

4D Study of Right Heart Anomalies

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Abstract: In this chapter, the 2D, color Doppler and four dimensional (4D) features of major right heart abnormalities are described. In particular, the echocardiographic views on which the various lesions are present are reported. The diagnostic role of 4D echocardiography in allowing a spatial demonstration of the defects with the possibility of getting new views into the heart is outlined. Videos of major diagnostic features are also provided, to facilitate the understanding of the text.

Key Words: 4D Sonography, Fetal Echocardiography, Right Heart Anomalies, Prenatal Diagnosis.

INTRODUCTION

This chapter will cover the most significant anomalies of the right side of the heart that can be observed prenatally. For each condition after a brief anatomical description, the criteria of echocardiographic diagnosis will be provided. In particular we will focus on the four dimensional (4D) acquisition and postprocessing modalities in fetal hearts with right side anomalies, demonstrating their use through case examples. A classification of right heart anomalies is reported in Table 1

Table 1: Fetal right heart anomalies

Anomalies of the inlet
Tricuspid atresia
Tricuspid dysplasia, regurgitation
Ebstein's anomaly
Anomalies of the outlet (without septal defect)
Pulmonary stenosis
Pulmonary atresia
Anomalies of the outlet (with septal defect)
Tetralogy of Fallot
Double outlet right ventricle
Truncus arteriosus

The anomalies of the outlet with associated a ventricular septal defect (the so called conotruncal anomalies) will be described in chapter 15.

TRICUSPID ATRESIA

Tricuspid atresia is a congenital heart disease (CHD) where there is direct connection between the right atrium and right ventricle. A communication with the right ventricle is usually present and occurs through a ventricular septal defect.

The prevalence of tricuspid atresia has been between 2-3% of infants with CHD and 1-2% prenatally [1,2,3]. Due to the relative hypodevelopment of the right ventricle tricuspid atresia is frequently diagnosed in utero under the hypoplastic right heart syndrome and this may explain the minor incidence reported in prenatal series.

Diagnosis is usually performed by the 4 chamber view of the fetal heart [4] where an asymmetric development of the right ventricle is usually evident and a patent tricuspid valve is not identified (Fig. 1 [video 1](#))

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Figure 1: ([video 1](#)) A four chamber view of the fetal heart demonstrating tricuspid atresia. The right ventricle (RV) is hypoplastic and there is no opening tricuspid valve.

The associated ventricular septal defect is usually visible, even though it may be of variable size (Fig. 2 [video 2](#)).



Figure 2: ([video 2](#)) Evidence of the ventricular septal defect (VSD) from the 4 chamber view of the fetal heart.

Color Doppler allows to confirm diagnosis by showing the lack of flow velocities across the tricuspid valve, a single ventricular flow inlet across the mitral valve and an unidirectional left to right shunt across the ventricular septal defect (Fig. 3 [video 3](#)).

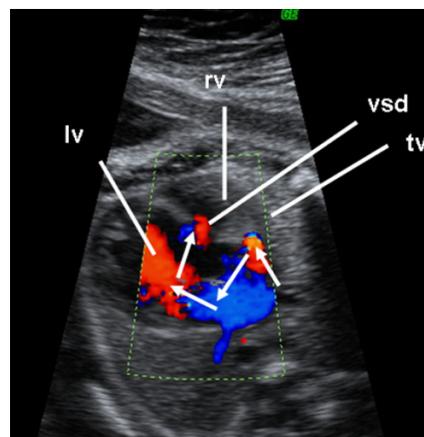


Figure 3: ([video 3](#)) Color Doppler shows absence of flow across the tricuspid valve (TV) and unidirectional shunt from the left ventricle (LV) to the right ventricle (RV) across the ventricular septal defect (VSD). Arrows indicate the abnormal blood flow direction due to the cardiac anomaly.

Intracardiac associated anomalies are frequent and in about 50% of the cases an obstructed right outflow is present. Arterial connections are usually concordant but transposed great arteries are present in about 20% of the cases. Less frequently an obstruction to the left ventricle outflow and anomalies of the pulmonary venous connections are present

Echocardiographic diagnostic criteria in tricuspid atresia are reported in Table 2.

Table 2: Sonographic features of tricuspid atresia

Small right ventricle
No opening tricuspid valve
Ventricular septal defect
No flow from right atrium to right ventricle
Unidirectional left to right shunt across the ventricular septal defect.

Tricuspid atresia is rarely associated with extracardiac anomalies. However it was reported a 5% incidence of chromosomal anomalies (trisomy 21, 18 deletion of chromosome 8 and 22q11 microdeletions). Unfrequent is also the association with extracardiac anomalies but an association with VATER e VACTEREL syndromes has been reported [1, 3].

Although diagnosis is usually possible by routine screening of fetal heart with the 4 chamber view (4) population-based studies have shown low detection rates, with more than 50% of tricuspid atresia remaining undetected on routine second-trimester fetal ultrasound examination [5-6].

The recent introduction of 4D ultrasonography to clinical practice provided an important advance in imaging technology. With a 4D acquisition of the fetal heart from the 4 chamber view it is possible to obtain several additional informations on the cardiac anatomy in tricuspid atresia.

