Interactive CardioVascular and Thoracic Surgery (2018) 1–4
doi:10.1093/icvts/ivy165

**BRIEF COMMUNICATION**



Cite this article as: Manuri L, Calaciura RE, De Zorzi A, Oreto L, Raponi M, Lehner A *et al*. Atrial flow regulator for failing Fontan circulation: an initial European experience. Interact CardioVasc Thorac Surg 2018; doi:10.1093/icvts/ivy165.

# Atrial flow regulator for failing Fontan circulation: an initial European experience

**Lucia Manuri[a,*], Rita Emanuela Calaciura[a], Andrea De Zorzi[a], Lilia Oreto[a], Massimiliano Raponi[b], Anja Lehner[c], Nikolaus Haas[c] and Salvatore Agati[a]**

[a] Mediterranean Pediatric Cardiology Center, Bambino Gesù Children's Hospital, Taormina, Messina, Italy
[b] Medical Direction, Bambino Gesù Children's Hospital, Rome, Italy
[c] Department of Pediatric Cardiology and Intensive Care, Ludwig Maximilians University Munich Campus, Munich, Germany

* Corresponding author. Mediterranean Pediatric Cardiology Center, Bambino Gesù Children's Hospital, Contrada Sirina-Taormina, Messina, Italy. Tel number +39-34-87225861, e-mail: lucia.manuri@opbg.net (L. Manuri).

Received 20 September 2017; received in revised form 28 March 2018; accepted 22 April 2018

## Abstract

Transcatheter creation or enlargement of an atrial septal defect has been used to promote adequate blood flow and mixing in some forms of congenital heart defects or as a relief valve in right or left atrial hypertension, resulting in better cardiac output and/or systemic saturation. We report a case of a 4-year-old male affected by complex congenital heart disease who was admitted for management of severe cyanosis following a staged pericardial fenestrated Fontan procedure. Transoesophageal echocardiogram showed a wide fenestration of 9 mm in size with a severely dilated pericardial Fontan system. To avoid a new surgical procedure and as part of a compassionate use programme, we decided to implant an atrial flow regulator device (4 mm in diameter) with percutaneous approach with the goal of reducing the right-to-left shunt and increasing the pulmonary flow. Preprocedural oxygen saturation was 75%, whereas after 2 months of follow-up, we observed a progressive increase of up to 95% with significant reduction in the pericardial Fontan system dimensions at echocardiography.

**Keywords:** Atrial flow regulator • Failing Fontan • Congenital heart defects • Hybrid procedure

## INTRODUCTION

The Fontan operation has improved survival of a generation of children born with congenital heart disease, resulting in a functional single ventricle physiology; however, it cannot recreate a physiological circulation. Fontan patients have been known to experience postoperative systemic venous hypertension, which in turn is associated with pleural effusions and protein-losing enteropathy, leading to a decreased duration and quality of life.

Despite the ongoing debate on its benefits, a circular fenestration hole (typically 4 mm) establishing a right-to-left shunt from the Fontan system to the common atrium is traditionally employed to relieve venous pressure in the Fontan conduit and improve early postoperative haemodynamics.

However, the haemodynamic improvements are obtained at the cost of reduced oxygen saturation due to right-to-left shunting, which may be excessive if the fenestration is permanent or even increases in size. The ideal fenestration would, therefore, limit shunt flow at tolerable systemic venous pressures, while allowing increased flow when pressures rises [1].

We present the first European experience of a child in whom an atrial flow regulator (AFR) device was implanted to reduce the shunt size and the right-to-left shunt, thereby leading to improved oxygenation. The boy was affected by tricuspid atresia, hypoplastic right ventricle, ventricular-arterial discordance and hypoplastic aortic arch. He had previously undergone a pericardial Fontan operation with an initial fenestration of 5 mm; during follow-up, severe desaturation developed due to enlargement of the fenestration. We decided to modulate the flow by percutaneous implantation of the AFR device, as part of a compassionate use progamme.

## CASE REPORT

A 4-year-old child with tricuspid atresia, hypoplastic right ventricle, ventricular-arterial discordance and hypoplastic aortic arch, who had previously undergone a pericardial fenestrated Fontan operation, was admitted for severe cyanosis.

The surgical history included hybrid palliation [2] at 30 days of life and comprehensive I–II stages [3] at 7 months. The postoperative period was complicated by acute Glenn thrombosis, and it was successfully treated using intravenous urokinase at the initial dosage of 4400 international units/kg IV over 20 min followed by a maintenance dosage (4400 international units/kg/h IV for 12 h); in addition, oral anticoagulation was initiated.

