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Short of breath, short of air, short of mechanics

INTRODUCTION

If you ask a neurologist about neurological disorders and respiratory failure, you will get a more or less adequate list of the Guillain–Barré syndrome, myasthenia gravis, amyotrophic lateral sclerosis and even the exceedingly rare Ondine’s curse, but probably nothing much else. Respiratory failure is not our specialty’s forté, and neuromuscular respiratory failure is not common. However, every once in a while, an undiagnosed patient is encountered with laboured breathing. The response should be swift. Not only can apnoea be imminent, but pulmonary aspiration is equally life threatening.

Neurological disease can impair respiration at multiple levels (Fig. 1) (Kelly & Luce 1991). The interconnections between the respiratory centres, motor neurones, and the respiratory muscles provide a functional system that moves air in and out of the lungs. If it fails, it causes hypercapnia. The alveoli and pulmonary capillaries permit efficient gas exchange by diffusion through a paper-thin barrier; if it fails, it causes hypoxemia. Both conditions may occur simultaneously, or one disorder may lead to the other when reduced airflow leads to poor alveolar recruitment followed by collapse.

Figure 1 Causes of respiratory failure in neurological disease.

In the initial evaluation, it is important to ask three major questions: does the patient generate breaths; is air getting to where it needs to be; and is the pulmonary apparatus intact: short of breath, short of air, or short of mechanics (Table 1)? This focused review will discuss what neurologists – in my opinion – need to know about acute respiratory failure.

Table 1 Causes of respiratory failure

Short of breath (drive)
opioids, barbiturates, anaesthetic drugs (e.g. propofol)
pontomedullary brainstem lesion (haemorrhage, infarct, trauma)
brainstem death
cervical spinal cord transection above C3
Short of air (conduit or transport)
upper airway obstruction (tongue, mucus, tooth fragment)
pneumothorax
massive pulmonary aspiration
severe anaemia
Short of mechanics (bellows)
flail chest (rib fracture)
reduced neuromuscular junction traffic (myasthenia gravis, Eaton-Lambert, organophosphates, botulism, tick paralysis).
diaphragm failure (trauma, Guillain-Barré, amyotrophic lateral sclerosis, myopathies)

THE ARCHITECTURE – WELCOME TO THE MACHINE

The muscles involved with respiration are not always all active at once. During quiet breathing, the diaphragm contracts synchronously with the intercostal and scalene muscles (inspiration), and the abdominal muscles barely assist the passive recoil of the rib cage (expiration). The inspiratory and expiratory muscles cycle but are simultaneously activated when we speak or sing.

The activation of muscles in the upper airway, such as the pharyngeal constrictor muscles and genioglossus, keep the pharynx patent. The diaphragm controls most of inspiration and dyspnoea is expected when it is not working properly. When it contracts, the rib cage expands due to its cephalo-caudal muscle fibre orientation. When the diaphragm is weak, accessory muscles – such as sternocleidomastoid, pectoralis, trapezius, and latissimi dorsi – which harness the rib cage, are recruited and their active contractions can be palpated by the examiner. Expiration is assisted by contraction of the abdominal muscles.

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In fact, when the thoracic volume is minimized after forceful exhalation, elastic energy is built up, which aids inspiration. Coughing requires closure of the glottis and contraction of both the diaphragm and the abdominal muscles. A patient with a forceful cough rarely has significant neuromuscular respiratory failure. The abdominal muscles can become severely affected in any neuromuscular disorder, reducing the effectiveness of coughing, and so predisposing to atelectasis.

THE BASICS – DO'S, DON'TS, DON'T-YOU DARE

It is almost routine for neurologists to call on the skills of emergency room physicians, anaesthesitists, or respiratory physicians to deal with a patient with an acute neurological disease and poor breathing efforts. Yet neurologists, usually more interested in the primary pathology than in what it does to respiratory physiology, may still sometimes find themselves in the awkward position of having to manage a patient in respiratory failure. Although many neurologists have

Figure 2 Maintaining the airway in an emergency (from Wijdicks & Borel 1998).

been disillusioned after their first attempt at intubation, or trying to understand a mechanical ventilator, basic airway management should be part of any physician's armamentarium, including neurologists. Securing the airway appropriately is not complicated (Wijdicks 1997; Wijidicks & Borel 1998). Figure 2 shows the initial manoeuvres.

