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**Title:** Home non-invasive ventilation (HNIV) improves survival in hypercapnic patients with cystic fibrosis

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**Body:** Background: The clinical benefits of home non-invasive positive pressure ventilation (HNIV) have not been well established in adult patients with cystic fibrosis (CF). Objective: The purpose of this matched case-control study was to compare the effects of HNIV and long-term oxygen therapy (LTOT) on survival of adult CF patients with chronic hypercapnic respiratory failure (CHRF). Methods: Twelve patients receiving HNIV were matched with 12 patients receiving LTOT regarding age, FEV1, PaCO2 and BMI. Results: There were no differences between groups of patients in demographics, clinical and functional characteristics (HNIV patients: 24.9±4.8 years, BMI 16.1±3.3 kg/m<sup>2</sup>, FEV1 15.3±3.4%, PaCO2 59.8±7.7 mmHg; and LTOT patients: 23.9±4.6 years, BMI 15.8±4.2 kg/m<sup>2</sup>, FEV1 16.1±4.4%, PaCO2 61.4±7.9 mm Hg). HNIV settings were: ST mode, IPAP 20.1±4.2 cm H<sub>2</sub>O, EPAP 4.6±0.7 cm H<sub>2</sub>O (nasal mask – 4, oronasal mask – 8). Compared with LTOT, HNIV significantly reduced breathing rate (p=0.036), PaCO2 (p=0.038) and number of exacerbations (p=0.028). Survival was significantly better in HNIV group in comparison with LTOT group (survival time 12.5 [95%CI 0.6-16.4] months vs 6.0 [95%CI 0.7-7.3] months; log-rank test, p=0.024). Conclusions: The survival of adult CF patients receiving HNIV was better than that of patients treated with LTOT alone. We suggest HNIV is a first-line treatment for adult CF patients with CHRF.