

A case report: stent implantation to treat coarctation of persistent 5th aortic arch associated with interrupted 4th aortic arch

Atsuko Kato *, Naoki Ohashi, and Hiroshi Nishikawa

Department of Paediatric Cardiology, Japan Community Health Organization Chukyo Hospital, 1-1-10 Sanjo, Minami-ku, Nagoya 457-8510, Japan

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Background

Persistent 5th aortic arch is a rare cardiac anomaly that is usually surgically corrected during infancy or early childhood if it is associated with coarctation of the aorta. Here, we report an adult with coarctation of the 5th aortic arch who was successfully treated by stent implantation.

Case summary

An asymptomatic 32-year-old woman presented with hypertension and a significant arm–leg difference in pressure. On suspicion of coarctation of the aorta, a chest computed tomography was performed, leading to a diagnosis of an interrupted 4th aortic arch with coarctation of a persistent 5th aortic arch. Percutaneous catheter intervention using a PALMAZ large stent dilated to 12 mm resulted in a minimal peak-to-peak pressure gradient. The patient was discharged home after a 2-day monitoring without hypertension and arm–leg blood pressure difference. She remained normotensive with a patent aortic arch on echocardiography performed 10 months after treatment.

Discussion

As for simple coarctation of the aorta, stent implantation was feasible and effective in an adult patient with coarctation of the 5th aortic arch.

Keywords

Case report • Persistent 5th aortic arch • Coarctation of the aorta • Stent implantation • Adult congenital heart disease

Learning points

- Stent implantation is a safe and effective treatment for management of a coarcted 5th aortic arch in adult patients and can be offered as an alternative to surgical management.
- In cases of refractory systemic hypertension in adults, it is important to measure the four-limb blood pressure to rule out coarctation of the aorta.

Introduction

Persistent 5th aortic arch is a rare congenital heart disease first described in 1969 by Van Praagh in a patient with a ‘congenital double-lumen aortic arch’.¹ Subsequent cases have exhibited variable anatomy and pathophysiology.^{2,3} The most common abnormalities are interruption or severe coarctation of the 4th aortic arch with or without coarctation of the 5th aortic arch.³ It is usually diagnosed and repaired in paediatric patients, especially when coarctation of the sys-

* Corresponding author. Tel: +81 52 691 7151, Email: atsukatou-ngi@umin.ac.jp

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temic arch is clinically significant. Only a few adult cases have been reported.^{4,5} We report an adult case who was successfully treated by intravascular stenting.

Timeline

Time	Events
22 years old	Hypertension was detected at the time of orthopaedic surgery. No investigation was performed.
24 years old	She was found to be hypertensive at a routine medical check-up. Antihypertensive medication (details unknown) was started. She was eventually lost to follow-up because she thought drug was not effective.
31 years old	She was again found to be hypertensive at a routine medical check-up. She was referred to a cardiology clinic, where the arm–leg blood pressure gradient was measured for the first time. Computed tomography (CT) angiography was performed in a tertiary cardiac centre. The diagnosis of coarctation of the aorta was made. She was referred to our hospital for surgical treatment, and surgery was planned by the cardiovascular surgical team.
32 years old	Paediatric cardiologists reviewed the CT angiography, and diagnosed persistent 5th aortic arch with coarctation of the aorta a week before surgery. Cardiac catheterization for stent implantation was offered to the patient. She agreed to undergo catheter intervention. Stent implantation was successfully performed using a PALMAZ large stent, leaving an arm–leg blood pressure gradient of 5 mmHg.
33 years old	She remains normotensive without an arm–leg blood pressure gradient at 10 months follow up.

Case presentation

A 32-year-old woman was found to be hypertensive at a routine medical check-up. She had a 10-year history of hypertension but had stopped taking medication a few years previously because she was asymptomatic and believed that the drug was not effective. A clinic physician noted a significant blood pressure gradient between the arms and leg and referred her to an adult cardiology centre. Computed tomography (CT) angiography of the chest suggested coarctation of the aorta, and she was referred to our hospital for surgery. On admission, the patient's blood pressure was 154/88 mmHg in the right arm, 148/89 mmHg in the left arm, and 110/84 mmHg in the right leg, an arm–leg pressure gradient of 44 mmHg. She had a



Figure 1 Chest X-ray showing bilateral rib notching (arrows).

long 2/6 systolic ejection murmur best heard at the left subclavian area, her femoral pulse was delayed and was weaker than the radial pulse. Chest X-ray demonstrated bilateral rib notching (Figure 1). Echocardiography revealed flow velocity acceleration with diastolic runoff was observed in the distal 5th aortic arch (Figure 2). She had good biventricular systolic function, mild hypertrophy of the left ventricle, and no other cardiac anomalies. She had been scheduled for surgery; however, following review of the images of CT angiography (Figure 3) by paediatric cardiologists, the diagnosis was persistent 5th aortic arch with coarctation and an interrupted 4th aortic arch, which might be amenable to stent implantation. Both surgical and catheter intervention options were subsequently offered, and the patient chose the catheter intervention option.

