Airway clearance in neuromuscular weakness

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Impaired airway clearance leads to recurrent chest infections and respiratory deterioration in neuromuscular weakness. It is frequently the cause of death. Cough is the major mechanism of airway clearance. Cough has several components, and assessment tools are available to measure the different components of cough. These include measuring peak cough flow, respiratory muscle strength, and inspiratory capacity. Each is useful in assessing the ability to generate an effective cough, and can be used to guide when techniques of assisting airway clearance may be effective for the individual and which are most effective. Techniques to assist airway clearance include augmenting inspiration by air stacking, augmenting expiration by assisting the cough, and augmenting both inspiration and expiration with the mechanical insufflator-exsufflator or by direct suctioning via a tracheostomy. Physiotherapists are invaluable in assisting airway clearance, and in teaching patients and their families how to use these techniques. Use of the mechanical insufflator-exsufflator has gained popularity in recent times, but several simpler, more economical methods are available to assist airway clearance that can be used effectively alone or in combination. This review examines the literature available on the assessment and management of impaired airway clearance in neuromuscular weakness.

In neuromuscular weakness, chest infections cause significant morbidity and are often the eventual cause of death.[1] They generally result from impaired airway clearance at the time of an upper respiratory tract infection or an episode of aspiration, or in the postoperative period.[2–6] Chronic respiratory deterioration can lead to impaired airway clearance and secondary infection without an acute precipitant.[2,7]

Respiratory muscle weakness may primarily involve the muscles of expiration, as occurs in spinal muscular atrophy, or may involve both those of inspiration and expiration as occurs in Duchenne muscular dystrophy.[7,8] Deterioration in respiratory muscle function occurs in progressive diseases, but there may be acute declines at times of illness, leading to an acute on chronic deterioration.[3] Cough is a major mechanism used to expel secretions from the airways.[9–11] The muscles of inspiration and expiration are required to generate an effective cough.[1,4,8,9]

Neuromuscular disorders involving the thoracic cage lead to a reduction in chest wall muscle contraction. These muscles eventually become fibrotic, shortened, and stiffened.[12] Articular contractures develop, reducing chest wall compliance.[12–14] Spinal deformity may contribute to the reduced chest wall compliance.[15] Expansion of the lungs becomes incomplete and areas of microatelectasis occur. Chronic microatelectasis or infections may lead to scarring and reduced pulmonary compliance.[13,16,17] Reduced pulmonary compliance is compounded by reduced elasticity and surfactant production from breathing at lower lung volumes.[13,16,18–20] These changes lead to an alteration in gas distribution and ventilation-perfusion mismatch.[18,19] Impaired airway clearance leads to secretion retention, potentiating atelectasis, secondary infections, and further reductions in lung compliance.[9] Reduced lung and chest wall compliance places the respiratory muscles at a

mechanical disadvantage and may lead to an acute worsening of respiratory failure and death.

There are many available methods to assist with airway clearance. In recent times there has been a push from many patient groups to offer the use of some of the newer and more expensive techniques to assist airway clearance early in the course of disease, and often before other techniques are tried. This review details the currently available evidence for using various treatment options for impaired airway clearance in clinical practice.

ASSESSMENT OF AIRWAY CLEARANCE

To generate an effective cough, children need to inspire to 80 to 90% of total lung capacity, rapidly and firmly close the glottis, and coordinate glottic opening with rapid, forceful contraction of the muscles of expiration.[7,21] Each step of this process may be impaired in neuromuscular weakness. Subjective assessment of cough is not accurate.

