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# Assessment of the Severity of Primary Respiratory Muscle Failure

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**SUMMARY**

Respiratory muscle weakness results either from disorders of the respiratory motor unit (primary type) or from muscle fatigue due to severe underlying pulmonary disease (secondary type). Blood gases reflect the effects of muscle fatigue but are too insensitive to be relied upon in primary muscle weakness. The reasons for this are discussed. It is emphasized that the ventilatory reserve should be monitored in patients with primary respiratory muscle weakness and that a decision to provide assisted ventilation should be based upon it, not on blood gas estimates.

**History**

The origin of modern critical care medicine can be traced to Lassen's 1953 description of the management of patients with paralytic poliomyelitis during an epidemic in Copenhagen the previous year. At the suggestion of an anaesthetist, Dr. B. Ibsen, 315 patients with respiratory muscle paralysis were ventilated during a six-month period by means of intermittent positive pressure breathing (IPPB) through cuffed tracheostomy tubes. This reduced mortality to 40 per cent, from the 80 per cent of previous years when only cuirass ventilators (iron lungs) were used. No IPPB machines were available at that time and patients were ventilated manually for periods as long as 3 months at a time by up to 200 medical students each day, working in relays. Respiratory muscle failure remains an important cause of respiratory insufficiency today and anaesthetists continue to play a leading role in intensive care.

**Classification**

Primary respiratory muscle failure is produced by neuromuscular disorders affecting the motor unit. The Guillain-Barré syndrome, myasthenia gravis, snake bite, hypokalaemia, peripheral neuropathy and acute myositis are important causes. Fortunately, paralytic poliomyelitis is now rare but other viral infections may produce similar effects. Secondary respiratory failure occurs from muscle fatigue in response to excessive work loads imposed by severe airway obstruction or lung disease. Primary respiratory muscle weakness is relatively easy to recognise but the assessment of its severity is rendered difficult by the fact that blood gases are relatively insensitive to its presence. It is difficult to detect secondary respiratory muscle weakness (muscle fatigue) but since its effects are reflected by the blood gases and since it will recover with the underlying cause, it is rarely essential to make a specific diagnosis of the condition.

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**Pathophysiology**

The function of the respiratory system is to secure gas exchange between blood and air so that the arterial blood gas tensions are kept within certain limits. Respiratory failure has been defined as an arterial blood $PaO_2$ below 60 mm/Hg (8,0 kPa) or a $PaCO_2$ above 49 mm/Hg (6,6 kPa). However, blood gases are more affected by disease of the lower airways and gas exchange units (acini) than by extrapulmonary disorders such as upper airway obstruction, abnormal central drive or weakness of the respiratory muscles. Since the lungs are normal in these conditions and gas exchange is minimally affected until the disease has reached a very advanced stage, the severity of the respiratory impediment can be seriously underestimated if blood gases alone are relied upon.

An insight into the reasons for the relative insensitivity of blood gas estimations in respiratory muscle weakness can be gained by examining the serial changes that occur in lung volume in a patient with progressive muscular weakness (Figure 1). Total lung capacity, vital capacity and inspiratory and expiratory reserve volumes fall steadily with progressive muscular weakness (Figure 1B). The tidal volume is however maintained at near normal levels. Any decrease in tidal volume is made up for by an increased respiratory frequency so that minute ventilation and alveolar ventilation ($PaCO_2$) are maintained until the respiratory weakness is extreme. In fact, patients with acute muscle weakness usually hyperventilate (low $PaCO_2$) as a result of reflexes that arise in the chest wall, even when very weak. In severe

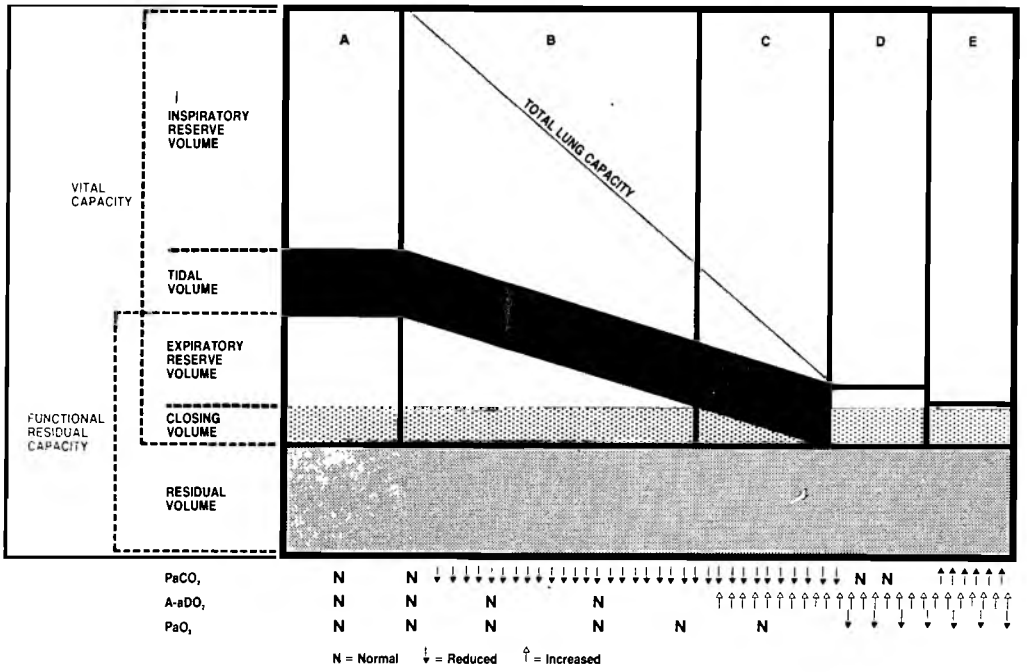


*Fig. 1. Sequential lung volume changes in progressive respiratory muscle paralysis.*

weakness the inspiratory and expiratory reserve volumes have been lost, no ventilatory reserve remains and the patient uses his entire vital capacity for each breath. It is only when the tidal volume falls so low that it can no longer be compensated for by an increased respiratory frequency that the $PaCO_2$ will begin to rise. A normal $PaCo_2$ is thus an ominous sign in a patient with clinically obvious respiratory muscle weakness, being indicative of the loss of the entire ventilatory reserve and any elevation in the $PaCO_2$ is an indication for immedicate intervention.

