




A case series of three patients with unilateral disconnected pulmonary artery supplied by an ipsilateral patent ductus arteriosus: neonatal ductal stenting as palliation to preserve pulmonary arterial patency

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Background

Disconnected branch pulmonary arteries with a systemic arterial origin of the disconnected vessel is a rare, but well-described entity. Most will have ductal tissue connecting the pulmonary artery to the aorta.

Case summary

We describe in this paper the haemodynamic result in three neonates presenting with ductal origin of a single branch pulmonary artery in the context of trans-catheter stenting procedures to maintain or re-recruit vessel patency. All were faced with potential or actual ductal closure and proceeded to trans-catheter stenting to re-cannulate the duct-dependent pulmonary artery. Two patients with otherwise normal anatomy struggled post-procedure with pulmonary hypertension and right ventricular dilatation. Both required surgical re-anastomosis of the disconnected pulmonary artery during the same admission—one 26 days post-stenting following failure to wean from high-flow respiratory support and the second 8 days post-stenting following failed extubation. In contrast, a patient with tetralogy of Fallot born at 2.5 kg underwent sequential stenting of the right ventricular outflow tract and then the left-sided ductus. He had a good post-procedural course and thrived for several months before complete repair.

Discussion

We describe the clinical courses and discuss the resultant haemodynamics, highlighting the importance of flow to each lung, the resulting haemodynamic implications and the compounding effects of additional lesions.

Keywords

Patent arterial duct • Pulmonary artery • Stenting • Case series

Learning points

- A branch pulmonary artery found to be arising from the aortic arch has a significant probability of being duct-dependent.
- Stenting of an aberrant pulmonary artery with systemic arterial origin with an otherwise normal heart may lead to significant early- and medium-term haemodynamic instability.
- Presence of an atrial septal defect can significantly complicate the haemodynamics.

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Introduction

A unilateral disconnected pulmonary artery supplied by an ipsilateral patent ductus arteriosus is rare. Understanding the incidence is difficult due to varying terminology (hemitruncus,¹ unilateral anomalous origin of a pulmonary artery,^{2,3} or ductal origin of a pulmonary artery^{4,5}). Additionally, some with an absent branch pulmonary artery may have undergone disconnection of a ductal origin of the pulmonary artery following duct closure.⁶ Association with a number of other lesions further complicates estimates of incidence.^{7,8} Surgical re-anastomosis is well-established.^{1,3,7}

Trans-catheter stent implantation to maintain ductal patency was performed first in 1992,^{9,10} and developments in stent technology as well as evolution of the technique have contributed to improved outcomes.¹¹

Within the heterogeneous group presenting for duct stenting is a small subset with duct-dependency of a single lung. Here we compare three such cases to illustrate the challenging haemodynamic situation that may result. In the first two, the right pulmonary artery (RPA) arose from a right ductus originating at the base of the innominate artery in the context of bilateral patent arterial ducts, initially managed with trans-catheter stenting of the occluded right ductus. This resulted in marked instability requiring early surgical reconnection of the disconnected pulmonary artery. We contrast the clinical course with another patient with tetralogy of Fallot (TOF) and a disconnected left pulmonary artery (LPA), in whom stenting of the right ventricular outflow tract (RVOT) and left arterial duct were performed sequentially, resulting in an excellent haemodynamic state for several months.

Timeline

Case 1	
Day 2	Diagnosis of right aberrant pulmonary arising from innominate artery
Day 5	Computed tomography scan demonstrating disconnection of right pulmonary artery (RPA). Prostaglandin infusion started
Day 9	Stenting of occluded ductus supplying RPA
One month	Surgical re-anastomosis of RPA to main pulmonary artery
Case 2	
Day 2	Diagnosis of right aberrant pulmonary arising from innominate artery
5 weeks	Right pulmonary artery found to have disconnected
6 weeks	Stenting of occluded ductus supplying RPA
7 weeks	Surgical re-anastomosis of RPA to main pulmonary artery
Case 3	
11 days	Right ventricular outflow stenting
20 days	Stenting of ductus supplying left pulmonary artery (LPA)
6 months	Repair of tetralogy of Fallot with stent removal and reconnection of LPA to main pulmonary artery

