

Hybrid approach in two cases with hypoplastic left heart syndrome

Hibrid yaklaşımı uygulanan hipoplastik sol kalp sendromlu iki olgu

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Abstract

Hypoplastic left heart syndrome (HLHS) encompasses a spectrum of congenital cardiac defects which mostly include left ventricular hypoplasia or atresia, mitral valve hypoplasia or atresia and ascending aortic hypoplasia and atresia. Hypoplastic left heart syndrome is a leading cause of neonatal mortality. In recent years, popularity of hybrid approach has increased. Hybrid procedures which requires close cooperations between paediatric cardiologists and cardiac surgeons includes a less invasive primary procedure; integrating interventional cardiology approaches and surgical techniques. Herein, we report two cases with hypoplastic left heart syndrome. We performed hybrid approach including ductal stenting and bilateral pulmonary banding on these patients.

Key words: Infant, newborn, hypoplastic left heart syndrome, treatment

Öz

Hipoplastik sol kalp sendromu; çoğunlukla sol ventrikül hipoplazisi veya atrezisi, mitral kapak hipoplazisi veya atrezisi ve çıkan aorta hipoplazisi veya atrezisini içeren doğumsal kalp defektleri spektrumunu kapsar. Hipoplastik sol kalp sendromu neonatal mortalitenin önemli bir nedenidir. Son yıllarda hibrid yaklaşımın popülaritesi artmıştır. Hibrid uygulama, pediatrik kardiyolog ve kalp cerrahı arasında yakın işbirliği gerektiren, girişimsel kardiyoloji yaklaşımları ve cerrahi teknikleri birleştiren ve daha az invaziv olan birincil bir uygulamadır. Bu yazıda, hipoplastik sol kalp sendromu olan ve duktal stent ve iki taraflı pulmoner arter bantlamayı içeren hibrid yaklaşımı uygulanan iki olguyu sunduk.

Anahtar kelimeler: Bebek, yenidoğan, hipoplastik sol kalp sendromu, tedavi

Introduction

Hypoplastic left heart syndrome (HLHS) encompasses a spectrum of congenital cardiac defects which mostly include left ventricular hypoplasia or atresia, mitral valve hypoplasia or

atresia and ascending aortic hypoplasia and atresia (1, 2). Postnatal clinical course of neonates with HLHS depends on the presence and magnitude of shunts-ductus arteriosus and foramen ovale (3). The first successful palliative surgical treatment of

neonates with HLHS was performed by Dr. William Iman Norwood and colleagues in 1981 (3). Norwood operation (stage I) connects right ventricle to both the systemic and pulmonary circulations. Bi-directional cavopulmonary anastomosis (BCPA) and Fontan procedures should be performed by 4-6 months and 2-4 ages, respectively to unload systemic right ventricle. During the last decade, alternative options such as hybrid palliation became available. Adequate interatrial communication with septostomy +/- stent, ensuring the patency of the ductus arteriosus to maintain systemic blood flow and protecting the pulmonary vascular bed by performing bilateral pulmonary arterial banding is aimed for this approach (4,5). In this report, we describe two neonate with HLHS who had hybrid approach.

Case 1

A 39-week neonate weighing 3815 gr was referred to our unit for the evaluation of tachypnea and low saturation. On physical examination, patient was cyanotic with a pulse oximeter reading 78%. His precordium was active with a hyperdynamic right ventricular impulse with normal peripheral pulses, and a grade 2/6 systolic ejection murmur heard along the mesocardiac area. A mild hepatomegaly was noted, and lung auscultation was normal. An echocardiogram showed a hypoplastic left ventricle, large secundum atrial septal defect and patent ductus arteriosus. Left ventricular end diastolic dimension was 7 mm, aortic valve annulus was 4 mm, isthmus and descending aorta was 5.5 mm and 7 mm, respectively. There were also mitral and aortic valve atresia. There was no antegrade flow in the aorta, but retrograde flow from the ductus to arcus aorta and its branches. We measured the annulus of atretic aortic valves. Prostaglandin E1 infusion and diuretic therapy

