February 26, 2013

PROVIDER

Pediatric Specialties

Attn: Dr.

ADDRESS

CITY, STATE ZIP

**Re: CoughAssist T70 - Letter of Medical Necessity**

*Patient: NAME LASTNAME*

*MRN/ID#:*

*DOB:*

*Sex:*

*Diagnosis: Spinal Muscular Atrophy, Type*

Dear PROVIDER:

On behalf of NAME LASTNAME, we request the Phillips Respironics CoughAssist **T70** cough stimulation device, including the following:

1. Detachable battery pack
2. Detachable battery charger
3. Oximetry interface kit
4. DC auto adapter
5. Roll stand
6. Foot pedal

Notably, NAME currently uses the CoughAssist CA-3000 device. We request switching from the CA-3000 model to the T70 model. The CoughAssist **T70** is medically necessary, as detailed herein.

**Medical Background**

NAME LASTNAME was born on DATE. She was diagnosed with Spinal Muscular Atrophy (SMA) Type 1 on or about DATE, at an age of approximately AGE months old. SMA Type 1 is a severe genetic neuromuscular disorder, described more fully below.

 Due to SMA, NAME requires 24 hour monitoring and care. She needs intensive hour-long respiratory treatments at least three times daily and additionally, as may be needed, to maintain basic pulmonary health. These treatments include aggressive chest percussion and mechanical cough assistance. NAME is dependent on noninvasive bi-level positive airway pressure (BiPAP) ventilation for respiratory support. NAME requires frequent suctioning of mouth and nose throughout the day and also requires feeding exclusively via gastronomy tube due to her lack of swallow function and protective airway reflexes.

As a consequence of SMA, NAME is especially susceptible to RSV and other severe respiratory infection. If she contracts RSV or other respiratory complication, she is likely to require extensive hospitalization.

**Spinal Muscular Atrophy, Type 1, Generally**

SMA is a recessively inherited neuromuscular disorder characterized by degeneration of spinal cord motor neurons, resulting in progressive muscular atrophy and weakness. The clinical spectrum of SMA ranges from early infant death to normal adult life with only mild weakness. These patients often require comprehensive medical care involving multiple disciplines. There is, however, no published practice standard for the care of these patients. (Wang, et al., Consensus Statement for Standard of Care in Spinal Muscular Atrophy, *J Child Neurol.* 2007; 22(8):1027.)

The weakness is usually symmetrical and more proximal than distal. Sensation is preserved. Tendon reflexes are absent or diminished. The severity of the weakness generally correlates with the age of onset. The most severe type presents in infancy. The infant may appear normal at birth. Weakness evolves in the first few months of life. (*Id.* at 1029.)

Individuals manifesting different levels of weakness due to SMA have been divided into four groups defined by functional ability. SMA Type 1, also known as Werding-Hoffmann Disease, is a severe form SMA where the highest function attained is never being able to sit independently (“nonsitters”). The highest function attained for Type 2 is the ability to sit up but not stand (“sitters”). The highest function attained for Type 3 is the ability to stand and walk, but that ability is lost over time. The highest function attained for Type 4 is the ability to stand and walk where that ability is not lost. Types 3 and 4 are “walkers.” (*Id.* at 1029-1030.)

Children with SMA Type 1 have impaired head control, with a weak cry and cough. Swallowing, feeding, and handling of oral secretions are affected before one year of age. Weakness and hypotonia in the limbs and trunks are eventually accompanied by intercostal muscle weakness. Infants exhibit chest wall collapse. (Wang at 1030.)

The natural age of death of children with SMA Type 1 is *less than 2 years*. Early morbidity and mortality are most commonly associated with bulbar dysfunction and *pulmonary complications*. (*Id.* at 1033.)

The key respiratory problems in SMA are as follows: (1) impaired cough, resulting in poor clearance of lower airway secretions; (2) hypoventilation during sleep; (3) chest wall and lung underdevelopment; and (4) recurrent infections that exacerbate muscle weakness. Pulmonary disease is the major cause of morbidity and mortality in SMA Type 1. Individuals progress to daytime respiratory failure via a sequence of *recurrent chest infections*, among other things. (*Id.* at 1033.) Noninvasive ventilation as a means of respiratory support is generally advised. (*Id.* at 1036.)