Cardiac catheterization was performed under general anaesthesia. A haemodynamic study revealed blood pressures of 80/45 mmHg in the ascending aorta and 63/48 mmHg in the descending aorta and a sluggish pressure waveform. Angiography showed a small 5th aortic arch of 11.7 mm diameter with discrete distal narrowing to 3.3 mm (Figure 4A). A PALMAZ[®] Large stent P3008 (Cords, Miami Lakes, FL, USA) was mounted on a 12 mm × 3.5 cm BIB[®] stent placement catheter (NuMED, Hopkinton, NY, USA) and delivered through a 10-Fr long sheath. The stent was successfully deployed at the lesion and angiography showed an increase of the diameter of the distal arch (Figure 4B). The pressure gradient across the stent was 5 mmHg. The patient experienced no complications and was discharged home in 2 days. On discharge, her blood pressure was 110/58 mmHg in the right arm and 112/64 mmHg in the leg. She remained normotensive with no arm–leg blood pressure gradient at 10-months follow-up without any antihypertensive medication.

Discussion

Persistent 5th aortic arch is a rare anomaly that was described by Gupta et al.⁶ as 'an extra-pericardial vessel arising from the ascending

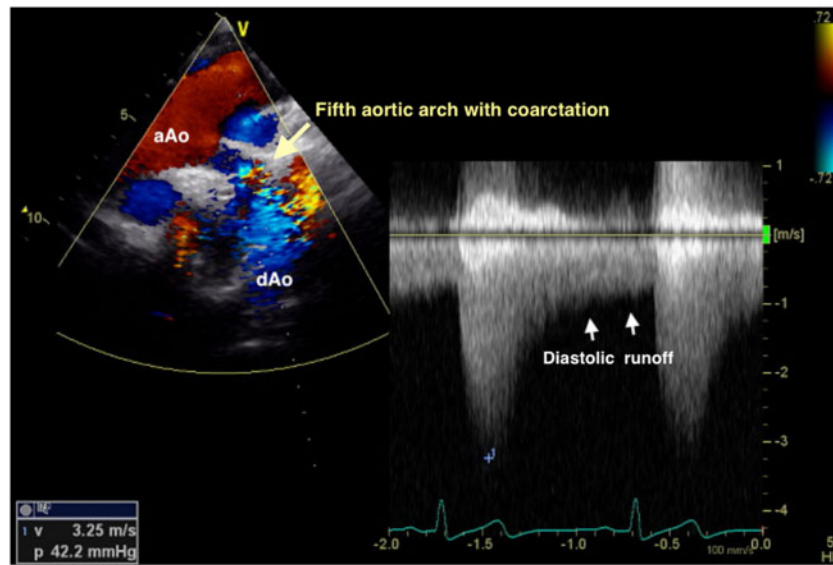


Figure 2 Suprasternal view in transthoracic echocardiography showing flow velocity acceleration with diastolic runoff (white arrows) in the distal 5th aortic arch. aAo, ascending aorta; dAo, descending aorta.

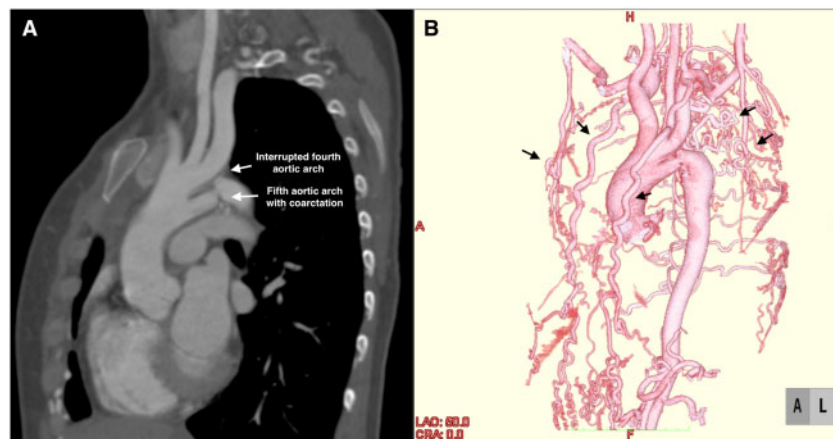


Figure 3 Computed tomography multiplanar reconstruction showing the interrupted 4th aortic arch after the branching of three major neck vessels and the 5th aortic arch with coarctation (A). Computed tomography three-dimensional reconstruction showing significant collateral arteries (arrows) connecting the proximal and distal aorta (B).

aorta proximal to the origin of the brachiocephalic arteries, terminating either in the dorsal aorta or in the pulmonary arteries via the persistently patent arterial duct'. As described by Freedom,⁷ our case had a Type 1 anomaly characterized by a double-lumen aortic arch associated with an interrupted 4th aortic arch and 5th aortic arch coarctation. Some Type 1 patients remain asymptomatic for a lifetime if the 5th aortic arch is relatively patent. The other patients can become critical if the 5th aortic arch becomes coarcted by constriction of

prostaglandin-sensitive ductal tissue that has infiltrated into the artery.⁸ Indeed, the previously reported cases often occurred in infants or young children who needed early surgical intervention for symptomatic coarctation at the initial diagnosis.^{9,10}

