HAUTE AUTORITÉ DE SANTÉ

RECOMMENDING
BEST PRACTICES

GUIDELINE

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) Secretary: Ms Laëtitia Gourbail
Literature search	From January 2007 to August 2018 (see literature search strategy described in annex 1 of the evidence report)

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Contents

Key messages

The following recommendations were considered as being those that should be implemented as a priority to improve the quality of care delivered to patients at the time of publication of the best practice guideline.

- It is recommended that tracheostomy only be envisaged following the failure of non-invasive ventilation (NIV) and cough assistance techniques. It requires the written informed consent of the patient, their person of trust or their legal guardian, following a multidisciplinary team meeting to consider the issue.

- Except in emergency situations, it is recommended that tracheostomy be exclusively surgical and performed by a practitioner with expertise in the procedure, either a surgeon or a resuscitation specialist.

- The post-surgical follow-up of tracheostomy should be carried out in an intensive care unit until the first tube change.

- Any early post-surgical bleeding requires immediate assessment by a competent professional and as soon as possible by the practitioner having performed the procedure.

- After the first tracheotomy tube change, transfer of the patient to a high-dependency unit or equivalent structure may be envisaged and rehabilitative management for swallowing and speech is necessary.

- In situations of bronchial congestion, bronchial drainage techniques are recommended in addition to tracheal suctioning. These are systematically tested and optimised in hospital first.

- Patient education and caregiver training in everyday procedures and measures to be taken in the event of certain emergency situations must be delivered before the patient is discharged from hospital.

- Tracheotomy tubes should be changed at least every 28 days.

Introduction

Referral

The *Association française contre les myopathies*-Téléthon (AFM (French myopathy association)-Téléthon), a nationally accredited patient and healthcare user association, asked the HAS to conduct a study relative to the role of tracheostomy in the management of ventilator dependence in neuromuscular diseases.

The *Société de pneumologie de langue française* (SPLF - French-language Pulmonology Society), the *Société de réanimation de langue française* (SRLF - French-language intensive care society), the *Filière nationale de santé des maladies neuromusculaires* (FILNEMUS - French national health network for neuromuscular diseases) (which groups together all the players in the care network dedicated to neuromuscular diseases) and the *Association nationale pour les traitements à domicile, les innovations et la recherche* (ANTADIR - French national association for home care, innovations and research) (which represents the majority of not-for-profit home care support services) support the request for inclusion in the HAS work programme.

The request was included in the HAS work programme.

The objective is to harmonise practices and improve the care of ventilated patients with neuromuscular diseases.

Context for development of the guidelines

The term “neuromuscular diseases” designates diseases that affect the muscles or their innervation system (motor unit disorder), which can appear at any age of life, be temporary or permanent, and progress at variable rates.

There are more than 200 different neuromuscular diseases (including muscular dystrophies, peripheral neuropathies, dysimmune neuropathies, spinal amyotrophies, myasthenia, myositis, amyotrophic lateral sclerosis), with various causes and consequences. Although most of these diseases are genetic, they can also be of immune, inflammatory, endocrine, medicinal or infectious origin, etc. All of these conditions are uncommon and are therefore considered to be “rare diseases”. However, all neuromuscular diseases combined, tens of thousands of individuals are affected in France.

In some diseases, the respiratory muscles are affected. Respiratory capacity and cough are reduced, leading to bronchial congestion and a risk of lung infection. Swallowing difficulties related to upper airway muscle dysfunction represent an additional risk factor for respiratory obstruction and lung infection.

Evolving knowledge about neuromuscular diseases and the management of patients with these diseases has led to real improvements in terms of both life expectancy and quality of life for these patients.

Respiratory physiotherapy aims to prevent congestion and help maintain muscle capacities. In the event of congestion, specific techniques are put in place, mechanically supported if necessary. In the event of respiratory failure, ventilatory support may be necessary.

Initially, all positive-pressure mechanical ventilation was proposed via tracheostomy (invasive ventilation). In the 1980s, night-time ventilation using a nasal mask (non-invasive ventilation) was proposed, reserving tracheostomy for patients unable to breathe alone and hence also ventilated during the daytime. But in the last few years, an alternative technique has been being used by some national and international teams, who have opted to offer patients with neuromuscular diseases

continuous non-invasive ventilation, supplementing night-time non-invasive ventilation with daytime mechanical ventilation using a mouthpiece, or nasal mask when the patient is unable to hold the mouthpiece, thereby enabling 24-hour ventilatory support. The use of this mouthpiece provides patients with greater freedom to read, speak, eat or drink than a mask, and prevents the development of skin injuries due to pressure points from the mask, by limiting its use to night-time only. Similarly, nasal cannulas generally provide better vision than a mask and prevent skin injury. It is possible to alternate between these two interfaces.

Ventilator dependence and upper airway muscle dysfunction can result in swallowing and feeding difficulties, and thereby exacerbate the respiratory failure. Similarly, an ineffective cough promotes bronchial congestion, which, in turn, can cause episodes of acute respiratory decompensation. Gastrostomy and cough assistance techniques can be effective solutions for these difficulties. These techniques further reduce recourse to tracheostomy, which could be reserved for patients in whom non-invasive ventilation has failed and for those patients and families who find the constraints of non-invasive ventilation too great, bearing in mind that tracheostomy also leads to other constraints.

It is essential to take into consideration patient preferences and quality of life.

In order to better understand the needs of patients, the AFM-Téléthon conducted a survey in 2014 in 445 patients with Duchenne muscular dystrophy, 30% of whom used non-invasive ventilatory support and 17% invasive ventilation (tracheostomy). Tracheostomy had been an elective procedure in over 50% of cases, 79% of patients noted a change in social life, with 56% of these considering this change to be positive and 39% negative.

Epidemiology

Since epidemiological data are almost non-existent, it is not possible to accurately assess the number of people with neuromuscular diseases, or the proportion of these patients requiring non-invasive or invasive ventilatory support.

In France, the number of patients with neuromuscular diseases eligible for full cover of healthcare costs for a long-term illness (ALD in French - chronic condition) is around 40,000.

Challenges

Although this subject concerns a relatively small population, these are patients who are severely affected by their disease and whose management is extensive and consumes significant amounts of healthcare. 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' prognosis in terms of survival.

Objectives

The aim is to address the challenges outlined above. To this end, the recommendations aimed at the professionals concerned are designed to support decision-making in terms the choice of care and to improve and harmonise practices. The ultimate objective is to improve the management of patients, and therefore the quality and safety of their care, as well as their quality of life.

Scope

- Patients or users concerned by the subject

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.

- Professionals concerned by the subject

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.

Definitions

Non-invasive ventilation

Non-invasive ventilation (NIV) refers to all ventilatory support techniques that take over all or part of the work of breathing in the absence of an endotracheal device in order to ensure satisfactory alveolar ventilation.