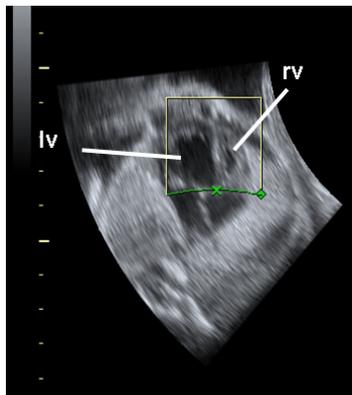


Figure 4: (video 4) Apical four chamber view of the fetal heart in a case of tricuspid atresia showing the disproportion between the left (LV) and right (RV) ventricle.

The advantages are:

a) look at the integrity of the septal wall using tomographic technique (7) (Fig. 5 video 5).

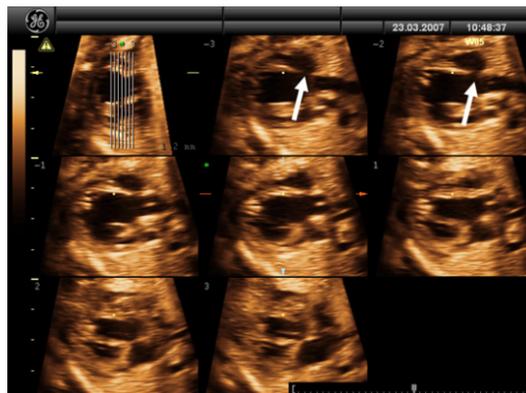


Figure 5: (video 5) Tomographic view of a case with tricuspid atresia allowing to evidence a small ventricular septal defect (arrows)

b) rendering the atrioventricular valve showing the tricuspid valve defect (Fig. 6 [video 6](#))

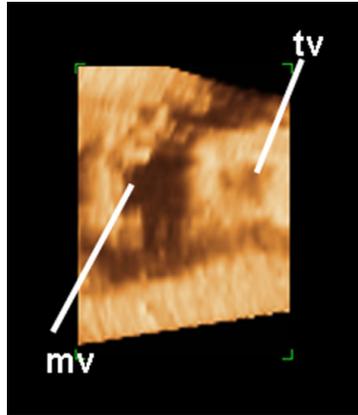


Figure 6: ([video 6](#)) From the apical four chamber view of the fetal heart shown in Fig. 4 the rendering function is used. The figure shows a coronal view of the atrioventricular valve during diastole showing the normal opening of the mitral valve (MV) and the “no opening” of the tricuspid valve (TV)

c) rendering the fetal heart to provide a better view of the right ventricle (RV) dimensions and contractility (Fig. 7, [video 7](#))

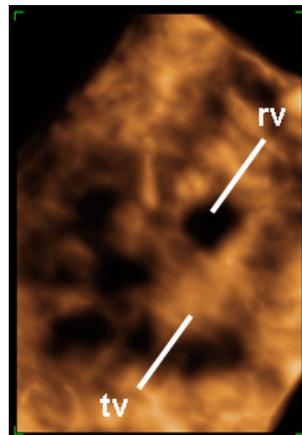


Figure 7: ([video 7](#)) Rendering of the 4 chamber view showing the deep size of tricuspid valve and the small right ventricle (RV)

d) providing a “live” view of the abnormal hemodynamic using inversion mode rendering techniques (Fig. 8 [video 8](#)) or rendering with color Doppler or B-flow.

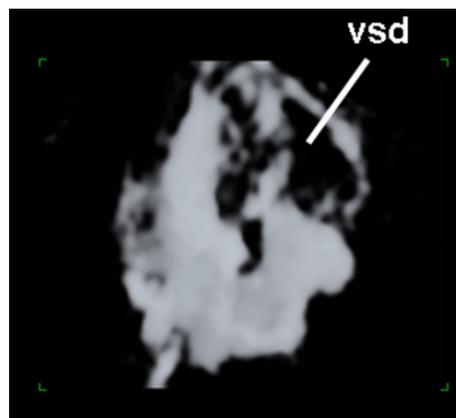


Figure 8: ([video 8](#)) Rendering of the 4 chamber view with inversion mode showing the abnormal hemodynamic characterized by the absence of flow across the tricuspid valve and the left to right shunt across the ventricular septal defect (VSD)

TRICUSPID DYSPLASIA AND REGURGITATION

This anomaly include a wide spectrum of diseases ranging from transient findings to a more severe forms characterized by thickened valve leaflets. Tricuspid regurgitation occurs in about 5% of second trimester fetuses [8] and it is generally induced by a relative immaturity of valve leaflets faced to high value of right ventricular afterload occurring during the first half of pregnancy. With advancing gestation the afterload falls due to reduction in placental impedance to flow, the valvular function improves and as a consequence the tricuspid regurgitation disappears.

Tricuspid dysplasia is distinguished from Ebstein's anomaly on the basis of the normal insertion of the valve leaflets to the ventricular wall.

Diagnosis may be difficult and it is usually performed by the 4 chamber view of the fetal heart were an enlarged right atrium with a tricuspid valve with increased echogenicity may be evidenced. By Color Doppler is possible to document the tricuspid regurgitation (Fig. 9 [video 9](#)).

