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Brief Report

Shunt reduction to pulmonary arteriovenous malformations using a modified covered stent

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Abstract Pulmonary arteriovenous malformations in patients with congenital heart disease have been associated with interruption of hepatic venous return to the lungs. We report a novel technique to improve systemic saturation using a modified covered stent in a patient with unilateral left pulmonary arteriovenous malformations in the setting of a Fontan circulation.

Keywords: Paediatric interventions; intrapulmonary shunt; haemodynamics; right-to-left shunt

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A VARIETY OF TRANSCATHETER INTERVENTIONS have been described to modify blood flow.[1–3] In patients with pulmonary arteriovenous malformations, eliminating the intrapulmonary shunt improves oxygenation.[4,5] We report a novel technique to treat an iatrogenic increase in right-to-left shunting by restricting flow to an affected lung using a modified covered stent. The patient had undergone extracardiac Fontan repair and developed diffuse unilateral pulmonary arteriovenous malformations.

Case report

A 12-year-old male, Jehovah's Witness, was admitted to the hospital with fatigue that worsened with exercise and hypoxaemia (oxygen saturation 92%). The patient was diagnosed with hypoplastic left heart syndrome at birth and underwent Stage 1 palliation at 2 days of life, followed by a bi-directional cavopulmonary anastomosis at 10 months of age and a non-fenestrated extracardiac Fontan at 2 years of age. Cardiac catheterisation revealed a severe long-segment stenosis of the left pulmonary artery (Fig 1a). This finding was assumed to be the cause of the patient's symptoms because of ventilation–perfusion mismatch and resultant dead space ventilation. To improve left lung perfusion, a stent was placed, increasing the diameter from 4.5 to 16 millimetres and resulting in increased flow to the left lung. There was no change in oxygen saturation acutely. Pressure within the left pulmonary artery improved to 16 millimetres of mercury and the patient was discharged home without complications.

At the 6-month follow-up, the patient reported poor exercise tolerance and was found to be cyanotic with oxygen saturations of 70–75%. Re-evaluation of the initial angiograms resulted in the diagnosis of pulmonary arteriovenous malformations in the left lung, which were not appreciated during the first catheterisation. The pulmonary arteriovenous malformations may have increased in size as a consequence of the increase in blood flow to the left lung, resulting in further decrease of oxygen saturation. The patient was observed over the following months without improvement in symptoms.

Therapeutic interventions were carefully discussed with the parents who were reluctant to consent to a surgical procedure with the possibility of blood transfusion. A transcatheter intervention

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was performed 12 months after initial stent placement. Catheterisation demonstrated diffuse left pulmonary arteriovenous malformations (Fig 1b). Temporary balloon occlusion of the left pulmonary artery resulted in an acute increase in oxygen saturation without significant haemodynamic alteration in the Fontan circulation. Owing to their large size, two pulmonary arteriovenous malformations were coil embolised, with no resultant change in oxygen saturation. The plan was then to re-restrict flow to the left pulmonary artery. A covered stent (iCAST™, Atrium Medical Corporation, Hudson, New Hampshire, United States of America) 10 millimetre/38 millimetre was modified as follows: the stent was partially inflated to free it from its delivery system. It was inflated to 4 millimetres over a standard 4 millimetre × 20 millimetre balloon (Johnson & Johnson, OPTA Pro, Cordis Corporation, Miami, Florida, United States of America). At the mid-portion of the stent, a 3.0 absorbable vicryl suture was secured to one of the struts, thus puncturing the covering, and then wrapped circumferentially. The integrity of the suture, limiting central expansion to 4 millimetres, was tested by expanding the stent with its original 10-millimetre balloon (Fig 2a). The stent was then hand-crimped over a 16 millimetre/40 millimetre Z-Med™ angioplasty balloon (NuMED Inc. Hopkinton, New York, United States of America). It was implanted into the left pulmonary artery, using a 12 French long sheath for deployment and the balloon was inflated to 11 atmospheres (Fig 2b). The narrowest diameter of the bow-tie-shaped stent measured 4 millimetres and longitudinally it shortened considerably. There was an immediate improvement in oxygen saturation from 80% to 93%, with predominant flow to the right lung. The patient tolerated the procedure well and he was discharged home the following day.

Discussion

We describe a patient who developed unilateral pulmonary arteriovenous malformations despite having achieved complete repair within 2 years of life. The long-segment stenosis of the left pulmonary artery observed at the first catheterisation was assumed to be the cause of this patient's symptoms and therefore it was stented. Pulmonary artery stenting resulted in an iatrogenic increase of intrapulmonary right-to-left shunting and worsening of the patient's symptoms. Upon discovering the arteriovenous malformations, an interventional procedure was developed with the intent to decrease the intrapulmonary right-to-left shunting and preserve lung tissue.