Case presentations

Case 1

A term, 3.1 kg male infant with 22q11 microdeletion syndrome presented in on Day 2 of life with dyspnoea. Examination yielded a soft murmur and moderate increase in work of breathing. Echocardiography demonstrated an RPA arising from the base of the innominate artery of a left-sided aortic arch (*Figure 1A*). A left-sided arterial duct arose from the aortic isthmus to the LPA, shunting bidirectionally. The intra-cardiac anatomy was normal with the LPA connected to the main pulmonary artery (MPA). A computed tomography (CT) scan performed on Day 6 of life showed the RPA had disconnected (*Supplementary material online, Figure S1*), indicating the connecting vessel was a ductus. The left ductus had closed.

Cardiac catheterization demonstrated the previous ductal origin from an ampulla at the origin of the innominate artery (*Figure 1B*). The right duct was crossed easily and two 3.5 × 9 mm Medtronic Integrity bare-metal stents (Medtronic Plc, Minnesota, USA) placed with a good angiographic result (*Figure 1C*).

Although extubated immediately, urgent reintubation was required due to torrential acute ipsilateral pulmonary oedema, with white-out of the right lung on chest X-ray (*Supplementary material online, Figure S2*). He was successfully extubated 48 h later, but over the coming weeks struggled with dyspnoea and poor weight gain. Echocardiography demonstrated both right ventricular dilatation and evidence of systemic pressure in the right ventricle (and thus the LPA, *Figure 1D*) with a bidirectional, but predominant left-to-right inter-atrial shunt.

Following failure to wean from high-flow respiratory support, he proceeded to surgical re-implantation of the RPA to the pulmonary trunk 26 days after his stent procedure. The stent was excised and the RPA reconstructed with a porcine pericardial patch, leaving a good surgical and haemodynamic result. His pulmonary hypertensive changes and right ventricular volume load resolved, and he was discharged home.

Five months post-repair, the reconnected RPA was stented with a 6 mm × 16 mm Cook Formula stent, re-dilated 1 year later. No further re-intervention has been required to date at follow-up of 3.5 years.

Case 2

Case two was a 2.7 kg term infant presenting with dyspnoea on Day 2 of life. An echocardiogram demonstrated an RPA originating from the base of the innominate artery of a left-sided aortic arch. Clinical examination demonstrated mild increase in work of breathing and a soft murmur. There was a moderate-sized atrial septal defect (ASD) shunting left to right, and a ductus from the usual site connected to the LPA. The LPA arose normally from the MPA off a normal right ventricle. Diuretics were started, but over the subsequent weeks, he struggled with poor weight gain and ongoing dyspnoea.

At early follow-up, no flow was seen in the RPA on echocardiography and a prostaglandin infusion commenced. Cardiac CT confirmed the vessel had disconnected (*Supplementary material online, Figure S3*). The left ductus had also closed. Given the combination of relatively low weight and significant distance to the MPA, a decision

