Service Specification No.	A14/S/01 Revision
Service	Respiratory: Complex Home Ventilation (Adult)
Commissioner Lead	Kathy Blacker
Period	12 months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

This specification relates to the hospital infrastructure necessary to enable the safe and sustainable establishment and maintenance of home care for patients with complex ventilation needs. This infrastructure includes:

- Inpatient critical care beds for the assessment of new patients and care of established patients with acute or chronic deterioration.
- Diagnostic services to assess patients with suspected complex ventilatory needs.
- Multidisciplinary teams (MDTs) to provide holistic assessment of care for patients with multi-system disease. MDTs should include dedicated respiratory physician with a major interest in ventilatory support, clinical nurse specialists, ward manager physiology technicians, physiotherapists, dieticians, speech and language therapy and occupational therapy.
- 24/7 emergency availability of advice for existing patients and to local centres
- 24/7 emergency response in the event of equipment failure.

What is Home ventilation?

- Home ventilation is the provision of respiratory support at home to treat patients with chronic respiratory failure.
- It is typically delivered non-invasively using a mask and ventilator, but

includes invasive ventilation via tracheostomy.

- Home ventilation is indicated in a number of diagnostic conditions. The main diagnostic groups include (but are not limited to) chest-wall disorders, neuromuscular disorders, COPD, and Obesity-Hypoventilation Syndrome.

What is 'complex' home ventilation?

Long term ventilation is used in a number of clinical situations. These range from its use in patients with sleep-disordered breathing to the need for continuous invasive support via tracheostomy in patients with complex conditions. In the former patients, ventilation is used during sleep periods only and can be discontinued for more than 24 hours without clinical harm. In the latter patients, there may be complete ventilator-dependence and the patient requires complex, high-cost care. One-to-one support is needed for such patients as inadvertent disconnection from the ventilator for more than a minute or two could prove to be fatal.

The overarching principle is that care needs to be provided on a patient-centred basis. This Complex Home Ventilation Service Specification relates to the hospital infrastructure that is necessary to enable the safe and sustainable establishment and maintenance of home care for patients with 'complex' ventilation needs. It is therefore defined by the characteristics of the patients who require this service and by the infrastructure of the service itself.

The complexity of an individual's care needs may vary over time, sometimes becoming more complex, and sometimes less so. Regardless of diagnosis, care requirements may change over time. The overall provision of home ventilation care needs to recognise this via an organised network approach to care. Access to complex care should be facilitated, as should a return to local units or shared-care approach when appropriate.

In general terms, the principles that define the 'complexity' of care relate to the characteristics of the patient groups who may benefit from home ventilation and the significant variation in care needs. Appendix 3 provides these characteristics in further detail. However, the most important patient-related factors comprise;

1. **Diagnosis**

- Grouping patients according to diagnosis is easier for coding and commissioning purposes. However, the complexity of care needs cannot be defined by diagnosis alone on an individual patient level.
- Nevertheless, there are some patient groups that can be identified as 'complex,' since they will typically meet some or all of the criteria outlined below.
- Patients with neuromuscular conditions, such as Motor Neurone Disease (MND), Duchenne Muscular Dystrophy (DMD), and Myotonic Dystrophy where diagnosis alone may identify the need for 'complex' input given the

speed of progression and the need for discussions of possible interventions. For progressive neuromuscular conditions that are characterised by death due to respiratory failure, early referral for respiratory monitoring is recommended.

2. Degree of dependence upon assisted ventilation

- Patients who require assisted ventilation for 14 hours or more during a 24 hour period are at risk of significant clinical harm if ventilation is interrupted.
- Back-up ventilators and consumables are required.
- Such patients (and their carers) require 24/7 nursing and technical support that provides the ability to trouble-shoot emerging problems (via telephone initially), the provision of further ventilator(s) and consumables via courier if necessary, and access to clinical advice as necessary.
- Hence, regardless of relative clinical stability, ventilator-dependent patients need to be able to access the above support via the clinical service that is responsible for their care.
- Patient groups at greater risk of ventilator-dependence are summarised in appendix 3.

3. **Need for complex physiotherapy input**.

- Assisted ventilation is only one aspect of respiratory care.
- Patients with complex medical problems frequently require expert physiotherapy support (for example, use of cough-assist devices). The importance of skilled multidisciplinary care has been proven for patients with Motor Neurone Disease (MND).
- The requirement for any form of assisted-cough intervention is a helpful marker that the individual is 'at risk' in that their ability to clear their own secretions is compromised. Such patients need access to prompt expert advice and intervention in the event of clinical deterioration.

