British Thoracic Society

Quality Standards

for the Respiratory Management of Children with Neuromuscular Weakness

JANUARY 2014 ISSN 2040-2023

BRITISH THORACIC SOCIETY REPORTS
VOL. 6 NO. 1 2014

BTS Quality Standards for the Respiratory Management of Children with Neuromuscular Weakness should be read alongside the BTS Guideline for the Respiratory Management of Children with Neuromuscular Weakness 2012 (www.brit-thoracic.org.uk)

British Thoracic Society Reports, Vol 6, No 1, 2014

ISSN 2040-2023

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Quality Standards for the Respiratory Management of Children with Neuromuscular Weakness

The British Thoracic Society has been at the forefront of the production of Guidelines for best clinical practice in respiratory medicine since the Society was established over 25 years ago. In recent years, the methodology for the production of evidence-based Guidelines has evolved considerably and a manual setting out the details policy for the production of BTS Guidelines was approved in July 2010.

A statement on quality standards based on each BTS Guideline is a key part of the range of supporting materials that the Society produces to assist in the dissemination and implementation of a Guideline's recommendations.

A quality standard is a set of specific, concise statements that:

- act as markers of high-quality, cost-effective patient care across a pathway or clinical area, covering treatment or prevention.
- are derived from the best available evidence .

NICE Quality Standards were used as a model for the development of BTS Quality Standards and further information on the NICE Quality Standards process is available here: http://guidance.nice.org.uk/qualitystandards/qualitystandards.jsp

This document contains Quality Standards to be used in secondary and tertiary care for the respiratory management of children with neuromuscular weakness (NMW), including those with:

- Congenital muscular dystrophies
- Spinal muscular atrophy
- Congenital myopathies
- Myotonic dystrophy
- Mitochondrial myopathy
- Congenital myasthenic syndromes
- Charcot Marie Tooth disease
- Pompe disease

The Quality Standards do not apply to children with cerebral palsy, myasthenia gravis or Guillain-Barre syndrome. The Quality Standards apply to children seen in paediatric units/ under the care of paediatric physicians and the upper age limit may vary according to the unit. The Quality Standards are based on the BTS Guideline published in 2012 (http://www.brit-thoracic.org.uk). The evidence base in this field is weak and the majority of guideline recommendations and Quality Statements are based on observational studies and expert consensus.

Acute respiratory failure associated with respiratory infection is the most frequent reason for unplanned hospital admission in children with NMW, and chronic respiratory failure is a frequent cause of death. With appropriate intervention, the incidence of unplanned hospital admission can be reduced and life expectancy can be improved. The BTS Guideline on the Respiratory Management of Children with Neuromuscular Weakness, published in 2012, summarises the available evidence in this field and provides recommendations to aid healthcare professionals in delivering good quality patient care.

The purpose of the Quality Standards document is to provide commissioners, planners and patients with a guide to standards of care which should be met for all children with NMW, together with measurable markers of good practice.

BTS quality standards are intended for:

- Health care professionals to allow decisions to be made about care based on the latest evidence and best practice.
- Families and carers of children with neuromuscular weakness and older children with neuromuscular weakness, to enable understanding of what services they should expect from their health and social care provider.
- Service providers to be able to quickly and easily examine the clinical performance of their organisation and assess the standards of care they provide.
- Commissioners so that they can be confident that the services they are purchasing are high quality and cost effective.

Method of Working

A Quality Standards Working Group was convened on 13th February 2013, with the following membership:

Jeremy Hull (chair)	Consultant in paediatric respiratory medicine Children's Hospital, Oxford
Michelle Chatwin	Consultant Physiotherapist in Respiratory Support Royal Brompton Hospital, London
Jayne Gallagher	Clinical nurse specialist Children's Hospital, Oxford
Neil Gibson	Consultant in paediatric respiratory medicine Royal Hospital for Sick Children, Glasgow
Jill Gordon	Consultant community paediatrician Suffolk Primary Health Care Trust
Imelda Hughes	Consultant paediatric neurologist Royal Manchester Children's Hospital
Julian Legg	Consultant respiratory paediatrician Southampton General Hospital
Renee McCulloch	Consultant in paediatric palliativemedicine Great Ormond Street Hospital for Children, London
Rob Ross Russell	Consultant in paediatric respiratory medicine and paediatric intensive care Cambridge University Hospitals
Anita Simonds	Consultant in respiratory medicine Royal Brompton and Harefield Hospital, London
Jo Ilott	Lay member and parent

Members of the Quality Standards Group submitted Declaration of Interest forms in line with the BTS Policy and copies of forms are available on request from BTS Head Office.

