**Diaphragm Pacing (DP) in congenital central hypoventilation syndrome (CCHS) – The Swedish experience**

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**Background:** Since the early 70's different options for long-term respiratory support have been examined and led to the development of various types of mobile ventilators (1). One of the major challenges has been to find a non-traumatic but safe connection between the ventilator and the patient. The golden standard has been endo-tracheal tubes placed through a tracheotomy. Efforts have also been made to apply non-invasive ventilation by using different facemasks, moulded nasal prongs or nasal CPAP prongs. In the late 70's a new clinical option was developed where electrical stimulation of the phrenic nerve made the diaphragm contract i.e. DP, producing more physiological ventilation (2). The advantages consisted of avoiding the negative effects of positive pressure ventilation on pulmonary circulation and lung tissue, achieving an increased mobility of the patients and providing a non-invasive connection of ventilatory assistance i.e. avoiding traumatic application on the face or upper airways.

**Aim/Methods**: To present a 20-year Swedish experience of DP with special focus on patients with CCHS.

**Results**: A total of 16 patients have received DP since 1989 on the clinical indications CCHS (n=7), meningomyelocele/Arnold-Chiari (n=4), spinal trauma (n=5) of which 2 patients acquired their injury due to birth trauma (tetraplegic n=1; specific trauma to the medulla oblongata n=1). Median age at implantation of DP in CCHS patients 8 years (range= 4-17 years). Prior to DP, non-invasive ventilation applied by mask/prongs (n=6), and only one patient with invasive ventilation by tracheotomy. The longest duration of non-invasive ventilation by mask has been 17 years, resulting in minor face deformity. Median duration of DP in CCHS patients 9 years (range=2-23 years). Partial failure of DP in one patient with CCHS occurred due to upper airway obstruction and therefore the patient combines DP with nIPPV on mask; the same patient has had a successful pregnancy with a healthy infant. Complications with DP in CCHS has been repositioning of the subcutaneous receivers in one patient and rupture of electrode to receiver connection due to manipulation by one patient. Implantation in children at 4-5 years of age (n=3) has been uncomplicated. Only minor adjustments of stimulatory amplitude have been made over the years, mainly postoperatively and during infections.

Conclusion: Our experience of DP in CCHS has shown good long term patient compliance and few complications. DP in CCHS seems to be a safe and attractive option for these patients, where further investigations of quality of life compared to other long-term ventilatory modes are warranted.

Ref:

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