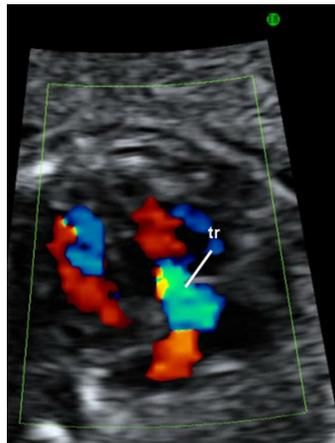


Figure 9: ([video 9](#)) Four chamber view of the fetal heart with color Doppler function superimposed showing tricuspid regurgitation (tr) in a second trimester fetus.

Pulsed or continuous Doppler allows to quantificate the severity of regurgitation. It is particularly important to quantify the duration of the regurgitation that may last all the systole (olossystolic) or its initial part (so called trivial) (Fig. 10).

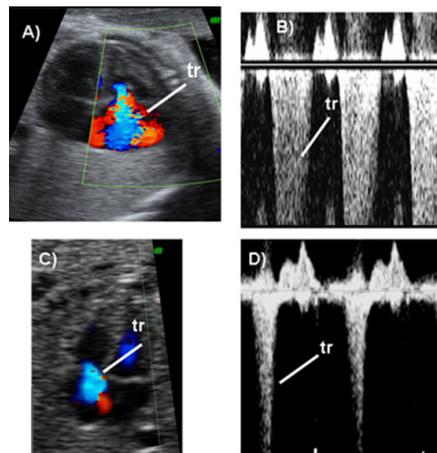


Figure 10: Quantification of tricuspid regurgitation (tr) with color and pulsed Doppler techniques. In panel A and B a case with olossystolic regurgitation and C and D a case of "trivial" regurgitation.

In order to avoid a misleading diagnosis of trivial regurgitation instead due to signals generated by physiological valve closure a duration > 70 msec is considered diagnostic [8].

Diagnosis of tricuspid regurgitation at 11+0 to 13+6 weeks of gestation is of clinical interest since 60% of fetuses with trisomy 21 and 30% of fetuses with trisomy 18 have this anomaly [9]. In presence of a normal karyotype (5% of euploid fetuses have this finding) tricuspid regurgitation may be an early marker of more complex CHD (10) (Fig. 11 [video 10](#))

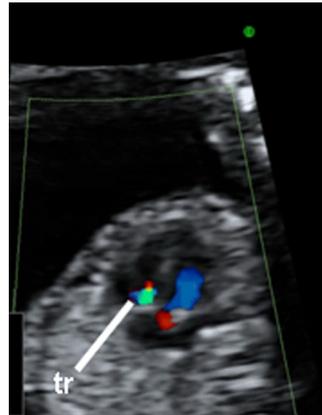


Figure 11: ([video 10](#)) Four chamber view of the fetal heart with color Doppler function superimposed showing tricuspid regurgitation (tr) in a fetus at 12 weeks of gestation.

Associated anomalies other than aneuploidies (trisomy 21,18, 13 and 45 X) may include secondary pulmonary stenosis/atresia due to reduced anterograde flow in the right outflow tract-

4D ultrasound provides two major advantages:

a) rendering the tricuspid valve allowing the visualization of the thickened leaflets (Fig. 12 [video 11](#))

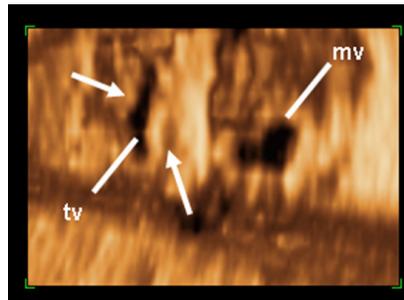


Figure 12: ([video 11](#)) Coronal view of the atrioventricular valve showing the normal the mitral valve (MV) and the thickened leaflets (arrows) of the tricuspid valve (TV)

b) exact localization of the color jet of the tricuspid regurgitation by multiplanar navigation of cardiac volume acquired with color Doppler. Indeed as shown in Fig. 13 the jet may be missed in one section (Fig. 13a [video 12](#)) and visualized only after the selection of the proper plane by multiplanar navigation (Fig. 13b [video 13](#)).

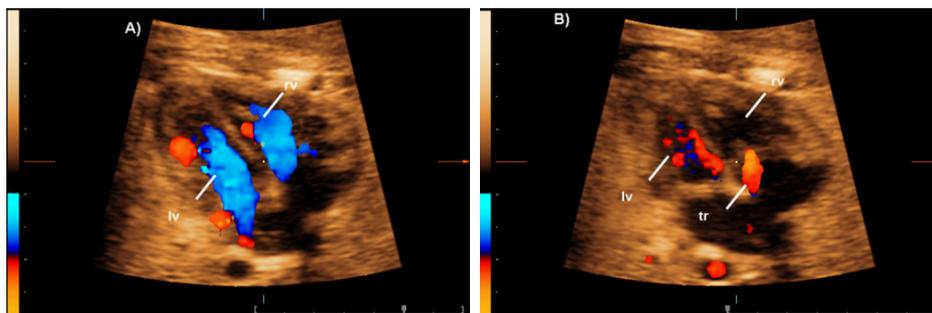


Figure 13: A ([video 12](#)) Four chamber view with color Doppler reconstructed from a 4D cardiac volumes showing apparent normal filling of left (LV) and right ventricle (RV). B ([video 13](#)) The plane is slightly scrolled cranially and tricuspid regurgitation (tr) is evidenced.

