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# Persistent fifth aortic arch: a comprehensive literature review

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Persistent fifth aortic arch (PFAA) is an extremely rare congenital cardiovascular anomaly resulting from the failure of the fifth aortic arch to degenerate during embryonic development; it is often associated with various other cardiovascular anomalies. Despite being first reported by Van Praagh in 1969, there have been only a few individual case reports. Owing to its rarity and lack of comprehensive understanding, PFAA is often misdiagnosed or missed diagnosed during clinical. Thus, this review aimed to summarise the embryonic development, pathological classification, imaging diagnosis, and clinical treatment of PFAA to improve its overall understanding, ultimately helping in accurate diagnosis and treatment.

## KEYWORDS

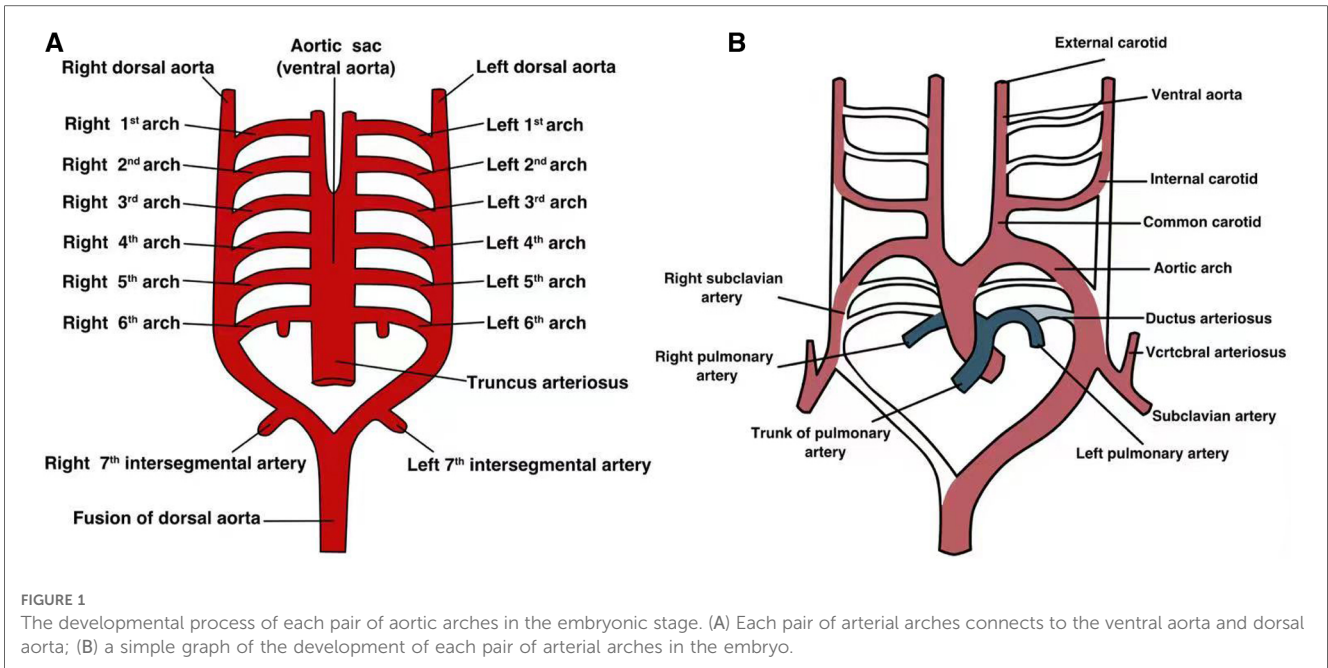
persistent fifth aortic arch, classification, diagnosis, imaging, treatment

## 1. Introduction

Persistent fifth aortic arch (PFAA) is widely recognised as a rare congenital cardiovascular malformation that results from the failure of the fifth aortic arch to degenerate during embryonic development (1). It was first reported by Van Praagh through autopsy in 1969 (2). PFAA has an extremely low incidence rate, with Gerlis (3) reporting an incidence of only 0.3%. Clinical symptoms of PFAA are nonspecific and mainly depend on the combination of other deformities and their haemodynamic changes. PFAA is usually confirmed during the neonatal or infant period, with only a few cases reported in adults (4). PFAA is earliest classified by the Weinberg classification (5). Subsequently, some new classifications have been reported according to the proximal and distal connections of the vessel and the direction of blood flow (6, 7). Due to the rarity of PFAA and the fact that only individual cases are reported, misdiagnosis and missed diagnosis are common in clinical practice (8–10). Thus, this review aimed to improve the comprehensive understanding of PFAA by summarising its embryonic development, pathological classification, imaging diagnosis, and clinical treatment to improve diagnostic accuracy and treatment.