NIV requires a home ventilator or one or more appropriate ventilatory support methods, along with interfaces fitted to the patient (morphology, leak control, etc.) and adapted to night-time and day-time conditions, enabling the following:

- control of daytime and night-time hypoventilation assessed by:
  - the absence of symptoms,
  - normalisation of morning and evening carbon dioxide levels, measured by blood gases,
  - normalisation of pulse oxygen saturation (SpO_2) and nocturnal transcutaneous CO_2 pressure ($PTcCO_2$);
- good tolerance, reflected by:
  - absence of skin injury;
  - absence of gastric dilatation or eye irritation related to leaks;
  - acceptance by the patient;
- minimisation of complications, such as facial deformity related to the interface;
- good compliance with treatment assessed on the basis of ventilator data;
- resumption of growth in children and improved nutritional status in adults.

Invasive ventilation

Invasive ventilation uses an endotracheal interface, which may be an intubation catheter or a tracheotomy tube. Only invasive ventilation by tracheotomy tube can be used at home.

Tracheotomy or tracheostomy

Tracheotomy or tracheostomy, is a procedure which consists of making an incision on the front of the neck to obtain a direct opening in the trachea. The resulting stoma can serve as a site for a tracheal tube to be inserted in order to allow a person to breathe without the use of the nose or mouth.

Surgical (or open) tracheostomy involves the dissection of the pretracheal tissue and the insertion of a tracheotomy tube (or cannula), under direct visualisation of the trachea. It is preferably performed in an operating theatre or at the patient's bedside in an intensive care unit (ICU).

Percutaneous tracheostomy is non-surgical and involves the gradual introduction of a guidewire and dilators using the Seldinger technique.

1. Tracheostomy indications

1.1. Indications in the absence of acute respiratory failure

Tracheostomy can only be envisaged following the informed consent of the patient, their person of trust or their legal guardian, after providing information on the advantages and disadvantages of invasive and non-invasive ventilatory support techniques.

Tracheostomy is never proposed as a first-line approach unless expressly requested by the patient (expert consensus).

Hence, it is recommended that tracheostomy be considered following the failure of non-invasive ventilation (NIV) and cough assistance techniques.

These failures may be related to the following situations (expert consensus).

- Impossible to put in place NIV (not feasible in practice):
  - patient refusal to comply with non-invasive ventilation;
  - intolerance to NIV (such as uncontrollable leaks around the interface or claustrophobia);
  - behavioural and cognitive disorders;
  - failure of invasive ventilation weaning carried out in a neuromuscular reference or expert centre.

- Inefficacy of NIV after its introduction:
  - persistent (two consecutive assessments in stable condition) and symptomatic daytime hypercapnia ($PaCO_2$ > 44 mmHg) despite optimised and well-managed NIV (as defined above and for a duration of 20 hours or more per day);
  - concomitant disorders (persistent nutritional problems [weight loss or weight stagnation in children] despite gastrostomy, gastro-oesophageal reflux with inhalation, severe bulbar dysfunction);
  - anatomical and functional causes (impossible to find an interface that fits and/or facial morphological abnormality);
  - repeated acute respiratory decompensation, despite the combined use of cough assistance techniques with NIV, requiring unscheduled hospitalisations,
  - ineffective mucus clearance.

Methods - Elective planning

Except in emergency situations, it is recommended that the indications for tracheostomy be discussed and validated at a multidisciplinary team (MDT) meeting (expert consensus) (see composition of multidisciplinary team meetings in annex 3).

Specific cases in which the indication may be discussed

Indication for temporary perioperative tracheotomy (particularly when the surgical procedure causes a reduction in respiratory capacity in the immediate post-operative period, the recovery of which is

gradual over a prolonged period of time, such as for spinal arthrodesis, for example) (expert consensus).

Early and immediately severe infantile neuromyopathies

Progressive profile

Early and immediately severe infantile neuromyopathies can be divided into three groups based on their progressive profile:

- progressive diseases (example: type-I spinal amyotrophy);
- stable diseases or with progressive improvement (examples: congenital muscular dystrophies, structural myopathies);
- diseases with rapid possibility of ventilator-free breathing (example: congenital myasthenic syndrome).

The choice of respiratory management (NIV or tracheostomy) depends on the progressive profile of the disease and the respiratory (grade B) and/or bulbar impairment. This choice is made during a review by a multidisciplinary team attached to a neuromuscular reference or expert centre (expert consensus).

In infants (aged from more than 28 days to under 3 years), tracheostomy should be considered when NIV needs exceed 16 hours per 24 hours (for progressive and/or stable diseases and in the absence of any additional acute episode increasing NIV needs (expert consensus).

NIV limitations

It is recommended that the indication for tracheostomy be discussed on a case-by-case basis for the following obvious reasons (expert consensus).

In infants, NIV is currently limited by the choice of interface. Mouthpiece ventilation is not possible in infants and young children up to around the age of 8 years. Mucus clearance techniques are also more limited in infants due to their lack of ability to actively cooperate.

The prolonged use of NIV (daily wear time and total NIV duration, in months or years), impedes physiological facial growth. NIV can therefore be associated with detrimental maxillo-facial deformity. Long-term NIV can cause hypoplasia of the midface, with retromaxilla, which can itself cause:

- ➔ upper airway obstructive syndrome;
- ➔ a dental articulation disorder compromising chewing, swallowing and speech;
- ➔ aesthetic damage.

The current nose masks (less deforming to the face) may be discussed on a case-by-case basis from the age of 6 years.

In infants and young children, the benefits of NIV are counterbalanced by the negative impact on their cognitive and psychomotor development, as well as their socialisation when the duration of NIV exceeds the child's physiological sleeping time (night-time and nap time) (expert consensus).

1.2. Non-elective indications (acute respiratory failure)

It is recommended that at-risk patients be identified as *"patients remarquables"* ("special patients") with an up-to-date emergency liaison file (ELF) including their advance directives (expert consensus) (see annex 4).

A tracheostomy is performed non-electively - i.e. in an emergency - when there is respiratory distress that is life-threatening and tracheal intubation is impossible or contraindicated (e.g. anatomical or morphological obstacles, arthrodesis [neck stiffness]). The physician should inquire about the existence of advance directives relating to the potential refusal of tracheostomy, knowing that he or she may refuse to apply them in the event of a life-threatening emergency, for the time needed to assess the situation, or when they appear inappropriate or inconsistent with the medical situation (article L1111-11 of the French public health code as amended by Law No. 2016-87 of 2 February 2016 - art. 8).

2. Contraindications for tracheostomy

It is recommended that contraindications be discussed at a multidisciplinary team meeting (expert consensus).

2.1. Absolute contraindications

- refusal of an adult and capable patient, expressed or otherwise by advance directives;
- refusal of legal guardian;
- anatomical obstacles without therapeutic alternatives (example: tumours, vascular problem) (expert consensus).

2.2. Relative contraindications

It is also recommended that the following relative contraindications be taken into consideration (expert consensus):

- absence of care facility options or life project;
- rapidly progressing infantile neuromuscular diseases with no prospect of improvement, such as type-1 SMA refractory to new biologic therapies;
- comorbidities that are life-threatening in the short term (example: severe heart disease).

3. Performance of tracheostomy

3.1. Elective tracheostomy

It is recommended that tracheostomy be electively planned and performed in coordination with the neuromuscular reference or expert centre (expert consensus).

It is recommended that tracheostomy in patients with neuromuscular diseases be exclusively surgical and performed by a practitioner with expertise in the procedure, who is either a surgeon or a resuscitation specialist (expert consensus).

