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PRENATAL DIAGNOSIS OF INTERRUPTED AORTIC ARCH: A NARRATIVE REVIEW OF ECHOCARDIOGRAPHIC FINDINGS AND DIAGNOSTIC CHALLENGES

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ABSTRACT

Interrupted aortic arch (IAA) is a rare but severe congenital heart defect characterized by the absence of luminal continuity between the ascending and descending aorta. Early prenatal diagnosis of IAA enables genetic evaluation, delivery planning and appropriate postnatal surgical management, thereby significantly improving neonatal outcomes. This narrative review aims to summarize current knowledge regarding the embryology, classification, associated anomalies, and prenatal echocardiographic diagnosis of fetal IAA, with particular emphasis on clinically relevant imaging features and diagnostic challenges. A literature-based review of previously published studies and clinical reports concerning fetal IAA was performed. Prenatal detection relies primarily on detailed echocardiography with four-chamber and three-vessel views. The second trimester appears to provide the most optimal conditions for diagnosis, as fetal size and position at that gestational age facilitate improved imaging. Characteristic findings of IAA include V-shaped, Y-shaped or W-shaped configurations of the aortic arch and lack of visible continuity between the ascending and descending aorta. Additional findings may include an abnormally shaped ductus arteriosus described as the "broken hockey stick" sign. Quantitative parameters suggestive of IAA include elevated pulmonary artery-to-aorta ratio (PA/Ao) and ventricular disproportion. IAA exhibits strong association with ventricular septal defect (VSD) and 22q11.2 deletion syndrome (especially type B IAA); therefore, detection of these abnormalities should prompt detailed assessment of the fetal aortic arch. Despite advances in fetal echocardiography, prenatal diagnosis remains challenging due to the rarity of IAA, variable presentation and overlapping echocardiographic features with other aortic arch anomalies, particularly coarctation of the aorta.

KEYWORDS

Interrupted Aortic Arch (IAA); Fetal Echocardiography; Prenatal Diagnosis; 22q12.2 Deletion Syndrome; Aortic Arch Anomalies; Congenital Heart Disease; Coarctation of The Aorta (CoA)

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1. Introduction

Pathologies of the fetal cardiac system are the most common congenital defects [1, 2, 3]. The prevalence of congenital heart disease (CHD) is around 1% of the live-born newborns and is estimated to appear 3 to 5 times more often in fetuses [1, 3, 4, 5]. Detection of the congenital defects by fetal echocardiography is continuously expanding and the right diagnosis has a significant impact on the postnatal proceeding [4]. Therefore, it is crucial to gather the existing data to allow for easier detection of the CHD. One of the severe cardiac diseases in fetuses is interrupted aortic arch (IAA) which is characterized by a lack of luminal continuity between the ascending and descending aorta [6, 7, 8, 9, 10, 11, 12, 13]. In terms of fetuses, the Polish National Registry for Fetal Cardiac Malformations (ORPKP) classifies IAA as a severe defect, indicating the need for surgical treatment during the neonatal period [14]. This narrative review aims to synthesize current knowledge regarding prenatal diagnosis of IAA, with particular emphasis on characteristic echocardiographic findings and diagnostic challenges. In addition, the review provides a comprehensive literature-based overview of the embryology, prevalence, classification, and associated anomalies of IAA.

2. Methodology

A narrative review of literature regarding IAA was conducted. Relevant publications were identified through a literature search of electronic medical databases, including PubMed and Cochrane. Articles addressing fetal IAA, prenatal echocardiography, embryology, associated anomalies, and differential diagnosis were reviewed. Due to the rarity of IAA and the limited number of available publications, no strict restriction regarding publication date was applied; however, priority was given to the most recent findings and contemporary sources. Original articles, systematic reviews, and clinical guidelines published in English and Polish were included.

3. Results

3.1 Prevalence of IAA

IAA is a rare but serious cardiac disease [6, 10, 11, 12, 13]. There are 2 cases per 100.000 live births and it comprises 1-1,5% of all congenital heart defects [8, 10, 13, 15, 16]. Isolated IAA is very uncommon, with 95-98% of patients having an associated cardiac anomaly [6, 8, 17, 18, 19]. Some studies classify atresia of the aortic arch, where continuity of segments is maintained by an imperforate fibrous strand as IAA, while others define IAA only as a complete discontinuity [8, 17, 19, 20, 21]. This variability does not significantly affect diagnosis, and further clinical management of the patient [8, 19].