## 2. Mechanisms of embryonic development

The aortic arch develops from the aortic sac. During normal embryonic development, six pairs of branchial arches originate from the pharyngeal arch and extend to the dorsal aorta (Figure 1). However, these arches do not always persist and undergo a sequence of orderly digestion and absorption processes between embryonic weeks 3–8 (11). The embryonic development of the arch is asymmetrical. Typically, the first and second branchial arches evolve into the transcranial artery (maxillary and hyoid arteries), whereas the third



branchial arch exists permanently, forming the left and right common carotid arteries and part of the internal carotid artery, with their roots connected to the aortic sac. The right fourth branchial arch connects to the right seventh intersegmental artery and then to the right third branchial arch, ultimately forming the brachiocephalic trunk. The left fourth branchial arch connects to the distal and proximal left dorsal aortas to form the horizontal aorta. The right sixth branchial arch forms the right pulmonary artery, whereas the left sixth branchial arch forms the ducts or arterial ductal ligament.

During normal embryonic development of the arterial system, the bilateral fifth branchial arch is immediately absorbed after its development. However, PFAA develops if any unilateral and/or

bilateral branchial arches fail to degenerate or completely degenerates and participates in vascular ring formation (12), resulting in hypoplastic blood vessels (13, 14).

### 3. Clinical classification

As PFAA can occur unilaterally or bilaterally, it can present as various clinical types. The Weinberg classification is commonly used in clinical practice to categorise PFAA based on different abnormal vascular connections (5) (Figure 2). Weinberg type A is characterised by a double-lumen aortic arch, where the upper arch is the fourth arch, and the lower arch is the fifth arch,

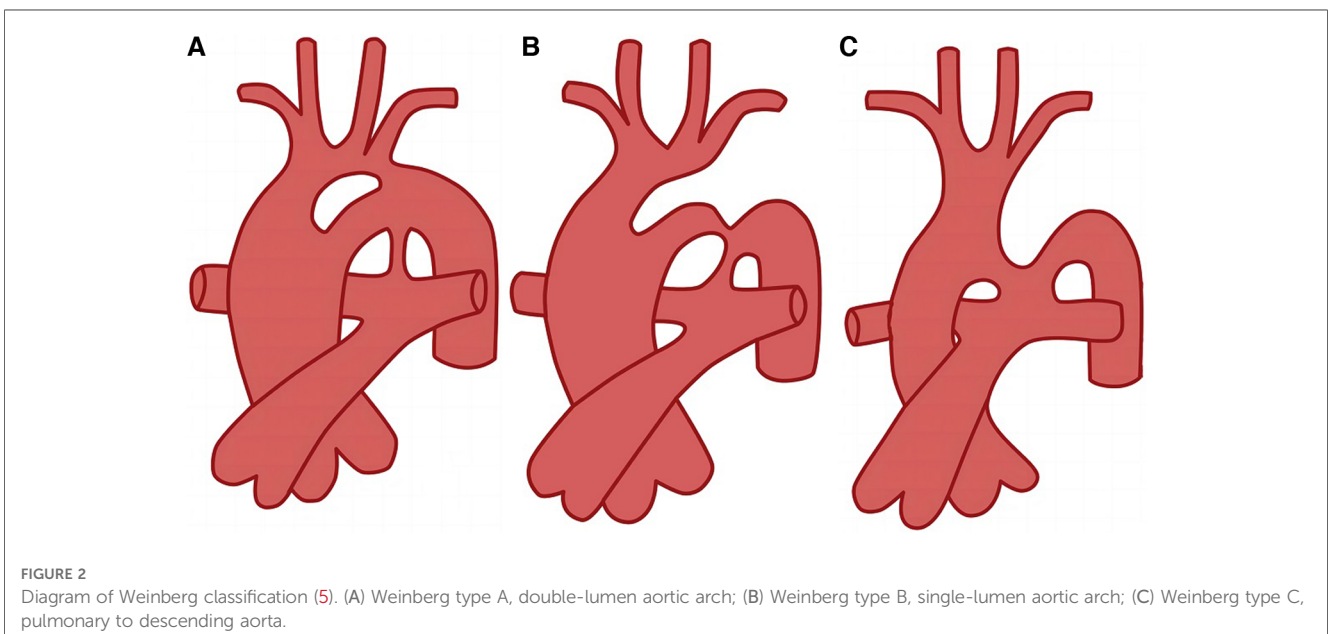


TABLE 1 Comparison of improved classification and Weinberg classification of PFAA.

