Original Paper

First Trimester Ultrasound Diagnosis of Right Aortic Arch (RAA)

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ABSTRACT: Objectives. In the present research we proposed to evaluate the cases diagnosed in the first trimester of pregnancy by ultrasound with RAA, knowing the fact that although, in most cases the disease is isolated and asymptomatic, in some cases the presence of RAA can be associated with other fetal structural abnormalities that must be detected and monitored during pregnancy. We established correlations between the postnatal or anatomopathological examination (in cases ended by therapeutic abortion) and the presence of RAA detected in the first trimester. Material and method. We conducted a retrospective analytical study that investigated the role of the RAA early detection (isolated or associated with other cardiac abnormalities) for a correct pregnancy monitoring and postpartum management. Between 2012 and 2018, patients admitted in the first Obstetrics and Gynecology Clinic-the Prenatal Diagnostic Unit-of the Emergency County Hospital from Craiova, were evaluated in the first trimester of pregnancy for genetic abnormalities and early morphology. The study material was represented by the patient's medical records (observation sheets, surgical protocol records, anatomopathological diagnostic records). The obtained information was stored in Microsoft Excel files and statistically processed. Results. During the study period, 14 cases with right aortic arch were diagnosed in the first and second trimester of pregnancy. 4 cases were detected in the first trimester: 2 cases (50%) presented left ductus arteriosus (DA)-RAA type 2 ("U" sign) and 2 cases (50%) presented right DA-RAA type 1 (mirror image-"V" sign). RAA type 1 associated Tetralogy of Fallot in one case (25%) and in another one case (25%) the anomaly was isolated. RAA type 2 associated atrio-ventricular septal defect (AVSD) in one case (25%) and in another one case (25%) the anomaly was isolated. There were no fetal extracardiac structural abnormalities associated with the RAA diagnosis in the first trimester. Conclusions. Over a seven years study period (2012-2018), 14 cases with RAA in the first and second trimester of pregnancy were detected. In the low-risk pregnancies group, the first trimester incidence of the RAA was 0.11% and the association of congenital heart abnormalities was 50%.

KEYWORDS: *Ultrasound diagnosis, right aortic arch, first trimester*

Introduction

The aortic arch is the segment of the aorta that connects the ascendant and descendent aortas. The normal position of the aortic arch, to the left of the trachea, can be early assessed during the first trimester ultrasound [1] on the three vessel and trachea (3VT) view [2,3,4].

Literature describes 2 types of right aortic arch (RAA): type 1, mirror image RAA where the "V" sign appears to the right of the trachea, between the aortic arch and the right ductus arteriosus (DA) and type 2 characterized by the "U" sign formed between the aortic arch that lays to the right of the trachea and the left DA.

As literature cites different incidences for cardiac and extracardiac associated abnormalities [4], differentiating these types of RAA is important.

The RAA is a rare congenital abnormality and is the one of the least diagnosed cardiac abnormality [5,6]. The incidence is very low, about 0.1% in low risk pregnancies [6]. The association with congenital heart diseases is considered to be around 50% [7].

Material and method

We conducted a retrospective analytical study that investigated the cases diagnosed with RAA, admitted between January 2012 and December 2018, in the Prenatal Diagnostic Unit of the First Obstetrics and Gynecology Clinic of the Emergency County Hospital from Craiova.

The patients were evaluated using the following data: personal data (name, surname), epidemiological data (age, place of residence, occupation, blood group, Rh), etiopathogenic data (family history of congenital malformations and prevention of maternal conditions-diabetes, lupus). The possible pregnancy exposure to toxic substances or infections was also evaluated.

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The study included 14 cases diagnosed with RAA isolated or associated with other cardiac and extracardiac abnormalities in the first and second trimester of pregnancy.

The study inclusion criteria were: pregnancies evaluated in the first trimester for genetic and morphological abnormalities.

The exclusion criteria included the patient's refusal to submit to the investigation protocol.

All patients gave a written informed consent, and the study was approved by the Ethics Committee of the University of Medicine and Pharmacy of Craiova.

