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Pulmonary arterial hypertension diagnosed during pregnancy – echocardiography as a tool for management

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Introduction: Despite advanced therapies for pulmonary arterial hypertension (PAH), maternal mortality in women with PAH and their offspring remains high (30-56% and 11-28%) and is especially high during the post-partum period[1,2].

Case report: 39-years-old woman was presented in 24th week of pregnancy with moderate dyspnea and cyanosis without peripheral edema. ECG showed right ventricular strain and NT-proBNP was 1300 ng/L. Echocardiogram showed severe precapillary pulmonary hypertension with systolic pulmonary pressure (PAP) 103 mmHg, reduced stroke volume (SVI 29 ml/m^2), normal cardiac output (CI 2.6 L/min/m^2) and mildly reduced systolic function of the right ventricle (FAC 30%, PMI TDI 0.8) with normal central venous pressure (CVP) 3 mmHg. Right heart catheterization confirmed echocardiographic hemodynamic measurements (mean pulmonary pressure 61 mmHg, pulmonary vascular resistance (PVR) 9.4 WU) and the vasoreactivity test was negative. Epoprostenol was initiated, titrated up to 22 ng/kg/min and on 30th week planned caesarean section was performed with NO inhalations, noradrenalin and dobutamine. Fortunately, standby supportive therapy - ECMO and high urgency lung transplantation, was not needed. The baby did well. The patient was extubated on the same day and sildenafil was added. The patient was discharged after 4 weeks. After 2 months epoprostenol was switched to treprostinil up to 26 ng/kg/min and macitentan was added. NT-proBNP stabilized at 170 ng/L. Echocardiography was performed weekly to monitor CI, CVP and right systolic function indexes. Favorable echocardiographic dynamics after therapy were observed: mean PAP 60 mmHg to 45-50 mmHg, PVR from 10 WU to 7 WU, stroke volume normalized and CVP remained normal. However, right ventricular systolic function improved but did not normalize (**Table 1**).

In **conclusion,** with complete echocardiographic hemodynamic assessment and thorough clinical assessment high-risk pregnancies in patients with severe PAH can be managed. In addition, FAC, MPI TDI, 3D ejection fraction and RV free strain better assess right ventricular systolic function than TAPSE.

TABLE 1. Echocardiography variables before and after therapy.

sPAP mmHg	106	64
mPAP mmHg	64	42
PVR (WU)	10	7
RAP mmHg	3	3
SV ml	51	69
SVI ml/m^2	29	40
CO L/min	4.6	4.7
CI L/min/m^2	2.6	2.7
RA cm^2	26	26
RV TAPSE cm	1.8	2.4
RV FAC %	24	30 (3D EF 36%)
RV MPI TDI RV free strain %	0.8 -12	0.6 -24
LV Eccentricity index	2.4 / 3.9	1.5 / 2.1
Pericardial effusion	minimal	minimal

sPAP = systolic pulmonary artery pressure, mPAP = mean pulmonary artery pressure, PVR = pulmonary vascular resistance, RAP = right atrial pressure, SV = stroke volume, SVI = stroke volume index, CO = cardiac output, CI = cardiac index, RA = right atrial area, RV = right ventricle, TAPSE = tricuspid annular plane systolic excursion, FAC = fractional area change, MPI TDI = myocardial performance index, 3D EF = three-dimensional ejection fraction, RV free strain = right ventricular free wall longitudinal strain.

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LITERATURE

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