In Poland, the data of the cases of CHD in fetuses is collected by Polish National Registry for Fetal Cardiac Malformations (ORPKP) created in 2004 [ORPKP]. Participants upload the data about pregnant patients and the Registry allows the ultrasonographers to assess their results [22, 23].

3.2 Embryology and pathogenesis of IAA

Development of the heart starts around 18-19 days of fetal life and the arterial system at the beginning of the 4th week [24, 25]. Formation of the great vessels occurs between the 6th and 8th weeks. In the 4th and 5th weeks of fetal development, six pairs of pharyngeal arches arise. Their arteries are called aortic arches [25]. The transverse aortic arch develops from the left fourth aortic arch [6, 25].

IAA results from abnormal development of the pharyngeal arch arteries, particularly involving defective remodeling of the fourth aortic arch. This process is strongly linked to impaired neural crest cell migration and differentiation, and is frequently associated with 22q11.2 deletion syndrome [26]. Abnormal regression of the left fourth aortic arch is especially linked to IAA-B [26].

3.3 Classification of IAA

IAA was first classified into three types by pathologists Celoria and Patton in 1959 [27], based on the anatomical site of interruption. Type A refers to interruption distal to the left subclavian artery, type B to interruption between the left common carotid and left subclavian arteries and type C to interruption between the innominate artery (brachiocephalic trunk) and the left common carotid artery. A further division into subtypes 1 and 2 is based on the origin of the right subclavian artery, depending on whether it arises normally or anomalously distal to the left subclavian artery [8].

Different developmental mechanisms are thought to be the underlying causes of different types of IAA [28]. Type B is strongly associated with anomalies related to neural crest cell dysfunction and is therefore believed to result from defective neural crest development. Type A is less frequently associated with these anomalies and is thought to origin from a combination of altered fetal blood flow and, to a lesser extent, these neural crest–related mechanisms [28]. There is not much data available about the origin of type C IAA.

Type A is only rarely associated with 22q11.2 deletion syndrome, whereas type B shows a strong association, with reported prevalence ranging from approximately 50% up to 70–80% in some cohorts [6, 11, 29, 30]. Therefore, identification of IAA type B should prompt genetic testing for 22q11.2 deletion [11, 13, 31, 32, 33]. Conversely, the presence of 22q11.2 deletion should raise suspicion for conotruncal anomalies, including IAA [11].

3.4 IAA and associated anomalies

In a study by Vogel et al. [11], 14% of fetuses prenatally diagnosed with IAA had a family history of structural or chromosomal anomalies. Around 30-50% of IAA cases are associated with a malformation syndrome or chromosome abnormality [11, 31]. Among these, the most common is 22q11.2 deletion syndrome (DiGeorge syndrome) [12, 13, 34, 30]. The TBX1 gene, located within the deleted region, plays a critical role in pharyngeal arch development and its haploinsufficiency leads to defects of the aortic arches [35] thus increasing the risk of IAA. It should be emphasized that the type of IAA most strongly linked to 22q11.2 deletion syndrome is IAA-B [36]. The anomalies associated with IAA are presented in Table 1.

Table 1. Anomalies associated with IAA.