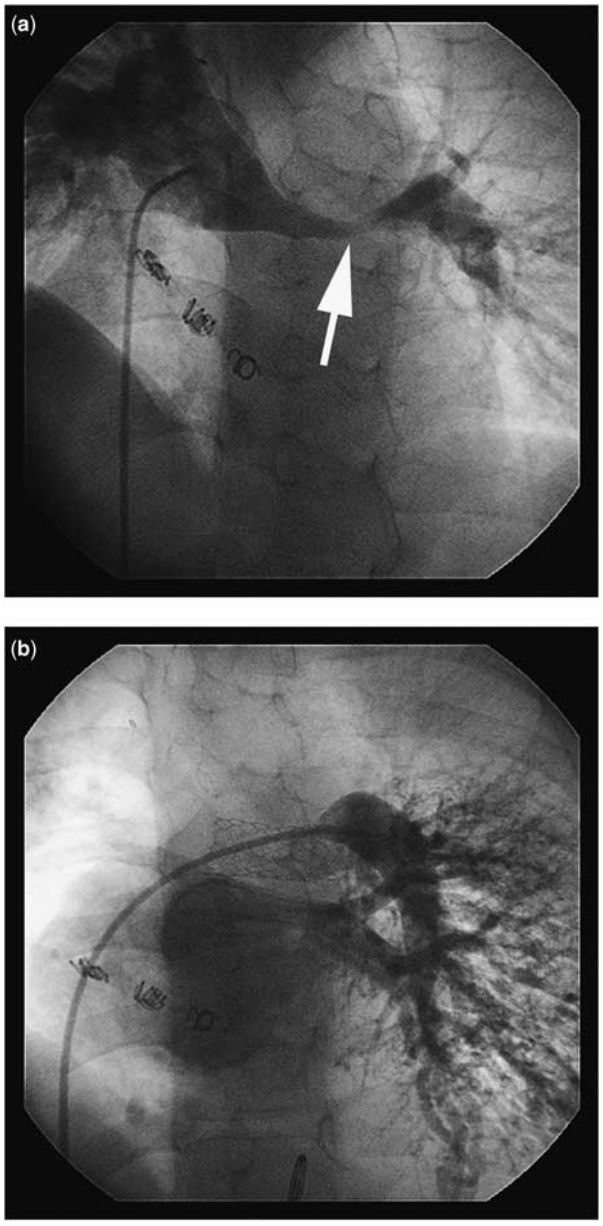

Figure 1.
*(**a**) White arrow indicates the long-segment stenosis of the left pulmonary artery. (**b**) Simultaneous arterial and venous phases indicate a diffuse process of pulmonary arteriovenous malformations in the left lung.*

To the best of our knowledge, this is the first report using a self-modified covered stent in a pulmonary artery to reduce intrapulmonary shunting. The use of uncovered "bow tie" stents to create interatrial communications has been previously reported.[2] The principles of this concept were applied to modify a covered stent by fashioning a vicryl suture around the mid-portion of the stent in order to preserve a waistline after deployment. The maximum diameter of the modified stent was 10 millimetres and, although it is recognised

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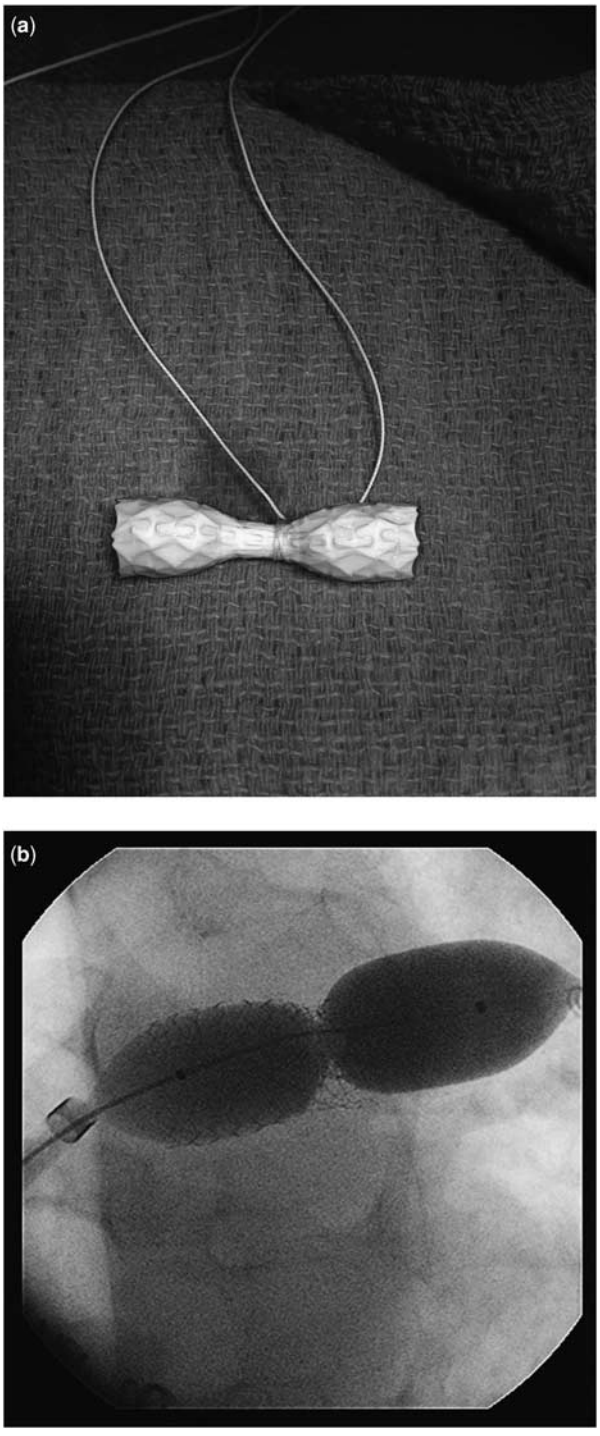