Improved classification		Weinberg classification
<b>A systemic to systemic connection</b>		
A1 Double-lumen aortic arch (13, 14, 15–22)	<b>A</b>	Double lumen aortic arch
A2 Single-lumen aortic arch (8, 23–28)	<b>B</b>	The fifth arch remains, and the fourth arch is interruption
A3 Abnormal brachiocephalic artery origin (29)		
<b>B systemic to pulmonary connection</b>		
B1 With pulmonary obstruction (30–33)	<b>C</b>	The fifth arch originates from the proximal innominate artery of the ascending aorta and connects to the pulmonary via the sixth arch
B2 Without pulmonary obstruction (13, 30, 34–37)		
B3 With unrestricted systemic and pulmonary flows (9)		
<b>C pulmonary to systemic connection</b>		
With Aortic atresia (38)		
<b>D hybrid</b>		
Bilateral PFAA (39)		

PFAA, persistent fifth aortic arch; AAO, aorta ascendens.

known as PFAA. The right innominate, left carotid, and left subclavian arteries typically arise from the fourth aortic arch. Weinberg type B is characterised by a single-lumen aortic arch, with the fourth aortic arch being interrupted, and the fifth arch originating from the ascending aorta and connecting to the descending aorta. Weinberg type C is characterised by the fifth arch originating from the proximal innominate artery of the ascending aorta and connecting to the pulmonary artery (Table 1).

In the present study, we conducted a literature search and collected data on 104 reported cases of PFAA from 77articles. We accurately classified these cases according to the Weinberg classification, excluding cases with unclear descriptions of the images and arterial connections. Among the 104 cases, Weinberg type A was the most common type of PFAA, accounting for 40.4% (42/104) of the cases, followed by Weinberg type B, accounting for 38.5% (40/104) of the cases. Weinberg type C was the least common, accounting for 18.3% (19/104) of the cases; the remaining 3 cases could not be classified using the Weinberg classification.

Subsequently, as more cases of PFAA were reported and a deeper understanding of the condition was gained, many scholars began to

suggest that the Weinberg classification could not sufficiently cover the various types of PFAA. Therefore, they proposed an improved classification based on the anatomical origin and haemodynamic changes of PFAA (6, 7, 40). This improved classification divides PFAA into four types (Figure 3 and Table 1).

Table 1 shows that the A1 type of the improved classification is equivalent to Weinberg type A, as both are characterised by a double-lumen aortic arch. The A2 type of the improved classification is equivalent to Weinberg type B, which is a single-lumen aortic arch, with the fourth arterial arch occluded or interrupted and the fifth aortic arch serving as the arterial arch supplying blood between the aorta and descending aorta. The B type of the improved classification is equivalent to Weinberg type C, where PFAA is connected from the ascending aorta and terminates in the pulmonary artery. Table 2 shows that the modified classification could cover all of our cases, including the 3 cases that were not defined by Weinberg classification, demonstrating that the relationship between the improved and Weinberg classifications is not a purely one-to-one correspondence. The improved classification covers the Weinberg

