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TWO-DIMENSIONAL (2D) AND FOUR-DIMENSIONAL (4D) ULTRASOUND WITH SPACIO-TEMPORAL IMAGE CORRELATION (STIC) IN DIAGNOSIS OF RIGHT HEART ABNORMALITIES

DVODIMENZIONALNI (2D) I ČETVERODIMENZIONALNI (4D) ULTRAZVUK SA SPACIOTEMPORALNOM SLIKOVNOM KORELACIJOM (STIC) U DIJAGNOZI ABNORMALNOSTI DESNOGA SRCA

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Original paper

Key words: Ebstein's anomaly, hypoplastic right ventricle, pulmonary stenosis

SUMMARY. Four cases of right heart anomalies are presented, where it was used 4D and STIC for improving detection and complete evaluation of anomaly. Prenatal diagnosis being associated with better obstetric management. Fetal echocardiography was performed with a 4D ultrasound system (Voluson E8, GE Healthcare, Kretztechnik, Zipf, Austria) equipped with a 4–8 MHz transabdominal transducer and STIC. All pregnant women underwent comprehensive evaluations with STIC, these views were stored and were shared for expert review, interdisciplinary consultation and parental counseling.

Izvorni članak

Ključne riječi: Ebsteinova anomalija, hipoplastična desna komora, pulmonarna stenoza

SAŽETAK. Prikazana su četiri slučaja s anomalijom desnog srca u kojih su rabljeni 3D ultrazvuk i STIC, koji poboljšavaju otkrivanje i točno vrednovanje anomalije. Prenatalna dijagnoza je potrebna za bolji opstetrički postupak. Fetalna ehokardiografija je obavljena 4D ultrazvučnim sustavom (Voluson E8, GE Healthcare, Kretztechnik, Zipf, Austrija), koji sadrži 4–8 MHz transabdominalnu sondu i STIC. Sve su trudnice bile podvrgnute opsežnom istraživanju pomoću STIK-a, a dobivene slike su bile spremljene i za konzultaciju i mišljenje eksperta.

Introduction

The characterization of complex CHD is challenging in the prenatal period representing one of the most difficult issues in pediatric cardiology and STIC may be considered to be one of the best tools available for optimizing the consultation of the maternal-fetal medicine expert with the cardiac surgeon and the pediatric cardiologist.[1] The integrated STIC software permits a new approach for clinical assessment of the fetal heart. The duration of the acquisition can be adjusted by the examiner (7.5, 10, 12.5 or 15 s) and the acquisition angle sweep ranges from 15° to 40°. The system analyzes information that could then be displayed and presented in rendering or multiplanar mode.[2] The examiner can navigate within the heart, and produce all of the standard images necessary for a comprehensive diagnosis. A reference dot that can be placed anywhere on the image by the physician, could help in the visualization of the spatial orientation of the three orthogonal planes. During live 2D real-time, the examination of the outflowtracts may be difficult, because of fetal orientation and movement. Using STIC, that has the ability to rotate the data volume around a single reference point, it is easy to reveal the outflow tracts. STIC enables the reconstruction of a 3D rendered image that contains depth and volume which may provide additional information that is not available from the thin 2D image slices. It also present gray-scale or color Doppler information or both for diastole and systole. The visualization of color Doppler information can also be changed gradually by manually changing the 'color threshold' button. STIC has the potential to shorten the evaluation time, when complex heart defects are suspected, permits offline analysis, and improved ability to describe to the patient the fetal cardiac anatomy, when a malformation is suspected. The major limitation of the STIC is in the advanced gestational age, the increased mineralization of the ribs and the sternum and the necessity of having a volume that includes the whole fetal thorax, from the transverse abdominal view to the upper mediastinum.

