JACC: CASE REPORTS VOL. 4, NO. 15, 2022

MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

ADVANCED

CASE REPORT: CLINICAL CASE

Device Occlusion of Native Pulmonary Blood Flow After Cavopulmonary Anastomosis With Persistent Pleural Effusions

Sophia Khan, MBChB, MD,[a] Abdulla Tarmahomed, MBChB,[a] Salim Jivanji, MBChB[a,b]

ABSTRACT

Native pulmonary tract flow after a cavopulmonary anastomosis may promote pulmonary artery growth but can lead to undesirable consequences. We report the case of a 17-month child with prolonged pleural effusions after cavopulmonary anastomosis in whom a ventricular septal defect occluder device was placed in the native right ventricular outflow tract. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:924-928)

A 17-month-old boy (weight 11 kg), with antenatally diagnosed congenitally corrected transposition of the great arteries, a noncommitted ventricular septal defect (VSD), and a dysplastic pulmonary valve with pulmonary valve stenosis presented with persistent pleural drainage more than 3 months after superior cavopulmonary anastomosis surgery.

LEARNING OBJECTIVES

- To understand potential consequences (both beneficial and adverse) of having a dual pulmonary blood supply in patients with cavopulmonary anastomosis.
- To review the rationale and decision making, including hemodynamic assessment, for occlusion of a native pulmonary blood flow in patients with cavopulmonary anastomosis.
- To consider the potential for hemolysis when there is residual flow through an occluder device.

MEDICAL HISTORY

At 6 weeks of age he was admitted because of cyanosis and underwent an emergency right modified Blalock-Taussig shunt. Twelve months later he underwent a superior cavopulmonary anastomosis, and the right modified Blalock-Taussig shunt was taken down, leaving the native pulmonary outflow tract open to augment pulmonary blood flow and promote growth of the pulmonary arteries (**Figure 1**). The cavopulmonary anastomisis was an intermediate step with a future plan to perform an atrial switch and a Rastelli procedure: in essence a "double switch."

After his cavopulmonary anastomosis, the initial postoperative course was uneventful, resulting in his being discharged home 7 days after surgery to receive diuretics. He subsequently experienced recurrent

From the [a]Alder Hey Children's Hospital, Liverpool, United Kingdom; and the [b]North West, North Wales, and Isle of Man Adult Congenital Heart Disease Network, Liverpool Heart and Chest Hospital NHS Foundation Trust, Liverpool, United Kingdom.
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 February 10, 2022; revised manuscript received April 10, 2022, accepted May 10, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.05.011

pleural effusions, requiring 2 readmissions to the hospital for chest drain insertion.

DIFFERENTIAL DIAGNOSIS

Persistent or recurrent pleural effusions after bidirectional cavopulmonary connection is a well-described phenomenon. Contributory factors to consider include raised capillary permeability secondary to infection, increased systemic venous pressures secondary to superior vena cava obstruction, and hypoalbuminemia leading to decreased oncotic pressure. Respiratory factors resulting in an increased negative intrathoracic pressure such as postoperative atelectasis or conditions that increase pulmonary artery pressure can also lead to persistent pleural drainage.[1]

In this case it was believed that the dual pulmonary blood supply was causing raised pulmonary artery pressures and contributing to his prolonged pleural drainage.

INVESTIGATIONS

Diagnostic cardiac catheterization demonstrated the mean pulmonary artery pressure to be 16 mm Hg (16/18 mm Hg), with the hemodynamic trace showing pulsatile flow. On test occlusion there was proven reduction of the pulmonary artery pressure to a mean of 10 mm Hg (11/9 mm Hg).

Angiography at the time demonstrated good right ventricular systolic function, with a large VSD, and subpulmonary and valvar pulmonary stenosis with poststenotic dilation of the main pulmonary artery (**Figure 2,** Video 1). The branch pulmonary arteries were confluent, and there was good arborization of both lungs, with normal unobstructed pulmonary venous return. There were no other contributory findings to explain the recurrent pleural effusions.