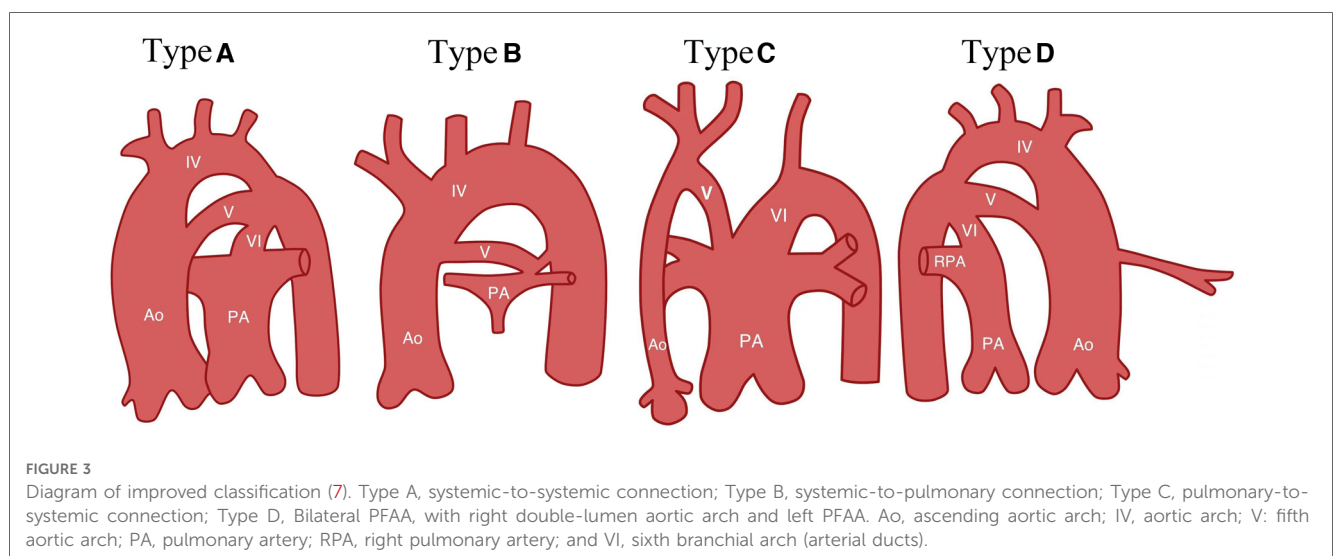


TABLE 2 Characteristics of the 104 cases of PFAA reported in the literature.

Age (case)	Sex (case)	Weinberg Classification (case)	Improved classification (case)	Diagnosis (case)	Treatment (case)
≤10 years (88)	Male (59)	Type A (42)	A1 (42)	US (18)	Surgical repair (52)
		Type B (40)	A2 (40)	CTA (39)	PGE1 + surgery (12)
	Female (39)	Type C (19)	A3 (1)	CAG (23)	Balloon Dilatation (4)
>10 years (12)	Unclear (7)	Cannot define (3)	B1 (5)	MRA (5)	Stent implantation (5)
			B2 (12)	Surgery (10)	No intervention (8)
			B3 (2)		PGE1 (2)
			C(1)	Post-mortem (9)	Drug therapy (1)
Unclear (4)			D(1)		Balloon + Stent (1)
					Unclear (19)

The numbers in parentheses represent the number of cases.

classification and is relatively more comprehensive and specific. Therefore, the improved classification is a supplement and derivative based on the Weinberg classification, which can more completely summarise different common and rare types of PFAA. The Weinberg classification only covers the common PFAA types corresponding to the improved classification.

## 4. Clinical symptoms and complications

The clinical symptoms of PFAA are nonspecific and mainly depend on the combination of other deformities and their haemodynamic changes (41). Patients usually do not exhibit any obvious clinical symptoms when PFAA is not associated with other vascular malformations, and they may even be asymptomatic. This condition is more common in Weinberg type A patients, who are often diagnosed during physical or accidental examinations (12, 19, 42).

Patients with congenital heart disease, such as aortic atresia, interruption of aortic arch (IAA), coarctation of the aorta, patent ductus arteriosus (PDA), pulmonary atresia (PA), tricuspid atresia, atrial septal defect, and ventricular septal defect (VSD), may experience a range of symptoms, including heart murmur, cyanosis, cough, shortness of breath, pneumonia, and bronchitis. For instance, Li et al. (43) reported a 50-day-old boy with Weinberg type B PFAA who had IAA and PDA and presented with shortness of breath and abnormal heart murmur. Interestingly, for some patients with PFAA who are associated with PA, the presence of PFAA can be beneficial, as the fifth arch serves as an important system for pulmonary shunt (31, 33). However, severe cases may result in congestive heart failure, cardiogenic shock, and multiorgan failure (35, 44). For instance, Sinha et al. (33) reported the case of a 2-month-old boy with Weinberg type C PFAA who had a VSD and PA. The patient presented with central cyanosis, poor feeding, and failure to thrive.

Furthermore, some patients with PFAA may exhibit distinct facial and bodily abnormalities in clinical practice, which may be linked to genetic and chromosomal abnormalities, such as 22q11.2 chromosome deletion (16, 20, 45, 46), PHACE (25), and Cornelia de Lange syndromes (CdLS) (47). Based on our literature review, six cases of PFAA were associated with 22q11.2 chromosome deletion syndrome, and they primarily presented

with scoliosis (48), palate abnormalities, facial deformities, hypocalcaemia (46), T-cell-mediated immune deficiency (49), and mild-to-moderate growth retardation. Growth retardation and poor feeding were the most common symptoms. In addition, two PFAA patients were found to have CdLS (14, 50) and were classified as Weinberg type A. CdLS is a genetic syndrome typically characterised by intellectual disability, distinct facial features, upper limb abnormalities, and atypical growth (51). In contrast, only one PFAA patient has been reported to have PHACE syndrome. The patient was classified as Weinberg type B and was associated with aneurysmal dilatation of the fourth aortic arch (25). The patient exhibited typical clinical manifestations of PHACE syndrome, such as frontotemporal cutaneous haemangioma, posterior fossa malformation, arterial abnormality, cardiac defects, and ocular abnormalities (52). Furthermore, the repeats of 22q11.2 (20) and 9q (47) are associated with PFAA. Therefore, clinicians should consider the possibility of PFAA in cases where a patient exhibits distinct facial features and abnormal cardiac haemodynamics.