The draft document was considered in detail by the BTS Standards of Care Committee on 14th March 2013. Following revision, the document was considered by both the BTS Professional and Organisational Standards Committee and the Public Liaison Committee in April – July 2013.

The document was made available on the BTS website for public consultation for the period from 4 July – 19 August 2013.

Following further revision the document was submitted for approval to the BTS Standards of Care Committee on 13 November 2013.

The Quality Standards document will be reviewed in 2018 or following the publication of a revised Guideline whichever is the sooner.

Each Quality Standard includes the following:

- A **quality statement** which describes an important marker of high-quality, cost-effective care for this condition.
- **Quality measures** which aim to improve the structure, process and outcomes of health care.

The quality measures are not intended to be new sets of targets or mandatory indicators for performance management which need to be collected. The quality measures are specified in the form of a numerator and a denominator which define a proportion (numerator/denominator). It is assumed that the numerator is a subset of the denominator population. The suggested numerator and denominator are provided to allow health care professionals and service providers to examine their clinical performance in relation to each quality standard. It is recognised that no national quality indicators will be available for this condition, and institutions will need to agree locally what information is required for the denominator to be used in each case, and what the expected level of achievement should be, given local circumstances. A brief description about the quality standard in relation to each audience is given.

The BTS Guideline for the management of children with neuromuscular weakness (2012) is the main reference for the Quality Standards. There is no specific order of priority associated with the list of quality standards.

Summary of Quality Statements

A specialist respiratory physiotherapist for the purposes of these quality statements is defined as: a physiotherapist who has greater than three years experience in respiratory physiotherapy, including experience of the respiratory management of children with neuromuscular weakness, and in particular is familiar with secretion clearance methods in this patient group, including the use of non-invasive ventilation, augmented cough techniques and mechanical insufflation-exsufflation devices.

Quality Statement

Quality statement	Respiratory assessment of children with NMW to take place at each planned medical clinic visit.
Quality measure	**Structure:** Evidence of local arrangements to ensure that respiratory assessments are recorded at each outpatient visit. **Process:** Proportion of children with NMW who have a respiratory assessment at each planned outpatient visit. Numerator – the number of planned medical clinic visits for children with NMW where there was a recorded respiratory assessment. Denominator – the total number of planned medical clinic visits for children with NMW.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to undertake and record the respiratory assessments and that these reports are available to health care professionals when required. **Healthcare professionals** ensure respiratory assessments are undertaken and recorded at each outpatient visit. **Commissioners** ensure that sufficient facilities, staff and equipment are available to allow respiratory assessments to take place in the outpatient setting. **Children with NMW** have a respiratory assessment at each planned medical clinic visit.
Relevant existing indicators	None identified.
Other possible national data sources	None identified
Source references	BTS Guideline for the Respiratory Management of Children with NMW 2012.
Rationale	Acute respiratory failure associated with respiratory infection is the most frequent reason for unplanned hospital admission and chronic respiratory failure is a frequent cause of death in children with NMW. Simple clinical assessment based on history taking, examination, and where appropriate tests of respiratory muscle strength and cough effectiveness is essential to identify children at risk of respiratory complications. These children may need more detailed evaluation and subsequent intervention where appropriate. An example of a clinical assessment tool that could be used for making respiratory assessments is given in Appendix 1.