4. Need for specialised monitoring and treatment, including the coordination of non-respiratory Multidisciplinary Team care

- Published evidence shows that patients with selected neuromuscular conditions (such as DMD and MND) benefit from coordinated specialist multidisciplinary care.
- Where possible, these should be coordinated via a single centre (e.g. combined respiratory and cardiac monitoring for patients with DMD).
- In other patients, referral to the 'complex' centre may be required for detailed assessment or optimisation of ventilation (for example, prior to Bariatric surgery or for a specific procedure such as Percutaneous Endoscopic Gastrostomy (PEG) placement).
- Patients supported with long-term invasive ventilation via tracheostomy.
- Patients who are transitioning from Paediatric services.

5. Difficulties in starting or continuing assisted ventilation

- Patients should be referred to the 'complex' centre from other home ventilation units when aspects of their care fall outside the expertise of such units.
- This includes patients who fail to respond to initial therapeutic strategies and those who deteriorate unexpectedly during treatment.
- The supervising clinical team should consider early discussion and onward referral for specialist assessment and management in such circumstances.

A 'Complex Home Ventilation Service' is defined as a clinical service that provides the necessary infrastructure for all aspects of patient care as outlined above. Essential characteristics of the complex home ventilation service infrastructure are described in Section 3.2.

Evidence base

Evidence supports the use of long term ventilation for selected patients with chest-wall disorders (such as scoliosis and post-thoracoplasty), neuromuscular disorders, parenchymal lung diseases (such as chronic obstructive pulmonary disease), obesity-hypoventilation syndrome, and selected patients with sleep-disordered breathing.

Appendix 2 describes this evidence base, the evolution of home ventilation services for adults within the NHS, and the characteristics of care that define 'complexity.'

National Drivers

- NICE guidance in managing patients with MND emphasises that Non Invasive Ventilation (NIV) should be delivered by competent respiratory professionals.

- National Patient Safety Agency (NPSA) alerts have identified cases where problems with administering NIV caused at least moderate harm. Failures in training and competence are recurring themes.

- Providing assisted ventilation at home can result in discharge delays and the requirement for a care package. Patients who are discharged from hospital to continue invasive ventilation via tracheostomy in the community require significant care. Costs are very high as a consequence. In UK practice, transfer to a complex ventilation / weaning centre improves weaning rates (removal of tracheostomy), reducing the need for high-cost community care. This has major implications for health and social care budgets. It also makes a major difference to patients and those close to them.

- National Guidance for the provision of aspects of specialist non-ventilation services to patients exists for some individual patient groups, e.g. MND, DMD, and around specific technologies e.g. diaphragmatic pacing and tracheostomies. There are some national standards available and some specialist society guidance.

- Providing complex home ventilation services also falls within the NHS Outcomes Framework Domains 1 (preventing people from dying prematurely) and 2 (enhancing quality of life for patients with long term conditions). Domain 5 (long-term care at home) is generally safer than in a hospital or other institutional environment. Improvement Area 1a specifically identifies reducing mortality from respiratory disease.

2. Outcomes

2.1 NHS Outcomes Framework Domains & Indicators

Domain 1	**Preventing people from dying prematurely**	√	
Domain 2	**Enhancing quality of life for people with long-term conditions**	√	
Domain 3	**Helping people to recover from episodes of ill-health or following injury**	√	
Domain 4	**Ensuring people have a positive experience of care**	√	
Domain 5	**Treating and caring for people in safe environment and protecting them from avoidable harm**		

1. Providers of home ventilation services should keep robust service-level data to show that they continue to meet the requirements of the above NHS Outcomes Framework Domains and Indicators. There is no current national registry, but a uniform approach to monitoring is recommended using data that should be available to each centre. Examples could include;

- Number of patients treated with assisted ventilation within that service (all patients, and categorised into the following groups; ventilation via tracheostomy, neuromuscular disease, NIV-dependence (defined as either use of ventilator >14 hours, or provision of 2 ventilators), patients <19years old).

- Clinical review* (% of patients who receive a formal clinical review within 3 months of starting assisted ventilation).

- Annual clinical review* to assess the adequacy of ventilation (% of patients who receive a formal annual review that includes measurement of the adequacy of ventilation, ideally via blood gas analysis).

- NIV compliance (annual measure) (% of patients using NIV for an average of 4 hours or more / 24 hours).

- Patient survival after starting ventilation** (all patients and patients with neuromuscular disease).

2. Appropriate training provided to the patient and carers prior to community discharge and a service infrastructure that is appropriate for the patient's needs (Domain 5).

* Clinical review – Clinical review is one that is undertaken by an appropriately trained professional and therefore includes allied health professionals with specific expertise in the management of patients who are treated with home mechanical ventilation.

** Patient survival – Confounders such as patient selection and extent of clinical stability at the time of starting NIV mean that patient survival alone is an unhelpful metric if all patients within the service are included. However, an important aim of the specification is to improve survival in individuals with chronic respiratory failure. This data should be collected annually to assess trends. Survival of patients with MND may be less prone to such confounders and should be reported separately.

3. Scope

3.1 Aims and objectives of service

Aims

- To improve survival and quality of life for individuals with chronic respiratory failure.

