Review Article:

**Pseudocoarctation of the arch and the abdominal aorta: A review.**


1. MBBS, MD, DM. *Primary and Corresponding author:* Assistant Professor, Dept. of Cardiology, JSS Hospital, JSSAHER, Mysuru, Karnataka, India, 570004.
2. MBBS, PGDCC. Clinical Cardiologist, Dept. of Cardiology, JSS Hospital, JSSAHER, Mysuru, Karnataka, India, 570004. Email: gurudmcardio@gmail.com

Email: docpk77@gmail.com
(Formerly Assistant Prof, Dept of Cardiology, JSSMC Hospital, Mysuru 04).

4. MBBS, MD (Anatomy). Assistant Professor, Dept of Anatomy, JSSMC, JSSAHER, Mysuru-5700015

Email: nb.pushpa@gmail.com

5. MBBS, MD (Radiology). Associate Professor, Dept. of Radiology, JSS Hospital, JSSAHER, Mysuru, Karnataka, India, 570004. Email: vikrampatil@jssuni.edu.in

**Abstract:** Pseudocoarctation of the arch of the aorta is a very rare congenital anomaly, occurring in isolation or with other congenital heart diseases. The anatomical basis of the condition is linked to an elongated, redundant aorta which may affect the arch, and very rarely the abdominal aorta giving rise to kink and buckling without causing any significant functional stenosis. It should be carefully differentiated from the more common true coarctation of the aorta. Although asymptomatic in the majority, few patients can have nonspecific symptoms and or complications due to aneurysm formation, dissection, or rupture of the aorta. Hence Pseudocoarctation should be closely followed for the onset of symptoms or possible complications. In the absence of recommendations, no specific therapy is indicated in asymptomatic patients, although symptoms and complications warrant definitive treatment. In this article, we are discussing embryology, anatomical basis, signs, symptoms, complications, treatment options, and follow-up of patients with pseudocoarctation of the aorta.

**Conclusion:** Pseudocoarctation of the arch and abdominal aorta is an extremely rare congenital anomaly occurring in isolation or associated with other congenital heart diseases. No clinical features are specific to this condition and often diagnosed incidentally. Although, in the absence of symptoms no therapy is required, the occurrence of symptoms and complications guide definitive therapy. As the natural history of the disease is unknown, the condition when diagnosed should be closely followed up for the occurrence of any complications such as aneurysm formation, dissection, or rupture of the aorta.

**Abbreviations:** CoA: Coarctation of Aorta,

**Keywords:** pseudocoarctation; coarctation; aneurysm; dissection; kinking.
1.1. Introduction:
Aortic pseudocoarctation is a very rare congenital anomaly affecting either the aortic arch or the abdominal aorta and can coexist with other congenital heart diseases. After the first description of what is currently called ‘pseudocoarctation’ of the thoracic aorta by Rosier and White in 1931, [1] a few cases and case series have been reported. As reported by Sujatha Singh et al, aortic pseudocoarctation has a slight male preponderance and presents at an average age of 43 years [2] and can be associated with other congenital heart diseases such as bicuspid aortic valve, aortic and subaortic stenosis, patent ductus arteriosus, ventricular septal defect, single ventricle, atrial septal defect, and anomalies of the branches of the aortic arch as explained by I Steinberg. [3] Pseudocoarctation has been reported as a manifestation of Takayasu’s arteritis and in patients with Turner’s syndrome. [4] It may affect either the arch of the aorta or very rarely the abdominal aorta. The incidence of the thoracic aortic pseudocoarctation is unknown, while that of abdominal aortic pseudocoarctation is said to be one in every 200 coarctations of the thoracic aorta. [5] Although the exact relationship between true coarctation and pseudocoarctation is unclear as of now, they are however considered distinct entities by most, and some believe that they are variations of the same abnormality. [6,7] Clinically they are differentiated by the following features: [5,6,8]

- Presence of a high aortic arch (may arise higher than clavicle)
- Elongated, redundant aortic arch with a kink and buckling
- Minimal or no luminal narrowing of the aorta or obstruction to the flow of blood (no functional stenosis)
- Minimal or no pressure gradient across the involved segment, usually less than 25mmHg
- Absence of collateral arteries, rib notching, and features of left ventricular hypertrophy.

