Persistent fifth aortic arch stenosis associated with type A interruption of the aortic arch: a report of six cases

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To the Editor: Persistent fifth aortic arch (PFAA) is a rare congenital cardiovascular malformation that occurs when the pharyngeal fifth aortic arch does not degenerate during the embryonic period. The first case of PFAA was described in an autopsy specimen in 1969.[1] In 1973, the persistence of a left fifth aortic arch was first reported.[2] Since then, several case reports have described PFAA in different forms. PFAA stenosis associated with type A interruption of the aortic arch (type-A IAA) is most common in clinical practice and requires surgical intervention. From 2013 to 2018, six cases [Tables 1 and 2] were diagnosed using echocardiography and computed tomography angiography (CTA). Five cases were successfully treated with surgery and followed-up. The purpose of this study was to summarize the clinical characteristics and diagnostic features of PFAA stenosis associated with type-A IAA to improve its diagnostic accuracy and allow for complete pre-operative preparation and proper treatment.

Informed consent was obtained from their parents of the six individuals described here. The severity and timing of PFAA clinical symptoms vary. Five patients' symptoms appeared in the first 3 months of life. The sixth patient had no symptoms until age 4. The five infant patients were in critical condition because of pneumonia, respiratory failure, and congestive heart failure. Three of them were neonates in the intensive care unit, and their symptoms also included jaundice, difficulty feeding, and ductal shock. Physical examination revealed hypertension, systolic murmur, and differences in blood pressure in the upper and lower extremities. Echocardiography showed the structure of the great arteries and cardiac function allowing a correct initial diagnosis of PFAA stenosis. The diameter of PFAA stenosis ranged from 2.0 to 3.2 mm, and the pressure gradient ranged from 64 to 73 mmHg. Changes in the intra-cardial structure included left atrioventricular dilatation, thickened left ventricular wall, and decreased left ventricular function.

The 4-year-old patient with PFAA was admitted to the cardiology department after the discovery of hypertension (highest blood pressure was 150/120 mmHg and lowest blood pressure was 107/70 mmHg). Routine echocardiography revealed PFAA stenosis with type-A IAA. The inner diameter of the fifth arch stenosis was 3.6 mm, and the pressure gradient was 50 mmHg.

Five patients underwent surgical treatment according to their specific conditions. Three patients underwent resection of the fifth aortic arch using end-to-end or end-to-side anastomosis of the descending aorta. One patient underwent resection of the fifth aortic arch, subclavian artery transplantation, and subclavian artery anastomosis of the descending aorta. The 4-year-old patient had a fourth artificial pipe connection and retained the fifth aortic arch. The pressure differences between the upper and lower limbs of these five cases before surgery were more than 40 mmHg, and the pressure difference after surgery was essentially the same. Post-operative follow-up revealed stable conditions.

During embryological development of the great artery, the bilateral fifth primitive arches usually degenerate early after appearing or do not appear at all. However, for some reason, the fifth arch may persist unilaterally or bilaterally and completely or partially, producing a wide variety of appearances. Because of its embryologic location, the persistent fifth arch is an extra-pericardial vessel arising from the ascending aorta proximal to the origin of the brachiocephalic arteries, ending either in the dorsal aorta or in the pulmonary arteries via the sixth arch. For practical purposes, some scholars have classified the proximal and distal connections of the vessel and the blood flow direction into four types: systemic to systemic,

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Table 1: General clinical performance.

Case	Sex	Age (months)	Initial symptoms	Body temperature (°C)	Breathing (times/ min)	Heart rate (beats/min)	LULBP (mmHg)	RULBP (mmHg)	LLLBP (mmHg)	RLLBP (mmHg)	Heart murmur
1	M	1	Jaundice, pneumonia	36.8	45	135	92/54	94/55	71/34	70/35	Yes
2	M	1	Pneumonia, congestive heart failure	37.6	50	192	109/79	133/85	78/54	81/49	Yes
3	M	3	Severe pneumonia	36.7	30	130	102/85	98/79	62/35	59/33	Yes
4	F	3	Heart murmur	36.6	36	146	86/49	86/67	68/34	69/32	Yes
5	M	48	Hypertension	36.7	20	108	118/80	120/80	70/35	68/35	Yes
6	M	0	Poor response, difficulty breathing	36.1	46	147	93/65	94/58	76/53	77/52	Yes

M: Male; F: Female; LLLBP: Left lower limb blood pressure; LULBP: Left upper limb blood pressure; RLLBP: Right lower limb blood pressure; RULBP: Right upper limb blood pressure.

