Catheterization and Cardiovascular Interventions 84:1157–1162 (2014)

# Hybrid Procedure of Bilateral Pulmonary Artery Banding and Bilateral Ductal Stenting in an Infant With Aortic Atresia and Interrupted Aortic Arch

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**We report an infant with aortic valve atresia, interrupted aortic arch, ventricular septal defect, confluent pulmonary arteries, bilateral arterial ducts, absent common carotid arteries, and anomalous coronary arteries arising from main pulmonary artery. Hybrid procedure consisting of bilateral pulmonary artery banding and bilateral arterial duct stenting was performed at 4 weeks of age. Hybrid procedure can be an alternative palliative approach in an infant with this complex cardiac anatomy.** © 2014 Wiley Periodicals, Inc.

**Key words: Hybrid procedure; pulmonary artery banding; bilateral arterial ducts; absent carotid arteries; stent placement**

## INTRODUCTION

Persistence of bilateral arterial ducts (AD) is an uncommon congenital malformation, which is most frequently associated with pulmonary atresia and nonconfluent pulmonary arteries [1]. The occurrence of bilateral AD with aortic atresia and interrupted aortic arch (IAA), in which the systemic circulation is supported by bilateral AD, is even rarer [2]. Management of patients with bilateral AD is difficult and the condition is associated with significant mortality, due to the frequent coexistence of heterotaxy syndrome and complex congenital heart disease [3]. Palliative stent placement has been performed in patients with bilateral AD, to stabilize pulmonary and systemic circulation and promote subsequent growth in these critical infants [4,5].

We report an infant with aortic valve atresia, IAA, ventricular septal defect (VSD), confluent pulmonary arteries, bilateral AD, absent common carotid arteries, and anomalous coronary arteries arising from the main pulmonary artery. He underwent a Hybrid procedure which consisted of bilateral pulmonary artery banding and stent placement on bilateral AD at 4 weeks of age.

## CASE REPORT

A 2.5 kg male infant was born at 38 weeks gestation to a 20-year old G3P2 mother. Antenatal echocardiography showed a large perimembranous VSD, severely hypoplastic ascending aorta and aortic valve atresia, widely patent ductal arch, and adequate size of right and left ventricles. Postnatal echocardiography confirmed the diagnosis with left AD arising from confluent pulmonary arteries to the descending aorta. Ascending aorta was severely hypoplastic (2.1 mm in diameter). He required subambient oxygen therapy with FiO2 of 19% for pulmonary over-circulation preoperatively. Fluorescent in situ hybridization confirmed the diagnosis of 22q11.2 deletion syndrome. Cardiac magnetic resonance further delineated his complex cardiac anatomy [6]. IAA was seen with left AD extending to the left sided descending aorta. The left subclavian artery arose from the proximal descending aorta and gave rise to the left vertebral artery. Right AD originated from the proximal right pulmonary artery and extended to the right subclavian artery, which gave rise to the right vertebral artery. Magnetic resonance angiography of neck and head demonstrated absent bilateral common carotid arteries with a normal

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Conflict of interest: Nothing to report.

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Received 4 March 2013; Revision accepted 5 February 2014

DOI: 10.1002/ccd.25424
Published online 8 February 2014 in Wiley Online Library (wileyonlinelibrary.com)

