HAUTE AUTORITÉ DE SANTÉ

RECOMMENDING
BEST PRACTICES

EVIDENCE REPORT

Role and management of tracheostomy in the care of ventilator-dependent patients with slowly progressive neuromuscular diseases

Validated by the HAS Board on 5 November 2020

Best practice guidelines (BPCs) are defined in the health field as methodically developed proposals to assist the practitioner and the patient to find the most appropriate care in given clinical circumstances.

BPCs are rigorous summaries of the state-of-the-art and scientific data at a given time, described in the evidence report. They do not exempt health professionals from exercising discretion in the patient's treatment; this must be the treatment considered to be most appropriate depending on their own findings and the patient's preferences.

These best practice guidelines were developed according to the method summarised in the evidence report and in the HAS methodological guide available on-line: Development of best practice guidelines – Clinical practice guidelines method.

The objectives of this guideline, the population and the professionals concerned by its implementation are summarised on the last page (information sheet) and described in detail in the evidence report.

The latter and the guideline summary can be downloaded from www.has-sante.fr.

Guideline grades

A	**Established scientific evidence** Based on studies with a high level of evidence (level of evidence 1): randomised controlled trials with high power and without major bias or meta-analysis of randomised controlled trials, decision analysis based on well conducted studies.
B	**Scientific presumption** Based on scientific presumption provided by studies with an intermediate level of evidence (level of evidence 2) such as low power randomised controlled trials, well conducted non-randomised controlled studies, cohort studies.
C	**Low level of evidence** Based on studies with a lower level of evidence such as case-control studies (level of evidence 3), retrospective studies, case-series studies, comparative studies with major biases (level of evidence 4).
EC	**Expert consensus** In the absence of studies, the guidelines are based on agreement between experts in the working group after consultation of the review group. The absence of grading does not mean that the guidelines are not relevant and useful. However, it should prompt additional studies.

Description of the publication

Title	Role and management of tracheostomy in the care of ventilator-dependent patients with slowly progressive neuromuscular diseases
Work method	Clinical practice guidelines (CPG)
Purpose(s)	To aid the decision-making process in terms of the choice of care, to improve and harmonise practices. The aim is to improve the management of patients, and therefore the quality and safety of their care, as well as their quality of life, bearing in mind that tracheostomy increases the cost of care and limits the number of downstream care facilities able to accommodate these patients. In addition, it is an important ethical issue insofar as the choice of treatment affects quality of life, as well as patients' survival prognosis.
Targets concerned	Any individual with respiratory failure due to a slowly progressive neuromuscular disease. Slowly progressive neuromuscular diseases are understood as myopathies that lead to slowly progressive respiratory failure, primarily Duchenne muscular dystrophy. They also include type I spinal muscular atrophy and non-progressive conditions such as sequelae of poliomyelitis, chronic inflammatory demyelinating polyneuropathy and myasthenia. All age groups are concerned, with the exception of neonates (< 28 days). Patients with spinal cord injuries and rapidly progressing neurodegenerative conditions, such as amyotrophic lateral sclerosis, are excluded from the scope of this best practice guideline. The professionals concerned are those involved in the care of patients with neuromuscular diseases: paediatricians (paediatric neurologists, paediatric respiratory medicine specialists), respiratory medicine specialists, breathing and swallowing therapists, sleep specialists, anaesthesia and resuscitation specialists, intensive care and resuscitation specialists, surgeons, ear, nose and throat (ENT) specialists, radiologists, physical medicine and rehabilitation specialists, neurologists, general practitioners, nurses, speech therapists, physiotherapists, mechanical ventilation technicians.
Requester	*Association française contre les myopathies* (French myopathy association, AFM) - Téléthon
Sponsor(s)	*Haute Autorité de santé* (French National Authority for Health) (HAS)
Project management	Coordination: Cédric Paindavoine, project manager, HAS Department for Good Clinical Practice (head of department: Dr Pierre Gabach) Secretarial duties: Ms Laëtitia Gourbail

Literature search	From January 2007 to August 2018 (see literature search strategy described in annex 1 of the evidence report) Performed by Mr Aurélien Dancoisne, with the help of Ms Juliette Chazareng (head of Documentation-Monitoring department: Ms Frédérique Pagès)
Authors	Mr Ghilas Boussaïd, methodologist, epidemiologist, Colombes – project officer
Declaration of interests	The members of the working group communicate their public declarations of interest to the HAS. They may be consulted on the website https://dpi.sante.gouv.fr. They were analysed according to the analysis grid of the HAS guidelines for the declaration of interests and management of conflicts of interest. The interests declared by the working group members were considered to be compatible with their participation in this work.
Validation	Version dated 5 November 2020
Other formats	Recommendations

This document and its bibliographic reference are available to download at www.has-sante.fr

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Contents

1. Indications and contraindications for tracheostomy

A peer-reviewed Cochrane review (1) compared the efficacy of non-invasive ventilation with invasive ventilation in people with neuromuscular disease and/or chest wall disorders. A database literature search was performed by two independent experts. (**Expert opinion**)

The advantages described are as follows:

- invasive mechanical ventilation is safer than non-invasive ventilation since it routinely uses volume control mode, which, in contrast with the pressure control mode most commonly used in non-invasive ventilation, maintains the same insufflated volume irrespective of the mechanical loads imposed by the respiratory system on the ventilator;
- invasive mechanical ventilation, even in the absence of an inflated cuff to enable speech, reduces the risks of inhalation of secretions from the upper airways or of oesophageal reflux;
- invasive mechanical ventilation also facilitates the clearance of bronchial secretions by direct tracheal suctioning;
- by bypassing the upper airways, invasive mechanical ventilation reduces the risks of inefficiency observed in non-invasive mechanical ventilation due to leaks associated with non-invasive interfaces and opening of the mouth, and therefore offers better control of efficient minute ventilation;
- however, non-invasive mechanical ventilation decreases the risks of lung infection and barotrauma and does not cause tracheostomy-related complications such as tracheal stenosis and tracheal haemorrhage;
- it should always be offered as a first-line approach.

Comments

This literature review adopted a good methodology with a database search conducted by two independent experts. However, this study confirms - probably for ethical reasons - that no randomised trials exist enabling a clear comparison of the benefits of invasive ventilation versus non-invasive ventilation, bearing in mind that tracheostomy may have life-changing consequences and significantly modify the patient's needs.

A non peer-reviewed literature review (2) described the management of long-term non-invasive ventilation in patients with neuromuscular diseases. This review makes the following recommendations:

- exclusion of the use of tracheostomy as a long-term ventilation method, except in the ALS population;
- ventilator-dependent patients should be decannulated to return to non-invasive ventilation.

Comments

This review is based on the opinion of a single expert, with a potential bias in terms of selection of articles from the international literature. It is difficult to include this review in support of these guidelines as some of the international literature was not covered; this amounts to a substantial loss of information.

As regards decannulation, very few studies have assessed this practice in patients with progressive neuromuscular diseases. The literature available on this subject primarily concerns the studies conducted by the team of Dr Bach JR described below.

A single-centre retrospective study (3) reported decannulation cases between 1999 and 2013. Decannulation was performed using mechanical insufflation-exsufflation, before and after decannulation, to prevent atelectasis and thereby improve thoraco-pulmonary compliance, reduce oxygen desaturation and promote autonomous breathing. Three weeks after decannulation, 61 patients (age > 16 years), of whom 36 were totally ventilator-dependent, thus considerably increased their unassisted spontaneous breathing time ($p < 0.001$). (**Level 4**)

Comments

Study limitations: experience of a single centre. In addition, measurement of vital capacities is not described. Is this performed immediately post-insufflation or post-exsufflation? During unassisted breathing? Before tracheostomy?

The baseline respiratory function of the patients is not reported, and it is not specified whether the patients were tracheostomised as an emergency or as a preventive measure.

A literature review with independent peer reviewers (4) determined the success of extubation using cough augmentation techniques compared to no cough augmentation in all adults and children with acute respiratory failure with no diagnostic exclusions. Ultimately, this review only retained one trial in 75 subjects without neuromuscular disease and one case-control study of neuromuscular patients described below.

A randomised trial (5) evaluated the efficacy of mechanical insufflation-exsufflation in the context of an extubation protocol. In this trial, 75 patients without neuromuscular disease (including 26 women) with an average age of 61.8 (± 17.3) years were randomised, 40 to the control group (mean simplified acute physiology score II (SAPS II) of 47.8 ± 17.7), and 35 in the insufflation-exsufflation group (n = 35; mean SAPS II 45.0 ± 15.0). In the 48 hours following extubation, 20 control patients (50%) and 14 patients having undergone mechanical insufflation-exsufflation (40%) required NIV. Patients in the insufflation-exsufflation group had a significantly lower reintubation rate to the controls; 6 patients (17%) *versus* 19 patients (48%) respectively, $p < 0.05$. Considering only the subgroup of patients having used NIV, the reintubation rates related to NIV failure were significantly lower in the insufflation-exsufflation group compared to the controls; 2 patients (6%) versus 13 (33%) respectively; $p < 0.05$. The mean ICU length of stay following extubation was significantly shorter in the group treated with insufflation-exsufflation compared to the control group (3.1 ± 2.5 *vs* 9.8 ± 6.7 days; $p < 0.05$). No significant differences were noted in total ICU length of stay. The authors conclude that the use of

mechanical insufflation-exsufflation may reduce reintubation rates, resulting in a reduction in ICU length of stay following extubation. (**Level 2**)

Comments

This study was well conducted. However, it included patients with obstructive respiratory failure, and therefore cannot be applied to patients with neuromuscular diseases.

A case-control study (6) evaluated the use of mouthpiece/nasal intermittent positive-pressure ventilation (IPPV) as an alternative to intubation or to permit extubation for patients with primarily neuromuscular ventilatory impairment. Four of ten patients who presented acute ventilatory failure were managed without intubation, despite becoming dependent on continuous ventilator use. The six intubated patients, despite having no ventilator-free breathing ability, were successfully extubated using a pressure-control ventilator to NIV once normal arterial oxygen saturation levels could be maintained without additional O_2. (**Level 3**)

General comment

There are a limited number of studies providing sufficient data as well as analysis and publication biases. The overall methodological quality and hence the levels of evidence as concerns the efficacy of cough augmentation techniques with a view to decannulation in patients with neuromuscular diseases are very weak. The randomised trial was not conducted in patients with neuromuscular diseases, but in predominantly obstructive patients.

Similarly, a **Hispanic expert committee without independent peer reviewers (7)** conducted a review of the literature concerning the respiratory management of patients with chronic respiratory failure treated by non-invasive ventilation, concluding that non-invasive ventilation techniques improve life expectancy and avoid the need for invasive ventilatory support. (**Expert opinion**)

Comments

There is a lack of European representativity (no French, British, Italian or Belgian teams) having already published papers on this subject.

The British Thoracic Society (8) conducted a peer-reviewed literature review concerning the respiratory management of children with neuromuscular weakness, based on 177 articles on the management of tracheostomy as a long-term ventilation method, analysed using a clinical practice assessment grid. (**Expert opinion**)

The review revealed that tracheostomy may be indicated:

- in the event of severe bulbar dysfunction resulting in frequent aspiration (tracheostomy may be necessary to allow more effective airway clearance);
- in the event of failure to extubate, despite optimal management for 2 weeks or more;
- when ventilatory support is needed for more than 16 hours per day;
- in the event of failure to correct hypoxaemia or hypercapnia with NIV;

- when there is severe mid-face hypoplasia not correctable by adjusting the NIV interface;
- in the event of the preference of the family or the patient, and based on the experience of the clinical team.

To perform this:

- tracheotomy tubes should be carefully sized and positioned in order to prevent the distal end of the tube being off-centre and rubbing against the tracheal wall;
- the caregivers and parents of children with severe neuromuscular diseases must be trained in the delivery of homecare (endotracheal suctioning).