Any discussion must begin with close attention to any upper airway obstruction. The airway may be blocked by vomit, mucus, blood, or other foreign objects and they need to be removed quickly. In some patients with significant facial trauma, the face is grotesquely swollen, making it virtually impossible to inspect the mouth. In these patients, often with paroxysms of stridor, emergency tracheostomy can be immediately successful.

(In the field, a laryngeal mask can be used, particularly when the larynx is obscured by blood and secretions. This mask is easily placed on the face and can be used as a bridging device before the endotracheal tube; Fig. 2). The upper airway should first be opened by tilting the patient's head backwards. This is called 'the sniffing position' and it will secure a normal conduit through the trachea and pharynx. Extension of the neck might be compromising in a patient with a spinal cord injury, and it remains important to keep the neck immobilized. The tongue jaw lift/finger sweep method involves placing the thumb in the mouth with the other fingers pulling the chin forward, while cleaning the mouth using the opposite hand. The tongue may not only lie frozen due to a stroke but the fleshy muscle may fatally obstruct the upper airway anytime the patient lapses into stupor. The neurologist should then stand behind the patient placing the thumb and index finger on the mask and the other fingers behind the vertical part of the mandible, lifting the jaw upwards to open the collapsed airway. The thumb and index finger press the ventilating mask onto the face. The success of the maintained airway can be guided by the patient's colour, and also by readings of the pulse oximeter, which should show more than 90% oxygen saturation (by the way, nail polish makes it a useless device).

Mask ventilation using a resuscitation bag that supplies 10–15 L/min of oxygen can maintain the airway for hours, until a skilled anaesthetist arrives. After the airway is secured, endotracheal intubation should follow. Although the neurologist may not perform the procedure, he should be flagrantly provocative by warning that succinylcholine may raise intracranial pressure (Lanier *et al.* 1986) and by asking for fiberoptic intubation when neck injury is likely (Rhee *et al.* 1990). Moreover, even though there is no strict definition, 'a difficult airway' should be anticipated before introducing the laryngoscope blade. Immediate red flags are mandibular hypoplasia, history of rheumatoid arthritis, short muscular neck, inability to visualize any pharyngeal structures other than the hard palate, and finally inability to insert three closed and extended fingers into the open mouth. The predictive value of all these bedside warnings has not been tested, but their presence should at least change the mind of an enthusiastic novice.

THE NEUROLOGY OF BREATHING – BE MINDFUL OF THE THREE ‘BS’: BITE, BELLOWS AND BELLY

Acute lesions of the cerebral hemisphere or brain stem impact on respiratory control. Voluntary control of breathing originates in the cortex that connects to all spinal cord levels and the motor neurones send connecting fibres to the diaphragm, intercostal and abdominal muscles. The automatic control of respiratory drive is in the primary ventilatory nuclei of the brain stem, where lesions can cause apnoea during sleep (so-called Ondine’s curse, but this is so extremely rare that it does not merit monitoring every brain stem stroke patient, except with pulse oximetry). Impaired voluntary breathing involves an entire gamut of respiratory disorders and is not discussed further.

Certain abnormal breathing patterns result in hypoxaemia and can barely be observed due to much-needed intervention with the mechanical ventilator. These include apneustic breathing or cluster breathing, both common in brain stem strokes. Cheyne–Stokes breathing (oscillating cycle of up to 2 min of hyperpnoea separated by apnoea) is very frequent, and should be left alone, but may lead to brief oxygen desaturation. Acute spinal cord lesions affecting C3–C5 result in ventilator dependency. It is a misconception, however, to think that this stops patients speaking, because special speech tracheal cannulas are available. Lesions below the level of C₅ spare the nerve connections to the diaphragm, but expiratory effort is markedly reduced due to involvement of the abdominal and intercostal muscles. Placing the patient in a supine position improves expiration due to pressure of the abdominal contents on the thorax. Breathing becomes laboured when the patient is sat in a chair and so abdominal binders are required (compare with the Guillain–Barré syndrome with exactly opposite responses).

