

# Transposition of Great Arteries and Ventricular Septal Defect in Jehovah's Witnesses

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## =Abstract=

An infant having parents of Jehovah's Witnesses was 4 months old and 5.6 kg in weight. Echocardiographic diagnosis was complete transposition of great arteries(TGA), ventricular septal defect(VSD), atrial septal defect(ASD), patent ductus arteriosus(PDA), and bilateral superior vena cava(SVC). The preoperative hemoglobin level was 14.9 g/dl. We performed an arterial switch operation(ASO) with VSD closure without transfusion and he was discharged uneventfully 16 days after the operation. At that time the hemoglobin level was 12.8 g/dl. We report that we could successfully correct the complex congenital heart disease without transfusion by the combined use of erythropoietin and aprotinin, intraoperative meticulous hemostasis, and postbypass ultrafiltration.

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**Key words:** 1. Transposition of great vessels  
2. Jehovah's Witnesses  
3. Arterial switch operation

## CASE

The cardiac surgery of the Jehovah's Witnesses patients have many problems such as overhemodilution and bleeding. Especially in early infants, they have very small blood volumes and there are many difficulties in performing the intracardiac operation without transfusion. Recently, we successfully performed the ASO of complete TGA in Jehovah's Witnesses sibling without transfusion and so report it.

A four month old infant(5.4 kg in weight) was admitted because

of heart failure and mild cyanosis. All of the patientsfamily were Jehovah's Witnesses. On admission, his oxygen saturation was 80% in room air, and hemoglobin and hematocrit were 14.9 g/dl and 45%, respectively. Echocardiography presented a sequential analysis of {S,D,D} and complete TGA with VSD. The VSD was the muscular trabecular type and with dimensions of 6×5.4 mm. Associated anomalies were PDA, secundum ASD(6.9 mm in diameter), and bilateral SVC. Eight days before the operation he was on a regimen of iron 6 ml (17.8 mg/ml) orally and 1500 units(300 units/kg) of erythropoietin injected subcutaneously 3

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times a week. The operation was done with a median sternotomy and in order to prevent hemodilution the heart-lung machine prime volume was kept to a minimum of 310 ml. The priming solution was composed of 230 ml of plasma solution(Normosol-R), 50 ml of 20% albumin, and 30 ml of 15% mannitol. Furthermore, in order to reduce the prime volume, the length of the bypass circuit was shortened(Each diameter of venous and arterial bypass circuit were 1/2" and 3/8" pump pack). Before cardiopulmonary bypass 500,000 U of aprotinin was injected and during cardiopulmonary bypass(CPB) 150,000 U were introduced into the heart-lung machine. A 10 Fr arterial cannula was inserted into the distal portion of the ascending aorta and after a 16 Fr single venous cannula was inserted into the right atrium a vent catheter was then inserted into the right upper pulmonary vein. A hematocrit of 20% was maintained during cardiopulmonary bypass with hemofiltration.

As cardiopulmonary bypass ensued the patent ductus arteriosus was ligated and divided, thereafter, mobilization of the pulmonary artery was done into the hilum. CPB was simplified due to the bilateral SVCs and in order to shorten bypass time, total circulatory arrest was done at a rectal temperature of 15°C and the VSD was closed with glutaraldehyde-fixed autologous pericardium. After cardiopulmonary bypass was restarted(low flow cardiopulmonary bypass with cardiotomy sucker) the atrial septal defect was closed and the right atrium was closed. The ASO was done with the usual methods<sup>1)</sup>. Because the discrepancy in the size of the aorta and pulmonary artery was significant, wedge resection of the valsalva sinus of the pulmonary artery(non-facing sinus of the neo-aorta) was done and in front a fold was created and sutured. The distal pulmonary artery was anastomosed to the neopulmonary artery with only autologous tissue(Pacifico method<sup>1)</sup>). Weaning from the heart-lung machine was done uneventfully and after completion of cardiopulmonary bypass a modified method of ultrafiltration was performed

The total cardiopulmonary bypass time was 217 minutes, the aortic cross clamp time was 132 minutes, and the total circulatory arrest time was 23 minutes. The postoperative hematocrit was 25%. 1500 units of erythropoietin was injected subcutaneously until 6th postoperative day and iron and folic acid were given orally until 7th postoperative day. The postoperative course was uneventful and the patient was discharged on the postoperative 16th day with a hemoglobin level of 12.8 g/dl and a hematocrit of 44.4%. Currently after 3 months of outpatient follow up the patient is well without medication.

## DISCUSSION

There are many considerations in performing open heart surgery without transfusion in early infants. The first one is hemodilution. Marked hemodilution occurs with initiation of CPB in infants. During CPB there are marked loss of oncotic activity and increased tissue edema.

And significant reduction in plasma proteins and coagulation factors is present. For preventing hemodilution we chose methods using a neonatal oxygenator, decreasing circuit tube size, and shortening the circuit length to decrease the priming volume and hemodilution.

The second one is bleeding. In neonates and infants preexisting coagulopathies are not uncommon. Furthermore, cyanotic children with significant polycythemia have a baseline abnormality of platelet function and coagulation factors<sup>2)</sup>. Shapira et al<sup>3)</sup>. also reported that low prime volume was the only independent predictor of decreased transfusion requirements and that low preoperative hematocrit was identified as a strong independent predictor of allogenic blood transfusion. In order to decrease bleeding and not to transfuse, we used aprotinin and erythropoietin. The subsequent increase in red cell mass, usually seen in the 5 to 6 days after erythropoietin stimulation, could then be taken advantage of increased potential for intraoperative autologous donation, decreased hemodilution on bypass, and faster postoperative recovery from anemia<sup>4)</sup>. Also aprotinin protects platelet function and its extracorporeal circulation activates the intrinsic coagulation pathway, fibrinolysis, and complement. As a result the improved hemostasis occurs and it is related to protection of platelets from the damaging effect of CPB<sup>5)</sup>.

In summary, in an infant of Jehovah witness family, the anatomical correction of complete TGA with VSD could be performed safely without blood transfusion by using a low prime hemodilution, combined use of erythropoietin and aprotinin, and meticulous operative hemostasis.

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**=국문초록=**

여호와의 증인을 부모로 둔 환아는 생후4개월, 5.6Kg이었다. 심초음파상 완전 대혈관 전위와 심실 중격 결손, 심방 중격 결손, 동맥관 개존증 및 양측 상대 정맥이 관찰 되었다. 수술전 혈색소 값은 14.9 g/dl이었다. 수혈 없이 심실 중격 결손 교정과 대혈관 치환술을 시행하였으며, 별문제 없이 수술 후 16일에 환아는 퇴원하였다. 퇴원 당시 혈색소 값은 12.8g/dl였다. 복잡 심기형을 가진 영아에서 수술 전 erythropoietin의 투여, 수술중 철저한 지혈 및 초여과법 등의 방법으로 수혈 없이 완전 교정술이 가능하였기에 보고하는 바이다.

- 중심 단어: 1. 대혈관 전위  
2. 여호와의 증인  
3. 대혈관 치환술