The advantages of tracheostomy are as follows:

- they leave the face free, making social interaction and eating easier and providing direct access for tracheal suctioning;
- they provide a more secure and more tightly sealed interface for ventilator-dependent patients;
- when children require ventilation for more than 16 hours in each 24-hour period, a tracheostomy may provide a more satisfactory interface between the child and the ventilator than non-invasive ventilation, which children find almost impossible to accept.

Comments

When children require mechanical ventilation for more than 16 hours per day, a non-invasive interface may be difficult to accept or poorly tolerated (specificity: facial deformations in children).

A literature review with an international peer review committee composed of 84 experienced clinicians (9) concerned the multidisciplinary management, in particular respiratory, of patients with Duchenne muscular dystrophy (DMD), and focused partially on the criteria for use of invasive ventilation. (**Expert opinion**)

The indications for tracheostomy in patients with Duchenne muscular dystrophy (DMD) depend on the following factors:

- patient and clinician preferences;
- inability of the patient to successfully use non-invasive ventilation;
- inability of the local medical infrastructure to care for a non-invasive ventilation-dependent patient;
- three failures to achieve extubation during an acute episode despite optimum use of non-invasive ventilation and mechanically assisted cough;
- failure of non-invasive methods of cough assistance to clear lung secretions and oxygen saturation below 95% or equal to the patient's baseline, necessitating frequent direct tracheal suctioning via tracheostomy.

Comments

The methodology for these guidelines is solid. This international literature review is based on a selection of 489 articles. The articles were selected on the basis of the experience of the expert committee using the RAND/UCLA Appropriateness Method, i.e., a critical analysis of the literature and an anonymous vote. However, very few randomised controlled trials were found, which can be explained by the ethical impossibility of randomising patients (invasive *vs* non-invasive ventilation).

The German Medical Association of Pneumology and Ventilatory Support (10) published guidelines for home mechanical non-invasive and invasive ventilation. These guidelines are based on an exhaustive review of the literature, with peer reviewers. (**Expert opinion**)

Before the initiation of ventilation via tracheostomy, it is necessary to:

- fully inform the patient and his/her family about the course of the disease prior to the decision being made;
- obtain consent from the patient or legal representative (parent, legal guardian, etc.).

If the patient so requests or gives his/her consent, tracheostomy should be indicated in the following situations:

- inability to fit an appropriate interface for effective NIV;
- NIV intolerance;
- NIV inefficacy;
- severe bulbar symptoms with recurrent aspiration;
- inefficacy of bronchopulmonary congestion management using non-invasive cough methods;
- impossible to switch to NIV after intubation and invasive ventilation, in the absence of acute complications.

In emergency situations, a decision to perform a tracheostomy can be made even in the event of advance healthcare directives opposing this, since it can always be reversed.

An update of these guidelines on mechanical ventilation, and, in particular, the specificities with respect to non-invasive ventilation, was recently published (11), without modifying the indications for invasive ventilation.

A taskforce composed of representatives of 10 member countries of the Pan-American and Iberic Federation of Medical Societies (12) defined the indications and contraindications for tracheostomy in patients with neuromuscular weakness.

The main indications for tracheostomy include protection and access to the airway to remove bronchopulmonary secretions, prolonged mechanical ventilation, upper airway obstruction, and reduction of dead space to facilitate ventilatory weaning.

The main contraindications are coagulation disorders, short neck (neck circumference of 46 cm, with a distance between the cricoid cartilage and the sternal notch of 2.5 cm), enlarged thyroid gland and/or isthmus, infection of the soft tissues in the neck, presence of pulsatile vessels in the tracheal region, local malignancy, or complex ventilatory support required (fraction of inspired oxygen 70%, positive end-expiratory pressure 10 cm H_2O). However, the contraindications listed above can be managed and overcome by an experienced professional. (**Expert opinion**)

1.1. Paediatric specificities

A cross-sectional retrospective study (13) quantified the side effects of nasal mask use in 40 children with respiratory failure, including 14 patients with neuromuscular disorders. Seven (18%) presented transient erythema, nine (23%) presented prolonged erythema, and three (8%) presented skin necrosis. Skin injury was associated with age over 10 years (OR = 16) and use of a commercial mask (OR = 15) and was less frequent in patients with obstructive sleep apnoea. Replacing a commercial mask with a custom-made mask was associated with a reduction in the skin injury score.

In addition, global facial flattening was present in 68% of the patients and maxillary retrusion was present in 37%. (**Level 4**)

A multicentre, prospective observational study (14) was conducted in 60 children with type-1 SMA treated with nusinersen in seven neuromuscular centres in Germany. After six months of treatment, 47 children (77%) improved their CHOP INTEND score by 4 points. The mean change in CHOP INTEND score was 9.0 ± 8.0 points. Nineteen patients (31.1%) demonstrated an improvement in HINE-2 motor milestones. (**Level 2**)

A literature review (15) documented the indications, preoperative considerations, and procedure types for tracheostomy in children . This study reports that tracheostomy is a much less common procedure in paediatric intensive care units (less than 3% of patients), that there is no definite consensus about the length of time a child should remain endotracheally intubated before the placement of a tracheostomy, and that tracheostomy in children continues to remain a predominantly surgical procedure. The authors conclude that children with tracheostomy have a higher risk of adverse events and mortality, which are largely secondary to their comorbidities rather than the tracheostomy. (**Expert opinion**)

2. Complications of tracheotomy

2.1. In children

A retrospective observational study (16) examined the long-term mortality of 426 children undergoing tracheostomy between 2001 and 2011. The mortality and decannulation rates were compared based on tracheostomy indication and age. The median patient age was 1.5 years (3 days to 24 years). The primary indications for tracheostomy included: airway obstruction, congenital neurological disease, acquired neurological disease, congenital respiratory disease, and acquired respiratory disease. Overall, 98 (23%) died during the study period and the Kaplan-Meier survival curve demonstrates 75% survival at 5.9 years (95% confidence interval [3.0-8.0]). Patients undergoing a tracheostomy for airway obstruction were the least likely to die; while patients with acquired neurological disease were most likely to die. A total of 163 patients (38%) were decannulated, 50% of whom 1.2 years after tracheostomy [0.9-1.5]. Patients with congenital neurological disease were the least likely to undergo decannulation. (**Level 4**)

A retrospective observational study (17) assessed a population of children presenting adverse outcomes in the first two years following tracheostomy, between 2009 and 2011. A total of 502 patients underwent tracheostomy. Median age at tracheostomy was 8 years (interquartile range: 1-16 years), and 62.7% had a complex chronic condition. The two-year rates of in-hospital mortality and tracheostomy complication were 8.9% and 38.8%, respectively. In multivariable analysis, the highest likelihood of mortality occurred in children aged less than 1 year compared with 13 years and older (OR = 7.3; [3.2–17.1]; the highest likelihood of tracheostomy complication was in children with a complex chronic condition versus those without a complex chronic condition (OR 3.3, [1.1–9.9]. Total healthcare spending in the two years following tracheostomy was $53.3 million, with hospital, home and primary care constituting 64.4%, 9.4%, and 0.5% of total spending, respectively. (**Level 4**)

A retrospective observational study (18) assessed the risk of death following tracheostomy in a paediatric population. A total of 513 children (≤ 18 years) underwent a tracheostomy between 1984 and 2015. The highest mortality rate (27.8%) was observed in patients aged 13 to 18 years; their mortality rate was significantly higher when compared to the lowest mortality risk group patients (aged 1 to 4 years, $p = 0.03$). Patients with cardiopulmonary disease had an increased risk of mortality compared with patients with airway obstruction (HR = 3.53, [1.72-7.24], $p < 0.001$) and other indications. Adjusted hazard ratios for bronchopulmonary dysplasia (BPD) and congenital heart disease (CHD) were 2.63 and 2.61, respectively ($p < 0.001$). (**Level 4**)

A retrospective observational study (19) determined predictive factors for complications following tracheostomy placement in 206 patients younger than 2 years, followed up between 2012 and 2013 in 61 surgical facilities in the USA. Fifty patients (24.3%) experienced a major complication within 30 days following tracheostomy. The most common complications were pneumonia (16 [7.8%]), postoperative sepsis (12 [5.8%]), death (12 [5.8%]), and deep or organ space surgical site infections (8 [3.9%]). The predictive factors were: neonatal age (OR = 2.38, [1.06-5.37], $p = 0.04$), intraventricular haemorrhage (OR = 2.72, [1.01-7.32], $p = 0.048$). The authors conclude that young children undergoing tracheotomy tube placement have high rates of morbidity. (**Level 4**)

2.2. In adults

A prospective study (20) with reference to the literature compared the complications of translaryngeal endotracheal intubation and tracheotomy in 150 ICU patients. Complications were observed in 62% of endotracheal intubations and 66% of tracheostomies. The most frequent complications were secondary to excessive cuff pressure (19%), self-extubation (13%) and inability to seal the airway (11%). The most common problems of tracheostomy included infectious complications (36%), local haemorrhage (36%), the consequences of excessive cuff pressure (23%) and subcutaneous emphysema complicated or otherwise by pneumomediastinum (13%). The complications of tracheostomy were generally judged to be more severe than those of endotracheal intubation. There was a higher prevalence of tracheal stenosis after tracheostomy than after endotracheal intubation (65% *vs* 19%, $p < 0.01$). Autopsy of patients who died revealed laryngotracheal injury in 39/41 (95%) intubated patients and 20/22 (91%) tracheostomised patients. It also revealed ulcers on the posterior aspect of the vocal cords in 51% of patients of patients having died after endotracheal intubation. (**Level 2**)

Comments

This historical prospective study is cited in several studies that serve as references in the respiratory management of neuromuscular disorders. However, it currently presents a retrospective bias, insofar as the performance and hygiene conditions for the procedure have improved significantly in the past 40 years. It therefore over-estimates the frequency and severity of current complications.

The German Medical Association of Pneumology and Ventilatory Support (10) highlights the following adverse effects of invasive ventilation via tracheostomy: increase in secretions and respiratory infections, dysphagia, development of granulomas and tracheo-arterial fistulas with cataclysmic haemorrhage. These guidelines are based on the articles described below.

A descriptive study (21) conducted an autopsy evaluation of tracheobronchomalacia (TBM) in seven patients with Duchenne muscular dystrophy (DMD) receiving long-term ventilation via tracheostomies over a period of 5 to 30 years. Five of the seven patients presented variable degrees of tracheobronchomalacia. Two of these patients also had tracheal perforations, one of which resulted in a fatal haemorrhage from a tracheovascular fistula. The authors conclude that, over time, patients receiving positive-pressure ventilation (also receiving non-invasive ventilation) can develop airway thinning and dilation; and on invasive ventilation, tracheal erosion, which can lead to fatal haemorrhage in contact with an adjacent artery. (**Level 4**)

A retrospective study (22) examined tracheal haemorrhages associated with the use of tracheostomy in 49 patients with Duchenne muscular dystrophy. A total of seven patients died. Of those, six patients presented a massive tracheal haemorrhage. This haemorrhage was the cause of death for five of these patients. (**Level 4**)

Another prospective study (23) focused on home ventilator failure and its consequences. The study concerned 150 patients ventilated by tracheostomy for 15.4 hours per day on average, and

demonstrated that the most common complications were secondary to defective equipment or mechanical failure of the ventilator, unrelated to tracheostomy. (**Level 4**)

An examination of the FDA (Food and Drug Administration) database in 2010 (24) detailed 11 deaths related to home ventilation. These deaths concerned patients on invasive ventilation and included two cases of tracheotomy tube displacement, two cases of presumed ventilator malfunction and four cases presumed to be secondary to alarm failures. In other cases, the causes of death were multifactorial or inadequately supported. These results illustrate the need to train patients, families and home caregivers in the risks associated with home ventilator care.