## 5. Diagnostic imaging

Imaging technology plays a crucial role in the diagnosis of PFAA. Echocardiography is a non-invasive and cost-effective imaging method that allows real-time evaluation of dynamic anatomical structures, haemodynamics, and cardiac functions. It is recognised as the first-line diagnostic tool for PFAA, even in fetuses, owing to its ability to provide detailed information. Furthermore, echocardiography can be used to assess the systolic function of blood vessels preoperatively and postoperatively. However, complex extracardiac anatomy may lead to missed or incorrect diagnoses (8, 10). Computed tomography angiography (CTA) is considered to be more accurate than ultrasound in showing vascular origination, branching, aortic arch complexities, surrounding blood vessels, and their spatial structure. However, CTA is less effective in assessing intravascular blood flow. Magnetic resonance angiography (MRA) provides a clearer anatomical structure of the arch of aorta and its relationship with surrounding structures, thus making it a useful diagnostic tool before surgery or cardiac catheterisation. MRA has gradually replaced cardiac catheterisation as an effective non-invasive method in clinical practice. However, MRA application in clinical

practice is limited by its high cost. Digital subtraction angiography is considered the gold standard for the diagnosis of PFAA (53); however, it involves radiation exposure and intervention (54).

Of the 104 cases we reviewed, almost all patients underwent echocardiography. According to our statistical analysis (Table 2), 18(17.3%) patients were initially diagnosed using echocardiography, 39(37.5%) patients were initially diagnosed using CTA, 23(22.1%) patients were initially diagnosed using cardiac catheterisation, 5(4.8%) patients were confirmed using MRA (29, 30, 38, 55, 56), 10(9.6%) patients were diagnosed during the operation, and 9(8.7%) patients were confirmed post-mortem (2–4, 9, 31, 57). The results indicate that various imaging techniques have great diagnostic values in identifying cardiovascular malformations. Echocardiography can serve as the first-line diagnostic tool for PFAA, and when combined with CTA and/or MRA, it can provide a more accurate evaluation of the pathological anatomy, clinical type, and haemodynamics of PFAA.

## 6. Treatment of PFAA

Patients with PFAA and no clinical symptoms may not require intervention. However, when PFAA is associated with other cardiovascular malformations caused severe haemodynamic change, surgical treatment and prostaglandin E1 (PGE1) are often used to manage severe clinical symptoms in infancy (28). PGE1 is used to dilate blood vessels, reduce blood pressure, and prevent platelet aggregation (58). The surgical procedure typically involves resection of the narrow part or ligation of the fifth arch to reconstruct the fourth arch, patch enlargement of the arch stenosis, and patch augmentation of the fifth arch to replace the fourth arch, sometimes using the Gore-Tex tube grafts or stenting (8, 47, 59–61).

Of the 104 cases, 64 have undergone surgery and six patients underwent resection of the stenotic segment of the fifth arch and end-to-end anastomosis of the fifth arch with the descending aorta. In the present review, 14 patients received PGE1 treatment and 12 of them were subsequently treated surgically. Among the 14 patients who received PGE1 treatment (8, 38), except for the two patients who showed ineffective for PEG1 treatment, 12 patients showed good results in dilating the narrowed blood vessels (3, 15, 21, 23, 28, 30, 34, 58, 62–65). In our reviewed 104 cases, five patients underwent interventional balloon dilation; among them, two patients experienced long-term restenosis and

required secondary balloon dilation or surgery (66, 67). Six patients received stent implants, and follow-up results showed normal ventricular ejection function and no definitive stenosis (24, 42, 47, 66, 68, 69).

## 7. Conclusion

PFAA is a rare congenital cardiovascular malformation typically reported as individual case studies. The clinical symptoms of PFAA are nonspecific and mainly depend on its complications. Due to the inadequate understanding of PFAA, patients with this condition are often misdiagnosed or missed diagnosed. In this review, we collected literature on reported cases of PFAA and summarised its embryonic development, pathological classification, imaging diagnosis, and clinical treatment. We aimed to provide a comprehensive understanding of PFAA and improve its diagnosis and treatment.

## Author contributions

YH and LC conceived the review. SH performed the literature review and the first draft. GZ contributed to the study's conception. XD drew the diagrams. SH and HY worked on the revisions. All authors contributed to the article and approved the submitted version.

## Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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