

# Assessment of persistent fifth aortic arch by echocardiography and computed tomography angiography

Haiyan Yang, MD<sup>a,b,e</sup>, Xu Zhu, MD<sup>a,e</sup>, Chun Wu, MD<sup>c,e</sup>, Xiaodong Zhao, PhD<sup>d,e,\*</sup>, Xiaojuan Ji, PhD<sup>a,e,\*</sup>

## Abstract

To evaluate the utility of echocardiography (echo) in the diagnosis of persistent fifth aortic arch (PFAA), a very rare congenital aortic arch anomaly, and to compare echo and computed tomography angiography (CTA) imaging findings to improve our understanding of this anomaly.

Data on the clinical diagnosis, imaging findings, and clinical management of PFAA were retrospectively analyzed in 10 suspected cases of PFAA admitted to our hospital between January 2012 and February 2017. We compared echo as a first line examination modality, and CTA and surgery results as the gold standard. Weinberg's classification was used to classify the type of PFAA.

All patients (100%) received echo examination, eight patients (80%) received CTA examination, and four patients (40%) received sternotomy surgery; all recovered well after surgery. According to Weinberg's classification, 2, 6, and 2 cases (20%, 60%, and 20%) were classified as Type A, B, and C, respectively. Echo was able to diagnose 5 cases of PFAA (1 Type A case and 4 Type B cases) in the first instance. The diagnostic conformance rate of echo was 62.5% after comparisons with CTA and surgery results.

The clinical manifestation of PFAA was atypical, and its diagnosis depended primarily on medical imaging. Echo has a relatively high diagnostic accuracy for PFAA, which is very valuable for its early detection.

**Abbreviations:** 2D = 2-dimensional, AA = aortic atresia, APW = aortic pulmonary window, CAG = cardiac catheterization angiography, CDFI = color Doppler flow imaging, CE-MRA = enhanced magnetic resonance angiography, CHD = congenital heart disease, CoA = coarctation of aorta, CTA = computed tomography angiography, DAA = double aortic arch, DSA = digital subtraction angiography, ECG = electrocardiogram, echo = echocardiography, IAA = interruption of the aortic arch, MRA = magnetic resonance angiography, PA = pulmonary atresia, PDA = patent ductus arteriosus, PFAA = persistent fifth aortic arch, PFO = patent foramen oval, PGE1 = prostaglandin E1, TA = tricuspid atresia, TGA = transposition of the great arteries, TOF = tetralogy of fallot, VSD = ventricular septal defect.

**Keywords:** CTA, diagnosis and treatment, echocardiography, PFAA

Editor: Kanhua Yin.

The work was supported by National Natural Science Foundation of China (Grant no. 81301300); Project of Chongqing Science and Technology Bureau (cstc2016shmszx 130009); China Postdoctoral Science Foundation funded project (2018T110949); and Project of Chongqing Municipal Human Resources and Social Security Bureau (cx2019065).

The authors have no conflicts of interest to disclose.

<sup>a</sup> Department of Ultrasound, Children's Hospital of Chongqing, <sup>b</sup> State Key Laboratory of Ultrasound in Medicine and Engineering, College of Biomedical Engineering, Key Laboratory of Biomedical Engineering, Chongqing, <sup>c</sup> Department of Cardiothoracic Surgery, <sup>d</sup> Department of Pediatric Research Institute, <sup>e</sup> Ministry of Education Key Laboratory of Child Development and Disorders, National Clinical Research Center for Child Health and Disorders; China International Science and Technology Cooperation base of Child development and Critical Disorders; Chongqing Key Laboratory of Pediatrics, Children's Hospital of Chongqing Medical University, Chongqing, P.R. China.

\* Correspondence: Xiaojuan Ji, Chongqing Medical University Affiliated Children's Hospital, Chongqing, Chongqing China (e-mail: 400146@cqmu.edu.cn); Xiaodong Zhao, Chongqing Medical University Affiliated Children's Hospital, Chongqing, Chongqing, China (e-mail: zhaoxd530@aliyun.com).

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How to cite this article: Yang H, Zhu X, Wu C, Zhao X, Ji X. Assessment of persistent fifth aortic arch by echocardiography and computed tomography angiography. *Medicine* 2020;99:9(e19297).

Received: 10 September 2019 / Received in final form: 23 January 2020 / Accepted: 23 January 2020

<http://dx.doi.org/10.1097/MD.00000000000019297>

## 1. Introduction

Persistent fifth aortic arch (PFAA) is widely recognized as an exceedingly rare congenital cardiovascular anomaly of the aortic arch, which is due to the fifth aortic arch not degenerating in the embryo, and is frequently associated with other cardiovascular anomalies.<sup>[1–3]</sup> The embryological mechanism of PFAA was described by Congdon in 1922,<sup>[1]</sup> and first reported by Van Praagh at autopsy in 1969.<sup>[2]</sup> The first two living cases of PFAA were reported by Izukawa in 1973.<sup>[3]</sup> Since then, PFAA has been described mostly in individual case reports because of its rarity. To our best knowledge, there have been very few studies of PFAA.

