Rerouting Surgery of Cardiac Type Total Anomalous Pulmonary Venous Return in a Premature Newborn with Very Low Birth Weight

En-Ting Wu, Shu-Chien Huang,† Mei-Hwan Wu, Jou-Kou Wang, Chung-I Chang‡*

Intracardiac repair for complex congenital heart defects in premature neonates with very low birth weight (VLBW) is still a challenge to pediatric cardiac surgeons. We report the successful rerouting of cardiac type total anomalous pulmonary venous return (TAPVR) in a premature newborn (36th gestational week) with VLBW (1250 g). She had severe hypoxemia and low cardiac output despite medical treatment. Rerouting surgery of TAPVR was performed under deep hypothermia circulatory arrest at the age of 20 days. The sternum was left open and approximated 2 days later. Follow-up echocardiography showed good ventricular function without pulmonary venous obstruction. The endotracheal tube was removed 7 days postoperatively. She was then discharged without complications. In conclusion, with improved cardiopulmonary bypass technique and perioperative care, open heart surgery can be performed in premature newborns with VLBW. [J Formos Med Assoc 2007;106(2 Suppl):S23–S26]

Key Words: prematurity, total anomalous pulmonary venous return, very low birth weight

Open heart surgery in premature neonates with very low birth weight (VLBW) is considered to carry a higher risk of mortality. Therefore, many centers prefer conservative medical treatment. However, while awaiting weight gain, many premature infants experience sepsis, poor weight gain, and failure to thrive. In recent years, several reports have described successful repair of heart defects in premature infants. With advances in neonatal care, cardiologists and cardiovascular surgeons have more chances to manage premature or LBW infants with congenital heart disease. Here, we report a premature infant with total anomalous pulmonary venous return (TAPVR) to coronary sinus and restrictive patent foramen ovale (PFO). She had progressive cyanosis, poor weight gain, and low cardiac output since birth and was referred to our institute after diagnosis. Urgent open heart surgery was performed at the age of 20 days when her body weight was 1308 g. Postoperatively, the patient recovered well without complications. Our review of the literature found that this was the lowest body weight in a patient with successful open cardiac surgery reported from Taiwan.

Case Report

This premature girl was born at 36 weeks of gestation and had a birth weight of 1250 g and cyanosis and tachypnea since birth. Vital signs showed heart rate 180 beats/minute, respiratory rate 80/minute,
and oxygen saturation of 70% under room air. Physical examination found fine crackles, regular heart beat with grade II/VI systolic murmurs, and hepatomegaly. Chest radiography showed mild cardiomegaly and increased pulmonary infiltration. Arterial blood gas showed pH 7.39, pO₂ 40 mmHg, pCO₂ 50 mmHg, HCO₃⁻ 31.3 mmol/L under room air. Hemogram revealed hemoglobin 18.8 g/dL, white blood cells 15,500/μL, and platelets 305,000/μL. Echocardiography showed TAPVR, draining into coronary sinus, restrictive PFO (pressure gradient from right atrium [RA] to left atrium [LA], 10 mmHg), enlarged RA and right ventricle, and moderate degree of tricuspid regurgitation. She was intubated with mechanical ventilation. Weight gain was poor, and she had progressive hypoxemia and oliguria. Cardiomegaly and diffuse infiltration of bilateral lung fields were noted on chest radiography (Figure 1). She was sent to the operating room at the age of 20 days when her body weight was 1308 g. Vitamin K₁ was administered to prevent bleeding tendency. A percutaneous untunneled central venous catheter was not inserted as in routine cardiac surgery, and the fluid and medication were administered mainly via two peripheral lines. After disinfection, median sternotomy was performed. Cardiopulmonary bypass (CPB) was instituted with ascending aorta cannulation (6 Fr, DLP, Medtronic, Minneapolis, MN, USA) and right atrial cannulation (14 Fr right-angled cannula, Medtronic). Extracorporeal circuits (Liliput 901, Dideco, SpA, Mirandola, Italy) with 1/4 inch tubing were primed with irradiated fresh red blood cells, lactate Ringer’s solution, heparin, albumin, and bicarbonates, achieving a hematocrit value of 30%. After systemic cooling to 18°C, the aorta was crossclamped, and blood cardioplegia was infused into the aortic root. Then, circulatory arrest was undertaken and the RA was opened. The coronary sinus was unroofed and PFO was enlarged. Rerouting of TAPVR to LA and closure of the interatrial septal defect were accomplished with an autologous pericardial patch pretreated with 0.6% glutaraldehyde for 10 minutes. The patient was placed back on partial CPB and rewarmed. The heartbeat resumed normal sinus rhythm spontaneously. The total bypass time was 87 minutes and circulatory arrest was 17 minutes. A 5.5 Fr three-lumen central venous catheter was inserted through the RA appendage. Three chest tubes were placed into the pericardial and the bilateral pleural cavities, respectively. After hemostasis, the sternum was kept open and stented with a partial syringe. A silicone membrane was sutured to the skin edges with continuous prolene stitches and covered with iodine impregnated adhesive plastic drape. The postoperative condition is illustrated in Figure 2. Milrinone and dopamine infusion were given in the postoperative period. The heart rate was
144 beats/minute, blood pressure 68/39 mmHg, and central venous pressure was 7 mmHg on arrival at the intensive care unit. Arterial blood gas showed pH 7.38, pCO₂ 45.2 mmHg, pO₂ 45.2 mmHg, and lactic acid 2.5 mmol/L. The patient had good urine output. She underwent approximation of the sternum 2 days postoperatively. Inotropic agent was tapered off gradually and the RA line was removed 2 days after sternum closure. The weaning process was smooth, and she was extubated on the 7th postoperative day. Chest radiography showed normal heart size and clear lung field (Figure 3). Feeding program was advanced smoothly and she had good weight gain after the operation. Follow-up echocardiography showed normal heart function without pulmonary venous obstruction. Brain sonography showed normal brain parenchyma without intracranial hemorrhage. The patient was discharged 45 days postoperatively with the body weight of 2350 g.

