may be advisable as it would decrease the shear stress on the fragile erythrocytes. We used a roller pump, safe use of which has been reported by Yoshimura et al[5]. Splenectomy before cardiac surgery has also been described. Kaminishi et al has described splenectomy before surgical repair of double outlet right ventricle (DORV)[2]. However, splenectomy in infants is not recommended due to the increased risk of the procedure and greater infection risk after the procedure[1]. Kawahira et al reported a 15- month-old child who underwent open-heart surgery without a previous splenectomy, and they conclude that splenectomy before cardiac operations in children with HS may not always be necessary[3]. Use of non-ionic antihemolytic detergent poloxamer 188 and haptoglobin has been reported[2,3,5]. Poloxamer 188 protects the red cell membrane and prevents the increase of serum-free hemoglobin. Haptoglobin helps in decreasing the serum-free hemoglobin.

The present case had mild grade hereditary spherocytosis characterized by the absence of anemia and laboratory evidence of hemolysis. Open heart surgery was performed at 7 months of age because of the presence of a large shunt and severe mitral regurgitation. Perioperatively, the patient did not have any hematuria, and postoperatively patient did not require blood transfusions. There was no progression in the grade of hereditary spherocytosis by the lab parameters as well, and the patient was discharged uneventfully.

To conclude, congenital heart surgeries on CPB can be safely performed in infants and other pediatric populations with hereditary spherocytosis. Adequate precautions should be taken to prevent hemolysis and secondary organ damage following CPB.

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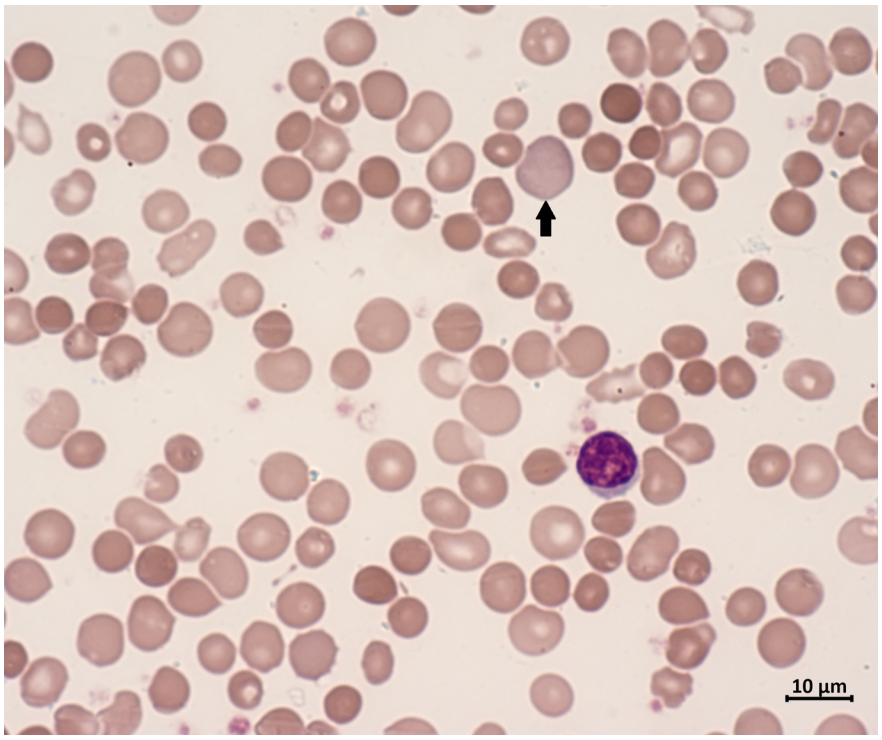

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