At the age of 3 years and 5 months, he underwent a pre-Fontan cardiac catheterization showing good morphology of the Damus–Kaye–Stansel anastomosis with venovenous collaterals directed

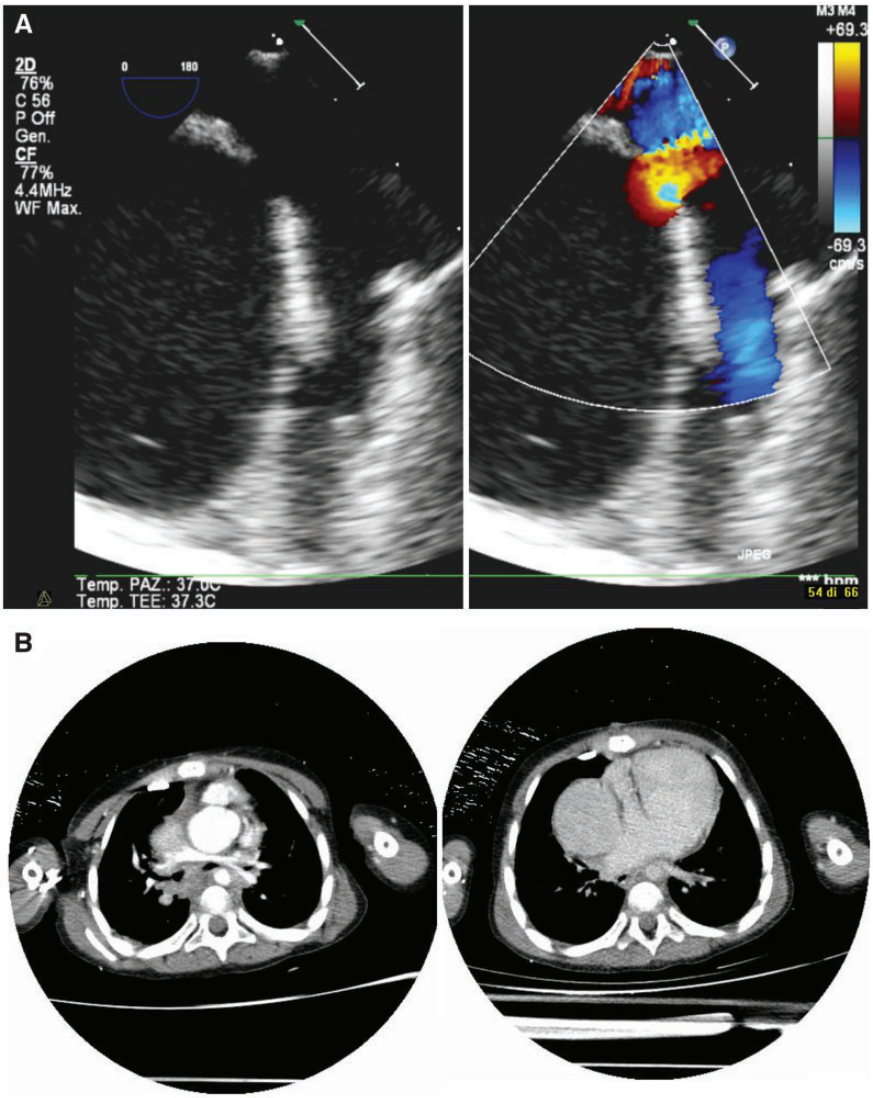


**Figure 1:** (**A**) Transoesophageal echocardiography showing a dilated fenestration. (**B**) Computed tomography showing the right pulmonary artery and dilation of the extracardiac Fontan circuit.

towards the inferior vena cava. Haemodynamic data were as follows: right atrial pressure 7 mmHg, left atrial pressure 6 mmHg, Glenn circuit pressure 13 mmHg, cavopulmonary anastomosis oxygen saturation 68%, systemic oxygen saturation 88%, with a cardiac index of 3 l/m/m$^2$, a pulmonary index of 1.8 l/m/m$^2$ and a calculated *Qp*/*Qs* ratio of 0.62. Pulmonary vascular resistance was 3 WU/m$^2$ with a predicted pressure in the Fontan system of 16 mmHg.

The patient subsequently underwent a fenestrated pericardial Fontan operation. The postoperative period was characterized by long-term haemodynamic instability and intensive care support due to pericardial effusion and partial seizures. Infantile neuropsychiatric consultation was performed, and medical therapy with levetiracetam was started; the treatment was completely withdrawn during follow-up.

