So-Called Hypertrophic Emphysema

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Before discussing the pathogenesis and clinical aspects of this disease, clarification of some of the pertinent terminological definitions may be in order. Hypertrophic emphysema is the most commonly used designation of this disease. Also, it is known under the following names: genuine emphysema, idiopathic emphysema, diffuse vesicular emphysema, obstructive emphysema, dystrophic emphysema, pulmonary hypertrophy, pulmonary dilatation, chronic large lung, and pneumonectasis. Inasmuch as loss of the elastic fibers and more or less extensive destruction of the alveoli are cardinal aspects of this disease, one may designate it as destructive emphysema. Still a more appropriate term, pseudohypertrophic emphysema, may be offered for the nomenclature of this condition. Hypertrophy of the lung in genuine emphysema is far from being similar to muscular hypertrophy of the extremities seen in men engaged in athletics or in occupations which entail heavy physical work. Although in genuine emphysema the lung is larger than normal, there is neither an increase in the number of functionally competent alveoli nor a proportionate augmentation in the number, size and functional capacity of corresponding capillary vessels.

During the course of development of genuine emphysema a great many interalveolar septums are destroyed. Consequently, large, cyst-like air spaces appear in the lung. Cyst-like formations of this type are designated as blebs when they are localized subpleurally. The ones localized in other parts of the lung are conventionally referred to as bullae.

Concepts vary relative to the pathogenesis of genuine emphysema. I am of the opinion that the following factors are of cardinal importance in this respect: (1) Increased intrapulmonary pressure during strenuous coughing. (2) Infections and other pathologic changes which result in extensive degenerative alterations in the elastic elements of the lung. (3) Partial bronchial occlusion of the check-valve type. Familiarity with the role of each of these items in the pathomechanics of genuine emphysema is helpful in the satisfactory management of this disease.

During the compressive phase of cough which is strenuous, the intrapulmonary pressure may be as high as from 80 to 200 mm. of mercury over and above atmospheric pressure. This aerodynamic trauma, when sustained for a prolonged period of time, is likely unduly to stretch and to damage the elastic fibers of the lung.

Chronic lung infections may lead to destructive changes in the elastic elements of the lung in two ways: (1) through direct toxic influences; (2) through consequent fibrosis. The latter causes constriction or complete obliteration of some of the nutritional blood vessels. The consequent diminished blood flow exerts an untoward influence upon the implicated lung
structures. Similar sequels may follow prolonged, massive inhalation of noxious fumes, gases and dusts.

Widespread spasm of the peribronchial and peribronchiolar smooth muscles is of more common occurrence than generally realized. Its presence in allergic bronchial asthma is well known. Also, it may be provoked by infections of the lower respiratory tract, by inflammatory hyperemia, by congestion due to failure of the left ventricle and by extensive pulmonary fibrosis. Bronchospasm, particularly in the presence of congestion and edema of the bronchial mucosa and instances where inflammatory exudate narrows the bronchial lumen, is likely to set up a check-valve mechanism. The latter permits the ingress of air to parts of the lung distal to the partial occlusion. Inasmuch as the egress of the entrapped air is prevented, there develops in widespread areas of the lung what may appropriately be called pneumatic alveolar hypertension.

The deleterious effect of pneumatic alveolar hypertension can be more readily appreciated if the following changes are borne in mind. (1) Air currents are subject to the same physical laws as water currents. They move from a site of higher pressure toward areas of lower pressure. While during quiet respiration the intraalveolar pressure cannot be higher than the atmospheric pressure which prevails in the bronchi, during the compressive phase of strenuous coughing the intraalveolar pressure may rise to 200 mm. of mercury over and above atmospheric pressure. (2) The physical law of communicating vessels applies to the relationship between respiratory bronchioles and their respective cluster of alveoli. The same pressure which exists in the narrow respiratory bronchiole is transmitted undiminished to the entire perimeter of all of the respective alveoli. (3) At the termination of the compressive phase of cough, when there is a precipitous drop in the intrapulmonary pressure, evacuation of the air is slower from alveoli attached to spastic, partially occluded bronchi than from alveoli connected to bronchi of normal lumen. This exerts an appreciable distending influence upon the alveoli implicated.

The normal configuration and size of the thorax are dependent on anatomic as well as on physiologic factors. The physiologically stretched out position of the lung is maintained (1) by the pressure of the atmospheric air; (2) by the centrifugal suction effect of the intrapleural negative pressure. The thoracic cage is larger than the lung. Even so, the lung is held in apposition to the inner surface of the chest wall by the traction of the intrapleural negative pressure. The latter is an expression of the difference between the pressure of the atmospheric air reaching the lung through the lower air passages and the innate centripetal (hilusward) retractility of the elastic elements of this organ.

