Cardiology in the Young

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Hybrid approach for end-stage heart failure treatment in a 6-month-old baby

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Brief Report

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Abstract

A 6-month-old baby girl, with a history of 2 months of intensive care management and two episodes of cardiac arrest, was transferred from another European country to initiate the "Giessen approach" for end-stage heart failure in children. At the admission, left ventricular ejection fraction was 20%. Severe mitral valve regurgitation and severe left atrial dilatation were present. Right ventricular function was preserved, and tricuspid valve regurgitation was mild. As a result, the patient underwent surgical pulmonary artery banding. Additionally, unloading of the left atrium was achieved by implanting an 8 mm atrial flow regulator device through a hybrid per-atrial approach. Two months after the procedure, the patient was progressively weaned from the inotropes and transferred to the ward.

Heart failure is a major cause of mortality and morbidity worldwide.[1,2] Based on the assumption that ventricular–ventricular interaction may benefit patients with dilated left ventricular failure, Schranz et al.[3] proposed an innovative application of pulmonary artery banding as an effective treatment for end-stage heart failure in dilated cardiomyopathy in young children (<6 years of age) with preserved right ventricular function. A multicentre experience with pulmonary artery banding for dilated cardiomyopathy has been recently reported in selected paediatric patients,[4] with satisfactory results.

In patients with severe mitral valve regurgitation and/or severe left atrial enlargement, left atrial decompression improves atrioventricular interaction and global haemodynamic function, reducing left atrial pressure and pulmonary congestion.[5] Therefore, atrial fenestration might be helpful in a sub-group of patients undergoing pulmonary artery banding.[6] On the other hand, extracorporeal circulatory support is needed to create a surgical atrial fenestration, making weaning from extra-corporeal membrane oxygenator after surgery more challenging.

Since 2021, the Occlutech atrial flow regulator (AFR) device received the CE mark for the treatment of end-stage heart failure and severe pulmonary hypertension in adults. This device can create an interatrial communication after trans-septal puncture and balloon dilatation of the implantation site. Two sizes are available: 8 mm and 10 mm. The size is referred to the maximum diameter of the fenestration. The disks' diameter is 21 and 23 mm, respectively.

In this paper, we report a hybrid approach to create an atrial septal defect in a 6 kg, 6-month-old infant with end-stage heart failure undergoing pulmonary artery banding. Her medical history begun at 5 months of life, when she was admitted to the intensive care unit abroad for cardiogenic shock in dilated cardiomyopathy. She experienced two cardiac arrests which required resuscitation manoeuvers. Unfortunately, the patient was still depending on inotropes 4 weeks after her admission. Thus, she was referred to our institution to initiate the Giessen approach for end-stage heart failure and to consider potential heart transplantation.

At the admission to the intensive care unit, the echocardiography scan showed a severe left ventricular ejection fraction impairment (11%), severe mitral valve regurgitation, severe left atrial dilatation, mild impairment of right ventricular function, and mild tricuspid valve regurgitation. She experienced a third cardiac arrest few hours after the admission. After optimisation of the inotropic support, the left ventricular-ejection fraction increased to 23% and the left ventricular end-diastolic diameter to 41 mm (Z-score +6.5). dP/dT was 370 mmHg/s, while mitral valve regurgitation remained severe as well as left atrial dilatation. The right ventricular function improved to a normal value (TAPSE 13 mm, fractional area change 40%). Blood tests and cardiac MRI excluded myocarditis and congenital anomalies.

Given the normal right ventricular function, pulmonary artery banding was indicated for palliation of end-stage heart failure. Following the Giessen protocol, in patients with severe left atrial enlargement and mitral valve regurgitation, an interatrial communication was indicated to unload the left atrial and decrease left atrial pressure. To perform both procedures off-pump, we planned to implant an 8 mm Occlutech atrial flow regulator (Occlutech LTD, Helsinborg, Sweden) device to create an interatrial fenestration.

