Technical Article:

PULMONARY EMPHYSEMA

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Pulmonary emphysema can be defined as a condition of distention of the alveoli and can, be classified as follows:

Classification of Pulmonary Emphysema

(1) Large Lunged or Hypertrophic
(2) Small Lunged or atrophic
(3) Compensatory
(4) Acute Vesticular

Large lunged or hypertrophic emphysema is by far the most important and consideration of its natural history, pathophysiology, complications and management will be the purpose of this communication. The remaining types will be considered very briefly indeed.

Atrophic emphysema is not associated with airway obstruction, is seen in a few old people, is symptomless, and may be considered a variant of the aging process.

Compensatory emphysema compensates space only — not function. If a lobe of lung be removed or becomes shrunken to a fraction of its normal size by atelectasis or fibrosis the space it formerly occupied is filled by the adjacent lung which is stretched to fill the space. This increases the pull on the mediastinum which is drawn to the affected side.

Acute vesicular emphysema occurs during acute asthmatic attacks. The lungs may be quite ballooned by air but when the attack subsides, the lungs return to their normal state.

For a proper understanding of the natural history, symptoms and, hence, the management of hypertrophic emphysema, a review of some of the features of the physiology of normal breathing and the pathophysiology of emphysema is desirable.

On normal inspiration the thoracic cage enlarges and in so doing, enlarges the lungs which can be considered as two elastic bodies stretched, from the time of the first infant breath, between the mediastinum and the chest wall. Although mildly stretched on expiration, the lungs are further stretched by inspiration and the recoiling force of the stretched lung accounts for the sub-atmospheric intra-pleural pressure greater on inspiration than on expiration.

Inspiration is a powerful active movement produced by contraction of the diaphragm and other inspiratory muscles. The lungs are further stretched with resultant enlargement of the air sacs, dilatation and elongation of the bronchi. Normal expiration on the contrary is a weak passive movement largely accomplished by the elastic recoil of the lungs and results in decrease in size of the air sacs, shortening and narrowing of the bronchi.

Forced inspiration is an exaggeration of normal inspiration but forced expiration is accomplished by the abdominal and other expiratory muscles forcing the diaphragm up and the ribs down and in. Force is therefore exerted on the alveoli and bronchi from without, tending to compress them and empty them of air. In the absence of airway obstruction there is no dilating force exerted on the alveoli. Thus, in the absence of other factors, the blowing of wind instruments cannot cause any dilating force on the alveoli.

If there is narrowing of the airway from any cause the strong inspiratory movement will largely overcome the impediment but the weak normal expiratory force is inadequate to empty the air sacs. The obstruction is further aggravated in expiration by the expected narrowing of the bronchus which has been described. The result is gradual ballooning of the alveoli distal to the obstruction. Such air trapping may be overcome for a while by forced expiration but if the narrowing persists,

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the lungs become enlarged as seen in a temporary and reversible form in the acute asthmatic attack. While such narrowing is at the level of the bronchioles in the asthmatic it could occur at any level in the bronchial tree.

If, to airway obstruction at any level weakening of the bronchial wall is added, the situation is aggravated. During forced expiration the bronchial obstruction is now increased not only by the expected narrowing of the bronchi but by the collapse of the weakened wall which virtually produces a flap valve permitting entry of air on inspiration but grossly impeding its exit on expiration.

It is of interest to note that chronic pulmonary emphysema (hypertrophic emphysema) does not develop in uncomplicated asthma. It is found in patients with chronic bronchitis, asthmatics who develop bronchitis, and in a few cases with distal airway obstruction without evidence of either bronchitis or asthma, so called primary emphysema.

Herzog[1,2] in Europe, Robert Fraser[3] in Montreal and Rainer[4,5] and his colleagues in Denver, Colorado have drawn attention to the weakening of the walls of the trachea and main stem bronchi in some patients with emphysema. They have demonstrated the collapse of these weakened tubes on forced expiration and the consequent severe airway obstruction. Most recently Makelem, Fraser and Bates[6] have demonstrated a similar but very localized defect in the lobar bronchi and their main branches of some emphysematous patients. While more difficult to demonstrate, it appears not unlikely that a similar situation may develop in much smaller ramifications of the bronchial tree and produce similar severe airway obstruction. It seems probable that a major factor in emphysema is weakening of the bronchial walls at various levels with consequent flap valve collapse of the bronchial wall on forced expiration.

While airway obstruction due to weakening of the bronchial walls at various levels is an important factor in emphysema and is irreversible, nevertheless it must be remembered that obstruction of smaller air passages by spasm, edema or viscid secretions are major contributing factors which are amenable to treatment.