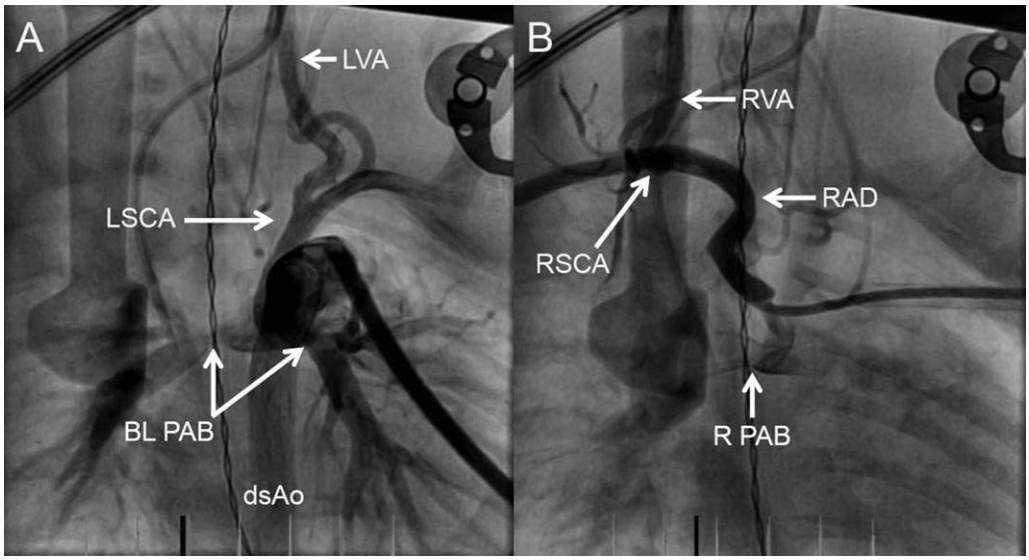


**Fig. 1. Cardiac catheterization, pre-stent (antero-posterior view). A 7-Fr sheath is placed in the main pulmonary artery. A: Angiography from the sheath shows bilateral branch pulmonary arteries, left descending aorta (dsAo), and left subclavian artery (LSCA). Left vertebral artery (LVA) arises from LSCA. Note the banding of bilateral branch pulmonary arteries (BL PAB). B: Selective angiography in the right arterial duct (RAD) shows a small caliber tortuous vessel connected to the right subclavian artery (RSCA). The right vertebral artery (RVA) arises from RSCA. Note the banding in right pulmonary artery (R PAB).**

circle of Willis supplied by a large basilar artery formed by left dominant large vertebral arteries [6]. Head ultrasound was normal. Doppler study of neck vessels showed normal waveforms and patency.

The management options were extensively discussed. We thought that neonatal arch reconstruction and vascular modification was at very high risk and unlikely feasible, given the extremely complicated vascular anatomy. His family requested a possible lowest risk procedure which would allow his family to take him home. Before making the decision to select the Hybrid approach as a palliation, we spent a few weeks on making a correct diagnosis, unifying the inputs from multiple providers, obtaining second opinions from other centers, and discussing care with his family. At the age of 4 weeks, Hybrid procedure was performed to stabilize the systemic circulation and restrict pulmonary blood flow by bilateral pulmonary artery banding and stent placement on bilateral AD. Intraoperative inspection through median sternotomy revealed that a hypoplastic ascending aorta provided no branches and appeared to be filled retrogradely. There were no coronary arteries arising from the ascending aorta. No electrocardiographic change was noted with clamping ascending aorta for 3 min. In fact, both coronary arteries anomalously originated from the main pulmonary artery. Bilateral pulmonary artery banding was performed utilizing a 3 mm Gore-Tex tube graft. After pulmonary artery banding, oxygen saturation decreased from 90% to 85% and diastolic systemic pressure increased by 20 mm Hg. A 7-Fr sheath was placed in the main pulmonary artery. Angiography from the sheath showed complex vascular connections (Fig. 1A). Selective angiography in right AD showed a small caliber tortuous right AD (3.3 mm in diameter) extending to the right subclavian artery, which gave rise to the right vertebral artery (Fig. 1B).

Stent placement on right AD was performed. A 0.014″ Choice wire was positioned through right AD. Firstly, a 4 mm × 15 mm Multilink vision coronary stent (Guidant Corp., Santa Clara, CA) was deployed in the distal right AD (Fig. 2A). Secondly, another 4 mm × 12 mm Multilink Vision coronary stent was deployed in the proximal right AD (Fig. 2B). These two stents were positioned partially overlapped. Stents were carefully positioned to cover the both edges of right AD but maintain non-obstructive flow into the right subclavian artery and right pulmonary artery. Post-stent angiography showed good flow into the right pulmonary artery and stented right AD (Fig. 2C and D).

