CASE REPORT

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Temporary left ventricular assistance for extreme postoperative heart failure in two infants with Bland-White-Garland syndrome

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Abstract

Anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland syndrome – BWG) is a serious congenital cardiac anomaly leading to myocardial ischemia with severe heart failure. Immediate surgical correction is the treatment of choice, and the risk of postoperative complications depends on the degree of myocardial injury. The authors present two cases of infants with BWG, in whom long-term (175 and 26 days) left ventricular assistance with a Berlin Heart device was used, resulting in successful weaning from the support and subsequent hospital discharge. Because of serious hemorrhagic complications and their neurological consequences observed in the first patient, the anticoagulation protocol was modified in the second patient, providing more stable support and allowing the device to be removed after a shorter period of time.

The Berlin Heart left ventricular assist device may be treated not only as a bridge for transplantation but also, considering the shortage of donors in this age group, as a bridge to recovery.

Key words: Bland-White-Garland syndrome, left ventricular assist device, infant, intensive care.

Streszczenie

Zespół Blanda-White'a-Garlanda (BWG), czyli nieprawidłowe odejście lewej tętnicy wieńcowej od pnia płucnego, jest wadą, która prowadzi do niedokrwienia lub zawału mięśnia sercowego. Leczeniem z wyboru jest operacyjne odtworzenie fizjologicznego ukrwienia wieńcowego, a ryzyko wystąpienia powikłań pooperacyjnych zależy od stopnia uszkodzenia mięśnia sercowego. Autorzy przedstawiają dwa przypadki zespołu BWG, w których zastosowano w okresie pooperacyjnym długoterminowe (175 i 26 dni) wspomaganie lewej komory za pomocą komory typu Berlin Heart. Uzyskano poprawę wydolności mięśnia sercowego z możliwością usunięcia układu wspomagającego i wypisano pacjentów do domu. Ze względu na wystąpienie u pierwszego z niemowląt powikłań krwotocznych i poważnych następstw neurologicznych protokół antykoagulacyjny u kolejnego pacjenta został zmodyfikowany, co spowodowało stabilniejszy okres wspomagania i szybsze jego zakończenie.

Wspomaganie lewokomorowe pompą Berlin Heart może być traktowane u najmniejszych niemowląt nie tylko jako pomost do transplantacji serca, lecz przy ograniczonych możliwościach transplantacji w tej grupie wiekowej także jako metoda regeneracji mięśnia sercowego.

Słowa kluczowe: zespół Blanda-White'a-Garlanda, urządzenie do mechanicznego wspomagania lewej komory, niemowlę, intensywna terapia.

Introduction

Bland-White-Garland syndrome (BWG), i.e., abnormal origin of the left coronary artery from the pulmonary trunk, is a rare congenital defect that is at times difficult to diagnose. Its first symptom, appearing during the neonatal period or infancy, is the development of circulatory insufficiency in the course of ischemia or myocardial infarction, and the treatment of choice is prompt as possible surgical reconstruction of physiological (two-vessel) coronary blood supply [1, 2]. Due to myocardial injury, the patients may require

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postoperative mechanical assistance of cardiac function, which is treated as a bridge to heart transplantation [3].

The aim of this study is to present the treatment of two infants with BWG, admitted with myocardial infarction and dilated cardiomyopathy, in whom, due to extreme circulatory insufficiency, left ventricular assistance with a Berlin Heart pump was applied postoperatively [4].

Case reports

Case study 1

A 3-month-old girl (weight: 4.3 kg) was urgently admitted to the department with signs of heart failure. Electrocardiogram (ECG) showed signs of previous myocardial infarction, while X-ray examinations demonstrated an enlarged cardiac silhouette. Echocardiographic examination revealed BWG, a significantly enlarged spherical left ventricle with ejection fraction (EF) of 10–20%, mitral valve regurgitation (MVI) +\++, and atrial septal defect/patent foramen ovale (ASD/PFO) (ProBNP > 35 000 pg/ml). The child was qualified for urgent surgical correction of the defect, but despite the correct implantation of the coronary artery, myocardial contractility did not improve. In view of this lack of improvement, a left ventricular assist device (LVAD) in the form of a Berlin Heart pump (10 ml) was implanted after three weeks as a bridge for heart transplantation. Gradually, the patient's circulation stabilized. During 5 months of left ventricular assistance, stabilization of the international normalized ratio (INR) level during the administration of antithrombotic agents was not achieved due to problems with feeding the child and periodic infections. On the 48th day of assistance, an intracranial hemorrhage occurred during an infection (Fig. 1). An attempt to disconnect the LVAD was unsuccessful. Due to the need to reduce anticoagulation, the ventricle was replaced twice during the treatment because of thrombus formation. On the 160th day of treatment, control examinations confirmed the existence of progressive atrophic cerebral changes (Fig. 2). On the 174th day of treatment, in view of improved heart contractility and neurological contraindications for heart transplantation, the assist device was removed after a gradual reduction of the assistance parameters. Catecholamines were discontinued, and the child was extubated within the first week after the discontinuation of assistance. The ejection fraction of the heart was 70% after the assistance was ended (Fig. 3). Subsequently, the child was transferred to the Maternal and Child Health Center in Katowice for further treatment and rehabilitation. After the conclusion of treatment (hiatus hernia surgery, percutaneous gastrostomy), the child was discharged; she remains home and her circulation is fully functional, but her development is limited due to severe neurological injury.