Quality Statement

Quality statement	Children with NMW who have an unplanned hospital admission for a respiratory illness (referred to below as selected children) to be assessed by a specialist respiratory physiotherapist and where clinically appropriate, the child and their carers to be shown how to carry out effective airway clearance, and provided with the necessary equipment for this purpose. Clinical services that care for children with NMW should review all such admissions on an annual basis.
Quality measure	**Structure:** Evidence of local arrangements to ensure that all selected children with NMW are reviewed by a specialist respiratory physiotherapist. **Process:** Proportion of selected children with NMW who are reviewed by a specialist respiratory physiotherapist to assess and manage effective airway clearance Numerator – the number of selected children with NMW who have been reviewed by a specialist respiratory physiotherapist. Denominator – the total number of selected children with NMW.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to allow selected children with NMW to see a specialist respiratory physiotherapist; also to ensure that necessary equipment including lung volume recruitment bags, and mechanical insufflation/exsufflation devices are available if required. **Healthcare professionals** ensure that all appropriate children are referred to a specialist respiratory physiotherapist to be taught airway clearance and augmented cough techniques. **Commissioners** ensure that access to specialist respiratory services is available, and that necessary equipment including lung volume recruitment bags, and mechanical insufflation/exsufflation devices are available if required. **Children with NMW** and their carers, where appropriate, are taught airway clearance and assisted cough techniques.
Relevant existing indicators	None identified.
Other possible national data sources	None identified
Source references	BTS Guideline for the Respiratory Management of Children with NMW, 2012
Rationale	• Difficulty with airway clearance, particularly during respiratory tract infection, is the most likely reason for episodes of acute respiratory failure and unplanned hospital admission. • The definition of a specialist respiratory physiotherapist is given on page 4. • Airway clearance methods may include augmented cough techniques. Necessary equipment may include lung volume recruitment bags and/or mechanical insufflation-exsufflation devices.

Quality Statement

Quality statement	Children with NMW who have an ineffective cough or a history of recurrent respiratory exacerbations (referred to below as selected children) to have an individualised written management plan to deal with respiratory exacerbations.
Quality measure	**Structure:** Evidence of local arrangements to ensure that all selected children with NMW are provided with an individualised written respiratory exacerbation management plan. **Process:** Proportion of selected children with NMW who have an individualised written respiratory exacerbation management plan. Numerator – the number of selected children with NMW who have an individualised written respiratory exacerbation management plan. Denominator – the total number of selected children with NMW.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to provide selected children with NMW with an individualised written respiratory exacerbation management plan. **Healthcare professionals** ensure that all appropriate children have an individualised written respiratory exacerbation management plan. **Commissioners** ensure that services record the provision of an individualised written respiratory exacerbation management plan. **Children with NMW** and their carers, where appropriate, to be involved in developing and using an individualised written respiratory exacerbation management plan.
Relevant existing indicators	None identified
Other possible national data sources	None identified
Source references	BTS Guideline for Respiratory Management of Children with NMW, 2012.
Rationale	The development of an individualised written respiratory exacerbation management plan is intended to help children with NMW and their carers to recognise, respond to and reduce the severity of respiratory exacerbations. An example of a respiratory management plan is provided in Appendix 2. The plans should include a description of the care pathway a child should follow during a respiratory exacerbation, and the contact details of the health professionals involved. Up to date copies of the plan should be held by the patient (and/or carer) and their clinical teams in primary, secondary and tertiary care.