PFAA is classified as either type A, B, or C according to different abnormal vascular connections.<sup>[4]</sup> Subsequently, some new classifications have been described by relating the proximal and distal connections of the vessel and the direction of blood flow.<sup>[5]</sup> PFAA stenosis associated with type B interruption of the aortic arch (type-B IAA) is most common in clinical practice and requires surgical intervention.<sup>[6]</sup> Currently, digital subtraction angiography (DSA) with cardiac catheterization angiography (CAG) is considered the gold standard for diagnosis of congenital aortic arch anomalies. However, conventional DSA with CAG requires the use of a contrast agent, which remains controversial with regard to ionization radiation and potential allergic reactions.<sup>[7]</sup> Echo is an indispensable diagnostic tool because of its noninvasive repeatability, low cost, and ability to show vascular malformations.<sup>[8]</sup>

In the present study, we report 10 PFAA cases confirmed at our hospital from January 2012 to February 2017. We retrospectively analyzed the patients' clinical data, including their clinical classifications, imaging features, treatment, and follow-up. The aim of this study was to evaluate the accuracy of echo for the diagnosis of PFAA and to compare it with computed tomography angiography (CTA).

**2. Methods**

We retrospectively investigated 10 cases of PFAA and calculated the incidence of each type (the ratio of PFAAs for each type vs the total number of patients). This study was based on data obtained between January 2012 and February 2017 in our hospital. The study protocol was approved by the institutional ethics committee. The requirement for informed consent was waived due to the retrospective nature of the research. Data were collected from medical records, including physical manifestations, echocardiograms, CTA, and surgery reports.

All patients received echo examination (Philips IE 33 [Philips; Bothell, MA] and GE Vivid E9 [GE Healthcare; Milwaukee, MI]) with a frequency of 2 MHz to 8 MHz using 2-dimensional (2D) and color Doppler flow imaging (CDFI) through the conventional left parasternal, subxiphoid, apical, and suprasternal notch views. CTA was performed on a GE Light Speed VCT (GE Healthcare) scanner. A Medrad Vistron high pressure syringe (Medrad Inc., Indianola, PA) was used for high pressure injection of Omnipaque 300 (Daiichi-Sankyo; Tokyo, Japan) (350 mg/ml) through a median antebachial vein.

In this study, we categorized PFAA patient according to Weinberg type: Type A is a double-lumen aortic arch with or without arch hypoplasia or coarctation; Type B is atresia or interruption of the fourth aortic arch with a PFAA; Type C is a systemic-to-pulmonary arterial connection with or without pulmonary or systemic arterial obstruction.<sup>[4]</sup>

**3. Results**

There were 7 males (70%) and 3 females (30%). The youngest patient was 12 days old and the oldest patient was 11 years old. The median age was 6.50 months. The average birth weight was 3.09 ± 0.35 kg, and all the patients were born at full-term.

Nine patients (90%) were first admitted to the respiratory department for coughing, wheezing, cyanosis, or poor activity; almost all of them had heart murmurs (grade 2 or 3 systolic murmurs). Only 1 patient (10%) had a family history of congenital heart disease (CHD), but the details were unclear. One patient (10%) was referred to the outpatient clinic due to myasthenia gravis and dyspnea. Chest X-rays on the remaining nine cases showed pneumonia or cardiac enlargement. The electrocardiograms (ECG) showed sinus arrhythmia; however, one case showed ST-T change. Six patients (Weinberg type B, combined with IAA) had obvious blood pressure differences, as high as 30 to 50 mmHg, between the arms and lower limbs upon physical examination (Table 1).

According to Weinberg classification,<sup>[4]</sup> Type A, B, and C accounted for 2, 6, and 2 of the 10 cases (20%, 60%, and 20%), respectively. Weinberg type B was the most common type of PFAA in this cohort, and 50% of the patients were associated with IAA (type A, 40%; type C, 10%). Two patients (20%) had a patent foramen oval (PFO), two patients (20%) had a right aortic arch, and the remaining eight patients (80%) had a left aortic arch.

Among the 10 patients, 8 (80%) received CTA examination and 4 (40%) received sternotomy surgery. Two patients only received echo examination and were diagnosed as PFAA. Among the 8 patients who received CTA examination, 5 cases of PFAA (1 type A case and 4 type B cases) were first diagnosed by echo, with a diagnostic accuracy rate of 62.5%. One case of type A and one case of type B PFAA were not detected; hence, the rate of misdiagnosis was 25.0%. In addition, one case (12.5%) of type C PFAA was misdiagnosed as an aortic pulmonary window (APW) (Table 2).

