Respiratory Problems of the Chronically Ill

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• Functional impairment of the respiratory system and subsequent disability may frequently complicate the course and treatment of chronic diseases. The spectrum of physiologic disturbances includes muscle dysfunction, decreased chest wall compliance, decreased pulmonary compliance, increased airway resistance, impaired diffusion, and defective control of ventilation. Early clinical manifestations are easily overlooked or attributed to the primary disease. Special tests are necessary to measure the functional capacity of the respiratory system. Some tests can be performed in the office or at the bedside, while others require a respiratory function laboratory. Management includes the use of respiratory stimulants, mucolytic enzymes, surgical procedures, postural exercises, mobilization and stretching, special breathing technics, and postural drainage. Work simplification and work-saving devices may be needed to help reduce the oxygen demand. Mechanical respiratory aids are used for patients with severe hypoventilation to extend their functional abilities.

The physical restoration of the chronically ill person is aimed at increasing mobility, increasing strength, and increasing functional ability. When we fail, we can often explain it away by saying that the patient was poorly motivated, that he could not tolerate the prescribed exercise, or that his cardiac reserve was inadequate for participation in the rehabilitation program.

Rarely are we aware that many chronically ill persons are also suffering from hypoventilation. This may not only restrict their physical capacity but may also limit their ability to cooperate. Hypoventilation is most often recognized as a result of primary diseases of the lungs such as emphysema, asthma and chronic bronchitis; however, it is most often overlooked when it occurs in conjunction with other chronic diseases. Respiratory functional impairment and subsequent disability may complicate many chronic illnesses, not primarily respiratory, and contribute significantly to the patient's total problem. Defective pulmonary ventilation, though a secondary complication of some chronic diseases, may be severe enough to cause neurologic, psychologic and metabolic disturbances.[1] Unfortunately, the symptoms of hypoventilation are too often passively accepted by both the patient and physician as being the inevitable progression of the primary disease.

Functions of the Respiratory System

The most important function of the respiratory system is to supply oxygen for use by the body cells and to eliminate carbon dioxide. In addition to the exchange of gases between the organism and its environment, the respiratory apparatus must also accommodate such activities as speaking, coughing, singing, sneezing, laughing, sighing and straining, without at the same time interfering with its primary function. Breathing itself depends on the proper functioning of the following four mechanisms:

1. *Respiratory Pump*

The thoracic cage and the muscles of respiration provide the bellows action required for drawing air into the lungs. About 60 per cent of the inhalation force of ventilation is provided by the diaphragm. The other 40 per cent comes from the external intercostals and the scaleni. During stress or after loss of primary inspiratory muscles, the sternocleidomastoids, trapezius, serratus anterior and even the platysma participate in the inspiratory effort. Expiration itself

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internal pressure distribution within the tissue. While soft tissue is deformable by external pressure and thus will diminish the steepness of the pressure gradient within the tissue, any bony prominences are relatively immovable and will exert reactive forces on the tissue between the bone and skin. If the area of the prominence is small, these forces may lead to pressures considerably in excess of that observed externally and thus become the more significant factor in formation of decubiti.

The experience gained with this "Bed of Springs and Nails" and the results obtained provided background and stimulus for the design and development of a pliable device for measurement of pressure distribution over many points using electrical contacts in a matrix of air cells. A "Pneumatic Cell Matrix Measuring the Distribution of Contact Pressure Over Human Body" was developed at the Engineering Design Center of Case Institute of Technology[6,7] and the results obtained with this device will be reported in the next paper of this series.

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is passive and depends mainly on the recoil of the musculoskeletal framework of the thorax and of the lung tissue. The internal intercostals are also used during expiration when more than tidal volume is needed. The abdominal muscles, however, are used for additional expiratory force during strenuous exercise in both normal individuals and in patients with altered ventilatory mechanics from muscle weakness. They are important for coughing, sneezing, and raising intra-abdominal pressure.

2. *Alveolar-Capillary System*

Hundreds of millions of air chambers provide a semipermeable surface of about 1000 square feet where the exchange of oxygen and carbon dioxide takes place.

3. *Pulmonary Circulation*

This system maintains a flow of blood through the lungs, permitting the essential gas exchange to occur between the internal and external environment.