Other studies (7 retrospective studies, 1 national survey, 3 case studies, 4 literature reviews) focus on the complications of tracheostomy in a general population and are summarised in annex 1.

3. Tracheostomy methods and performance

The German Medical Association of Pneumology and Ventilatory Support (10) makes the following recommendations to prevent or minimise the risks of complications (**Expert opinion**):

- perform a tracheostomy with a view to home ventilation when the patient is in a stable condition;
- use a flexible and adjustable tracheotomy tube so that it can be centred in the trachea;
- check using a flexible endoscope that the end of the tracheotomy tube is properly centred in the trachea and that the tracheal mucosa is normal.

A taskforce composed of representatives of 10 member countries of the Pan-American and Iberic Federation of Medical Societies (25) developed recommendations relative to the management of adult tracheostomy patients. The group identified 23 relevant questions among 87 issues that were initially identified. In the initial search, 333 relevant publications were identified of which 226 publications were chosen. The task force generated a total of 19 recommendations, including the following (**Level 1**):

- it is preferable to use a percutaneous method aimed at reducing infectious complications (**Grade 1B**);
- since different percutaneous methods exist, the choice of method should take into consideration the operator's experience, clinical judgement and local practice (**Grade 2D**);
- a percutaneous method at the patient's bedside is preferable to the surgical method in the operating theatre, in order to reduce costs (**Grade 2C**);
- it is suggested that the specialist performing the percutaneous method should receive training (**Grade 2D**).

These guidelines are based on three meta-analyses and a randomised, controlled clinical trial summarised in the articles below:

The meta-analysis (26), which included 17 randomised, controlled trials and 1,212 patients, demonstrated a significant reduction in wound infection using a percutaneous method compared to the surgical method ($p < 0.0005$). A possible explanation for this difference is the minimally invasive nature of percutaneous techniques. There was no difference in terms of bleeding complications.

Another meta-analysis (27), including 15 prospective, randomised controlled trials involving nearly 1,000 patients, revealed fewer complications in the "percutaneous group" with respect to wound infection ($p = 0.0002$) and "unfavourable scarring" ($p = 0.01$). However, decannulation and obstruction complications were significantly more common with the percutaneous technique.

Finally, the results of the most recent meta-analysis (28) confirm that there is a lower risk of minor stoma inflammation ($p = 0.006$) and infectious complications ($p < 0.00001$) with percutaneous techniques. The reduction in risks with percutaneous methods is primarily related to the gradual abandon of the multiple dilator technique. The risk of postoperative bleeding was also lower with the percutaneous technique ($p = 0.04$), but this finding comes from a single study conducted with the modern percutaneous tracheostomy technique proposed by Fantoni (39% in terms of weight). Apart from this study, no difference between the two methods was observed with respect to bleeding complications.

A randomised controlled study (29) having compared percutaneous versus surgical tracheostomy is of particular interest. No significant difference was found in the combined primary outcome measure (bleeding, infection, pneumothorax, accidental decannulation and other major surgical complications).

The overall complication rate was low, in the region of 3.5%. However, there were fewer stoma infections in the “percutaneous technique” group.

The French-language Intensive Care Society and the French Society of Anaesthesia and Intensive Care Medicine (30) developed recommendations for the use of tracheostomy in intensive care units, in adults and outside emergency contexts. The recommendations were developed using the GRADE method. The expert panel formulated the following recommendations:

Before tracheostomy

- The experts suggest that the indication for tracheostomy in patients with chronic respiratory failure should be the subject of multidisciplinary discussion.
- Tracheostomy in intensive care should not be performed before the fourth day of mechanical ventilation. (**Grade 1A**)
- The experts suggest that tracheostomy (percutaneous or surgical) should not be performed in intensive care units in situations at high risk of complications (haemodynamic instability, intracranial hypertension > 15 mmHg; severe hypoxaemia: PaO_2/FiO_2 < 100 mmHg with PEP > 10 cmH_2O; uncorrected bleeding disorders [platelets < 50,000/mm^3 and/or INR > 1.5 and/or PTT > 2 times normal]; refusal by the patient and/or family; patient in the terminal stage or active treatment withdrawn). (**Expert opinion**)
- The experts suggest that medical and surgical teams should discuss and decide upon the tracheostomy technique to be used when there is a risk of complications. (**Expert opinion**)

Concerning the technique used

- Percutaneous tracheostomy should be preferred as the standard method in intensive care patients. (**Grade 1+**)
- Percutaneous dilatational tracheostomy using a single dilator should be preferred as the standard method in intensive care patients. (**Grade 2+**)
- Fibre-optic bronchoscopy should be performed before and during percutaneous tracheostomy. (**Grade 2+**)
- A laryngeal mask airway should not be used during percutaneous tracheostomy in intensive care. (**Grade 2A**)
- Cervical ultrasound should be performed with percutaneous tracheostomy in intensive care. (**Grade 2+**)
- The experts suggest that antibiotic prophylaxis should not be prescribed for tracheostomy. (**Expert opinion**)
- The experts suggest that a standardised procedure should be implemented in intensive care units that perform percutaneous tracheostomy. (**Expert opinion**)

Concerning tracheostomy decannulation methods

- Le experts suggest that a multidisciplinary decannulation protocol should be available in intensive care units. (**Expert opinion**)
- The tracheotomy tube cuff should be deflated when the patient is breathing spontaneously. (**Grade 2+**)
- A pharyngolaryngeal examination should be performed at or following decannulation. (**Grade 2+**)

Percutaneous tracheostomy is recommended as the first-line approach standard method; however, it has contraindications.

A prospective study (31) compared obese patients (body mass index > 30) and non-obese patients who underwent percutaneous tracheostomy at a tertiary medical-surgical intensive care unit between May 2004 and October 2005. During the study period, 227 percutaneous tracheostomies were performed (50 in the obese patient group and 177 in the non-obese patient group). Major complications were significantly higher in obese patients (12% versus 2%, p = 0.04). (**Level 2**)

A retrospective study (32) reviewed the charts of all morbidly obese patients (BMI > 35) having undergone either percutaneous tracheostomy (PT) or open tracheostomy (OT) over a period of 58 months. A total of 1,062 tracheostomies were performed between 2000 and 2004. One hundred and forty-three patients had a BMI ≥ 35 kg/m^2. Eighty-nine patients underwent PT and 53 patients underwent OT. Events were reported in 5 patients (5.6%) undergoing PT (4 conversions to OT, 1 malpositioning) and in 3 patients (5.6%) undergoing OT (malpositioning resulting in hypoxia, bleeding requiring surgical intervention, aborted attempt at open surgery). (**Level 4**)

The French-language Intensive Care Society and the French Society of Anaesthesia and Intensive Care Medicine (30) suggested the following contraindications for percutaneous tracheostomy: an unstable cervical spine, an anterior cervical wound or infection, a neck that has been treated (surgery or radiotherapy), difficulty in identifying anatomical landmarks (e.g., obesity, short neck, thyroid hypertrophy), or stiffness of the cervical spine, prompting the use of surgical tracheostomy. (**Expert opinion**)

The Danish Society of Intensive Care Medicine (33) developed guidelines for percutaneous dilatational tracheostomy. This working group recommends that surgical tracheostomy be a second-line approach for the general population, with percutaneous tracheostomy being used as a first-line approach (**Expert opinion**). The expert group makes the following recommendations.

Absolute contraindications to percutaneous tracheostomy:

- unstable fractures of the cervical spine;
- severe local infection of the anterior neck;
- uncontrollable coagulopathy;

Relative contraindications to percutaneous tracheostomy:

- controlled local infection or wounds or burns or local radiotherapy;
- controllable coagulopathy;
- complex mechanical ventilation (high PEEP and/or FiO_2 requirements);
- difficult anatomy (e.g. tracheal deviation);
- haemodynamic instability;
- elevated intracranial pressure.

4. Post-tracheostomy follow-up

4.1. Management of care (hygiene, care, equipment, personnel, training/certification of caregivers and families in routine care and emergency procedures)

The French-language Intensive Care Society and the French Society of Anaesthesia and Intensive Care Medicine (30) developed recommendations for the use of tracheostomy in intensive care units, in adults and outside emergency contexts. The recommendations were developed using the GRADE method. The expert panel formulated the following recommendations concerning tracheotomy monitoring and maintenance (**Expert opinion**):

- the experts suggest that intensive care units should have a tracheotomy management protocol;
- they also suggest that special attention should be paid to securing the tracheotomy tube, maintenance of a corrugated tube, and prevention of repeated local trauma caused by the moving and weight of the tubes (avoid pulling the tracheotomy tube);
- the experts consider it useful to check the position of the tracheotomy tube (chest X-ray, ease of tracheal suctioning, absence of dyspnoea) and, if necessary, to use fibre-optic bronchoscopy to look for injury or stenosis, without specifying the frequency or timing.

The American Thoracic Society (34) developed clinical practice guidelines for paediatric patients on home invasive ventilation. The grading of recommendations assessment, development, and evaluation (GRADE) methodology was used to formulate and grade the following recommendations relative to discharge to home post-tracheostomy (**Expert opinion**):

- the child must be medically stable for discharge:
  - no significant change to ventilator settings or oxygen requirement for at least several days and preferably several weeks before discharge,
  - no acute decompensation events within the few days to weeks before discharge,
  - home respiratory equipment trialled and tolerated in the hospital for at least 24 to 48 hours before discharge,
  - transport tolerated by the child;
- family caregivers must demonstrate their willingness and ability to care for the patient, which involves the following:
  - delivering all prescribed therapies,
  - demonstrating competency in the care and replacement of their child's tracheotomy tube (caregiver education must include recognising and responding to urgent issues such as tube obstruction, decannulation, and bleeding from tracheotomy),
  - at least two family caregivers must be fully trained in all aspects of the child's care,
  - regular hand washing (this is essential and its importance cannot be overemphasised),
  - caregivers must be able to safely transport the child in both routine and urgent situations (a "Go Bag" with all necessary travel items, including an extra tracheotomy tube and obturator, a size smaller tracheotomy tube, suction catheters, scissors, tracheotomy tube ties, and lubricant, will remain with the child at all times),
  - family caregivers must be instructed not to smoke near their ventilated child;

- a home care service provider must be available and able to provide the required equipment and technical support:
  - perform a home inspection to confirm that the home environment and electrical systems are adequate for the necessary medical equipment,
  - provide 24-hour availability as a resource and to service the equipment, including same-day replacement of malfunctioning equipment,
  - respiratory technicians should visit patients at least monthly and more often as needed;
- home professional caregivers (nurses, physiotherapists, etc.) must be trained.

4.2. Management of mucus congestion in tracheostomy patients

4.2.1. Regulatory framework

It is important to refer to the regulatory framework introducing the conditions for the performance of home care in a tracheostomy patient by healthcare professionals and home or family caregivers.

Nurses

(French Decree No. 2004-802 of 29 July 2004).

Book III - Title I: Nursing profession Chapter I Practising the profession - Section 1 Professional procedures.

Article R.4311.5: As part of their specific role, nurses perform the following procedures or deliver the following care aimed at identifying risks and ensuring the comfort and safety of patients and their environment, including informing patients and their families:

15° Suctioning of secretions, irrespective of whether or not the patient is intubated or has a tracheostomy;

18° Administration of non-medicinal products by aerosol;

28° Mouth care with application of non-medicinal products;

38° Participation in the disinfection and sterilisation procedure for reusable medical devices.

Article R.4311.7: Nurses are authorised to perform the following procedures, either in application of a medical prescription which, except in emergencies, is written, qualitative and quantitative, dated and signed, or in application of a previously defined qualitative and quantitative written protocol, dated and signed by a physician:

22° Care and monitoring of intubated or tracheostomised patients, the first tracheostomy tube change being performed by a physician;

24° Administration of medicinal products by aerosol or spray;

25°Mouth care, with application of medicinal products and, if required, instrumental assistance;

30° Checking that assisted ventilation or monitoring devices are working properly, control of the various settings and monitoring of patients on these devices;

31° Placement of an oxygen tube;

38° Sampling and collection of secretions and excretions.