Abdominal pseudocoarctation is similar in form to the thoracic aortic pseudocoarctation, with marked redundancy of a segment of the aorta with no functional stenosis. The redundant tortuous segment does not have collateral circulation. [9]

In this article, we are discussing embryology, anatomical basis, symptoms and signs, possible complications, treatment options, and follow-up of patients with this very rare congenital anomaly. This review was undertaken given the ambiguity surrounding this rare congenital anomaly and to obtain clarity regarding its diagnosis and management from the available literature.

1.2. Illustrative case narration:
Eighteen years old, male, patient with no known comorbidities, significant family history, or prior hospitalization, presented with exertional breathlessness NYHA (New York Heart Association functional class) class II, with intermittent upper backache unrelated to the exertion of two weeks duration. Clinical examination revealed a pulse rate of 84 bpm, and blood pressure of 134/86 mmHg, peripheral arterial oxygen saturation was 98% on room air. Peripheral pulses in all four limbs were equally felt with no significant discrepancy in the recorded blood pressures. ECG (Electrocardiography) was unremarkable. A 2-D ECHO (2-dimensional Echocardiography) showed CHD (congenital heart disease) with features suggestive of CoA (coarctation of the aorta). Peak doppler gradient was 25mmHg across the affected segment with normal LV (left ventricle) function. CT (computerized tomography) aortogram done revealed mild narrowing of the distal part of the arch of the aorta, with elongated, redundant, buckled aortic arch. Proximal descending aorta immediately distal to the buckled segment showed mild prominence in caliber, and the features were consistent
with pseudocoarctation of the arch of the aorta. Other abnormalities evident on CT were the “variant origin” of the left vertebral artery from the arch of the aorta, fusion and segmentation anomalies involving the upper thoracic vertebra, and bilateral hypoplastic first ribs. Investigations such as routine blood counts, renal, liver, thyroid function tests, CXR (chest x-ray) PA (posteroanterior) view, ultrasound abdomen, and pelvis were within normal limits.

Image 1: 2-D ECHO images in (a) apical four-chamber (A4C) view showing normal chambers (LV-left ventricle, RV-right ventricle, LA-left atrium, and RA-right atrium) and absence of left ventricular hypertrophy (white hollow arrows), (b) Suprasternal view showing features of coarctation of the arch of the aorta with mild dilatation of the descending aortic segment (AA-ascending aorta, PA-pulmonary artery, P.arch-proximal arch, D.arch-distal arch, p.desc.aorta-proximal portion of the descending aorta) (c) suprasternal view showing color doppler across the lesion demonstrating mild turbulence (AA-ascending aorta, DA-descending aorta, PA-pulmonary artery, white hollow star-shows ‘coarcted’ segment, white hollow arrow shows mild turbulence across the aortic arch), and (d) in the suprasternal view continuous wave doppler showing a gradient of 25mmHg.
Image 2. CT-image: 3-dimensional reconstruction of the ascending, arch, and descending aorta. The distal portion of the arch showing slightly reduced caliber, with redundancy and buckling. The descending aorta immediately distal to the buckled segment shows mild prominence in caliber, (a) Yellow line-Proximal arch measuring 2.6 cms, Light blue line-caliber of the kinked segment 2.4cms, and white line- the caliber of the distal portion of the aortic arch, 2.7cms. The pink arrow shows the kinked segment of the aortic arch, (b) the deep blue line shows the mildly prominent proximal portion of the descending aorta, 2.8cms, just distal to the distal portion of the kinked aortic arch. The pink arrows show the kinked, buckled aortic arch.

Video Clips 1 2D ECHO.
Video Clips 2 CT Aortogram.