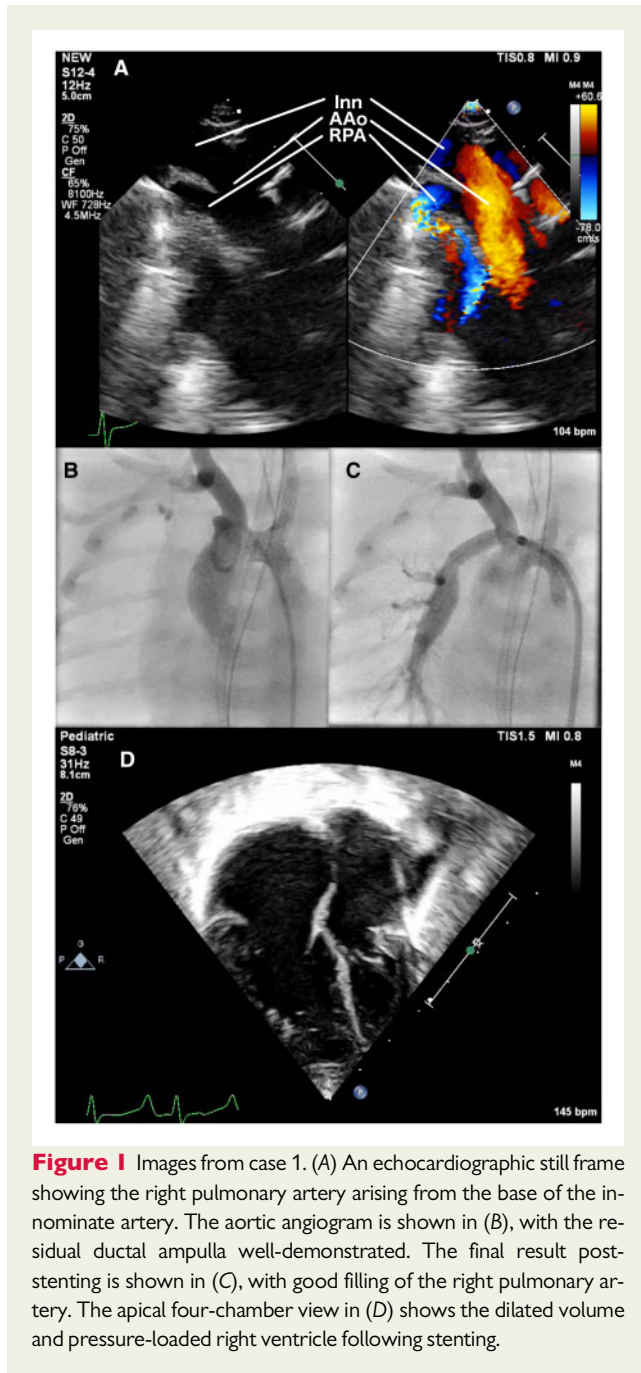


Figure 1 Images from case 1. (A) An echocardiographic still frame showing the right pulmonary artery arising from the base of the innominate artery. The aortic angiogram is shown in (B), with the residual ductal ampulla well-demonstrated. The final result post-stenting is shown in (C), with good filling of the right pulmonary artery. The apical four-chamber view in (D) shows the dilated volume and pressure-loaded right ventricle following stenting.

was made for him to proceed to trans-catheter stenting of his occluded right-sided duct.

Angiography demonstrated a small ampulla at the origin of the innominate artery but no flow into the RPA (Figure 2A). The right-sided ductus was crossed (Figure 2B) and stented with 3.5×12 mm and 3×8 mm Medtronic Integrity bare-metal coronary stents (Figure 2C).

Although initially extubated, within hours he deteriorated with a marked fall in his mixed venous saturations and severe right-sided chest X-ray changes (Supplementary material online, Figure S4A) requiring reintubation and a milrinone infusion.

Over the coming days, he experienced significant haemodynamic instability. Echocardiography demonstrated dilatation of the right

ventricle with evidence of high right ventricular (and thus LPA) pressures (Figure 2D). There was generous flow down the stent into the right lung with diastolic flow reversal in the arch. Flow across the ASD was significant with a large left-to-right shunt.

Although the chest X-ray changes resolved (Supplementary material online, Figure S4B), he failed to extubate, and with a deteriorating clinical state, surgical anastomosis of the aberrant RPA to the MPA with fenestrated ASD closure was performed 8 days after his catheter intervention using an autologous pericardial patch to augment the vessel. Pre-discharge echocardiography demonstrated a normally sized right ventricle with normal pulmonary artery pressures.

One year post-intervention, no further interventions have been required.

Case 3

A male infant was born at term with a birthweight of 2.5 kg following an antenatal diagnosis of TOF. His baseline saturations were in initially in the 80s with a soft murmur and normal work of breathing. The LPA arose from the base of a left innominate artery off a right-sided aortic arch. Although initially well, he developed hypercyanotic spells over the first few days. Prostaglandin infusions and propranolol were commenced. The LPA remained patent, but there was marked flow acceleration across a narrow RVOT, so he was put forward for RVOT stenting to secure antegrade flow to the RPA.

He proceeded to the catheterization laboratory at 11 days of age. The RVOT was crossed and stented with a single 4.5×15 mm Medtronic Integrity bare-metal stent from a right internal jugular approach (Figure 3A and B).