Physiotherapists

Articles 8 and 9 of French Decree No. 2000-577 of 27 June 2000 relative to the professional procedures and practice of the physiotherapy profession qualify physiotherapists to intervene in the respiratory rehabilitation of patients, on medical prescription and on condition that a physician can intervene at any time:

- to perform rhinopharyngeal and tracheal suctioning in tracheostomised or intubated patients;
- to administer non-medicinal or medicinal products prescribed by the physician by aerosol prior to application of clearance and supportive methods;
- to measure the peak respiratory flow.

Healthcare assistants

Healthcare assistants work under the responsibility of nurses, within the framework of the role given to them by the latter, defined by articles 3 and 5 of French Decree No. 2002-194 relative to the professional procedures and practice of the nursing profession. This therefore concerns care related to medical devices or equipment: oxygen therapy (assembly and maintenance of equipment), patient monitoring, assembly and maintenance of endotracheal suctioning equipment, administration of non-medicinal aerosols; care of dependent persons: non-medicinal mouth care.

Home or family caregivers

Legislative rules relating to tracheal suctioning.

French Order of 27 May 1999 relative to the training of persons authorised to perform endotracheal suctioning.

Article 1: The length of training for the persons mentioned in article 1 of the abovementioned French decree of 27 May 1999 is five days, including two days of theoretical training followed by three days of clinical training in a department caring for tracheostomised patients.

Article 4: Family members of tracheostomised patients may follow the training mentioned in article 1 of the present Order in the department in which the tracheostomised patient was cared for. The head of department assesses the theoretical and clinical knowledge of the parties concerned and delivers a certificate accordingly.

French Decree No. 2015-495 of 29 April 2015 relative to the certification of home caregivers to perform endotracheal suctioning stipulates the regulatory provisions relating to the missions of home assistance and support and patient care services, indicating that their employees may perform tracheal suctioning as long as they meet certain training conditions scheduled by the French Public Health Code and French Decree No. 99-426 of 27 May 1999 qualifying certain categories of persons to perform endotracheal suctioning.

4.2.2. Suctioning (advantages and disadvantages)

The *Haute Autorité de santé* (French National Authority for Health) (35) assessed the benefit of the tracheal suctioning devices included on the list of products and services approved for reimbursement (LPPR), with a view to their public funding. The aim of this assessment was to determine the indications and conditions for prescription and use of tracheal suctioning devices, drawing on the articles indicated below.

The American Association for Respiratory Care (36) developed guidelines relative to nasal, oropharyngeal and endotracheal suctioning at home in patients with or without tracheostomy/laryngectomy. However, the development methodology was not clearly described (**Level 4**):

- the main indication for suctioning in patients cared for at home is patients' inability to effectively clear their airways by coughing;
- the suctioning procedure should only be envisaged when this indication is clearly present;
- pre-oxygenation should not be performed routinely. In patients with neuromuscular diseases, experience appears to show that when the vital capacity is < 1.5 l, hyperinflation may replace tracheal suctioning, which generally becomes pointless in this case. Pre-oxygenation may be indicated in paediatric patients with a reduced respiratory reserve, patients in whom oxygen desaturation has been documented, patients with cardiac dysrhythmias, ventilated patients;
- the routine use of normal saline installation is not recommended.

Comments

Recommendations of poor methodological quality since not explained.

The American Association for Respiratory Care (37) produced new guidelines on endotracheal suctioning in mechanically ventilated patients based on an electronic literature search for articles published between January 1990 and October 2009, using the MEDLINE, CINAHL and Cochrane Library databases. A total of 114 clinical trial, 62 literature reviews and 6 meta-analyses on tracheal suctioning were analysed. The following recommendations were developed using the criteria of the GRADE method.

- Suctioning frequency and duration:
  - it is recommended that endotracheal suctioning should be performed only when secretions are present, and not routinely (**Grade 1C**);
  - it is suggested that the duration of the suctioning event be limited to less than 15 seconds (**Grade 2C**);
  - use of shallow suction is suggested instead of deep suction, based on evidence from infant and paediatric studies (**Grade 2B**).
- Pre-oxygenation:
  - it is suggested that pre-oxygenation be considered if the patient has a clinically important reduction in oxygen saturation with suctioning (**Grade 2B**).
- Normal saline instillation:

    - normal saline instillation prior to endotracheal suctioning is not recommended (**Grade 2C**).
- Suction tube:
    - it is recommended to perform suctioning without disconnecting the patient from the ventilator (**Grade 2B**);
    - it is suggested that a suction catheter be used that occludes less than 50% of the lumen of the endotracheal tube in children and adults, and less than 70% in infants (**Grade 2C**).

The American Thoracic Society (38) developed a consensus relative to the care of children with long-term tracheostomy, based on an international multidisciplinary working group, reviewed by a learned society committee. (**Expert opinion**)

- A clean technique is recommended for home care. All caregivers should thoroughly wash their hands before and after each suctioning procedure. Alcohol or disinfectant foam is an acceptable substitute when soap and water are unavailable for handwashing. Non-sterile, disposable gloves should be worn for the protection of any caregiver that is not a family member or by anyone who is concerned about infection.
- After suctioning is complete, the catheter is flushed with tap water until secretions are cleared from the lumen. The outside of the catheter is then wiped with alcohol and allowed to air dry. A hydrogen peroxide flush is useful when particularly adherent secretions are present. The catheter is then stored in a clean, dry area.
- Catheters can be reused by the same patient as long as the catheter remains intact and allows inspection of removed secretions.
- There are a number of methods available for more thorough cleaning including commercial products, alcohol, and a vinegar-and-water soak. There are no data to suggest the frequency of “more thorough” cleaning or that one method is better than another.
- The premeasured technique is recommended for all routine suctioning.
- Suctioning should be done on the basis of clinical assessment. In children with no evidence of secretions, a minimum of suctioning, in the morning and at bedtime, to check for patency of the tube is recommended.
- The routine instillation of normal saline is not recommended.
- The largest size catheter that will fit inside the tracheostomy tube is recommended, because a large-bore tube will remove secretions more efficiently than the previously recommended smaller size tube.
- Suction pressure: pressures of 80 to 100 mm Hg are typically used for paediatric patients. Suction should be applied both while inserting and removing the catheter. The suction should be adequate to efficiently remove secretions with a rapid pass of the catheter. Before patient discharge, the home tracheal suction machine should be tested to ensure that it is adequate to clear the patient's secretions.
- A rapid technique that is completed in less than 5 s is recommended. This is vital when using a large suction catheter, relative to the tracheostomy tube size, to prevent atelectasis.

Comments

The recommendations are of mediocre methodological quality (no published systematic review is available) and are based on expert opinions. The evidence levels and grading of the recommendations are not explained. And the authors’ interests and the funding are not specified.

The Canadian Thoracic Society (39) developed a clinical practice guideline for the management of patients on home mechanical ventilation, based on a peer-reviewed literature review. Some of these recommendations concerned patients on invasive ventilation: (**Expert opinion**)

- minimally invasive rather than deep suctioning is recommended when possible;
- clean, as opposed to sterile, conditions are adequate for home secretion clearance and suctioning.

Comments

The methodology is adequately detailed (several data sources, study selection criteria mentioned and restriction to English-language articles). However, little information is given about tracheal suctioning.

The Australian Agency for Clinical Innovation (40) developed a clinical practice guideline on endotracheal suctioning in mechanically ventilated ICU patients by searching several databases (PubMed, CINAHL and Cochrane), between 2006 and June 2012. The recommendations are as follows. (**Expert opinion**)

Indication criteria:

- The decision to suction must be made on the basis of the clinical need to maintain the patency of the tracheobronchial tree. A tracheal tube should only be suctioned when clinically indicated by signs which could include:
  - visible, palpable or audible secretions (such as sputum, gastric or upper airway contents or blood);
  - respiratory symptoms: desaturation, rising peak inspiratory pressure (during volume-controlled mechanical ventilation), decreasing tidal volume (during pressure-controlled ventilation), increased respiratory rate, increased breath sounds on auscultation;
  - cardiovascular symptoms: increased heart rate and blood pressure;
  - other reasons: restless/agitated or diaphoretic patient;
  - desaturation, a “saw-tooth pattern” on the ventilator graphics.

Suctioning time:

- The total suction procedure (from insertion to removal of catheter) should take a maximum of 15 seconds with negative pressure applied continuously as the catheter is being withdrawn from the tracheal tube.

Suction tube:

- The size of the suction catheter should be less than half the internal diameter of the tracheal tube.
- Tracheal tubes with subglottic secretion drainage should be used for mechanically ventilated patients expected to require ventilation for more than 72 hours.

Normal saline instillation:

- To prevent the occurrence of adverse events, instillation of normal saline should not be routinely used prior to suctioning.

Comments

Clinical practice guideline with the development methods described.

A peer-reviewed literature review (41), based on the GRADE method, identified seven controlled trials in the Medline and CINAHL, databases concerning the value of normal saline instillation in ICU patients. The authors conclude that the procedure significantly decreases oxygenation (**Grade B**). However, the effects on haemodynamics and pneumonia incidence remain controversial.

Comments

Additional studies are required to determine the efficacy of instillation of solutions other than normal saline in the management of these secretions.

Furthermore, it is important to highlight the discomfort, pain and distress associated with tracheal suctioning described in the articles below.

A retrospective observational study (42) examined pain associated with clinical procedures routinely performed in intensive care units, including endotracheal suctioning. This study was conducted in 6,201 patients (176 patients aged under 18 years and 5,957 adults, 68 missing data). The data concerning endotracheal suctioning concern 784 patients. In adults, the mean pain intensity score for tracheal suctioning was 3.94 ± 3.32 on a scale of 0 to 10 (with 10 being the most intense pain), and procedural distress was 3.15 ± 3.27 on a scale of 0 to 10 also (10 being the most intense distress). The scores were the highest (5.00 ± 3.16 for pain and 4.57 ± 3.10 for distress) in the 13-17 year age group. (**Level 4**)

A retrospective observational study (43) examined the care given paediatric tracheostomy patients in their homes (use of equipment and frequency of care, etc.) and the incidence of pneumonia. To this end, 60 patients completed a questionnaire sent out between May 1995 and June 1996. This study provided descriptive information, including the fact that 40.7% of patients systematically performed suctioning every 1 to 4 hours, not as needed. The study also analysed the occurrence of pneumonia and noted that 60.6% (20/33) of patients reusing their tracheostomy tubes reported pneumonia within the previous year, versus 25.9% (7/27) of patients who systematically changed the tracheostomy tube. (**Level 4**)

Comments

The Bahng, 1998 article is of low methodological quality. However, it identifies the performance of systematic tracheal suctioning, rather than on an as-needed basis, despite the fact that this procedure can cause adverse events.

Mechanical cough assistance methods such as insufflation-exsufflation are reported as being less dangerous and can replace tracheal suctioning in the studies below.