Case study 2

A 3.5-month-old boy (weight: 5.9 kg) was admitted in severe condition: ECG indicated an infarction, and BWG was suspected. Echocardiography showed indirect signs of an abnormal origin of the left coronary artery from the pulmonary trunk (left ventricular dilatation, EF 20%, massive mitral regurgitation, dilated right coronary artery; the origin of the left coronary artery from the left coronary sinus was not visualized). On the 3rd day of the patient's stay, aortography was conducted: the left coronary artery was obscured by a shadow from the side of the right coronary artery and, subsequently, entered the pulmonary trunk. Urgent surgical correction of the defect was per-

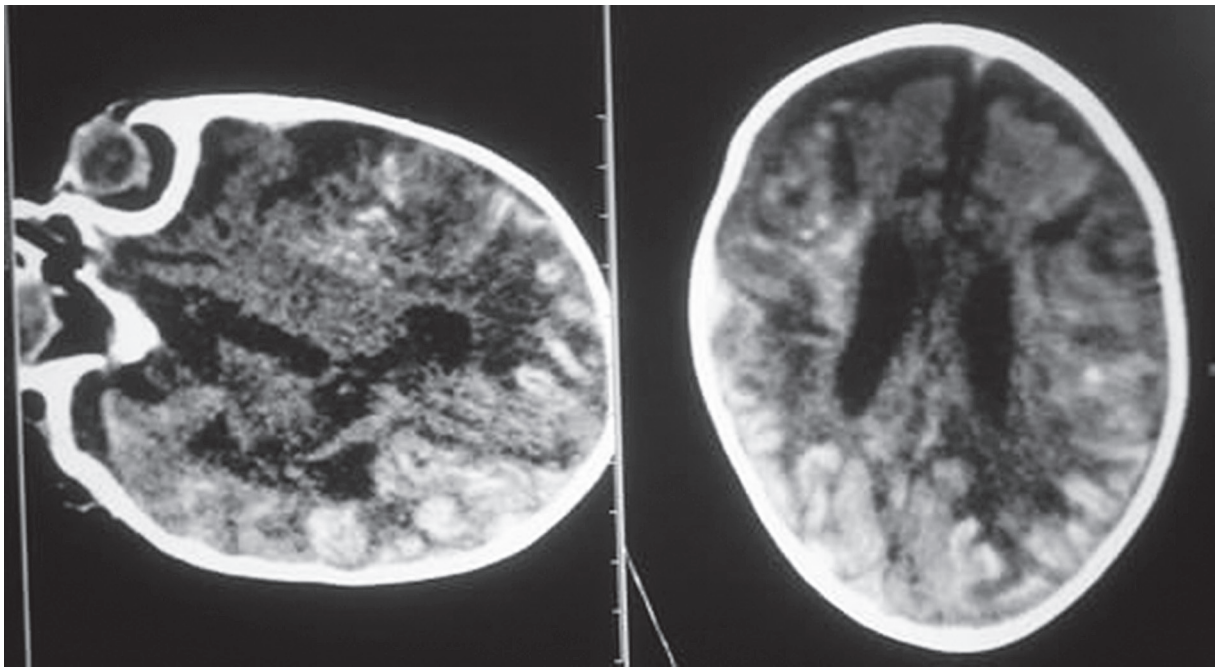

Fig. 1. Image of intracranial bleeding in the first patient

formed. The discontinuation of extracorporeal circulation was possible using adrenaline (0.3 µg/kg/min), milrinon (0.7 µg/kg/min), and inhaled nitric oxide (iNO) (10 ppm), with the chest left open. During the first hours, the child remained stable; however, on the 2nd day, cardiac dysrhythmias appeared, causing circulatory decompensation. A Berlin Heart LVAD (10 ml ventricle) was urgently implanted. Later in the postoperative course, the chest was closed, and the catecholamine doses were gradually reduced. On the 6th postoperative day, the child was successfully extubated. After 2 weeks of assistance, in view of improved myocardial contractility, the process of increasing the load on the left ventricle was started, and the LVAD was removed on the 26th day of assistance. On the next day, catecholamines were discontinued, and the child was extubated. Echocardiography demonstrated normal left ventricular systolic function (50%) and hemodynamically significant mitral insufficiency (2nd degree). After 56 days of hospitalization, the boy was discharged home with a recommendation to undergo rehabilitation. At present, the functioning of the patient's circulation is normal, as is the child's development.

