JACC: CASE REPORTS VOL. 16, 2023

IMAGING VIGNETTE

ADVANCED

CLINICAL VIGNETTE

Staged Acclimatization in a Failing Fontan by AFR in AFR

Ever-Decreasing Circles

Jenny E. Zablah, MD, Natalie Soszyn, MBBS, BMedSci, MMed, Gareth J. Morgan, MB, BAO BCh, MPhil

ABSTRACT

We describe serial implantation of atrial flow regulator (AFR) devices in the Fontan fenestration of a 4-year-old patient. Initially, the fenestration size was decreased using a 6/5 AFR, resulting in improved saturations and hemodynamics. One year later, further improvement was achieved by placing a 4/10 AFR inside the original device. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;16:101868) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 2-year-old boy weighing 15.1 kg with hypoplastic left heart syndrome demonstrated a low cardiac output state coupled with increasing oxygen saturations secondary to acute fenestration occlusion confirmed by transthoracic echocardiography 12 hours following an uncomplicated Fontan completion. He had favorable hemodynamics prior to Fontan completion (**Figure 1A**).

The patient was anticoagulated, ventilated, and then transferred to the catheterization lab. Transesophageal echocardiography and angiography confirmed total occlusion of the fenestration. Fenestration recanalization was achieved by serial dilation with increasing high-pressure balloons. He was treated aggressively with warfarin and discharged home.

He experienced significant desaturation (78%) over the subsequent months. A 4-dimensional flow cardiac magnetic resonance demonstrated that 30% of his cardiac output passed through his fenestration (**Figure 1B**). He was brought to the catheterization laboratory 6 months following his Fontan procedure for elective fenestration reduction with an atrial flow restrictor (AFR) device. A 6-mm fenestration diameter/5-mm waist length AFR was selected, as it would restrict the right-to-left flow while allowing adequate Fontan decompression and augmentation of cardiac output as physiologically required. Under general anesthetic and using transesophageal echocardiography, fluoroscopic, and angiographic guidance, the device was implanted using a previously described technique and achieved immediate improvement in saturations (from 78% to 86%) (**Figure 1A**).[1]

The patient returned 17 months later due to persistent desaturation. Complete balloon fenestration occlusion resulted in improvement in systemic saturations (from 75% to 89%) but an unacceptable 33% reduction in cardiac output (from 3.86 L/min/m^2 to 2.57 L/min/m^2) (**Figure 1A**). Bench testing determined that placing a

From the Division of Cardiology, Department of Pediatrics, Heart Institute, Children's Hospital Colorado, University of Colorado Denver Anschutz Medical Campus, Aurora, Colorado, USA.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received January 31, 2023; revised manuscript received March 6, 2023, accepted April 11, 2023.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2023.101868

ABBREVIATIONS AND ACRONYMS

AFR = atrial flow regulator

4-mm fenestration diameter/5-mm waist length AFR within the previously implanted device would create a significant decrease in flow with only a small decrease in the effective diameter (Figure 1C).

A 4/5 AFR was successfully positioned within the previous device. Multimodality imaging demonstrated that in vivo the device was more compressed compared with the benchtop model. This device was therefore removed and replaced with a 4-mm fenestration diameter/10-mm waist length AFR. Repeat imaging demonstrated this device had more favorable disc apposition, waist expansion and achieved more satisfactory hemodynamics (Figures 1D and 1E).

FOLLOW-UP

One-year following this last intervention, his saturations were 92% to 95% on room air with a patent fenestration on echocardiography.

DISCUSSION AND CONCLUSIONS

The AFR is a centrally fenestrated, double-disc device designed to maintain the patency of a newly created or already existent atrial communication allowing a variable level of pressure and volume offloading.[2] It has been used by our group and others to achieve a predictable, low-profile Fontan fenestration in preference to stent implantation.[2,3]