If tracheostomy is performed by a resuscitation specialist, a surgeon must be available and present on-site, in coordination with a neuromuscular reference or expert centre (expert consensus).

The patient's informed consent must be obtained before the multidisciplinary team meeting and after a reflection period compatible with their treatment.

An anaesthesia consultation must be completed before the tracheostomy.

A cerivothoracic angio-CT scan enabling diagnosis of tracheobronchomalacia, as well as personalised modelling of the trachea and its relationship with the thyroid gland and vessels, are recommended in adults (expert consensus). This examination will also be very useful for the choice of tracheotomy tube.

A multidisciplinary team meeting will then define (expert consensus):

- whether tracheostomy should be preceded by other additional investigations (cervical ultrasound, flexible tracheal fibroscopy/endoscopy depending on the diseases);
- if tracheostomy can be performed by a resuscitation specialist at the patient's bedside, or if it must be performed by a surgeon in the operating theatre;
- if tracheostomy must be performed under general anaesthesia and following intubation or whether it can be performed under NIV and local anaesthesia.

3.2. Non-elective tracheostomy

In extreme emergencies, if the patient cannot be intubated, tracheostomy may be performed percutaneously or surgically, depending on the expertise of the practitioner and the local conditions (expert consensus).

3.3. Surgical tracheostomy technique

The choice of tracheotomy tube depends on the results of the physical examination and, if applicable, the imaging exam (expert consensus).

In adults, it is recommended that the first tube preferably be a tube with a low-pressure cuff (expert consensus).

This is a routine procedure, for which the technique is perfectly described in the various textbooks, and which can be performed in the operating theatre or at the patient's bedside in the intensive care unit: patient installed in the dorsal decubitus position, neck in extension (block under the shoulders).

The approach consists of making a transverse incision, the size of which must be tailored to the age and size of the patient (3 to 4 cm in adults, 1 cm below the cricoid cartilage or 2 cm above the sternal

notch), then proceeding to open the linea alba vertically between the strap muscles, while the thyroid isthmus can be retracted upwards, if necessary, or sectioned.

For the tracheal opening, the incision can be made horizontally between two rings (usually between the second and third ring) or vertically in adults. A monofilament type suture is immediately placed on each tracheal margin to facilitate the introduction of the cannula during the creation of the tracheotomy, and then subsequently during recannulation. These sutures are knotted together and taped to the torso using adhesive tape, identifying the side corresponding to each suture. They are removed when the tube is changed for the first time, usually after a week (expert consensus).

It is recommended that the intubation tube be raised just above the level of the tracheal incision without extubating, and then a tracheotomy tube of a size appropriate to the internal diameter of the trachea be inserted (equivalent to the intubation tube) (expert consensus).

It is recommended that the distal end of the tube ideally stop two to three rings above the carina to limit carinal irritation (expert consensus).

It is recommended that the tube be fastened in place (preferably using a tie), ensuring that the knot is tied while the patient is seated, with the head held, in order to assess the neck circumference as accurately as possible (expert consensus). To ensure that the tie is not too tight, a finger should be able to pass under it. The tube should not be sutured to the skin to avoid the risk of accidental decannulation being overlooked.

It is recommended to check the position of the tracheotomy tube by X-ray and/or fibroscopy (expert consensus).

It is recommended that the patient remain in intensive care until the first tube change (expert consensus).

If there is a high risk of decannulation or difficulty in recannulating, the cartilage edges may be sutured to the skin and a stoma may be created.

Paediatric specificities must be highlighted. A vertical midline incision of the second, third and fourth tracheal rings is recommended, rather than a horizontal incision between two rings. However, using this technique, there is a risk of laryngo-tracheal disinsertion in a very young child of low weight. The anterior part of the tracheal arch is not resected in children (expert consensus).

Neonatal-size tubes are usually shorter. The risk of decannulation is higher. It is therefore recommended to prefer paediatric tubes when the neck length allows this (expert consensus).

4. Post-tracheostomy follow-up and prevention of complications

4.1. Initial follow-up in the ICU

As regards initial follow-up in the intensive care unit, the following recommendations are made (expert consensus):

- monitor the tube cuff pressure at least every 8 hours and maintain it at the minimum pressure preventing leaks, without ever exceeding 25 mmHg;
- systematically have the practitioner perform a local examination 48 hours after the procedure and at any time in the event of bleeding or any another anomaly;
- have the first tube change performed in the intensive care unit by a physician qualified in the procedure (on D10 at the latest) (the second tube can be a tube with or without cuff depending on the tolerance to leaking ventilation [deflated cuff] with the first tube);
- remove the retrieval sutures after the first tube change;
- take particular care not to displace the tracheotomy tube when installing or moving the patient and during mucus clearance techniques;
- for any mobilisation, have one person designated to hold the tube in position;
- move the patient to a chair whenever the local examination by the practitioner so allows;
- if the patient is ventilated, centre the tubes properly to prevent any pulling on the tracheotomy tube;
- systematically carry out specific monitoring of swallowing, particularly of liquids, to verify the absence of signs of aspiration;
- in the absence of swallowing difficulties, resume oral feeding after the local examination conducted after 48 hours;
- monitor the presence of congestion, and conduct bronchial clearance if necessary, using manual or instrumental techniques, whenever the local examination by the practitioner so allows.

Specific cases of children and agitated patients

The major risk is decannulation, and the asphyxiating consequences of this.

It is therefore recommended to systematically put in place effective physical restraint including wrist straps (in infants, made-to-measure restraining sleeves can replace wrist straps (expert consensus).

If the patient is ventilated it is recommended that the tubes be placed well away from the hands and feet, to prevent any pulling on the tracheotomy tube (expert consensus).

In children, until the first tracheotomy tube change, the following should be preferred:

- ➔ restraint measures (harness and sleeves);
- ➔ washing in bed (no baths).

Installation of the infant in a baby seat, even fastened in, should be avoided.

4.1.1. Identification and management of early complications

Early complications generally occur before the first tracheotomy tube change.

Bleeding complications

- due to bleeding at the thyroid isthmus, particularly in the event of a transisthmic approach, or of the cervical vessels lateral to the tracheotomy;
- due to erosion of the trachea, then of the brachiocephalic artery, by the tip of the tube or at the level of the cuff, or direct erosion by the lower surface of the tube before it enters the trachea.

Any early surgical bleeding requires immediate assessment by a competent professional and as soon as possible by the practitioner having performed the procedure (expert consensus).

Mild bleeding of the tracheotomy opening can be controlled by straightforward compression (expert consensus).

Heavy bleeding requires urgent surgery (injury to the brachiocephalic artery (BCA), inferior thyroid artery, innominate vein [IV], etc.) (expert consensus).

Infectious complications of the surgical wound

This is a genuine local infection requiring local or systemic treatment in accordance with the applicable guidelines. No prophylactic antibiotic therapy is recommended (expert consensus).

Tube obstruction due to bleeding or thick secretions

Preventive measures include routine humidification of the inspired air and, when necessary, suctioning limited to the length of the tube (expert consensus).

In the event of obstruction, the tracheotomy tube should be changed immediately.

