**Living with Diaphragm Pacing. A Multinational Quality of Life (QoL) study of children with Congenital Central Hypoventilation Syndrome (CCHS) using a phrenic nerve stimulator**

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**Aim**

The purpose is to assess the QoL in children with CCHS who are currently implanted with a phrenic stimulator as ventilatory support. A secondary aim is to define the follow-up care and administration around these patients and investigate if there is a correlation between QoL and health care accessibility in this cohort of patients.

**Background**

To this day, children with CCHS have been using a variety of ventilation methods to sustain adequate ventilation when their natural breathing regulation fails them. None of these methods have been ideal due to problems with infections, malformations of the facial skeleton and freedom of movement. In the late 1970s phrenic nerve stimulation (DP) was proven successful for people suffering from central hypoventilation, and was subsequently applied on children some years later (1,2).

**Method**

We have identified 145 patients world-wide implanted with a phrenic stimulator on CCHS indication, 49 being below 18 years of age. A web-based survey was distributed to all the identified patients under the age of 18 (n=49). The survey is divided into three parts. The first assesses the regularity of follow-ups, distance to doctor’s office, differences in terms of symptoms, diagnostic procedures and genetics. The other two parts are made one self-assessment and one proxy-assessment version of the validated QoL assessment tool, Disabkids®.

**Results**

Deadline for participation is set for March 9th 2012 and statistical analysis of data will be made with SPSS ® non-parametric and parametric values where appropriate. Type of questions and preliminary results will be presented accordingly.

**Conclusion**

Our hypothesis is that this study will indicate that the use of DP for ventilatory support enables an easier and freer life for the CCHS patient. National and international differences in follow-up procedures, which might have impact on the QoL for these patients, will be assessed and evaluated.

Ref

1. Glenn WWL, Holcomb WG, Gee JBL, Barh R. Central hypoventilation syndrome: long-term ventilatory assisance by radio-frequency electrophrenic respiration. Ann Surg 172: 755-773, 1970
2. Chen ML, Tablizo MA, Kun S, Keens TG. Diaphragm pacers as a treatment for congenital central hypoventilation syndrome. Expert review of medical devices. Vol 2 5:577-585. 2005