In breathlessness due to neuromuscular disease, the focus of examination should be on bulbar dysfunction (**bite**), paradoxical breathing (**bellows**), and impaired coughing (**belly**). Careful inspection and testing of the oropharyngeal muscles may point to a diagnosis. Long-standing dysfunction, such as in amyotrophic lateral sclerosis, is often evident by the wrinkled, fasciculating slowly moving tongue and hyperactive jaw reflex. In myasthenia gravis, muscle weakness is prominent in the masseters and repetitive forceful biting on a tongue depressor is

soon followed by inability to close the teeth. Passage of air through the nose when asked to blow out the cheeks against counter pressure reveals palatal as well as facial weakness. On the other hand, jaw weakness is a late phenomenon in the Guillain–Barré syndrome and often absent. The assessment of oropharyngeal weakness not only hints at involvement of the respiratory mechanisms, but ineffective swallowing leads to aspiration. In a recent study, bulbar dysfunction predicted later requirement of mechanical ventilation (Lawn *et al.* 2001).

The symptoms of acute neuromuscular respiratory failure (**bellows**) are often very subtle indeed. When initially confronted with these patients, an initial cursory observation of breathing, blood gases, and chest X-ray indicates no danger to the patient at all. However, after striking up a conversation, it becomes obvious that the patient frequently pauses during sentences to take a breath, sweats at the hairline, has a mild tachycardia, and when asked, confirms a sense of discomfort. Also, when asked if they have to work to breathe, these patients will be glad you asked and nod yes. And yet the arterial blood gases can be entirely normal because the patient is still able to compensate for threatening hypoxaemia by breathing faster. The typical response in other medical disorders is increased tidal volume, but in neuromuscular disorders this is actually reduced because of respiratory muscle fatigue. The tachypnoea may be subtle and the respiratory rate is often increased to 20–30 breaths per minute and quickly rises. Remarkably, studies on the normal respiratory rate are virtually nonexistent, but it is around 15 breaths per minute.

The classic textbook features of inspiratory paradox is inward movement of the abdomen during inspiration. When the diaphragm stops working, the positive downward pressure on the abdominal contents is replaced by a negative pressure, which causes the inward sucking movement of the abdomen (with normal inspiration the diaphragm expands the chest, the lungs fill with air, and the abdomen is pushed out). However, inspiratory paradox due to diaphragmatic weakness is a late phenomenon and, particularly when observed in a patient with an acute neuromuscular disorder, indicates that an early opportunity for intubation has been missed. These patients are on the verge of apnoea and many of them stop breathing in the middle of the night (Lawn *et al.* 2001). A useful bedside

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test is to have the patient count out loud to 20 in one breath after maximal inhalation. If the patient can count at least one number per second, the vital capacity is probably still normal and outside the danger zone. It is important to note here that both hypoxaemia and hypercarbia are late phenomena. Hypoxaemia occurs due to significant shunting associated with collapse of multiple alveoli that are not recruited during breathing. Thus, patients who have a mild tachycardia displaying evidence of hypoxaemia on a pulse oximeter, and have evidence of increased work of breathing, or sweat beads at the hairline, need to be intubated pre-emptively (Fig. 3). The abdominal muscles (**belly**) barely contribute to the mechanics of respiration. However, when the diaphragm fails, they may be recruited and then fulfil a major role in expiration and coughing. The maximum expiratory pressure is an important value to assess and a decrease from normal (< 40 cm H₂O) means the patient will accumulate secretions, develop microatelectasis and even bronchial branch obstructions.

THE DEVICES – SIMPLIFY, SIMPLIFY

Pulmonary function tests are very useful and easy to do with simple mechanical bedside devices. The commonly-used peak flow metre (for asthma) is unreliable because expiratory flow rates are normal. In neuromuscular respiratory failure, the airway is patent and lung recoil is actually increased. The most simple test is assessment of vital capacity, maximal inspiratory (PI_{max}), and maximal expiratory (PE_{max})

pressures. The patient's position in which these values are obtained is rather critical because clinically-relevant diaphragmatic fatigue may become obvious in the supine position (Allen *et al.* 1985). The technique of assessment of respiratory muscle function is important and scuba dive mouthpieces may reduce leakage, particularly if there is bilateral facial palsy.

There is little data of the usefulness of pulmonary function testing in clinical assessment, except in the Guillain-Barré syndrome. A retrospective analysis of 114 patients noted possible critical values of vital capacity of less than 20 mL/kg, PI_{max} less than 30 cm H₂O, PE_{max} less than 40 cm H₂O (the so-called 20/30/40 rule), but also any reduction in vital capacity of more than 30% from baseline (Lawn *et al.* 2001).