Peak cough flow

Peak cough flow (PCF) is a measure of the maximum airflow generated during a cough and is normally 360 to 1200l/min. The PCF correlates well with the ability to clear airway secretions. Bach et al.[1] have suggested from clinical observations that a PCF of at least 160 l/min is required for effective airway clearance. This group have not found any patients with an assisted PCF >270l/min who developed acute respiratory distress during upper respiratory tract infections.[1] When the PCF is <270l/min, it is likely to fall below 160l/min during times of illness.[22] There has been little specific examination of the alterations in PCF during times of illness published in the literature. PCF is related closely to forced vital capacity (FVC) in Duchenne muscular dystrophy and the relative risk for PCF <270l/min when FVC is <2.1l is 4.8 (1.72–13.40).[23] It is suggested that regular monitoring of the PCF be undertaken when the FVC is <2.1l.[23]

Donha-Schwake et al.[24] performed a retrospective review of 46 children and adolescents (mean age 12.7y, SD 3.7y, range 6–20y, 28 males) with neuromuscular disease. They were able to show that PCF <160l/min was a sensitive (75.2) and specific (79.2) predictor for a history of severe chest infections requiring admission.

In patients with neuromuscular weakness and an endotracheal or tracheostomy tube, the ability to extubate (remove the endotracheal tube) or decannulate (remove the tracheostomy tube) is related to the PCF. Bach and Saporito[25] documented the PCF in 62 extubation/ decannulation attempts in 49 patients with neuromuscular weakness. Extubation was to non-invasive ventilation. The 43 attempts with a PCF >160 l/min were all successful. Fifteen attempts with a PCF <160 l/min all failed, and of four patients with a PCF of 160 l/min, two were successful and two failed.

Measures of respiratory muscle strength

Maximal expiratory pressures measured from total lung capacity correlate well with the ability to generate adequate flow for an effective cough.[26] If the maximal expiratory pressure is >60cmH$_2$O, the cough is effective. Cough transients are the bursts of increase in flow above the normal forced expiratory flow that occur during coughing. Cough transients are required to be greater than the normal forced expiratory flow to effectively expel air and clear airway debris. When patients with neuromuscular weakness are asked to perform a forced expiratory flow followed by a single partial inspiration and then successive coughs, effective cough transients are generated in those with maximal expiratory pressure >60cmH$_2$O and are absent in those with maximal expiratory pressure <45cmH$_2$O.[26]

Dohna-Schwake et al.[24] measured peak expiratory pressure after inspiration to inspiratory vital capacity (IVC) and found a peak expiratory pressure <3kPa to be sensitive (85.7%) but not specific (47.3%) for severe chest infections. Peak inspiratory pressure had only a weak influence on PCF.

Inspiratory vital capacity

The IVC is the maximum capacity of air that can be inspired during normal respiration (i.e. without air stacking). Donha-Schwake et al.[24] retrospectively reviewed 46 children and adolescents with neuromuscular disorders for a history of severe chest infections requiring admission. The IVC was the strongest risk factor for severe chest infections. An IVC <1.1l was a sensitive (90.5) and specific (70.8) predictor for a history of severe chest infections requiring admission. This group propose that increasing the IVC by air stacking to the maximum insufflation capacity should improve the PCF and airway clearance and prevent future severe chest infections.

Maximum insufflation capacity

The IVC can be increased by actively stacking air behind the glottis beyond the volumes that the unaided muscles of inspiration are able to achieve. The maximum insufflation capacity is a measure of inspiratory volumes achieved by air stacking.[27] Various methods of air stacking are available and will be discussed in the following section.

Kang and Bach[27] used air stacking with a manual resuscitator bag or portable volume ventilator to stack air forcibly behind the glottis until no further increases in capacity were seen (maximum insufflation capacity). If oropharyngeal and laryngeal function are adequate to hold air behind the glottis, the maximum insufflation capacity is larger than

the vital capacity. The amount of air that can be stacked is limited by chest wall and lung compliance and is related to the vital capacity. In patients with lower vital capacities, the maximum insufflation capacity is less, but the lower the vital capacity, the greater the percentage increase in volume from the vital capacity to the maximum insufflation capacity. Air stacking potentiates cough so that those patients who are able to stack air have a greater increase in their PCF than those who cannot (78 vs 21%).