- To develop an equitable national complex home ventilation service whereby

patients will have access to specialist multi-disciplinary diagnosis, treatment and follow up. The service aims to reduce current variation in the accessibility of such services.

- To establish and support patients with chronic respiratory failure onto the most appropriate and least intrusive mode of ventilation that is possible.

Objectives

The Complex Home ventilation Service will deliver these aims by;

- Providing a specialist multi-disciplinary service for the diagnosis and treatment of complex ventilatory failure.
- Preventing premature death by ensuring equitable access to appropriate specialist treatment.
- Maximising the possibility of patients being able to live in their own homes on the most appropriate and least invasive mode of ventilatory support. The maximum amount of time possible each day should be spent breathing spontaneously, hence improving the quality of life of both patient and carers (if required), and maximising the chances of returning to work.
- Delivering high-quality training and support for family members, carers and the wider healthcare network in the care of a ventilator-dependent patient.
- Where appropriate, the Complex Home Ventilation Service will provide specialist outreach to support and educate patients carers and the healthcare network
- Ensuring the cost-effective use of expensive resources.
- Ensuring that the method of delivery and settings used to deliver assisted ventilation are titrated to optimal levels via appropriate monitoring and assessment. This includes the availability of dedicated inpatient beds in which continuous respiratory physiological monitoring can be conducted.
- 24/7 emergency availability of advice for existing patients and to local centres
- 24/7 emergency response in the event of equipment failure
- Providing access for patients receiving treatment in other home ventilation units within their network when aspects of care fall outside the expertise of such units. Following assessment / management, such patients' may or may not continue to require input from the complex home ventilation service. If care requirements become 'non-complex,' then further care delivery should return to the original unit.
- Developing and sharing regional (network) and national protocols and guidelines. The complex home ventilation service should work with local teams (and vice versa) to ensure that all patients treated with home mechanical ventilation within

their network receive high-quality care. For some patients, a shared care approach is recommended if this results in a more effective and convenient service. Development of effective regional networks should improve access into and out of the complex centre. For this reason, relevant outcome measures, as described in Section 2, should apply to ALL providers of home ventilation services, including those who do not manage patients with complex requirements.

3.2 Service description/care pathway

This specification relates to the hospital infrastructure necessary to enable the safe and sustainable establishment and maintenance of home care packages for patients with complex ventilation needs.

Patient identification;

1. Via diagnosis

- For example Neuromuscular diseases

2. Via degree of dependence upon assisted ventilation

- Requirement for assisted ventilation for 14 hours or more / 24 hour period.
- Referral to a 'complex' centre is necessary if the existing centre is unable to provide 24-hour nursing and technical cover.

3. Via need for complex respiratory physiotherapy

- Patients who require expert, multidisciplinary physiotherapy input (e.g. provision of cough-assist advice and devices for patients with Muscular Dystrophies and MND) should be referred to the 'complex' centre.
- Most patients who require this assessment will have other complex care needs and therefore may remain under the care of the 'complex' centre. However, if this is not the case, then care could return to the existing centre.

4. Via need for specialised monitoring and treatment

Individuals with progressive conditions for whom timely specialist intervention improves outcomes. Such patients require long-term specialist multidisciplinary monitoring (e.g. cardiology, neurology, physiotherapy). Conditions include;

- DMD
- Other muscular dystrophies
- MND
- Myotonic Dystrophy
- Long-term invasive ventilation via tracheostomy
- Transition from Paediatric service

Where possible, care for patients should be delivered at home or as close to home as possible. The balance is to provide care closest to home without compromising access and delivery of the best possible care; service models may differ in their approach to achieve this.

5. Via difficulties in starting or continuing assisted ventilation

- Individuals who have failed to respond to initial therapeutic strategies (or deteriorate unexpectedly) at their local centre.
- Depending upon local inpatient bed provision, patients with Chronic Obstructive Pulmonary Disease (COPD) (or other patients who continue to require ventilatory support following a period of acute ventilation) may benefit from transfer to a Complex Ventilation Centre to assist with NIV optimisation prior to community discharge.
- Once established on long-term NIV, care may return to the local centre.

Key Complex Home Ventilation Service infrastructure identifiers;

Clinical Personnel;

- Care led by a consultant team with specific expertise in home ventilation.
- A nursing team with specific expertise in the provision of home ventilation.
- A physiotherapy team with expertise in the management of this patient group (for example, expertise in providing advanced secretion clearance techniques)
- Continuity of care provided by all key professionals above.
- Access to on-site Speech and Language Therapy, Occupational Therapy, and Dietetic support, when clinically indicated.

Infrastructure;

- Dedicated inpatient beds to enable bedside monitoring of ventilatory function, especially during sleep
- On-site critical care is necessary to provide higher level care in the event of patient deterioration.
- Resident 24/7 on-call cover of sufficient expertise and seniority to manage common airway emergencies.
- Access to ward-based bronchoscopy as required on an urgent basis 24/7.
- Access to Pulmonary physiology assessment, including the ability to measure lung function and muscle strength and to perform sleep studies.