Severe respiratory muscle weakness is very rarely an isolated feature. Subtle limb muscle weakness is also often detected. An important correlation, or lack thereof, is reduction of respiratory muscle strength with either PaO₂ or PaCO₂. Only with 50% reduction in strength, is there some rise in PaCO₂ (Braun *et al.* 1983), although low flow (0.5–2 L/min) O₂ administration may worsen hypercarbia substantially (Gay & Edmonds 1995). There are no mechanisms to increase alveolar ventilation, and in patients with longstanding carbon dioxide retention and dependency on 'hypoxaemic drive', the additional administration of oxygen may cause hypercarbic coma.

After intubation has been performed, it is important to place the patient on the mechanical ventilator to provide adequate oxygenation. Virtually all patients are well served with an initial ventilator order that includes intermittent mandatory ventilation mode. The positive pressure breaths that are delivered by the mechanical ventilator are triggered by the patient, usually by generating a small pressure difference in the patient-ventilator circuit. The patient is able to breathe in between and have entirely unsupported breaths. This ventilator order is particularly useful in neurological patients because it allows for spontaneous breathing and yet it can deliver hyperventilation if needed. A typical ventilator order for neurologically stable patients is an SIMV (synchronized intermittent mandatory ventilation) mode of FIO₂ (fraction of inspired oxygen) of 0.4–1.0, respiratory rate at 8–12 breaths/minute, tidal volume of 10–15 mL/kg, PEEP (positive end – expiratory pressure) of 2–5 cm of H₂O, and an inspiration/expiration ratio of 1–3 (Slutsky 1993; Wijedicks & Borel 1998).

Figure 3 Breathless with forehead sweating and tachycardia – bring out the scope.

Remember,
blue is bad,
white is
worse

After the patient has remained on the mechanical ventilator for a time, a tracheostomy should be considered if prolonged mechanical ventilation is anticipated. There is an unexplained urge to do an early tracheostomy, probably because of comfort for the patient, the claim of more effective tracheal suctioning, and – even more unproven – the alleged considerable risk of tracheolaryngeal stenosis (Marsh *et al.* 1989). These concerns typically apply to patients with underlying chronic obstructive pulmonary disease, who populate medical intensive care units. But patients with acute neurological disease (Guillain–Barré or myasthenia gravis) usually have normal lungs and do not appreciate a tracheostomy scar as the only tell-tale sign of having been in an intensive care unit (Wijdicks *et al.* 2001). Many patients with a central nervous system catastrophe can be weaned off the mechanical ventilator in three weeks, and tracheostomy can be avoided. In patients with the Guillain–Barré syndrome or myasthenia gravis, improvement is also expected within the first 20–30 days, obviating the use of a tracheostomy (Lawn & Wijdicks 1999). Tracheostomy is undoubtedly indicated in patients (Lanza 1990) who remain comatose following head injury, patients who have a persistent spinal cord injury and decide to maintain this level of support, and in patients in which the neurological disorder is irreversible such as motor neurone disease. If tracheostomy is considered, traditional surgical tracheostomy is standard although there is renewed interest in the use of percutaneous dilatational tracheostomy (Ciaglia & Craniero 1992; Malthaner *et al.* 1998; Suh *et al.* 1999). This procedure is usually performed by general surgeons and therefore continues to be criticized by some otolaryngologists. It may be preferred in patients with the Guillain–Barré syndrome or myasthenia gravis because the cosmetic effects are substantially less and complications such as haemorrhage, false passage, pneumothorax, and tracheal stenosis continue to be very low.

CONCLUSIONS

This brief review emphasizes some of the roles neurologists can play in airway management. Neurologists do not need a sophisticated knowledge of mechanical ventilators, how to intubate, or initiate a weaning procedure. Mechanical ventilators are constantly changing, and their operation is best handled by experts in the field. In the USA, respiratory therapists are specially trained to maintain the mechanical ventilator but, due to

the marked turnover of machines, even they can be caught thumbing through the manual.

Neurological disease can strike at different sites of the nervous system and for any patient who is not breathing well, a systematic approach is indicated after securing an adequate airway and oxygenation. Early intervention is necessary before circulatory collapse. Remember, blue is bad, white is worse.

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