The alveolar-arterial oxygen tension gradient ($A\text{-}aDO_2$) is more sensitive than either the $PaCO_2$ or $PaO_2$ for demonstrating the presence of abnormal gas exchange. Unfortunately it is seldom estimated. The $A\text{-}aDO_2$ remains normal in patients with respiratory weakness until (Figure 1C) the tidal volume enters the closing volume range. The closing volume is a subdivision of the expiratory reserve volume and is so named because small airways begin to close off and impair gas exchange when the expiratory reserve volume falls to its level. The greater the fraction of the tidal volume that falls in the closing volume range, the greater will the $A\text{-}aDO_2$ be. Closing volumes are related to age. They are at their lowest, approximately 10 per cent of vital capacity, between the ages of 12-24 years. They are highest in the newborn and elderly. The lower the closing volume, the smaller will be the effect of the declining expiratory reserve volume on gas exchange and the less sensitive will the $A\text{-}aDO_2$ be.

Arterial oxygen tension ($PaO_2$) can be quite misleading. Hyperventilation results in the arterial oxygen tension being maintained within the normal range despite an increasing $A\text{-}aDO_2$ gradient and the $PaO_2$ will only fall appreciably when between 30-50 per cent of the tidal volume occurs within the closing volume (Figure 1D).

The interpretation of blood gases in patients with respiratory muscle weakness is thus difficult and changes indicative of overt respiratory failure only occur with very advanced disease. I therefore discourage the use of blood gases as a criterion for intervention in these patients.

**Ventilatory reserve**

It can be seen from the above that primary effect of respiratory muscle weakness is a decrease of the patient's ventilatory reserve (inspiratory plus expiratory reserve volumes). When the loss of ventilatory reserve is so great that the ability to cough effectively is lost, the patient's life is in danger. The cough is a major defence mechanism of the respiratory tract. Its suppression will inevitably be followed sooner or later by pneumonia with progression to overt respiratory failure and death unless ventilatory assistance is given. **Pneumonia is the major cause of morbidity and mortality in patients with primary respiratory muscle failure and it should be the objective of therapy to identify patients who have lost their ventilatory reserve before this complication supervenes.** This can be done either on the basis of certain measurements or by clinical observation.

**Criteria for ventilatory intervention**

**Measurements.** A vital capacity below 12 ml/kg, a ventilatory dead space to tidal volume ratio 0,6 or greater, or inability to generate inspiratory negative pressures less than –20 cm of water (2 kPa) are given as criteria for intervention. Ideally one would like to be able to base a decision to intervene on numerical criteria, in practice ill patients cannot be relied upon to make the maximal efforts needed to enable vital capacity or maximal inspiratory force to be determined with confidence. The measurement of dead space to tidal volume ratio (VD : VT) is non-invasive and needs no patient co-opeation but it does require equipment to measure end-tidal and mixed exhaled air that is not readily available. Peak expiratory flow rates (PEF) are an extremely insensitive measure of muscle weakness, are no substitute for vital capacity determinations, and cannot be used as a criterion for intervention.

**Clinical.** In practice, clinical criteria for intervention are used. Clinical signs can be as objective and reliable as the numerical values described above provided that specific signs are used and that one does not base the assessment on superficial impressions of the patient's well-being.

**A cough is the best clinical test of integrated respiratory muscle function.** It requires a maximal inspiration produced by the diaphragm, followed by transient glottic closure (bulbar function) and then a forced expiration using the abdominal muscles. The cry of a child carries the same significance. If the cough, cry or voice are weak the mechanism should be sought for further clinical examination.

Poor inspiratory effort causing a weak cough is almost invariably due to diaphragmatic paralysis. The expiratory reserve volume will be low and can be detected clinically by elevation of the upper border of the liver on percussion above the 5th intercostal space. Indrawing of the abdomen during inspiration (paradoxical abdominal motion) establishes the diagnosis of diaphragmatic paralysis. It can be confirmed by radiographic screening in cases of doubt. When unable to cough, such patients require ventilatory assistance even though they may still appear to be pink and comfortable. Similarly, a pneumonia in a patient with respiratory muscle weakness indicates that the ability to protect the airway has been lost and indicates the need for ventilatory assistance.

A soft voice or cry is seldom, in my experience, due solely to respiratory muscle weakness and usually implies concomitant vocal cord paresis, often associated with malfunction of other bulbar muscles. If good inspiratory and expiratory efforts are present such patients will be able to keep their lungs expanded and clean and need only to be given nasogastric tube feeds to avoid aspiration. In order to protect the airway, a tracheostomy should be performed if there is difficulty swallowing or associated pooling of oral secretions.

Uncommonly, a poor cough is found to be due to abdominal muscle weakness alone with intact diaphragmatic and global function. In this instance, physio-

therapy with abdominal pressure during expiration will, by mimicking a cough, enable one to keep the lungs clean without resorting to intubation or tracheostomy.

**Conclusion**

Blood gases are an insensitive index of the severity of primary respiratory muscle weakness. Affected patients who are unable to cough effectively require assistance that should not be withheld because blood gases are still found to be in the normal range.

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