IAA isolated	IAA and other cardiac anomalies	IAA and extracardiac anomalies
2-5% [6, 8, 17, 18]	95-98% [6, 8, 17, 18, 37]	30-50% [11, 31, 37]
	VSD – 90% [8, 10, 18, 28, 38], 30-75% with isolated VSD [17, 39, 40, 41] (2-4 times more frequently muscular than membranous[10, 42])	DiGeorge syndrome: 30-50% of IAA overall [13, 15, 18, 29, 39, 40, 44, 32] >50% of IAA-B [13, 47]
	ASD (usually present in the form of a stretched patent foramen ovale) [8, 18, 43]	polyhydramnios, IUGR, two-vessel umbilical cord, and hyperechogenic multi-lobed placenta [10, 18, 48]
	large PDA - 90-100% [8, 9, 17, 38]	CHARGE syndrome [49] Turner syndrome, VACTERL and Klippel-Feil syndrome [11, 15]
	Truncus arteriosus – 10-15% [8, 17, 29, 40, 41, 43, 44]	deviation of septum or hypoplasia/aplasia of the infundibular septum - 75% [6, 10, 12, 18, 40, 44, 50]
	Transposition of the great arteries - 6-8% [9, 17]	ARSA [17, 29, 39, 40, 45] Type A IAA – 5% [19] Type B IAA – 50-100% [10, 29, 43]
	Single ventricle – 3-20% [17, 40, 41, 43, 45]	Williams syndrome [51]
	Aortic valve bicommissural – 30% [8, 17, 46]	Apert syndrome [52]
	Taussig-Bing - 12% [41], DORV might also appear [6, 29, 40, 41, 44, 45]	Aortopulmonary window - 4% [9, 29, 40, 41, 44, 45, 53] (Berry syndrome - in >90% IAA is present [54])
	subaortic stenosis - 25% [46]	Jakobsen syndrome [49]

**VSD – Ventricular Septal Defect; ASD – Atrial Septal Defect; PDA – Patent Ductus Arteriosus; DORV - Double Outlet Right Ventricle; IUGR - Intrauterine Growth Restriction; ARSA - Aberrant Right Subclavian Artery.*

Due to the strong association of VSD and IAA (especially type B), when VSD is detected, it should prompt detailed evaluation of the fetal aortic arch [11]. VSD is commonly associated with posterior malalignment of the outflow septum [55, 17]. Deviation of outflow septum or hypoplasia/aplasia of the infundibular septum is present in 75% of patients with IAA, especially type B, and may lead to subaortic stenosis and aortic annular hypoplasia resulting in varying degrees of left ventricular outflow tract (LVOT) obstruction [6, 10, 12, 18, 40, 44, 50, 55, 56]. LVOT is observed to be further reduced in size in cases where the right subclavian artery originates anomalously (ARSA), particularly when associated with a hypoplastic aortic annulus [43, 55]. ARSA most commonly arises either distal to the left subclavian artery or high in the neck from the right common carotid artery (CORSA) or from the arterial duct (isolated right subclavian artery) and is associated with an increased risk of subaortic stenosis [10, 43, 55].

3.5 Prenatal diagnosis of IAA

IAA is a challenging condition to diagnose prenatally, however detection rates have been increasing [11, 48]. The primary method used to diagnose IAA and associated cardiac anomalies is echocardiography [6, 39]. Variation in the prenatal detection of IAA across trimesters is presented in Table 2.

Table 2. Variation in the prenatal detection of IAA across trimesters.

IAA detection in 1st trimester	IAA detection in 2nd trimester	IAA detection in 3rd trimester
discrepancy between the right and left atria and ventricles can be visualized [1, 12]	optimal visualization conditions that facilitate diagnosis - favorable size of the heart, position of the fetus, the quality of the images, undeveloped lungs and uncalcified ribs [57]	limited visualization conditions - due to e.g. well-developed lungs, calcified ribs, placenta on anterior wall, and decreased volume of the amniotic fluid [58]
VSD can be detected with the use of color Doppler imaging [12, 13]	the mean diagnosis of IAA is most commonly obtained around 25th weeks of pregnancy [10, 11, 12]	some studies reported diagnosis of IAA at median gestational age of 31-32 weeks [42, 48]
the three vessels view (3VV) in the upper mediastinum can present a relatively smaller ascending aorta than the pulmonary artery [12, 13] however the diameters of AoA and PA may still be within normal range limits in early gestation [11]	discrepancy between the diameters of AoA and PA becomes more visible [11]	dfferences in great vessel diameters may become less pronounced compared to the 2nd trimester [11, 59]