A prospective (before/after) study (44) compared the effects of mechanical insufflation-exsufflation versus suctioning via tracheostomy tubes on respiratory variables for six amyotrophic lateral sclerosis patients. The baseline values were 93.50 ± 2.26% for pulse oxyhaemoglobin saturation, 18.50 ± 4.23 cm H_2O for peak inspiratory pressure, 4.67 ± 1.37 cm H_2O for mean airway pressure, and 1.03 ± 0.25 J/litres for work of breathing performed by the ventilator. Only work of breathing performed by the ventilator decreased after tracheal suctioning ($p < 0.05$), whereas all variables improved significantly after mechanical insufflation-exsufflation. (**Level 2**)

A cross-sectional study (45) evaluated the preferences of spinal cord injury patients concerning secretion management techniques. The survey was conducted in 18 patients with a spinal cord injury, with an average age of 34 years. On the ASIA (American Spinal Injury Association) impairment scale (AIS), 72% of patients had an ASIA A score, 22% and ASIA B score and 6% an ASIA C score, with a neurological level of injury ranging from C1 to T3. The results indicate that patients found mechanical insufflation-exsufflation significantly less irritating ($p < 0.001$), less painful ($p < 0.001$), less tiring ($p = 0.01$) and less uncomfortable ($p < 0.001$) than endotracheal suctioning. In a direct comparison, 89% of patients preferred mechanical insufflation-exsufflation to suctioning. In addition, 89% of patients found insufflation-exsufflation faster, 78% found it more convenient, and 72% found it more effective than suctioning. (**Level 4**)

A retrospective study (46) evaluated the safety, tolerance, and effectiveness of mechanical insufflation-exsufflation in a population of 62 mechanically ventilated patients with neuromuscular disease , of whom 34 were boys. The median age at initiation of mechanical insufflation-exsufflation use was 11.3 years (range: 3 months to 28.6 years). Mechanical ventilation via tracheostomy was used in 29 patients, and 25 patients used non-invasive ventilation. The median duration of use was 13.4 months (range: 0.5 and 45.5 months). Five patients chose not to continue mechanical insufflation-exsufflation treatment. Complications were reported in two patients, but ultimately they used the mechanical insufflation-exsufflation device. Chronic atelectasis resolved in four patients after beginning mechanical insufflation-exsufflation therapy, and five patients experienced a reduction in the frequency of pneumonia. The authors conclude that in 90% of cases, the use of mechanical insufflation-exsufflation was safe, well-tolerated, and effective in preventing pulmonary complications. (**Level 4**)

A prospective study (47) examined the safety and tolerability of mechanical insufflation-exsufflation in patients ventilated by tracheostomy. Mechanical insufflation-exsufflation was applied 26 times to seven male and six female subjects in whom endotracheal suctioning was indicated. The mean age was 62.6 ± 20 years. The mean insufflation tidal volume was 1,043.6 ± 649.9 ml. No statistically significant differences were identified between baseline and post-procedure variables. No complications were detected. All except one of the mechanical insufflation-exsufflation sessions were productive, showing secretions in the proximal artificial airway, and were well tolerated. (**Level 2**)

Comments

These studies report mechanical cough assistance methods such as insufflation-exsufflation as being less dangerous and better tolerated by patients than endotracheal suctioning. However, there

are no studies comparing insufflation-exsufflation with endotracheal suctioning in patients with slowly progressive neuromuscular diseases.

4.3. Management of speech

The following observational studies describe the use of speaking valves in tracheostomy patients in order to restore speech during mechanical ventilation.

A prospective study (48) examined the effect of a voice tracheostomy tube (VTT) in which the cuff deflates on expiration to enable tracheostomy patients to speak on expiration. A total of 16 patients on invasive ventilation were included and were switched to this tube. All the patients except one were able to speak on expiration without any change in PaO_2. Bronchoscopy showed that the cuff of the VTT did not damage the tracheal mucosa. The authors conclude that the VTT enables patients to speak during mechanical ventilation without causing any complications. (**Level 4**)

A physiological study (49) examined the characteristics of speaking valves in tracheostomised patients with neuromuscular diseases during periods of free breathing. Six commercially available speaking valves were studied in a dynamic set-up simulating a respiratory rate of 20 breaths/min, a tidal volume of 0.5 l and a peak flow of 0.5 l/sec. Assessments were performed in 10 tracheostomised patients, using no speaking valve, and with a speaking valve considered to be the most resistant and one considered to be the least resistant among the valves identified. The valves presented a wide range of resistance, ranging from 1.3 to 5.9 $cmH_2O/l/sec$. The additional work of breathing value varied with a ratio of 4.4 between the best and the worst valve. A significant effect on inspiratory difficulty (Borg scale) was observed between the absence of a speaking valve and the most resistant valve. The authors conclude that the variety of aerodynamic characteristics of speaking valves should be considered when choosing the device, according to the underlying condition of the patients benefiting from their use. (**Level 2**)

Studies have described the impact of adjusting mechanical ventilator parameters on speech:

A prospective study (50) compared the effects of two ventilatory modes (assist-control ventilation versus bilevel positive-pressure ventilation) on speech in tracheostomised patients with neuromuscular disease. Nine patients with neuromuscular disease were included. At rest, the ventilatory parameters were similar with both modes. Speech induced an increase in inspiratory time during bilevel positive-pressure ventilation (or bilevel positive airway pressure: BIPAP), with a greater increase in the volume released by the ventilator during speech as compared to with assist-control ventilation (ACV) (172 ± 194, versus 26 ± 31 ml). Consequently, speech duration was longer during inspiration with BIPAP, with speech production able to extend into expiration. Three patients could speak continuously during several respiratory cycles while receiving BIPAP. (**Level 4**)

A physiological study (51) examined the impact of making adjustments to ventilator settings on the speech of tracheostomised patients receiving positive-pressure ventilation. Fifteen adults with spinal cord injuries or neuromuscular diseases were included. The ventilator was adjusted using lengthened

inspiratory time (Ti), positive end-expiratory pressure (PEEP), and combinations thereof. When Ti was lengthened (by 8 to 35% of the ventilator cycle), speaking time increased by 19% and pause time decreased by 12%. When PEEP was added (5 to 10 cm H_2O), speaking time was 25% longer and pauses were 21% shorter. When lengthened Ti and PEEP were combined (with or without reduced tidal volume), their effects were additive, increasing speaking time by 55% and decreasing pause time by 36%. The combined intervention improved speech timing, loudness, voice quality, and articulation. (**Level 4**)

A prospective study (52) compared the effects of a speaking valve versus external PEEP in tracheostomised ventilator-dependent neuromuscular patients. Between December 2008 and April 2009, 10 patients with neuromuscular disorders were included. The text reading time, perceptive score, intelligibility score, speech comfort, and respiratory comfort were similar with PEEP and the speaking valve. During speech, the respiratory rate increased by at least 3 cycles/min above the backup rate in seven patients with PEEP and in none of the patients with a speaking valve. The authors conclude that low-level PEEP is as effective as the speaking valve to ensure good speech quality. (**Level 2**)

5. Alternatives to tracheostomy

Historically, tracheostomy was indicated when night-time ventilation was no longer sufficient, marking the start of ventilator dependency. Since the mid-1980s, the reference alternative to tracheostomy has been non-invasive ventilation, combined with cough management.

5.1. Daytime non-invasive ventilation

5.1.1. Indications for daytime non-invasive ventilation

An analysis of the literature (53) on the use of mechanical ventilation in patients with slowly progressive neuromuscular disease such as Duchenne muscular dystrophy (DMD) resulted in the development of recommendations validated by an independent multidisciplinary working group.

The recommendations concerning night-time, then daytime non-invasive ventilation are as follows:

- in Duchenne muscular dystrophy and, by extension, in all other aetiologies of slowly progressive myopathies, night-time non-invasive ventilation is usually proposed if the patient presents daytime hypercapnia of > 45 or > 50 mmHg (depending on the studies) and oxygen saturation < 92% or < 95% (depending on the studies) or night-time hypercapnia with signs of hypoventilation;
- in the event of progression of chronic respiratory failure, daytime non-invasive ventilation becomes complementary to the initial night-time non-invasive ventilation in the presence of daytime symptoms and/or persistent daytime hypercapnia despite well conducted night-time ventilation.

Comments

The recommendations are not clearly defined with respect to ventilator dependency. Daytime ventilation methods such as mouthpiece ventilation are not cited. These recommendations are primarily based on studies conducted in the context of Duchenne muscular dystrophy (DMD).

In 2011, the Quebec Ministry of Health and Social Services, along with all Canadian learned societies (39) developed guidelines concerning the long-term use of non-invasive ventilation in a population similar to that of the HAS. The optimal time to begin daytime ventilation has not been clearly defined. Studies are in favour of extending night-time ventilation into the daytime when daytime hypercapnia reappears despite properly conducted night-time ventilation. The Quebec Ministry of Health and Social Services drew on the following studies.

In a prospective study (54), daytime ventilation was introduced when transcutaneous $PtcCO_2$ was above 45 mmHg for 2 hours preceding the start of night-time ventilation, thereby demonstrating the efficacy of mouthpiece ventilation on daytime hypercapnia and enabling a median survival of 33 years without tracheostomy when used in combination with cough augmentation and airway clearance techniques. (**Level 4**)

A retrospective study (55) confirmed these results by comparing three groups: patients monitored before 1984 not receiving mechanical ventilation; patients ventilated by tracheostomy between 1984

and 1991; and, since 1991, patients receiving non-invasive ventilation combined with assisted coughing methods. The mean ages of death for the first and second groups were 18.6 ± 2.9 years and 28.1 ± 8.3 years respectively. The last group had a survival median of 39.6 years. (**Level 4**)

In a **prospective study (56)** of 8 patients with Duchenne muscular dystrophy (DMD) followed up for 39 months, the authors demonstrate that once daytime hypercapnia has been controlled by NIV, despite the progression of respiratory muscle failure (decrease in vital capacity of 33% compared to their initial value), the efficacy of NIV (daytime normocapnia) was then maintained thanks to extension to daytime and the gradual increase in the mean duration per day (up to 18 ± 2 hours per day). (**Level 4**)

Studies demonstrate a significant reduction in hospitalisation rates when patients are receiving NIV.

A retrospective study (57) analysed pneumonia and hospitalisation rates in patients living at home, either receiving oxygen therapy, or with tracheostomy tubes but not ventilated, or ventilated by tracheostomy or non-invasive methods. A total of 684 patients out of 1,200 were ventilated. Among the non-DMD neuromuscular patients on NIV, the pneumonia and hospitalisation rates were reduced when ventilation was more than 16 hours per day compared to less than 16 hours per day. (**Level 4**)

A retrospective study (58) assessed the long term physiological and clinical outcome in 79 patients with musculoskeletal disorders (73 patients with neuromuscular disorders, 6 patients with disorders of the chest wall) who received non-invasive ventilation for chronic respiratory failure over a period of 46 years. Hospital admission rates increased nearly eightfold in patients receiving mechanical ventilatory support compared to before mechanical ventilation ($p < 0.01$). However, after the initiation of daytime mouth ventilation, the hospitalisation rates fell by 36%. (**Level 4**)

Comments

The current literature is poor and of low methodological quality. However, it suggests that switching to daytime non-invasive ventilation reduces the hospitalisation rate.

After conducting a review of the MEDLINE databases, **an American Thoracic Society consensus committee (59)** developed the following recommendations relative to daytime NIV:

- need for monitoring of patients at least annually: blood gas exchange or SaO_2 + end tidal CO_2 levels ($PETCO_2$);
- indication for daytime NIV: $PETCO_2$ > 50 mmHg or when oxygen saturation remains < 92% during the daytime.

A physiological prospective study (60) demonstrated the benefit of adding daytime NIV to night-time NIV in a population of patients with neuromuscular diseases when the present daytime clinical signs of poor respiratory tolerance, predominantly at the end of the day. It demonstrated, in 50 patients with DMD presenting these symptoms, that a 2-hour session of daytime non-invasive ventilation in the

middle of the day improved dyspnoea at the end of the day (Borg score), respiratory muscle endurance and tension-time index (representing respiratory muscle work). (**Level 2**)

An international expert group led by Newcastle developed recommendations on the overall management, including respiratory, of DMD patients (9) using the RAM method (RAND Corporation–University of California Los Angeles Appropriateness Method), based on a literature review and the opinion of 84 international experts. (**Expert opinion**)

Concerning the indication for daytime ventilation, the recommendations are as follows:

- swallowing difficulty (feeding) due to dyspnoea, relieved by ventilatory support;
- inability to speak a full sentence without dyspnoea;
- symptoms of daytime hypoventilation with $SpO_2 < 95\%$ and/or $PaCO_2 > 45$ mmHg.