Decannulation and early displacement

The tube must be carefully repositioned without forcing to prevent any risk of incorrect routing. In the event of failure, temporary emergency oral intubation should be performed (expert consensus).

In the event of obliteration of the tip of the tracheostomy tube due to its displacement into the surrounding tissues (incorrect routing) or due to the distal tip abutting against the tracheal wall, confirmed by fibroscopy, which also looks for the presence of granuloma, repositioning or a change of tube is usually necessary (expert consensus).

Subcutaneous or extensive erythema

- due to an excessively tight skin suture;
- due to positive-pressure ventilation or coughing with air leaks not being released due to a very tight cervical suture;
- due to incorrect routing of the tracheotomy tube.

Prevention consists of ensuring the skin suture around the tube is not too tight (expert consensus).

Thoracic imaging enables pneumothorax and pneumomediastinum to be eliminated (expert consensus).

4.1.2. Identification and management of late complications

- swallowing and bronchial congestion problems (see below);
- tracheal injury: the choice of surgical technique and tracheotomy tube (see above) can prevent some of these complications (grade B);
- tracheal necrosis: tracheal intubation followed by tissue debridement (grade B). Management depends on the location of the area of necrosis and its extent, and where possible the tube cuff should be placed below the area of necrosis.

It is recommended to prefer the use of an uncuffed tube or to continue monitoring of the cuff pressure (expert consensus).

- Tracheomalacia: adaptation of the tube length (grade B).
- Tracheal stenosis due to post-devascularisation ischaemia and mechanical erosion: caused by cuff over-inflation, angulation of the tube. It is recommended to check the cuff pressure to limit cartilage ischaemia and to discuss the indication for an endoscopic dilatation procedure (grade B).
- Tracheo/BCA fistula (caused by direct contact between the tube and the brachiocephalic artery due to a tracheotomy that is too low, an over-inflated cuff, excess movement of the head or tube) suspected following haemoptysis, irrespective of abundance, which requires urgent surgical treatment (grade B).
- Tracheo-oesophageal fistula, usually related to erosion of the tracheal membrane by the tube cuff, but also to incorrect tube angulation. This complication is more frequently observed in the presence of a gastric feeding tube. It usually requires deflation of the cuff or the use of an uncuffed tube (grade B) and removal of the oesophageal tube. Surgical treatment must systematically be discussed according to the benefit-risk balance as soon as the diagnosis is made, and also discussed again based on the evolution in the event of initial surgical abstention (from interposition flap alone to anastomosis resection with interposed flap, etc.).
- Formation of granulomas: suctioning into the tracheotomy tube must not go beyond the length of the tube to limit irritation of the tracheal mucosa and granuloma formation. In order to limit this risk, it is recommended to adjust the length of the suction tube on a tracheotomy tube model (grade B).

4.2. In a high-dependency unit or equivalent structure

Monitoring of vital signs may remain necessary, then can be gradually reduced and stopped depending on the patient's evolution (expert consensus).

The emergency tube change kit must include a spare tube of identical diameter and a tube with a smaller diameter, an intubation tube, Laborde forceps (3 branches), a laryngoscope and a bag valve mask (BVM) (expert consensus).

In some cases, a made-to-measure tube can be envisaged (expert consensus).

If not already done in the ICU, a swallowing assessment should ideally be conducted by the ENT specialist or speech therapist, if possible in a sitting position, head upright, cuff inflated, then deflated, starting with swallowing saliva, followed by thickened water or yoghurt, then gaseous fluid, then water or other coloured fluid to better detect any aspiration and stasis.

If this assessment is conclusive, feeding tests can be proposed in order to adapt meal textures if required. In the event of difficulties, it is recommended that a speech therapy assessment and ENT opinion be requested (expert consensus).

In parallel with speech therapy information and rehabilitation, it is recommended to explore speech improvement techniques using medical devices: speaking valve during “leaky” ventilation (example: deflated cuff), and respirator adjustment (lengthening of inspiratory time [Ti], initiation of positive expiratory pressure [PEP]) (expert consensus).

After these device changes and ventilatory adjustments, good control of daytime and night-time gas exchange should be checked (expert consensus).

Several ventilation programmes with or without leaks, depending on day/night cycle or in the event of respiratory exacerbation may be defined to ensure correction of daytime and night-time hypoventilation (expert consensus).

4.2.1. Rehabilitation of speaking

Rehabilitation via speech therapy is essential.

During mechanical ventilation, speaking is possible during inspiration in the event of “leaky” ventilation, since a proportion of the insufflated volume is directed towards the upper airways.

During expiration, speaking is only possible if there is positive pressure in the trachea.

Patients need to understand the anatomical and physiological changes in their speech. Speech therapy will help patients:

- understand and use techniques enabling the restoration of positive subglottic pressure required for speech (expert consensus);
- adapt the speed of their speech and coordinate it with the times when subglottic pressure is positive, which depends on ventilation conditions. Therefore, depending on whether the patient is ventilated or not, on the setting of the ventilator and/or on the presence or absence of a speaking valve, the subglottic pressure may be positive on inspiration only, on expiration only or both (expert consensus);
- coordinate the swallowing of saliva with speaking (expert consensus).

The team should be provided with information explaining the different types of rehabilitation possible for speaking/communicating with the tracheotomy tube. For example, it is important to explain the benefit of deflating the cuff and using a speaking valve to enable and optimise communication with family members, or to explain to the patient how to synchronise speech with the intermittent positive subglottic pressure generated by the ventilator.

In liaison with the speech therapist or occupational therapist, the implementation of augmentative and alternative communication (AAC) methods is recommended when the patient cannot communicate verbally or to supplement verbal communication (examples: alphabet charts, symbols or pictures, laser glasses, eye-tracking, etc.) (expert consensus).

It is recommended that all possible efforts be made to restore the patient’s spontaneous speech (expert consensus). This can be achieved by adaptations to ventilation based on the introduction of ventilation with intentional leaks in ventilation-dependent patients, i.e. ventilation with a deflated or uncuffed tracheostomy tube. The leaks thus obtained during the inspiratory phase of the respiratory cycle allow the patient to speak when breathing in. However, without additional adaptations, speech may remain of poor quality, since the patient depends on the ventilator pressurisation during inspiration to speak.

During expiration, the decrease in subglottic pressures, due to the opening of the expiratory circuit of the ventilator connected to the tracheostomy tube, does not allow effective speech. Patients can improve their speech by increasing their respiratory rate, thus shortening the duration of the expiratory time (which is not conducive to speaking), which requires paying careful attention to the trigger sensitivity of the ventilator so that patients can trigger it without any risks of unwanted self-triggering. To further improve speaking, two techniques that restore the possibility of speech during expiration and a degree of independence from the ventilator can be considered. These two techniques enable restoration of subglottic positive pressure and an expiratory flow towards the upper airways.