Four of the 10 patients (40%) received sternotomy to correct their cardiovascular malformations and were discharged on day 17 ± 2 after surgery. Their postoperative history was uncomplicated, and their blood pressures were restored within the normal range after surgery, without cardiac insufficiency, obstruction, pulmonary hypertension crisis, or serious lung infection. Preoperative and postoperative aortic pulmonary window echocardiographic and CTA images of a representative case for each type are shown in Figures 1 to 5. Patients were mainly followed up using echo after surgery, with the longest follow-up time being nearly 5 years. Upon follow up, all patients

**Table 1**  
General clinical performance of 10 cases of persistent fifth aortic arch.

Case	Age	Sex	Clinical symptoms	Breathing (times/min)	Heart rate (beats/min)	LULBP (mmHg)	RULBP (mmHg)	LLLBP (mmHg)	RLLBP (mmHg)	Heart murmur	ECG	Outcome
1	12 days	F	Anorexia	56	140	128/70	134/65	103/55	106/72	-	sinus arrhythmia	Lost to follow-up
2	26 days	F	Cough, Choking milk	45	143	139/58	127/68	102/73	108/81	-	ST-T	Surgery (good recovery)
3	1 month	M	Short breath, cough, cyanosis	69	145	123/68	121/73	84/46	82/51	+	sinus arrhythmia	Lost to follow-up
4	6 months	M	Cough, Pneumonia	50	139	139/58	127/68	82/43	78/31	+	sinus arrhythmia	Surgery (good recovery)
5	7 months	M	Heart murmur	34	128	107/80	105/71	106/69	110/75	+	sinus arrhythmia	Lost to follow-up
6	10 months	M	cyanosis	30	125	136/61	134/66	73/46	80/48	-	sinus arrhythmia	Surgery (good recovery)
7	1 year	M	cyanosis	38	138	128/70	134/65	75/34	60/32	+	sinus arrhythmia	Lost to follow-up
8	11 years	F	Weakness of eyelids and legs	51	146	108/72	104/61	96/64	79/45	-	sinus arrhythmia	Died
9	2 months	M	Pneumonia	55	129	121/68	114/77	105/84	110/72	+	sinus arrhythmia	Lost to follow-up
10	9 months	M	Cough, Pneumonia	57	135	118/71	124/55	95/54	90/62	+	sinus arrhythmia	Surgery (good recovery)

Note: ECG = electrocardiogram, F = female, LLLBP = left lower limb blood pressure, LULBP = left upper limb blood pressure, M = male, RLLBP = right lower limb blood pressure, RULBP = right upper limb blood pressure.

+ indicates yes; - indicates no.

**Table 2**  
**Echocardiology imaging results.**

Case	LVD	TLVW	LVEF	LVFS	PH	PFAA (diameter, stenosis, velocity, pressure gradient)	Weinberg type	Anomalies	Confirmation
1	+	+	60%	36%	+	16.5mm, 3 mm, 4.09 m/s, 68 mmHg	Type A	PDA/ASD/VSD/persistent L SVC	Only Echo
2	+	+	47%	29%	+	missed diagnosis	Type A	PDA/CoA/right aortic arch	CTA + surgery
3	+	+	31%	14%	-	7 mm, 3.2 mm, 4.05 m/s, 63 mmHg	Type B	IAA (type A)/PTA/VSD/ASD/TGA	CTA
4	+	+	45%	21%	+	missed diagnosis	Type B	AS/CoA/LPAS/PFO	CTA + surgery
5	+	+	65%	35%	+	14 mm, 3.5 mm, 4.15 m/s, 59 mmHg	Type B	IAA (type A)/PDA	CTA
6	+	+	64%	34%	+	12 mm, 2.8 mm, 4.56 m/s, 83 mmHg	Type B	IAA (type A)	CTA + surgery
7	+	+	50%	31%	+	10.8 mm, 3.1 mm, 3.98 m/s, 56 mmHg	Type B	IAA (type C)/PTA/VSD/ASD/TGA	CTA
8	+	+	65%	29%	-	13.4 mm, 3.2 mm, 4.26 m/s, 71 mmHg	Type B	IAA (type A)	Only Echo
9	-	-	71%	38%	+	9.8 mm, 3 mm, 4.06 m/s, 65 mmHg	Type C	PDA/ASD/persistent L SVC	CTA
10	-	-	68%	32%	-	misdiagnosed as APW	Type C	PFO/right aortic arch	CTA + surgery

Note: AS = aortic stenosis, ASD = atrial septal defect, CoA = coarctation of aorta, CTA = computed tomography angiography, IAA = interrupted aortic arch, LPAS = left pulmonary artery stenosis, L SVC = left superior vena cava, LVD = Left ventricular dilatation, LVEF = left ventricular ejection fraction, LVFS = left ventricular fractional shortening, PDA = patent ductus arteriosus, PFAA = persistent fifth aortic arch, PFO = patent foramen oval, PH = pulmonary hypertension, PTA = persistent truncus arteriosus, TGA = transposition of the great arteries, TLVW = thickened left ventricular wall, VSD = ventricular septal defect. + indicates yes; - indicates no.