was begun, and decision of hybrid approach was made. Cardiac catheterisation supported the findings of echocardiography. Bidirectional shunt through atrial septal defect with a 9 mm and dilated pulmonary artery three times aorta and Krichenko type C ductus with a 6 mm diameter were noted. 8x16 mm balloon-expandable stent (RX Herculink Elite) was deployed in the ductus. Afterwards, bilateral pulmonary banding procedure was performed. No complications were noted after procedure. Tracheostomy was needed for optimal respiratory care. After one week of procedure, patient had 32 mmHg gradient through the ductal stent showing restrictive blood flow. We did not perform angiography due to sepsis and deterioration in the clinical picture. Klebsiella pneumonia was isolated in blood culture. Cultures of urine, tracheal aspirate and tip of the catheter were negative. The patient was died on the 39th day postoperatively after sudden deterioration in general condition and hypotension low cardiac output due to possible septicaemia.

Case 2

A 38-week old neonate with a suspicion of congenital heart disease on obstetric ultrasound was referred to our unit. His birth weight was 3800 gr., baby was pallor with a heart rate of 174/min and respiratory rate of 55/min. Blood pressure was 54/31 mmHg. Patient had 3/6 systolic murmur with a pulse oximetry reading of % 95, and femoral pulses were bilaterally palpable. There was no abnormality in blood tests and biochemistry parameters. Transthoracic echocardiography revealed hypoplastic left heart syndrome, secundum atrial septal defect and patent ductus arteriosus. Left ventricular end diastolic diameter was 9 mm, aortic annulus was 4.5 mm, isthmus was 5 mm, and descending aorta was 7.5 mm. Inotropic support, diuretic and PGE1 infusion was started. We started

PGE1 infusion with a dose of 0.01 mcg/kg/min after immediately birth in case who prenatally diagnosed. On the second day of hospitalisation, patient was intubated due to tachypnea and respiratory acidosis in blood gas analysis. Since, this baby is term neonate; PGE1 infusion may lead to abdominal distention, biliary and bloody gastric residue, which developed on postnatal day 2. We decided to stop enteral feeding and start antibiotic therapy due to abdominal distention and biliary, bloody gastric residues. PGE1 infusions was also stopped because of necrotizing enterocolitis. The diagnosis of sepsis and disseminated intravascular coagulopathy were considered based on increase of acute phase reactants and alterations in blood diathesis tests. On day 11 of hospitalisation, the decision of hybrid approach was made. The length and diameter of the ductus were determined by echocardiography and angiography. The angiography showed atretic mitral and aortic valves, dilated pulmonary artery, tubular ductus with a 5 mm. There was bilateral shunt through ASD with a 7 mm diameter. The same procedures including 6x18 mm balloon-expandable ductal stenting (RX Herculink Elite) and bilateral pulmonary banding were applied in this patient, and no complications after procedures were observed. Postoperative echocardiography demonstrated 12 mmHg peak instantaneous gradient in stent area. *Candida glabrata* was isolated from blood, but urine and tracheal culture were negative. Patient was lost after 23 days of procedure due to sepsis with sudden onset of bradycardia and hypotension.

DISCUSSION

HLHS is a fatal neonatal congenital heart disease which accounts for 25% of deaths during the first

week of life (4). The first successful palliative surgery in patients with hypoplastic left heart syndrome has been succeeded by Norwood et al. in 1981 (3). Stage I Norwood includes atrial septectomy, anastomosis of proximal pulmonary artery to aorta, and separation of pulmonary and systemic circulation using either a right ventricular to pulmonary artery conduit or a Blalock-Taussig shunt. Significant success has been recorded in three staged procedure during the last decade. However, general morbidity and mortality of the initial Norwood procedure and its influence on the long-term success of the following Fontan procedure remain suboptimal (6). Therefore, during the last decade, hybrid approach with bilateral pulmonary arterial banding, stenting of patent ductus arteriosus to restore unobstructed systemic output and creation of unobstructed atrial septal defect has been emerged as an alternative to Norwood operation in children with HLHS. The decision concerning the individual strategy in patients with HLHS has changed with increasing experience in making prenatal and postnatal diagnosis as well as in surgical palliations and is now based on medical and social considerations (7). Compared to almost 30 years of experience with the Norwood stage I operation the hybrid approach is relatively new. Originally reported in 1993, hybrid procedures for initial palliation of hypoplastic left heart syndrome were devised as an alternative to the stage 1 Norwood procedure. Hybrid procedures which requires close cooperation between the paediatric cardiologists and cardiac surgeons includes a less invasive primary procedure; integrating interventional cardiology approaches and surgical techniques (5).