Quality Statement

Quality statement	Children with NMW to have access to inpatient services which include specialist respiratory physiotherapy and expertise in the use of non-invasive ventilation.
Quality measure	**Structure:** Evidence of local arrangements to ensure that all children with NMW have access to inpatient services which include specialist respiratory physiotherapy and expertise in the use of non-invasive ventilation. **Process:** Proportion of clinical services caring for children with NMW that have established clinical pathways to inpatient services which include specialist respiratory physiotherapy and expertise in the use of non-invasive ventilation. Numerator – the number of clinical services which care for children with NMW that are aware of the clinical pathway that will provide prompt access to specialist respiratory physiotherapy and expertise in the use of non-invasive ventilation. Denominator – the total number of clinical services that care for children with NMW.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to allow all children with NMW to have access to inpatient services which include specialist respiratory physiotherapy and expertise in the use of non-invasive ventilation. **Healthcare professionals** ensure that all children with NMW, when necessary, are referred to inpatient services which include specialist respiratory physiotherapy and expertise in the use of non-invasive ventilation. **Commissioners** ensure that access to inpatient services which include specialist respiratory physiotherapy and expertise in the use of non-invasive ventilation is available. **Children with NMW** and their carers, when necessary, have access to inpatient services which include specialist respiratory physiotherapy and expertise in the use of non-invasive ventilation.
Relevant existing indicators	None identified
Other possible national data sources	None identified
Source references	BTS Guideline for Respiratory Management of Children with NMW, 2012.
Rationale	Specialist respiratory physiotherapy and the use of non-invasive ventilation are both critical in the management of episodes of acute respiratory failure that can occur during respiratory exacerbations in children with NMW. Not all clinical units who care for children with NMW will have specialist physiotherapists or medical and nursing staff familiar with the use of non-invasive ventilation. All clinical units should be aware of the need for these services during respiratory exacerbations, and the referral pathways that ensure rapid access when required. The definition of a specialist respiratory physiotherapist is given on page 4.

Quality Statement

Quality statement	Assessment for sleep-disordered breathing to take place no less than annually for all children with NMW who have symptoms of obstructive sleep apnoea or hypoventilation, children with clinically apparent diaphragmatic weakness and children with rigid spine syndromes (referred to below as selected children).
Quality measure	**Structure:** Evidence of local arrangements to ensure that selected children with NMW have access to services able to carry out assessments for sleep-disordered breathing. **Process:** Proportion of selected children with NMW who have at least annual assessments for sleep-disordered breathing. Numerator – the number of selected children with NMW who have had at least an annual assessment for sleep-disordered breathing. Denominator – the total number of selected children with NMW.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to allow selected children with NMW to have access to services able to carry out at least annual assessments for sleep-disordered breathing. **Healthcare professionals** ensure that all selected children with NMW have at least annual assessments for sleep-disordered breathing. **Commissioners** ensure access to services with the necessary expertise, equipment and capacity to carry out at least annual assessments for sleep-disordered breathing in selected children with NMW. **Children with NMW,** where appropriate, have at least annual assessments for sleep-disordered breathing.
Relevant existing indicators	None identified
Other possible national data sources	None identified
Source references	BTS Guideline for Respiratory Management of Children with NMW, 2012.
Rationale	Early identification of sleep-disordered breathing is necessary to prevent morbidity associated with obstructive sleep apnoea and sleep-associated hypoventilation. Please refer to the BTS Guideline for Respiratory Management of Children with NMW, 2012 for further details of assessments for sleep-disordered breathing.

Quality Statement

Quality statement	Children with NMW using mechanical respiratory support (referred to below as selected children) to be assessed regularly to ensure the continued effectiveness of the respiratory support. The frequency of review will vary according to clinical circumstances but will not be less often then every 12 months.
Quality measure	**Structure:** Evidence of local arrangements to ensure that selected children with NMW have access to services able to carry out assessments of the effectiveness of their respiratory support. **Process:** Proportion of selected children with NMW who have at least annual assessments of the effectiveness of their respiratory support. Numerator – the number of selected children with NMW who have had at least an annual assessment of the effectiveness of their respiratory support. Denominator – the total number of selected children.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to allow selected children with NMW to have access to services able to carry out at least annual assessments of the effectiveness of the respiratory support. **Healthcare professionals** ensure that all selected children with NMW have at least annual assessments of the effectiveness of the respiratory support. **Commissioners** ensure access to services with the necessary expertise, equipment and capacity to carry out at least annual assessments of the effectiveness of the respiratory support in selected children with NMW. **Children with NMW**, where appropriate, have at least annual assessments of the effectiveness of their respiratory support.
Relevant existing indicators	None identified
Other possible national data sources	None identified
Source references	BTS Guideline for Respiratory Management of Children with NMW, 2012.
Rationale	Effective mechanical respiratory support is necessary to prevent symptoms and to reduce the frequency of unplanned hospital admissions. The amount and type of respiratory support each child requires will be affected by growth and disease progression.