Figure 2.
*(**a**) Covered stent preparation. The stent has been modified with a suture to produce a central restriction upon full balloon inflation (see text). (**b**) Stent implantation demonstrating central restriction despite balloon expansion.*

that the covering around the stent may tear with overdilation, a 16-millimetre balloon was chosen for deployment because that was the diameter of the stent already in place in the left pulmonary artery. Stent implantation resulted in a rapid decrease in the intrapulmonary shunt and subsequent increase in the oxygen saturation. An absorbable vicryl suture was used knowing that it will be reabsorbed, thus allowing for future balloon dilation to increase the diameter of the stent if the pulmonary arteriovenous malformations regress over time. It is recognised that overdilation of a covered stent will result in tearing of the cover, particularly if it has been punctured by the suture's needle; however, in this patient the aim was to restrict flow, and hence the integrity of the cover was not as important clinically.

In patients requiring a bi-directional cavopulmonary anastomosis as part of staged repair,[4,6] deficiency of a hepatic factor has been proposed as the cause for developing pulmonary arteriovenous malformations. Our patient had a long-standing period with impaired flow to the left lung possibly resulting in decreased exposure to the hepatic venous flow and subsequent development of arteriovenous malformations.

Currently, the patient is being followed up as an outpatient and is stable with oxygen saturations above 85%. The treatment remains expectant as long-term exposure of lung tissue to hepatic venous flow may eventually cause regression of the arteriovenous malformations.[7–10] The option to gradually increase blood flow to the left lung by serial dilation of the stent allows time for regression of the arteriovenous malformations. Recognising the limitations of a single case report with short-term follow-up, we believe that this strategy may be suitable for the treatment of other vascular anomalies where flow restriction is required.

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References

1. Nazarian GK, Qian Z, Vlodaver Z, et al. Evaluation of a new vascular occlusion device. Invest Radiol 1993; 28: 1165–1169.
2. Pedra CA, Pihkala J, Benson LN, Freedom RM, Nykanen D. Stent implantation to create interatrial communications in patients with complex congenital heart disease. Catheter Cardiovasc Interv 1999; 47: 310–313.
3. Kretschmar O, Sglimbea A, Corti R, Knirsch W. Shunt reduction with a fenestrated Amplatzer device. Catheter Cardiovasc Interv 2010; 76: 564–571.
4. Van Den Bogaert-Van Heesvelde AM, Derom F, Kunnen M, Van Egmond H, Devlon-Blancquert A. Surgery for arteriovenous fistulas and dilated vessels in the right lung after the Glenn procedure. J Thorac Cardiovasc Surg 1978; 76: 195–197.

5. Farra H, Balzer DT. Transcatheter occlusion of a large pulmonary arteriovenous malformation using the Amplatzer vascular plug. Pediatr Cardiol 2005; 26: 683–685.
6. Freedom RM, Yoo SJ, Perrin D. The biological "scrabble" of pulmonary arteriovenous malformations: considerations in the setting of cavopulmonary surgery. Cardiol Young 2004; 14: 417–437.
7. AboulHosn J, Danon S, Levi D, Castellon Y, Child J, Moore J. Regression of pulmonary arteriovenous malformations after transcatheter reconnection of the pulmonary arteries in patients with unidirectional Fontan. Congenit Heart Dis 2007; 2: 179–184.
8. McElhinney DB, Kreutzer J, Lang P, Mayer JE Jr, del Nido PJ, Lock JE. Incorporation of the hepatic veins into the cavopulmonary circulation in patients with heterotaxy and pulmonary arteriovenous malformations after a Kawashima procedure. Ann Thorac Surg 2005; 80: 1597–1603.
9. Sernich S, Ross-Ascuitto N, Dorotan J, DeLeon S, Ascuitto RJ. Surgical improvement of hepatic venous mixing to resolve systemic arterial hypoxemia associated with post-Fontan pulmonary arteriovenous fistulae. Tex Heart Inst J 2009; 36: 480–482.
10. Kim SJ, Bae EJ, Lee JY, Lim HG, Lee C, Lee CH. Inclusion of hepatic venous drainage in patients with pulmonary arteriovenous fistulas. Ann Thorac Surg 2009; 87: 548–553.