ABBREVIATIONS AND ACRONYMS

APBF = antegrade pulmonary blood flow

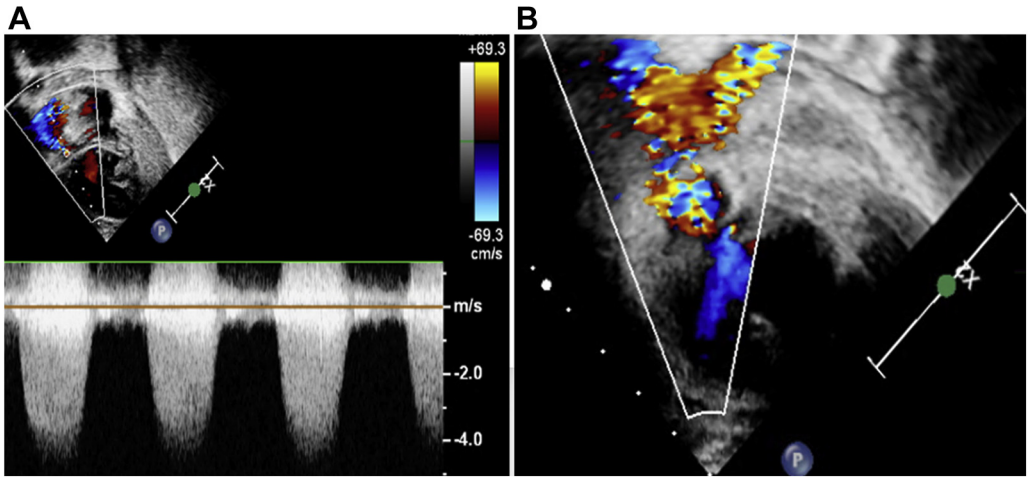
VSD = ventricular septal defect

MANAGEMENT

The right ventricular outflow tract was occluded with a 12/7-mm Amplatzer muscular VSD occluder (Abbott Cardiovascular). There was good conformity of the device and minimal flow through the device on angiography (**Figure 3,** Videos 2 and 3). The hemodynamics were similar to those described during the diagnostic catheterization, confirming a significant reduction in mean pulmonary artery pressures.

In view of the initial residual forward flow through the device, the patient was regularly followed up with echocardiography (**Figure 4,** Video 4) and monitored for signs of hemolysis.[2] He had preliminary evidence of hematuria on urinalysis and raised lactate dehydrogenase levels. This was managed conservatively and resolved within 3 weeks. The pleural effusions

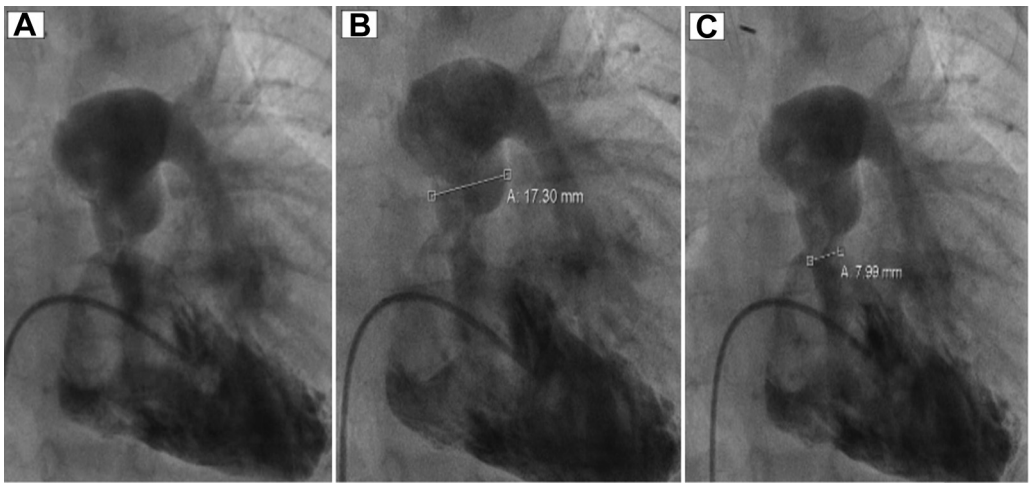
FIGURE 1 Baseline Echocardiography Images



Echocardiography images showing spectral and color Doppler of the forward flow arising from the morphologic left ventricle through the pulmonary valve. There is pulmonary stenosis at the valvular and subvalvular levels. The maximum velocity on the spectral Doppler trace is measured at 4 m/s.

