

Increasing isolated right heart chambers in second trimester ultrasound: always a warning sign?

Aumento isolado das câmaras cardíacas no ultrassom de segundo trimestre de gestação: sempre um sinal de alerta?

Milena Carvalho Carrilho¹, Fernanda Silveira Bello de Barros¹, MD; Edward Araujo Júnior¹, MD, PhD

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Congenital heart disease (CHD) is one of the leading causes of birth defects and occurs in approximately 1 in every 100 live births^[1]. This condition has been attracting the attention of the medical community in terms of screening and diagnosis of CHDs. Because of the constant movement of the fetus during ultrasound examinations and the small size of the heart, it is challenging to obtaining good quality images and performs accurate analysis of intracardiac structures and outflow tracts. The increasing ability of medical professionals allied to recent technological advances, from the development of sector transducers with real-time imaging to two-dimensional studies and color mapping of blood flow to the advent of three-dimensional (3D) technology, has enabled new perspectives for earlier and more accurate diagnoses.

Despite these medical advancements, analysis of the enlargement of the right cardiac chamber of the fetus remains a major challenge for sonographers and echocardiographers. In the normal fetal heart, physiological enlargement of the right atrium and a simultaneous increase in the right ventricle can be observed. Moreover, the right atrium is the only cardiac chamber that receives the entire cardiac output^[2]. This situation demands special attention and caution when determining the boundaries between physiological and pathological processes, particularly when evaluating aortic coarctation, which can be erroneously considered as normal.

Since 1988, the enlargement of the right cardiac chambers has drawn attention from specialists in fetal cardiology. Allan et al.^[3] conducted a prospective study involving approximately

2000 pregnant women, and enlargement of the right ventricle and pulmonary artery were found in 24 fetuses. Coarctation or interruption of the aorta was clinically suspected in 18 of these fetuses and confirmed in 10. Five cases of aortic coarctation were not detected by fetal echocardiography but were confirmed postnatally^[3]. Starting in 2001, Hornung et al.^[4] studied 43 fetuses with right ventricular enlargement over a 5-year period and verified that this condition had both cardiac and noncardiac origins. Fifteen patients presented enlargements of cardiac origin, 14 of which were caused by structural abnormalities; one patient had tachycardia. These structural abnormalities primarily included coarctation of the aorta (N=4) and ventricular septal defects (VSD; N=4). Nine patients with enlargement had associated extracardiac malformations, particularly chromosomal disorders, and 19 others had associated enlargement of the right ventricle without intra- or extracardiac anomalies^[4].

Differential diagnosis should always involve the assessment of aortic coarctation when there is disproportionate enlargement of the right ventricular chamber, considering the low sensitivity and predictive values of the ventricular disproportion, corresponding to 62% and 33%, respectively^[5]. The sensitivity and specificity values can decrease even further after the 34th week of gestation, when there is physiological enlargement of the right cardiac chamber^[6].

Aortic coarctation accounts for 2%-6% CHD patients. Nonetheless, channel-dependent congenital cardiopathies are the least diagnosed in the prenatal period because of the high rate of false-negative results and the low diagnostic specificity; consequently, many newborns are considered healthy and are discharged. The prenatal diagnosis of aortic coarctation allows early intervention and improved prognosis^[2,7].

After right chamber enlargement is diagnosed, physicians should initially ensure that it is an isolated occurrence and

¹Federal University of São Paulo (UNIFESP), Department of Obstetrics, Fetal Cardiology Unit, São Paulo, SP, Brazil.
E-mail: araujojred@terra.com.br

Abbreviations, acronyms & symbols

3D	Three-dimensional
3VT	Three-vessel and tracheal
CHD	Congenital heart disease
VSD	Ventricular septal defects

eliminate the possibility of occurrence of other conditions, including right ventricular outflow tract obstruction, hypoplasia of the left chambers, and restrictive atrial septum. An additional differential diagnosis of this isolated enlargement includes vein of Galen aneurismal malformations and the noncardiac diagnoses of aneuploidy, anemia, and restricted intrauterine growth caused by placental insufficiency^[2,8].

A thorough assessment of the aorta through the three-vessel and tracheal (3VT) view is critical to determine the disproportion in the diameter of the great arteries. To decrease the high incidence of false-positive results related to the prenatal diagnosis of aortic coarctation, Pasquini et al.^[6] included the measurement of the aortic isthmus in the 3VT method on the basis of the hypothesis that aortic arch hypoplasia is more common in the fetus, thus avoiding diagnostic errors arising from analysis of the aortic arch as a whole. Considering that the site of narrowing of the aortic isthmus is located immediately before the entry of the ductus arteriosus to the proximal descending aorta, this location can be used to measure the isthmus (Z-score) associated with gestational age^[6].

Other cardiovascular conditions should be accurately identified in patients with enlargement of the right cardiac chambers, such as the presence of a persistent left superior vena cava, VSD, and a bicuspid aortic valve. These diagnostic findings may be overlooked and often mask the diagnosis of aortic coarctation^[9,10].

In summary, the isolated enlargement of the right cardiac chamber is a normal finding in the most cases. However, aortic coarctation is a serious disease, and its early detection can improve patient recovery. For this reason, we recommend specialized fetal echocardiography in all pregnant women whose fetuses present with isolated enlargement of the right cardiac chamber detected by ultrasound during the second trimester.

Authors' roles & responsibilities

MCC	Drafting of the manuscript or revising its content critically
FSBB	Drafting of the manuscript or revising its content critically
EAJ	Final approval of manuscript

REFERENCES

1. Hoffman JI, Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol.* 2002;39(12):1890-900.
2. Lopes LM. Anomalias do arco aórtico. In: Lopes LM, ed. *Atlas comentado de cardiologia fetal.* São Paulo: RR Donnelley America Latina; 2003. p.258-68.
3. Allan LD, Chita SK, Anderson RH, Fagg N, Crawford DC, Tynan MJ. Coarctation of the aorta in prenatal life: an echocardiographic, anatomical, and functional study. *Br Heart J.* 1988;59(3):356-60.
4. Hornung TS, Heads A, Hunter AS. Right ventricular dilatation in the fetus: a study of associated features and outcome. *Pediatr Cardiol.* 2001;22(3):215-7.
5. Matsui H, Mellander M, Roughton M, Jicinska H, Gardiner HM. Morphological and physiological predictors of fetal aortic coarctation. *Circulation.* 2008;118(18):1793-801.
6. Pasquini L, Mellander M, Seale A, Matsui H, Roughton M, Ho SY, et al. Z-scores of the fetal aortic isthmus and duct: an aid to assessing arch hypoplasia. *Ultrasound Obstet Gynecol.* 2007;29(6):628-33.
7. Paladini D, Russo M, Teodoro A, Pacileo G, Capozzi G, Martinelli P, et al. Prenatal diagnosis of congenital heart disease in the Naples area during the years 1994-1999: the experience of a joint fetal-pediatric cardiology unit. *Prenat Diagn.* 2002;22(7):545-52.
8. Head CE, Jowett VC, Sharland GK, Simpson JM. Timing of presentation and postnatal outcome of infants suspected of having coarctation of the aorta during fetal life. *Heart.* 2005;91(8):1070-4.
9. Pasquini L, Fichera A, Tan T, Ho SY, Gardiner H. Left superior caval vein: a powerful indicator of fetal coarctation. *Heart.* 2005;91(4):539-40.
10. Paladini D, Volpe P, Russo MG, Vassallo M, Sclavo G, Gentile M. Aortic coarctation: prognostic indicators of survival in the fetus. *Heart.* 2004;90(11):1348-9.