This working group updated these recommendations in 2018 (61) without modifying the respiratory management. (**Expert opinion**)

5.1.2. Impact of NVI via mouthpiece (or nasal)

A peer-reviewed Cochrane review (1) compared the efficacy of non-invasive ventilation with invasive ventilation in people with neuromuscular disease and chest wall disorders. A literature search in the databases was performed by two independent experts. (**Expert opinion**)

The authors report the following advantages of non-invasive ventilation:

- NIV can be delivered using a broad variety of patient/ventilator interfaces, such as nasal, buccal or full-face masks and mouthpieces;
- evidence demonstrating that daytime ventilation can be effective via a mask or mouthpiece (rather than tracheostomy) is based on observational studies conducted in DMD patients indicated below.

A retrospective study (62) reported the use of continuous ventilation via a mouthpiece in 257 people with acute or chronic respiratory failure, with follow-up for 13.4 years. No significant complications were demonstrated. The authors concluded that for individuals with chronic respiratory muscle insufficiency but adequate bulbar muscle function, NIV via a mouthpiece can be an effective alternative to tracheostomy. (**Level 4**)

In the previously cited study (57), regarding pneumonia and hospitalisations, no significant difference was observed in tracheostomised and non-tracheostomised DMD patients. (**Level 4**)

A previously cited prospective study (54) assessed the impact of daytime ventilation via mouthpiece on the survival of DMD patients. A total of 42 DMD patients aged 15 to 33 years were studied. The survival rates were 88% after 1 year, 58% after 5 years and 51% after 7 years. The mean age of death was 31 years. The vital capacity stabilised for 5 years, with an improvement in clinical condition and normalisation of $PTcCO_2$ during the daytime (8.17 ± 2.22 to 5.78 ± 0.73 kPa). No accidents or minor adverse effects were observed. The authors conclude that daytime mouthpiece ventilation is safe, prolongs survival and stabilises vital capacity in DMD patients. (**Level 2**)

Two other studies concerning a population of mouthpiece ventilation-dependent DMD patients support the conclusions of the abovementioned article.

A prospective study (63) in 12 ventilation-dependent DMD patients having used night-time mask NIV and mouthpiece NIV during the daytime. The mean age of initiation of nocturnal NIV was 17.8 ± 3.5 years and the mean vital capacity was 21% of the predicted value. The mean age of initiation of daytime NIV was 19.8 ± 3.4 years and the mean vital capacity was 13.2% of the predicted value. The results of this study demonstrated an improvement in CO_2 levels. The mean survival on 24-hour NIV was 5.7 years (range 0.17 to 12 years). Of the 12 patients, two deaths occurred after 3.75 and four years, respectively. (**Level 2**)

A retrospective study (64) reported the benefits of daytime mouthpiece ventilation in DMD patients, consulting the records of 300 patients followed up between 1995 and 2013. Out of 300 patients, 79 used night-time NIV (8 hours or more) and 20 became dependent on daytime mechanical ventilation. The patients were aged from 24 to 51 years. Four of them died from cardiac causes and one patient switched to invasive ventilation via tracheostomy. The authors conclude that NIV can prolong survival without resorting to tracheotomy and without hospitalisation. (**Level 4**)

As regards the life expectancy of ventilation-dependent neuromuscular patients, six observational studies (three prospective and three retrospective/historical studies) assessed the impact of ventilation methods (non-invasive versus invasive), on the survival of patients with neuromuscular diseases.

A prospective observational study (65) compared morbidity and causes of death in 42 DMD patients on full-time mechanical ventilation, either via tracheostomy (n = 16) with a mean age of 32.7 years, or using non-invasive methods (n = 26) with a mean age of 27 years, over a 5-year period (2002-2006). The patients were receiving similar treatments (airway clearance and medicinal). The authors report more frequent morbidity, mucus hypersecretion and tracheal injury in tracheostomy patients than in NIV patients. However, weight loss and the need for gastric feeding appeared less frequent in the invasive ventilation group. The authors conclude that mortality is comparable between the two groups. (**Level 2**)

A retrospective observational study already cited (55) reported the survival of non-ventilated DMD patients or those receiving invasive or non-invasive ventilation. Three groups were compared: non-ventilated patients before 1984 (group 1), those undergoing tracheostomy from 1984 to 1991 (group 2), and those on non-invasive mechanical ventilation after 1991 combined with cardioprotective medication (n = 58) (group 3). In total, 187 patients were included in this study. The mean age of death of the 56 patients in group 1 was 18.6 ± 2.9 years, that of the 18 patients having died in group 2 was 28.1 ± 8.3 years, while three subjects were still alive at the end of study, and the 88 patients on non-invasive ventilation in group 3 had a median survival of 39.6 years ($p < 0.001$). (**Level 4**)

A prospective observational study (66) described the survival of 144 children requiring long-term respiratory support, including patients with DMD (n = 18), type-1 (n = 6) or type-2 SMA (n = 14), and other congenital myopathies (n = 18). Among these patients, 28 were on invasive ventilation and 116 on non-invasive ventilation. This study demonstrated that the 5-year survival was 94% overall in the population and was significantly higher in patients on NVI (97%) than in patients with invasive ventilation (84%). The 10-year survival was 91% overall in the population. (**Level 2**)

A retrospective observational study (67) in 194 infants with type-1 SMA demonstrated that, compared to invasive ventilation, NIV with mechanical cough assistance was associated with lower survival rates at 24 months (68% versus 95%, $p < 0.001$) and 48 months of age (45% versus 89%, $p < 0.001$). In parallel, recourse to invasive ventilation decreased from 50% between 1992 and 1998 to 12.7% between 2005 and 2010 ($p < 0.005$) with a non-significant increase in recourse to NIV from 50% to 65%. (**Level 4**)

A retrospective observational study (68) analysed the role of mechanical ventilation and causes of death of DMD patients over a period of 30 years. Between 1981 and 2011, 119 adult DMD patients were cared for at the AFM Yolaine de Kepper de Saint-Georges-sur-Loire centre in France. Life expectancy without or with ventilatory support was 22.16 and 36.23 years, respectively. Similarly, the life expectancy of patients born after 1970 (mostly with ventilatory support) was 40.95 years old and 25.77 years old for those born before 1970. In parallel with the increase in life expectancy of these patients, deaths of cardiac origin increased by 8% to 44%. This study did not demonstrate any difference between the ventilation methods (invasive versus non-invasive). (**Level 4**)

A prospective observational study (69) assessed survival at 12 years, as a function of the mechanical ventilation method used (invasive versus non-invasive taking into account switches to invasive methods), in 150 DMD patients. The authors report that the risk of death was associated with the presence of swallowing disorders and cardiac failure but not with the type of mechanical ventilation (invasive versus non-invasive). (**Level 2**)

Comments

These are single-study centres, with clinical and administrative contexts and practices that differ depending on the country.

The study conducted by Ishikawa in 2011 (55) highlights a better survival of patients under NIV. However, this study has several biases. In particular, it compares patients having undergone tracheostomy before 1991 and without cardioprotective treatment (50% of patients died of cardiac causes) with patients having received NIV and cardioprotective treatment.

The study conducted by Gregoretti in 2013 (67) demonstrates lower survival in patients under NIV. However, it is important to note that this is a population of patients with type-1 SMA and therefore generally suffering from bulbar syndrome leading to swallowing difficulties, requiring frequent suctioning, and difficult to manage using non-invasive cough assistance methods.

The study by McDougall in 2013 (66) demonstrated better survival in children on NIV than in children on invasive ventilation. However, the analyses are univariate and were not adjusted on the basis of clinical status on initiation of mechanical ventilation or the disease, which are potential prognostic factors.

The studies by Soudon in 2008, Kieny in 2013 and Boussaïd in 2016 (65,68,69) demonstrate that patients undergoing daytime assisted ventilation using NIV have a long-term survival similar to that of patients ventilated by tracheostomy.

No studies, except one (Boussaïd 2016) (69), discuss the switching from NIV to invasive ventilation following an episode of acute respiratory failure leading to intubation or ventilation by tracheostomy.

5.2. Management of mucus congestion

Mucus congestion (definition and complications)

Weakness of the respiratory muscles, particularly the expiratory muscles, results in a weak and ineffective cough leading to repeated bronchopulmonary congestion and infections. Congestion is defined as obstruction of the airways by an accumulation of uncleared mucus. Bronchial congestion can result in fatigue on effort, difficulty coughing, atelectasis and oxygen desaturation. To help secretions move upwards and aid their clearance, manual bronchial drainage with increased expiratory flow and mechanical/instrumental cough assistance techniques can be combined.

Adjuvant cough assistance techniques

Several guidelines exist concerning the use of instrumental cough assistance techniques in patients with neuromuscular disorders. These guidelines are based on analyses of the published literature and are presented below.

The American Association for Respiratory Care (70) described guidelines relative to the efficacy of nonpharmacologic airway clearance techniques based on a review of the English-language published literature from 1990 to 2012, extracted from two databases (MEDLINE, CINAHL) (**Expert opinion**). Hence, cough assistance techniques (mechanical insufflation-exsufflation) are recommended in patients with a neuromuscular disease, particularly when the peak cough flow is less than 270 l/min.

Comments

Recommendations of good methodological quality (published and available systematic review, search of two databases, a single language selected, article selection criteria and levels of evidence indicated). No level 1 studies were found by Andrews J et al. The recommendations are based on low levels of evidence and on other recommendations, in particular those of Finder in 2004; themselves based on low levels of evidence.

Recommendations about new approaches and therapies available for the management of the respiratory complications of DMD patients were published **in 2004, by the American Thoracic Society (ATS) previously cited (59)**, based on a review of the "pertinent' literature by their review committee following a search of the MEDLINE database and selection of publications from 1966 to 2003. **(Expert opinion)**

The recommendations are as follows:

- DMD patients and their caregivers should be trained in airway clearance techniques, and know how to employ those techniques early and aggressively;
- assisted cough techniques should be used in patients whose clinical history suggests difficulty in airway clearance, or whose peak cough flow is less than 270 l/min and/or whose maximal expiratory pressures are less than 60 cm H_2O;

- the committee strongly supports the use of mechanical insufflation-exsufflation in patients with DMD and also recommends further studies in this area.

Comments

Recommendations of poor methodological quality (searching of only one database, no indication of the languages selected, a single study selection criterion indicated, no grading of levels of evidence and recommendations, etc.) and based on studies with a low level of evidence (observational studies).

Pressure-control ventilators and other hyperinflation techniques

As regards the use of pressure-control ventilators in patients with neuromuscular diseases, seven publications concerning clinical practice guidelines or consensus conferences were identified: an **international consensus (71)**, **a British Thoracic Society guideline specifically for children with neuromuscular weakness (8), the Canadian Thoracic Society (39), a collaboration between the British Thoracic Society and the Association of Chartered Physiotherapists in Respiratory Care (72), a French consensus conference, the guidelines of the *Association française contre les myopathies françaises* (French myopathy association) in partnership with the HAS (73) and a collaboration between the *Société française de neurologie* (French neurology society) and the *Association des neurologues libéraux de langue française* (French-language association of non-hospital neurologists) in partnership with the HAS (74).**

Comments

All the recommendations highlight the low level of evidence of the literature on this subject. Most of the literature is based on observational studies, case series, and a few randomised and non-randomised controlled studies with numerous methodological limitations.

The recommendations are therefore primarily based on expert opinions.