Limited symptomatic adult cases have been reported, both of whom presented with hypertension.^{4,5} Our patient had a long history of refractory hypertension without any anatomical evaluation. It is critical to consider secondary hypertension, especially

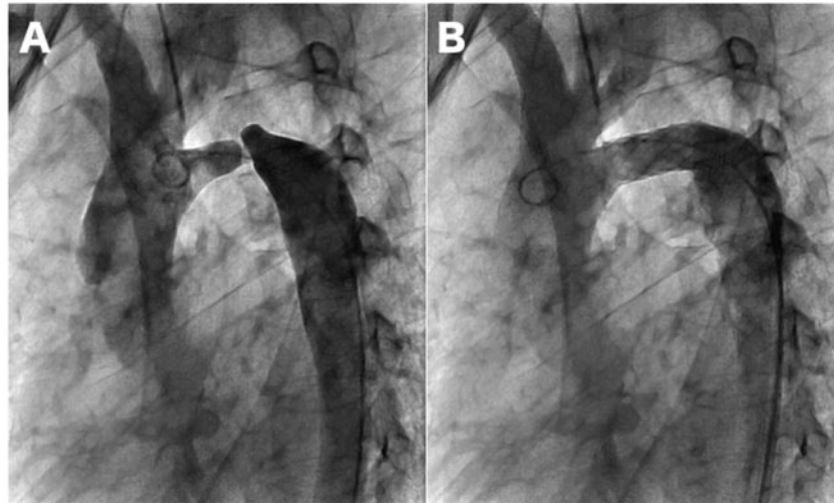


Figure 4 Catheter angiography of the 5th aortic arch (A) before and (B) after implantation of a PALMAZ large stent (P3008E) stent at the coarctation site.

when seeing young healthy adults with ‘treatment-resistant’ hypertension. Coarctation of the aorta is one of the ‘treatable’ diagnoses that can be made by measuring the blood pressure in both the upper and lower extremities or palpating femoral pulse, even though they might be subtle in the presence of collateral arteries. Subsequent cardiac imaging, such as CT angiography or magnetic resonance imaging, could result in a definitive diagnosis.^{2,11}

Both surgery and interventional treatment have been reported for persistent 5th aortic arch with coarctation.^{4,5,8,12} Considering the highly invasive nature of surgery in the presence of the possible numerous collateral arteries in the chest especially in an adult cohort, stent implantation can be a favourable alternative. The recent evidence of its safety and effectiveness strongly support it as the first option in adults with aortic coarctation.^{13,14} Although covered stents are superior to bare metal stents in such patients to avoid aortic wall injury,¹⁴ balloon-expandable covered stents are not available in Japan. We used a bare metal stent, assuming that the risk of aortic wall injury was relatively low because of her small, but acceptable, aortic arch that would not require extensive dilation.¹⁵ Previously, covered and bare CP-stents have been used in two patients with Type 1 persistent 5th aortic arch with coarctation, resulting in good short-term outcomes^{5,12}; we can speculate that the treatment of this disease should be aimed to relieve the obstruction of the 5th aortic arch, like simple coarctation of the aorta, regardless of the presence of interruption or coarctation of the 4th aortic arch. Nonetheless, reintervention of stenting in coarctation of the aorta due to re-stenosis is not uncommon.¹³ In addition, aortic wall injury as well as stent fracture may develop months after successful stent implantation in the aorta.^{13,15} Long-term follow-up is necessary, and its outcome needs to be elucidated in this particular cohort with persistent 5th aortic arch.

In conclusion, stent implantation is a safe, effective, and less-invasive treatment for the patients with a coarcted 5th aortic arch that should be offered to an alternative to surgery.

Lead author biography



I, Atsuko Kato, MD, has been a staff physician at the Department of Paediatric Cardiology at Japan Community Healthcare Organization Chukyo Hospital, Nagoya, since 2017. She graduated from Nagoya City University, School of Medicine in 2005 and started medical internship at Chubu Hospital, Okinawa. After completing the paediatric residency at Nanbu Child Medical Center, Okinawa in 2011, I

had general paediatric cardiology training at Shizuoka Children’s Hospital, Shizuoka for 2 years, then at the Hospital of Sick Children, Toronto, Canada in 2013 for another 2 years. Subsequently, she completed the interventional fellowship in Toronto in 2016. She is board certified in paediatrics and paediatric cardiology. She is currently a member of secretaries of Japanese Pediatric Interventional Cardiology society and its database working group.

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 patient consented to the publication of her case after as complete explanation of its educational value and that publication may help improve the care of other patients in the future. She agreed not to receive any financial benefits.

Conflict of interest: none declared.

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