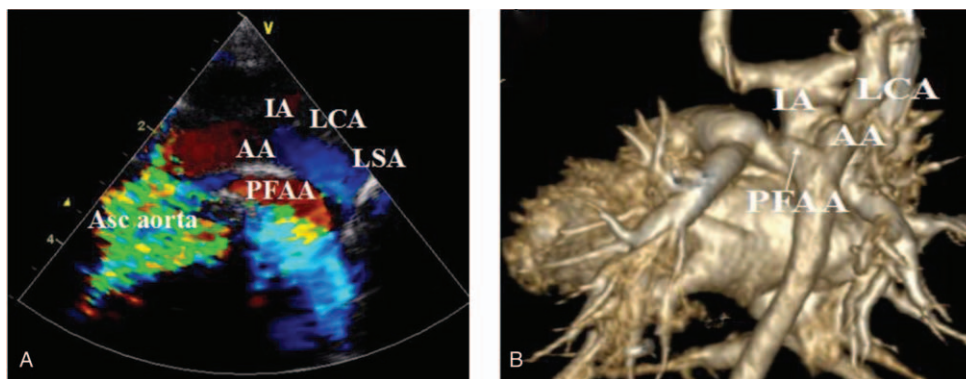
were in good clinical condition without residual arch stenosis or dilatation. Five patients were lost to follow-up after hospital discharge.

**4. Discussion**

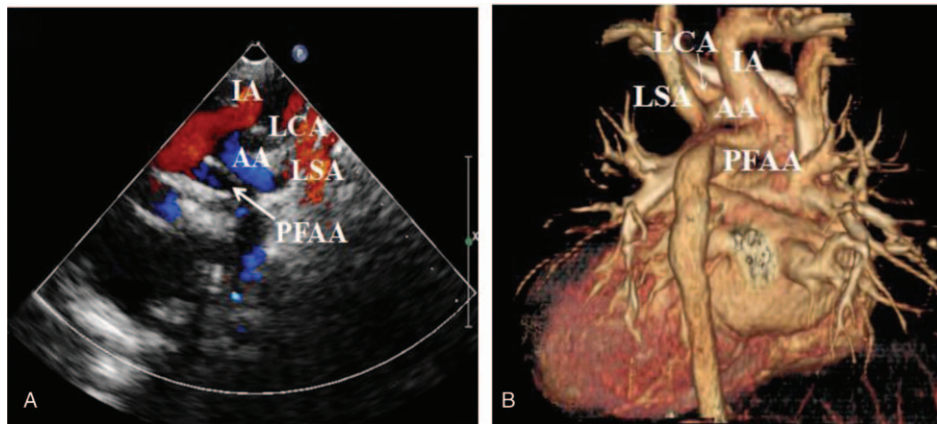
During normal embryonic development of the great arteries, the bilateral primitive fifth aortic arches are rudimentary vessels that degenerate at approximately the 4th week of embryonic life.<sup>[6]</sup> However, in rare cases, one or both fifth arch (es) persist in the process of arterial loop formation, creating the anomaly of PFAA.<sup>[9]</sup> Weinberg classified this type of malformation into 3 categories,<sup>[4]</sup> and Lloyd reviewed the new classification based on anatomical and physiological characteristics.<sup>[10]</sup> In our study, type B PFAA was the most common, using Weinberg classification.

PFAA is usually diagnosed during the neonatal or infant period, with only a few adult cases reported.<sup>[11]</sup> The oldest reported patient with PFAA was a 51-year-old female,<sup>[11]</sup> and the

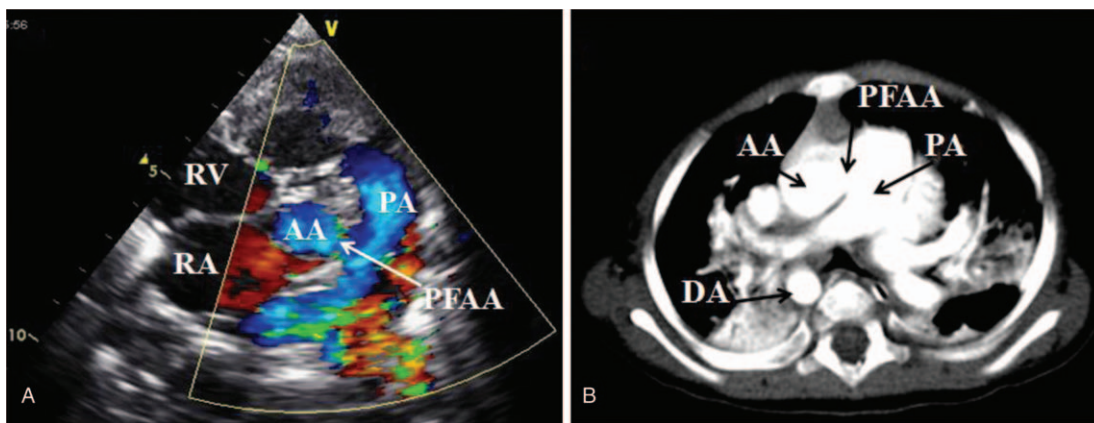
youngest was a fetus at 20 weeks of gestation.<sup>[12]</sup> Although Gerlis reported an incidence rate of 0.3% for PFAA,<sup>[13]</sup> the prevalence in our hospital was about 0.01% over the 5 year study period. This may be because some patients were asymptomatic or the characteristic clinical symptoms of PFAA were absent. The clinical symptoms and diagnosis of PFAA are strongly affected by associated CHD and its associated hemodynamic changes.<sup>[14]</sup> Such individuals are occasionally diagnosed because of obvious blood pressure differences between the arms and lower limbs, or incidentally because of unrelated disorders.<sup>[15]</sup> For example, although one patient (case 8, Weinberg type B) also had IAA (Type A), she survived until 11 years-of-age without serious hemodynamic symptoms and it was determined that PFAA was incidental to her severe nervous system disease. Usually there are severe clinical symptoms in infants or young children, even newborns, such as heart murmurs, cyanosis, pneumonia, bronchitis, and cough (the remaining nine cases listed in this paper).



