INDICATIONS FOR URGENT OPERATION IN PULMONARY TENSION DISORDERS IN CHILDHOOD*

BY

I. SZÖTS and T. JAKAB

From Second Surgical Clinic of University of Budapest, Hungary

Pulmonary tension disorders or the tension cyst phenomenon are well-known conditions. The increase of intrathoracic pressure distends the bronchi, the lung tissue or the pleural space. Its pathogenesis must be a ball-valve mechanism somewhere in the bronchial tree, that admits air but prevents its egress. The aetiology of this condition varies within wide limits, but all the forms have in common dyspnoea, cyanosis and heart failure. The tension changes of the lungs are classified according to their aetiology. We distinguish between developmental cysts and obstructive (valvular) emphysema. Many authors consider that infective cysts form a third group, but as both forms may become infected, we do not consider them as a separate group.

This report is based on the material of the second Surgical Clinic of the University of Budapest and gives an account of our observations in four cases. In the past 12 years 441 intrathoracic operations have been carried out on infants and children. Of these, 45 were for the treatment of pulmonary cystic disease: in six of these emergency thoracotomy had to be carried out. The following reports describe four of these.

Case Reports

Case 1. A 2-month-old baby presented with respiration that had been difficult since birth with cyanosis on crying. At the age of 1 month he was admitted to a hospital where the condition was considered to be pneumothorax. Chest needling and drainage were given without any effect. After four weeks of continuous drainage he was transferred to our department still with dyspnoea and cyanosis. A chest radiograph showed obstructive emphysema of the left lung (Fig. 1). Because the infant's condition was rapidly deteriorating, an emergency thoracotomy was performed. On opening the left chest, a distended and emphysematous ballooned upper lobe was found that occupied the whole of the left chest. This was eviscerated through the thoracotomy

wound. The left lower lobe was atelectatic and could not be re-aerated. As the atelectasis might have been present for weeks, a pneumonectomy was performed. After an uneventful convalescence the infant made a complete recovery. He is now 4 years old.

The pathological report showed obstructive emphysema of the left upper lobe with dilated alveoli and bronchioli.

A radiograph of the thorax four years after the operation revealed that the remaining right lung had herniated over to the left side (Fig. 2).

Case 2. A 14-month-old girl, at 10 months of age, had developed a chronic non-productive cough with dyspnoea. A radiograph revealed increased translucency of the left lung (Fig. 3). After a few days the left lung

FIG. 1.—Case 1. Obstructive emphysema of the left upper lobe.

* A paper read at a meeting of the British Association of Paediatric Surgeons in Sheffield, July 1963.

FIG. 2.—Case 1. Four years later. Right lung has herniated over to the left side.

FIG. 4.—Case 2. The same patient after the left lung became atelectatic.

FIG. 3.—Case 2. Chest radiograph showing increased radiolucency of the left lung.

FIG. 5.—Case 2. Tomogram gave rise to the suspicion of a coin lesion at the bifurcation.

became atelectatic and the mediastinum was displaced to the left (Fig. 4). Bronchoscopy revealed compression of the left main bronchus from the outside, resulting in almost complete obstruction. A tomogram gave rise to the suspicion that a coin lesion might be present at the bifurcation (Fig. 5). A few days later the left lung became emphysematous again. Thoracotomy revealed that at the bifurcation there was an egg-sized thin-walled cyst distended with fluid. This compressed the left main bronchus. The removal of the cyst released the compression of the main bronchus. The post-operative course was uneventful.

The pathological report confirmed a thin-walled bronchial cyst. We saw this girl four years after the operation and she was developing normally.

Case 3. A girl, 10 months of age, experienced respiratory difficulty at 7 months of age. She was admitted to a paediatric department where a chest radiograph showed a homogeneous opacity in the right hemithorax, posteriorly. The chest was aspirated, and yellow fluid was found. In

spite of this, the shadow increased in the next few days and she became febrile (Fig. 6). One night she developed acute dyspnoea with cyanosis, and an emergency thoracotomy had to be performed. When the right pleura was opened it revealed a distended, bluish transparent cyst as large as an infant's head, which was extirpated. Convalescence was uneventful. Three years after the operation a chest radiograph was normal.

The pathological report showed that this was an enterogenous cyst (Fig. 7). There is no mention in the published reports available to me that such enterogenous cysts may cause acute compression of the intrathoracic organs by their sudden increase in size. We believe, however, that our case rightly belongs to this group.