Intercostal muscle weakness is responsible for triangular chest deformity, with falling ribs, and results in recurrent atelectasis and bronchopulmonary infections. Respiratory episodes of pulmonary congestion, aspiration pneumonia, and atelectasis are frequent. False passages of saliva with swallowing disturbances increase pulmonary congestion and the risk of aspiration pneumonia and respiratory distress. (Ioos, C., *et al*., Respiratory Capacity Course in Patients with Infantile Spinal Muscular Atrophy, *Chest*, 2004;126:831-837.)

Patients with SMA Type 1 show a marked progressive and regular decrease in lung capacity. SMA is a progressive disease with a progressive decline in lung capacity. Studies show severely impaired respiratory function. Lung capacity diminishes continuously. (*Ibid*.)

The risk of pulmonary complication increases as lung capacity decreases. Therapies are essential to limit pulmonary congestion and atelectasis, and to limit the risk of respiratory distress. (Ioos at 837.)

Children with neuromuscular disorders that result in an inability to clear secretions are at risk of more severe infections after viral infection. Increase in secretion volume and thickness can overwhelm a compromised swallowing function and lead to a risk of aspiration pneumonia, atelectasis, and congestion of the upper and lower airways. Further, these infections cause acute deterioration in muscle strength. Children with neuromuscular weakness are at risk for more severe infections and such children at a higher risk factor for severe RSV disease. (Pantich, H., Viral Respiratory Infections in Children with Technology Dependence and Neuromuscular Disorders; *Pediatr Infec Dis J*, 2004:23: S222-227.)

**CoughAssist is Medically Necessary**

CoughAssist is a portable, electrically powered device that uses a blower and valve to alternately apply positive then negative pressure to the patient’s airway in order to assist the patient in clearing retained bronchial secretions.  Air is delivered to the patient via a breathing circuit incorporating a flexible tube, a bacterial filter and either a facemask or mouthpiece.  Based on the Cofflator from O.E.M. Corporation used during the polio epidemics in the 1950s, the CoughAssist is a proven technology that fell into disuse in the early '60s after tracheostomy and suctioning became more popular.  It has regained acceptance as a noninvasive alternative for clearing airway secretions.  By avoiding invasive surgery (tracheotomy) when noninvasive alternatives are viable, both quality of life and mortality rates are improved as documented in *Chest*, *Am J Phys Med Rehabil*, *Arch Phys Med Rehabil*, *Rehabilitation R&D Progress Reports* (Veterans Health Administration), *Distrofia Muscolare*, *Bull N Y Acad Med*, and other medical sources.

Use of the CoughAssist is indicated for the management of retained secretions where the patient is unable to generate a sufficient unassisted cough flow rate to clear secretions (< 160 to 270 L/min).

The adoption of a routine, daily noninvasive protocol (the use of CoughAssist in conjunction with noninvasive positive pressure ventilation) has been shown to increase patient's quality of life, decrease respiratory-related hospitalization from 21 days/year to less than 2 days/year, and prolong life, in many cases by over 50 years. (*Prevention of Pulmonary Morbidity*).

In a child without SMA, bronchial secretions are removed from the lungs via a “productive cough” which moves the secretions up the trachea and out the mouth.  A cough is produced when the diaphragm and chest muscles work to expand the lungs, filling them with air, then rapidly contract to expel the air and secretions (explosive exhalation). However, due to SMA, NAME lacks the muscle strength to create the full inhalation/explosive exhalation cycle required to remove secretions from her lungs.  If these secretions are not removed, bacterial infection (pneumonia) and oxyhemoglobin desaturation (low blood oxygen) can occur, resulting in respiratory failure, hospitalization and death.

 In addition, NAME’s poor bulbar (swallowing) muscle control increases her susceptibility to aspirate saliva, foods or liquids into her lungs.  If the aspirated material is not removed via coughing, bacterial infection (pneumonia) and oxyhemoglobin desaturation (low blood oxygen) can occur, resulting in respiratory failure, hospitalization and death.

 SMA causes NAME to have insufficient cough flow rate to clear her secretions. She has diminished bulbar function resulting in high risk of aspiration of saliva and other respiratory contaminants.

PROVIDER has deemed the CoughAssist to be medically necessary, as evidenced by the fact that PROVIDER has been providing NAME a CoughAssist model CA-3000 since approximately August 2010. Prompt and emergency access to a CoughAssist is medically necessary for removal of bronchial secretions and aspirated matter.