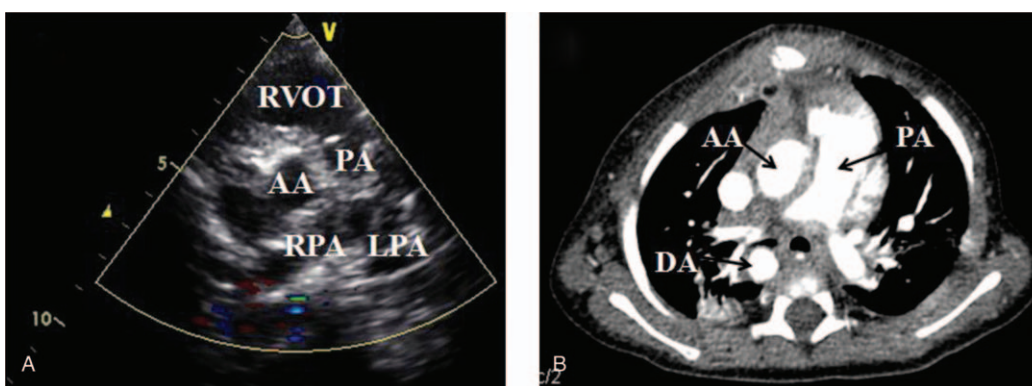
**Figure 1.** Case 2. A. Preoperative suprasternal views on echocardiography reveal Weinberg type A persistent fifth aortic arch (PFAA). Color doppler flow imaging (CDFI) shows the “double-lumen aortic arch”, the aortic arch is divided into superior and inferior channels, with the right innominate artery (RIA), left carotid artery (LCA) and left subclavian artery (LSA) arising from “the upper lumen”. The lower arch, which is of similar size, arises from the ascending aorta (Asc aorta) at the level of the RIA and joins the upper arch. The lower arch is a PFAA. The fourth arch and PFAA connect to the descending aorta together. AA = aortic atresia, Asc aorta = ascending aorta, IA = innominate artery. B. Surface volume rendering technique (SVR) also shows Weinberg type A PFAA with a right aortic arch and “double-lumen aortic arch”, the fifth arch originates from the ascending aorta (Asc aorta) and is parallel with the fourth arch (aortic arch). There is a tiny branching vessel originating from the distal ascending aorta connected to the proximal descending aorta, and the innominate artery (IA), left carotid artery (LCA), and left subclavian artery (LSA) originate from the aortic arch. The fourth arch and fifth arch connect to the descending aorta together. AA = aortic atresia, PFAA = persistent fifth aortic arch. White arrow = PFAA.



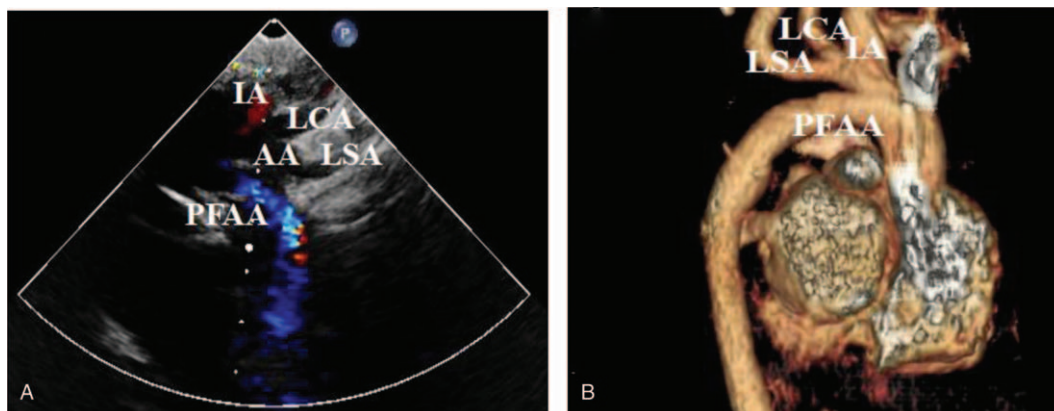
**Figure 2.** Case 4. A. Preoperative suprasternal views demonstrating Weinberg type B persistent fifth aortic arch (PFAA). Color doppler flow imaging (CDFI) shows that the fourth aortic arch is interrupted; the fifth arch originates from the ascending aorta and connects to the descending aorta, but with coarctation. The innominate artery (IA), left carotid artery (LCA), and left subclavian artery (LSA) originate from the aortic arch. AA = aortic atresia, PFAA = persistent fifth aortic arch. White arrow = LCA. B. Surface volume rendering technique (SVR) also demonstrates that the fourth aortic arch is interrupted, and the fifth arch originates from the ascending aorta and connects to the descending aorta with coarctation. The innominate artery (IA), left carotid artery (LCA), and left subclavian artery (LSA) originate from the aortic arch. There is abundant collateral circulation. AA = aortic atresia, PFAA = persistent fifth aortic arch. White arrow = LCA.