EBSTEIN'S ANOMALY

Ebstein's anomaly is a rare entity with a prevalence of 0.3-0.5% of CHD [1, 11]. Ebstein's malformation is a CHD in which the septal and mural (posterior) leaflets of the tricuspid valve are displaced downward into the inlet portion of the right ventricle. Tricuspid valve is always incompetent and there is a right atrium enlargement. The right ventricular size is reduced as its function is inversely related to its dimension [11].

Diagnosis is performed by cardiomegaly with dilatation of right atrium, low insertion of the septal and mural leaflets while the anterior usually appears moving in an abnormal way (spinnaker effect) (Fig. 14 [video 14](#)).

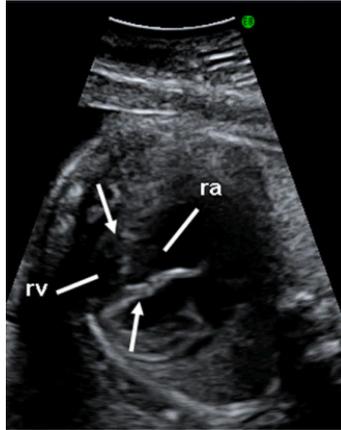


Figure 14: ([video 14](#)) Four chamber view of the fetal heart showing an enlarged right atrium (RA), the low insertion (arrows) of the septal and posterior leaflets in the right ventricle (RV) and the “spinnaker effect” ([video 14](#)) and the anterior leaflet.

At color Doppler a massive regurgitant jet is evident (Fig. 15 [video 15](#)).

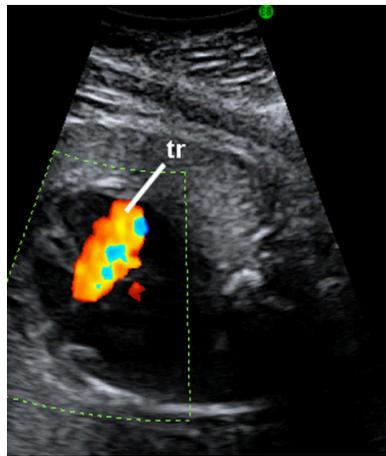


Figure 15: ([video 15](#)) Some fetus of Fig. 14 on color Doppler a severe tricuspid regurgitation is shown.

Intracardiac associated anomalies are frequent and include anatomic or functional (due to reduced anterograde flow) pulmonary stenosis-atresia, restrictive foramen ovalis, atrioventricular and ventriculo-arterial discordance [11]. The presence of associated anomalies affects the prognosis.

The presence of chromosomal anomalies is unusual but Ebstein's disease can be associated with Apert's, Noonan's and CHARGE syndromes [11].

4D echocardiography has been shown to be useful in predicting the results of surgical valvuloplastic [12]. Indeed it is possible to:

a) obtain an assessment of the commissures and leaflet surfaces by rendering the tricuspid valve (Fig. 16 [video 16](#)).

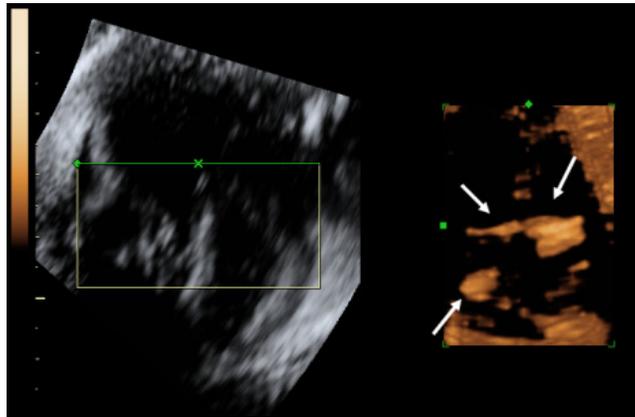


Figure 16: ([video 16](#)) Coronal view of the tricuspid valve in Ebstein' disease showing the rendering and the relationship of dysplastic leaflets(arrows)

b) providing a “live” view of the abnormal hemodynamic using rendering with color Doppler by showing the amount of retrograde and anterograde perfusion of the right ventricle (Fig. 17 [video 17](#)).