4. *Respiratory Control Center*

The control for respiration is located in the brain stem and contains cells which rhythmically discharge nerve impulses to the respiratory muscles. The frequency of discharge is controlled by the local arterial pH, the pCO_2, and by reflexes originating at peripheral chemoreceptors and muscle proprioceptors. The center maintains an arterial oxygen saturation of 95 per cent or higher, an arterial carbon dioxide pressure of 40 mm. Hg, and a pH of 7.40.

Etiology

Hypoventilation is practically always present in those chronic diseases which are accompanied by:

1. *Respiratory Muscular Dysfunction*

The primary and accessory muscles of respiration, both inspiratory as well as expiratory, are frequently affected by the weakness or incoordination of conditions such as Parkinson's syndrome, cerebral palsy, high spinal cord injuries,[2] multiple sclerosis, amyotrophic lateral sclerosis,[3] Guillain-Barré syndrome, myasthenia gravis, and muscular dystrophy.[4]

2. *Decreased Chest Wall Compliance*

Tightness of the soft tissues of the thorax, or rigidity of its bony framework, will limit chest excursion and increase the work of breathing. Aging, obesity, kyphoscoliosis, ankylosing spondylitis,[5] scleroderma, scarring from severe chest burns and postsurgical complications[6] all may cause a decrease in ventilatory efficiency.

3. *Decreased Pulmonary Compliance*

Inflammatory infiltration and fibrosis of the lung tissue itself will cause lung rigidity and stiffness. Collagen diseases, sarcoidosis, and diffuse interstitial fibrosis may cause hypoventilation by limiting lung tissue expansion.

4. *Increased Airway Resistance*

Interference with airflow by constriction or obstruction of the lower airway is a common cause of pulmonary insufficiency. Besides asthma, emphysema and chronic bronchitis, which always cause an increase in airway resistance, there are many other chronic conditions where bronchial secretions are retained or where there is a narrowing of the lumen of the bronchial tree. Cystic fibrosis, connective tissue disorders, or any disease which requires long periods of bed rest, or wheel chair confinement, may impair airflow by the presence of viscous mucus, bronchospasm or large amounts of secretions.

5. *Impaired Diffusion*

Poor air exchange is caused by the failure of the oxygen tension in the alveoli to reach equilibrium with the oxygen tension in the blood. Reduction of the diffusing capacity of oxygen is the main feature of the alveolar-capillary blockade syndrome. Pulmonary congestion and edema as well as chronic diseases associated with interstitial fibrosis reduce diffusion by increasing the thickness of the alveolar membrane. Pulmonary embolism

and thrombosis, and diseases associated with a vasculitis such as the collagen diseases, may reduce the size of the capillary bed so that the red cells are unable to remain in the gas exchange vessels sufficiently long to become fully saturated with oxygen.

6. *Defective Control of Ventilation*

Alterations in the brain stem usually adversely affect respiration. These respiratory center changes occur in conditions such as encephalitis, cerebrovascular disorders, brain injuries, and tumors. Obesity[7] and myxedema may also interfere with the central control of respiration.

Usually where hypoventilation does exist as a secondary complication of a chronic disease more than one of the above etiologic factors contribute to the condition. For example, in scleroderma there can be a decrease in both chest wall and pulmonary compliance, as well as an impairment in diffusion.

Clinical Manifestations

The symptoms of hypoventilation, though often present in chronically ill patients, are rarely recognized unless the respiratory impairment is severe and prolonged. Those with mild impairment will show only slight breathlessness during light activity. In others, headaches, lassitude, somnolence, and easy fatigability may be observed. The patient may talk with a low voice and may not be able to produce an effective cough or blow his nose or clear his throat efficiently. The mental confusion, irritability and poor memory, so frequently found in chronically ill patients, are in many instances the result of carbon dioxide narcosis. When the impairment is severe and prolonged, neurologic complications and heart failure will usually occur.

Clinical Assessment

The routine examination of the respiratory system, which includes auscultation, percussion, and roentgenography, may reveal a primary respiratory disease; however, unless specific respiratory function tests are performed in all chronically ill patients, secondary hypoventilation may go undetected.