Quality Statement

Quality statement	Surgery in children with NMW to be undertaken by paediatric surgeons and paediatric anaesthetists and in units with specialist physiotherapists and facilities for paediatric high dependency care after surgery and experience in the use of non-invasive ventilation.
Quality measure	**Structure:** Evidence of local arrangements to ensure that all children with NMW requiring surgery have access to services which include paediatric surgeons, paediatric anaesthetists, specialist physiotherapy services and facilities for paediatric high dependency care and non-invasive ventilation. **Process:** Proportion of children with NMW who require surgery who have access to services which include paediatric surgeons, paediatric anaesthetists, specialist physiotherapy services and facilities for paediatric high dependency care and non-invasive ventilation. Numerator – the number of children with NMW who underwent surgery and had access to inpatient care which include paediatric surgeons, paediatric anaesthetists, specialist physiotherapy services and facilities for paediatric high dependency and non-invasive ventilation. Denominator – the total number children with NMW who underwent surgery.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to allow children with NMW requiring surgery to have access to services which include paediatric surgeons, paediatric anaesthetists, specialist physiotherapy services and facilities for paediatric high dependency care and non-invasive ventilation. **Healthcare professionals** ensure that all children with NMW who require surgery are referred to inpatient services which include paediatric surgeons, paediatric anaesthetists, specialist physiotherapy services and facilities for paediatric high dependency care and non-invasive ventilation. **Commissioners** ensure access to services which include paediatric surgeons, paediatric anaesthetists, specialist physiotherapy services and facilities for paediatric high dependency care and non-invasive ventilation. **Children with NMW** who require surgery have access to services which include paediatric surgeons, paediatric anaesthetists, specialist physiotherapy services and facilities for paediatric high dependency care and non-invasive ventilation.
Relevant existing indicators	None identified
Other possible national data sources	None identified
Source references	BTS Guideline for Respiratory Management of Children with NMW, 2012.
Rationale	Children with NMW are at increased risk of morbidity associated with surgery. This can be minimised by undertaking surgery in paediatric centres with the necessary expertise and facilities, which should include a paediatric high dependency unit. It should be recognised that any major surgical procedure, particularly scoliosis surgery, and any surgery on children with profound weakness or bulbar dysfunction, or children already using non-invasive support, should take place in a paediatric unit with intensive care facilities for post-operative care. The definition of a specialist respiratory physiotherapist is given on page 4.

Quality Statement

Quality statement	Children with NMW who require scoliosis surgery to have a pre-operative respiratory assessment by a multidisciplinary team including paediatric surgeons, paediatric anaesthetists, respiratory paediatricians and specialist physiotherapists.
Quality measure	**Structure**: Evidence of local arrangements to ensure that all children with NMW requiring scoliosis surgery have a pre-operative respiratory assessment by a multidisciplinary team. **Process:** Proportion of children with NMW who require scoliosis surgery who have a pre-operative respiratory assessment by a multidisciplinary team. Numerator – the number of children with NMW who underwent scoliosis surgery who had a pre-operative respiratory assessment by a multidisciplinary team. Denominator – the total number children with NMW who underwent scoliosis surgery.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to allow children with NMW requiring surgery to have access to services which include a pre-operative respiratory assessment by a multidisciplinary team. **Healthcare professionals** ensure that all children with NMW who require surgery are referred to services which include a pre-operative respiratory assessment by a multidisciplinary team. **Commissioners** ensure access to surgical services which include a pre-operative respiratory assessment by a multidisciplinary team. **Children with NMW** who require surgery to have a pre-operative respiratory assessment by a multidisciplinary team.
Relevant existing indicators	None identified.
Other possible national data sources	None identified.
Source references	BTS Guideline for Respiratory Management of Children with NMW, 2012.
Rationale	Children with NMW have an increased risk of respiratory morbidity associated with scoliosis surgery. Effective management of respiratory problems associated with scoliosis surgery requires a pre-operative assessment by a multidisciplinary team including paediatric surgeons, paediatric anaesthetists, respiratory paediatricians and specialist physiotherapists and should include a nutritional assessment. The definition of a specialist respiratory physiotherapist is given on page 4.