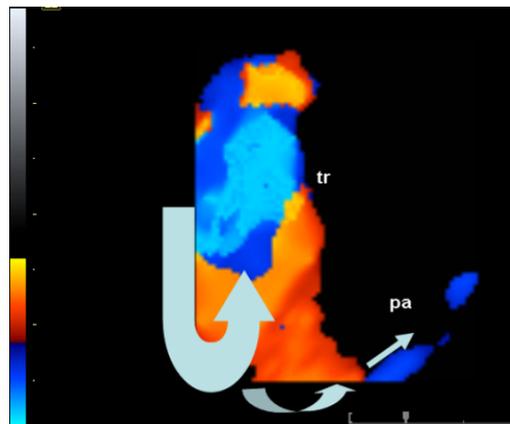


Figure 17: ([video 17](#)) Color Rendering of right ventricle hemodynamics in Ebstein's anomaly showing the prevalence of retrograde flow with tricuspid regurgitation (tr) and the marked reduced anterograde flow in pulmonary artery (pa)

c) allowing an absolute volumetric quantification of the functional ventricle and enlarged atrium. This may be obtained manually on acquired cardiac volumes or using an semiautomatic segmentation approach of cardiac cavities (13,14) as shown in Fig. 18.

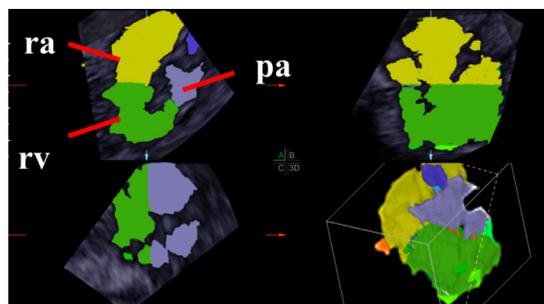


Figure 18: Semiautomatic segmentation of the right heart showing the enlarged right atrium (ra yellow), the small right ventricle (rv green) and pulmonary artery (pa grey) outflow shown in the 3 orthogonal planes and rendered. (3D).

PULMONARY STENOSIS

Isolated pulmonary stenosis represent 0.8% of CHD [1] and covers a wide spectrum in terms of presentation and severity and must be distinguished from critical stenosis and atresia of the valve.

The 4 chamber view is usually normal and diagnosis is frequently missed in screening program. Right ventricular outflow tract show the sonographic features shown in Table 3.

Table 3: Sonographic features of isolated pulmonary stenosis.

Restricted pulmonary valve leaflets
Dilatation after stenosis (occasionally)
Turbulent flow across the PA valve
Increased Doppler velocity

An example of isolated pulmonary stenosis is reported in

Fig. 19 and [video 18](#).

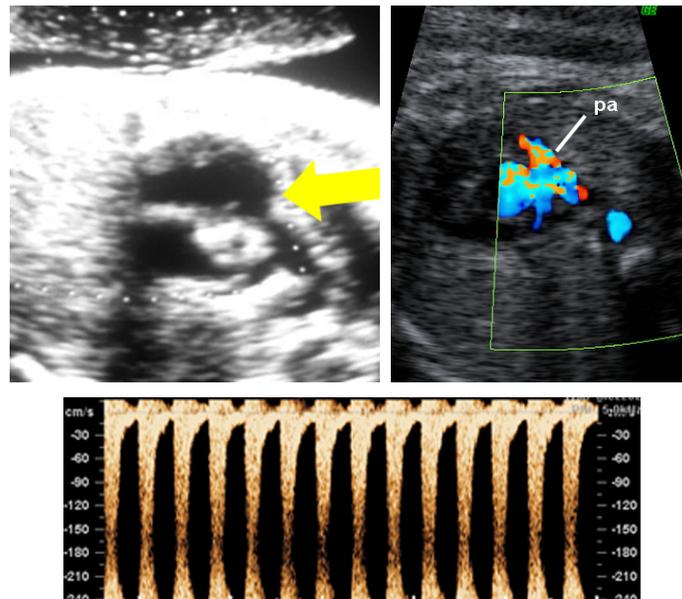


Figure 19: ([video 18](#)) Short axis view of the fetal heart showing the narrowing of the pulmonary artery (pa) (arrow upper panel) and the turbulent flow (middle panel) and the increased flow velocities at pulsed Doppler (lower panel).

Pulmonary stenosis can occur as a part of Noonan's, Alagille's and Williams syndromes and it is unfrequently associated with chromosomal anomalies.

In isolated pulmonary stenosis 4 D echocardiography is useful in evaluating the thickened restricted pulmonary valve leaflets (Fig. 20 [video 19](#)).

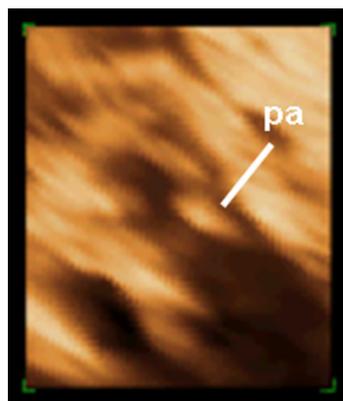


Figure 20: ([video 19](#)) rendering of the short axis view of the fetal heart showing the thickened pulmonary artery (PA) leaflets.

PULMONARY ATRESIA

Pulmonary atresia and critical stenosis with intact ventricular has an incidence of about 10%, of all CHD [1]. There is continuity between the two forms and critical stenosis may develop in an atresic valve. According to tricuspid valve there are two different anatomic variants. In absence of tricuspid regurgitation an hypoplastic right ventricle is present while in presence of tricuspid regurgitation the dimension of the right ventricle are normal and the right atrium enlarged. The former situation is more frequent and accounts for about the 70% of cases.