WHEN TO ASSIST AIRWAY CLEARANCE

Patients with neuromuscular weakness usually have their respiratory status monitored with regular spirometry. Prediction equations for normal spirometry values have been developed using ulna length measurement to overcome the difficulties in measuring height or arm span in this group of children.[28] PCF is a simple, easily performed test that should be monitored regularly once the FVC is <2.1l.[23] There is not good evidence in the literature for when regular monitoring of IVC or respiratory muscle strength should begin.

Airway clearance becomes impaired and the risk of serious chest infection increases when the PCF is <160l/min, the maximal expiratory pressure is <45cmH$_2$O, and the IVC is <1.1l. A PCF <270l/min is likely to fall below <160l/min during acute illnesses. Techniques to assist airway clearance should be taught when these levels are approached.[1,22,24,26] In general, teaching the techniques required when patients are well allows them to master the techniques for use when they are unwell or in the perioperative period.

TECHNIQUES TO ASSIST AIRWAY CLEARANCE

Airway clearance can be assisted by augmenting inspiration, expiration, or both, by aiding mucous mobilization, or by direct suctioning of the airway.

Augmenting inspiration

Augmenting inspiration generally involves air stacking. This can be done by glossopharyngeal breathing or by the use of a manual resuscitator bag or a ventilator (invasive or non-invasive) to force air into the airway generally without expiring between breaths and holding the stacked air behind a closed glottis. After air is stacked, a cough can be generated by coordinating rapid forced expiratory effort with glottic opening, or it may be combined with an assisted cough. Air stacking may not be as effective when glottic incompetence is present.[27]

Glossopharyngeal breathing uses the glossopharyngeal muscles to gulp air repeatedly through the glottis, which is then closed to stack air.[29] The patient does not expire between gulps. It does not require the assistance of a carer.

Once air is stacked, a cough or assisted cough may be used to clear the airways. Its effectiveness is limited by poor laryngeal function, as glottic closure between gulps is required to hold the stacked air. It cannot be used if a tracheostomy is present because of air escape.[29]

A manual resuscitator bag with a one-way valve may be used to force air through the glottis, which is then closed to hold the stacked air. Expulsion of the air can then occur forcefully with an unassisted or assisted cough. Air stacking using a combination of glossopharyngeal breathing and the manual resuscitator bag can increase the unassisted PCF from 108l/min (SD 61.8) to 256l/min (SD 77.4).[30] Use of the manual resuscitator bag requires the assistance of a carer. The manual resuscitator bag may be used with a face mask or may be attached to a tracheostomy or endotracheal tube.

A ventilator may be used to stack air mechanically in a similar way to the manual resuscitator bag. It can be applied non-invasively with a nasal or oronasal mask or a mouthpiece. It can also be applied to a tracheostomy or endotracheal tube. The ventilator pressure may be increased to 30 to 50cmH$_2$O for several breaths without expiring between breaths to attain air stacking. This may then be followed by an unassisted or assisted cough.

The intermittent positive pressure breathing device may be used to augment inspiration. Dohna-Schwake et al.[21] used intermittent positive pressure breathing via a mouthpiece or oronasal mask in a group of 29 patients with neuromuscular diseases to air-stack. The patients had a PCF <160l/min or a history of chest infections. Air stacking was performed by titrating pressures from 10 to 40mbar until further volume increases were no longer seen or patients did not tolerate further increases in pressure. The mean pressure to obtain maximum insufflation capacity was 32.6mbar (SD 4.9). The IVC increased from 0.68 (SD 0.40) to a maximum insufflation capacity of 1.05 (SD 0.47; $p<0.001$). The unassisted PCF rose from a mean 119.0l/min (SD 57.7) to 194.5l/min (SD 74.9; $p<0.001$). Patients with lower IVCs had greater improvements in their PCF.