Discussion

In patients with TAPVR, obstruction at any level of the pathway by which pulmonary venous blood enters the LA will cause cyanosis and low cardiac output. It is one of the surgical emergencies of congenital heart disease, because unique to this anomaly is the absence of palliative treatment for the critically ill neonate. Surgical rerouting is the only treatment even in small neonates.¹

The New England Regional Infant Cardiac Program revealed clearly that children with congenital heart disease are more susceptible to LBW, often associated with prematurity, and also that the presence of a cardiac malformation is an important cause of death among these patients.²,³ With advances in obstetrical and neonatal care, the survival rate of VLBW infants, defined as infants weighing less than 1500 g at birth, has reached 76.2% in Taiwan.⁴ Management of these newborns with congenital heart disease is an immense challenge for the pediatric cardiologist and cardiovascular surgeon. Heat labiality, vulnerability to infection, limited hemodynamic reserve, and immaturity of organ systems, particularly myocardium, lung, and liver, are factors that make surgery and perioperative care more difficult in premature than in term infants. Although the outcome of neonatal cardiac surgery has improved dramatically in the last two decades, several reports of low weight and premature age being significant risk factors for poor outcome after correction of congenital heart disease support the perception that these particularly fragile infants are too small to withstand major cardiac operation, including the eventual use of CPB.⁵,⁶ However, Chang et al⁷ reported a study of 100 neonates with LBW in which the hospital survival rate was 82% in the complete repair group, 78% in the palliated group, and 77% in the delayed surgery group. Thus, deferring an early intervention to wait for weight gain is not beneficial since the outcome of prolonged medical therapy does not exceed that of early surgery.⁸,⁹ Intensive medical support and continuous prostaglandin infusion may achieve clinical stability for delaying surgery, but this principle does not seem to be associated with a better outcome because prolonged hospitalization may increase the risk of sepsis and patients may have failure to thrive due to advanced heart failure.⁷ This is especially true for neonates with compromised hemodynamics.
In recent reports, open cardiac surgery in patients with body weight less than 2500 g could be performed with good results.\textsuperscript{10} Oppido et al\textsuperscript{11} performed cardiac surgery in 60 infants weighing less than 2500 g, and 35 of them were operated on CPB. The mortality rate was 15%, and the survivors did not have neurologic complications. Age, weight, prematurity, type of surgery, and use of CPB did not influence early mortality.\textsuperscript{11} In Taiwan, we have successfully performed arterial switch operation in a LBW infant (weight, 1900 g) with transposition of the great arteries and necrotizing enterocolitis.\textsuperscript{12}

Cardiac surgery in LBW neonates requires the collaborative work of pediatric cardiologists, cardiac surgeons, anesthetists, and intensive care specialists. Diagnosis relies on echocardiography only, and cardiac catheterization should be avoided to prevent stress, osmotic load, and possible pulmonary hypertensive crisis.\textsuperscript{13} During cardiac surgery, the CPB should be primed with fresh blood product without electrolytes imbalance, and low-pressure and high-flow CPB achieved with vasodilators. Central vein access usually could be achieved by right atrial line, and the line was maintained after the patient recovered. Delayed sternal closure was usually utilized because the edematous heart after CPB might be compressed; hence, cardiac tamponade could be avoided.\textsuperscript{14} In the intensive care unit, the fluid balance must be meticulously managed, and peritoneal dialysis can be used to augment fluid removal. The sternum was closed when the edema subsided. The patient could recover well since adequate cardiac physiology was achieved after the surgery. The aim of nutrition therapy was to achieve good body weight gain, and enteral feeding was the preferred route.

In conclusion, with the advances in cardiac surgery and perioperative management, LBW is not a contraindication for open cardiac surgery. We successfully performed an intracardiac repair for TAPVR in a 1308 g neonate, without complications.

References