Along with the ballooning and rupture of alveoli, the consequences of airway obstruction are far reaching. Its first result is uncomplicated pulmonary emphysema. The patient in this early stage of the disease will present the symptoms of the underlying cause of the disease, bronchitis or asthma complicated by bronchitis and in addition, will have some shortness of breath on exertion even between attacks of asthma or bronchitis.

Although many physical signs of emphysema are described such as hyperresonance on percussion, increased antero-posterior diameter of chest, mid-dorsal kyphosis, diminished chest expansion, indrawing of the sub costal margin on inspiration, distant breath sounds or harsh vesicular breath sounds with prolonged expiratory phase, the sine qua non in the clinical diagnosis of emphysema is the demonstration of big lungs. This can only be done on physical examination by demonstrating low lung borders. It is accomplished easily by percussion at the lower borders posteriorly, at the lower border of the right lung anteriorly and by demonstrating the encroachment of the enlarged lung on the area of superficial cardiac dullness. The physical signs of airway obstruction to expiration are prolonged expiration, expiratory rhonchi and ballooned lungs.

From the laboratory side, the most valuable evidence of airway obstruction is reduction in the one second timed vital capacity which gives an indication of airway obstruction to expiration and of air trapping. Normally 75% of the total vital capacity is expelled in the first second. This is reduced in proportion to airway obstruction. Radiological examination at this stage is likely to be uncertain. The diaphragms may appear low but since chest X-rays are taken during full inspiration, this is unreliable. The lateral plate showing an increased area of translucency between the heart and sternum is perhaps the most valuable indication. Radiolucency, slender vertical heart shadow, lessened obliquity of the ribs, widened intercostal spaces, increased antero-posterior diameter and mid-dorsal kyphosis may be present in the more advanced stages.

The early diagnosis of emphysema will be made by the physician who maintains a sharp watch on patients who have recurrent attacks of bronchitis or asthma and utilizes all aids to detect the disease. It is evident that patients will be seen who have chronic airway obstruction without emphysema. That the exact recognition of early emphysema is difficult is indicated by the poor correlation of

clinical with post mortem findings. The important thing is to consider all patients with chronic airway obstruction as in danger of developing emphysema and treating them accordingly. At all stages of the disease resulting from airway obstruction the aim should be prevention of progression. The importance of early diagnosis and treatment lies in the opportunity provided for such prevention or delay of the progression of the disease. In the untreated or unsuccessfully treated patient the progression is remorseless.

Airway obstruction due to { *Bronchospasm*, *Bronchial Edema*, *Bronchial Secretions*, *Bronchial Collapse*

Air Trapping

leads to *Maldistribution of air*

and *Impaired perfusion due to compression of pulmonary capillaries and increased vascular resistance*

The result is an abnormal ventilation-perfusion ratio which gives rise to inadequate oxygenation of the blood and to retention of carbon dioxide - the condition of respiratory acidosis.

This clinical state can be recognized by the altered function produced. Hypoxia results in an increase in the tone of the small pulmonary arterioles with resultant pulmonary hypertension which aggravates the perfusion defect already present and increases the load on the right ventricle. The process is accentuated by hypercarbia. The effect of hypoxia on the brain is to cause drowsiness, confusion, hallucinations, papilledema and eventually coma. All these sysmptoms may be confused with primary neurological disease. When papilledema is present, which is not uncommon, the problem is even greater.

Hypercarbia, in addition to potentiating the effect of hypoxia on the pulmonary circulation, causes a profound vasodilatation of the systemic arterioles which results in increased venous return, increased cardiac filling and increased output per beat. The high output state thus produced can be recognized by warm, pink extremities, increased digital throb, capillary pulsation, full arm veins, water hammer pulse, high systolic blood pressure with low diastolic pressure, active left ventricle.

In the presence of chronic hypercarbia the respiratory centre becomes insensitive to stimulation by carbon dioxide - the hypoxia alone remains as a stimulus to respiration. If in such circumstances oxygen is given by tent, mask, or nasal catheter the hypoxic stimulus is removed, the shallow respirations characteristic of respiratory acidosis will cease and unless the situation is rapidly corrected coma and death will follow.

Laboratory examination will reveal a low pO_2, high p CO_2 and high bicarbonate. In those laboratories lacking the necessary equipment for these valuable tests the recognition of the combination of a high carbon dioxide combining power with an acid should help in diagnosis.