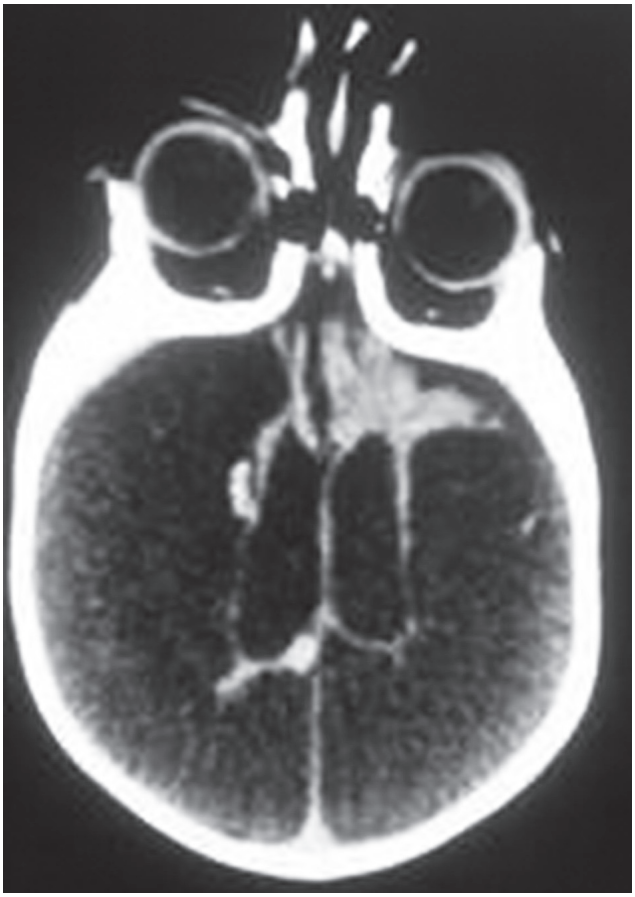

Fig. 2. Image of the first patient's brain before the end of left ventricular assistance

Discussion

Ventricular assistance with the Berlin Heart LVAD has enabled the survival of infants after BWG procedures performed during or after left ventricular infarction and during severe circulatory insufficiency. This management is treated as a bridge for heart transplantation and also, lately, as short-term postoperative assistance [4]. Conducting long-term assistance in children weighing < 10 kg is associated with 40% mortality, and bleeding complications are the most common cause of death [5]. In the first of the described cases, the occurrence of an intracranial hemorrhage on the 48th day and the observed progressive cerebral atrophy precluded the child from being qualified for a heart transplant. Disconnecting the assist device in view of serious contraindications for further therapy is a difficult decision, and no precise management guidelines have yet been formulated [6]. In this case, the decision was made after a medical consultation and many conversations with the parents. After the assistance was discontinued, the functioning of the circulatory system proved to be satisfactory. It was acknowledged that, due to the difficulties in stabilizing the INR values during therapy (infections, feeding disorders), the treatment strategy in infants should be modified [6, 7]. In the case of the second infant, the administration of oral coumarin-type drugs was abandoned completely; the child was receiving heparin infusion and aspirin or