- The first technique is the use of positive expiratory pressure (PEP). Maintaining this pressure during expiration can make it possible to maintain sufficient pressure to allow speech. The effective level of PEP varies depending on the patient. The usual initial level is 5 cmH20, without exceeding 10 cmH20, but it may be necessary to increase the PEP to obtain good-quality speech. If the PEP is increased, the ventilator trigger sensitivity should be reassessed to make sure it is not triggered automatically. In ventilator-dependent patients, a high PEP level is not necessary at night. Hence two different ventilation programmes - one for daytime and one for night-time - can be envisaged.
- The second technique involves using a one-way speaking valve placed on the ventilator circuit. However, this technique demands complete expiration via the upper airways, which is not possible in all patients, leading to a potential risk of active expiration, hyperinsufflation and barotrauma. It is essential to check the patient's tolerance to this system in a medical setting before prescribing it at home, in order to ensure the patency of the upper airways.

If the patient is not totally ventilator-dependent and can therefore breathe alone, the use of tracheotomy occlusion can be envisaged in the absence of an inflated cuff, preferably using a one-way speaking valve, rather than a cap placed over the tracheotomy opening. The two techniques restore expiratory subglottic positive pressure and hence redirect the expiratory flow toward the upper airways, restoring the possibility of speech. If it is well tolerated, a cap enables humidification of the air inspired by the upper airways. However, the cap requires a full respiratory cycle via the natural airways with increased resistance due to the presence of the tube in the trachea, whereas the speaking valve enables the advantages of the tracheotomy to be maintained during inspiration (reduced dead space and work of breathing), but at the risk of potential drying out of secretions and the upper airways.

In patients who cannot be ventilated with a deflated cuff, tracheotomy tubes equipped with a channel enabling the potential creation of a subglottic air flow to allow speaking are available. However, these were originally designed for subglottic suctioning making their efficacy uncertain, and the need for an alternative air source to supply the channel make their home use complicated (expert consensus).

All these techniques can modify the ventilation conditions (increase in dead space, increased resistance, drying out of mucosa, obstruction of tracheotomy tube). In addition, they are contraindicated in the event of bronchopulmonary congestion and increased pneumothorax risk.

It is therefore recommended that these various techniques and their tolerance be tested under medical supervision and monitored before being prescribed at home (expert consensus).

4.2.2. Dysphagia

The main problems following intubation and tracheostomy are ear-nose-throat related ones: pharynx and larynx sensitivity problems, immobilisation of the larynx by the tracheotomy tube, paresis of the soft palate and vocal cords, laryngeal oedema/ankylosis/ulceration, loss of swallowing reflex.

After the ENT specialist has excluded a detrimental effect of the tracheotomy tube on swallowing, it is recommended that a swallowing assessment be performed by a speech therapist to evaluate the mobility and sensitivity of the ENT structures (assessment of praxia, voice, mouth, pharyngeal and soft palate sensitivity) (expert consensus).

Swallowing tests are proposed following a medical opinion. On the advice of the team, the tests can be performed:

- preferably while on mechanical ventilation, which is the most favourable condition (grade B);
- with the cuff deflated (in this case, it is important to check for the existence of aspiration by suctioning the tracheotomy tube immediately after swallowing);
- with cuff inflated (lower immediate aspiration risk).

Where possible, it is recommended that tests be performed with the cuff deflated to avoid pressure stresses on the upper oesophagus and to allow the larynx to rise during swallowing.

When testing on a deflated cuff or in the absence of a cuff, the use of coloured liquids or semi-liquids makes checks for aspiration more reliable during suctioning under the same conditions as during the initial ENT examination.

The amounts of liquid are gradually increased, testing the content of a teaspoon (5 ml) then a tablespoon (15 ml), and finally, sips from a glass.

Safety postures, such as chin tipped slightly forward, help to slow down the food bolus during propulsion in the pharyngolarynx and contribute to better closure of the airways.

Instructions are then issued following the tests and disseminated to the team for patient re-feeding and re-hydration. These are validated by the physician.

The speech therapist also plays a role in informing the medical and paramedical team about the consequences of tracheostomy on swallowing. It is essential that this information be provided in advance of the instructions that will be left after the tests.

Self-assessment of the dysphagia-related handicap can be offered to the patient in order to specify the impact of the dysphagia. Tools exist for this purpose (in particular: the deglutition handicap index, and the Sydney Swallow Questionnaire, for which a French-language version has also been validated).

In the event of suspected inhalation, all oral feeding and hydration must be suspended and an ENT exam must be requested.

4.2.3. Rehabilitation of dysphagia

Following the speech therapy assessment and the ENT assessment, the speech therapist - or the physiotherapist - intervenes to rehabilitate the disrupted ENT functions.

Oral mobility exercises (tongue, lips, cheeks, soft palate) are proposed in order to reprogramme the oral phase of swallowing. Sensitivity may be disrupted by the re-routing of air. Taste and smell are diminished and should be stimulated by restoring the respiratory flow in the upper airway by deflating the cuff or using an uncuffed tube.

The laryngeal region is hypomobile due to the insertion of the tube. The risk of choking and aspiration is increased. The lack of elevation of the larynx should be compensated for by safe postures (sitting if possible, head tilted forward), and thickening of textures so that the food bolus arrives less quickly in the pharyngolarynx and the protective structures of the airways close. Communication between the

speech therapist, dietician and caregiving team is important to ensure that the volume, texture and posture constraints identified during the assessment are implemented.

These same instructions should be passed on when the patient is discharged (speech therapy assessment and report) in order to ensure continuity in the recovery and monitoring of swallowing.

The presence of PEP during feeding and deflated cuff/uncuffed ventilation enables positive subglottic pressure to be maintained at the time of swallowing (protective effect), regardless of the time in the respiratory cycle when swallowing occurs.

When the patient is able/willing to be disconnected from the ventilator at mealtimes, the use of a speaking valve should be considered (if the patient's tracheotomy tube is uncuffed or the cuff is deflated) in order to reduce the risks of inhalation (the speaking valve enables apnoea and positive subglottic pressures to be maintained during swallowing), and thus to enable the restoration of optimal ventilation-swallowing synchronisation (inhalation-swallowing-exhalation). It is also recommended that patients be asked to take a deep breath in before starting voluntary swallowing, if they have the respiratory capacity to do so, in order to increase the subglottic pressure at the time of swallowing (grade B).

4.3. Prevention of complications by nursing care throughout patient follow-up in the hospital setting

The management of neuromuscular diseases requires specific training of nursing personnel.

4.3.1. Changing the tracheotomy tube

The frequency of tube changes depends on the density of secretions, the complications observed and the type of tube. Tracheotomy tubes should be changed at least every 28 days, or once a week in paediatric patients. Subsequent tube changes after the first one may be performed by a nurse on medical prescription (expert consensus). The main risks are the inability to recannulate the patient and incorrect routing.

4.3.2. Local care

It is the responsibility of nurses, and sometimes physiotherapists, to:

- check correct attachment of the tube by the tie in order to prevent accidental decannulation (expert consensus);
- check the cuff pressure using a pressure gauge every 8 hours in the hospital department, as well as after mobilisation of the tube or in the presence of air and fluid sounds (expert consensus);
- clean the tracheotomy opening at least once daily, change the “Y”-slot compress under the neck pad of the tube, and clean and dry the internal lining, change the heat and moisture exchanger (HME) filter and the humidification water (expert consensus).