In Fig. 21 ([video 20](#).) an example with hypoplastic right ventricle is reported.

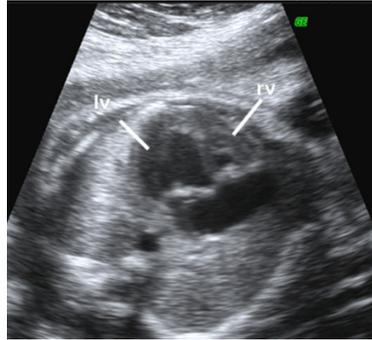


Figure 21: ([video 20](#)) Four chamber view showing an hypoplastic right ventricle (rv) when compared to the left ventricle (lv).

Color flow mapping confirms the abnormal filling of the ventricle (Fig. 21 [video 21](#))

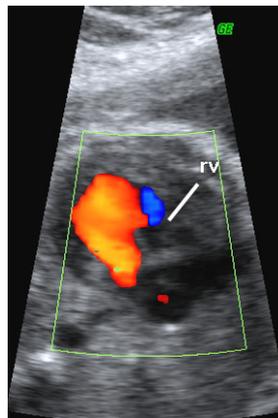


Figure 22: ([video 21](#)) Four chamber view with color Doppler function superimposed showing the absent filling of the right ventricle (rv).

The pulmonary artery can vary in size from being near normal to severe hypoplastic and the valve leaflets are thickened and do not disappear during systole (Fig. 23 [video 22](#)).

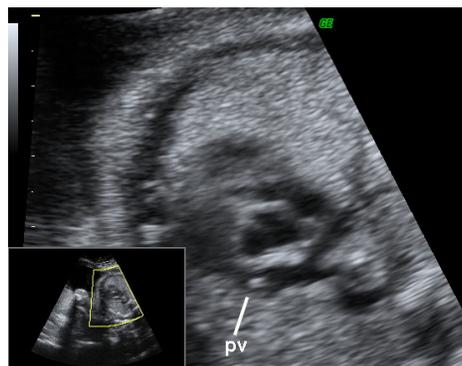


Figure 23: ([video 22](#)) A short axis view of the fetal heart showing a pulmonary trunk severely hypoplastic. The pulmonary valve (pv) is also demonstrated and appears to thickened and dysplastic.

Minimal or no forward flow is present across the pulmonary valve or in main pulmonary artery and reverse flow is the ductus arteriosus (ductal dependency).

Ductal dependency may be evidenced from the 3 vessel view of the fetal heart showing flow in the pulmonary trunk in the opposite direction to the flow in the aorta (Fig. 24 [video 23](#))

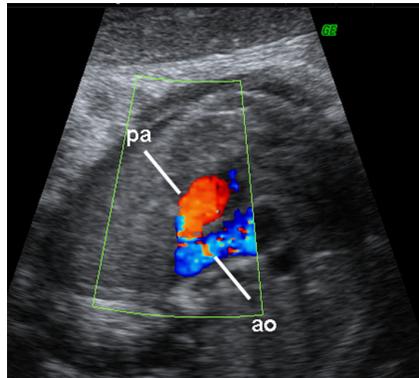


Figure 24: ([video 23](#)) 3 vessel view of the fetal heart in a fetus with pulmonary atresia. Color flow mapping shows that in the pulmonary artery (pa) the flow is directed from the spine toward the anterior chest wall, rather than being directed toward the spine as is normal and occurring in aorta (AO).

In presence of a concomitant anomaly of the tricuspid valve (valvular dysplasia or Ebstein's malformation) there is an increase of heart dimension mainly due to an enlargement of the right atrium (Fig. 25 [video 24](#)).

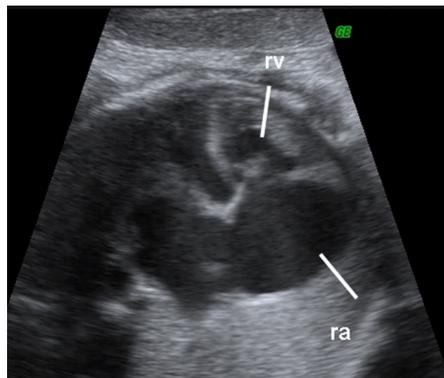


Figure 25: ([video 24](#)) Four chamber view of a fetus with pulmonary atresia and associated tricuspid dysplasia. The right atrium (ra) is enlarged and the right ventricle (rv) is hypertrophic and of reduced size.

The dimension of right ventricle can vary from severely hypoplastic to almost normal size. Color flow mapping allows to demonstrated marked tricuspid regurgitation as shown in Fig. 26 ([video 25](#)).

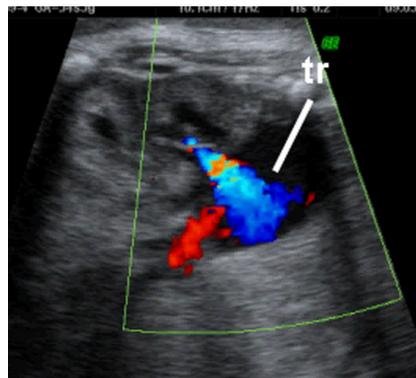


Figure 26: ([video 25](#)) Same fetus of Fig. 25 with color flow mapping showing the tricuspid regurgitation (tr).