**AoA – Aortic Arch; PA – Pulmonary Artery.*

A smaller diameter of aortic arch (AoA) compared to pulmonary artery (PA) may indicate coarctation or interruption of the aorta [10, 12, 13, 42, 48, 60, 61]. However, most of the screening examinations performed during the 1st trimester cannot confirm those diagnoses, as the diameters may still be within normal limits for this gestational age [62]. Therefore, a 1st trimester scan does not fully exclude the possibility of an aortic arch anomaly [57]. A definitive diagnosis is most commonly established in the 2nd trimester [11, 12], when the majority of fetal echocardiographic examinations are performed [1, 10, 11, 18, 22, 42, 57, 63]. However, it is important to note that the hemodynamics of the fetus may evolve during the second part of the pregnancy. That is why a detailed echocardiographic assessment of the fetus during the 3rd trimester is crucial [22, 23, 64]. Furthermore, normal results at the 1st and 2nd trimester do not exclude the presence of congenital heart disease and third-trimester evaluation may reveal previously undetected abnormalities [64, 65, 66].

Current screening recommendations include a 4-chamber view, all 4 valves, and the great vessels [11, 62, 67]. The four-chamber projection (4CV) in fetuses with IAA may initially present a normal view with preserved chamber morphology and correct connections between the chambers and great vessels [42]. In most cases, there is no significant discrepancy between the right and left ventricles, and overall cardiac size remains within normal limits [10, 11, 12, 18, 42, 48, 68]. The normal appearance of the heart may be explained by frequent co-occurrence of VSD, which allows blood flow into the left ventricle promoting its normal growth and resulting in normal LV dimensions in fetuses with IAA. Despite this, ventricular size discrepancy may still occur occasionally even in the presence of VSD [10, 11,12]. An abnormal left ventricle is more likely to be observed in cases with LVOT or aortic arch obstruction and restrictive VSD [10, 11]. When present, left–right ventricular disproportion visible on the 4CV should prompt further evaluation of the great vessels [13].

3.6 Fetal echocardiographic features of IAA

There are several morphological features visible in fetal echocardiography that may aid in the diagnosis of IAA. A common finding is that the ascending aorta appears straighter than normal, as it does not curve at the level of the first brachiocephalic vessel [11, 13, 69].

In type B IAA, this results in a characteristic Y-shaped or V-shaped configuration, in which the ascending aorta gives rise to the innominate and left common carotid arteries and continues cephalad without forming the normal aortic arch curvature [10, 11, 12, 13, 48, 60].

In type A IAA, the ascending aorta demonstrates a subtle curvature after the origin of the innominate artery. The left common carotid and left subclavian arteries arise in their normal sequence, reflecting preservation of the proximal portion of the aortic arch. A characteristic W-shape is observed [10, 13, 68].

In the sagittal view, the ascending aorta in type B IAA can be traced into the innominate and left common carotid arteries but not into the descending aorta. A similar finding is observed in type A IAA, where the subclavian artery originates from the ascending aorta; however there is no continuity with the descending aorta [10, 12].

The complete aortic arch is not visualized in transverse planes. However, blood flow in the distal aortic arch may be detected in the sagittal view as retrograde flow originating from the patent ductus arteriosus (PDA) [10, 12, 60]. Furthermore, reversed blood flow may be observed in the transverse aortic arch [60]. Słodki et al. [48] additionally described an abnormally shaped ductus arteriosus in some fetuses with IAA, referred to as the "broken hockey stick" sign.

Prenatal diagnosis of IAA via echocardiograms may be challenging due to the anatomical similarity between the ductus arteriosus and the aortic arch, as well as the potential misinterpretation of the left subclavian artery arising from the proximal descending aorta as a carotid vessel originating from the aortic arch [9, 14, 15]. These difficulties may be further exacerbated by unfavorable fetal position or maternal factors such as obesity [47, 56].