Augmenting expiration

The assisted cough involves the patient taking a deep inspiration and closing the glottis. As the patient opens the glottis, and forcibly contracts the muscles of expiration, to generate a cough, an upward thrust is applied to the epigastric area (Heimlich-type assisted cough) or the anterior chest wall by a carer. The benefit of the Heimlich-type assisted cough may be limited in those with obesity or severe scoliosis, where chest wall deformity can limit the benefit.[31] In those with osteoporosis, care must be taken when using the anterior chest wall-assisted cough. The

benefit of the assisted cough may be diminished by poor laryngeal function. If air cannot be held behind a closed glottis, then coordinating expulsion of air with a thrust from a carer is more difficult and if coordination is not ideal, the effectiveness of the assisted cough diminishes.[9,32]

In a group of eight patients with neuromuscular weakness and without scoliosis, the assisted cough resulted in a PCF increase to 185l/min (93–355) from an unassisted PCF of 104l/min (43–188) $p<0.01$.[31] The assisted cough can also be used with air stacking techniques, where air is stacked by the use of glossopharyngeal breathing, a manual resuscitator bag, or a ventilator (non-invasive or invasive) to stack air before performing the assisted cough.[27,30] Air stacking using glossopharyngeal breathing and the manual resuscitator bag and assisting the cough can increase the unassisted PCF from 108l/min (SD 61.8) to 256l/min (SD 77.4).[30]

Augmenting inspiration and expiration

The mechanical insufflator-exsufflator uses positive pressure to air-stack before reducing the ventilator pressure to a negative value as the glottis opens to draw the air out. The Cough Assist (JH Emerson Company; Cambridge, MA) is a compact mechanical insufflator-exsufflator that is gaining popularity. It can be used with a facemask or mouthpiece, or used via a tracheostomy or endotracheal tube. It uses a pressure of up to $+60cmH_2O$ to augment inspiration followed by a negative pressure of up to $-60cmH_2O$ to augment expiration. It can be used in automatic, manual, or combined modes.

Chatwin et al.[8] studied 22 adults and children with neuromuscular disorders who were clinically well. The patient group tended to have relatively severe respiratory muscle weakness, with 17 of the 22 patients requiring non-invasive nocturnal ventilation and all having recurrent chest infections. Comparison was made between PCF measured by the unassisted cough (mean 169l/min, 95% confidence interval [CI] 129–209), physiotherapy-assisted cough (mean 188l/min, CI 146–229), non-invasive ventilator-assisted cough (182l/min, CI 147–217), exsufflation-assisted cough (235l/min, CI 186–284), and insufflation-exsufflation-assisted cough (297l/min, CI 246–350). The expiratory pressures were set to patient comfort and had a mean insufflation pressure of $15cmH_2O$ (SD 3) and a mean exsufflation pressure of $-15cmH_2O$ (SD 9). There was a significant improvement in the PCF obtained from the unassisted cough and the exsufflation-assisted cough in adults ($p<0.01$) and between the unassisted cough and the insufflation-exsufflation-assisted cough in both adults and children ($p<0.001$).

Chatwin and Simonds[33] then studied the benefit of the mechanical insufflator-exsufflator in a group of eight patients with neuromuscular weakness who were unwell with chest infections. The trial was a 2-day randomized crossover treatment programme of non-invasive ventilator-assisted physiotherapy versus non-invasive ventilator-assisted physiotherapy with the mechanical insufflator-exsufflator. The use of mechanical insufflator-exsufflator was reported to reduce treatment times until the physiotherapist felt that airway clearance was adequate and the patients subjectively felt that clearance was better.

Vianello et al.[34] treated 11 patients with neuromuscular weakness and a respiratory tract infection with the mechanical insufflator-exsufflator along with chest physiotherapy. Comparison was made with a historical group of 16 patients who had been treated with postural drainage, the assisted cough, and suctioning. Treatment with the mechanical insufflator-exsufflator was not tolerated by one patient because of gastric distension and gastro-oesophageal reflux. Another patient had gastric distension without reflux and was able to continue treatment. Two out of the eleven mechanical insufflator-exsufflator patients and 10 out of the 16 control patients required minitracheostomy or intubation. The authors concluded that the mechanical insufflator-exsufflator improved airway mucous encumbrance and should be included in the non-invasive management of patients with neuromuscular weakness. There was no comparison with other available techniques to assist airway clearance.