2. Discussion:
2.1. Aortic pseudocoarctation in literature:
In 1931 Rosler and White published an article describing unusual variations of the roentgen shadow of the elongated thoracic aorta, which is probably the first documentation of what is currently known as pseudocoarctation. [1] Subsequently, in 1951, Dotter and Steinberg described two patients' aortic anomalies and commented on their resemblance to the true coarctation in their report on anomalies of the arch of the aorta. [10] In the same year, Robb [11] reported a case while Souders et al. [12] published data on three patients with mediastinal tumors but were later diagnosed as having ‘subclinical coarctation’. In 1952, Dotter and Steinberg reported another case of aortic deformity and proposed the term ‘pseudocoarctation’. [13] In 1955 DiGuglielmo and Guttadauro described two patients with a similar aortic arch deformity which they called ‘kinking of the aorta’. [14] In 1958 Stevens described a similar case and called it ‘buckling of the arch of aorta’. [15] However, currently, the term “pseudocoarctation” is being used to describe this form of aortic deformity as suggested by Dotter and Steinberg in 1952. [13] Subsequently, few case reports and series have been reported in the literature enumerating differing manifestations, complications, and surgical treatment options.

2.2. Anatomical types:
Although there is no formal classification there are two reported types of pseudocoarctation in the literature based on the segment of the aorta involved. In the more common type, the pseudocoarctation may affect the arch or thoracic aorta, and in the relatively rarer type, the abdominal aorta. In both types, the possible mechanism leading to pseudocoarctation remains the same, i.e., an elongated, redundant portion of the aortic segment due to incomplete or failure of compression of the embryologic segments, giving rise to kinking or buckling without causing functional stenosis.
2.3. Embryology:

2.3.1. The thoracic aortic pseudocoarctation:
Pseudocoarctation appears to be closely related to true coarctation and differs only in the absence of significant narrowing and gradient across the involved segment. The arch of the aorta develops from multiple structures. The portion of the arch proximal to the brachiocephalic trunk arises directly from the aortic sac. The medial area of the arch, between the brachiocephalic trunk and the left common carotid artery, arises from the left fourth aortic arch. The portion of the arch distal to the left common carotid artery arises from the dorsal aorta.[3] In organogenesis some arterial segments elongate while others become greatly contracted; one of these changes occurs in the seventh dorsal intersegmental arteries when they undergo a cephalad shift and become the first portions of the subclavian arteries. When the heart descends into the thorax, the subclavian arteries remain relatively far cephalad; there is associated compression of the third to seventh segments of the dorsal aortic roots and the left fourth aortic arch segments. [3] It is widely believed that failure of compression of these segments results in an abnormally long aortic arch that twists at the point of insertion of the ligamentum arteriosum. This would explain the kinking of the aortic isthmus, that is, the segment between the left subclavian artery and the ductus arteriosus, which is the primary area of involvement in all reported cases of pseudocoarctation thus far. [16] Another explanation attributes the thoracic aortic pseudocoarctation to a short, taut, ligamentum arteriosum, or patent ductus arteriosus. This has been demonstrated in some of the cases, by dividing the adjacent patent ductus arteriosus which relieves pseudocoarctation. [3] However, it is not clear to what extent each of the described mechanisms plays a role in causing pseudocoarctation. The current understanding is one or both mechanisms may be involved. Son JS and associates reported an exceptional association of a pseudoacoarctation and the anomalous origin of the left vertebral artery directly from the aortic arch. [17] Embryologically, the aberrant origin of the left vertebral artery directly from the aortic arch is due to the persistence of the 8th intersegmental artery. [18] Our patient also had the origin of the left vertebral artery from the aortic arch as a “Variant anatomy”.

