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**Title:** Pulmonary hypertension due to alveolar hypoventilation: Functional impact and improvement of haemodynamics and exercise capacity under non-invasive positive pressure ventilation

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**Body:** Background: For pulmonary hypertension (PH) due to lung diseases guidelines recommend treatment of the underlying disease. Due to preserved cardiac index (CI) in alveolar hypoventilation the functional impact of elevated pulmonary artery pressure (PAP) is not clear. The impact of non-invasive positive pressure ventilation (NIPPV) on exercise capacity of patients with PH due to alveolar hypoventilation is unclear. Objective: To characterize the functional impact of PH due to alveolar hypoventilation and the haemodynamic and functional consequences of NIPPV. Methods: Analysis of hemodynamics and functional capacity of 18 patients with daytime PH due to alveolar hypoventilation. The analysis included the data from baseline after a complete diagnostic work-up and after 3 months of NIPPV. Results: Patients presented with a mean PAP (mPAP) of 49 +/- 12 mmHg, a CI (3,21 +/- 0,66), a 6-minute walking distance (6-MWD) of 303 +/- 133 m and severely elevated nt-pro-BNP levels. mPAP correlated negatively with maximal workload ( $R = -0,71667$ ,  $p = 0,029$ ) and six-minute-walking distance ( $R = -0,621$ ,  $p = 0,010$ ). Under NIPPV, we found a significant reduction of mPAP (-17,75 mmHg,  $p = 0,0005$ ), NT-pro-BNP serum levels (-2110 pg/ml,  $p = 0,0014$ ), improvement of the 6-MWD (+66 m,  $p = 0,0082$ ) and maximal workload (+18 W,  $p = 0,028$ ). CI did not change significantly. Changes of workload and mPAP correlated negatively ( $R = -0,7545$ ,  $p = 0,0305$ ). Conclusions: Despite preserved CI elevated mPAP has a functional impact for patients with PH due to alveolar hypoventilation. NIPPV leads to a significant reduction of PH and improvement of exercise capacity.