Technical Support;

- All patients within the complex home ventilation service should have direct telephonic access to the service on a 24/7 basis (a helpline that enables direct 24/7access to a member of the team who can triage clinical queries safely and appropriately). A consistent and safe response requires sufficient clinical experience (usually a senior member of the nursing team who follows a locally-agreed pathway).
- Reliable technical support for the servicing and maintenance of home ventilators, including the provision for ventilators couriered to patients' own homes on an urgent (same night) basis if this is clinically indicated.
- Back-up ventilators and batteries should be provided to patients who have significant ventilator-dependence (>14hrs/24).

A 'Complex Home Ventilation Service' therefore provides the necessary infrastructure for

all aspects of complex inpatient assessment and long-term home care for patients who require home mechanical ventilation.

- Provide a reference point for clinicians and commissioners to ensure a network approach to patient care.
- Patients should have access to specialist, complex care when indicated; care may return to an existing local centre if and when the "complex" intervention has been completed. A collaborative approach between specialised and local centres is required.

Service Entry Criteria

Patients who meet the following criteria;

- Assisted invasive ventilation via tracheostomy.
- Patients treated with NIV who are transitioning from Paediatric Services.
- Individuals treated with NIV who have complex requirements necessitating repeated specialist multidisciplinary input (e.g. Neurology, Cardiology DMD, spinal muscular atrophy, Becker's muscular dystrophy, Limb Girdle Muscle Dystrophy, and Myotonic Dystrophy).
- Individuals treated with NIV (or at risk of ventilatory failure due to the progressive nature of the disease concerned) who require expert, multidisciplinary physiotherapy input (e.g. provision of cough-assist advice and devices in those for example with Muscular Dystrophies and MND).
- Assisted non-invasive ventilation in patients with a requirement for complex ventilation assessment and/ or management.

Service Exit Criteria

- Patients referred from an existing provider of home ventilation services within a network should return back to that service once they no longer meet criteria for continued management in the 'complex' centre. To establish the general principles, examples here include but are not limited to
    - Patients referred with sub-optimal control of sleep disordered breathing (sleep apnoea syndromes / obesity hypoventilation) will be discharged back to the local NIV unit when ventilatory support has been optimised.
    - Patients with obesity-hypoventilation syndrome in whom Bariatric surgery is being considered; such patients may benefit from optimisation of NIV ahead of surgery, but may return to their original service at a later date.
    - Patients with COPD in whom there have been difficulties in establishing home ventilation or in whom there is a need for further assessment regarding the potential benefit of treatment. Once treatment has been established, future care may revert back to the original centre.

3.3 Population covered

The service outlined in this specification is for patients ordinarily resident in England*; or otherwise the commissioning responsibility of the NHS in England (as defined in '*Who Pays?: Establishing the responsible commissioner'* and other Department of Health

guidance relating to patients entitled to NHS care or exempt from charges).

* Note: for the purposes of commissioning health services, this EXCLUDES patients who, whilst resident in England, are registered with a GP practice in Wales, but INCLUDES patients resident in Wales who are registered with a GP practice in England.

3.4 Any acceptance and exclusion criteria and thresholds

Referral sources:

- Local acute care hospitals
- Local ventilation units or sleep services
- Consultant Respiratory Physicians
- Critical care units
- Consultant Neurologists
- Paediatric consultants to facilitate transition of patients already ventilated and surveillance of those who may need ventilating in the near future
- Rehabilitation consultants
- Other specialist teams (e.g. Endocrinology / Metabolic / Trauma/ Bariatric services).

Acceptance criteria:

- All long-term tracheostomy ventilated patients
- Patients with multi-system progressive disease needing multidisciplinary assessment of ventilator requirements.
- Young adults (aged 13-19 years) transitioning from specialist paediatric services, as described in the Paediatric Long Term Ventilation Service Specification (E07/S/c).
- Individuals with chronic respiratory failure likely to require more than 14 hours per day NIV including those with comorbidities
- Patients with progressive neuromuscular disease suspected to require or treated with NIV.

Exclusion criteria:

- Overnight NIV for uncomplicated sleep disordered breathing.
- Paediatric patients. As described in E07/S/c, transition may start from aged 13 years following contact from the Paediatric Long Term Ventilation team.
- Individuals who require ventilation as a consequence of Spinal Cord Injury (traumatic or non-traumatic).

3.5 Interdependencies with other services/providers

Co-located services

- Critical Care Unit
- Physiotherapy
- Speech and language
- Dietetics

- Occupational therapy
- Sleep diagnostics
- Radiology

Interdependent services

- Neurology/neurosciences
- Palliative care
- Gastroenterology (gastrostomy feeding tube insertion for progressive MND)
- Cardiology (for inherited progressive MND)

Related services

- Units within the same regional network providing acute NIV services
- Neurorehabilitation (medical, physiotherapy and occupational therapy)
- Surgery (spinal, ear nose and throat (ENT) and urology)
- Morbid obesity services
- Sleep services and ventilation units that lack the infrastructure to manage patients with complex care requirements (e.g. units providing NIV for patients with uncomplicated sleep disordered breathing).

