AIM
To review the practical aspects (initiation, adjustment and monitoring) of home noninvasive positive pressure ventilation (NPPV) via a nasal, oral or facial interface, when prescribed for patients with neuromuscular disease (NMD).

BACKGROUND
- There is a need for guidelines to bridge the gap between published results and real-life situations in patients' homes and during medical consultations.
- Clinical studies on NPPV have shown that rigorous management achieves high quality results. However, patients and their families often have problems with the practical aspects of NPPV.
- Published clinical studies rarely cover these practical aspects (initiating, adjusting and monitoring NPPV). If mentioned at all, they are described in the "Materials and methods" section of articles.
- Doctors (both general practitioners and specialists) and allied health professionals tend to be unfamiliar with NMD as it is relatively uncommon.
- There is a general lack of knowledge on use of NPPV equipment and methods in patients with NMD. In addition, terminology and equipment in the field of ventilation are constantly changing as a result of technological advances and competition between manufacturers.

POPULATION CONCERNED
These guidelines concern patients with genetic or non-genetic NMD who have been prescribed long-term NPPV. Patients with rapidly progressing forms of NMD and paediatric patients are the subject of either discussion or specific recommendations.
NPPV is the method of first choice when assisted ventilation is indicated in a patient with NMD and should be initiated in a specialist unit suited to the patient's age and disease.

As respiratory muscle weakness has repercussions on all phases of cough, long-term NPPV can only be prescribed in a patient with NMD if it is accompanied by cough assistance techniques for managing congestion.

Medical staff and allied health professionals (working either in hospital or the community) treating patients with NMD should be trained in the equipment and methods used in NPPV and cough assistance.

Initiation of long-term NPPV requires the informed consent of the patient and, if appropriate, their family. They should be given full, clear information about the respiratory disorder, its likely course, and the advantages and drawbacks of available treatment options, and should be trained in using ventilation equipment and in assisted cough techniques.

Once equipment and settings have been chosen, adjustments have often to be made. Frequent patient monitoring is required to check for possible causes of failure, and find solutions.

Continuity of care between the team initiating NPPV and the home care team is essential.

The patient's own doctor and allied health professionals from the service provider's organisation (i.e. the organisation supplying and maintaining the equipment) should monitor the patient's respiratory status from time to time during home visits, as well as during periodic visits to the unit where NPPV was initiated.

The service provider is responsible for the maintenance of the equipment, the supply and renewal of consumables, and repairs. Help should be available 24 hours a day, 7 days a week via a telephone helpline and/or home visits.

If respiratory status deteriorates under NPPV or problems remain despite changes to the equipment and/or settings, the doctor may, with the support of the patient's carers, propose the use of ventilation by tracheostomy.
Indication for assisted ventilation

Choice of NPPV as assisted ventilation method
(patient and patient's family given clear and complete information about the respiratory disorder, its course, treatment options and informed consent)

Initiation in a facility (usually hospital) suited to the patient’s age and disease
• choice of ventilation method
• choice of device
• choice of interface
• choice of circuit
• choice of initial settings
• initiation: protocol for one or more days, which need not be consecutive, including adjusting equipment and settings, to correct any hypoventilation while maintaining patient comfort (monitoring, tests)
• training the patient and the patient's family in using NPPV equipment and cough assistance techniques
• the team responsible for home monitoring should contact the patient and the patient’s family before the patient is discharged home.

Discharge home
• the patient's own doctor is given detailed information about the diagnosis, the patient's current condition, the potential risks, the likely course of the disease, and any treatment given
• an allied health professional from the service provider delivers home care at regular intervals; the technical department provides permanent support through regular visits and visits on request
• the medical unit (usually a hospital unit) where NPPV was initiated monitors the patient regularly; the monitoring pattern will depend on the course of the disease
• each professional passes on information collected to all other professionals involved
• the patient should be given a short summary of the history of their disease, ventilator settings, the risks associated with different situations and types of drug therapy, a list of equipment to be transported, and contact details for the referral centre
• the patient and their family should be given a document describing what to do in the event of
  ◦ technical problems (call to the service provider, use of emergency equipment, technical service available 24 hours a day, 7 days a week);
  ◦ or medical problems (call to the patient's own doctor, general emergency services, specialist emergency services available 24 hours a week, 7 days a week).

This Quick Reference Guide is an English summary of the main points of the French guidelines:
“Modalités pratiques de la ventilation non invasive en pression positive, au long cours, à domicile, dans les maladies neuromusculaires” – Clinical practice guideline – May 2006.