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Treatment of bronchopulmonary dysplasia-associated pulmonary hypertension with pulmonary vasodilators – single center experience.

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Introduction
Patients with bronchopulmonary dysplasia-associated pulmonary hypertension (BPD-PH) have significantly higher mortality (up to 50%) and morbidity compared to patients with bronchopulmonary dysplasia (BPD).
Objective
The aim of this study was to present single center experience and outcome of the treatment with pulmonary vasodilators patients with severe BPD-PH
Methods
BPD-PH risk factors (according to Nagiub et al), WHO-FC, NTproBNP, echo estimated RV systolic pressure before treatment and during follow-up, were chosen for retrospective analysis.
Material
Between 2009-2018 12 pts with severe BPD-PH diagnosed at age 9,4 mths (3-18,5) were accepted for treatment. Ten pts (83%) had severe BPD with oxygen blood saturation 70-90%HbO2. BPD-PH risk factors (2-7) were confirmed in all. Severe PH (RV systolic pressure >2/3 of systemic) was diagnosed in cardiac catheterisation in 8 pts and by echo in 4 pts. Seven pts were in III-IV WHO-FC and 5 pts in II WHO-FC. Median NTproBNP was 3000pg/ml (171-35000).
Congenital shunt coexisted in 6 pts: small PDA -1, ASD -2, VSD -2. Neonatal PDA closure was performed in 3 pts. 3 pts were diagnosed after shunt closure (2 ASD, 1 VSD). Pulmonary vein stenosis was excluded in all pts.
Treatment included oxygen therapy (11pts), sildenafil (11pts) and sildenafil + bosentan (1pts) with no side effects.
Results
Improvement in WHO-FC, NTproBNP after 3 mths of therapy was observed in 9 pts (75%). No changes in oxygen blood saturation were found.
4 deaths (33%) occurred 1,8-8,1 mths after beginning of therapy: 1 related to sepsis in patient without shunt and 3 in patients with VSD (1 after PAB, 2 after surgical VSD closure).
Pulmonary vasodilators were discontinued in 6 pts (50%) under echo/cardiac catheterization control after 10,3-40,7mths. 2 ASD were closed >1 year after treatment discontinuation. Two patients still require PH treatment.
Conclusions
BPD-PH treatment with pulmonary vasodilators is well tolerated, led to clinical improvement in majority of patients and normalisation of pulmonary hypertension parameters in half of survivals.
Decision on VSD surgery in patients with BPD-HP must take into account the high risk of postsurgical death.
Further studies with larger population are needed to establish outcome in patients with BPD-PH.