4. Applicable Service Standards

4.1 Applicable national standards e.g. NICE

Service Standards

- Care should be led by a respiratory consultant with specific expertise in complex ventilation. To provide a sustainable service, a minimum of three (full-time equivalent) consultants will be required. This is likely to require a caseload of at least 300 patients who are actively supported with home ventilation (not all of whom will be complex home ventilation patients) to ensure a viable service.

- Consultant work patterns must deliver continuity of care.

- A consultant with expertise in respiratory physiology and complex ventilation must be dedicated to the service and be available during working hours. Consultant input and review if needed should be available 24 hours per day, 7 days per week, with availability to attend on an on-call basis in-line with local policy (normally within 30 minutes of contact).

- Consultant-led ward rounds supported by the senior nursing team must occur every day, seven days per week and include daily input from the specialist nursing and physiotherapy teams.

- An identified Lead Nurse with overall responsibility for the nursing elements of the service.

- A robust Nurse training programme that enables the generation and maintenance of competencies in the management of patients who are invasively ventilated via tracheostomy and in the provision of assisted ventilation via NIV. It is expected that there will be a designated Nurse lead with responsibility for this role.

- An identified designated Lead Physiotherapist with overall responsibility for the respiratory physiotherapy and rehabilitation elements of the service.

- A physiotherapist with expertise in the management of this patient group (expertise in secretion clearance techniques) must be available immediately during working hours and be able to attend on an on-call basis in-line with local policy (normally within 40 minutes of contact).

- An on-site critical care unit is necessary to provide higher level care in the event of patient deterioration.

- There must be resident 24/7 on-call cover of sufficient expertise and seniority to manage common airway emergencies.

- It must be possible to undertake bronchoscopy on an urgent basis 24 hours per day, 7 days per week for inpatients.

- All patients should have on-site access to a Speech and Language Therapist, Occupational Therapist(s), and Dietetic support, when clinically indicated.

- All patients should have access to pulmonary physiology assessment, including ability to measure lung function and muscle strength and to undertake sleep studies.

- Reliable technical support must be available for the servicing and maintenance of home ventilators. When clinically indicated, there must be provision for ventilators to be couriered to patients' own homes on an urgent (same night) basis.

- All patients should have access to the Complex Ventilation Unit on a 24/7 basis (a helpline that enables direct 24-hour per day access to a member of the team who can triage clinical queries safely and appropriately – this may be achieved by providing direct telephone access to the inpatient ward).

Databases

Databases of patients are often held at a local level by units providing this service, but these are not standardised in terms of the information held, and do not provide a comprehensive picture of national provision.

Consideration needs to be given to database formation and maintenance with necessary technical expertise. Ideally, there should be a national registry.

4.2 Applicable standards set out in Guidance and/or issued by a competent body (e.g. Royal Colleges)

National and International Guidance

'*Motor neurone disease: the use of non-invasive ventilation in the management of motor neurone disease*' NICE clinical guideline 105, 2011

Respiratory Care of the Patient with Duchenne Muscular Dystrophy

American Thoracic Society (ATS) Consensus Statement, March 2004

NICE Interventional Procedure Guidance (IPG)307 *Intramuscular diaphragm stimulation for ventilator dependent chronic respiratory failure due to neurological disease*: guidance 2009

NICE quality standard for Chronic Obstructive Pulmonary Disorder, standard 11: Relates to provision for NIV. 2011

Non-invasive ventilation in chronic obstructive pulmonary disorder: management of acute type 2 respiratory failure - Royal College of Physicians/British Thoracic Society concise guideline, 2008 Updated version expected 2016

National Patient Safety Agency (NPSA) Difficult airway society, Intensive care society National Tracheostomy Safety Project currently underway: guideline development

NPSA Signal: non-invasive ventilation 2011

European Respiratory Society Task force Weaning from mechanical ventilation. Eur Respir J 2007; 29: 1033–1056. Statement of the Sixth International Consensus Conference on Intensive Care Medicine Organised jointly by the European Respiratory Society (ERS), the American Thoracic Society (ATS), the European Society of Intensive Care Medicine (ESICM), the Society of Critical Care Medicine (SCCM) and the Societe de Reanimation de Langue Francaise (SRLF), 2007

5. Applicable quality requirements and CQUIN goals

5.1 Applicable quality requirements (See Schedule 4 Parts A-D)

5.2 Applicable CQUIN goals (See Schedule 4 Part E) To be agreed with the commissioner.
6. Location of Provider Premises
The Provider's Premises are located at: These services are not nationally designated services.
7. Individual Service User Placement
Not applicable.