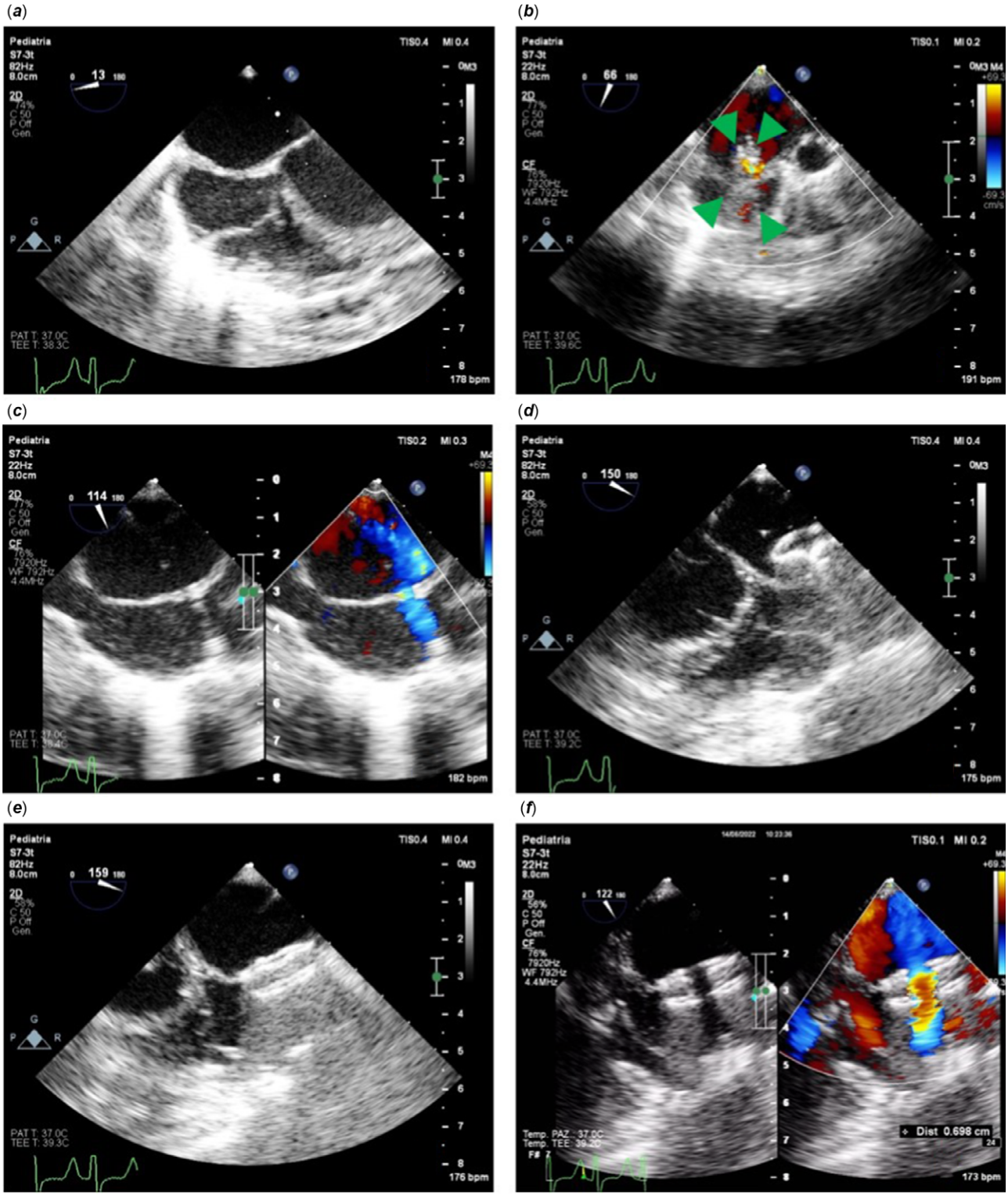

Figure 1. Hybrid approach for AFR implantation in 6-month-old infant. (***a***) Atrial septum from a transverse view. The total septal length was 30 mm. (***b***) The atrial septum was punctured with a 18 Ga, than stabilised a SV5 guidewire in the left atrium. On the guidewire, an 8x20 mm Sterling balloon was inflated (green arrows). (***c***) After the inflation, a 4 mm atrial septal defect was obtained. (***d***) An 11 Fr Terumo sheath was advanced in the atrial septal defect, and the left disk was deployed in the dilated left atrium. (***e***) By keeping a gentle traction on the system, the right disk was opened. Once verified the device stability, the device was deployed. (***f***) Color-Doppler demonstrated the left to right shunt. The diameter was 7 mm.

