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Right ventricular function evolution with pregnancy in repaired Tetralogy of Fallot

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ABSTRACT

This case illustrates the evolution of right ventricular 3D area strain during pregnancy in a patient with repaired Tetralogy of Fallot. This report highlights impairment in RV function with pregnancy, suggesting the importance of pre-pregnancy RV systolic function assessment, especially using 3D echocardiography.

Pregnancy is responsible for marked physiological changes in women: increased plasma volume and cardiac output essentially. These changes lead to an increase in the heart size, mainly through dilatation that might be significant in patients with congenital heart disease. Pregnancy in Tetralogy of Fallot (TOF) patients is considered as class II risk according to the modified WHO scoring system¹. Pre-pregnancy counseling includes a thorough evaluation of right ventricular (RV) function and assessment for pulmonary valve replacement. Up to 12% of patients will present either arrhythmia or heart failure. Data regarding RV evolution during pregnancy are conflicting^{2,3} and none of these magnetic resonance studies assess changes during pregnancy. Echocardiographic data, especially in 3D can provide insights into the right ventricular function.

We hereby present the evolution of a 35-years-old patient with repaired TOF before and during her fourth pregnancy. The patient underwent primary palliation at the age of 2 months and corrective surgery with trans-annular patching at the age of 3 years old. Pre-pregnancy counseling included a complete clinical assessment with a 24-hour ECG recording and a cardio-pulmonary exercise test. Cardiac magnetic resonance imaging revealed severe pulmonary regurgitation (regurgitant fraction 50%) with preserved RV function (RV end-diastolic volume 103 mL/m²; RV ejection fraction 52%). Echocardiography confirmed the severe pulmonary regurgitation associated with mild pulmonary stenosis (maximum pressure gradient 40mmHg). TAPSE was 17 mm, peak tricuspid valve S was 6.3 cm/s. Three-dimensional echocardiographic analysis using TomTec 4D RV Function 2.0 indicated a pre-pregnancy RV ejection fraction of 50% with an RV end-diastolic volume of 84 mL/m². Global RV area strain; as determined by the area change of each mesh triangular element and calculated using VTK (v7.10, Kitware, New York, US)⁴ was -26.6% (global RV longitudinal strain was -11.0% and circumferential strain was -16.9%). The patient was evaluated in our center a year later when she was 32 weeks pregnant. Her clinical examination revealed no

sign of heart failure. No increase in QRS duration was noted. The ultrasound revealed severe pulmonary regurgitation with lower RV ejection fraction (40%) using 3D echo and RV dilatation (RV end-diastolic volume 110 mL/m²). The RV-pulmonary artery maximum pressure gradient was 44 mmHg. TAPSE and peak tricuspid valve S did not change significantly. Global RV area strain was -11.5% at 32 weeks (global RV longitudinal strain -1.9%, circumferential strain -9.2%). The patient went into spontaneous labor at 39 weeks 2 days gestation and vaginal delivery occurred uneventfully under epidural anesthesia.

Figure 1 illustrates RV area strain changes between pre-pregnancy counseling and a year later, when the patient was 32-weeks pregnant. Area strain is represented at end-systole over the RV mesh exported from the quantification software. Despite almost no change in TAPSE and peak tricuspid valve S, this report illustrates a substantial alteration in RV systolic function with reduced RV ejection fraction, RV dilatation and impaired RV area strain. RV area strain changes predominate in the inlet and outlet part of the RV. As previously reported with systolic function⁵, those regions are probably the most sensitive to preload changes and myocardial stress.

Recent data seem to indicate that patients with mild RV dilatation are less likely to experience significant deterioration as a consequence of pregnancy, whereas patients with larger right ventricles are more likely to dilate significantly their RV. These data might help pre-pregnancy counseling in TOF and our report illustrates the important changes in RV strain occurring with pregnancy (even accounting for the variability of 3D echocardiography, including the inherent inter, intra-observer and inter-loop variability) as well as the uselessness of TAPSE and peak tricuspid valve S to assess RV function in TOF. However, conflicting data exist regarding RV systolic function. New tools such as 3D RV strain assessment seem promising to assess finer changes in RV function, especially during

pregnancy. Further studies and reports are needed to investigate long-term detrimental effects of pregnancy in TOF.

DISCLOSURES : None

FIGURE AND VIDEO LEGENDS:

Figure 1: RV area strain changes between pre-pregnancy counseling (baseline) and a year later (32-weeks pregnant). Area strain is represented at end-systole over the RV mesh. The third column represents area strain changes with pregnancy.

Video 1: RV area strain represented over the RV mesh along the cardiac cycle, before pregnancy (baseline).

Video 2: RV area strain represented over the RV mesh along the cardiac cycle, during pregnancy (32-weeks).

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SUMMARY

Pregnancy is responsible for marked physiological changes, particularly in congenital heart disease patients. Our report illustrates the changes in RV strain occurring with pregnancy in a patient with repaired tetralogy of Fallot. 3D strain assessment seems promising to finely assess changes in RV function, especially during pregnancy with increased RV preload.

Figure 1
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