JACC: CASE REPORTS, VOL. 4, NO. 15, 2022
AUGUST 3, 2022:924–928

FIGURE 2 Angiography Demonstrating Pulmonary Outflow Tract Anatomy



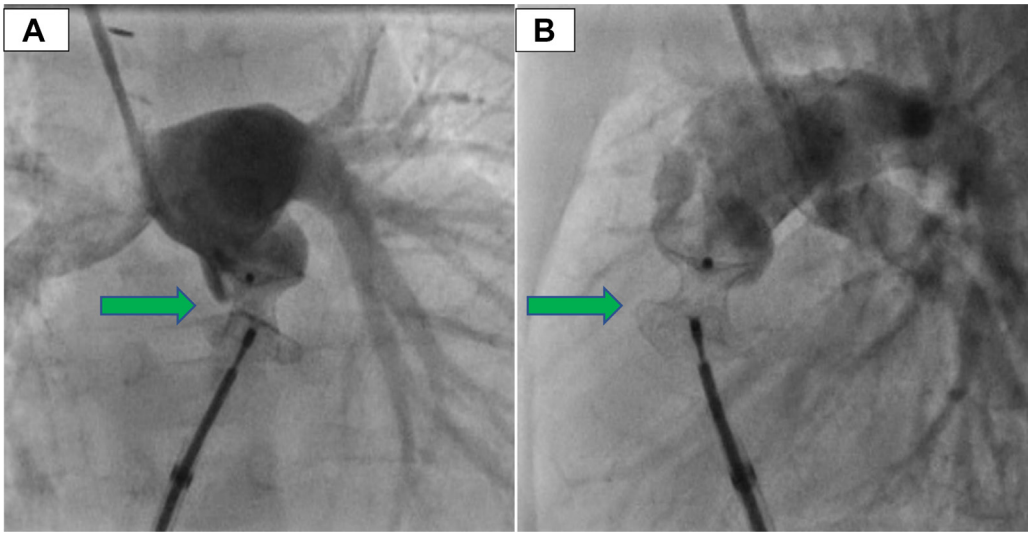
(A) Left ventricular angiography with measurements taken at **(B)** the widest and **(C)** the narrowest part of the outflow tract.

resolved after the right ventricular outflow tract occlusion.

DISCUSSION

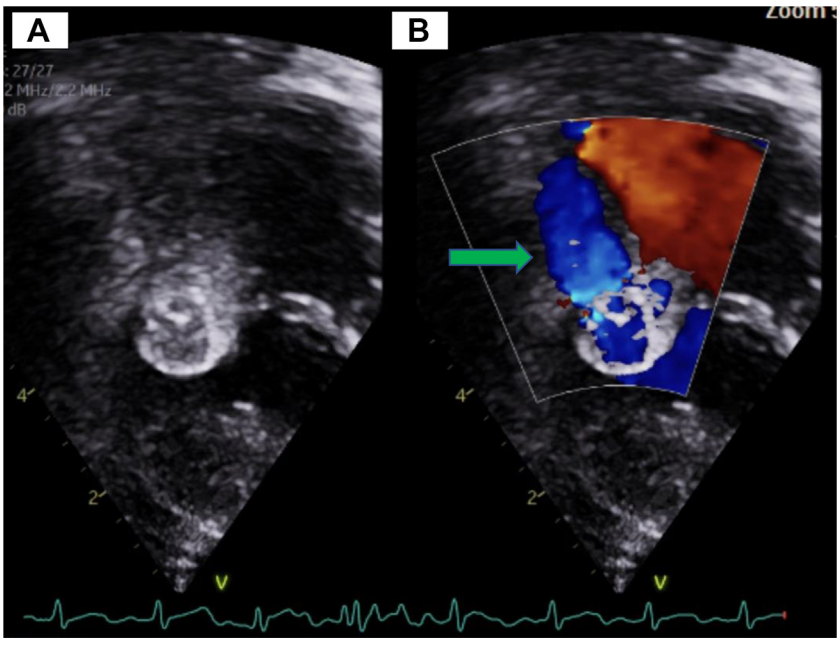
There is conflicting evidence regarding the effects of antegrade pulmonary blood flow (APBF) for patients undergoing cavopulmonary anastomosis. Some studies suggest that the additional pulsatile pulmonary blood flow pattern is beneficial for growth and development of the pulmonary arteries.[3,4] Others describe a detrimental effect of pulsatile APBF leading to excessively high pulmonary artery pressure and as such an increased risk

FIGURE 3 Angiograms Post Device Deployment



Anteroposterior **(A)** and straight lateral **(B)** projection confirming good device position **(green arrow)** with angiography in the main pulmonary artery demonstrating unobstructed flow to both branches.