**Figure 3.** Case 10. A. A left parasternal view demonstrates Weinberg type C persistent fifth aortic arch (PFAA). Color doppler flow imaging (CDFI) shows that an abnormal shunt vessel connects the ascending aorta to the pulmonary trunk with the right aortic arch. AA = aortic atresia, PA = pulmonary atresia, RA = right atrium, RV = right ventricle. White arrow = PFAA. B. The CTA reveals an abnormal shunt vessel connecting the ascending aorta to the pulmonary trunk. Bilateral pneumonia is also present. AA = aortic atresia, DA = descending aorta, PA = pulmonary atresia, PFAA = persistent fifth aortic arch. Black arrows = (DA, AA, PFAA, PA), respectively.



**Figure 4.** Case 2 following surgical correction. A. Left parasternal views show that the abnormal lumen is resected, and the descending aortic coarctation is augmented. AA = aortic atresia, LPA = left pulmonary atresia, PA = pulmonary atresia, RPA = right pulmonary atresia, RVOT = right ventricular outflow tract. B. A cross section of computed tomography also shows resection of the abnormal lumen. The bilateral pneumonia is resolved. AA = aortic atresia, DA = descending aorta, PA = pulmonary atresia. Black arrows = (DA, AA, PA), respectively.



**Figure 5.** Case 4 following surgical intervention. A. Suprasternal views show Weinberg type B persistent fifth aortic arch (PFAA) with the color doppler flow imaging (CDFI) demonstrating augmentation of the coarctation, which is no longer restrictive. AA=aortic atresia, IA=innominate artery, LCA=left carotid artery, LSA=left subclavian artery, PFAA=persistent fifth aortic arch. B. SVR further demonstrates successful repair. IA=innominate artery, LCA=left carotid artery, LSA=left subclavian artery, PFAA=persistent fifth aortic arch.

PFAA is rarely an isolated anomaly, and the arch malformation is usually associated with other CHD, such as IAA, ventricular septal defect (VSD), coarctation of aorta (CoA), patent ductus arteriosus (PDA), tetralogy of fallot (TOF), tricuspid atresia (TA), transposition of the great arteries (TGA), aortic atresia (AA), pulmonary atresia (PA), or descending aortic aneurysm.<sup>[16–20]</sup> Therefore, diagnoses of PFAA may be difficult or delayed. Case 10 (type C) was initially incorrectly diagnosed as APW. Case 4 (type B) was misdiagnosed as aortic stenosis, coarctation, and left pulmonary arterial stenosis. Case 2 was misdiagnosed as coarctation with PDA. The diagnoses of these three patients were ultimately correctly defined at the time of surgery. It is noteworthy that Weinberg type A PFAA may be misdiagnosed as double aortic arch (DAA),<sup>[21]</sup> Weinberg type B was usually misdiagnosed as IAA or CoA, and Weinberg type C may be diagnosed as APW. The evidence for PFAA diagnosis was the detection of an abnormal vascular connection on imaging.

In early years, DSA with CAG was the gold standard for the diagnosis of PFAA.<sup>[7]</sup> Currently, echo can provide a noninvasive, low-cost, real-time evaluation of dynamic anatomical structures, hemodynamics, and cardiac functions, including vascular stenosis and atresia, valve prolapse, and pulmonary hypertension, and malformations such as PDA, IAA, and DAA.<sup>[8]</sup> Echo is the first-line diagnostic tool for PFAA, even in fetuses. For example, Bhatla<sup>[11]</sup> reported a fetus at 20 weeks of gestation diagnosed with PFAA by echocardiography. Echo is used to assess preoperative anatomy and postoperative results; however, both missed and incorrect diagnoses may occur because of complex extracardiac anatomy. Among our cases, the accuracy of echo for PFAA diagnosis was 62.5%.