Failure to provide NAME with prompt access to a CoughAssist device will likely result in repeated oxyhemoglobin desaturation, bronchial infection, pulmonary failure, hospitalization, and death. The only viable alternative to the use of a CoughAssist device is invasive surgery to install a tracheostomy tube resulting in hospitalization, higher medical care and equipment costs, lower quality of life, and premature death.

Given the severity of SMA Type 1, NAME bears a substantial risk of coming into contact with at a variety of critical respiratory problems.  Lack of access to a CoughAssist can and will cause NAME more serious infection than in a child who is capable of clearing their secretions without mechanical help.

In addition, providing a Cough Assist to NAME for routine daily use is cost effective I that the device will greatly reduce the chances that NAME will get ill enough to require a lengthy, and expensive, hospitalization. The cost of the device is relatively small compared to the cost of admission to the PICU for respiratory failure.

In sum, CoughAssist is not just “medically necessary.” It is critical. Accordingly, the optimal CoughAssist device is medically necessary.

**CoughAssist T70 is Medically Necessary Relative to the CoughAssist CA-3000**

“What constitutes a minimum standard of care will naturally evolve with available innovations.” (Jacobson, P., Medical Liability and the Culture of Technology; *The Project on Medical Liability in Pennsylvania*, 2004:55.)

The CoughAssist T70 is a fairly new device. It is medically necessary as compared to the CoughAssist CA-3000 NAME currently uses.

Battery & Power

The most critical feature of the T70 over the CA-3000 is the battery. The CA-3000 has no battery. The T70 has a battery option that is medically necessary.

The lack of a battery in the CA-3000 limits its function, and patient safety, to the range of the power cord and the proximity of a power outlet. The CA-3000 limits NAME’s in-home mobility to areas close to power outlets. Power outlets are being used by lamps may force us to have to unplug a light and attempt to use the device in the dark to remove a sudden mucus plug. We will not be able to see her face to determine if she is struggling for air, and we will not be able to get a good alignment of the cough mask on her face if the lights are out.

Further, if we ever experience a power outage, NAME will lose use of the CoughAssist, a critical piece of equipment, until power is restored. As the hurricanes have shown, entire regions of tens of thousands of square miles can lose power for extended periods, stretching days and weeks, due to severe weather conditions. The CA-3000 puts NAME at severe risk that is easily avoided with the T70 and its battery pack.

The T70 battery pack allows 4 respiratory treatments on a single charge. If a power outage occurs, the battery pack will permit us to safely provide for NAME in-home until power is restored or emergency services arrives with an alternative source of power. SMA children are prone to sudden mucus plugs, and being without power is a great risk when critical devices lack batteries.

The T70 battery is removable and interchangeable with the Triology bipap ventilator battery. NAME has the Trilogy 100, and the interchangeable batteries provide added flexibility and security in emergencies.

Additionally, the CA-3000 creates an enormous problem when it comes to ambulance transport in that just about no ambulances carry a CoughAssist device. Further, few ambulances can accommodate a CoughAssist device for two reasons: (1) the CoughAssist CA-3000 requires a pure sine wave power source, which many ambulances lack; and (2) ambulance companies generally do not allow patient equipment to be plugged into the ambulance power system because of liability issues. Thus, with the CA-3000, NAME may be without a CoughAssist when she may need it most: in an ambulance with respiratory failure. The T70 does not require a pure since wave power source. Also, the T70 battery solves this problem in ambulances that do not permit powering patient equipment.

Size & Weight

Another vast difference between the CA-3000 and the T70 is size and weight. The CA-3000 measures 11.5 x 11 x 16.5 inches, for a total volume of nearly 2,100 in2 and a weight of 24 lbs. This model is a large, heavy, unwieldy box with rounded edges, no handles, and uneven weight distribution. The CA-3000 is hard to grip, and just moving it around our home is very difficult.

We must move the device around the home to follow NAME around the home and also to take it with us to NAME’s medical visits at PROVIDER. Moving an awkward device like that around requires both hands and puts it at risk of being dropped and damaged. A damaged CoughAssist device puts NAME in great danger of choking and/or inadequate emergency airway clearance until a replacement CoughAssist unit arrives from the DME provider. Damaged equipment also and puts PROVIDER and the DME provider at risk of added expense to replace damaged equipment as well as added risk that NAME will need to be admitted to the PICU for respiratory failure due to inadequate airway clearance.