When as the result of loss of the elastic elements of the lung, its centripetal (hilusward) contractility is greatly reduced, there is a proportionate decrease in or complete disappearance of the negativity of the intrapleural pressure. The inspiratory muscles of the chest wall, not being obliged to counteract the inward pull of the intrapleural negative pressure, are bound to distend the thoracic cage. This train of events is similar to that seen
when initial artificial pneumothorax is given. It is obvious, therefore, that
distention of the thorax as well as the enlargement of the lung in genuine
emphysema are brought about by the same factor, namely by the loss of
the elastic contractility of the lung.

The normal position of the diaphragm is the direct result of the upward
traction force of the intrapleural negative pressure. In genuine emphysema,
in consequence of the disappearance of the upward traction of the intra-
pleural pressure, the diaphragm occupies a constant low (inspiratory)
position. Clinically, it is easily demonstrable that the diaphragm occupies
a low position after the establishment of artificial pneumothorax. Also, it
is known that the respiratory excursions of the diaphragm are restricted
following the institution of artificial pneumothorax. The reason is that
in an abnormally low position the diaphragm is functionally handicapped
or completely defunctionalized. In emphysema, its respiratory motions are
slight, absent or may be paradoxical: rising on inspiration and descending
on expiration.

In addition to impaired diaphragmatic function, the following functional
changes are noted in so-called hypertrophic emphysema: (1) Lessened
negativity of the intrapleural pressure. (2) Decreased respiratory excursions
of the ribs. (3) Diminished return of venous blood to the right auricle
because of the less negative intrathoracic pressure. Consequently, the
venous pressure is elevated. (4) Decrease in or lack of dilatation of the
pulmonary vascular bed on inspiration. (5) Pulmonary hypertension.
(6) The pulmonary tidal air is normal or slightly reduced. (7) The com-
plemental air is reduced. (8) The functional residual air is from two to
three times that of normal. (9) The maximum breathing capacity is re-
duced. The reduction may be as much as 50 per cent or more. (10) The
vital capacity of the lung may be lowered by from 20 to 60 per cent.
(11) Carbon dioxide content of the alveoli is increased to 7-8 per cent
(50-60 mm. of mercury). (12) Oxygen saturation of the blood is below
normal. It may be as low as 60 per cent. (13) There is an increase in the
bicarbonate reserve in the blood. (14) Plasma chlorides are decreased.
(15) Polycythemia is present without pathologic bone marrow changes.
(16) The size of the erythrocytes is increased.

So as to avoid misguided therapeutic intervention, I wish to present
the salient differential diagnostic aspects of genuine emphysema and senile
emphysema. The term emphysema is derived from the Greek words which
mean "in" and "to blow". Applied to the lung, this term signifies inflated
lung. According to the derivation of this term, senile emphysema is a
misnomer, for in this condition the lung is small, with the exception of
the Kountz-Alexander type of senile emphysema. On postmortem exam-
ination when the chest is opened, the lung collapses in senile emphysema.
Some of the alveoli are dilated in this condition. In the aged, the alveoli
become flabby like the skin. The consequent dilatation of the alveoli is
the result of atrophic changes in the alveolar septums, corresponding to
the age of the individual. Similar degenerative changes prevail in all of
the senescent body tissues. Simultaneously, some of the elasticity of the
lungs is lost. But it is to be kept in mind that this loss is proportionate to the loss of the tone and strength of the respiratory muscles of the chest. Consequently, the normal functional balance remains undisturbed between the centripetal (hilarward) traction of the pulmonary elastic fibers and the centrifugal traction of the intrapleural negative pressure. The latter is actuated by the outward pull of the inspiratory muscles of the chest.