In patients where respiratory acidosis does not lead rapidly to coma and death but persists in less severe degree the hypoxia and hypercarbia persist in chronic form and so also do their effects on the pulmonary and systemic arterioles. While there is general agreement that hypoxia is a cause of pulmonary hypertension and that this effect is exaggerated by hypercarbia, there is some confusion in the literature with regard to the effects of respiratory acidosis on the systemic circulation. There is no doubt that the inhalation of carbon dioxide by the normal individual causes peripheral vasodilatation, all the clinical features of the high output state and that cardiac output per beat by actual measurement is increased. The high output state will not of itself cause right ventricular failure as this chamber handles increased volume well. It is, however, a poor force pump and working against high pulmonary resistance will hypertrophy and eventually fail. It may do so while the output of the left ventricle is still high and so present the picture of high output failure. Eventually the output of the left ventricle will decline to less than normal so that depending on when the determination is made high output or low left ventricle output may be found in patients with cor pulmonale and right ventricular failure.

The real cause of right ventricular failure in these patients is pulmonary hypertension due to increased vascular resistance. Since part of the increase is caused by increased

arteriolar tone, due to hypoxia and hypercarbia and hence reversible - it is important to remember this when considering treatment.

Another phenomenon which must be considered is the development of polycythemia in a small minority of patients with chronic respiratory failure. Although it is well recognized that when healthy individuals live at altitudes of 10,000 ft. or more above sea level, polycythemia develops, most patients with emphysema, even though hypoxia may be greater, do not develop polycythemia. This facet of the disease requires further study to determine if the stimulus to red cell production provided by hypoxia is countered by acidosis either by interference with the production of erthropoietin by the kidney or blocking the effect of erthropoietin on the bone marrow. Yet another explanation might lie in the masking of the increased red cell mass by increased plasma volume or the cause may lie in other as yet underterminal factors.

Treatment

The progression of the disease is clear and untreated tends to be remorseless. All too often the physician considering the problems presented by such a patient is overcome by a sense of hopelessness which is conveyed all too easily to the patient. There is no place for despair. To treat this condition successfully a physician knowledgeable in the pathogenesis and natural history of the disease and the factors which influence it adversely must carefully assess the status of his patient and then exert all his skill to guide his patient's life into paths which will achieve maximum pulmonary function and retard progression of the disease to the greatest degree possible. Much can be achieved and the patient should be brought to an appreciation of this fact.

In the early stages respiratory failure is not a problem and the physician's efforts should be directed toward gaining all knowledge possible of the underlying causes of the disease and attempting to relieve them.

If bronchitis is at the root of the trouble efforts to protect the patient from recurrent respiratory infections should be made and should include vaccination against those respiratory viruses for which effective vaccines are available. These include the influenza viruses group A, A1, A2 Asian and P.R. 8 (Porto Rican 8), influenza B1 and adenoviruses No. 3, No. 4 and No. 7. Such measures cannot be expected to prevent all attacks of bronchitis. The common cold in an otherwise healthy individual is relatively innocuous but in a patient with emphysema may easily precipitate bronchitis, increased airway obstruction and all its sequelae. When this happens, it is important to determine as soon as possible the presence of secondary bacterial invaders and deal with them by the exhibition of suitable antibiotics. There is now ample evidence to indicate that the prophylactic use of antibiotics in these patients is inadvisable. Where broad spectrum antibiotics are used to lessen the incidence of infection from non-specific organisms they have failed. The incidence of infection has not been reduced - only the type of the infecting organism is changed to one resistant to the particular drug used. A widespread custom of advising the patient to start antibiotic therapy when the sputum becomes yellow or green falls in between the two programs just discussed. Green or yellow sputum cannot be equated with bacterial infection. Such sputum occurs in virus pneumonia without the presence of significant secondary invaders. In the writer's opinion any procedure short of determining the invading organism and dealing with it falls far short of ideal treatment. This does not mean undue delay since once the sputum sample has been obtained treatment with penicillin can be started pending report on the organism present and its sensitivity.

The removal of lung irritants provides the next most important means of treatment. While in a few cases industrial fumes, dust or smog may be at fault, by far the commonest offender is tobacco smoke. Too often the patient will not accept advice in regard to giving up tobacco smoking but nevertheless it remains the physician's responsibility to indicate clearly to his patient that tobacco smoking is nothing more or less than a self-inflicted wound to the patient with emphysema. If need be, this can be demonstrated easily by doing a timed vital capacity before and after the inhalation of a few puffs of tobacco smoke. It has been shown repeatedly that cessation of smoking has resulted in decreased cough, sputum, wheezing, and dysponea and improved pulmonary function.

Bronchodilator drugs should be tried whenever evidence of airway obstruction is found. If a pulmonary function laboratory is available, the most effective drug for the

individual concerned can be determined readily. Bronchodilators can be given by mouth, per rectum, by inhalation or by parenteral injection. In some instances their administration by using a nebulizer combined with a positive pressure respirator is highly effective.