The study material was represented by the patient's medical records: observation sheets, surgical protocol records and pathological diagnostic records.

The data were collected in Microsoft Excel files for descriptive statistical analysis.

For the ultrasound diagnosis of RAA, an extended heart protocol was applied (Fig.1). This included the 3 vessel and trachea view (3VT) in conventional two-dimensional (2D) ultrasound and Colour Doppler mode in order to confirm the correct position of the aortic arch.

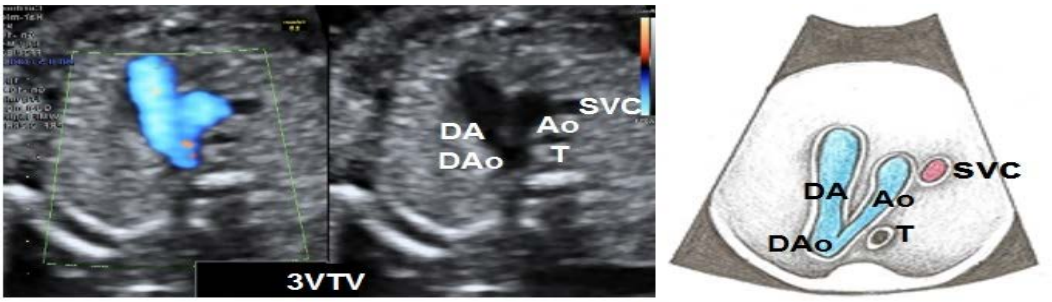

Fig.1. 3VT: aorta-Ao, trachea-T, descending aorta-DAo, spine-Sp, superior vena cava-SVC, ductus arteriosus-DA

The examiner started with the four-chamber view in the axial section and moved the ultrasound probe in a cranial direction to emphasize the left and the right outflow tracts, the three-vessel view and the 3VT view. The normal position of the aortic arch was demonstrated by the presence of the "V" sign to the left of the trachea and anterior and to the left of the spine. The normal "V" sign is formed by the aortic arch and the left DA. This image is diagnostic for the normal left aortic arch.

An abnormal "U" sign indicates a RAA with left DA. In this situation the trachea is situated between the two vessels. An apparent normal "V", but to the right of the trachea, indicates a mirror image RAA. This sign forms between a RAA and a right DA (Fig.2).

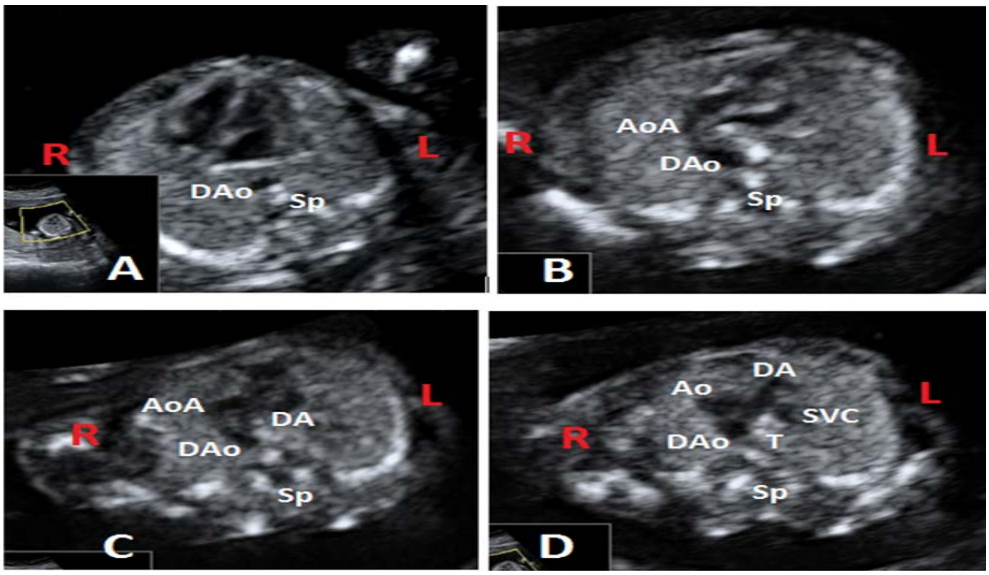

Fig.2. First trimester fetal heart: Four chamber view-A; Emergence and course of the right sided aortic arch (AoA)-B; Arterial arches heading toward the right side of thorax-C; 3VT showing a right-sided 'V'-sign. Ao-aorta, T-trachea, AoA-ascending aorta, DAo-descending aorta, Sp-spine, SVC-superior vena cava, DA-ductus arteriosus-D

