**Network of Pediatric Palliative Care: a model for care of children with CCHS and their families**

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**Background**

Children with Congenital Central Hypoventilation Syndrome (CCHS) are eligible for pediatric palliative care (PPC), because they have a long-life dependence on mechanical ventilation and possible various anatomical and functional abnormalities of the autonomic nervous system. Therefore they requires an interdisciplinary management to assure comprehensive and continuous care including clinical, psychological and social aspects during all the phases of their disease (i.e. acute events, stability).

We present the application of Palliative Care Network Model for children with CCHS at University of Padua.

**Methods**

The PPC Center of the University of Padua has managed patients with CCHS since 2003 with the aim to assure a family-centered home-care for these patients. The Center coordinate a network of care-providers composed by palliativist pediatricians, general pediatricians, hospital pediatricians, social assistants, school personnel, psychologists, to organize and maintain an individualized project of care for each patients. The network provides 24/24 hours on call availability, ventilation management, medical aids provision at home, periodic home visits, planned admissions for follow-up, algorithms for emergency management at home, training for emergency management for families and operators, respite admissions, assistance about civil rights and economical benefits.

**Results**

Since 2003 8 patients with genetic diagnose of CCHS have been followed (4m/4f), mean age 6.4 years old (range:2.6-11). 1 patient was diagnosed also with Hirschsprung disease.

All children were submitted to a biannual follow-up till 4 years old and then to an annual evaluation. Major clinical problems during the follow up were: cardiac rhythm abnormalities (3 patients), seizures diagnosis and treatment (6 patients), endocrine-metabolic dysfunctions (hypoglycemia episodes 3 patients, diabetes insipidus 1 patients).

6 out of 8 children had hospital admissions for acute problems (mean: 1admission/patient/year, range: 0-3.2). A mean of 20 call/patient/year was realized. More common problems concerned tracheostomy management and respiratory tract infections. Most part of these problems were solved at home following phone indications or requiring home visits by Network personnel (palliativists or general pediatricians).

All children in school age (6 children) go to the primary school with a devoted operator for sanitary assistance (25-30 hrs/week); each operator was chosen by the family and underwent a theoretical and practical training about ventilation support and emergency management. No one acute episode occurred at school requiring medical intervention. 2 families obtained economical support for home attendance. For all children ventilator, circuits, tracheostomy tubes, aspiration devices are warranted by health system with no charge for families. All families were satisfied about the network service.

**Conclusions**

PPC network of care assures a complete multidisciplinary assistance to meet complex clinical needs of children with CCHS. It allows families to manage their children at home in a safe and efficacious way.