Pulmonary atresia is not commonly associated with extracardiac anomalies or aneuploidies.

The outcome is affected by the possibility of postnatal treatment with two-ventricle repair that allows a good long term outcome. On the other end those who will end in an univentricular repair face a less optimistic long term prognosis.

Classical criteria of poor prognosis and subsequent univentricular repair are the presence of fibroelastosis in the right ventricle (Fig. 27 [video 26](#)) and the presence of sinusoids (coronary-right ventricle fistulas) (Fig. 28 [video 27](#)) [15, 16].

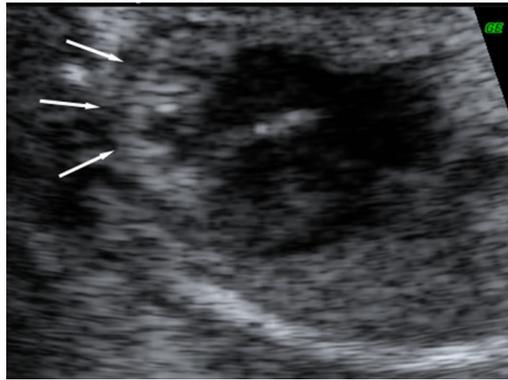


Figure 27: ([video 26](#)) Four chamber view of a fetus with pulmonary atresia. The right ventricle size are reduced, the myocardium appears trabeculated and with an increased echogenicity of the free walls (arrows) suggesting the presence of fibroelastosis.

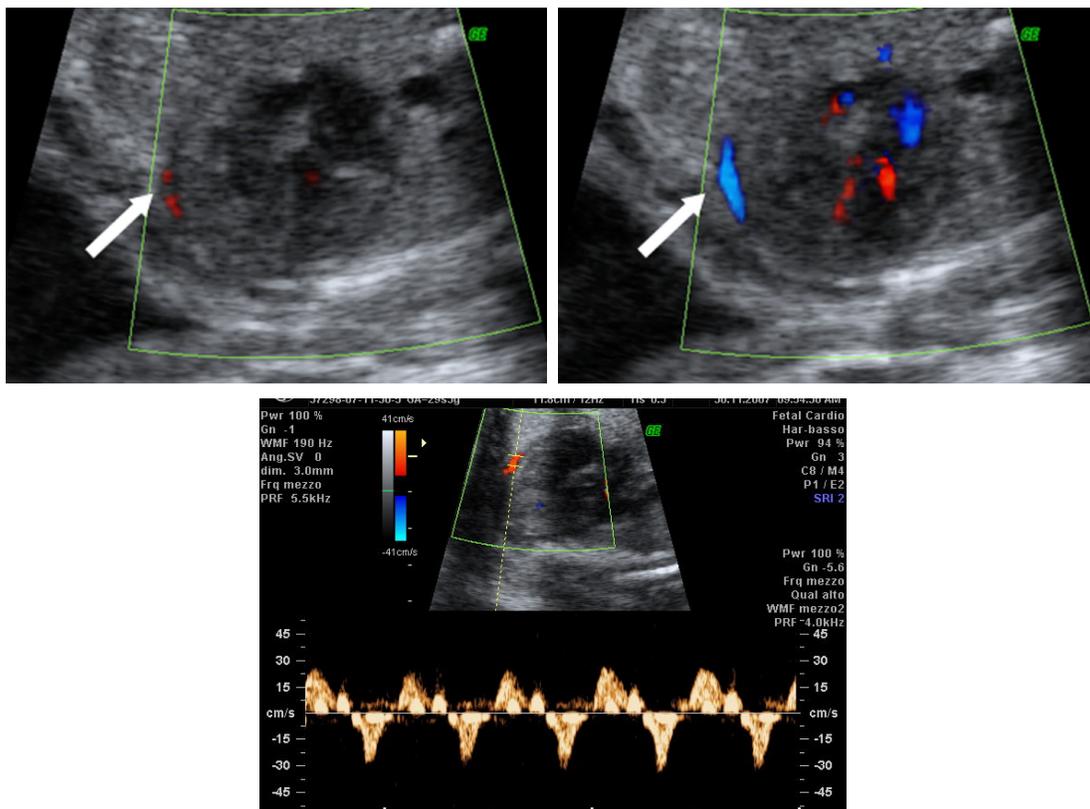


Figure 28: ([video 27](#)) Same fetus of Fig. 27 showing the presence of coronary right ventricle fistula (sinusoid). Color flow mapping shows the dilated right coronary artery (arrows) on the surface of right ventricle showing biphasic flow direction (retrograde flow panel A, forward flow panel B) and corresponding spectral Doppler tracing panel C).

More recently further prognostic criteria has been suggested as useful markers to predicts early in gestation the possibility of two-ventricle repairs such as the degree of reduction of ventricular size or changes in the ratios

between left and right ventricle size or atrioventricular valves and the presence absence of tricuspid regurgitation [16, 17]. Their clinical significance remains to be established

In fetuses showing poor prognosis intrauterine treatment has been suggested. The rationale of such approach is that the severity of the disease can further evolve in utero, and an early intervention may improve the outcome by altering the natural history of such conditions by performing fetal cardiac intervention intrauterine by balloon valvuloplastic or radiofrequency perforation [18]. The risk/benefit ratio of this approach is not yet established.