Appendix One

Quality standards specific to the service using the following template:

Quality Requirement	Threshold	Method of Measurement	Consequence of breach
Domain 1: Preventing people dying prematurely			
Providing patients with competency training to enable safe discharge to community care.*	To be agreed	Audit of practice	
Provision of a personal care plan with specific guidance on emergency management and escalation plans.	To be agreed	Audit of practice	
Domain 2: Enhancing the quality of life of people with long-term conditions			
Follow up plan agreed on starting home ventilation, with this initial clinical review to take place within 3 months of starting ventilation.	To be agreed	Audit of practice / annual report	
Annual clinical review that includes an objective measure of adequacy of ventilation (e.g. blood gas measurement).**	To be agreed	Audit of practice / annual report	
Domain 3: Helping people to recover from episodes of ill-health or following injury			
Agreed managed hospital discharge pathway that facilitates discharge home or care closer to home (e.g. if rehabilitation is required prior to discharge home)	To be agreed	Audit of practice	
Domain 4: Ensuring that people have a positive experience of care			
Local patient satisfaction survey	To be agreed	Audit of practice	

Domain 5: Treating and caring for people in a safe environment and protecting them from avoidable harm			
Providing access to 24-hour support for advice and facilitating early transfer to the unit when clinically indicated.	To be agreed		

*When the responsibility for community care is adopted by an external provider, then further training of carers becomes the responsibility of that community agency. The training of carers to use ventilators, cough assist devices and other devices remais the responsibility of the Complex Home Ventilation Service/

**Clinical review may be undertaken by a doctor or allied health professional with suitable training.

Appendix 2

Explanatory text for service commissioners

Evolution of UK home ventilation services

- Prior to the 1980s, few patients received ventilatory support at home. Those who did used negative pressure devices (such as cuirass).
- Nasal continuous positive airway pressure (CPAP) devices were developed in the 1980s to treat patients with Obstructive Sleep Apnoea-Hypopnea Syndrome (1). This led to the recognition that mask ventilation (NIV) could be used in acute respiratory failure in the hospital setting. NIV was also used on a long-term basis in selected patients with chronic respiratory failure.
- NIV proved to be simpler and more effective than the existing negative pressure approaches and soon became the main method for long-term support.
- The number of adults treated with home ventilation is recorded by each centre (at least via an equipment database). However, collation of this data nationally is not available and accurate UK home NIV prevalence data are lacking.
- In 2000, the Eurovent study (2) reported a UK prevalence of 4.1 patients per 100,000 population receiving NIV. Since Eurovent, data from existing units show a 6-10 fold increase in home NIV usage.
- In 2012, a UK survey (3) showed an increase in the number of providers of home ventilation services (increasing from 26 to 40 units in a 10-year period), alongside an increasing range of indications, spanning neuromuscular disorders, lung disease, chest-wall disorders and obesity-hypoventilation (with improved recognition of the benefits of NIV).
- The Paediatric Long Term Ventilation Service Specification (E07/S/c) notes that 'recent medical literature from the UK demonstrates a significant rise in the number of children on long term ventilation, although the population remains a relatively small volume, specialised and high cost one.' It is estimated that there are approximately 250 tracheostomy-ventilated children managed out of hospital and a further 1000-1300 children with complex needs dependent upon NIV under specialist follow up. Whilst the majority will have conditions that are significantly life-limiting, advances in care mean that an increasing proportion are surviving childhood to transition to adult services.

Evidence base

Chest-Wall disorders

- Patients with restrictive chest-wall disorders, such as scoliosis were the earliest cohort to use home NIV. In selected patients, it provides a survival benefit compared to long-term oxygen therapy (4-7).

Neuromuscular disorders

- In selected patients with neuromuscular disorders, such as Duchenne Muscular Dystrophy and Motor Neurone Disease (MND), home NIV improves symptoms, quality of life and survival (8,9).
- Muscle weakness (pump failure) is the mechanism of respiratory failure in most patients with neuromuscular diseases. In such circumstances, NIV provides effective breathing support when respiratory muscle function is limited. These principles are similar (and therefore apply) for patients with other neuromuscular conditions.

Chronic Obstructive Pulmonary Disease (COPD)

- NIV improves quality of life and survival in selected patients with hypercapnic COPD (e.g. those in whom adequate oxygenation via long-term oxygen therapy results in uncontrolled hypercapnia).
- NIV provides cost-effective care in hypercapnic patients who experience frequent exacerbations that require acute NIV.
- Randomised studies have provided conflicting results (10-14). Reasons for differences in trial outcomes include patient selection and adequacy of NIV treatment used. Some earlier studies suggested a lack of benefit, but were limited by the ineffective delivery of ventilation. Recent data (12) shows that long-term NIV provides benefit, providing that efforts are made to reduce hypercapnia upon starting NIV.
- A major UK study (HOT-HMV) should help our understanding. Recruitment is complete and follow-up is in progress. The trial has been configured to represent UK clinical practice.
- Obstructive Sleep Apnoea Syndrome (OSAS) may occur in patients with COPD. This is termed 'Overlap Syndrome.' CPAP use in patients with moderate to severe OSAS is supported by good quality trial data. CPAP or NIV can provide successful outcomes for patients with Overlap Syndrome. Choosing between CPAP and NIV is a clinical decision, often according to the degree of impairment in ventilation.

