57.00

PRIMARY PULMONARY HYPERTENSION IN NEONATES - ADVANCED PATOHYSTOLOGICAL CHANGES

A. Dasovic Buljevic, V. Benjak, D. Jelasic, N. Rojnic Putarek, I. Malcic
University Hospital Rebro, Department of Neonatal, Intensive Care Unit and Department of Paediatric Cardiology, Zagreb, Croatia
ivanmalcic@hotmail.com

We are referring three neonates (2f, 1m) admitted to NICU because cyanosis. The primary pulmonary hypertension (PPH) was suspected after all other causes of cyanosis were ruled out. Patient 1., female, impossible to accomplish satisfactory oxygenation by mechanical ventilation. ECHO; R-L shunt on the PFO, TV regurgitation, typical M-mode movement of PV. Chest X-ray normal. Radioangioscintigraphy; R-L, slowed pulmonary flow. Treatment by conventional mechanical ventilation, prostaglandin E1, tolazoline. Deep hypoxemia sustained, the baby died at the age of 13 days. Patient 2, female, intracranial haemorrhage of the third degree and perinatal infection, ECHO; PDA (medicamentously closed), large ASD II., but bidirectional shunt on the atrial level persisted. Cyanosis progressive and severe, PH by catheter measuring - PVR 12.5 Uxm2 Baby died from ventilator associated pneumonia at the age of three months. Patient 3, male with trisomy 21, diagnosed postnatally. Severe cyanosis and intracranial haemorrhage of the second degree was present. ECHO: dilated RV, TV insufficienty (gradient 70 mmHg), PV insufficienty (regurgitation wave of 2,4 m/s) and M-mode movement of the PV, hughe IVC, R-L shunt on the PFO, no structural hearth disease. The neonate was treated by mechanical ventilation, NO inhalation and calcium channel blockers, but died at the age of two months. In all three patient autopsies showed pulmonary plexyform lesions (V degree - Heath and Edwards). Advanced patohistological changes implicate early prenatal beginning of the disease, because of the absence of physiological remodelling of the PA because intrauterine pulmonary vascular endotel damage.