2.3.2. The abdominal aortic pseudocoarctation:
Said to have been first described by Quain [19] in 1847 as ‘partial coarctation’ which is currently termed as pseudocoarctation. It is generally localized between the superior mesenteric and renal arteries. [5] The abdominal aorta normally develops from the paired dorsal aortae by a process of fusion of the dorsal aortae during fetal development. The abdominal aortic pseudocoarctation results presumably due to an irregular fusion of the two dorsal aortas, with obliteration and loss of lumen in one of them. [20] This disordered fusion is proposed as the cause of a range of rare vascular anomalies in this region of the body, from abdominal aortic coarctation or hypoplasia to dual-channel abdominal aorta, aortic interruption, and pseudocoarctation. [21] We could only find around 7 reported cases/articles on abdominal aortic pseudocoarctation to date on PubMed search. The dilatation of the descending aorta just distal to the pseudocoarctation is probably due to the same pressure-flow relationships produced by hydraulic forces as described by Holman and Robicsek et al. [22, 23] However, in pseudocoarctation there is no significant stenosis and pressure gradient across the involved segment. Hence I. Steinberg concludes that the presence of post-stenotic dilatation distal to the lesion may suggest a malformation which is probably an associated anomaly rather than a resulting lesion. [24] Further I. Steinberg and associates in their article argue, since there is no stenosis in pseudocoarctation of the aorta, the term “post constrictive” dilatation is preferable to “post stenotic” dilatation and indeed would apply to all degrees of localized narrowing of the aorta, with or without a pressure gradient. As the embryologic explanation for true coarctation of the aorta becomes clear, so will it be for pseudocoarctation. [3]
2.4.1. Signs and symptoms of thoracic aortic pseudocoarctation:
Patients with isolated pseudocoarctation in the absence of associated congenital anomalies and complications are often asymptomatic and may be diagnosed incidentally on evaluation for other diseases. There are no clinical features specific to pseudocoarctation. However, patients may occasionally come with resistant hypertension, exertional breathlessness, dysphagia, chest discomfort, or back pain, which often indicate complications. Associated conditions such as aortic dissection, aortic insufficiency, and mitral valve prolapse may be present, and accordingly, various cardiac murmurs may be heard along the sternal borders, the base of the neck, or interscapular region. Wann LS et al. reported a case of pseudocoarctation presenting with hypotension and haemothorax due to spontaneous rupture of an associated aortic aneurysm. Recognition of this condition is important, as it may be mistaken for true coarctation, aneurysm, or mediastinal neoplasm.

2.4.2. Signs and symptoms of abdominal aortic pseudocoarctation:
Roya Etemad-Rezai et al. in 2009 reported a patient with Waardenburg syndrome presenting with chest and back pain, who subsequently diagnosed to be having associated abdominal aortic pseudocoarctation. Prabhakar M in 1972 reported a case of abdominal aortic pseudocoarctation with an x-ray abdomen showing calcified lymph nodes and calcifications on the left side suggestive of an aortic aneurysm. In the same case, they described loud abdominal bruit. Similar to thoracic aortic pseudocoarctation, no symptoms or signs are specific to this condition and the underlying abnormality is often diagnosed during routine evaluation for other illnesses.

2.5 Investigations:
In the case of thoracic pseudocoarctation, an x-ray of the chest may show abnormalities such as cardiomegaly (LV type), mediastinal widening or mass, double density in the left cardiac border, soft tissue density in the left superior mediastinum overlying aortic knob, etc depending on the morphology of the involved segment. Israel Steinberg and associates in their series of ten cases published in 1969, described cardiomegaly (LV type), the reversed figure “3” or the “E” sign in the esophagogram, and the “3” sign in the descending aorta. However, unlike true coarctation, even in the adult, there is no rib notching from collateral circulation suggesting no significant functional stenosis. In the case of abdominal aortic pseudocoarctation, a chest x-ray may be largely unremarkable. There may be calcification if there is an associated aneurysm in the x-ray abdomen. Nevertheless, an x-ray can not distinguish between a mild true coarctation from that of pseudocoarctation, and due to its low sensitivity and specificity x-ray is of limited value in establishing the diagnosis.

Two-dimensional Echocardiography is an invaluable tool in suspecting and diagnosing coarctation, besides identifying various associated LV hypertrophy, the gradient across the segment, and congenital heart lesions. However, a definitive diagnosis of pseudocoarctation by ECHO alone may not be feasible requiring additional investigative tools such as catheterization, CT, or MR angiography. Cardiac catheterization and angiography provide a definitive diagnosis of this condition. Besides delineating the lesion, the pressure gradient across the involved segment can definitively identify pseudocoarctation from true coarctation. Several investigators have used CT angiography and aortogram to identify and study morphology, focal deformity, elongation, kinking, stenosis, post stenotic dilatation, associated dissection, calcification, and collateral arteries if any. 3 D reconstructive images can identify the exact morphology, although the severity and functional significance of the stenosis can not be described.