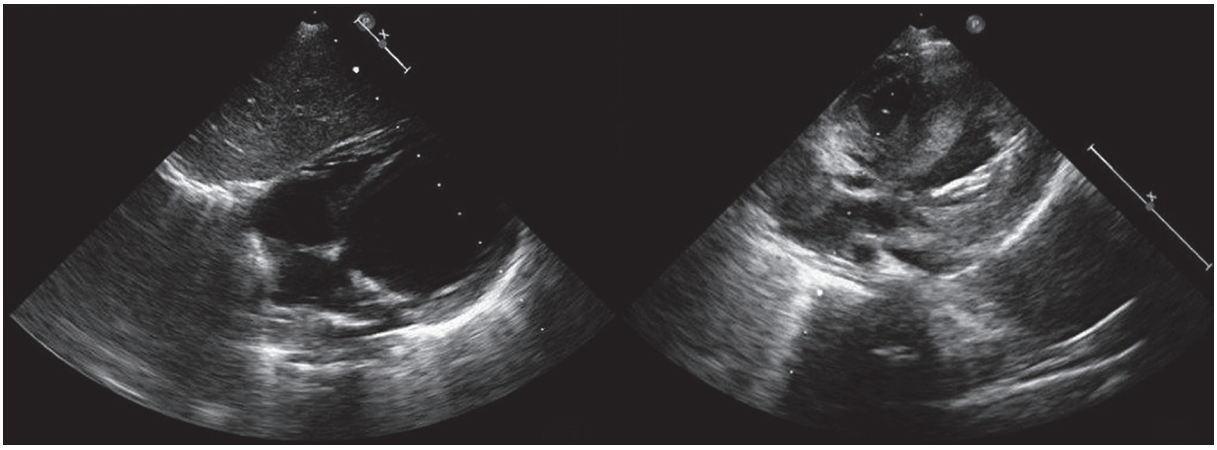

Fig. 3. Image of the first patient's heart before and after LVAD treatment

clopidogrel, while activated clotting time (ACT) and activated partial thrombaplastin time (APTT) were checked every 6 hours in order to achieve the values of 150–170 and > 80 seconds, respectively. Inflammatory, coagulation and biochemical parameters were monitored in order to detect any disorders or changes in the coagulation system; the pump was monitored as well in order to detect the development of deposits or thrombi, especially during infection. A new protocol was introduced, consisting in weekly assessments of cardiac contractility and attempts to reduce assistance in order to remove the LVAD as quickly as possible. These changes appear to have resulted in a more stable course of assistance in the second patient, who was ultimately discharged home in good general condition.

Conclusions

Left ventricular assistance with the Berlin Heart pump in small infants can be treated not only as a bridge to a heart transplant, but also, considering the limited transplantation options in this age group, as a bridge to recovery.

Establishing an efficacious and stable dose of oral antithrombotic agents in patients so small is extremely challenging and may be associated with additional risk.

Disclosure

Authors report no conflict of interest.

References

1. Haponiuk I, Rycaj J, Grzybowski A, Obersztyn A, Urbańska E, Białkowski J, Zembala M, Skalski JH. Leczenie operacyjne zespołu Blanda-White'a-Garlanda u noworodka z rozległym pozawałowym uszkodzeniem lewej komory. Czy korekcja jest bardziej korzystna od transplantacji serca? Folia Cardiol 2004; 11: 687-696.
2. Szmigielska A, Roszkowska-Blaim M, Gołąbek-Dylewska M, Tomik A, Brzewski M, Werner B. Bland-White-Garland syndrome – a rare and serious cause of failure to thrive. Am J Case Rep 2013; 14: 370-372.
3. Stege D, Kiski D, Tjan T, Wolf B, Scheld HH, Kehl HG. 1-year left ventricular assist device (LVAD) experience as bridge to heart transplantation in an infant with Bland-White-Garland syndrome. Thorac Cardiov Surg 2010; 58 Suppl. 2: S167-S169.
4. Qiu L, Yu X, Liu J, Zhang W. The left ventricular assistance device was used for anomalous origin of the left coronary artery from the pulmonary artery in perioperative period. Zhonghua Wai Ke Za Zhi 2015; 53: 430-435.
5. Conway J, Louis JS, Morales D, Law S, Tjossem C, Humpl T. Delineating survival outcomes in children < 10 kg bridged to transplant or recovery with the Berlin Heart EXCOR Ventricular Assist Device. JCHF 2015; 3: 70-77.
6. Hollander SA, Axelrod DM, Bernstein D, Cohen HJ, Sourkes B, Reddy S, Magnus D, Rosenthal DN, Kaufman BD. Compassionate deactivation of ventricular assist devices in pediatric patients. J Heart Lung Transplant 2016; 35: 564-567.
7. EXCOR® Pediatric VAD Ventricular Assist Device with Stationary Driving Unit Ikus Rev. 2.1 for Pediatric Use: Berlin Heart, Inc. www.fda.gov/downloads, www.berlinheart.com