When a RAA was suspected, the foetus was sequentially scanned for other cardiac or extracardiac abnormalities.

The antenatal diagnosis was confirmed by postnatal or pathological examination in cases ended up by therapeutic abortion.

Results

During the study period, 14 cases with right aortic arch were diagnosed in the first and second trimester of pregnancy. 4 cases were detected in the first trimester and 10 cases in the second trimester (Table 1).

Table 1. Patients distribution by maternal age group in first trimester (FT) and second trimester of pregnancy (ST)

Age group	15-19 years		Total	20-24 years		Total	25-29 years		Total	30-34 years		Total	**All groups total**		**Total**
Trimester (T)	FT	ST		FT	ST		FT	ST		FT	ST		FT	ST	
Patient number	-	2	2	2	4	6	2	3	5	-	1	1	4	10	14
Percentage	-	14.3	14.3	14.3	28.6	42.9	14.3	21.4	35.7	-	7.1	7.1	28.6	71.4	100

Thus, for the cases diagnosed with RAA in the first trimester, both aged groups that included this pathology had the same chance of developing it, ie the 20-24 age group and the 25-29 age group.

For the cases diagnosed with RAA in the second trimester, the most frequent maternal age was for the 20-24 years old group, then the incidence clearly decreased for both the 15-19 age group and the 25-29 age group. The 25-29 age group, had however, a higher incidence, and overall these two age groups showed the highest chance of presenting with the pathology, for both the first and second trimester.

The average maternal age for all cases was of 24±5 years.

Gestational age at diagnosis ranged between 12 weeks and 5 days and 21 weeks and 6 days (Table 2).

Table 2. Patients distribution by gestational age

	-13w		Total	13w+		Total	**All groups total**		**Total**
Trimester (T)	FT	ST		FT	ST		FT	ST	
Patient number	1	5	6	3	5	8	4	10	14
%	7.1	35.7	42.9	21.4	35.7	57.1	28.6	71.4	100

One case (7.1%) diagnosed in the first trimester had a gestational age of 12 weeks and 10 days and in 3 cases (21.4%) the gestational age was: 13 weeks and 2 days (one case-7.1%), 13 weeks and 6 days (one case-7.1%) and 13 weeks and 5 days (one case-7.1%).

5 cases (35.7%) diagnosed in the second trimester with RAA had a gestational age between 20-22 weeks of gestation and in the other 5 cases the gestational age ranged between 19 and 20 weeks of gestation.

From the 4 cases detected in the first trimester: 2 cases (50%) associated left arterial duct (DA)-RAA type 2 (U sign) and 2 cases (50%) associated right arterial duct-RAA type 1 (mirror image-V sign) (Table 3).

Table 3. Patients distribution by RAA type in the first trimester

RAA type	**RAA type 1**	**RAA type 2**	**Total**
Patient number	2	2	4
%	50,0	50,0	100,0

The cases diagnosed with RAA were further scanned for possible cardiac or extracardiac associated abnormalities. Two cases (50%) associated cardiac defects: one case diagnosed with RAA type 1 associated Tetralogy of Fallot (Fig.3) and one case diagnosed with RAA type 2 associated atrio-ventricular septal defect. It is of interest to note that both cases that associated cardiac defects were in the group above 13 weeks of gestational age, linking this to a possible limitation of imaging techniques in detecting these defects at younger ages.

The other 2 cases (one case of RAA type 1 and one case of RAA type 2) were isolated, without further detectable vascular abnormalities (Table 4).