A prostaglandin infusion was continued to maintain ductal patency into the LPA. He returned to the catheterization laboratory on Day 20 of life. The ductus supplying the LPA was severely stenosed and stented with a 3×12 mm Medtronic Driver and a 3×8 mm Abbott Multi-link (Abbott Laboratories, IL, USA) bare-metal stents (Figure 3C and D). He made excellent clinical progress and was discharged home 6 days later.

Complete surgical repair with VSD closure, removal of the right ventricular and ductal stents, reconnection of the LPA, and placement of a pulmonary trans-annular patch was performed at 6 months of age and at 6.5 kg.

He is well and at 1 year post-complete repair no further intervention has been required.

Discussion

Stenting of the arterial duct to both lungs is a regularly performed and well-understood procedure.^{10,11} Stenting to a single lung, however, is rarer and as such less well understood.^{4,7,12}

Our first two cases are examples of ductal origin of a branch pulmonary artery with confirmed bilateral arterial ducts in otherwise normal hearts, where one pulmonary artery arises normally from the right ventricle but the other solely supplied by a ductus. Diagnosis was made in the early postnatal period and supply lost to the aberrantly supplied lung following ductal closure. They were initially managed with stenting of the occluded ductus to re-establish flow and allow growth prior to definitive repair. Both suffered an adverse