Case 4. A 3-month-old boy became dyspnoeic, and a staphylococcal infection was suspected. A distended cavity in the left upper lobe completely compressed the left lung and partly compressed the right lung (Fig. 8). Needle aspiration proved pressure. This led to a temporary return of the mediastinum to its normal position. In

FIG. 6.—Case 3. Enterogenous cyst in the right hemithorax.

a few hours the tension in the cyst increased again. In spite of the introduction of a drainage tube and continuous aspiration, dyspnoea and cyanosis could not be arrested, and suddenly this became severe. An emergency thoracotomy was performed which allowed the air to escape with a whistling noise from the distended cyst. The left upper lobe containing the cyst was removed; in the lower lobe bronchopneumonic foci could be palpated. On the seventh post-operative day a bilateral bronchopneumonia led to cardiac failure and death. The pathologist reported that the large cyst in the apical segment resulted from a narrowing of the bronchus. This

FIG. 8.—Case 4. Obstructive emphysema in the left upper lobe.

was reduced to a slit and its cartilage was abnormally formed. No epithelium could be seen, only endothelium-like flat cells (Fig. 9). The diagnosis was pneumatocele.

Discussion

A diagnosis of pulmonary tension disorders is possible on the grounds of symptoms and radiological examination. The common feature of these dis-

FIG. 7.—Case 3. Photomicrograph of the enterogenous cyst.

FIG. 9.—Case 4. Photomicrograph of the resected bronchus.

orders is that there is a large cavity in the thorax, containing air or fluid that compresses the surrounding lung tissue and displaces the mediastinum. Bronchial cysts and valvular emphysema are not easy to differentiate from tension pneumothorax. Separation of the latter is important because treatment is different. It is important to differentiate this condition from diaphragmatic hernia (which can be done with a barium meal) because this requires a different operative procedure. In many cases the final diagnosis is only arrived at during thoracotomy. In established cases of tension pneumothorax a pleural puncture is indicated. This is free of danger; but if a puncture is performed in a case of bronchial cyst or of obstructive emphysema, the puncture may cause pneumothorax. Puncture and aspiration drainage are indicated in cases with infection when thoracotomy is contraindicated. This procedure cannot be expected to give complete recovery in the majority of cases. The choice of procedure is thoracotomy and resection. The diagnosis of tension cyst phenomenon does not necessarily mean an urgent operation; in the majority of cases it is possible to choose the optimum time. In children under 6 months we only operate if it is absolutely necessary. After 6 months of age the risk of thoracotomy is reduced, and in cases without severe symptoms we prefer to wait. In cases of tension cysts, if the symptoms are increasing during the observation period or if the cyst grows noticeably, the operation must be carried out quickly, and if it is performed in time it carries almost no risk. Prolonged observation and unsuccessful conservative measures are harmful. The risks are as follows.

- (1) Repeated punctures and prolonged drainage may cause infection of the pleural space and the cyst.
- (2) Inhalation bronchopneumonia may occur.
- (3) The prolonged compression of the surrounding lung may lead to permanent fibrosis.

(4) Cardiovascular disturbances due to the displacement of the heart and the great vessels may lead to cardio-respiratory embarrassment.

Development of the ball-valve mechanism is attributed to extra- and endobronchial factors. In our cases the main cause was a malformation of the bronchial cartilage. This is demonstrated by a photomicrograph of our case (Fig. 10). The rudimentary development of the bronchial cartilage, its insufficient elasticity and its weakness are further

FIG. 10.—Photomicrograph showing the malformation of the bronchial cartilage of the upper bronchus.

factors leading to the ball-valve mechanism. Thoracotomy and resection are tolerated by older infants very well. The infants, who were previously underdeveloped, began to grow rapidly. Their chests developed proportionately, and this happened even with our one patient who had a pneumonectomy. Thus, if there is an indication for the operation of a tension cyst, it must be performed without delay.

Summary

In infants, pulmonary disease due to acute tension disorders may be classified according to aetiology into two main groups: (1) developmental tension cysts; (2) obstructive or ball-valve emphysema. The best treatment for tension cysts is thoracotomy and lung resection. Prolonged observation is dangerous and may lead to serious pulmonary infections, lung fibrosis or sudden death due to suffocation or inhalation of infected material. Four typical examples have been described.

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