Stent placement on left AD was then performed. A 0.035″ Rosen wire was positioned through left AD. Pre-stent angiography (Fig. 3A) showed a large caliber left AD (7.1 mm in diameter). A 8 mm × 20 mm Protégé self-expanding stent was deployed in the left AD (Fig.

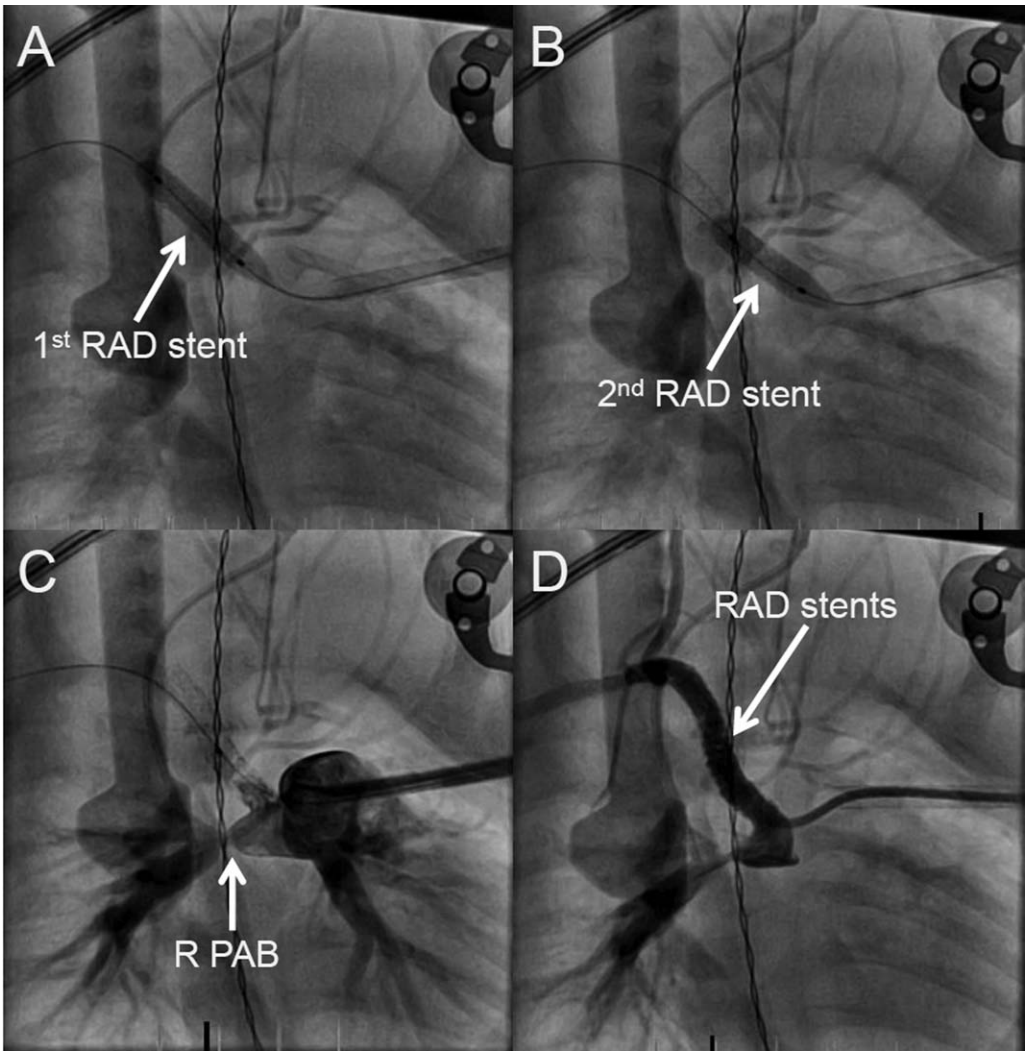


**Fig. 2. Stent placement on right ductus arteriosus (RAD) in antero-posterior view. A: First stent. A 0.014″ Choice wire is positioned in the distal right subclavian artery. A 4 mm × 15 mm Multilink Vision coronary stent is deployed in the distal RAD. B: Second stent. A 4 mm × 12 mm Multilink Vision coronary stent is deployed in the proximal RAD and partially overlapped with the distal stent. C: Post stent, angiography. The second stent covers the proximal portion of RAD without obstructing flow into the right pulmonary artery. Note the banding in right pulmonary artery (R PAB). D: Post-stent, selective angiography in RAD. Note good flow into the stented RAD.**

3B). Stents were carefully positioned to maintain non-obstructive flow into the left subclavian artery. Post-stent angiography showed good flow through the left AD (Fig. 3C). Fluoroscopy showed stable position of stents on bilateral AD (Fig. 3D). Following Hybrid procedure, oxygen saturations decreased to 85% on room air and diastolic BP increased by 20 mm Hg.

The infant was extubated one day after Hybrid procedure and required transient subambient oxygen therapy. He was discharged with aspirin home 28 days after Hybrid. He was followed every week in our clinic and showed a gradual weight gain with satisfactory cardiovascular examination. At 4 months of age, he was doing well with weight of 4,080 g and oxygen saturations of 76% on room air. Echocardiography showed good biventricular function with trivial tricuspid regurgitation. Stent on left AD was widely patent, although stent on right AD was not well visualized. The flow acceleration (4 m/sec) was seen at banding of both proximal branch pulmonary arteries. He was scheduled to undergo diagnostic cardiac catheterization in a few weeks. However, he died at home in his sleep the next morning.

## DISCUSSION

To our knowledge, Hybrid procedure consisting of bilateral branch pulmonary artery banding and stent