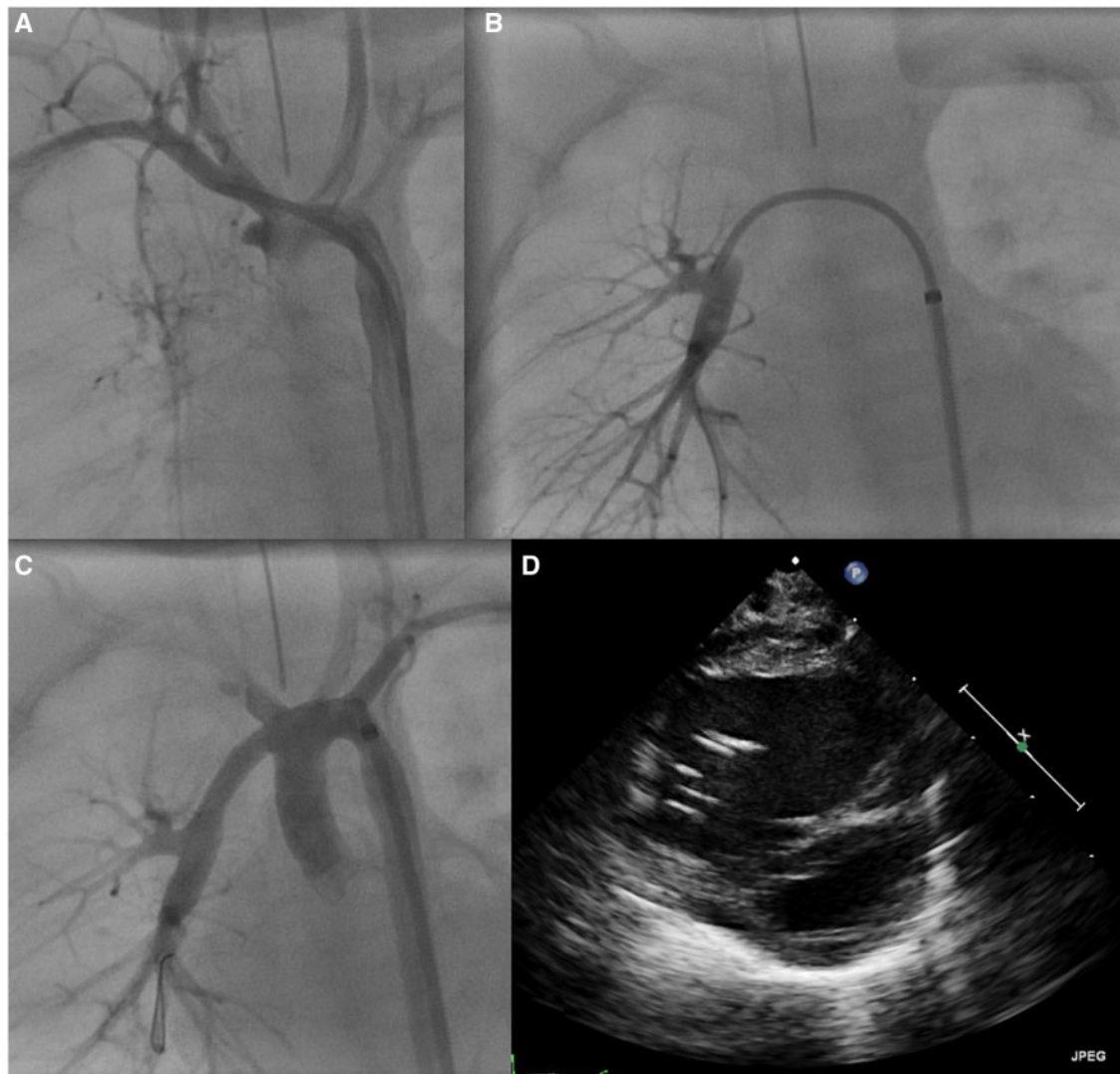


Figure 2 Images from case 2. (A) An angiogram from the innominate artery demonstrating the right-sided ductal ampulla. An angiogram in the right pulmonary artery is shown in (B). Trans-catheter stenting successfully recanalizes the right pulmonary artery (C). An echocardiographic still from the parasternal short axis demonstrates the dilated and both pressure- and volume-loaded right ventricle following stenting of the right pulmonary artery (D).

haemodynamic response necessitating semi-urgent surgical repair during the same admission.

In our patients, we observed two phases to the haemodynamic response following re-recruitment of the disconnected lung. There is an immediate response consisting of unilateral chest X-ray changes with pulmonary oedema in the stented lung, and a later phase with high right ventricular pressures and ventricular dilatation.

We propose that the early phase of unilateral pulmonary oedema results primarily from acute reperfusion injury to the stented lung, exacerbated by an associated increase in left ventricular end-diastolic pressure (LVEDP) from the acute increase in preload. With institution of mechanical ventilation and diuretic therapy, this reverses over a period of days.

Following recovery from this early phase and with resolution of the unilateral chest X-ray changes, both our patients continued to struggle, with evidence of high right ventricular pressures and marked right ventricular dilatation on echocardiography (Figures 1D and 2D). We suggest this results from a high pulmonary blood flow state to both lungs, leading to both pre- and post-capillary pulmonary hypertension, exacerbated by an atrial shunt.

In this anatomy, the normally connected lung receives a twice-normal blood volume, as all the systemic venous return is pumped from the right ventricle into this single lung (Figure 4A and B). In the stented lung, pulmonary flow is dictated by stent diameter and length. Without appreciation that the stented vessel supplies only one lung, excess flow results. In our patients, this may well be compounded by a relatively proximal source of blood flow from the aorta.