According to the international consensus (71), alveolar recruitment techniques or techniques such as air stacking in volume-control mode are recommended in patients with neuromuscular diseases when their vital capacity is less than 80% compared to normal. The authors indicate that if these techniques are ineffective due to severe glottal difficulties, passive insufflation techniques should be preferred. In the guideline specific to children with neuromuscular weakness, the authors indicate that manually assisted coughing and air stacking techniques to achieve maximum inspiratory volume are effective techniques to improve cough, to be used if applicable. (**Level 4**)

The BTS/ACPRC collaboration (72), concludes that it is useful to implement a maximal insufflation strategy to improve effective cough generation when vital capacity falls below 1,500 ml or 50% of the predicted value in patients with a neuromuscular disease. It encourages regular air stacking over several insufflation cycles (2 to 3 cycles) without breathing out between cycles (10-15 times in a row,

three times per day) in patients with vital capacity of less than 2,000 ml and on volume-control ventilation.

As regards the **HAS recommendations (73)** relative to the practical methods for continuous pressure NIV in patients with neuromuscular disorders, these stress that instrumental techniques of any type must imperatively be combined with NIV when patients have a peak expiratory flow (PEF) of less than 180 l/min

All the recommendations encourage further research to determine the efficacy of these strategies in patients with or without bulbar dysfunction to improve cough.

Comments

No systematic review, meta-analysis or randomised control studies were identified. The clinical studies are not very abundant and are of poor methodological quality. However the use of pressure-control ventilators is nonetheless recommended in patients with neuromuscular diseases for lung expansion, and also for cough augmentation.

Insufflator-exsufflators

All the recommendations (see table below) reach a positive conclusion with respect to the use of mechanical insufflation-exsufflation techniques in patients with neuromuscular diseases.

The recommendations were developed based on observational studies, case series, a few randomised or non-randomised controlled studies presenting numerous methodological limitations, and on expert opinions.

The various criteria cited for the implementation of mechanical insufflation-exsufflation techniques in these patients are detailed in the table below.

Table 1: Criteria for the use of mechanical insufflation-exsufflation in patients with neuromuscular diseases

There is a consensus concerning the value of using insufflation-exsufflation within all international learned societies. There are a few nuances, summarised in the table below.

Recommendations	Conditions	Criteria for the use of mechanical insufflation-exsufflation
Bach international consensus, 2013 (71) (**Level 4**)	Neuromuscular diseases	Patient with PEF < 300 l/min (invasive or non-invasive)
American Association for Respiratory Care, 2013 (75) (**Expert opinion**)	Neuromuscular diseases	Particularly in patients with PEF < 270 l/min
British Thoracic Society 2012 (8) (**Expert opinion**)	Children with neuromuscular weakness	Children with ineffective cough (including children over the age of 12 years with PEF < 270 l/min)
		Mechanical insufflation-exsufflation in very weak children, particularly with a loss of bulbar function
Canadian Thoracic Society, 2011 (39) (**Expert opinion**)	Other neuromuscular diseases	Patient with another neuromuscular disease with PEF < 270 l/min
British Thoracic Society / Association of Chartered Physiotherapists in Respiratory Care, 2009 (72) (**Expert opinion**)	Neuromuscular diseases	Treatment option to be considered in patients with bulbar dysfunction who are unable to breath stack
Swiss guidelines, Rosière, 2009 (76) (**Expert opinion**)	Neuromuscular diseases	No criteria
American College of Chest Physicians, 2006 (77) (**Expert opinion**)	Neuromuscular diseases	Patients with impaired cough
AFM/Haute Autorité de santé (French National Authority for Health), 2006 (73) (**Expert opinion**)	Neuromuscular diseases	PEF < 180 l/min, essential to combine assisted cough techniques (manual and/or instrument) with NIV
American Thoracic Society, 2004 (59) (**Expert opinion**)	Duchenne muscular dystrophy	Patients with airway clearance difficulties or PEF < 270 l/min and/or maximal expiratory pressure < 60 cm H_2O

5.3. Management of swallowing and feeding

In progressive neuromuscular diseases, the oropharyngeal muscles can weaken as the disease progresses, potentially leading to dysphagia. The term dysphagia means difficulty swallowing and covers all the symptoms presented in patients in whom the swallowing function is impaired (78). For non-tracheostomised and ventilator-dependent patients, meals are difficult because they necessarily involve repeated interruptions of ventilatory support, leading to prolongation of feeding time (nNardi), breathlessness, choking and, ultimately, undernutrition, creating a vicious circle (undernutrition/muscle weakness). To overcome this undernutrition and enable continuation of NIV without being forced to propose a tracheostomy, a nasogastric tube or, preferably, a percutaneous endoscopic gastrostomy (PEG) tube may be inserted.

Comments

No systematic review, meta-analysis or randomised control studies were identified. In the context of neuromuscular diseases (excluding SLA), there are few studies.

A cross-sectional study (79) assessed feeding difficulties in 14 children (aged 2 to 14 years) with merosin-deficient congenital muscular dystrophy. Of these 14 children, 12 were below the 3rd centile for weight. On questioning, all parents thought their child had difficulty chewing, 12 families modified the diet, and 13 children took at least 30 minutes to complete a meal. The mouth architecture was abnormal in 13 children. Nine children had an abnormal pharyngeal phase, with a delayed swallow reflex. Three of these also showed pooling of food in the larynx and three showed frank aspiration. Six children had a history of recurrent chest infections. Six of eight children who had pH monitoring also had gastro-oesophageal reflux. Five children had a gastrostomy, which stopped the chest infections and improved weight gain. (**Level 4**)

A physiological prospective study (80) studied the interactions between breathing and swallowing in neuromuscular disorders and evaluated the impact of mechanical ventilation on swallowing in patients who could breathe spontaneously, with or without tracheostomy. This study demonstrated breathing swallowing synchronisation anomalies during spontaneous breathing, facilitating the risk of aspiration. In addition, swallowing was fragmented (several swallows over several breathing cycles for each water-bolus size: 5, 10 and 15 ml). However, mechanical ventilation improved the swallowing parameters in 10 tracheostomised patients since tracheostomy enabled independent mechanical ventilation and swallowing. (**Level 4**).

A prospective study (81) assessed the effect of a speaking valve on breathing–swallowing interaction in tracheostomised and non-ventilated patients with neuromuscular diseases. Eight tracheostomised neuromuscular patients who were able to breathe spontaneously were studied with and without a speaking valve. Swallowing characteristics and breathing-swallowing synchronisation were not influenced by speaking valve use. However, expiratory flow towards the upper airway after swallowing, which protects against aspiration, was negligible without the speaking valve and was restored by adding the speaking valve. (**Level 4**)

A prospective study (82) studied the tolerance of gastrostomy feeding in 25 patients aged between 11 and 38 years with DMD, between 1997 and 2007, including 20 on NIV. The nutritional status improved with a weight-for-age (W/A) ratio increasing from 69% (45-128%) initially to 87% (49-164%) after 22 months of follow-up. Complications occurred in 21 patients (84%), but were not life-threatening. (**Level 2**)

A multicentre cohort study (25 Japanese centres) (83) evaluated the efficacy and tolerance of gastrostomy feeding in 144 muscular dystrophy patients between 2007 and 2009. Among these patients, 53 were on non-invasive ventilation and 46 on invasive ventilation. Many benefits, including an improvement in nutritional status, swallowing and respiratory status were observed after the introduction of gastrostomy feeding. In particular, in patients with Duchenne muscular dystrophy, mean body weight significantly increased. The most significant complications were respiratory failure and peritonitis. (**Level 2**)

An independent French working group (84) developed guidelines on gastrostomy in neuromuscular patients, independently of the presence of a tracheostomy, based on articles published between 2000 and 2014 (**Level 4**):

- the main gastrostomy methods used are percutaneous endoscopic gastrostomy (PEG) and percutaneous radiologic gastrostomy (PRG);
- the indications are swallowing disorders, weight loss, insufficient spontaneous food intake, feeding difficulties.

The PEG technique is preferable if the forced vital capacity (FVC) is higher than 50%. PRG should be preferred if the FVC is less than 50%, if the PEG technique fails or in the event of poor general condition of the patients.

Still independently of tracheostomy, **a non peer-reviewed literature review (78)** (based on the originator studies cited above) concerning the indications for the placement of gastrostomy in DMD patients was conducted by a Belgian team without a real peer reviewer committee (**Level 4**). The authors reach the following conclusions:

- gastrostomy should be considered if a high-calorie diet cannot be introduced or if it is unsuccessful (absence of weight stabilisation or gain) after a 6-month trial;
- the degree of respiratory impairment and cardiac dysfunction should also be considered;
- the provision of early information to patients and their families is necessary;
- the trial is considered to be unsuccessful when the weight still decreases with high calories;
- oral food and drinks are allowed with the exception of solid (including minced and mashed) food. The gastrostomy tube, supplied with its protection button, can be inserted by external surgery for which general anaesthesia is required. Fortunately, PEG placement at the bedside is possible and desirable. PEG placement requires local anaesthesia of the pharynx, a very mild sedation and NIV during the procedure(85) and the prior administration of a medicinal product that stimulates digestive motricity to increase the transit of food through the stomach. Feeding with PEG is ideally nocturnal which allows low-flow filling of the stomach (1 l/8 h). The type of formula is the same as the formula given via nasogastric tube. An adult DMD patient uses a mean of 1 to 1.5 L/24 h but this can be adjusted to the volume of food and drink still taken orally. When the decision to place the PEG tube is evident, nasogastric tube feeding, such as ultra-

thin mini-type tube, is proposed. After a few weeks, a PEG is placed, with the advantage of its discretion and hygiene. As specified above, oral feeding can be maintained, but without the social pressure or the need for performance. PEG is reported as an effective intervention to improve weight status and to decrease the rate of chest infections. It is generally well tolerated although complications such as peritonitis are reported. When aspiration is evident and PEG feeding is dictated by the critical need to protect the lungs from inappropriate intrusion of food, normal feeding is prohibited;

- drinks are given via the PEG catheter. In patients with tracheostomy presenting with severe aspiration, lungs can be protected by inflating the cuff.

6. Quality of life

As regards the quality of life of patients with neuromuscular diseases, two studies assessed patient choice in terms of NIV or ventilation by tracheostomy.

A cross-sectional study (86) concerned 168 adult patients on home ventilation (mean age 55 years), mostly poliomyelitis patients with non-progressive disease, having used ventilation by tracheostomy and NIV for at least one month each. At the time of the survey, 111 patients were using tracheostomy after having used NIV and 59 patients were using NIV after having used tracheostomy. Of the 111 patients having switched to tracheostomy ventilation after NIV, 55 patients expressed a preference for NIV, 42 for tracheostomy, and 14 had no preference. (**Level 4**)

A cross-sectional study (87) assessed quality of life using questionnaires sent by post to 91 adults (mean age 59 years) using home mechanical ventilation for a restrictive lung disease, including 16 patients with a neuromuscular disease. Sixty patients used NIV and 31 had a tracheostomy. Two out of three quality of life scores demonstrated no difference between the "NIV" and "tracheostomy" groups. For the third quality of life measurement, the score was significantly better in the tracheostomy group but the difference between the groups was small. (**Level 4)**

A cross-functional study (88) evaluated factors related to the care burden. A questionnaire was sent to 8 specialised centres. Retrospective data for 792 patients still alive and on mechanical ventilation were examined. Irrespective of mechanical ventilation method used, patients with neuromuscular diseases need more home care. Patients with neuromuscular diseases are hospitalised less often than patients with COPD. Patients with neuromuscular diseases more often had a tracheostomy. Independently, underlying disease, level of dependency, number of hours spent under mechanical ventilation, presence of tracheotomy, home distance from hospital, and hospital accesses are the main measures enabling assessment of care burden. (**Level 4**)

A recent cross-sectional study (89) assessed quality of life and life satisfaction in patients with long-term invasive ventilation following ICU treatment and unsuccessful weaning. This study included 25 patients, including 11 patients with neuromuscular diseases. A quality of life comparison was conducted on the basis of underlying disease (neuromuscular diseases versus chronic obstructive pulmonary disease [COPD]) and the individual's attitude towards tracheostomy and invasive ventilation (no regret versus regret). The data demonstrated that the patients with COPD were more often dissatisfied than the patients with neuromuscular diseases. The area of dissatisfaction most often cited was mobility. In particular, the COPD patients (85.7%) were more often dissatisfied with their mobility compared to the neuromuscular disease patients (45.5%). Finally, 18.2% of patients with neuromuscular diseases (n = 2) indicated that they would have refused to have a tracheotomy if they had to choose again. (**Level 4**)

Annexe 1. Literature search

Information sources

Bibliographic databases

Medline (National Library of Medicine, United States);

Embase (Elsevier, Netherlands);

Cochrane Library (Wiley Interscience, United States);

HAS *(Haute Autorité de santé* (French National Authority for Health), France).

