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Conference highlights report

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Presentation 1 – Progression from infancy to childhood

Presenter: Dr Alessandro Amaddeo

The number of children treated at home with Long Term Home Ventilation (LTHV) has increased significantly over the last 15 years, with an accompanying 10-fold increase in prevalence[1] thus the interest in this topic has grown in recent years.

Data about this growth has been published in different studies, for example, the researches in the Center for Home Mechanical Ventilation (HMV) of Utrecht found out that during the 1979–1988 period only 8 patients received HMV but this number increased to 122 in the 1999–2008 decade[2].

There are multiple explanations for this increase: non-invasive ventilation (NIV) is now an accepted treatment option for children with sleep-related breathing disorders and with neuromuscular diseases (NMD) and it's now considered an acceptable option both by health professionals and by patients and their families[1].

The prevalence of diseases may slightly vary among different studies, time periods, and geographical locations, but the two main indications are NMD and cranial-facial abnormalities/obstructive sleep apnea.

A study,conducted by McDougall et al, in a single center over a 15-year period well describes this population. In this research, the most frequent indications were neuromuscular disorders (47%) followed by craniofacial abnormalities/obstructive sleep apnea (OSA) (16%). Less common indications were spinal injury (6%), abnormal ventilatory control (acquired or congenital, 13%), airway malacia (8%) with the remaining 10% having a various range of other conditions including diaphragm paresis postcardiac surgery, scoliosis, pulmonary hypoplasia and pulmonary involvement of Langerhans cell histiocytosis[1].

Along, with an increase in prevalence, treatment with NIV is started earlier than before in fact there has been a 1.5-fold increase in older children (12–17 years), a 2-fold increase in children aged 6–11 years and a 3-fold increase in the youngest[2].

The rationale for home non-invasive ventilation in these patients varies significantly according to the underlying condition, NDM patients have a chronic disease that has a natural tendency to worsen progressively; on the other hand, for children with complex OSA NIV is used when surgery is contraindicated or ineffective, or as a supportive treatment until an appropriate age is reached for surgical intervention[3].

There are some important differences between adults and children when it comes to NIV.

In both groups NIV is often used during sleep, however, the amount of time spent sleeping varies consistently according to age: a child younger than 2 years sleeps and consequently uses a ventilator for about 12–14 h/day far more than the average adult patient.

Clinicians must carefully consider which ventilator to choose, in particular the presence of an internal battery and the use of life support ventilators. In fact, some children are too young to help themselves in emergencies such as ventilator malfunction or electrical power failure hence the necessity to choose life support ventilators especially for the youngest. However these ventilators are more expensive, heavier and may not be available in low-income countries.

Another important issue to consider is weights influence on the flow detection hardware and algorithm; this becomes particularly important during the follow-up and data analysis from the ventilator.

One big limitation related to the application of NIV in infants and children are the interfaces and their appalling paucity. Contrary to adults, nasal masks seem to be the preferred type, both in the chronic and acute setting, even in children that have passed the age of obligatory nose breathing[4]. In case of severe anatomical anomalies face masks might be the only choice but the risk of vomiting and consequence aspiration must be carefully considered.

Another unique aspect of children is that they grow and even though their height/weight varies it is not routinely needed to change the pressures delivered at least in patients affected by NMD. In fact all age groups of pediatric subjects receiving NIV require similar ventilator pressure levels which don't need to be adjusted over time; the only parameter that requires modifications is the backup respiratory rate because of the physiological decrease in breathing frequency[5].

Since the patients are still growing masks can alter the development of the facial bones causing some degree of deformity with global facial flattening being the most common[6].

As for adult patients, skin injuries (erythema, skin necrosis) are also common and mostly observed on the forehead and the glabella[6].

Another fundamental issue in the field of pediatric NIV treatment is the possibility of weaning as the child grows and becomes a young adult; about 25% of patients can, at some point, be weaned. The possibilities of success are highly dependent on the underlying disease. Patients with upper airway anomalies may improve spontaneously with age or after surgical[7] interventions and thus can be weaned successfully; on the other hand, weaning from NIV is unlikely to occur in children with NMD's.

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Presentation 2 – Decannulation

Presenter: Dr Lara Pisani

Professor Pisani highlighted that long-term ventilation (LTV) is associated with an improvement in survival in a vast range of conditions, thus resulting in an increasing number of patients surviving to adulthood. More than 85% of the children will need to transition to adult services. These patients can be broadly stratified according to their level of needs: 5% have complex needs; 25% have complex chronic conditions; while the remaining 70% are patients with chronic conditions with good control. Children on LTV tend to fall into the first two categories[1].