Fauroux et al.[35] studied 17 children with neuromuscular weakness who had been clinically stable for the preceding month. All were using intermittent positive pressure ventilation. The effects on physiological parameters before and 1 minute after the use of the mechanical insufflator-exsufflator were investigated. Mechanical insufflation and exsufflation were set at + and $-15cmH_2O$, $30cmH_2O$, and $40cmH_2O$ respectively. Children did not reach the machine's set pressures for inspiration or expiration. Inspiratory and expiratory flows were greater than those during tidal breathing and expiratory volumes were greater than those during tidal breathing. Compared with a forced expiratory manoeuvre, the expiratory volumes were greater only at pressures of + and –30 and $40cmH_2O$. There was a statistically significant decline in the end tidal carbon dioxide level at all pressures, but this is probably clinically insignificant as all levels were within the normal range, and alterations were small. There was also an increase in the sniff nasal inspiratory pressure (29 [SD 19] vs 31 [SD 20], $p=0.046$) and PCF or peak expiratory flow (162 [SD 97] vs 192 [SD 99], $p=0.02$). The mechanical insufflator-exsufflator was well tolerated at all pressures and the subjective respiratory comfort improved after using pressures of + and $-40cmH_2O$. The vital capacity was unchanged. To my knowledge, this is the first study to have examined

the physiological effects of the mechanical insufflator-exsufflator in children.

There have been case reports[4] and retrospective reviews[7,30] of the clinical use of mechanical insufflation-exsufflation but no further prospective comparisons with other methods of assisting airway clearance. The mechanical insufflator-exsufflator is generally well tolerated and considered safe.[7,30] Intolerance has been attributed to inadequate caregiver training, but some patients did not find it subjectively effective or thought that it contributed to chest wall or abdominal pain.[7]

Mucous mobilization

The intrapulmonary percussive ventilator delivers high-frequency (100–300 cycles/min) bursts of gas to the lower airway to reduce the viscosity of secretions during acute illnesses. In a case series of four patients with neuromuscular disease and atelectasis resistant to standard physiotherapy who used the intrapulmonary percussive ventilator, one had a third-degree atrioventricular block resulting in hypoxaemia and bradycardia during two treatments, and this treatment modality has now fallen out of favour.[36]

Tracheostomy

A tracheostomy provides the means for direct airway suctioning and clearance of secretions from the large airways.[25] Techniques of air stacking such as using the manual resuscitator bag or ventilator to stack breaths and augmenting expiration with the assisted cough or mechanical insufflator-exsufflator can be used via a tracheostomy.[27,30] A tracheostomy is not without risks and can limit the patient's ability to communicate.[25]

CONCLUSION

In patients with neuromuscular weakness, impairment in airway clearance may occur gradually with disease progression, or more acutely at times of respiratory tract infections or aspiration or postoperatively. Cough is a major mechanism of airway clearance and assessment of inspiration and cough can be used to guide the introduction of techniques to assist airway clearance. If the IVC is <1.1l, the PCF is <160l/min, maximal expiratory pressure is <45cmH$_2$O, or peak expiratory pressure is <3kPa, patients are at risk of severe chest infections.[1,24,26] A PCF <270l/min is likely to fall below 160l/min at times of illness.[22] The risk of PCF <270l/min rises when the FVC is <2.1l.[23] At these levels, techniques to assist airway clearance should be considered.

Techniques to assist airway clearance include air stacking, assisting the cough, or using the mechanical insufflator-exsufflator. The available techniques may be used alone or in various combinations to obtain effective clearance for an individual. The combination of two or more techniques may be effective where either technique alone is not adequate.[27,30] When choosing which techniques to teach individuals, the severity of airway clearance impairment, patient comfort, and ease of performance of the available techniques need to be considered. The mechanical insufflator-exsufflator has gained increasing popularity in recent times, but other techniques may be very effective and do not impose the same financial costs. Simple techniques may also be effective adjuncts to the use of the mechanical insufflator-exsufflator.

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