Six months after Fontan completion, the patient developed severe desaturation; the mean pressure in the Fontan system at catheterization was 13 mmHg, and severe expansion of the Fontan circuit and right pulmonary artery stenosis were also documented.

Transoesophageal echocardiography (TOE) showed dilated fenestration, measuring 6–8 mm in size with evidence of severe dilatation of the Fontan circuit (Fig. 1A). Computed tomography was performed to define the morphologies of the right pulmonary artery and the extracardiac Fontan circuit (Fig. 1B).

After multidisciplinary discussion, the surgical risk was deemed very high, and therefore, it was decided to modulate the fenestration through the implantation of a 4-mm AFR device (Occlutech®, Sweden), aiming at increasing pulmonary blood flow by reducing the amount of right-to-left shunt between the Fontan circuit and RA.

The AFR device is currently under European Community registration for compassionate use in patients with severe right heart failure due to pulmonary hypertension or left heart failure and under the United States Food and Drug Administration's (FDA) emergency use guidance.

Our patient's family was provided with detailed information about risks and benefits of the procedure, including complications of general anaesthesia, TOE, cardiac catheterization and the device itself.

As for the conventional technique with intact interatrial communication, a trans-septal puncture is followed by static balloon

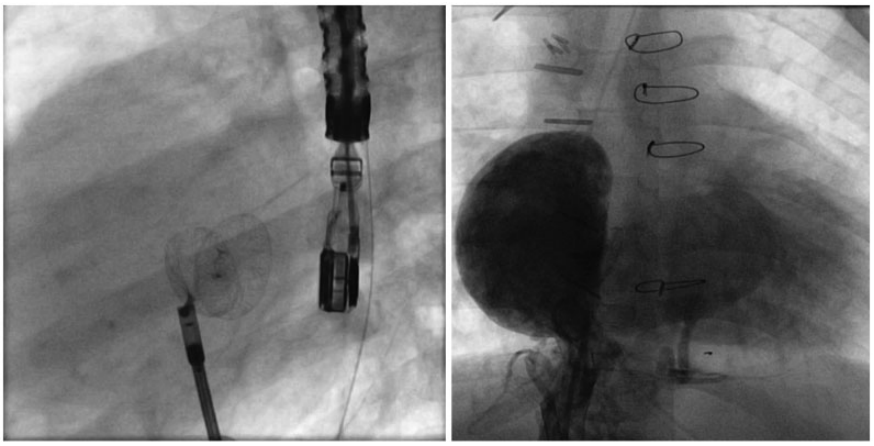

**Figure 2:** Technique of stent deployment (with kind permission from Prof. Dr. Nikolaus A. Haas, Munich).

dilatation. Thereafter, the delivery sheath is introduced, and the device is implanted in a similar fashion to a conventional double-disk ASD closure device. The measurement of the enlarged fenestration was performed by inflating a sizing balloon through it until the shunt disappeared, which revealed a diameter of 9 mm. After balloon occlusion, systemic oxygen saturation increased to 95%, whereas pressure in the Fontan circuit increased to 18–20 mmHg, such that complete closure was not an option. Therefore, a 9-Fr delivery system was advanced to the atrium. Under fluoroscopic (Fig. 2) and TOE guidance, the left-sided disc was first deployed on the atrial aspect of the fenestration, then the assembly was carefully retracted leaning on the Fontan tunnel wall, and finally, the right-sided disc was deployed.

Under fluoroscopic and TOE control, the position and stability of the device were verified, and a careful wiggle manoeuvre was performed, with subsequent successful release.

Oxygen saturation immediately increased from 73 to 80% with a mild increase in Fontan circuit pressure from 13 to 15 mmHg. The procedure was uneventful, and the patient was discharged after 3 days.

In addition, the patient was already treated using a Type-5 phosphodiesterase inhibitor, which was increased to a dosage of 2 mg/kg/dose 4 times a day.

At the 2-month follow-up, there has been progressive increase in peripheral oxygen saturation of up to 95%, and echocardiography showed significantly reduced dimensions of the Fontan circuit. Until now, 4 months after the intervention, a continuous right-to-left shunt is detectable, and there is no sign of haemolysis.