Case reports

Case 1

A 34-year-old Caucasian primigravida was referred to our unit at 34 weeks gestation after routine dating ultrasound, that revealed the heart anomaly. The patient had no prenatal care prior to this time. The family history was negative for congenital anomalies or genetic abnormalities and the patient denied exposure to drugs or toxins. Our ultrasound examination revealed a single fetus with BPD, HC, AC and FL measurements ade-

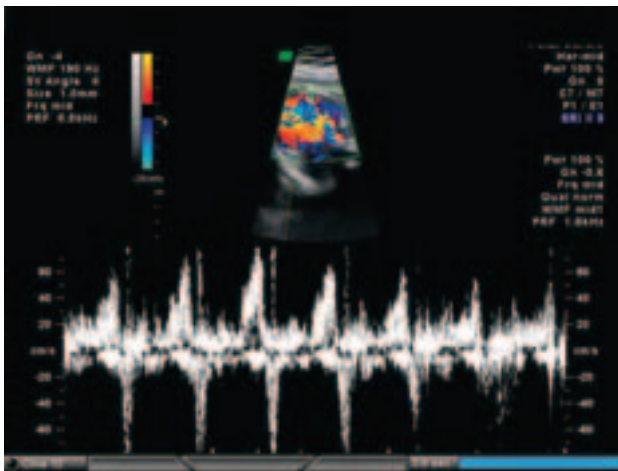*Figure 1.* Right atrium enlarged
Slika 1. Desni atrij je povećan

Figure 4. Ebstein anomaly
Slika 4. Ebsteinova anomalija

Figure 2. Retrograde flow in right atrium
Slika 2. Povratni tijek u desnom atriju
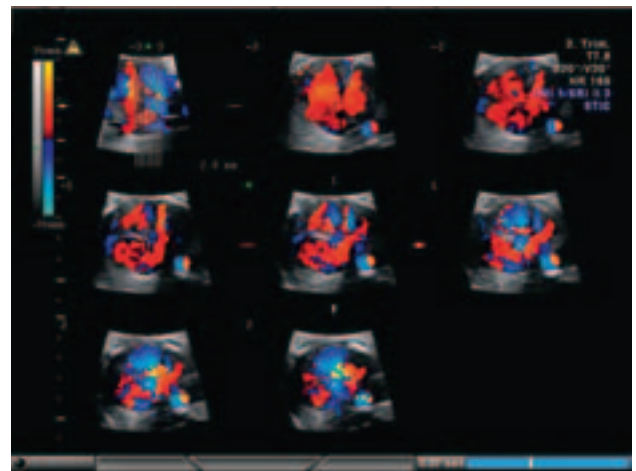
Figure 5. STIC tricuspid insuficiency
Slika 5. Trikuspidalna insuficijencija STIK-om

After STIC acquisition, we noticed normal ventriculo-arterial connections and signs of fetal hydrops: pericardial effusion, pleural effussion. Doppler echocardiography had demonstrated the retrograde flow into the right atrium *(figure 2)*. Pulmonal artery was dilated and blood flow suggested pulmonary stenosis. Also STIC had helped us to find that there was a reverse flow in ductus arteriosus *(figure 3)*. Diagnosis was Ebstein disease, severe form. After consulting with paediatrician and pediatric surgeon, it was decided delivery. A female infant of 2400 g, Apgar 8, had been resuscitated by paediatrician (administration of prostaglandin to mantain ductal patency, bicarbonate infusion for metabolic acidosis and treatment of arrhytmia were necessary) and unfortunately, didn't survive neonatal period, due to congestive heart failure.