FIGURE 4 Echo Post Device Occlusion of Pulmonary Outflow Tract



Echo demonstrating the **(A)** device in good position and **(B)** with color Doppler demonstrating residual flow **(green arrow)** across the device.

of superior vena cava syndrome and pleural effusions.[5,6] There are, however, further published studies that have returned ambiguous results with no clear advantages or adverse effects from APBF.[7]

An increase in central venous pressure leads to an increase in the capillary filtration of fluid and is a recognized risk factor for persistent pleural effusions.[8] In the case described, after occlusion of the APBF there was a significant reduction in mean pulmonary artery pressures, complete resolution of the patient's persistent pleural effusions, and no major adverse events.

This case is unique because of the young age and size of the child and the complete occlusion of APBF with a device. There are published reports of off-label use of muscular VSD occluder devices, but those are predominately in adults. Among those reported uses are occlusion of patent ductus arteriosus, closure of perforated sinus of Valsalva aneurysm, and closure of a pulmonary artery to left atrium fistula.[9]

CONCLUSIONS

This case demonstrates the role of catheter hemodynamic assessment and intervention in patients with persistent pleural drainage after cavopulmonary anastomosis who have native APBF. In our case we showed a demonstrable change in pulmonary pressures by occluding the APBF, a clear improvement in the patient's clinical condition, and swift resolution of his persistent pleural effusions. This case also highlights the importance of monitoring for hemolysis after device insertion.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr. Salim Jivanji, Department of Cardiology, Alder Hey Children's Hospital, East Prescot Road, Liverpool L12 2AP, United Kingdom. E-mail: salim.jivanji@alderhey.nhs.uk. Twitter: @salimjivanji.

JACC: CASE REPORTS, VOL. 4, NO. 15, 2022
AUGUST 3, 2022:924–928

REFERENCES

1. Talwar S, Agarwala S, Mittal CM, Choudhary SK, Airan B. Pleural effusions in children undergoing cardiac surgery. *Ann Pediatr Cardiol*. 2010;3(1):58–64.

2. Amoozgar H, Soltani R, Edraki M, et al. Hemolysis and its outcome following percutaneous closure of cardiac defects among children and adolescents: a prospective study. *Ital J Pediatr*. 2019;45(1):128.

3. Kowatari R, Suzuki Y, Daitoku K, Fukuda I. Long-term results of additional pulmonary blood flow with bidirectional cavopulmonary shunt. *J Cardiothorac Surg*. 2020;15(1):279.

4. Ferns SJ, El Zein C, Multani K, et al. Is additional pulsatile pulmonary blood flow beneficial to patients with bidirectional Glenn? *J Thorac Cardiovasc Surg*. 2013;145(2):451–454.

5. Frommelt MA, Frommelt PC, Berger S, et al. Does an additional source of pulmonary blood flow alter outcome after a bidirectional cavopulmonary shunt? *Circulation*. 1995;92(9):240–244.

6. Talwar S, Sandup T, Gupta S, et al. Factors determining early outcomes after the bidirectional superior cavopulmonary anastomosis. *Indian J Thorac Cardiovasc Surg*. 2018;34(4):457–467.

7. Berdat PA, Belli E, Lacour-Gayet F, Planché C, Serraf A. Additional pulmonary blood flow has no adverse effect on outcome after bidirectional cavopulmonary anastomosis. *Ann Thorac Surg*. 2005;79(1):29–37.

8. Bai L, Feng Z, Zhao J, et al. Risk factors and long-term prognosis for chylothorax after total cavopulmonary connection in children: a retrospective study from a single center. *Front Pediatr*. 2021;9:744019.

9. Bansal N, Aggarwal S, Turner DR. Closure of insufficient, native right ventricular outflow tract with AMPLATZER™ muscular ventricular septal defect occluder in a patient with tetralogy of Fallot post-Melody(®) valve. *Ann Pediatr Cardiol*. 2019;12(2):159–162.

KEY WORDS Cavopulmonary, pediatric, pleural effusions, right pulmonary outflow tract, ventricular septal defect occluder

APPENDIX For supplemental videos, please see the online version of this paper.