We found that CTA is more accurate than echo for showing vascular origination, branching, aortic arch complexities, surrounding blood vessels, and their spatial structure.<sup>[22]</sup> However, CTA is less helpful for intravascular blood flow assessment. Magnetic resonance angiography (MRA) is a potential supplementary approach, but it is more expensive and is influenced by heart rate. Zhu et al<sup>[23]</sup> reported that enhanced MRA (CE-MRA) can provide more specificity than angiography in diagnosing congenital arch deformity.

Four of our patients underwent successful repair with extracorporeal circulation through a median sternotomy,

including ligation of the fifth arch, patch enlargement of the CoA, and/or patch augmentation of the PFAA. Our first patient to receive surgery had a persistent fifth arch with a right aortic arch and PDA, a little coarctation at its junction with an ascending aorta, and unaffected hemodynamics. The surgery mainly focused on ligation of the PDA, and patch enlargement of the CoA was sufficient. Our second and third cases both involved severe coarctation at its junction with an ascending aorta; the persistence of the fifth aortic arch in aorta obstruction may be an embryonic compensatory measure. Thus, in many such cases, the persistent arch is found to be beneficial, acting as an extra systemic-to-systemic and systemic-to-pulmonary shunt, which helps to maintain favorable hemodynamics. The required surgery patched the enlargement of the coarctation and augmented the PFAA to recover the hemodynamics. Our fourth patient was misdiagnosed as APW before surgery as the echo was difficult to interpret. Due to the existence of the PFAA, the hemodynamics showed a large amount of left-to-right shunt. The patient underwent repair of the fifth arch by ligation. All four patients recovered well, with a maximum follow-up time of 5 years.

As noted in the literature, some patients with PFAA and no clinical symptoms may not require intervention. However, most PFAA patients do require intervention due to complex CHD, including treatment with prostaglandin E1 (PGE1),<sup>[24]</sup> although Carroll et al<sup>[25]</sup> reported PGE1 was ineffective with PFAA. Our four patients were given PGE1 routinely before and after surgery. For most PFAA patients, the main surgical needs are resection of the narrow part or ligation of the fifth arch to reconstruct the fourth arch,<sup>[26]</sup> patch enlargement of the arch stenosis,<sup>[27]</sup> and patch augmentation of the fifth arch to replace the fourth arch,<sup>[28]</sup> sometimes using Gore-Tex tube grafts<sup>[29]</sup> or stenting.<sup>[30]</sup> The surgical method of choice depends on the associated CHD and the hemodynamic change. Uysal et al<sup>[31]</sup> recently reported the first case of PFAA with CoA treated with balloon angioplasty. Balloon angioplasty may be a better way to repair PFAA in the future.

There were several limitations in this study. First, we included a relatively small number of PFAA cases because of the rarity of this disease. Without doubt, more information would be obtained with a larger sample size. Second, due to the high incidence of lost follow-up, we cannot report the long-term prognosis of the

patients in detail. Third, none of our 10 patients received an MRI examination, which may have presented more information. In future studies, stricter follow-up and MRI examinations should be considered.

In conclusion, our results suggest that PFAA has a low incidence and does not have a typical clinical manifestation because of its associated complex anomalies. Weinberg type B may be most common (60%) and is inevitably associated with IAA or CoA. PFAA may be diagnosed more accurately and promptly using echo with typical imaging features, and its diagnostic accuracy can be as high as 62.5%, or even higher. CTA can confirm or correct the diagnosis and show the origin, course, and type of deformity more clearly. Patients with PFAA usually receive surgery immediately after diagnosis, and the prognosis depends upon the severity of the associated anomalies.