The patient underwent pulmonary artery banding through a midline sternotomy under continuous transesophageal echocardiography monitoring, as described elsewhere.[3] The atrial flow regulator device was implanted first (Fig 1). We performed a purse-string suture on the right atrial appendage, then we used a 20 Ga needle to puncture the interatrial septum under

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transesophageal echocardiography guidance. Once passed in the left atrium, a 0.018" SV5 guidewire (Cordis, Miami, FL) was placed in the left atrial, and the septum was pre-dilated with an over the wire 8 mm Sterling balloon (Boston Scientific, Marlborough, MA) (Fig 1b-c). On the same guidewire, an 11 Fr Terumo Sheath (Terumo LTD, Tokyo, Japan) was advanced into the left atrium. Finally, the 8 mm atrial flow regulator device was successfully implanted and eventually released under transesophageal echocardiography guidance (Fig 1d-e). The effective atrial septal defect diameter was 6 × 7 mm (Fig 1f). The mean echographic gradient was 4 mmHg. Once the device effectiveness and stability were verified, we proceeded with pulmonary artery banding in order to obtain a right ventricle pressure equal to 60% of systemic blood pressure. Due to acute bi-ventricular dysfunction, the patient was supported by extra-corporeal membrane oxygenator for the following 48 h. The chest was left open for 5 days. After extra-corporeal membrane oxygenator removal, the pulmonary artery banding was tightened to increase right ventricular pressure and to maintain the right ventricular/left ventricular pressure ratio after the improvement of biventricular function. Myocardial biopsy performed during the surgical procedure confirmed the diagnosis of idiopathic dilated cardiomyopathy.

The patient was progressively weaned from inotropes and ventilation. She tolerated oral therapy for chronic heart failure (Captopril 2 mg × 3/day, Bisoprolol 0.9 mg twice/day, Furosemide 6 mg × 3/day) and started feeding by mouth.

Echocardiography performed two months after the procedure showed an increase in left ventricular-ejection fraction up to 35%, dP/dT 630 mmHg/s, moderate mitral valve regurgitation, left atrial unloading, moderate dilatation of the right ventricular with a preserved TAPSE (11.5 mm), and mild tricuspid valve regurgitation. Pulmonary artery banding peak gradient was 30 mmHg.

The management of end-stage heart failure in infants and children is challenging. The lack of donors and the unavailability of intracorporeal ventricular assist devices force patients and caregiver to very long hospital stays.[6] Despite optimal medical care, morbidity and mortality rates in infants with end-stage heart failure remain high.[7] Infants with end-stage heart failure (who cannot be stabilised with medical therapy) can be effectively supported by pulmonary artery banding.[4] In some cases, left atrial unloading may be indicated to improve symptoms, optimise medical therapy, and to reduce the left ventricular filling pressures.[5] A right heart catheterisation is usually indicated to achieve data on left atrial pressure, cardiac index, pulmonary vascular resistances, and to perform an endomyocardial biopsy. In very high-risk settings, once confirmed the diagnosis and decided the therapeutic plan, some data might be obtained during surgery. In our case, the myocarditis was excluded by blood tests and by a cardiac magnetic resonance.

Recently, the atrial flow regulator device became available for treatment of end-stage heart failure in adults.[8–9] However, the large sheath needed (12–14 Fr) may impede its use in children weighting <15 kg. In these patients, a hybrid approach might be considered when an associated surgical manoeuver is planned to complete the procedure off-pump. In small infants, transatrial puncture ensures a safe access to the right atrium and allows a rapid atrial septum perforation despite severe left atrial enlargement causing bulging of the interatrial septum to the right. Atrial flow regulator implantation enables a calibrated, stable, and smooth interatrial septal communication compared to balloon atrial septostomy (isolated or associated to stent implantation). Furthermore, this device can be safely crossed for diagnostic or interventional purposes. Therefore, the atrial flow regulator implantation can also be considered in peculiar settings, when an atrial septal defect is needed for long time (like in cardiomyopathies, alternatively to stent implantation), or when future percutaneous procedures in the left atrial (or on the pulmonary veins) are planned after a first surgical approach (staged or hybrid approaches).

In conclusion, in very selected paediatric patients with end-stage heart failure, left ventricular unloading through atrial flow regulator implantation, and pulmonary artery banding could be performed safely with a single off-pump hybrid procedure and may lead to haemodynamic stabilisation of the patient. In small children, the atrial septum might be more compliant. Thus, a smaller balloon diameter (5–7 mm) might be considered to achieve a restrictive atrial septal defect, with final diameter <5 mm.

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Conflicts of interest. None.

Disclosures. BC is a proctor for Occlutech devices. The remaining authors have nothing to declare.

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