Quality Statement

Quality statement	Children and young people with NMW who are undergoing regular assessment for sleep disordered breathing or who are using respiratory mechanical support (referred to below as selected children and young people) to have a planned process of transition from paediatric to adult respiratory services.
Quality measure	**Structure:** Evidence of local arrangements to ensure that a process is in place for selected children with NMW to have a planned transition from paediatric to adult respiratory services. **Process:** Proportion of selected children with NMW moving from paediatric to adult respiratory services that have a planned transition process. Numerator – the number of selected children with NMW moving from paediatric to adult respiratory services who have a planned transition process. Denominator – the total number of selected children with NMW moving from paediatric to adult respiratory services.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to allow selected children and young people with NMW moving from paediatric to adult respiratory services to have a planned coordinated transition process with age appropriate care throughout which acknowledges adolescent developmental changes. **Healthcare professionals** ensure that all selected children and young people with NMW moving from paediatric to adult respiratory services have a planned, coordinated transition process with age appropriate care throughout which acknowledges adolescent developmental changes. **Commissioners** ensure access to young person friendly services which include a planned transition process between paediatric and adult respiratory services. **Selected children** with NMW who are moving from paediatric to adult services to have a planned transition process for their respiratory care.
Relevant existing indicators	None identified
Other possible national data sources	None identified
Source references	BTS Guideline for Respiratory Management of Children with NMW, 2012.
Rationale	Moving from paediatric to adult services is difficult for young people with NMW and for their carers and may coincide with disease progression and increased respiratory morbidity. A planned process of transition reduces anxiety in the patients and carers and ensures continued high quality care. Please refer to the BTS Guideline for respiratory management of children with NMW, 2012 for further details of the transition process.

Quality Statement

Quality statement	All children with life-shortening NMW (referred to below as selected children) to have access to paediatric palliative care services.
Quality measure	**Structure:** Evidence of local arrangements to ensure that selected children with NMW have access to paediatric palliative care services. **Process:** Proportion of clinical services caring for selected children with NMW which have access to paediatric palliative care services. Numerator – the number of clinical services caring for selected children with NMW which have access to paediatric palliative care services. Denominator – the total number of clinical services caring for selected children with NMW.
Description of what the quality statement means for each audience	**Service providers** ensure systems are in place to allow selected children with NMW to have access to paediatric palliative care services. **Healthcare professionals** ensure that all appropriate selected children with NMW are referred to a paediatric palliative care service. **Commissioners** ensure access to paediatric palliative care services **Children with NMW** who have a life-shortening condition, and their carers, are able to benefit from the services that a paediatric palliative care service can offer.
Relevant existing indicators	None identified
Other possible national data sources	None identified
Source references	BTS Guideline for Respiratory Management of Children with NMW, 2012.
Rationale	Use of paediatric palliative care services, particularly children's hospices and children's community nursing services, can provide valuable support to children with life-shortening NMW, including respite care and expertise in symptom control.

REFERENCES

1. BTS Guideline for the Respiratory Management of Children with Neuromuscular Weakness, 2012, Thorax Vol 67, Supplement 1

http://www.brit-thoracic.org.uk

Appendix 1

Example of a respiratory assessment tool

The purpose of the respiratory assessment is to determine whether a child with neuromuscular weakness child has any of the following:

- night time hypoventilation
- obstructive sleep apnoea
- an ineffective cough
- a risk of aspiration

Night time hypoventilation can cause morning nausea, headache and a poor appetite. It can also lead to poor sleep quality and daytime sleepiness and lethargy. Snoring is the cardinal feature of obstructive sleep apnoea. An ineffective cough sounds weak and quiet. Ineffective coughing will result in prolonged respiratory illnesses, which will usually start as a simple cold. More severe episodes will result in hospital admissions. Children who aspirate tend also to eat slowly, and may cough or splutter on eating or drinking, although this may not always be apparent. Respiratory effort may increase after eating if aspiration has occurred. Unexplained and persistent respiratory symptoms should raise the possibility of aspiration.