Long-term ventilation can be performed either through tracheostomy or with face masks though most patients can be managed through NIV some will require tracheostomy. In a study that followed 933 patients during a 10-year period (1998–2008) 22% had a tracheostomy performed, mostly the ones affected by NDM diseases[2].

NMD's are a broad spectrum of conditions that vary in their progression, prognosis, and level of lung function impairment. NMD can compromise the gas exchange and pump functions of the respiratory system, the upper airway muscle tone and airway protection, and the efficiency of secretion clearance. Tracheostomy in these patients should be considered in five different conditions according to the British Thoracic Society guidelines:[3]

- Severe bulbar dysfunction resulting in frequent aspiration.
- After an acute exacerbation has led to a period of invasive ventilation and extubation has failed despite optimal management for 2 weeks or more.
- When ventilatory support is needed for more than 16 h per day.
- When there is failure to correct hypoxemia or hypercapnia with NIV.
- Severe mid-face hypoplasia not correctable by adjusting the NIV interface.

The key point is to identify the patients in which the tracheostomy must be performed/kept and those that can be decannulated and eventually switched to non-invasive ventilation.

Typically, patients with progressive neuromuscular disorders develop acute respiratory failure (ARF), are intubated, and when failing spontaneous breathing trials undergo a tracheotomy and receive tracheostomy mechanical ventilation.

But this must not always be the case, in fact, different centers have demonstrated that continuous noninvasive ventilatory support along with mechanical insufflation-exsufflation can be used indefinitely instead of performing tracheostomy and "unweanable patients" can be extubated using the same technique[4].

In general, there is no consensus on how the decannulation process should be done but two steps are considered primordial for the initiation of the decannulation process, cuff deflation and assessment of airways permeability.

After performing the previous steps, it is suggested to evaluate the patient's ability to protect the lower airways, assessing first swallowing capability, and the patient's capacity to manage secretions and expel them through cough. Some studies suggest cough training could help the patients who failed this step[5].

The final step of the decannulation process is the permanence of the occluded tracheostomy; in this period the patient should be able to breathe spontaneously and sufficiently through the upper airway, maintaining stable oxygen saturation. Only after this long and comprehensive evaluation the patient can be finally decannulated. In conclusion, decannulation is a complex process that includes a multidisciplinary approach and that varies greatly between centers since there are no universally accepted guidelines.

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Presentation 3 – Transition from adolescent to adult care

Presenter: Dr Alessandro Amaddeo

The transition from pediatric to adult healthcare systems is one of the most challenging periods for patients. Blum defined "transitional care" as "the purposeful, planned movement of adolescents, and young adults with chronic physical and medical conditions from child-centered to adult-oriented healthcare systems"[1].Adolescence is recognized as a vulnerable period due to the physiological, emotional, and psychosocial changes. Teens with chronic diseases have to deal with their health problems as well as with the mentioned problems of adolescence itself.

This is also a period when health risk behaviors peak (alcohol, tobacco, and drugs) and chronic conditions tend to exacerbate or worsen. Patients with NMD often transition at a time when their disease progresses resulting in loss of independence, an increasing reliance on LTV, and the emergence of a raft of new health problems.

Therefore, it is essential to consider the "transition" from pediatric to adult services in the global context of a period of multifactorial change for the patient.

The lack of a smooth transition can have a negative impact on compliance to therapies, as demonstrated in a study that found that medication adherence deteriorates in pediatric transplant recipients who transition to adult services[2]. This finding is consistent with others that found reduced medication adherence, increased risk of discontinuity of care and higher emergency department and hospital use[2].

Even though most patients described adult health care transition as a normal life process and a welcome marker of pending adulthood there are a big number of obstacles[3].

The most prominent barrier mentioned by youth with special health care needs and caregivers is the difficulty in leaving their pediatric clinicians with whom they have had a long-standing relationship. Health care transition barriers may contribute to a sense of medical homelessness and increased caregiver burden[4].

But also clinicians identify many transition barriers. The most common obstacles reported by pediatric and adult care clinicians are the lack of communication and coordination and the different practice styles between healthcare professionals[4].

In 2011, the American Academy of Pediatrics published a clinical report on the health care transition from adolescence to adulthood. It proposed an algorithm containing action steps such as discussion of transition policy, initiation of a transition plan, and review of this plan at specific time points.

Despite this, when pediatric pulmonary program directors in the US were surveyed on the transition process in 2015, 78% of respondents reported that their workplace did not use a standard protocol for transition, and of those 78%, 41% had no process in place at all[13]. These results revealed just how much progress still needs to be made in this group of patients.

