of knowing whether it is in primary or secondary apnoea. If in primary apnoea it will recover without intervention, but if analeptic drugs are given it will start gasping more quickly and vigorously, giving rise to the clinical impression that the drug is life-saving. Effective I.P.P.V., though also unnecessary, would not be harmful and would similarly hasten recovery.

If the baby is in secondary apnoea it will die unless given I.P.P.V., and the administration of analeptics will be of no benefit. What evidence there is suggests that lobeline or ethamivan will seriously jeopardize the chances of resuscitation by I.P.P.V. because they induce hypotension at this stage.

We feel strongly that analeptic drugs have no place in obstetric or neonatal departments, as they are of no benefit in primary apnoea and potentially lethal in secondary apnoea. The availability of personnel trained in intubation of small babies is of paramount importance, as recommended by the Central Midwives Board (February 1968). The practice of putting a drop or two of ethamivan on the tongue while waiting for the anaesthetist or to see what happens should be discouraged.

We wish to thank Mrs. S. Moonsaffie for her invaluable technical help and Riker Products for supplies of Vandid (ethamivan) and Numotac (taloximine) and for information on dosage.

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Medical Memoranda

Pulmonary Tuberos Sclerosis Presenting with Pleural Effusion

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Tuberos sclerosis in all its forms has been calculated to occur with an incidence of 1:150,000 in the general population of the United Kingdom and an incidence of 1:300,000 for those forms of the disease with mental defect (Dawson, 1954).

Pulmonary complications are uncommon, only 17 cases having been described. Dawson (1954) reviewed the nine cases then reported in the literature and described four others. Green (1968) described four more. The pulmonary complications in these cases usually presented with dyspnoea, haemoptysis, and spontaneous pneumothorax. The following case is of interest in that, although having typical signs and symptoms and classical radiological appearances, the predominating feature was a large pleural effusion, the nature of which is described.

PATHOLOGY

Tuberos sclerosis usually affects several systems, embracing the brain and eye, skin, viscera, bones, and lungs. These may be briefly summarized as follows: sclerotic areas in the brain consisting of glial fibres with few cell bodies which may calcify and be visible radiologically. The patients usually present with epilepsy or mental defect. The retinal lesions, phakomata, consist either of single white glistening masses protruding into the vitreous or of multiple white or grey flat plaques. Skin changes include subungual fibromata, the adenoma sebaceum of the face, and "peau chagriné". This last is a sheet of thick nodular skin in the lower thoracic or lumbar region. The facial eruption consists of papules extending across the nose on to both cheeks with small superficial adenomata. There may also

be "café au lait" spots. The commonest visceral abnormalities are renal tumours of mixed epithelial, fatty, fibrous, and vascular tissue—occasionally malignant. Tumours are also found in other viscera; rhabdomyomata of the heart, polypi in the duodenum and small intestine, and occasionally tumours in the liver, uterus, and larynx. The limb bones and the skull may show cyst-like spaces and periosteal thickening on radiological examination. The lungs when involved usually show multiple pea-sized cysts, air filled, with brown walls consisting of bundles and tumours of smooth muscle with angiomatous new formations containing excessive iron pigment. A mucus-secreting adenoma has also been described in the lung.

CASE REPORT

A married woman aged 46 was first referred to the outpatient department in 1959, when she complained of shortness of breath and was found to be myxoedematous. She had adenoma sebaceum and had been aware of the facial rash for 10 years. She was noted to be of slightly reduced intelligence. She responded well to thyroxine and remained well until 1966, when she again complained of shortness of breath and fatigue and was found to have a large left pleural effusion. She was admitted for pleural aspiration. Intracerebral calcification and subungual fibromata were noted at this time and laboratory investigations were as follows: haemoglobin 12.7 g./100 ml.; white cell count 8,400/cu.mm. (neutrophils 62%, eosinophils 2%, lymphocytes 34%, monocytes 2%); E.S.R. 24 mm. in one hour; urea and electrolytes normal; cholesterol 418 mg./100 ml.; sputum: no malignant cells. Chest x-ray examination confirmed the presence of a large left pleural effusion with cystic mottling throughout both lungs (Fig. 1). Pleural aspiration: 880 ml. of slightly blood-stained fluid, protein 5.1 g./100 ml., no organisms, culture sterile, no malignant cells, scanty red cells, lymphocytes, and endothelial cells; L.E. cells were not seen and pleural biopsy was normal. After pleural aspiration her breathing returned to normal.

FIG. 1.—Left pleural effusion with cystic mottling in lung fields.

FIG. 2.—Left hydropneumothorax.

FIG. 3.—Enlarged view of collapsed lung showing a group of cysts.

She remained well for 18 months, but was readmitted in December 1968 with a reaccumulation of the pleural effusion. On this admission the facial adenomata, subungual fibromata, and cerebral calcification were as before; she was also found to have retinal phakomata and for the first time gave a history of petit-mal epilepsy and cough productive of purulent sputum. Laboratory investigations resulted as follows: haemoglobin 16.8 g./100 ml.; E.S.R. 3 mm. in one hour; white cell count 11,800/cu.mm. (neutrophils 82%, eosinophils 1%, lymphocytes 11%, monocytes 6%); urea 47 mg./100 ml., electrolytes normal; cholesterol 222 mg./100 ml.; L.E. cells were not found and latex test was negative. Chest x-ray examination again showed a left pleural effusion with cystic mottling throughout the remainder of the lung fields. Aspiration was repeated and 600 ml. of pink creamy fluid was withdrawn. This contained 4 g. of protein per 100 ml. The differential cell count showed neutrophils 1%, lymphocytes 74%, eosinophils 24%. There were no organisms and none were cultured. There were no malignant cells.

After aspiration the chest x-ray picture showed fully expanded lungs, a small residual effusion, and the small cysts already mentioned. Ten days later physical signs indicated that the effusion was increasing in size, though she had no new symptoms. Repeat x-ray films showed a left hydropneumothorax (Figs. 2 and 3). This re-expanded satisfactorily without aspiration and at the time of writing the patient was free of respiratory symptoms and had been discharged from hospital.

COMMENT

In the 17 cases of pulmonary tuberous sclerosis recorded the major features have been chronic progressive dyspnoea, re-

current pneumothorax, and haemoptysis occurring predominantly in an adult woman. These differ from the cutaneous and cerebral features, which are usually established by the second decade. Other pulmonary lesions have been one case of solitary adenoma, one case of lobar consolidation, and three cases where typical x-ray appearances—that is, cystic lung fields—were found without respiratory signs or symptoms. In only two of the 17 descriptions is there mention of pleural effusion—one in association with a pneumothorax and the other an almost terminal event. The above case is typical of those previously described by Dawson and Green—that is, progressive dyspnoea, spontaneous pneumothorax, and cystic lungs occurring in an adult woman with minor mental impairment and late onset of pulmonary infection. It would, however, appear to be unique in having a large recurring pleural effusion.

I should like to thank Dr. G. P. Baker for permission to report this case.

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