3.7 Quantitative echocardiographic parameters suggestive of IAA

3.7.1 Pulmonary artery-to-ascending aorta ratio (PA/Ao)

Generally, in fetuses with IAA, a typical anatomical arrangement of the three vessels in the upper mediastinum is preserved. However, the ascending aorta (Ao) and the aortic valve (AoV) are usually significantly smaller than the main pulmonary artery (PA) and pulmonary valve [10, 11, 12, 13, 32, 39, 42, 48]. That developmental regression results from VSD and large ductus arteriosus, which redirect blood flow away from the ascending aorta [39]. Quantitative differences in vessel size have been reported. In a study by Yang et al. [68], the diameter of the Ao at 18 weeks of gestation was 1.7 mm, compared to 4.6 mm for the PA. Similarly, Wójtowicz et al. [12] reported an Ao diameter of 2.2 mm at 21 weeks. In comparison, the Ao of a healthy fetus at corresponding gestational ages measures approximately 4.0 mm [60].

In a normal pregnancy, the fetal pulmonary artery/ascending aorta (PA/Ao) ratio is approximately 1.1 at 20 weeks and increases to 1.2 at 40 weeks [59, 70]. Słodki et al. suggested [48] that while this ratio increases slightly with advancing gestation in normal pregnancies, it may be further elevated in case of IAA due to altered hemodynamics resulting from the lack of continuity between the ascending and descending aorta. This change in hemodynamics leads further to more progressive dilatation of the PA. Thus, isolated dilation of the pulmonary artery in the upper mediastinum of the fetus may suggest the presence of IAA [71].

Significantly increased pulmonary artery-to-ascending aorta (PA/Ao) diameter ratios, reaching values of 3.0–3.4, have been reported in type B IAA, whereas such high ratios are typically not observed in type A [42]. As noted by Słodki et al. [48], these markedly elevated PA/Ao ratios may be highly suggestive of type B IAA.

3.7.2 Ventricular disproportion - left ventricle-to-right ventricle ratio (LV/RV) and aortic valve-to-pulmonary valve ratio (AoV/PV)

In type A IAA the left ventricle (LV) is typically smaller than the right ventricle (RV), with an average reported LV/RV ratio ranging from 0.44 to 0.78. In contrast, in type B IAA, there is usually no significant difference between ventricular dimensions, and the LV/RV ratio is approximately 1.0 [10, 42].

The aortic valve-to-pulmonary valve (AoV/PV) diameter ratio in fetuses with IAA typically ranges from 0.4 to 0.6. The average AoV/PV ratio in fetuses with no IAA but with an isolated VSD is approximately 0.8 [11, 42]. Therefore, Hirano et al. noted [42] that presence of VSD combined with reduced AoV/PV should prompt evaluation for IAA.

Quantitative parameters suggestive of IAA obtained in 2nd and 3rd trimesters in selected studies are presented in Table 3.

Table 3. Pulmonary artery-to-ascending aorta (PA/Ao) and left ventricle-to-right ventricle (LV/RV) ratios in IAA reported in selected studies.

Year	Author	Pa/Ao ratio	LV/RV ratio
2021	Yang et al. [68]	2.7	1.0
2016	Hirano et al. [42]	1.8 – 2.7	0.61 ± 0.17 (type A) 1.0 (type B)
2015	Vogel et al. [11]	1.7 – 2.4	<1.0
2014	Wójtowicz et al. [12]	2.0	<1.0
2011	Słodki et al. [48]	2.2 - 3.0	Not quantitatively reported, abnormal four-chamber view
2002	Volpe et al. [13]	>1.4	<0.77

Values represent reported ranges or mean measurements depending on the study methodology.

3.8 Differential diagnosis: IAA vs CoA

A decrease in LV/RV ratio, as well as discrepancies in PA/Ao and AoV/PV ratios may also be observed in fetuses with coarctation of the aorta (CoA) [11, 48, 61, 72, 73], thereby limiting the specificity of these echocardiographic parameters in differentiating between IAA and CoA. Additionally, morphological features such as the "W-shape" configuration may be seen in both conditions, further complicating differentiation [61]. According to data from the Congenital Heart Surgeons' Society [39], the ascending aorta tends to be smaller in IAA compared to CoA. However, distinguishing between IAA — particularly type A — and CoA remains challenging [11, 42]. Both conditions typically occur at a similar anatomical location, with the key difference being the absence of luminal continuity in IAA [61, 19]. Despite the similarities, an important distinguishing feature is that in IAA the ascending aorta follows a straight course toward its branches, whereas in CoA the aortic arch maintains its normal curvature and continuity with the descending aorta [13].