In conclusion, transition is a process, not a single event of transference of care. All patients, whether manifesting complex needs or not, require some degree of individualized planning during their transition. This can only happen if there is a systemic recognition of the need for greater collaborative care partnerships between pediatric and adult clinicians.

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2. Annunziato, Rachel A., et al. "Adherence and medical outcomes in pediatric liver transplant recipients who transition to adult services." Pediatric transplantation 11.6 (2007): 608-614.

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Presentation 4 – Growing into old age on NIV

Presenter: Professor Anita Simonds

The use of noninvasive ventilation (NIV) as first-line supportive therapy for acute respiratory failure (ARF) is increasing in the ICU. This technique in selected populations of critically ill patients reduces the need for intubation and decreases mortality. The use of non-invasive ventilation is frequently proposed for the respiratory support of patients with a do-not-intubate order (DNI) thus representing the only ventilatory support possible.

In an observational study, 1,696 patients were admitted to an ICU: of these, 60% required ventilatory support, and 37% received NIV during their ICU stay with the proportion of patients needing ventilatory support similar in very old and younger ones[1].

Interestingly failure rates were very similar between old (42%) and young patients (40%) but the in-hospital mortality of the older patients significantly higher than for younger patients.

Interesting data comes from the survival rate of old patients that have failed the NIV trial and do not have a DNI order. The data shows that the prognosis of these patients was not better than that in patients not intubated due to endotracheal intubation limitations. As a result endotracheal intubation after NIV failure in this population of patients seems of questionable benefit[1].

As the study just mentioned showed, NIV is a well-accepted treatment for acute episodes of respiratory failure in older subjects and as the European population is aging there has been an important increase in the number of patients over 75 years old treated domiciliary with NIV. These patients now represent more than 40% of ventilated adults in France, compared to 30% in 1996[2]. Despite the fact that elderly patients now represent a large part of the Chronic Respiratory Failure population, very few data on NIV adherence, efficacy and clinical outcomes are available in this specific population.

As demonstrated in various studies there are no significant differences between older and younger patients treated with NIV regarding technical aspects (interfaces and settings) or adherence to therapy[3]. Differences are routinely found in the number of comorbidities, higher in the older group, as expected. As for the indications for home NIV there is a higher prevalence of COPD diagnosis in the older age group; conversely NMD is rarely an indication for long-term NIV in the elderly.

As for the efficacy NIV is as efficient in the elderly as it is in the younger as indicated by the improvement of the arterial blood gasses (ABG) values and sleep quality scores[4].

All this data confirms the feasibility and the positive clinical impact of NIV in the old. However early reports did not show particularly favorable outcomes in the older patients treated with NIV. This was primarily due to a higher risk of neuropsychological impairment and difficulties adapting to the overall burden of home NIV when compared to younger individuals.

Many studies examined the impact on Quality of Life (QoL) of NIV in the elderly: *Adrien et al.*[5] did not find any improvement in the Short Form 36 (SF-36) questionnaire following NIV initiation even though the improvement of ABG values and sleep quality; similar results were found by *Couturier et al.*[6] that confirmed increased survival and improved gas exchanges in patients >80 yrs old but with no impact on QoL.

A possible explanation for these results is that there was no validated disease-specific score to evaluate the quality of life in HMV subjects since most of the studies conducted have used the SF 36, or measures specifically designed for patients with COPD such as the Saint Georges Respiratory Questionnaire.

In recent years, however, the Severe Respiratory Insufficiency Questionnaire (a 49-item self-administrated quality of life measurement) has been developed. This is an instrument specifically designed to assess the effects of home mechanical ventilation on QoL.

In a study performed in 2007 that used this new questionnaire the most important determinants of QoL were dyspnea, the presence of an obstructive disease and the number of hospitalizations in the previous year. The data supports the growing idea that the degree of impairment in quality of life is influenced by the underlying disease with COPD patients having the worst QoL[7].

But NIV is a chronic therapy that accompanies the patients for many years and QoL is usually re-evaluate 3–6 months after initiating therapy. The only study with a long follow-up (6 years) found improved QoL in the majority of patients and confirming what had previously been suggested by other studies: that patients with COPD tend to have lower QoL (measured through the SRI score) than other disease groups[8].

Since NIV to achieve its clinical benefit needs to be used consistently it increases the amount of time that patients dedicate to healthcare tasks with the risk of becoming overwhelming thus leading to poor compliance. For this reason it is important to educate the patients and outlining potential treatment burden before starting long-term home NIV[9].

In conclusion, long term ventilation in the elderly is a difficult topic and to date little is known regarding the use of NIV on a long-term basis in the very old.

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