The advantages of 4D fetal echocardiography are similar to those already described for the other right heart diseases and may be summarized as follows:

- a) evaluation of the morphology and dynamics of the pulmonary artery (Fig. 20, [video 19](#)).
- b) evaluation of the tricuspid valve (Fig. 12 [video 11](#)) and its regurgitation.
- c) direct visualization of right ventricle morphology with rendering technique (Fig. 29 [video 28](#)).

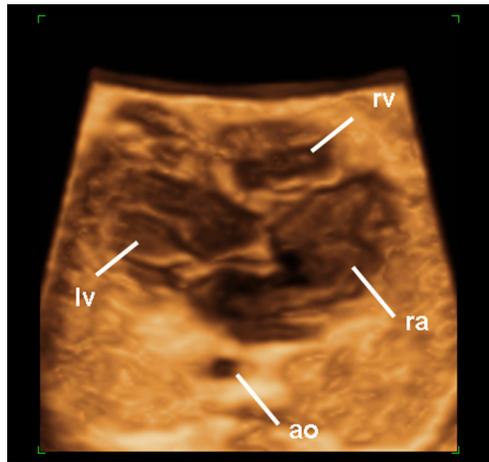


Figure 29: ([video 28](#)) Rendering of the fetal heart in a fetus with a hypoplastic right ventricle (RV). LV left ventricle, ra right atrium, ao aorta.

- d) providing a “live” view of the abnormal hemodynamic using inversion mode rendering techniques Fig. 30 ([video 29](#)).

e) evaluating ventricular volume, geometry and stroke volume (see also chapter 16). In particular the use of semiautomatic software for volume calculation [14] allows reliable measurements in short time interval compatible with clinical practice (Fig. 31).

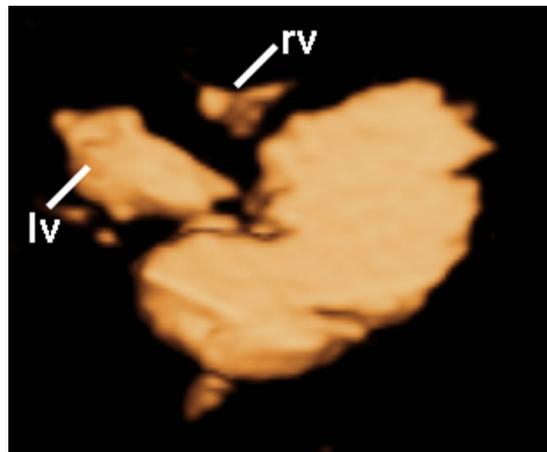


Figure 30: ([video 29](#)) Some fetus of Fig. 29, the small right ventricle (rv) is visualized with inversion mode. LV left ventricle.

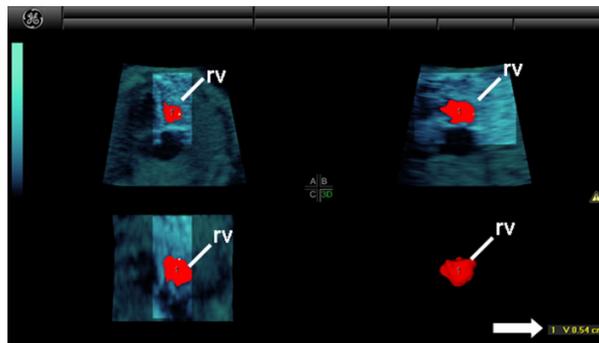


Figure 31: Example of quantification of ventricular volume in a fetus with a small right ventricle (rv) during systole using a semiautomatic system (sonoAVC see reference 14). The arrows indicates the absolute ventricular volume during systole.

It is also possible to obtain a simultaneous assessment of both ventricular volumes (Fig. 32 [video 30](#)) allowing to perform ratios between the size of the two ventricles.

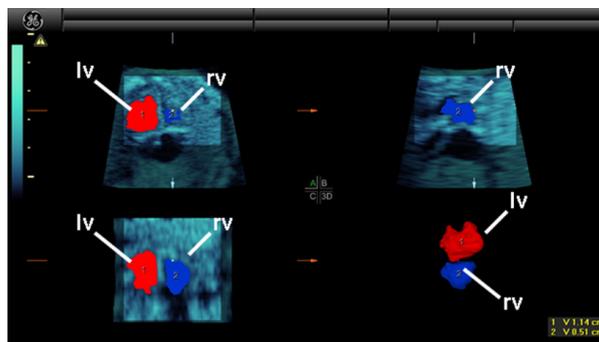


Figure 32: ([video 30](#)) Example of simultaneous assessment of right (rv blue) and left (lv red) ventricles using sonoAVC technique.

This approach has the potential to allow an absolute and reliable quantification of ventricular size and function (by assessing the stroke volume) thus allowing serial observations through pregnancy of such fetuses. This may allow to evidence the evolution of the CHD and identify fetuses that may benefit from intrauterine treatment or earlier delivery.

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