Case 2

A 31-year-old woman gravida 2 para 1, presented to the outpatient clinic for ultrasound exam. She was 37 weeks gestation and her medical history revealed that
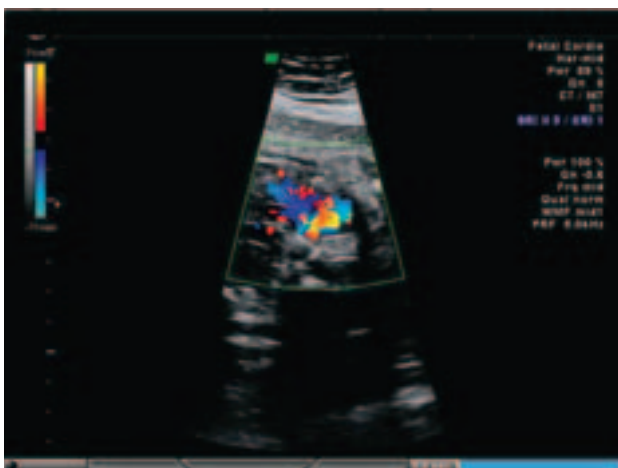

Figure 3. Reverse flow in ductus arteriosus
Slika 3. Obrnuti tijek u duktusu arteriozusu

quate with 34 weeks gestational age. We didn't observed any anomalies, except the heart. On subcostal four chamber view it was an enlarged right atrium *(figure 1)*.
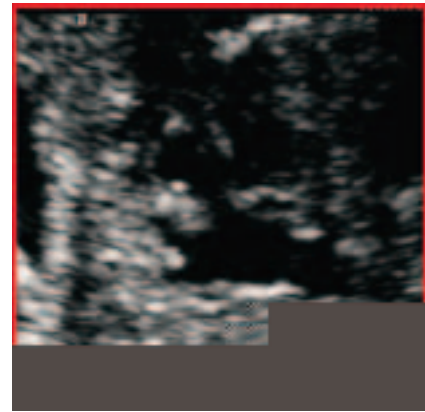
Figure 6a. Right ventricle hypoplasia
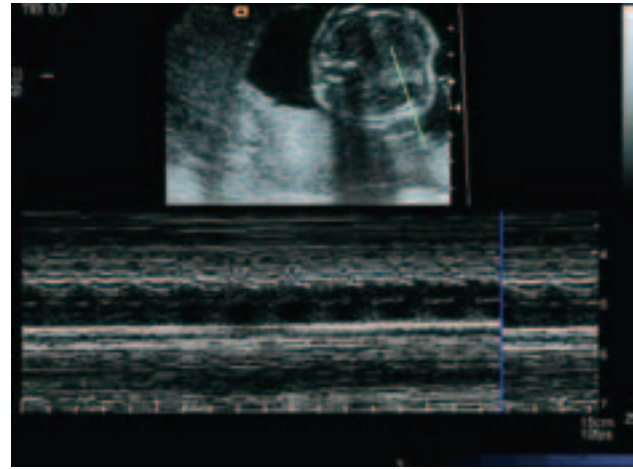*Slika 6a.* Hipoplazija desne komore

Figure 6b. Right ventricle hypoplasia mode M
Slika 6b. Hipoplazija desne komore načinom M

pregnancy was achieved after treatment for infertility. 13 weeks scan and bitest were normal, also 24 weeks scan was normal and amniocentesis was not performed. There no risk factors were identified. The right atrium on the four chambers image was large, the posterior and septal leaflets of tricuspid valve were displaced, and there was an enlarged right atrium on four chamber view *(figure 4)*. Pulmonary artery and aorta were normal and had normal ventriculoarterial connections. After STIC acquisition, we were able to see that there were not other cardiac anomalies associated. Noncardiac anomalies were not associated. Color Doppler revealed tricuspid insufficiency *(figure 5)* but there no other sign of cardiac dysfunction at 37 weeks of pregnancy. Diagnosis was Ebstein disease, mild form. After consulting with pediatrician, we sent this case to be delivered in an obstetric unit with a cardiac pediatric surgery department. The prognosis of this case was better, due to a complete evaluation with the use of STIC, and at first sign of cardiac decompensation and pulmonary maturity, it was decided to deliver the baby. He adapted well to extrauterine life after intensive neonatal care.