## DISCUSSION

A hybrid approach is the treatment of choice in our centre for neonatal patients who are candidates for Norwood palliation. In our experience, the hybrid approach and eliminating the risks for early open heart surgery and exposure to cardiopulmonary bypass, if associated with systemic vasodilator therapy and dedicated feeding protocol adherence, could reduce the incidence of gastrointestinal complications, which is significantly greater in neonates with CHD than in the normal newborn population and is even higher among patients with a single-ventricle physiology, particularly those with hypoplastic left heart syndrome [3].

During the past 5 years, we have adopted a technique of Fontan completion without a conduit, which involves the creation of a tunnel whose borders are the right atrial wall and inferior vena cava medially and inferiorly, the native pericardium and lung posteriorly and laterally, right pulmonary artery superiorly and heterologous pericardial patch anteriorly [2]. The main purpose of such a system would be that the expandable pericardium could, in part, compensate acute systemic venous hypertension and promote flow through the lung, enhanced by the right lung excursion and atrial contraction. In our series, the patient we report is the only one who received fenestration due to poor pulmonary vasculature, which can explain the progressive enlargement of the fenestration at follow-up.

Palliative transcatheter AFR implantation has been already described for the treatment of patients with symptomatic pulmonary artery hypertension to establish an adequate atrial communication, as an alternative approach to increase systemic ventricular output [4]. In some centres, AFR has also been applied to different pathologies, such as right atrial congestion [5], right heart failure, left atrial congestion and failing Fontan, all as part of a compassionate use programme. The AFR device facilitates controlled blood flow that allows the distended atrium to be decompressed without an uncontrolled and sudden change of *Qp*/*Qs*. AFR is manufactured in different sizes 4-, 6-, 8- and 10-mm diameter of the hole and different heights. The implantation technique is comparable to the implantation of the commonly used double-disc atrial septal defect devices from the same company, and thereby, is a standardized and safe procedure [6].

## CONCLUSIONS

Palliative transcatheter AFR implantation has been successfully used to relieve right or left atrial hypertension in selected groups of patients.

Furthermore, AFR implantation may represent an alternative approach in patients with failing Fontan circulation, providing increased pulmonary flow and better oxygen saturation with acceptable Fontan pressure, thanks to the decompressive effect of a fenestration.

This is, to our knowledge, the first example of an AFR placement in a young patient aged 4 years. Although the AFR device was originally developed to establish a connection between the left and right atrium, in a previously intact septum, it may be well suitable to reduce the size of an enlarged fenestration. We observed an immediate improvement in haemodynamics and symptoms following the procedure, with no complications. However, further studies will be required to evaluate efficacy and long-term outcomes of AFR in this specific subgroup of patients.

## ACKNOWLEDGMENTS

The authors thank specially and sincerely Dott. Michele Saitta for his hard work in the paediatric cathlab with very complex congenital cases and Dott. Maria Giulia Gagliardi who helped and supported the centre to become a referral department for interventional paediatric cardiology in the South of Italy.

**Conflict of interest:** none declared.

## REFERENCES

[1] Albal PG, Menon PG, Kowalski W, Undar A, Turkoz R, Pekkan K. Novel fenestration designs for controlled venous flow shunting in failing Fontans with systemic venous hypertension. Artif Organs 2013;37:66–75.

[2] Gundry SR, Razzouk AJ, del Rio MJ, Shirali G, Bailey LL. The optimal Fontan connection: a growing extracardiac lateral tunnel with pedicled pericardium. J Thoracic Cardiovasc Surg 1997;114:552–8; discussion 558–9.

[3] Manuri L, Morelli S, Agati S, Saitta MB, Oreto L, Mandraffino G *et al*. Early hybrid approach and enteral feeding algorithm could reduce the incidence of necrotising enterocolitis in neonates with ductus-dependent systemic circulation. Cardiol Young 2017;27:154–60.

[4] Patel MB, Samuel B, Girgis RE, Parlmer MA, Vettukattil JJ. Implantable atrial flow regulator for severe, irreversible pulmonary arterial hypertension EuroIntervention 2015;11:706–9.

[5] Haas NA, Laser KT, Bach S, Kantzis M, Happel CM, Fischer M. Decompressive atrioseptostomy (DAS) for the treatment of severe pulmonary hypertension secondary to restrictive cardiomyopathy. Int J Cardiol 2016;203:845–7.

[6] Haas NA, Soetemann DB, Ates I, Baspinar O, Ditkivskyy I, Duke C *et al*. Closure of secundum atrial septal defects by using the occlutech occluder devices in more than 1300 patients: the IRFACODE project: a retrospective case series. Catheter Cardiovasc Interv 2016;88:571–81.