4.3.3. Nasal passages and oropharyngeal care

- Nasal cleansing should be performed following the department’s protocol (expert consensus).
- If a nasogastric tube (NGT) is in place, its fastening system is changed and the tube is mobilised daily. In the presence of a nasal pressure ulcer and/or acute sinusitis, removal of the NGT is

recommended, with insertion of a tube in the other nostril (expert consensus). In the longer term, an alternative to the gastric tube should be considered, such as gastrostomy.

- Mouth care should be performed several times per day following the department's protocol (expert consensus).
- Secretions from the back of the throat may also be suctioned using a mouth tube by patients themselves, a third person or the nursing personnel (expert consensus).

4.3.4. Endotracheal suctioning

- Endotracheal suctioning using a flexible tube is essential in a patient with secretions. It limits congestion, improves haematosis and maintains tracheotomy tube patency. Aseptic conditions are recommended when handling the suction tube and performing the endotracheal suctioning procedure (expert consensus).
- Following the slow and gradual introduction of the suction tube without exceeding the length of the tracheotomy tube, suctioning should be carried out during a single withdrawal movement, which may be more or less rapid depending on the patient's tolerance and the extent of the secretions (expert consensus).
- It is recommended that instillation be avoided, since this increases the risk of infection (expert consensus). In addition, it causes oxygen desaturation (grade B). It is preferable to use a normal saline aerosol to make secretions more fluid and facilitate their drainage (expert consensus).

4.3.5. Humidification

Effective humidification limits obstruction of the tracheotomy tube by bronchial congestion/secretions. It is necessary to make sure that humidification is working (switched on, water level, temperature). The degree of humidification is tailored to the abundance and consistency of secretions, the patient's comfort and the situation (wheelchair use, day/night). During wheelchair use, an HME filter is the only option, whereas while sleeping or simply lying down, heating humidifiers may be preferred (expert consensus).

Once stabilised, after the first tracheotomy tube change and the switch from an intensive care ventilator to a home ventilator, patients can be transferred to high-dependency unit (HDU) or equivalent structure, or to an expert respiratory service, so that their caregivers can be trained, their ventilation modes adapted, and they can prepare to return to their place of residence (expert consensus).

4.4. Prevention of complications by care throughout patient follow-up in the home setting

The techniques are identical. Therefore family caregivers must be able to receive training in the care procedures in the hospital, bearing in mind that these must be adapted to the constraints of the home and the environment, while maintaining all the hygiene rules (hand washing, hand sanitiser).

- Checking of cuff pressure: this does not always need to be done using a pressure gauge and can be done using a syringe in which the air or water volume required to obtain the correct pressure based on the manufacturer's guidelines will have been measured in advance before the patient's discharge from hospital (expert consensus).
- Performance of mouth care several times per day (expert consensus).
- Different tubes for tracheal suctioning and mouth care, to be changed at least every 24 hours (expert consensus).

- The frequency of tracheotomy tube changes is indicated on a medical prescription, with a maximum of 28 days' use (also see manufacturer's recommendations). Depending on the models, and if required, tubes can be reused after decontamination for a limited duration and number of times (see manufacturer's recommendations) (expert consensus).
- In order of preference, sterile water, distilled water for medical use, or boiled water that has then be cooled to room temperature should be used for the ventilator humidification system. This should be changed daily (clean the reservoir with soapy water) (expert consensus).

4.5. Mucus clearance in addition to suctioning to be performed in hospital and at home

In situations of congestion, as a supplement to endotracheal suction, bronchial drainage techniques, particularly proximal, are recommended (expert consensus).

- The techniques are initiated, tested and optimised in hospital by the physiotherapist in order to determine which ones are most effective and best tolerated by the patient (expert consensus). They are then continued at home by physiotherapists or caregivers.
- The frequency of mucus clearance sessions, the choice and the adaptation of the techniques used are guided by the prior respiratory assessment, in particular taking into account the location and quality of secretions, as well as the patient's tolerance (dyspnoea, oxygen saturation, respiratory rate, heart rate) (expert consensus).
- Measurement of peak cough flow may also be used, in addition to the patient's perception of effectiveness and comfort, and the ease of use of the technique, in order to help in the choice of the different cough assistance techniques proposed, as well as their settings.
- Particular care must be taken if any instrumental techniques are used that require connection to the tracheotomy tube, making sure they are properly secured in place to avoid any pulling on the tracheostomy (expert consensus).

As regards proximal bronchial drainage:

- mechanical insufflation-exsufflation, combined or otherwise with manual compression, is the most recommended technique due to its expected efficacy and ease of use. It is particularly indicated in the event of an acute congestion episode and for patients incapable of cooperating. Mechanical insufflation-exsufflation can also be routinely use to limit the frequency of suctioning (expert consensus);
- manual chest-abdomen compression techniques, associated or otherwise with prior hyperinsufflation, may also be used if mechanical insufflation-exsufflation is impossible or as alternative techniques (expert consensus).

If necessary, peripheral bronchial drainage may be performed. Different techniques can be tested, such as postures or manual compressions that promote expiration towards the expiratory reserve volume or certain instrumental techniques supplemented by the proximal drainage described above (expert consensus).

Irrespective of the bronchial drainage technique used, routine endotracheal and oral suctioning using two separate tubes is recommended to finalise secretion clearance (expert consensus).

Outside periods of bronchial congestion, and in order to prevent the development of atelectasis, the use of hyperinsufflation, using a pressure-control ventilator, for example, or the use of mechanical insufflation-exsufflation can be routinely proposed (expert consensus).

It is recommended that patients and their natural caregivers be trained in the selected manual and instrumental techniques to enable subsequent management at home, bearing in mind that it will not always be possible to access the expertise of a physiotherapist (expert consensus). A re-evaluation of the indications and methods of mucus clearance techniques is recommended at each assessment in order to improve the recommendations made (expert consensus).

5. Patient education and caregiver training before patient discharge from hospital

Training concerns both natural and professional caregivers.

It is delivered in the intensive care unit or in the follow-on care and rehabilitation department in coordination with the neuromuscular reference and/or expert centre with the support of a checklist drawn up in advance and given to the patient on discharge.

It includes, as a minimum, instructions on:

- the equipment that it is essential to have available at home and when moving around;
- the ventilator and the circuits (use of the ventilator, batteries, circuits, humidification, heating, air filtration);
- tracheotomy tube care (verification of tube and cuff patency, opening, cuff, suctioning, tube changes, cleaning, consumable maintenance, liner and ties);
- speaking;
- swallowing;
- mucus clearance;
- use of the bag valve mask (BVM);
- measures to be taken in the event of bleeding, accidental decannulation, obstruction and ventilator failure;
- measures to be taken in the event of triggering of the ventilator alarms.

The equipment required for care must be stored in a clearly defined location and space. It must be readily accessible to all those involved in the patient's care. Stocks must be kept up-to-date. The internal batteries of the devices should be checked to ensure they are fully charged up, but also the external batteries. The back-up ventilator must always be connected and equipped with a circuit, ready to be used very quickly if required.

