Decannulation in congenital central hypoventilation syndrome

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Abstract

Rationale: Patients with congenital central hypoventilation syndrome (CCHS) require long-term ventilation to ensure gas exchange and to prevent deleterious consequences for neurocognitive development. Two ventilation modes may be used for these patients depending on their tolerance, one invasive by tracheostomy and the other noninvasive (NIV). For patients who have undergone a tracheostomy, transition to NIV is possible when they meet predefined criteria. Identifying the conditions favorable for weaning from a tracheostomy it critical for the success of the process. Objective: The aim of the study was to share our experience of decannulation in a reference center; we hereby describe the modality of ventilation and its effect on nocturnal gas exchange before and after tracheostomy removal. Methods: Retrospective observational study at Robert Debré Hospital over the past 10 years. The modalities of decannulation and transcutaneous carbon dioxide recordings or polysomnographies before and after decannulation were collected. Results: Sixteen patients underwent decannulation following a specific procedure for transition from invasive to NIV. All decannulations were successful. The median age at decannulation was 12.6 [9.7; 15.0] years. Nocturnal gas exchange was not significantly different before and after decannulation, while expiratory positive airway pressure and inspiratory time increased significantly. An oronasal interface was chosen in two out of three patients. The mean duration of hospital stay for decannulation was 4.0 [3.0; 6.0] days. Conclusion: Our study underlines that decannulation and transition to NIV are achievable in CCHS children using a well-defined procedure. Patient preparation is crucial to the success of the process.

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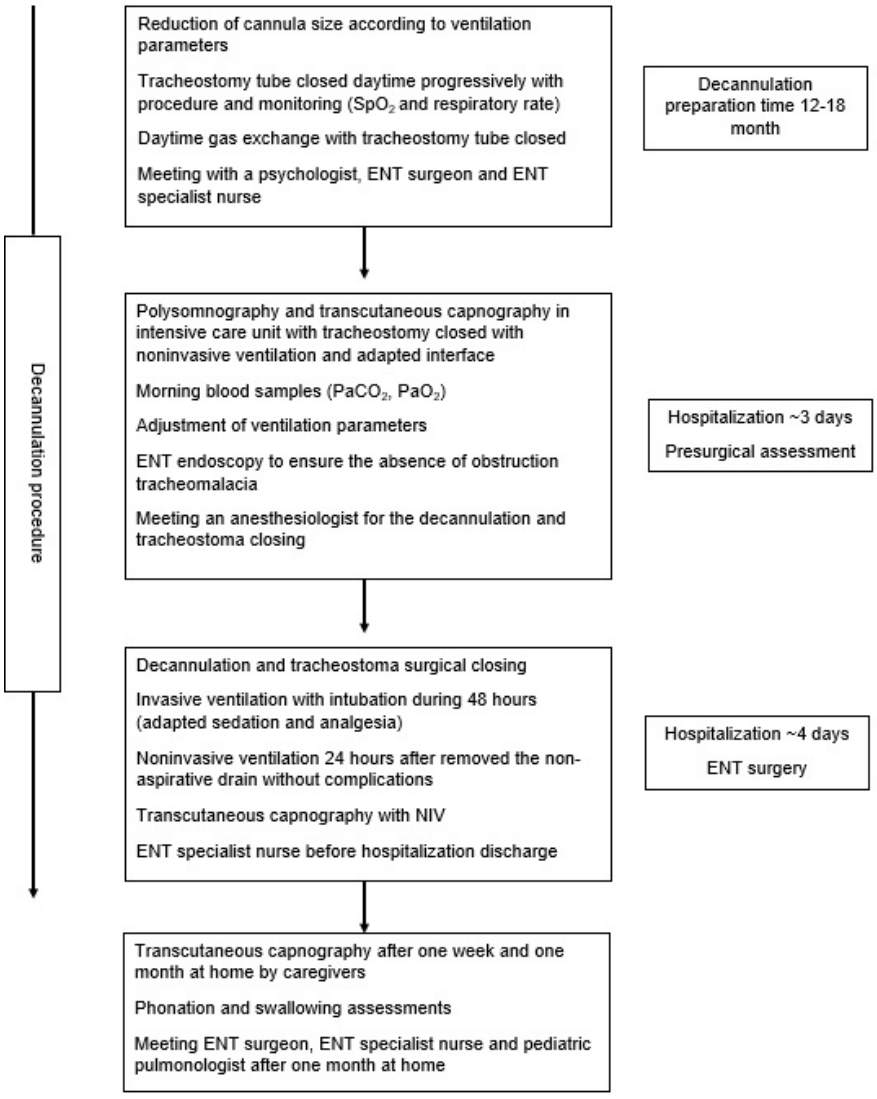

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