Postural drainage, particularly following bronchodilators on rising in the morning, often does much to provide the patient with a comfortable day and of course lessens the chance of serious pulmonary infections.

Patients in whom asthma is the underlying cause have already developed complicating bronchitis with damage to the bronchial walls before emphysema develops. Thus all the measures so far discussed will apply to them too, but obviously treatment of the asthmatic should be directed first to the prevention of bronchitis. If an extrinsic allergen is concerned, desenitization, elimination of the allergen from the environment as with cat hair, or the patient from the allergen containing atmosphere as with rag weed pollen are important measures. In rare instances corticosteroid therapy may be indicated.

Environment can sometimes be altered to the patient's benefit. Very few can uproot themselves and move to a dry climate like Arizona. But people living in cold damp houses can benefit from finding more suitable quarters. In the periods of exacerbation the provision of hospital rooms with controlled ventilation, with filtered air and controlled temperature and humidity has been shown to shorten the patient's stay.

Intelligent vigorous treatment combined with encouragement of the patient and inspiring in him a mood of confidence and optimism will do much to retard the progression of the disease and lessen the disability which it causes. Nevertheless, physicians will continue to be confronted with patients who have progressed to respiratory failure.

The treatment of respiratory acidosis provides one of the medical emergencies. As noted these patients suffer from hypoxia and hypercarbia caused by an exacerbation of their disease precipitated by one of the factors already discussed. Treatment must be directed toward keeping the patient alive until the precipitating factor - usually respiratory infection, can be controlled.

Respiratory stimulants of which nikethamide (Coramine - Ciba or Nikorin - Massenger) is the best, may be helpful in a few cases. Early enthusiasm for the use of carbonic anhydrase inhibitors has not been justified - the additional risk of adding a metabolic acidosis to the respiratory acidosis already present has proven to be too great a hazard.

The danger of the administration of oxygen by tent, mask, or nasal catheter has already been indicated. In this situation a means of delivering oxygen and removing carbon dioxide which is not dependent on the patient's own breathing must be provided. This means the use of some type of positive pressure respirator. It is not within the scope of this paper or the knowledge of the writer to discuss the virtues or inadequacies of the various available respirators. The Engstrom respirator in the experience of the Halifax group, is the most efficient - but costs nearly $5,000.

While a cuffed endotrachial tube introduced orally may be adequate in some patients, in many, tracheostomy is life saving. Not only does it reduce the physiological dead space and permit more adequate bronchial toilet, but it also protects the patient against the serious effects of laryngeal spasm induced by the irritation of secretions acting on a larynx which is in the same state of irritability as is seen in certain levels of pentothal anesthesia or certain levels of unconsciousness following head injury.

The treatment of cor pulmonale which is in effect right ventricular failure characterized by increased jugular venous pressure, enlargement of the liver, ascites and edema demands skill and knowledge on the part of the physician. Too often all efforts are directed toward treatment of the heart failure by conventional methods without considering and treating the underlying cause. It is true that digitalis may be of some value and that restriction of salt and the exhibition of diuretics may be helpful in ridding the body of excess water and sodium, but it must be remembered that the underlying cause of the right ventricular failure is lung failure with hypoxaemia and hypercarbia which has resulted in pulmonary hypertension. The situation then is a combination of heart failure and lung failure and treatment must be directed to correcting both. The correction of hypoxia and hypercarbia will do more to

correct right ventricular failure in such patients than will any other measure.

Polycythemia when found should not be regarded as a beneficial compensating measure but as a sequel to chronic lung failure developing in some patients for reasons as yet not clearly understood. The increased viscosity of the blood adds to the circulation difficulties and indicates its correction by repeated venesection.

The development of pulmonary treatment units in hospitals is progressing rapidly. In such units a trained staff is able to look after several patients in respiratory failure more efficiently than would be the case if each were in a private room. The need for trained personnel to work in such units provides a problem which must be faced in common with the provision of trained personnel for many other hospital activities. The physician's first thought is likely to be to ask the nursing service to provide such personnel. But is it necessary to take people who have already had three years training and will now require further training in this special work? It is submitted that the real training required is a knowledge of sterile technique, a knowledge of the respirators and other equipment used, laboratory training in the tests used in the unit and experience on the job. All this will require much less than three years and will spare nurses for the work for which they are trained.

In conclusion it is again emphasized that much can be done for the patient suffering from pulmonary emphysema if the physician deals with his patient with confidence based on knowledge and the optimism which the results justify.

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