Pulmonary hypertension and pregnancy. Is it time to reconsider recommendations in certain groups? Contemporary outcomes in a tertiary referral centre

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Background and aims: Pregnancy is as a major risk scenario for pulmonary hypertension (PH) females. Disease targeted therapy (DTT) has dramatically changed prognosis in certain PH groups, namely: pulmonary arterial hypertension (PAH), chronic thromboembolic PH (CTEPH) or segmental-PH; also facilitating pregnancy management. In spite a more benign course in the responders to calcium channel blockers subgroup (R-CCB), maternal mortality is thought to remain high and current recommendations still consider all PH-patients as very high risk (mWHO IV). We aimed to analyse pregnancy outcomes in PAH, CTEPH and segmental-PH in a contemporary cohort and to specifically assess pregnancy risks in the R-CCB subgroup.

Methods and results: All pregnant PAH, CTEPH and segmental PH patients and patients transferred within 8 weeks post-delivery between January 2000 and January 2020 to our centre were studied.

28 pregnancies were included. Mean age 28±6 years. Underlying aetiology: 10 (36%) Idiopathic-PAH, 4 (14%) Heritable-PAH (1 of them veno-occlussive disease-POVD-), 2 (7.1%)PAH-connective tissue disease, 9 (32,1%) PAH-congenital heart disease (1 Eisenmenger), 1 (3.6%) CPETH, 2 (7.1%) segmental-HP. 21% were R-CCB.

From the overall 28 pregnancies, 32% underwent early termination of pregnancy (ETP). When pregnancy was continued in non-responders, 5 (23%) were admitted from cardiac causes and up to 13% required inotropes. 66.7% of non-responders patients needed uptitration of PH-DTT along pregnancy (19% being discharged on prostacyclins). Considering the whole cohort, two patients (7.1%) died along the pregnancy period, both non-responders PAH (1 IPAH and 1 POVD). One died at ETP and the other one 48 hours after C-section at week 22. No R-CCB presented with maternal complications, and all continued on CCB without needing any change on treatment.

Regarding the new-born, average gestational age was 28 weeks and 53% suffered from some neonatal morbidity (only 1 R-CCB), including 1 neonatal death.

From a haemodynamic perspective, baseline mean pulmonary artery pressure (mPAP) was 41.9±15 mmHg, pulmonary vascular resistances (PVR) were at 7.1±4 WU. If haemodynamics were also available after pregnancy (53%), no significant increase in PVR was noted, although the required DTT was remarkably higher.

Survival-free of death or lung transplantation (Figure 1) was calculated at 78.57% over a median long-term follow-up of 6 years.

Conclusions: DTT has improved outcomes in pregnant PH females. Although mortality related to pregnancy appears high but not prohibited, maternal and neonatal morbidity is still very high. Moreover, ETP in this population is not without complications. In addition, DTT often requires uptitration suggesting disease progression. Nevertheless, R-CCB females appear to be able to complete pregnancy without events supporting our suggestion to reconsider their estimated pregnancy-associated risks.

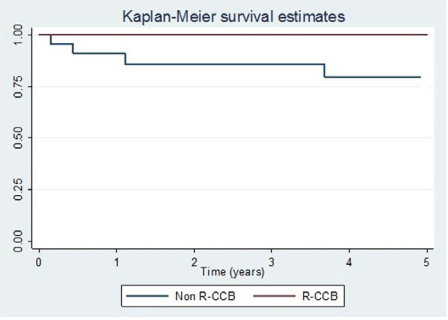

Figure 1. 5-year survival