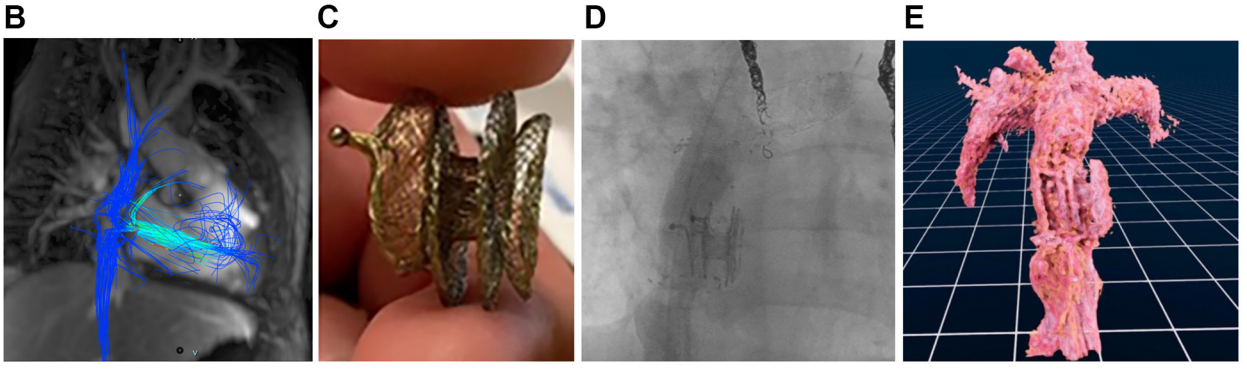
Serial reduction of fenestration size is feasible with the AFR-in-AFR technique and represents a good option for patients with failing Fontan physiology that require gradual matriculation from Glenn to Fontan physiology. Bench testing is required to determine appropriate device sizes but may not reflect in vivo performance. Hence, a range of device sizes should be available for these procedures.

FIGURE 1 Placement of an AFR-in-AFR in the Fenestration of a Failing Fontan

A Hemodynamic data

	Pre-Fontan	First study (1 day after Fontan)		Second study (6 months after Fontan)		Third study (10 months after Fontan)		Fourth study (27 months after Fontan)		
	Baseline	Baseline	Post-fenestration angioplasty	Baseline	Post-1st AFR placement	Baseline	Balloon occlusion of fenestration	Baseline	Balloon occlusion of fenestration	Post-2nd AFR placement
Pressures (mmHg)										
Glenn/Fontan	10	15	17	11	13	11	13	7	10	11
RFA	62/33 (46)	72/33 (43)	85/42 (55)	68/41 (52)	76/44/(57)	78/42 (56)	73/42 (54)	71/33 (49)	63/39 (48)	76/31 (48)
Saturations (%)										
Glenn/Fontan	55%	-	40%	61%	-	68%	74%	59%	64%	69%
RFA	80%	95%	87%	79%	90%	78%	91%	75%	89%	88%
TPG	7	10	10	4	-	7	7	6	6	6
Cardiac index (L/min/m²)	2.96	-	1.36	4.11	-	4.81	3.74 *(↓23%)*	3.86	2.57 *(↓33%)*	3.66
PVRi (Wood units/m²)	0.79	-	-	2.6	-	2.91	2.67	2.16	3.37	-
Qp:Qs	0.64	-	0.84	0.56	-	0.5	1	0.72	1.04	-

RFA, right femoral artery; TPG, trans-pulmonary gradient; PVRi, pulmonary vascular resistance index; Qp, pulmonary flow; Qs, systemic flow



(A) Hemodynamic data from cardiac catheterization prior to Fontan and at 1 day, 6 months, 10 months, and 27 months following Fontan. **(B)** Four-dimensional flow cardiac magnetic resonance demonstrating 30% of cardiac output passing through the fenestration. **(C)** Bench testing demonstrating the diameter of the fenestration of a 4/5 atrial flow regulator (AFR) device placed within a 6/5 AFR device. **(D)** A 3-dimesional rotational angiography (3DRA) performed following placement of an AFR in AFR. **(E)** A 3-dimensional model was created using virtual reality in which the 2 discs of the devices in the Fontan side are seen. Q_p = pulmonary flow; Q_s = systemic flow; RFA = right femoral artery; TPG = transpulmonary gradient.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

Drs Zablah and Morgan have served as consultants for Occlutech. Dr Soszyn has reported that she has no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Jenny Zablah, Children's Hospital Colorado, 13123 East 16th Avenue, Box 100, Aurora, Colorado 80045, USA. E-mail: jenny.zablah@childrenscolorado.org. Twitter: @jennyzablah.

REFERENCES

1. O'Callaghan B, Zablah JE, Ivy DD, Morgan GJ. Technical description, deployment technique and potential applications of the Occlutech atrial flow regulator device. *Interv Cardiol*. 2021;13(suppl 2):17-22.

2. Manuri L, Calaciura RE, De Zorzi A, et al. Atrial flow regulator for failing Fontan circulation: an initial European experience. *Interact Cardiovasc Thorac Surg*. 2018;27(5):761-764.

3. Zablah JE, Morgan GJ. Use of Occlutech® atrial flow regulator in a single ventricle patient: a 3D view of a successful intervention. *Eur Heart J Cardiovasc Imaging*. 2020;21(10):1151.

KEY WORDS AFR, failing Fontan