Obesity-Hypoventilation Syndrome (OHS)

- Rising obesity rates have led to an increase in the number of individuals who develop respiratory failure as a consequence of obesity (obesity-hypoventilation syndrome, OHS). OHS may occur as a result of uncontrolled severe OSAS (90%) or via hypoventilation alone (absence of OSAS on sleep study, approximately 10% of all patients with OHS).
- At least 5% adult population are likely to have significant OSAS, of whom 10-20% may have OHS.
- Untreated, outcomes are poor with up to 33% mortality over 18 months from recognition of chronic respiratory failure. OHS is now a common indication for NIV.
- Improvements in physiological outcomes can be shown in clinical studies (16). Long-term treatment with NIV improves quality of life and reduces mortality.

General principles determining the complexity of care

Chest-wall disorders

- For most patients with chest-wall disorders, starting NIV is uncomplicated. Long-term adherence and benefit are expected.
- Whilst top-up daytime use is needed for some, significant NIV-dependence during wakefulness is uncommon.

Neuromuscular disorders

- Starting NIV is usually straightforward for patients with neuromuscular conditions.
- However, for patients with rapidly progressive conditions, such as MND, the assessment, decision-making, and communication requirements can be complex.
- Trial data shows that some patients with MND are more likely to gain benefit than others (non-bulbar vs. Bulbar-onset disease). However, patients rarely present at either end of this spectrum and the rate of individual decline shows significant variability.
- Patients with progressive neuromuscular disorders may develop increasing dependency upon NIV with advancing disease. Daytime usage may be required and some become totally dependent upon assisted ventilation, occasionally via tracheostomy.
- Complexity of care is increased in the event of swallowing difficulties and chest infections. Access to specialist physiotherapy is essential.

Chronic Obstructive Pulmonary Disease (COPD)

- Starting home NIV in COPD can be complex. As noted above, trial evidence is conflicting. Patient selection is key.
- In UK practice, the need for home NIV may be identified following acute NIV treatment. The need for long-term NIV should be considered during a period of clinical stability. However, some patients fail to maintain satisfactory blood gases on stopping acute NIV. Starting home NIV may allow safe discharge home in such circumstances. Reassessing the need for NIV once stable is recommended.
- Improving ventilation (reducing carbon dioxide) is a necessary outcome when home NIV is started. Achieving this in patients with COPD can be challenging. In clinical practice, adapting to NIV and optimising settings may require a longer period of inpatient adaptation compared to some other diagnostic groups.
- Once home NIV is established, long-term care is less complex. Most patients with COPD require NIV only at night, although some with more advanced disease may use NIV for periods of the day.

Obesity-hypoventilation syndrome (OHS)

- Starting NIV is typically straightforward.
- Once established, NIV is required during sleep and treatment requirements are not complex for most patients, although more holistic care may be achieved via links with other multidisciplinary specialist services (such as bariatric services).

Complex Sleep Apnoea Syndrome

- Most patients with moderate to severe obstructive sleep apnoea are expected to benefit from simpler therapies, including continuous positive airway pressure (CPAP) or simple NIV.
  .
- A small proportion of patients develop central sleep apnoea whilst using CPAP therapy. This is termed complex sleep apnoea and is characterised by a combination of central and obstructive apnoeas.
- Referral for complex sleep / ventilation assessment may be required to optimise sleep disordered breathing in this (and similar circumstances). Once optimised, care may return to the original centre.

Transition from Paediatric services

- Transition is defined as a purposeful and planned process of supporting young people to move from children's to adults' services.

- Transition takes place at a pivotal time in the life of a young person. However, there is evidence that transition services are inconsistent, patchy and varied depending on the condition. A loss of continuity in care can be a disruptive experience and can result in clinical harm. Young adults with complex and multiple needs, such as long term ventilation, are at particular risk of falling into service gaps.
- As described above, there are approximately 1500 children on long term ventilation. Most of these children have complex, life-limiting conditions and require high-cost and complex community care. Transition to adult care is defined in the Paediatric Long Term Ventilation Service Specification (E07/S/c); transition may start from 13 years with complete transfer to adult services usually by the age of 19 years.
- Due to the complexity of care needs (often multidisciplinary in nature), all children managed within the paediatric long term ventilation service should transition to the adult complex home ventilation service.

Summary – what is complex home ventilation?