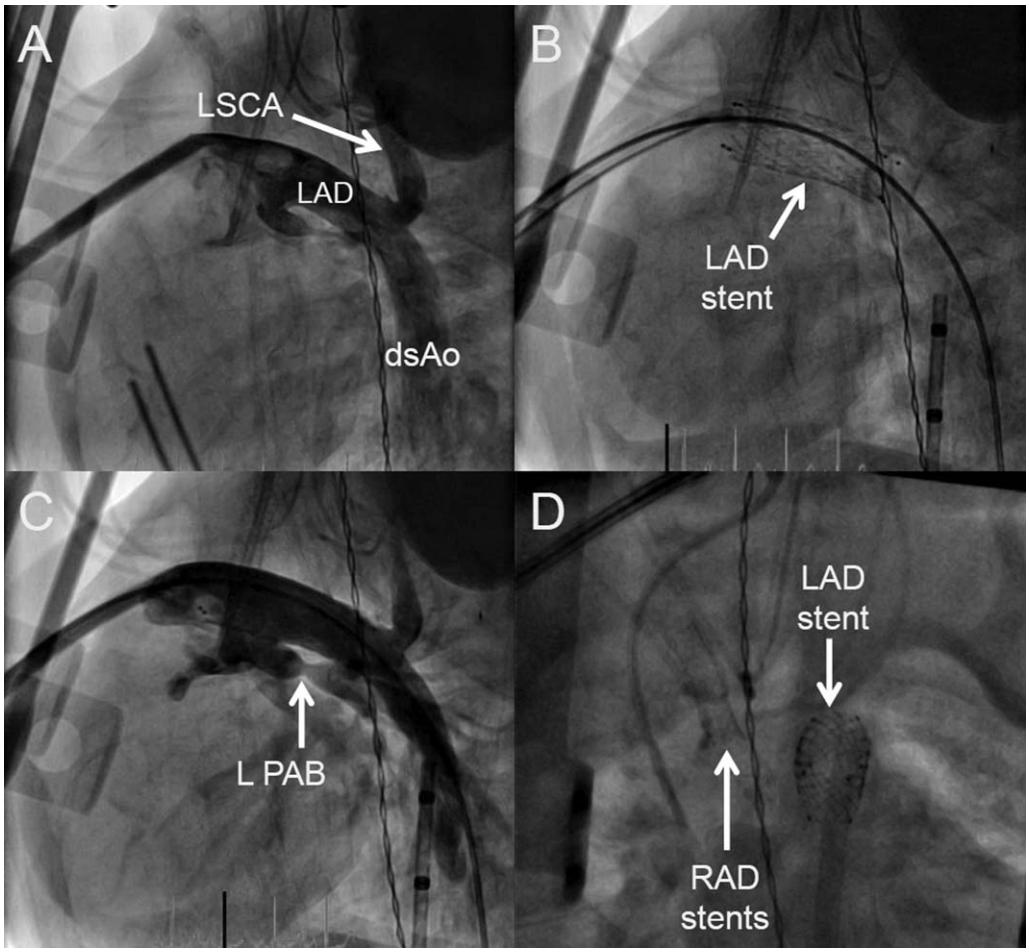


**Fig. 3. Stent placement on left arterial duct (LAD) in lateral views (A–C). A: Pre-stent angiography from the sheath in the main pulmonary artery. A large caliber LAD connects with the descending aorta (dsAo). Left subclavian artery (LSCA) arises from the junction of LAD and dsAo. B: Post-stent fluoroscopy. A Rosen wire is positioned in the dsAo through the stented LAD. C: Post-stent angiography from the sheath. Stent covers the proximal portion of LAD sufficiently but dose not cover the origin of the LSCA. Note the banding in left pulmonary artery (L PAB). D: Post-stent fluoroscopy in antero-posterior view. Note the two overlapped stents in RAD and one larger stent in LAD.**

placement on bilateral AD has not been reported. Magnetic resonance imaging played an important role in defining the extremely complex cardiovascular anomaly. Extra-caution was taken to position stent on bilateral AD not to obstruct flow into both subclavian arteries because of absent bilateral common carotid arteries. Hybrid procedure was successful, resulting in immediate hemodynamic improvement. We planned to discuss a potential next surgical step of aortic arch reconstruction with single or biventricular repair after the scheduled cardiac catheterization. Unfortunately, the infant died at 4 months of age. The cause of death remains unclear. Stent on right AD was not well visualized on echocardiography. Acute thrombus on smaller caliber two overlapping stents on right AD would have caused a potential impairment of cerebral or coronary perfusion.

Bilateral AD is most often found in patients with pulmonary atresia [3], but has been described in three patients with AA with type C IAA [2,7,8]. In this unique anatomy, left AD originates from left pulmonary artery and extends to descending aorta, whereas smaller right AD originates from right pulmonary artery and supports an isolated right subclavian artery. Systemic circulation is entirely supported by bilateral AD. The hypoplastic ascending aorta and coronary arteries are filled retrograde by blood flow from right AD. In contrast, our case had similar cardiac features with additional complexity (Fig. 4A). Bilateral common carotid arteries were absent [6]. Coronary arteries