Should NAME have a sudden airway blockage or mucus plug, then she is at risk of choking before we can move the CoughAssist to her for emergency airway clearance. If we elect to leave the CoughAssist stationary in-home until needed, then NAME is at risk of choking before we can move her to the device. This is especially problematic in that it renders her routine bath time a high risk endeavor due to wet slippery hands, a wet slippery child, and a lack of prompt CoughAssist access.

Alternatively, the T70 measures only 11.5 x 9.1 x 9.5 inches, for a total of only about 990 in2 and a weight of only 9.4 lbs. The T70 has a handle and can be carried easily and securely with one hand. This makes in-home mobility, bathing, and her routine doctor visits much safer and easier, with minimized risk to the device or to NAME. The T70 also comes with a carrying case for enhanced portability and carrying security.

Size and weight considerations are especially important in that we expect NAME to soon have a power chair for in-home mobility. A power chair offers no space a device the size of the CA-3000, especially given that NAME will need a suction device and ventilator with her on the power chair at all times.

Digital Interface & Programming

The CA-3000 has a small *mechanical* dial interface with pressure markers in increments of 2 cmH2O, mechanical timer knobs with no markings to indicate increments of less than 1 second, and only two settings for inhale flow: low and high. The CA-3000 lacks precision in establishing accurate pressure, timing, and inhale flow.

Alternatively, the T70 has a large *digital* interface with precise digital pressure settings in increments of 1 cmH2O, digital timer settings in increments of 0.1 seconds, and three inhale flow settings: low, medium, and high. Perhaps the most critical machine NAME has should have the most precise settings available to deliver optimal cough support.

Further, the CA-3000 has *loose* knobs to control timing and pressures. If the knobs are inadvertently bumped during a respiratory treatment, a sudden increase in pressure can damage NAME’s lungs. The T70 avoids this problem with digital settings and a screen lock to prevent accidental alteration of settings during a treatment. The T70 enhances NAME’s safety accordingly.

The T70 also allows digital programming of cough sequence in 3 presets. This allows variation of treatments depending on the goal of the treatment at any given time: prophylactic use, mucus plugs, lung volume recruitment, etc.. The CA-3000 has no programming and only allows one kind of treatment.

The T70 displays the peak cough flow, SpO2, heart rate, and tidal volume. Peak cough flow measures cough strength to adjust expiratory pressure to maximize effective coughs. Tidal volume helps determine proper inspiratory pressure needed to deliver deep inhalation. The CA-3000 lacks this capability.

The T70 has an SD Card to save data in an information log. This allows us and NAME’s pulmonologist to evaluate various aspects of the treatments to maximize effectiveness. The CA-3000 has no such capability.

The T70 has an LCD display, allowing us to safely perform cough treatments in its light in the event of darkness from a power outage. The CA-3000 has no lighting whatsoever.

**Conclusion**

NAME’s life depends on the CoughAssist every day. Given its importance and critical role in her life, it is medically necessary to have equipment that can best suit her needs. The CA-3000 lacks capabilities that NAME needs to survive and prosper. The T70 is a much more capable machine, providing increased safety, stability, and maximizing her respiratory treatments and health, especially in emergencies. We request approval of the CoughAssist T70 for NAME, including the following:

1. Detachable battery pack for enhanced safety and in-home mobility
2. Detachable battery charger to recharge the battery as needed
3. Oximetry interface kit to enhance treatment safety and monitoring
4. DC auto adapter to enhance safety when taking NAME to and from medical appointments
5. Roll stand to enhance in-home mobility and to allow movement of both the CoughAssist and Trilogy on the same stand
6. Foot pedal to allow effective treatments while holding the CoughAssist device itself, the cough mask interface, and/or NAME’s head for safety during cough treatments

Thank you for your consideration.

Sincerely,

Mark Storm

**CONTACTS**:

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| J.H. Emerson Company22 Cottage Park AvenueCambridge, Massachusetts 02140-16911-800-252-1414[www.coughassist.com](http://www.coughassist.com)www.jhemerson.com | Respironics, Inc.1501 Ardmore Blvd.Pittsburgh, PA 15221-4401    1-800-638-8208www.respironics.com  |

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