Case 3

A 25-years-old primigravida, 24 weeks gestation, was referred to our unit for second trimester ultrasound screening. There were no exposure to drugs or toxins or history of cardiac anomalies in her medical records. The ultrasound examination showed us a single fetus with biometric measurements corresponding to 24 weeks gestation. With exception of the heart, there are no other anomalies. The right side of the heart on the four chambers image was very small, especially the right ventricle *(figure 6a)*. After STIC acquisition we noticed atrioventricular and ventriculo-arterial concordance, the tricuspid valve that was small, but patent. Doppler echocardiography has demonstrated the lack of antegrade flow into the right ventricle. In M mode examination these data are confirmed *(figure 6b)*. We have performed and amniocentesis with fetal karyotype analize, that

didn't reveal anything abnormal. Diagnosis was hypoplasia of the right ventricle. Because it was a severe anomaly, after counseling, the parents have decided upon therapeutical abortion. The infant was female, dead during abortion. The results of autopsy confirmed pathological findings detected by ultrasound, the placenta and umbilical cord were unremarkable.

Case 4

A 31-year-old-woman presented to the outpatient clinic for routine ultrasound exam at 38 weeks gestation. First trimester and second trimester ultrasound screening were normal and there were no exposure to drugs or toxins or history of cardiac anomalies in her medical records. Four chamber view was normal. After STIC acquisition, looking at the great vessels, we observed a dilated pulmonal artery with turbulence flow of 70 cm/s, a normal aorta and no other cardiac anomaly. Extracardiac ultrasound exam was normal. We followed up ultrasonography and we didn't notice any sign of cardiac dysfunction. Diagnosis was pulmonary stenosis. She delivered at 39 weeks gestation, a female infant 2900 g, Apgar 9. Peadiatric echocardiography confirmed diagnosis. Baby adapted well to extrauterine life.

Discussion

4D ultrasound and STIC permit in normal fetuses and those with congenital heart disease (CHD), after volume acquisition from four chamber view, the examination of the fetal heart using a tomographic approach and the rendering of cardiovascular structures to visualize the relationships, size and course of the outflow tracts.[3] In cases of CHD, the advantage of 3D/4D systems is the possibility to store examination volumes for later analysis, away from time constraints.[4]

Definition, pathology and hemodynamics

Tricuspid valve abnormalities are: Ebstein's anomaly, tricuspid valve dysplasia and unguarded tricuspid valve.

Ebstein anomaly is defined as displacement of the septal and posterior leaflets of the tricuspid valve from their normal location at the atrioventricular junction, into the right ventricle, creating an »atrialized« portion of this ventricle. In *tricuspid valve dysplasia*, the tricuspid valve is not displaced, but has nodular thickening of leaflets that do not coapt, often with abnormal chordae and papillary muscles. *Unguarded tricuspid valve* is characterised by little if any tricuspid valve tissue. In all defects could be found right ventricular dysfunction.[5] Some degree of tricuspid insufficiency is always present in Ebstein anomaly, the most severe form has tricuspid atresia and is named the imperforate type. The output of the right ventricle could be diminished and the pulmonary circulation supplied by the ductus arteriosus in a retrograde manner. The very large heart could lead to lung hypoplasia. Sometimes, there is severe cardiac dysfunction with massive cardiomegaly, hydrops fetalis and tachyarritmia. Lithium exposion in the first trimester, determine a four hundred increasing in the incidence of these anomalies. *Hypoplastic right ventricle* represent atresia of the pulmonary valve, usually associated with hypoplastic pulmonary artery and right ventricle. *Pulmonary stenosis* definition is the obstruction in the output of the right ventricle, due to an anatomic lesion of the pulmonary valve, pulmonary arteries or right ventricular infundibulum.