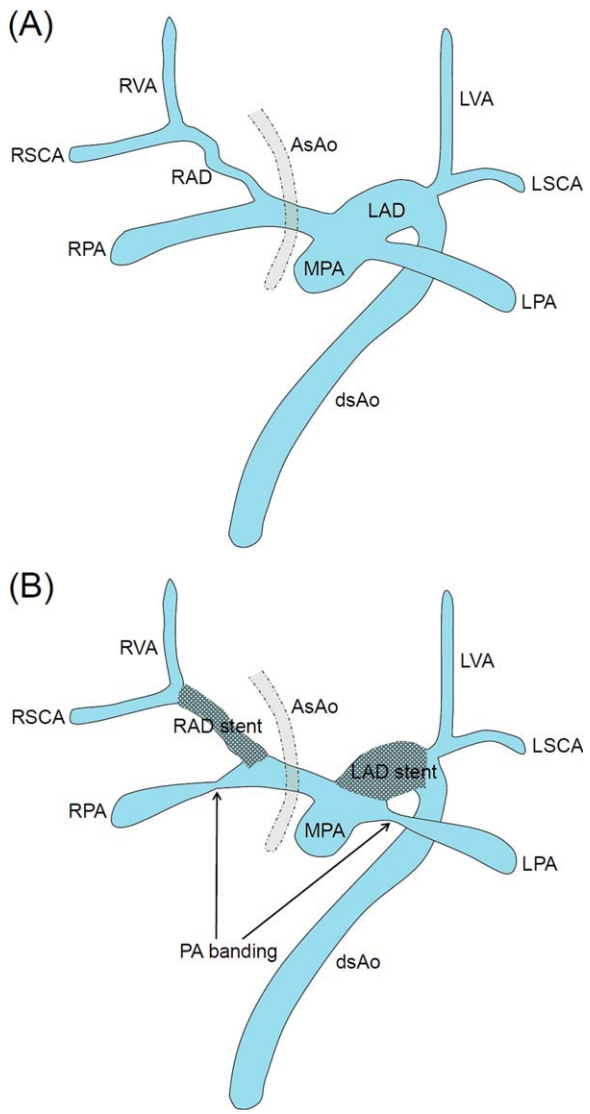


**Fig. 4. Anatomical diagrams in pre-stent (A) and post-hybrid procedure (B) showing an isolated hypoplastic ascending aorta (AsAo) with aortic valve atresia. Right arterial duct (RAD) originates from the proximal right pulmonary artery (RPA) and extends to the right subclavian artery (RSCA). Left arterial duct (LAD) extends to the left sided descending aorta (dsAo). Left subclavian artery (LSCA) arises from the proximal dsAo. Right and left vertebral arteries arise from the proximal RSCA and LSCA, respectively. Post-stent, bilateral pulmonary artery (PA) banding is seen distal to the origin of RAD and LAD. Stents are placed in both RAD and LAD. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]**

anomalously originated from the main pulmonary artery. Total anomalous origin of the coronary arteries from the pulmonary artery is extremely rare and has been reported in patients with septal defects, tetralogy of Fallot, and pulmonary atresia [9].

Percutaneous stent placement on bilateral AD utilizing carotid arteriotomy approach has been reported in two infants with pulmonary atresia and discontinuous pulmonary arteries [4]. The authors suggested certain advantages of AD stenting over surgery: (1) Unbalance of lung perfusion and distortion of the pulmonary arteries can be prevented by having AD stents conform to the size and angulations of the pulmonary arteries; (2) Repeat stent dilation potentially adjusts the shunt size to the pulmonary artery growth. Texter et al. reported stent placement on right AD followed by an additional stent in the pulmonary artery confluence for subsequent left pulmonary artery isolation, in a newborn with double inlet left ventricle, pulmonary atresia, confluent pulmonary arteries, and bilateral AD [5]. In contrast, our patient had aortic valve atresia with IAA, resulting in pulmonary over-circulation. Hybrid procedure allowed us to achieve a balanced pulmonary and systemic circulation; the bilateral pulmonary artery banding restricted the pulmonary blood flow and bilateral AD stenting established the stable systemic circulation.

## CONCLUSION

Hybrid procedure consisting of bilateral pulmonary artery banding and stent placement on bilateral AD can be an alternative palliative approach in an infant with this complex cardiac anatomy.

## AUTHOR CONTRIBUTION

DK is a first author who wrote the manuscript. DRT and RED critically edited the manuscript. SA is the senior author and critically edited and approved the manuscript.

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