Magnetic resonance angiography is a useful and powerful supplemental imaging test, with distinct diagnostic advantages for noninvasive assessment of pseudocoarctation. Besides
providing consistently excellent assessments of pseudocoarctation, the absence of radiation and the ability to appreciate the three-dimensional aspects of the abnormality are added advantages of the technique. Additionally, magnetic resonance imaging allows the assessment of adjacent structures that may also have abnormalities. [8]

2.6 Differential diagnosis:
In the case of thoracic aortic pseudocoarctation, it should be differentiated from true coarctation which is possible by the fact that there is no functional stenosis, no associated collateral arteries and rib notching, minimal or no significant gradient (<25mmHg) across the lesion, and absence of left ventricular hypertrophy. Post constrictive dilatation may or may not be present. Rarely, thoracic pseudocoarctation, like true coarctation may be complicated by aneurysm formation, dissection, or even rupture of the aorta. Due to redundant, kinked arch which is relatively more cephalad in the left superior mediastinum, may present with mediastinal mass effect and can be mistaken for an aneurysm or tumor. However, catheterization or modern imaging tools such as CT or MRI will give a clear diagnosis of the same.

Abdominal aortic pseudocoarctation is often diagnosed incidentally and like, thoracic pseudocoarctation, it may be mistaken for an abdominal aortic aneurysm or a tumor. Catheterization, CT, or MRI will help to diagnose the condition. Unlike thoracic aortic pseudocoarctation, there are no reported cases of complications such as aneurysm, dissection, or rupture in the case of abdominal aortic pseudocoarctation.

2.7 Complications associated with pseudocoarctation:
Although a majority of isolated pseudocoarctation, may go unnoticed, it may rarely be associated with complications. Due to its rarity, the exact incidence of complications in these patients is not known. Pseudocoarctation due to the aortic wall fragility is rarely complicated by aneurysm formation of the thoracic descending aorta leading to sudden aortic rupture or aortic dissection. [2, 28] However, the natural course of pseudocoarctation is unclear due to its rarity, and treatment indication remains controversial. [29] Mazzola A et al. in 2007 reported a case of pseudocoarctation with bicuspid aortic valve and aortic dissection who underwent successful surgical repair. [30] Mamoru Arakawa reported a case of thoracic endovascular repair of ruptured aortic pseudocoarctation. [29] Vallabhdas V et al. from India in 1996 reported a case of kinking of the aortic arch with aneurysmal dilatation of the aorta. [31] From the available literature it is clear that in isolated pseudocoarctation, the reported complications are aneurysm formation with or without associated mass effect, dissection, and rarely rupture.

2.8 Management:
Asymptomatic and mildly symptomatic patients are managed conservatively, but there is little data regarding long-term follow-up and treatment recommendations. Indications for surgery include symptoms, radiological features of aortic dissection, or impending aneurysmal rupture. Annual surveillance of thoracic aorta has been recommended for early diagnosis and intervention of aortic aneurysm [32] Makani and colleagues reported surgical repair via left thoracotomy for a patient with pseudocoarctation and multiple aneurysm formation. [28] Mamoru Arakawa and associates reported the first successful thoracic endovascular aortic repair for ruptured thoracic pseudocoarctation. [29] In our case, although the patient was initially symptomatic which subsided with beta-blockers and diuretics, we have not intervened further, and the patient is under observation for any possible recurrence of symptoms or complications.

2.9 Conclusion:
Pseudocoarctation of the arch and abdominal aorta is an extremely rare congenital anomaly occurring in isolation or associated with other congenital heart diseases. No clinical features are specific to this condition and often diagnosed incidentally. Although, in the absence of
symptoms no therapy is required, the occurrence of symptoms and complications guide definitive therapy. As the natural history of the disease is unknown, the condition when diagnosed should be closely followed up for the occurrence of any complications such as aneurysm formation, dissection, or rupture of the aorta.

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