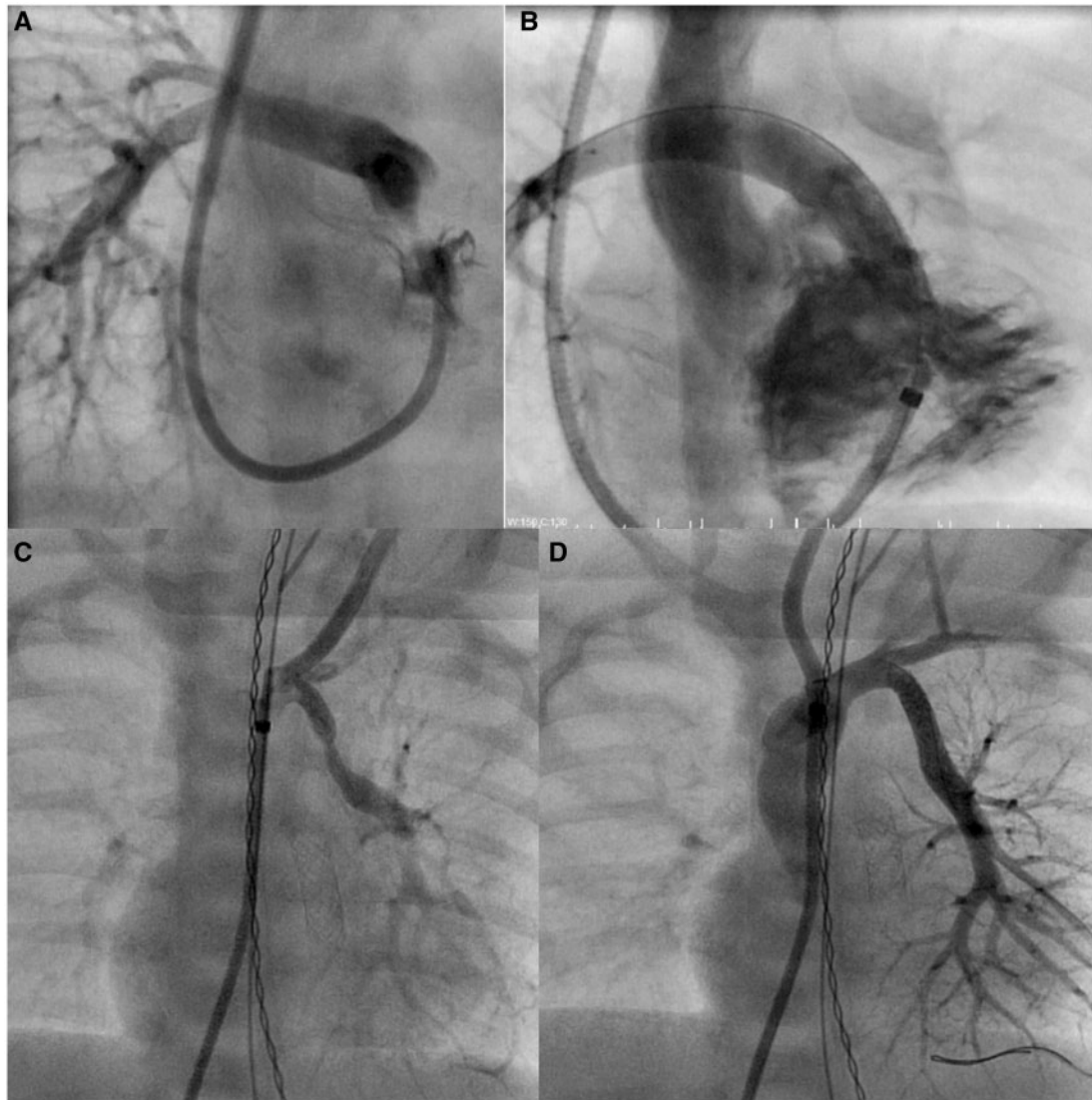


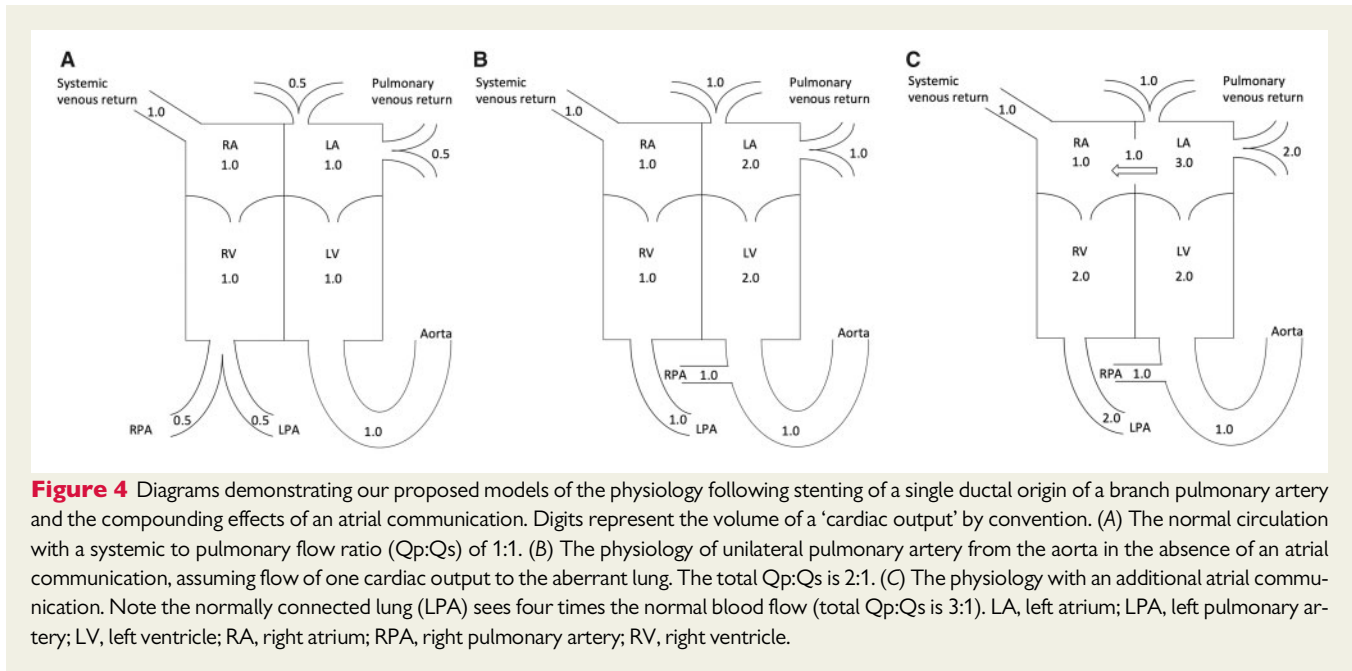
Figure 3 Images from case 3. (A) and (B) are taken from the right ventricular outflow tract stenting procedure, with the initial angiogram demonstrating severe stenosis of the right ventricular outflow tract in (A) and the final result with relief of stenosis post-stenting in (B). (C) An angiographic frame from the ductal stenting procedure with severe stenosis of the ductus feeding the left pulmonary artery, relieved following stenting with good anterograde flow in (D).

Therefore, we propose that a high-flow (and thus high preload) state exists to both lungs, and with a 3.5 mm stent in an otherwise normal heart, the minimum Qp:Qs is 2:1 (Figure 4B). With high pulmonary blood flow, pulmonary venous return is also increased, leading to both increased preload and LVEDP, driving post-capillary pulmonary hypertension.

Any additional left-to-right shunt (such as an ASD) has to be accommodated by the single normally connected lung. Given this lung sees twice-normal blood flow (as the only outlet from the right ventricle), this doubles the effect of any left-to-right shunt (Figure 4C). An ASD therefore increase pre- and post-capillary pressures to the normally connected lung and post-capillary pressure to the stented lung (Figure 4C).

Theoretically, therefore there results high pre-and post-capillary pressures to both lungs following stenting. Although the relative effects to each lung may well be discrepant, the resulting haemodynamic state is extremely challenging. Our patients were also neonates, and usual postnatal adaptation of the pulmonary vasculature is unlikely to have completed, compounding the induced pulmonary hypertension. Also, diastolic runoff to the stented lung can also affect both coronary and systemic blood flow, potentially generating a low cardiac output state.

There are three options from this point for the first two patients. Surgical re-anastomosis of the stented lung would immediately correct the physiology. Surgical reconnection of an aberrant pulmonary artery was first described with an interposition graft, followed soon



afterwards by a primary anastomosis. A number of techniques have been subsequently described.^{1,2,7}

Another option would be to close the atrial communication, although this would only partly correct the deleterious physiology. Interventionally, this has been performed in this context at 6 months of age¹² but did not significantly alter clinical course. Surgical closure would likely be challenging due to venous return from the stented lung on bypass in the context of a relatively small, pulmonary hypertensive patient.

The third option is to limit pulmonary blood flow. Banding or clipping the stent to reduce flow is likely to improve the haemodynamics,¹² albeit potentially again unpredictably, and increased risk of stent occlusion.