### Differential Diagnosis

<table>
<thead>
<tr>
<th>Size of the Chest</th>
<th>Pseudonyctrophitic Emphysema Increased</th>
<th>Senile Emphysema Not Increased*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shape of the chest</td>
<td>Barrel-like or box-like</td>
<td>Small, flat*</td>
</tr>
<tr>
<td>Position of ribs</td>
<td>Horizontal</td>
<td>Oblique*</td>
</tr>
<tr>
<td>Position of sternum</td>
<td>Elevated</td>
<td>Not elevated*</td>
</tr>
<tr>
<td>Intercostal spaces</td>
<td>Widened</td>
<td>Narrowed*</td>
</tr>
<tr>
<td>Dorsal spine</td>
<td>Kyphosis</td>
<td>Straight or convex</td>
</tr>
<tr>
<td>Neck</td>
<td>Short</td>
<td>Normal*</td>
</tr>
<tr>
<td>Shoulders</td>
<td>Thrown forward</td>
<td>Normal*</td>
</tr>
<tr>
<td>Accessory respiratory muscles</td>
<td>Visible function</td>
<td>No hyperfunction</td>
</tr>
<tr>
<td>Epigastric angle</td>
<td>Widened</td>
<td>Normal or narrowed</td>
</tr>
<tr>
<td>Motion of epigastrum</td>
<td>Not protruding on inspir.</td>
<td>Inspir. protrusion</td>
</tr>
<tr>
<td>Pulsation of epigastrum</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Motion of lower ribs</td>
<td>Inward on inspiration</td>
<td>Normal</td>
</tr>
<tr>
<td>Diaphragm: position</td>
<td>Low</td>
<td>Normal</td>
</tr>
<tr>
<td>Diaphragm: excursions</td>
<td>Decreased or absent</td>
<td>Normal</td>
</tr>
<tr>
<td>Type of respiration</td>
<td>Thoracic</td>
<td>Abdominal</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>Present</td>
<td>Infrequent or absent</td>
</tr>
<tr>
<td>X-ray: Costophrenic sinus</td>
<td>Shallow</td>
<td>Normal*</td>
</tr>
<tr>
<td>X-ray: Translucency</td>
<td>Increased</td>
<td>Normal or slight incr.</td>
</tr>
<tr>
<td>X-ray: Lung behind sternum</td>
<td>Increased</td>
<td>Not increased*</td>
</tr>
<tr>
<td>X-ray: Vascular markings</td>
<td>Indistinct</td>
<td>Normal or increased</td>
</tr>
<tr>
<td>X-ray: Shape of sternum</td>
<td>Anterior convexity</td>
<td>Normal</td>
</tr>
<tr>
<td>X-ray: A-P diameter of chest</td>
<td>Increased</td>
<td>Not increased*</td>
</tr>
<tr>
<td>Pulmonary function</td>
<td>Insufficiency</td>
<td>Normal</td>
</tr>
<tr>
<td>Arterial oxygen saturation</td>
<td>Low</td>
<td>Normal</td>
</tr>
<tr>
<td>Arterial CO₂ tension</td>
<td>High</td>
<td>Normal</td>
</tr>
<tr>
<td>Signs of right heart failure</td>
<td>Often present</td>
<td>Absent</td>
</tr>
<tr>
<td>Venous pressure</td>
<td>Increased</td>
<td>Normal</td>
</tr>
</tbody>
</table>

*Except in the Kountz-Alexander type of emphysema.
wall. In senile emphysema the respiratory excursions of the chest are reduced also because in the aged there are calcification of the costochondral junction, limitation in motion of the costovertebral joints, fibrosis and loss of elasticity of the thoracic ligaments, loss of elasticity of the bronchi due to calcification of their walls, possible pulmonary arteriosclerosis and concurrent heart failure. Pulmonary congestion associated with heart failure causes a decrease in the expansability of the lung.

The Kountz-Alexander type of emphysemas, according to the original definition of these clinicians, is not primarily a pulmonary disease. Changes in the lung are secondary to an increase in the size of the thoracic cage. One of the characteristic features of this condition is straightening and stiffness of the thoracic spine. These changes are brought about by degenerative alterations in the intervertebral disks. The nucleus or the entire disk becomes swollen. In the advanced degenerative phases some of the intervertebral disks become completely separated from the bone and undergo complete dissolution. With the progression of degenerative changes, the corresponding vertebral bodies become implicated, they thin out and finally, kyphosis may result. This, in turn, will lead to an outward flare of the ribs and to the development of barrel chest. The lung passively follows the distention of the thorax and thus it becomes somewhat enlarged in volume.

**Treatment**

It is almost a platitude to say that we as a nation are growing older. Since the turn of the century the population of the United States doubled, while the number of aged quadrupled. Today 8 per cent of the population, that is, more than 12 million out of 156 million, are over the age of 65. With this constantly lengthening life span, it is anticipated that the number of persons over 65 years of age will reach more than 13 per cent of the population of this country at the end of this century. All this being so and also, bearing in mind the pathogenesis of so-called hypertrophic emphysema, more and more instances of this disease are bound to come to the attention of practicing physicians.