Other sources

Websites of learned societies with expertise in the field studied;

Bibliography of selected articles;

Literature monitoring (AFM/HAS) conducted until completion of the project.

Literature search strategy

The search concerned the study types or subjects defined jointly with the project officer, the working group chair and the project manager.

A synopsis (see table) indicates the successive steps and highlights the results in terms of the number of references obtained by study type and subject over a given period. Only publications in French or English were used.

The search terms were either thesaurus terms (for example, MeSH descriptors for Medline) or terms from the title or abstract (free-text terms); when the search field is not specified in the table below, it is a descriptor field. These terms were combined in the number of steps required using Boolean operators.

Study type/Subject Terms used	Period	Number of ref.
TRACHEOSTOMY AND NEUROMUSCULAR DISEASES		

Guidelines and consensus conferences Step 1 (“Ventilator Weaning”[Mesh] OR “Respiration, Artificial”[Mesh] OR “Airway Management”[Mesh] OR “Airway Extubation”[Mesh] OR “Tracheostomy”[Mesh] OR “Respiratory Therapy”[Mesh] OR Lipseal[TIAB] OR mouthpiece[TIAB] OR positive pressure ventilation[TIAB] OR tracheotomy[tiab] OR Tracheostomy[tiab] OR “airway management”[tiab] OR noninvasive ventilation[tiab] OR non invasive ventilation[TIAB] OR “respiratory care”[tiab] OR Respiration Disorders[Majr:NoExp] OR Respiratory Insufficiency[Majr:NoExp] OR intratracheal Intubation[TIAB] OR Endotracheal Intubation[TIAB]) AND Step 2 “Motor Neuron Disease”[Majr:NoExp] OR “Bulbar Palsy, Progressive”[Majr] OR “Muscular Atrophy, Spinal”[Majr] OR “Fatigue Syndrome, Chronic”[Majr] OR “Muscular Diseases”[Majr] OR “Neuromuscular Junction Diseases”[Majr] OR “Peripheral Nervous System Diseases”[Majr] OR “Poliomyelitis”[Majr] OR “Stiff-Person Syndrome”[Majr] OR “Neuromuscular Diseases”[Majr:NoExp] OR Cochrane[tiab] OR dystrophy[tiab] OR dystrophic[tiab] OR dystrophies[tiab] OR neuromuscular diseasemetal[tiab] OR neuromuscular disorder*[tiab] AND Step 3 recommendation*[TI] OR guideline*[TI] OR statement*[TI] OR consensus[TI] OR position paper[TI] OR health planning guidelines[MH] OR practice guideline[PT] OR guideline[PT] OR Consensus Development Conference[PT] OR Consensus Development Conference, NIH[PT]	01/01/2007-07/08/2018 01/01/2007-07/08/2018	26 26
Meta-analyses, Systematic reviews Step 1 AND Step 2 AND Step 4 meta-analysis*[TI] OR meta-analysis*[TI] OR meta-analysis[TI] OR systematic review*[TI] OR systematic overview*[TI] OR systematic literature review*[TI] OR systematical review*[TI] OR systematical overview*[TI] OR systematical literature review*[TI] OR systematic literature search[TI] OR pooled analysis[TI] OR meta-analysis[PT] OR Cochrane database syst rev[TA]	01/01/2007-07/08/2018	138
Clinical trials Step 1 AND Step 2 AND Step 5 random*[TIAB] OR random allocation[MH] OR double-blind method[MH] OR single-blind method[MH] OR cross-over studies[MH] OR randomized controlled trial[PT] OR “Controlled Clinical Trial”[PT] OR multicenter study[PT]		

Article inclusion and exclusion criteria

In order to be selected, the studies were required to meet the following criteria:

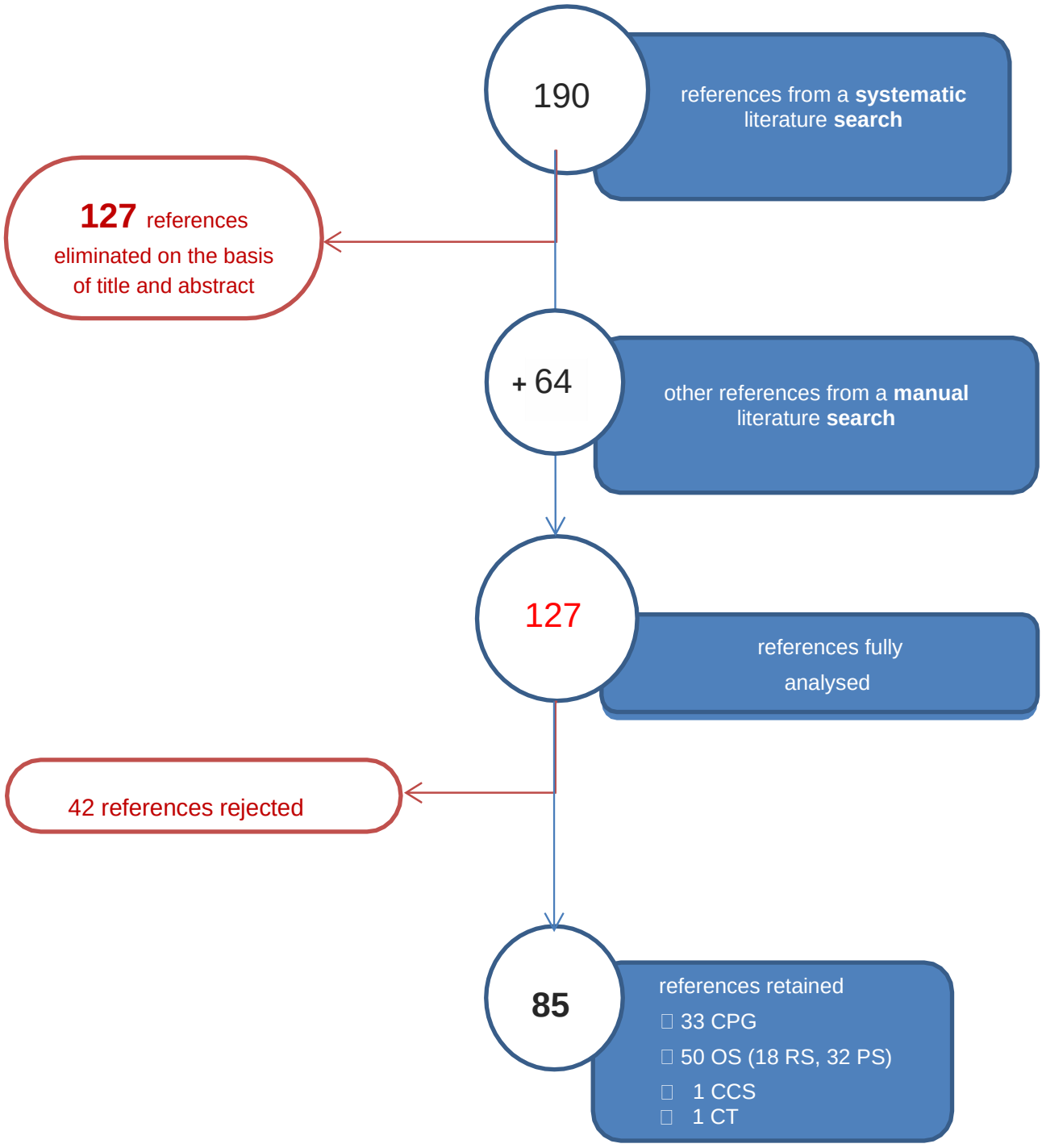
- Population: subjects with or without tracheostomy, with respiratory failure due to a slowly progressive neuromuscular disease. Slowly progressive neuromuscular diseases are understood as myopathies that lead to slowly progressive respiratory failure, primarily Duchenne muscular dystrophy.
- Study types selected: randomised controlled trials (RCT), clinical practice guidelines (CPG), health technology assessment reports (HTA), systematic reviews (SR), and meta-analyses (MA).
- Publications from 1990, only looking at the most recent publications when several exist from the same source and on the same subject.
- The search was restricted to publications in English and French.

The search excluded other rapidly progressive neurodegenerative diseases and those with a poor short-term prognosis, such as amyotrophic lateral sclerosis.

Search results

In total, 190 bibliographic references were identified from the systematic search conducted. An additional 64 references were identified following a manual search. Of these references, 127 were analysed fully and 85 were retained. They concern 33 clinical practice guidelines, 1 clinical trial, 1 case control study and 50 observational studies.

The selection process is illustrated in the figure below. Selection of publications based on their title and abstract and data extraction were performed by one person.



CCS = case control study; CT = clinical trial; OS = observational study; RS = retrospective study; PS = prospective study; CPG = clinical practice guidelines.

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Caisse nationale de l'assurance maladie (National health insurance fund for salaried workers) (CNAM)

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French college of general medicine

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French national council for thoracic and cardiovascular professionals

French national council for physical medicine and rehabilitation professionals

French national council for neurology professionals

French national council for paediatric professionals

French national council for pulmonology professionals

French national council for radiology and medical imaging professionals

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Société de réanimation de langue française (French-language Society for Intensive Care)

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Abbreviations and acronyms

AFM-Téléthon	*Association française contre les myopathies* (French myopathy association, AFM) - Téléthon
BIPAP	Bi-level positive airway pressure
BMI	Body mass index
COPD	Chronic obstructive pulmonary disease
TLC	Total lung capacity
FRC	Functional residual capacity
VC	Vital capacity
FVC	Forced vital capacity
PEF	Peak expiratory flow
DM1	Myotonic dystrophy type 1
DMD	Duchenne muscular dystrophy
EPAP	Expiratory positive airway pressure
FDA	Food and Drug Administration
FiO_2	Fraction of inspired oxygen
RR	Respiratory rate
PEG	Percutaneous endoscopic gastrostomy
PRG	Percutaneous radiologic gastrostomy
GRADE	The grading of recommendations assessment, development and evaluation
HAS	*Haute Autorité de santé* (French National Authority for Health)
IPAP	Inspiratory positive airway Pressure
LGMD	Limb girdle muscular dystrophy
LPPR	*Liste des produits et prestations remboursables* (List of products and services qualifying for reimbursement)
NMD	Neuromuscular disease
EP max	Maximum expiratory pressure
PEEP/PEP	Positive End Expiratory Pressure
$PETCO_2$	End tidal CO_2
IP max	Maximum inspiratory pressure
PSV	Pressure support ventilation
$PTcCO_2$	Transcutaneous carbon dioxide pressure
BPG	Best practice guideline
SaO_2	Oxygen saturation
ALS	Amyotrophic lateral sclerosis
SMA	Spinal muscular atrophy

SNIP	Sniff nasal inspiratory pressure
PNS	Peripheral nervous system
CNS	Central nervous system
ST	Surgical tracheostomy
PT	Percutaneous tracheostomy
PIT	Prolonged inspiratory time
ACV	Assist-control ventilation
HV	Home ventilation
FEV1	Forced expiratory volume per second
IV	Invasive ventilation
MV	Mechanical ventilation
NIV	Non-invasive ventilation
RV	Residual volume
REV	Residual expiratory volume

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