In contrast to patients one and two, our 3rd patient with TOF made excellent progress following stenting of the ductus. Pulmonary flow in the context of TOF is fundamentally different, as the starting Qp:Qs is <1:1, whereas the starting point in the first two patients with otherwise normal anatomy is Qp:Qs is 1:1 prior to stenting and augmented from there.

In patients with isolated ductal origin of a single pulmonary artery, haemodynamic instability and hypertension in the stented lung occur in a significant proportion of patients.¹² One multicentre report⁴ has described 10 patients developing pulmonary hypertension, proposing a number of mechanisms of failure. The experience from both our studies highlights the multifactorial haemodynamic problems in this setting.

In conclusion, the physiology following trans-catheter intervention for a single, isolated disconnected pulmonary artery is extremely challenging and can lead to marked haemodynamic instability. At diagnosis, the possibility of bilateral ducts and possible loss of flow to one lung must be considered, and at

intervention, care must be taken to limit stent size and the haemodynamic effects of an ASD considered.

Lead author biography



Following paediatric cardiology training in London, Southampton, and Leeds, Dr Andrew B. Ho works as a consultant paediatric cardiologist at Southampton General Hospital in the UK. His special interest is in congenital cardiac intervention in both children and adults.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patients' next-of-kin in line with COPE guidance.

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References

1. Nathan M, Rimmer D, Piercey G, del Nido PJ, Mayer JE, Bacha EA et al. Early repair of hemitruncus: excellent early and late outcomes. *J Thorac Cardiovasc Surg* 2007;**133**:1329–1335.
2. Garg P, Talwar S, Kothari SS, Saxena A, Juneja R, Choudhary SK et al. The anomalous origin of the branch pulmonary artery from the ascending aorta. *Interact Cardiovasc Thorac Surg* 2012;**15**:86–92.
3. Goldstein BH, Bergersen L, Powell AJ, Graham DA, Bacha EA, Lang P. Long-term outcome of surgically repaired unilateral anomalous pulmonary artery origin. *Pediatr Cardiol* 2010;**31**:944–951.
4. Agrawal H, Petit CJ, Miró J, Miranda CD, Kenny D, Justino H. Contralateral pulmonary hypertension following resuscitation of unilateral ductal origin of a pulmonary artery: a multi-institutional review. *Pediatr Cardiol* 2017;**39**:71–78.
5. Cox D, Quinn R, Moran A, Donnelly J. Ductal origin of the pulmonary artery in isolation: a case series. *Pediatr Cardiol* 2010;**31**:997–1001.
6. Pool PE, Vogel JH, Blount SG. Congenital unilateral absence of a pulmonary artery. The importance of flow in pulmonary hypertension. *Am J Cardiol* 1962;**10**:706–732.
7. Trivedi KR, Karamlou T, Yoo S-J, Williams WG, Freedom RM, McCrindle BW. Outcomes in 45 children with ductal origin of the distal pulmonary artery. *Ann Thorac Surg* 2006;**81**:950–957.
8. Bockeria LA, Podzolkov VP, Makhachev OA, Alekyan BG, Khiriev TK, Zelenikin MA et al. Palliative surgical treatment of congenital heart defects associated with unilateral absence of the pulmonary artery. *Interact Cardiovasc Thorac Surg* 2013;**16**:286–292.
9. Gibbs JL, Uzun O, Blackburn ME, Wren C, Hamilton JR, Watterson KG. Fate of the stented arterial duct. *Circulation* 1999;**99**:2621–2625.
10. Gibbs JL, Rothman MT, Rees MR, Parsons JM, Blackburn ME, Ruiz CE. Stenting of the arterial duct: a new approach to palliation for pulmonary atresia. *Br Heart J* 1992;**67**:240–245.
11. Bentham JR, Zava NK, Harrison WJ, Shauq A, Kalantre A, Derrick G et al. Duct stenting versus modified Blalock-Taussig shunt in neonates with duct-dependent pulmonary blood flow. *Circulation* 2018;**137**:581–588.
12. Krammoh EK, Bigras J-L, Prsa M, Lapierre C, Miró J, Dahdah NS. Therapeutic strategies in children with an isolated unilaterally absent proximal pulmonary artery. *Pediatr Cardiol* 2010;**31**:607–610.