Since mass x-ray surveys, preemployment x-ray examinations of the chest and chest roentgenograms on admission to general hospitals are accepted by the medical profession as well as by the public, it is justifiable to point out the possible recognition of subclinical forms of this disease. With its early detection, appropriate measures may be instituted for obviating its progression. At the same time, development of complications may be prevented.

In my personal experience, I have found artificial pneumoperitoneum a highly satisfactory method in the management of so-called hypertrophic emphysema. Technical details of this procedure are presented in the author's monograph, "Pneumoperitoneum Treatment". In addition, four points deserve particular emphasis.

1) The amount of air given with each treatment should be less than 1,000 cc. Larger amounts of air are bound to cause limitation in motion of the diaphragm. Thus the treatment would defeat its own purpose.
2) Pneumoperitoneum treatment can be started in the office or in the clinic. "Refills" are given at weekly intervals.

3) The patient should wear a snugly fitting abdominal girdle night and day. Elastic segments of the girdle should be small. Metal plates, padding or inflatable compartments are not essential. The girdle should not interfere with the respiratory motion of the lower ribs.

4) Artificial pneumoperitoneum is well tolerated by the patient, provided no extensive peritoneal adhesions are present. Treatments may be continued for years.

The following factors are responsible for the beneficial results of artificial pneumoperitoneum:

1) Artificial elevation of the diaphragm refunctionalizes this previously defunctionalized muscle.

2) Pneumoperitoneum emancipates the diaphragm from the gravitational pull of the liver and of the spleen.

3) Increased intraperitoneal pressure caused by pneumoperitoneum is associated with reflex relaxation of the peribronchial spastic smooth muscles.

4) In so-called hypertrophic emphysema, artificial pneumoperitoneum renders the intrapleural pressure more negative. This greater negativity draws the diaphragm upward and the chest wall inward. Thus the function of the corresponding inspiratory muscles is improved.

5) Pulmonary pneumatic dyskinesia (faulty distribution of the inhaled air) is corrected. Consequently, the oxygen-carbon dioxide exchange is improved.

6) Oxygen saturation of the arterial blood is increased and its carbon dioxide concentration is decreased.

7) The increased negativity of the intrathoracic pressure facilitates the return of venous blood from the periphery of the greater circulation to the heart, with consequent decrease in the venous pressure.

8) There is increased blood flow from the heart to the lung. This relieves strain on the right ventricle.

9) Cough mechanism is improved.

While my own experience as well as the findings of others offer convincing evidence of the efficacy of pneumoperitoneum in the management of so-called hypertrophic emphysema, this treatment should not be looked upon as a cure-all. Artificial pneumoperitoneum is of limited value or of no value at all in the following instances:

1) When there is anatomically and functionally irreversible, extensive loss of alveoli and elastic elements of the lung.

2) When adhesions prevent the elevation of the diaphragm.

3) When there is atrophy of disuse of the diaphragm in long standing emphysema.

4) In cases of heart failure which cannot be corrected.

5) When uncontrollable complications interfere with cardio-respiratory function.

While I advocate artificial pneumoperitoneum as the method of choice
for the treatment of genuine emphysema, I do recommend, whenever circumstances so require, the use of antimicrobial drugs, chemotherapeutic agents, bronchorelaxant medicaments, hyposensitization, the administration of digitalis preparations and diuretics and other forms of supportive and specific treatment.

For completeness' sake, I wish to mention other methods which have been advocated for the treatment of this condition.

1) Expiratory pressure breathing with pursed lips was first recommended by Schutz as a purposeful therapeutic measure in 1935.

2) Oxygen in gradually increased concentrations given by inhalation was first advocated by Barach in 1938.

3) Intermittent positive pressure breathing of oxygen was first introduced by Motley and his associates.

4) Manual compression of the lower anterior part of the chest, the upper part of the abdomen or both, rhythmically at intervals corresponding to the expiratory phase of the respiratory cycle, has been advocated by clinicians in this country and abroad during the past few decades.

SUMMARY

A review of the pathomechanics of so-called hypertrophic emphysema qualifies artificial pneumoperitoneum as a logical, preferential method of treatment of this disease.

RESUMEN

Una revisión de la mecánica patológica del llamado enfisema hipertrófico permite calificar al neumoperitoneo artificial como el método de elección para el tratamiento de esta enfermedad.

RESUME

L'auteur, après avoir passé en revue les facteurs physio-pathologiques de l'emphysème que l'on qualifie d'hypertrophique, montrent qu'ils permettent de considérer le pneumopéritone comme la méthode de traitement logique et supérieure à toute autre.

REFERENCES