Table 2: Imaging results of the patients.

Case	Left atrioventricular dilatation	Thickened left ventricular wall	LVEF	PH	IAA	PFAA (diameter, stenosis, velocity, pressure gradient)	Complications	CT confirmation	Surgery confirmation
1	+	+	45%	+	Type A	6.5–2.2, 4.15 m/s, 69 mmHg	ASD 6.8 mm, TR, PE	+	–
2	+	+	63%	+	Type A	8.0–3.2, 4.29 m/s, 73 mmHg	PFO 2.4 mm, MR, TR	+	+
3	+	+	53%	–	Type A	5.0–3.2, 4.09 m/s, 67 mmHg	PFO 3.7 mm, MR, TI	+	+
4	+	+	44%	–	Type A	5.5–2.9, 4.02 m/s, 65 mmHg	PFO 3.8 mm, MR	+	+
5	–	–	69%	–	Type A	7.3–3.6, 3.52 m/s, 50 mmHg	–	+	+
6	+	+	37%	+	Type A	5.7–2.0, 4.00 m/s, 64 mmHg	PFO 2.0 mm	+	+

+ indicates yes; LVEF: Left ventricular ejection fraction; PH: Pulmonary hypertension; IAA: Interrupted aortic arch; ASD: Atrial septal defect; CT: Computed tomography; MR: Mitral regurgitation; PE: Pericardial effusion; PFAA: Persistent fifth aortic arch; PFO: Patent foramen ovale; TR: Tricuspid regurgitation; TI: Tricuspid incompetence.

systemic to pulmonary, pulmonary to systemic, and bilateral; each type has different sub-types.[3,4] According to the sub-types and combinations of cardiovascular malformations, PFAA has different clinical manifestations. Currently, increasing understanding of PFAA and additional reports of special cases, the disease spectrum and clinical sub-types are broadening.[5] The incidence of PFAA is unknown. It has been labeled the "great pretender" as it mimics anatomically similar structures; therefore, a lack of understanding of the disease easily leads to misdiagnoses.[4,6]

PFAA stenosis associated with type-A IAA is considered a systemic-to-systemic connection and is most commonly found in clinical practice. In this disease, the (fourth) aortic arch is interrupted after the left subclavian artery arises; the fifth aortic arch originates from the ascending aorta opposite or proximal to the brachiocephalic artery and ends in the descending aorta, and its last part is always narrow. The fifth aortic arch has a critical role in survival because it is a unique channel connected to the ascending and descending aorta. Furthermore, the degree of stenosis determines the symptoms.

Echocardiography and CTA have important roles in the diagnosis of PFAA stenosis because they can accurately describe the location and connection of the PFAA [Figure 1A–1D]. Diagnoses include: interruption of the fourth aortic arch (type A), whereby the three brachiocephalic arteries are clusters arising from the fourth aortic arch unconnected to the descending aorta; the fifth aortic arch originating from the ascending aorta opposite or proximal to the brachiocephalic artery and ending in the descending aorta; and PFAA with descending aorta joints with different degrees of stenosis. Imaging is important for determining the appropriate treatment and prognosis.