Embriology. In Ebstein anomaly there is a faulty delamination process in the 12–16 weeks fetus, resulting in abnormal apical insertion of septal and posterior leaflets of tricuspid valve, with the origin in the inner lyers of the right ventricular inlet; on the contrary the anterior leaflet that has a muscular origin, it is formed much earlier in development and it is in normal position. Hypoplastic right ventricle is the result of an inadequate blood supply being able to enter right ventricle, that occurs after cardiac septation. Possible, the embryologic basis for pulmonary stenosis is a maldevelopment of the distal part of the bulbus cordis.[6]

Occurence rate. These anomalies are less common, representing 3–7% of congenital heart diseases.

Associated anomalies. A wide variety of cardiac, craniofacial and digital abnormalities are associated with Ebstein anomaly. More frequently present are: pulmonary stenosis, pulmonary atresia, atrial septal defect, ventricular septal defect, corrected transposition of the great arteries, tetralogy of Fallot, coarctation and supraventricular tachycardia. Hypoplastic right ventricle is also associated with many cardiac and extracardiac anomalies, and particulary with maternal rubella and enterovirus infection. Pulmonary stenosis is associated with right ventricular hypoplasia, atrial and ventricular septal defects, anomalous pulmonary venous connection, Noonan and Williams syndrome and trisomy 13.

Ultrasound diagnosis

In Ebstein anomaly an enlarged right atrium on subcostal four chamber view is the first sign of congenital disease. Some authors determined septal leaflet disease measuring the distance between the septal leaflet of tricuspid valve and the anterior leaflet of mitral valve.[7] Signs of tricuspid dysplasia are nodularity and valvular thickening. Color Doppler reveal tricuspid insufficiency that could lead to arrhytmia and hydrops fetal. The pulmonary artery is frequently small. In hypoplastic right ventricle there is a small right ventricle on four chamber view, atrio-ventricular and ventriculo-arterial concordance, normal left heart. The tricuspid valve could be small, but it is patent. Color Doppler will demonstrate either minimal or no flow across the tricuspid valve. Ultrasound diagnosis of pulmonary stenosis is based on poststeenotic dilatation of the pulmonary artery, a small right ventricle and an increased velocity distal to the valve.

Treatment

Ebstein anomaly is very rarely associated with chromosomal abnormalities, so fetal karyotip is not indicated. The fetus is monitorised for cardiomegaly, hydrops and tachyarrytmia. Delivery in a tertiary center with intensive neonatal care and pediatric surgery should be arranged. Administration of prostaglandin to mantain ductal patency, bicarbonate infusion for metabolic acidosis and treatment of arrhytmia may be necessary. Surgical procedures are: Blalock Taussig shunt, Glenn anastomosis and catheterization with radiofrequency ablation. Infants with hypoplastic right ventricle are ductus dependent, termination of pregnancy should be offered. Surgical neonatal procedures in these cases include: pulmonary valvulotomy, Blalock Taussig shunt, or even cardiac transplantation. Surgical treatment for pulmonary stenosis include: open transpulmonary valvotomy, transventricular valvotomy and percutaneos balloon valvuloplasty.

Prognosis

In Ebstein anomaly overall prognosis is poor. Fetus could develop hydrops from tricuspid valve regurgitation and/or right heart failure and perinatal death. Morphological features associated with severe outcome are: marked atrial enlargement, dysplasia of the atrialized portion of the right ventricle, left ventricular compression. Other cardiac and noncardiac anomalies associated determine a worse prognosis. As in Ebstein's anomaly, in hypoplastic right ventricle the prognosis is severe when cardiomegaly and important tricuspid regurgitation are present. Patients with mild pulmonary stenosis are asymptomatic, the prognosis is worse when hydrops fetalis and congestive heart failure develop in utero.

Conclusion

We present these cases, because right heart anomalies are relative rare conditions (1/15.000 live birts), representing about 3–7% of congenital heart anomalies. 4D and STIC could help for a complete diagnosis of the

cardiac anomalies, a much better evaluation of prognosis and a better obstetric management.

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