Many of the tests which measure the mechanical efficiency of the respiratory system can easily be included in a general examination. These tests require a minimal amount of equipment, take little time to perform and can be done at the bedside or in the office.[8] A considerable amount of pertinent information can be obtained from the use of one or more of the following:

1. Manual measurement of the strength, coordination and endurance of the respiratory muscles.[9]
2. Measurements of chest expansion.
3. Measurement of range of mobility of the cervical spine, thoracic spine and shoulder girdle.
4. Measurement of breath holding time.
5. Snider's match ventilation test.[10]
6. Cough efficiency test.
7. Exercise tolerance tests.
8. Spirometry.[11]

When respiratory impairment is suspected further investigation is advisable.[12] More specific tests require a special respiratory function laboratory with facilities for ascertaining:

1. Eveness of the distribution of ventilation.
2. Oxygen content and saturation of the arterial blood.
3. Carbon dioxide tension.
4. Arterial blood pH.
5. Oxygen consumption, carbon dioxide ventilation rate, oxygen ventilation rate and heart rate at graded work loads.
6. Pulmonary blood distribution.

Management

Occasionally the appropriate management of the primary chronic disease may be in conflict with the proper management of its respiratory component. Since barbiturates, muscle relaxants, tranquilizers and sedatives are all respiratory depressants, it is important that respiratory tolerance to these drugs be evaluated on an individual basis. The dosage could then be adjusted or the respiratory effects be counteracted by the administration of other drugs such as amphetamines, nikethamide,[13] nicotinic acid, or by respiratory center stimulants.[14]

There are also specific drugs such as mucolytic enzymes and expectorants which will reduce the viscosity of the

secretions, and thereby improve the patency of the airways and lessen the work of breathing.

Occasionally surgery may be necessary to relieve chronic hypoventilation. A tracheostomy may be required in the more severely disabled person to assist ventilation, reduce the dead space, and permit the removal of bronchial secretions. In selected patients spinal fusion will allow for more efficient ventilatory mechanics. Muscle and fascial transplants, nerve anastomosis and pneumoperitoneum have all been used experimentally to increase breathing ability.

There are a variety of physical medicine technics which can be used to treat secondary hypoventilation. These methods of necessity must be prescribed on an individual basis, and in addition must not conflict with the treatment of the primary disease. Graded general exercise can be used to improve muscle tone, build strength and endurance, and stimulate both circulation and respiration.

Postural exercises can be aimed at maintaining the thorax at its optimum position of efficiency. Mobilization and stretching of tight or rigid joints and soft tissues will allow for increased chest excursion and improve compliance. Special breathing technics such as diaphragmatic, abdominal or segmental breathing, will strengthen and coordinate the diaphragm, increase the power of exhalation, and ventilate selected lung areas. Exercises to increase the power of the accessory muscles of respiration will provide supplemental ventilatory force where the primary muscles are weak or inefficient. Glossopharyngeal breathing can inflate the lungs with a sufficient volume of air to allow for clearing of bronchial secretions. It can also increase tidal volume and vital capacity by improving pulmonary and chest wall compliance. Postural drainage will help remove bronchial secretions and maintain airway patency.

When the respiratory impairment is progressive or fails to respond to treatment, it then becomes necessary to reduce the patient's energy expenditure to a level that is consistent with his physical capacity. Therefore, it is important to analyze the activities which the patient performs and to eliminate all but the essential ones. Work simplification and work-saving devices may all be needed to help reduce the oxygen demands while on the job or while performing the activities of daily living. Mechanical respiratory aids such as respirators, intermittent positive pressure machines, rocking beds, pneumobelts,[15] and mechanical cough stimulators, which are commonly used during the acute respiratory failure in diseases such as poliomyelitis[16] and myasthenia gravis, may also be needed for chronically ill patients with severe hypoventilation. These aids are often essential when an upper respiratory infection develops. They should also be considered when the patient is febrile, in very hot weather, during surgery and in the postsurgical period.

Summary

Respiratory impairment, as a secondary complication of many chronic diseases, may often remain undetected and its early symptoms may be overlooked or attributed to the primary disease.

Conditions which tend to reduce mobility, such as ankylosing spondylitis, scleroderma and kyphoscoliosis, may limit ventilation by decreasing chest wall compliance. When the primary disease causes generalized muscle weakness, incoordination or paralysis, hypoventilation will also result. The respiratory center itself may be adversely affected by brain injuries, drugs, encephalitis or tumors.

Clinical manifestations include a number of somewhat vague complaints, but if the physician makes the appropriate tests, sufficient objective findings will clearly point to hypoventilation whenever it is present. Ascertaining pulmonary efficiency is certainly as important as determining cardiovascular status.

Management may include medical and surgical procedures to prevent further respiratory impairment or to improve a failing respiratory system. Special exercises and technics can be aimed at increasing ventilation. Mechanical assistance can give the necessary respiratory support to patients who are undergoing rehabilitation.

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