### Author contributions

**Conceptualization:** Haiyan Yang, Xu Zhu, Xiaojuan Ji.

**Data curation:** Haiyan Yang, Xu Zhu, Chun Wu, Xiaojuan Ji.

**Formal analysis:** Haiyan Yang, Xiaojuan Ji.

**Supervision:** Xiaojuan Ji, Xiaodong Zhao.

**Writing-original draft:** Haiyan Yang.

### References

- [1] Congdon ED. Transformation of the aortic arch system during the development of the human embryo. *Contrib Embryol Carnegie Inst* 1922;14:47–110.
- [2] Praagh RV, Praagh SV. Persistent fifth arterial arch in man: congenital double-lumen aortic arch. *Am J Cardiol* 1969;242:279–82.
- [3] Izukawa T, Scott ME, Durrani F, et al. Persistent left fifth aortic arch in man. Report of two cases. *Br Heart J* 1973;35:1190–5.
- [4] Weinberg PM. Aortic arch anomalies. *J Cardiovasc Magn Reson* 2006;8:633–43.
- [5] Freedom RM, Yoo SJ, Mikailian H, et al. The natural and modified history of congenital heart disease 2007.
- [6] Blake HA, Manion WC. Thoracic arterial arch anomalies. *Circulation* 1962;26:251–65.
- [7] Zheng L, Cao YL, Wu RC, et al. Persistent fifth aortic arch stenosis associated with type A interruption of the aortic arch: a report of six cases. *Chin Med J* 2019;132:1482–4.
- [8] Bernheimer J, Friedberg M, Chan F, et al. Echocardiographic diagnosis of persistent fifth aortic arch. *Echocardiography* 2007;24:258–62.
- [9] Hanneman K, Newman B, Chan F. Congenital variants and anomalies of the aortic arch. *Radiographics* 2017;37:32–51.
- [10] Lloyd DFA, Ho SY, Pushparajah K, et al. Persistent fifth aortic arch: The great pretender in clinical practice. *Cardiol Young* 2017;28:1–7.
- [11] Inoue T, Morita K, Tanaka K, et al. Distal aortic arch aneurysm associated with persistent fifth aortic arch. *Ann Thorac Cardiovasc Surg* 2014;20:878–81.
- [12] Bhatla P, Chakravarti S, Axel L, et al. prenatal diagnosis of a persistent fifth aortic arch, pulmonary-to-systemic type: An unusual association with evolving aortic coarctation. *Echocardiography* 2015;32:875–7.
- [13] Gerlis LM, Ho SY, Anderson RH, et al. Persistent 5th aortic arch—a great pretender: three new covert cases. *Int J Cardiol* 1989;23:239–47.
- [14] Ohashi N, Sakamoto T, Kosuka Y, et al. Persistent fifth aortic arch with coarctation of aorta: report of a case. *Kyobu Geka* 2012;65:158–60.
- [15] Tang X, Wang L, Wu Q, et al. Persistent fifth aortic arch with interrupted aortic arch. *J Card Surg* 2015;30:284–7.
- [16] Kim SH, Choi ES, Cho S, et al. Persistent fifth aortic arch with coarctation. *Korean J Thorac Cardiovasc Surg* 2016;49:39–41.
- [17] Wei X, Wang F, Cao R, et al. True bovine aortic arch combined with coarctation of persistent fifth aortic arch in a child. *European Heart Journal - Cardiovascular Imaging* 2019.
- [18] Murugan MK, Gulati GS, Saxena A, et al. Multi-detector computed tomography (MDCT) in Persistent Fifth Aortic Arch (PFAA). *Heart Lung Circ* 2014;23:71–3.
- [19] Furtado AD, Manohar SR, Pradhan SK, et al. Persistent left fifth aortic arch with pentalogy of Fallot. *Thorac Cardiovasc Surg* 2012;60:233–5.
- [20] Tamayo-Espinosa T, Erdmenger-Orellana J, Becerra-Becerra R, et al. Persistent fifth aortic arch with patent ductus arteriosus. *Arch Cardiol Mex* 2015;85:161–3.
- [21] Linhares RR, Silva CE, Monaco CG, et al. Double lumen aortic arch or persistence of fifth aortic arch? Report of a case with no associated cardiac defects and literature review. *Echocardiography* 2011;28:143–5.
- [22] Schicchi N, Agliata G, Giovagnoni A. CT imaging of a rare case of persistent fifth aortic arch in newborn. *BJR Case Rep* 2016;2:20150048.
- [23] Zhu M, Zhong YM, Li YH, et al. Diagnosis of congenital obstructive aortic arch anomalies in Chinese children by contrast-enhanced magnetic resonance angiography. *J Cardiovasc Magn Reson* 2006;8:747–53.
- [24] Zartner P, Schneider M, Bein G. Prostaglandin E1 sensitive persistent fifth aortic arch type 2. *Heart* 2000;84:141.
- [25] Carroll SJ, Ferris A, Chen J, et al. Efficacy of Prostaglandin E1 in relieving obstruction in coarctation of a persistent fifth aortic arch without opening the ductus arteriosus. *Pediatr Cardiol* 2006;27:766–8.
- [26] Cetrano E, Polito A, Trezzi M, et al. Neonatal repair of persistent fifth aortic arch coarctation and interrupted fourth aortic arch. *Ann Thorac Surg* 2017;103:e475–7.
- [27] Tittel PI, Kaldararova M, Olejnik P, et al. Interruption of the fourth aortic arch and a persistent fifth aortic arch with coarctation: a rare morphological finding. *Kardiol Pol* 2015;73:131.
- [28] Subramanyan R, Sahayaraj A, Sekar P, et al. Persistent fifth aortic arch. *J Thorac Cardiovasc Surg* 2010;139:e117–8.
- [29] Konishi T, Iizima T, Onai K, et al. Persistent fifth aortic arch Complicated by coarctation of the aorta and aneurysm of the left subclavian artery (author's transl). *Nippon Kyobu Geka Gakkai Zasshi* 1981;29:1243–8.
- [30] Valderrama P, Álvarez T, Ballesteros F, et al. Coarctation of persistent fifth aortic arch with interrupted fourth arch: First pediatric report of stent intervention. *Rev Esp De Cardiol* 2016;69:337–8.
- [31] Uysal F, Bostan OM, Cil E. Coarctation of persistent 5th aortic arch: First report of catheter-based intervention. *Tex Heart Inst J* 2014;41:411–3.