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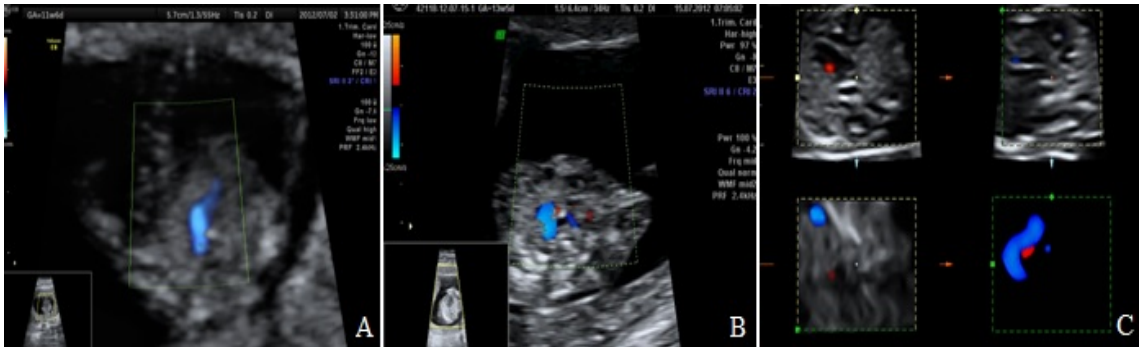

Fig.3. RAA associated with Tetralogy of Fallot: 11 weeks of gestation-A; 13 weeks of gestation-B and C

Table 4. Patients distribution by associated fetal structural abnormalities

	RAA associated with cardiac abnormalities		Total	Isolated RAA	Total	Total
Cardiac associated abnormalities	Tetralogy of Fallot	AVSD				
Patient number	1	1	2	2	2	4
%	25,0	25,0	50,0	50,0	50,0	100,0

Discussion

In most cases RAA is detected as isolated and asymptomatic [5]. However, the antenatal diagnosis of this anomaly is very important because of the possible associated structural abnormalities, especially congenital heart defects [8,9,10], that are considered medical emergencies in the first hours after birth. Therefore, an early detection of this abnormality is preferable. The RAA also presents a high risk for extra cardiac associated abnormalities [11,12,13,14].

In our study, in 2 cases that associated cardiac defects (TOF and AVSD), after the parents were extensively counseled on the importance of major cardiac abnormalities, they opted for therapeutic abortion.

The type of the RAA is important to diagnose because each type has a different prognosis. The literature cites an increased risk of severe congenital heart defects, especially associated with mirror imaging (RAA type 1) [15].

Specialized literature cites this as the most common associated cardiac abnormalities, TOF and AVSD. Our study detected 2 cases with RAA type 2 of which one isolated case (25%) and one case (25%) associated with AVSD; 2 cases were diagnosed with RAA type 1 of which one associated with TOF and one isolated case.

The literature reports an increased associated risk between RAA and extracardiac abnormalities [4].

The association with other extracardiac structural abnormalities may indicate an adverse prognosis, as none of our cases diagnosed in the first trimester associated extracardiac structural abnormalities and had normal postnatal outcome.

The first trimester morphological ultrasound can detect the position of the aortic arch [15,16].

A correct ultrasound diagnosis of the "V" sign cannot rule out the abnormality. The aortic arch should be noted as normal immediately after a correct fetal orientation, because the mirror image of a RAA with right DA forms exactly the same "V" sign, but on the right side [11].

A severe complication of RAA is the compression of the trachea and esophagus due to a tight ring made by the aortic arch. The literature mentions a rate between 13% and 26% of vascular compression in newborns with persistent apnea and airway obstruction [18].

However, in our study population, signs of compression were not detected and the postnatal follow-up was favorable.

Conclusions

Over a seven years study period (2012 and 2018), 14 cases with RAA in the first and second trimester of pregnancy were detected in our clinic.

In the low-risk pregnancies group, the first trimester incidence of the RAA was 0.11% and the association of congenital heart abnormalities was of 50%.

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