The grid below aims to quantify the relative complexity of care required in specific situations. RED indicates a highest-level of complexity, with reducing levels of complexity indicated by AMBER and then GREEN. It is by no means definitive on an individual level. It aims to provide service commissioners with a framework to show how varying care requirements may be highly complex, or less so;

	Assessment and monitoring	Starting ventilation	Discharge planning	NIV dependence likely	Advanced secretion (Physio)	Other MDT (SALT / dietetics)	Technical and clinical support (24/7)	Need for complex equipment or support	Other specialties (Gastro, Cardiology Urology etc)
Invasive ventilation via tracheostomy									
DMD and other dystrophies									
Transition from Paediatric Services									
Motor Neurone Disease									
Myotonic Dystrophy									
Other Neuromuscular									
COPD (post-acute AECOPD)									
COPD (chronic hypercapnia)									
COPD-OSA									
Chest-wall disease (Scoliosis)									
Obesity-hypoventilation (acute transfer)									
Obesity-hypoventilation (stable set-up)									
Complex Sleep Apnoea									

Whilst of importance, identifying areas of less complexity does not mean that providing safe and effective care requires limited expertise;

- Despite service expansion, patients treated with home NIV remain a small population (similar to the total number of adult patients with Cystic Fibrosis, for example).
- Significant expertise is required to assess and manage any patient who may require long-term support in the setting of chronic ventilatory failure.
- The current use of acute NIV provides evidence for the need for a cautious approach towards home NIV expansion. At present, acute NIV is delivered in almost all acute NHS Hospital trusts. Poor outcomes have been demonstrated in successive national audits, with mortality rates significantly higher than those that would be expected (>30% v >10%).
- Reasons for poor outcomes in patients treated with acute NIV include inadequate levels of training, staffing, and patient selection.
- Commissioners should use the specification to ensure appropriate integration of all home ventilation services within their regions so that there is equitable network access to the best possible care for all patients who require it.
- It is also recommended that commissioners apply the same outcome measures to **all** providers of home ventilation. The need to characterise clear network links with a regional complex ventilation service should be considered to be a requirement when commissioning **any** home ventilation service.

References

1. Sullivan CE et al. Reversal of obstructive sleep apnoea by continuous positive airway pressure applied through the nares. Lancet. 1981;1(8225):862-865.
2. Lloyd-Owen SJ et al. Patterns of home mechanical ventilation use in Europe: results from the Eurovent survey. Eur Respir J. 2005;25(6):1025-1031.
3. Mandal, S.; Suh, E.; Davies, M et al. Provision of home mechanical ventilation and sleep services for England survey. Thorax 2013;880-881.
4. Buyse B, Meersseman W, Demedts M. Treatment of chronic respiratory failure in kyphoscoliosis: oxygen or ventilation? Eur Respir J. 2003 Sep;22(3):525-8.
5. Simonds AK et al. Impact of nasal ventilation on survival in hypercapnic Duchenne muscular dystrophy. Thorax. 1998;53(11):949-952.
6. Annane D et al. Nocturnal mechanical ventilation for chronic hypoventilation in patients with neuromuscular and chest wall disorders. Cochrane Database Syst Rev. 2007.
7. Gustafson T et al. Survival of patients with kyphoscoliosis receiving mechanical ventilation or oxygen at home. Chest. 2006;130(6):1828-1833.
8. Bourke SC, Tomlinson M, Williams TL, Bullock RE, Shaw PJ, Gibson GJ. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. Lancet Neurol. 2006;5(2):140-7.

9. Ishikawa Y, Miura T, Ishikawa Y et al. Duchenne muscular dystrophy: survival by cardio-respiratory interventions. Neuromuscul Disord. 2011;21(1):47-51.

10. Kolodziej MA et al. Systematic review of noninvasive positive pressure ventilation in severe stable COPD. Eur Respir J. 2007;30(2):293-306.

11. McEvoy RD et al. Australian trial of non-invasive Ventilation in Chronic Airflow Limitation (AVCAL) Study Group. Nocturnal non-invasive nasal ventilation in stable hypercapnic COPD: a randomised controlled trial. Thorax. 2009;64(7):561-566.

12. Köhnlein T, Windisch W, Köhler D et al. Non-invasive positive pressure ventilation for the treatment of severe stable chronic obstructive pulmonary disease: a prospective, multicentre, randomised, controlled clinical trial. Lancet Respir Med. 2014;2(9):698-705.

13. Windisch W et al. High-intensity non-invasive positive pressure ventilation for stable hypercapnic COPD. Int J Med Sci. 2009;6(2):72-76.

14. Murphy PB, Hart N. Trials of home mechanical ventilation in COPD: what have we learnt? Thorax. 2014 Sep;69(9):787-8.

15. Owens RL et al. Sleep-disordered breathing and COPD: the overlap syndrome. Respir Care. 2010;55(10):1333-1344.

16. Borel JC, Tamisier R, Gonzalez-Bermejo J et al. Noninvasive ventilation in mild obesity hypoventilation syndrome: a randomized controlled trial. Chest. 2012 Mar;141(3):692-702.