According to French decree No. 99-426 of 27 May 1999 authorising certain categories of personnel to perform endotracheal suctioning, and the French Order of 27 May 1999 relative to the training of persons authorised to perform endotracheal suctioning, training in endotracheal suctioning is compulsory for patients' families and caregivers (home care assistants, educational and school support professionals [AEVS] for children). Family members of tracheostomy patients can complete this training in the department where the patient was cared for, while for professionals training is carried out by competent nursing training institutes.

In addition, in agreement with the *Haut Conseil de la santé publique* (HCSP - French National Council for Public Health), an application for inclusion on the list of "high risk to life" patients must be sent to the Regional Health Board by patients (or their legal representative) before they return home in order to ensure the electricity supplier is aware of their situation, and so that these patients benefit from a special information service.

6. Switching to the home environment

A return home can only be considered if the following conditions are met:

- both the patient and the family must be ready and willing to return home with continued psychological support if the family so wishes;
- a third person trained in tracheotomy-related procedure must be present 24 hours per day, seven days a week. The application for funding must be made;
- the home must be accessible for people with reduced mobility and include an individual room for the patient, a functional electrical system, access to a bathroom, and enable a third person (caregiver) to be accommodated 24 hours per day, seven days a week;
- a home assessment must be conducted by the service provider supplying the ventilation equipment before the patient returns home;
- the electricity supplier must be fully aware of the patient's reliance on electrical appliances.

The patient should be able to contact the emergency service linked to the neuromuscular reference or expert centre on which he/she depends, as well as the service provider, 24 hours per day and seven days per week in case of need.

The patient must have been identified as a “special patient” and therefore have an emergency liaison file including advance directives (cf. annex 4). This file is sent to the local emergency medical services (SAMU) where the patient lives before discharge from hospital, so that, in a life-threatening emergency, they can be identified whenever they call the service using a dedicated telephone, indicated by the patient and their family.

The reference or expert centre must organise the discharge in liaison with the patient’s primary care physician, nurses, physiotherapists, home care assistants, speech therapist (see annex 1), social worker, etc. Hospitalisation at home (HAH) may be envisaged if necessary during the initial period. There must be a direct link between these healthcare professionals and the department attached to the reference or expert centre responsible for the respiratory follow-up of patients.

The healthcare service provider designated by the patient or their family, from a list proposed by the physician, installs the equipment in advance. This provider intervenes before the patient’s discharge, to put in place two ventilators (with spare batteries), suction machines (with and/or without batteries) and the required consumables (enough tracheotomy tubes to ensure there is always a spare one, appropriate circuits, hydrophobic filters, sterile compresses and non-adherent foil-backed sterile compresses for tracheotomy, suction tubes, etc.). The provider takes the time to explain how the machines operate, and the daily hygiene measures to be applied, and to answer any questions from caregivers and healthcare professionals who will be involved in the patient’s care at home.

A follow-up booklet supplied by the health provider is completed and updated at each visit. The healthcare service provider must visit at least every three months to monitor treatment and at least every four months to monitor the equipment.

It is recommended that the neuromuscular reference or expert centre should discuss and schedule respite care with the patient and their family (expert consensus).

The resumption of a social life to avoid isolation must be facilitated.

A tracheotomy is not a contraindication to outings, going to school, using public transport, using an adapted vehicle, which implies adaptation of the ventilator to the wheelchair, the use of back-up batteries, the presence of an emergency kit (see annex 2), and an accompanying carer.

A tracheotomy therefore increases the burden of care in everyday life and can be an additional handicap to socialisation because of the problem of carers (close family or professionals sometimes extremely difficult to recruit). However, it does provide some satisfaction to patients with neuromuscular diseases in terms of improving the quality and safety of their ventilation in certain situations (feeding, bronchial congestion, communication, etc.). This means being able to envisage short-, medium- and long-term life projects depending on the environment (and in particular schooling for the youngest patients, then studies and the choice, for some, of living independently and working). The internet and social media also offer opportunities to avoid feeling isolated. It is recommended that aids be adapted to each patient, and at each stage, to limit disability.

On a social level, the team of professionals indicated above, as well as disability-related associations, are involved in helping to make life projects a reality.

In the event of travel to a foreign country or and/or air travel, this must be preceded by a medical check-up within 3 months prior to departure, the agreement of the neuromuscular reference or expert centre, as well as the opinion of the service provider in order to assess the feasibility of the trip and to prepare the necessary customs forms. It is then recommended to identify a medical facility that could be used if necessary in the country visited.

7. Future prospects

Evolutions appear to be necessary.

The implementation of this best practice guideline requires the mobilisation of resources (human resources, training, supervision, etc.).

The health authorities must continue to reflect on and support the implementation of this best practice guideline by promoting:

- the development of patient education programmes;
- research in the context of neuromuscular diseases causing ventilation-dependent respiratory failure;
- increase the number of places available in respite care centres for families and patients, in order to reduce the need for referral to neuromuscular reference or expert centres;
- extension of skills (performance of insufflation-exsufflation by nurses, tracheal suctioning by healthcare assistants, use of BVM, recannulation in extreme emergencies, etc.);
- the development of advanced practice nurses for the care of these patients, for hospital-community coordination.

Finally, the possibility of innovative therapies (biologic therapies) may lead to the continuous redefinition and adaptation of care guidelines on the basis of the evolution of knowledge concerning on the efficacy of these new therapies.

Table of appendices

Annexe 1. Speech therapy management: specific features of speech therapy in tracheotomy patients with neuromuscular diseases after their return home

During home rehabilitation, speech therapists need to adapt to the patient's situation, but the difference is that they are alone, without a medical team to support their rehabilitative procedures related to tracheotomy. When the patient leaves hospital, the speech therapist produces a discharge report indicating rehabilitation priorities and conditions, particularly whether the patient is ventilated or otherwise.

When they return home, patients may be on continuous or partial ventilatory support.

Communication assessment

- Assessment of communication needs with the family.
- Speaking options as a function of ventilation.

Swallowing assessment

In addition to motor, sensation and sensory evaluation, the assessment evaluates:

- the time it takes to eat a meal;
- body posture;
- the number and distribution of meals;
- swallowing tests.

Speech therapy priorities

As a function of the above assessments.

Communication/speaking rehabilitation:

- maintenance or implementation of communication methods (speaking alone and/or alternative communication tools);
- coordination work between the vocalisation that is possible (on inspiration and/or expiration) and the speech to be produced (sentence length, inform the family to wait until the end of sentences and not to interrupt). When there is no cuff (or it is deflated) and a speaking valve, patients will be able to speak more easily via their own breathing. If they are ventilator-dependent, they need to coordinate their speaking time with the ventilator.

Swallowing rehabilitation:

- if there are few swallowing difficulties, inflating the cuff is not necessary. In addition, speaking will be facilitated and this reduces the risk of tracheal mucosa injury;
- following the assessment, determine the type of texture for food;
- for swallowing liquids, if the assessment reveals choking, thicken liquids (thickening powder, water thickened to variable degrees). Also use chilled or sparkling liquids that stimulate swallowing and reinforce the airways' protective barriers;
- divide up meals in the event of fatigue;
- adapt the posture of the body and head during meals, as well as the environment (no distractions, such as television; prefer a quiet environment);
- monitor weight.

Adapt all speech therapy methods on the basis of the course of the disease and the disorders.