Ductal stenting is generally recommended particularly in patients with restrictive ductus

arteriosus not responding to PGE1 therapy. In our institute, we do not prefer only Hybrid procedure; we have also experience on Norwood stage I with either a Sano shunt or modified BT shunt. We decided to perform Hybrid procedure in these cases. In our cases, hybrid approach was successful with no procedure related complications. We prefer Columbus approach, which has evolved with the cooperative modifications of Galantowicz and Chetham (8). The palliation is performed by intraoperative, off-pump, a stent in the arterial duct positioned through a sheath in the pulmonary trunk followed by placement of bands on both pulmonary arteries. Bands are placed surgically round the right and left pulmonary arteries. Bilateral pulmonary artery banding and ductal stent implantation were performed concomitantly. We did not use angiography to show ductus arteriosus in both cases. Ductus diameter was measured by epicardial 2D- echocardiography. The hybrid approach to palliation of hypoplastic left heart syndrome can be complicated by the development of neointimal formation and in-stent stenosis of the PDA. This may obstruct retrograde aortic arch flow, decrease systemic circulation, and lead to interstage interventional procedures (9). Ventricular functions, degree of tricuspid regurgitation and peak instantaneous gradient through ductus arteriosus and pulmonary artery bands should be evaluated by echocardiography during postoperative period.

We performed hybrid procedure in the sterile operating room. In the first case, tracheostomy, which was performed after 32nd day of procedure, could be the major contributing factor to sepsis. In the second case, there was clinical picture of necrotizing enterocolitis before the procedure that

may help the progression of sepsis in the postoperative period. We tried to extubate patients earlier postoperatively. Several authors suggest that full mechanical ventilation lessens the burden of respiratory effort of the patient, and accordingly eliminates the metabolic demand that spontaneous respiratory function imposes. However, after 1 month of follow-up, our cases were lost due to sepsis. Both patients were intubated and had clinical findings consistent with general septicaemia. Preoperative hemodynamic findings and the presence of aortic and mitral atresia concomitantly in both cases could be associated with this unfavourable outcome.

Initial reports regarding early results of the hybrid approach were restricted by small cohorts of patients with heterogenous diagnoses and risk stratification, short follow-up period and the effect of the learning curve of this new method. In recent years, reports have been focused on the management of hybrid approach for high risk patient with HLHS. The results of new reports from several centres comparing the risks of hybrid approach and Norwood stage 1 and 2 are promising (10, 11). Several investigators addressing the potential advantage of Hybrid approach that there may be a dramatic improvement of patient's neurological development by delaying the age of the bypass. However, there is no definite answer what effect of hybrid approach will have on neurodevelopmental outcomes in future (12). In the majority of cases, total cardiac output and systemic extraction of oxygen are the most important, most variable, and most manageable determinants of systemic delivery of oxygen in the acute post-operative period following the initial stage. Our both cases had mitral/aortic atresia, which is a severe form of HLHS; we may argue that the prognosis could be better if they had mitral

stenosis/aortic stenosis.

In conclusion, Hybrid approach is an alternative to the Norwood stage 1 for children with high risk congenital heart disease. In this report, we describe two cases with hypoplastic left heart syndrome who had hybrid approach. Hybrid approach which mandates multidisciplinary team

including paediatric cardiologists and cardiac surgeons will have potential for being a primary treatment modality for high risk HLHS infants, in future. However, it should also be noted that long-term and multicenter prospective studies are needed to determine the clinical benefits and risks of this approach.

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