Norwood Procedure Performed on a Patient With Trisomy 13

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Summary

Trisomy 13 is associated with a variety of congenital anomalies, some of which are life-threatening and related to poor prognosis. Therefore, cardiac surgery is rarely offered to these patients, especially to those with complex cardiac anomalies. We report the case of a neonate weighing 2324 g who was born with severe congenital heart defects. Trans-thoracic echocardiography revealed the diagnoses of asplenia, single ventricle, aortic stenosis, coarctation of the aorta, hypoplastic aortic arch, and total anomalous pulmonary venous return. She was hemodynamically unstable. Palliative Norwood procedure with right ventricle–pulmonary artery conduit (RV–PA conduit) was performed at the age of 1 day to save her life. On postoperative day 7, chromosome analysis revealed trisomy 13. Echocardiography revealed good heart function; stable hemodynamic status was achieved with minimal amounts of inotropic agents. However, she developed anuria, which did not improve despite situational possible interventions, including peritoneal dialysis and continuous hemodiafiltration. On postoperative day 37, she succumbed to sudden cardiorespiratory failure. Nevertheless, this case indicates that a neonate with trisomy 13 can have a better chance at survival with cardiac surgery such as the Norwood procedure with an RV–PA conduit. (Int Heart J 2016; 57: 121-122)

Key words: Chromosome, Anomaly

The major physical characteristics associated with trisomy 13 are cleft lip and palate, microphthalmia, microcephaly, and polydactyly. Common medical conditions are congenital heart defects, respiratory difficulty, feeding difficulty, and kidney difficulty. The main cause of death is central apnea followed by heart failure. With such anomalies and causes, patients with trisomy 13 have poor prognosis; in fact, less than 10% survive beyond the first year.1 Consequently, surgery (for congenital heart disease) is rarely performed on these infants; indeed, only a few reports exist on cardiac surgery performed on patients with trisomy 13, especially surgeries with complex cardiac lesions. We report a rare case of a patient who, despite being diagnosed with trisomy 13, underwent the Norwood procedure with a right ventricle–pulmonary artery conduit (RV–PA conduit).

Case Report

At 37 weeks and 5 days of gestation, a girl was delivered by Caesarean section. She had been prenatally diagnosed with, as per fetal echocardiograms, a ventricular septal defect and coarctation of the aorta. Upon delivery, her heart rate dropped to 80 beats per minute after amniorrhexis. The Apgar score was 4 at 1 minute and 6 at 5 minute. Her weight was 2324 g, and length 46 cm. She had polydactyly and skin defects in the occipital region. Cardiac examination revealed a loud single second heart sound with a systolic murmur. Soon after birth, her blood pressure decreased and she became hemodynamically unstable. Dopamine and olprinone were administered intravenously and she was immediately intubated and placed on mechanical ventilation. On room air, an arterial blood gas assay revealed a pH of 7.12 and a lactate level of 97 mg/dL. Echocardiogram after birth led to a diagnosis of asplenia, single ventricle, aortic stenosis, coarctation of the aorta, hypoplastic aortic arch, and total anomalous pulmonary venous return into the right atrium. In an attempt to save her life, the Norwood procedure with an RV–PA conduit was performed at the age of 1 day. At a nasopharyngeal temperature of 28°C, duct tissue was resected, and the aortic arch was opened inferiorly from the ascending aorta above the aortic valve to the descending aorta. This was anastomosed to the posterior wall of the aortic arch. The neoaoorta was reconstructed by direct anastomosis of the proximal main pulmonary artery. The RV–PA shunt was performed by anastomosis with a 5-mm polytetrafluoroethylene ringed graft and a small right ventriculotomy to the distal end of the main pulmonary artery. Postoperatively, minimal amounts of inotropes were required, and her hemodynamics was stable. Chromosome analysis on postoperative day 7 revealed a diagnosis of trisomy 13. Postoperative
Echocardiogram demonstrated good univentricular heart function, and laboratory analysis showed a decline in the serum lactate levels. However, urine output gradually decreased; eventually, she developed anuria on postoperative day 9. Peritoneal dialysis and subsequent mechanical circulation for cardiac support with continuous hemodiafiltration were introduced. She was weaned off mechanical circulation support on postoperative day 14. However, anuria never recovered. On postoperative day 37, she became hemodynamically unstable and died.

**Discussion**

Trisomy 13 is associated with several life-threatening extracardiac disorders. Less than 10% of children reach their first birthday, and the most common cause of death is central apnea. Considering the short life expectancy, cardiac surgery for these patients is often not justified, especially where complex cardiac anomalies exist. Only very few patients with trisomy 13 undergo cardiac surgery. Maeda, et al reported 6 patients with trisomy 13 who underwent cardiac surgeries, only 2 of which were intracardiac repairs. Graham, et al reported 11 patients who received cardiac surgery. Notably, neither of these reports included patients who had undergone intracardiac repair for complex congenital heart disease. In the present case, our patient had two physical abnormalities, polydactyly and skin defects, and these cardiac anomalies were diagnosed prenatally, which might have suggested the underlying presence of trisomy 13. Since she had life-threatening congenital heart disease, there was no choice but to perform the palliative surgery. Less invasive palliative surgery including flow adjustable bilateral pulmonary artery banding could have been the choice for this patient in retrospect. After surgery, her hemodynamics stabilized, but she developed anuria, which did not recover despite treatment. Abdominal ultrasound demonstrated 2 kidneys and no hydronephrosis. Therefore, we speculate that she had congenital renal failure.

In conclusion, intensive cardiac and extracardiac management today may improve survival rates in patients with trisomy 13, but careful consideration must still be undertaken before performing cardiac surgeries, especially multi-staged operations, on trisomy 13 patients.

**References**