4. Discussion

4.1 Diagnostic limitations

Despite advances in fetal echocardiography, prenatal diagnosis of IAA remains challenging. Diagnostic limitations include similarity of echocardiographic features with coarctation of the aorta, dependence on fetal position, and operator experience [11, 42, 57, 74]. In addition, physiological changes in fetal blood flow and hemodynamics during gestation may mask any abnormalities, leading to false-negative results, particularly in the first trimester [57].

4.2 Screening and Clinical Management

As noted by Wójtowicz et al. [12], current guidelines of the Polish Society of Gynecology for routine fetal anomaly screening do not include systematic assessment of the aortic arch [67]. One of the primary indications for referral to a specialized cardiac center is increased nuchal translucency (NT) of the fetus, defined as greater than 3.5 mm [57, 67, 75]. However, in fetuses with IAA, increased NT accounts for only approximately 10% of referrals [10].

The most common reason for referral to tertiary care centers is the detection of abnormalities in the four-chamber view (4CV) or the three-vessel view (3VV) during routine screening. Other indications include extracardiac anomalies, polyhydramnios, persistent fetal arrhythmias (e.g., extrasystoles), intrauterine growth restriction (IUGR), and a positive family history of congenital heart disease [10, 13, 22, 42, 48].

The accuracy of prenatal diagnosis of aortic arch anomalies in specialized referral centers exceeds 80%. Diagnostic challenges primarily involve differentiating between severe coarctation, hypoplastic aortic arch, and complete interruption, as well as determining the specific type of IAA. Importantly, these distinctions do not substantially alter the postnatal management, as both hypoplastic aortic arch and IAA require the use of prostaglandins to maintain patency of the ductus arteriosus [10, 11, 42, 76 (56)].

Early detection of any fetal cardiac anomaly during routine screening is crucial, as subsequent evaluation in specialized centers provides high rates of further diagnostic accuracy. Prenatal diagnosis is associated with improved neonatal outcomes, including lower rates of metabolic acidosis and reduced risk of neurological complications [1, 2, 72]. Furthermore, it enables appropriate parental preparation regarding the prognosis and postnatal management as well as optimal planning of delivery and timing of surgical intervention [1, 62]. Systematic assessment of fetal heart throughout pregnancy and increased awareness of possible echocardiographic findings remain essential for quicker IAA diagnosis and further improvements of neonatal outcomes.

5. Conclusions

IAA is a rare but severe congenital cardiac anomaly that remains challenging to diagnose prenatally despite advances in fetal echocardiography. Early identification of IAA facilitates appropriate perinatal planning, including referral to specialized centers, genetic testing and surgical management, and increases the probability of neonatal wellbeing. Systematic echocardiographic assessment of the fetal heart is therefore essential, especially during second and third trimesters, when imaging conditions are most favorable.

Prenatal diagnosis of IAA relies on a combination of qualitative echocardiographic morphological features and quantitative parameters, including alterations in great vessel dimensions and ventricular proportions. Characteristic findings include absence of visible continuity between ascending aorta and descending aorta, abnormal V-shaped, Y-shaped, or W-shaped configurations of the aortic arch, and, in some cases, the presence of the "broken hockey stick" sign. Quantitative indicators suggestive of IAA include

increased pulmonary artery-to-aorta ratio and ventricular disproportion with reduced left ventricle-to-right ventricle ratio. However, many of these features overlap with other aortic arch anomalies, particularly coarctation of the aorta, which continues to limit diagnostic specificity. Additional diagnostic challenges include fetal position, operator-dependent variability and maternal features such as obesity.

IAA rarely occurs as an isolated defect and is frequently associated with other cardiac and extracardiac anomalies. The strong association of IAA type B, with genetic 22q11.2 deletion syndrome highlights the importance of integrated genetic and cardiac assessment.

Further refinement of screening protocols, increased awareness of subtle echocardiographic signs, and improved understanding of fetal hemodynamics may contribute to earlier and more accurate prenatal diagnosis of IAA, ultimately improving perinatal management and neonatal outcomes.

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