Annexe 2. Content of the mobile emergency kit

Tracheotomy-ventilated patient emergency bag;

- neuromuscular disease care and emergency card;
- spare tracheostomy tube of the same diameter, with dilator;
- tube with a smaller diameter;
- bag valve mask (BVM);
- tube ties;
- single-dose normal saline; bicarbonate 1.4% for instillation;
- suction tubes;
- portable suction machine (battery operated) with vacuum stop;
- sterile compresses, non-adherent foil-backed sterile compresses for tracheotomy;
- Luer 10 ml syringes if cough;
- pair of scissors;
- HME filters: heat and moisture exchanger filters;
- waterproof vinyl insulating and adhesive tape;
- if ventilatory support: charged spare battery, spare circuit, adhesive tape.

Annexe 3. Multidisciplinary team meeting

Chart specifying how multidisciplinary team (MDT) meetings are organised and operate in the field of tracheostomy in patients with neuromuscular diseases

- **Frequency**: each time a tracheostomy indication is being considered by the medical team, the patient or the family.
- **Coordinator**: the coordinator's role is to draw up a list of patients whose file has been analysed, to inform the MDT members, as well as the patient's referring doctor and, if necessary to invite representatives from disciplines useful for the discussions envisaged.
- **MDT members**

  * Essential permanent members:
    - resuscitation specialist (paediatric if the patient is a child);
    - surgeon (paediatric if the patient is a child);
    - neurologist (paediatric if the patient is a child);
    - psychologist;
    - social worker;
    - contact nurse;
    - physiotherapist.

  * Optional members:
    - as many as necessary depending on the sites.
- **Secretary** (of the coordinator): dated MDT meeting minutes are kept updated by the secretary, making it possible to note the names and qualifications of participants, the patients whose cases are being assessed and the treatment proposal made at each meeting.
- **Analysis of patient cases**: during the MDT meeting, each patient's case is presented by the referring physician and the treatment is defined jointly on the basis of the guidelines selected.
- **Traceability**: all the conclusions of the MDT meeting, at least one copy of which must be placed in the patient's file (paper or electronic), the name of the referring professional who is responsible for follow-up of the proposal (explanation to the patient and organisation of care).
- **Regular assessment** of MDT activity:
  - match between its conclusions and the updated guidelines;
  - consistency between the MDT treatment proposal and the treatment actually delivered. If the treatment actually delivered differs from the MDT proposal, the reasons must be justified by the referring physician and indicated in the patient's file;
  - improvement actions.

Annexe 4. Emergency liaison file

The emergency liaison file makes it possible to organise and anticipate an emergency care pathway dedicated to neuromuscular patients.

Flagging a patient as a *"patient remarquable"** ("special patient") means informing the emergency medical services (15) about the particular situation of a patient receiving home care via their "emergency liaison file", so that this information can be taken into account and transmitted to the doctor who will intervene at home: it may relate to the patient's condition or the specific arrangements requested by the patient.

It makes it possible to maintain consistency in the projects of the patient and his or her family, by avoiding emergency decisions that do not take into consideration the context and the choices expressed. It obviously requires the patient's agreement and may be revoked at any time.

*Definition of a *"patient remarquable"*: these are patients with an advanced and progressive serious disease or patients presenting multiple, non-curable conditions and disorders.

Participants

The following professional bodies and patient and user associations were consulted to propose experts invited individually to take part in working/review groups:

Institutional players and other agencies

(for the review phase)

Agence Nationale de Sécurité des Médicaments et des produits de santé (French National Agency for Medicines and Health Products Safety) (ANSM)

Caisse centrale de la mutualité sociale agricole (Central health insurance fund for agricultural workers) (CCMSA)

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

Direction de la sécurité sociale (French social security division) (DSS)

Direction générale de l'offre de soins (French Directorate General of Health Care Provision) (DGOS)

Direction générale de la santé (Ministry of Health) (DGS)

Learned societies and professional bodies

Association nationale pour les traitements à domicile, les innovations et la recherche (French national association for home care, innovations and research) (ANTADIR)

French college of physiotherapists

French college of general medicine

French national council for ENT and cervicofacial surgery professionals

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

Fédération nationale des orthophonistes (French national federation of speech therapists) (FNO)

Filière nationale de santé des maladies neuromusculaires (French national health network for neuromuscular diseases) (FILNEMUS)

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

Union nationale pour le développement de la recherche et de l'évaluation en orthophonie (French national union for the development of research and evaluation in speech therapy) (UNADREO)

Patient and user associations

Association de défense et d'entraide des personnes handicapées (Association for disabled rights and mutual help) (ADEP)

Association française contre les myopathies (French myopathy association, AFM) - Téléthon

Fédération française des associations et amicales de malades, insuffisants ou handicapés respiratoires (French federation of associations and patient groups for people with respiratory disabilities or failure) (FFAAIR)

Working group

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Ms Adeline Aguado, nurse, ventilation technician, Dijon

Prof. Djillali Annane, intensive care-resuscitation specialist, Garches

Ms Sylvie Arpin, speech therapist, Rosny-sous-Bois

Prof. Shahram Attarian, neurologist, Marseille

Prof. Isabelle Desguerre, paediatric neurologist, Paris

Dr Mostafa El Hajjam, radiologist, Boulogne-Billancourt

Prof. Brigitte Fauroux, paediatric respiratory medicine specialist, Paris

Dr Emmanuelle Jaillette, intensive care-resuscitation specialist, Lille

Mr Matthieu Lacombe, physiotherapist, Garches

Ms Catherine Lamouroux, nurse, Paris

Prof. Françoise Le Pimpec-Barthes, thoracic surgeon, Paris

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Abbreviations and acronyms

AEVS	*Acteur éducatif et vie scolaire* (educational and school support professionals)
AFM-Téléthon	*Association française contre les myopathies* (French myopathy association, AFM) - Téléthon
ALD	*Affection de longue durée* (chronic condition)
ANTADIR	*Association nationale pour les traitements à domicile, les innovations et la recherche* (French national association for home care, innovations and research)
BVM	Bag valve mask
AAC	Augmentative and alternative communication
cmH2O	Centimetres of water
ELF	Emergency liaison file
FILNEMUS	*Filière nationale de santé des maladies neuromusculaires* (French national health network for neuromuscular diseases)
HME filter	Heat and moisture exchanger filter
HAH	Hospitalisation at home
mmHg	Millimetres of mercury
ENT	Ear-nose-throat
$PaCO_2$	Partial pressure of carbon dioxide
PEP	Positive expiratory pressure
PRM	Persons with reduced mobility
$PTcCO_2$	Transcutaneous carbon dioxide pressure
BPG	Best practice guideline
MDT meeting	Multidisciplinary team meeting
SAMU	French emergency medical services
NGT	Nasogastric tube
SpO_2	Pulse oxygen saturation
SPLF	*Société de pneumologie de langue française (French-language Pulmonology Society)*
SRLF	*Société de réanimation de langue française* (French-language Society for Intensive Care)
BCA	Brachiocephalic artery
Ti	Inspiratory time
IV	Innominate vein (or brachiocephalic vein [BCV])
HDU	High-dependency unit
ICU	Intensive care unit
NIV	Non-invasive ventilation

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