As the "great pretender," PFAA stenosis associated with interruption of the fourth aortic arch may not be easy to recognize and may be more common than originally thought. If the definitive fourth aortic arch continues, then identifying the extra-arterial channel of the fifth arch may be straightforward. However, with interruption of the fourth arch, the fifth arch becomes the sole aortic arch making the diagnosis extremely challenging; Therefore, it is easily misdiagnosed as coarctation of the aortic arch (COA) in clinical practice because the hemodynamic

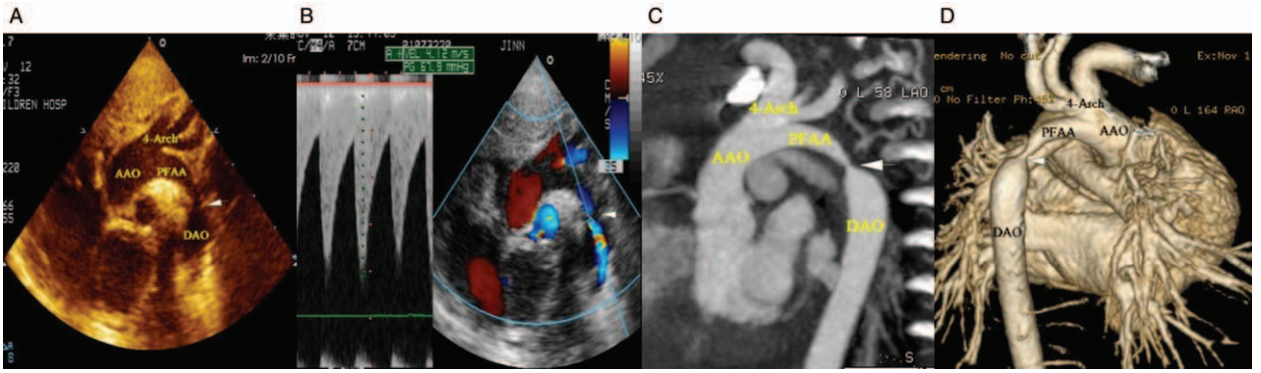

Figure 1: Echocardiography and CTA imaging. (A) Two-dimensional echocardiography showed type-A IAA, the three brachiocephalic arteries clusters arising from the fourth aortic arch unconnected to the descending aorta; PFAA originating from the distal ascending aorta and ending in the proximal DAO; white arrow indicating the location of PFAA stenosis; (B) Continuous Doppler echocardiography showed the velocity of PFAA stenosis was 4.12 m/s, pressure gradient was 68 mmHg; (C and D) CTA images further confirmed the echocardiography findings. 4-Arch: Fourth aortic arch; AAO: Ascending aorta; CTA: Computed tomography angiography; DAO: Descending aorta; PFAA: Persistent fifth aortic arch; Type-A IAA: Type A interruption of the aortic arch.

manifestations of these two diseases are identical. The fifth aortic arch is in an unusually inferior position; it approaches the same level as the main pulmonary trunk and arterial duct, with all brachiocephalic arteries appearing as a cluster originating from a single connection (the fourth aortic arch) to the ascending aorta that is unconnected to the descending aorta. Therefore, correct diagnosis is possible by focusing attention on these characteristic changes.

Reports have indicated that formation of the fifth aortic arch and its characteristics are different from the normal arterial walls, and pathological reports have also confirmed that the fifth aortic arch can be found in ductal tissue organization.[7-9] With PFAA, narrow elastic fibers with sensitivity to prostaglandins instead of smooth muscle fibers have been observed. With age, these can become narrower. Once diagnosed, this type of PFAA requires surgery.[7-9]

PFAA has been misdiagnosed as COA when discovered intra-operatively following left thoracotomy, and it has been treated with aortic arch advancement. Although the surgery was ultimately successful, it was potentially unsafe, thus underscoring the importance of correct diagnoses before pre-operative planning.[10]

In summary, a complete understanding of embryologic aortic arch development and the diagnostic features of PFAA stenosis are important for improving diagnostic accuracy. This allows for complete pre-operative preparation and correct treatment conducive to long-term outcomes in children.[11] PFAA can coexist with a variety of cardiovascular malformations that are difficult to diagnose with echocardiography alone; therefore, combining echocardiography with CTA greatly improves the diagnostic rate.

Conflicts of interest

None.

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