1=never; 2=occasionally; 3=sometimes; 4=often; 5=every day/night

Morning nausea:	1	2	3	4	5
Morning headache:	1	2	3	4	5
Poor appetite:	1	2	3	4	5
Poor sleep quality:	1	2	3	4	5
Snoring:	1	2	3	4	5
Cough or spluttering on eating or drinking:	1	2	3	4	5
Breathing noisy or more difficult after eating:	1	2	3	4	5

Respiratory exacerbations	
Number of chesty episodes per year:	None / 1-2 / 2-4 / 4-6 / >6
Usual duration of chesty episodes:	<5 days / 5-10 days / 10-20 days / > 20 days
Number of hospital admissions for chesty episode in last 12 months:	None / 1-2 / 2-4 / 4-6 / >6
Longest duration of any hospital admission for chesty episode:	<5 days / 5-10 days / 10-20 days / > 20 days
Was supplemental oxygen used for any chesty episode treated in hospital:	Yes / No

Airway clearance methods	
None used	
Manual assisted cough:	just with chesty episodes / most days / every day
Lung recruitment bag:	just with chesty episodes / most days / every day
Mechanical insufflation-exsufflation:	just with chesty episodes / most days / every day

For children using non-invasive ventilation (NIV)	
How many nights per week is the NIV used:	1 / 2 / 3 / 4 / 5 / 6 / 7
How much of each night is the NIV used:	<2 hours / 2-4 hours / 4-6 hours / 6-8 hours / > 8hours
What mask is being used:	nasal / full face
Any mask problems:	sore skin / facial hypoplasia / mask leak / other:

This template may be adapted for local use.

Appendix 2

Example of a management plan for respiratory exacerbations

Name of child:
Date plan started: Review date:
An Advance Care Plan exists for this child: yes/no. If yes, please make sure it corresponds with the advice given here.

When well: *text needs to be specific for each child – statements below are suggestions only*
Description of respiratory status and ability to clear secretions when well: Usual medication: Routine airway clearance (if any):
When less well: *text needs to be specific for each child – statements below are suggestions only*
Early warning signs may include: lethargy, worsening weakness, cough, increased breathing effort **Assessment** may include feeling the chest for rattles; whether the cough sounds effective; measuring oxygen saturation. **Action** may include: starting airway clearance methods, or increased frequency of existing airway clearance methods. These will need to be specified. If home pulse oximetry is used as a guide, parameters will need to be given – for example – airway clearance methods will need to be started/increased when oxygen saturations fall below 95% and continued until they improve above 95%. Other actions: eg starting oral antibiotics.
Failure to improve: *text needs to be specific for each child – statements below are suggestions onlyww*
If symptoms persist for more than (specify duration), medical assistance should be sought (see contact details) If home pulse oximetry is used: if oxygen saturations are not improved (above 94%) with airway clearance methods, medical assistance should be sought (see contact details)
Emergency: *text needs to be specific for each child – statements below are suggestions only*
If increased breathing effort persists, or there is a poor colour, or if oximetry is used it is very low (eg <90%) and does not improve with airway clearance, medical assistance should be sought immediately – either by calling an ambulance, or attendance at your nearest emergency department (specify which).

Hospital care: *text needs to be specific for each child – statements below are suggestions only*
Note for medical personnel: If this child needs hospital admission for a respiratory exacerbation, urgent assessment by a respiratory physiotherapist will be required. Frequent assisted airway clearance methods are likely to be needed. These are especially important if the child has oxygen saturations less than 95% in room air. Simply starting antibiotics and giving oxygen will not lead to improvement. For further advice about the respiratory management of this patient, please contact either the respiratory consultant, the respiratory nurse or the physiotherapist listed at the end of this document.

Your child's information
Name:
Hospital:
Hospital unit number:
NHS number:
School/Nursery:
Diagnosis:

	Name	Contact Number
Neurology consultant:		
Respiratory consultant:		
Community consultant:		
Respiratory nurse:		
General practitioner		
Respiratory physiotherapist:		

Who to contact for advice when your child is unwell:
in working hours:
outside working hours:

This template may be adapted for local use.

NOTES

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London WC1N 2PL
Registered as a Charity in England and Wales with number 285174
and registered in Scotland with number SC041209
Company Registration No. 1645201
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