causes of that condition. Zimmerman, however, has presented one that has the merit of locating the cause, where possible, to the three major divisions of the nerve supply to the bowel. Thus he divides the cases of spastic ileus into those due to (1) stimuli acting on the bowel wall directly, (2) stimuli reaching the bowel from distant lesions, and (3) stimuli reaching the bowel from the central nervous system. To these we would add a further group—(4) unknown. Of these four groups of cases, the second and fourth have a direct bearing on the cases we are reviewing.

The distant lesions, presumably acting through the extrinsic nerves, that may cause spastic ileus include lesions involving the coeliac plexus. Klett's (1923) case (quoted by Zimmerman, 1930) was due to carcinoma of the pancreas with retroperitoneal extension. Other reported cases have included acute pancreatitis and peptic ulcer penetrating posteriorly. In the 159 cases of spastic ileus collected by Zimmerman lesions involving the coeliac plexus were responsible for four. These lesions are very similar to those reported by Ogilvie, Dunlop, and Handley, in which malignant infiltration of the coeliac plexus was found. If it is accepted that such lesions can cause spastic ileus and that spastic ileus can produce the full clinical picture of mechanical ileus, then we feel that there are good grounds for believing that the cases reported by Ogilvie and others represent cases of spastic ileus.

Several cases of spastic ileus have been described in which no cause could be found. Zimmerman collected 39 cases. Aird (1949) also refers to this group. At operation an inexplicably dilated bowel is found, the spasm perhaps having been relieved by the anaesthesia. It is an obstruction without a cause. Our three cases, we believe, fit into this group, for they all presented the clinical picture of a mechanical ileus, whilst at operation there was distended bowel with no evidence of obstruction.

### Summary

Evidence is offered suggesting that the cases of false colonic obstruction described recently in the literature are merely cases of spastic ileus.

Three cases of idiopathic spastic ileus producing a picture indistinguishable from mechanical obstruction are described.

We would like to express our thanks to Mr. Brandon for his helpful advice in treating these cases and in the writing of this paper.

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Mr. C. Price Thomas, thoracic surgeon to the Brompton and Westminster Hospitals, and Dr. Robert Machray, his anaesthetist, are visiting Portugal under the auspices of the British Council, and two Portuguese nurses have returned after studying the care of Mr. Price Thomas's patients at the Brompton Hospital. The nurses will supervise the nursing of his patients at the Instituto de Oncologia, in Lisbon, where Mr. Price Thomas is demonstrating operations with Dr. Machray as anaesthetist. Mr. Price Thomas is lecturing on the surgical treatment of pulmonary tuberculosis and of carcinoma, and Dr. Machray on recent advances in anaesthesia.

# KARTAGENER'S SYNDROME

BY

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The association of complete transposition of the viscera with bronchiectasis was first noted by Siewert (1904) in the case of a 20-year-old male patient who had had a productive cough from the age of 1 month and at the age of 20 was found to have signs of cavitation at the left apex. Apart from brief case reports by Oeri (1909) and Guenther (1923), there is no further mention of the condition in the literature till Kartagener's (1933a) report of four cases, followed later by a further seven cases (Kartagener, 1933b; Kartagener and Horlacher, 1935). About the same time cases were also reported by Nüssel and Helbach (1934), Behrmann (1935), and Kautzky (1936).

Adams and Churchill (1937), surveying 23 cases of situs inversus totalis admitted to the Massachusetts General Hospital from 1886 to 1937, found that five of these had bronchiectasis. All five also had disease of the upper respiratory air passages. The incidence of bronchiectasis in this series of 23 was thus 21.7%, the incidence in all cases (232,113) admitted to the hospital during the same period being 0.306%. The most extensive survey is that of Olsen (1943), who examined the records of 85 cases of dextrocardia admitted to the Mayo Clinic during a period of 27 years and found evidence of bronchiectasis in 14 (16.5%). Ten of these had upper respiratory infection.

In some of the later cases, notably that of Richards (1944), mention is made of congenital absence of the frontal sinus, usually associated with ethmoid and maxillary infection. The case here presented showed congenital abnormality of the paranasal sinuses to an extent which has not hitherto been recorded.

### Case Report

A man aged 44 was referred to the medical out-patient department on October 8, 1947, with a history of productive cough of many years' duration. He was an only child, full-term delivery, without history of cyanosis at birth. His father died about 40 years ago, cause unknown, his mother is alive at 65 and is normal. Several cousins have been traced and are also normal.

The patient gave a history of measles at 3, whooping-cough at 5, and several attacks of pneumonia of increasing severity at the ages of 4, 10, and 16. The left side was always most involved in these attacks. For 20 years he had complained of pain in the left upper chest aggravated by coughing, deep breathing, or turning in bed. In 1927 he had five months in a sanatorium after what appears to have been an attack of left-sided pleurisy. Repeated sputum examination was negative for the tubercle bacillus. No mention was made of transposition on this occasion. His general condition deteriorated after discharge and the amount of sputum increased, with occasional slight haemoptysis. Up to twelve months ago he had suffered from severe frontal headache, which had now ceased.

Examination showed him to be a man of small stature (5 ft. 4 in. = 1.62 m.) and sallow complexion, weight 7 st. 13 lb. (50.35 kg.). He was right-handed and showed moderate finger-clubbing. The right testicle hung lower than the left. Situs inversus totalis was demonstrated by finding heart sounds and cardiac dullness to the right of the sternum, palpable descending colon in the right iliac fossa, and liver dullness on the left side. Confirmation was obtained by barium meal and E.C.G. examination.

Plain radiographs of the chest revealed a condition suggestive of multiple cysts in the left upper lobe. A bronchogram showed that the lungs were transposed, that the cysts seen in

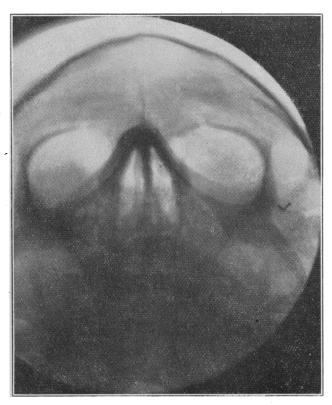
plain film were in communication with the bronchial tree, and that bronchiectasis was also present in the right lower lobe and lingula. Bronchoscopy showed much pus coming from the left upper bronchus. The left middle lobe bronchus looked inflamed and some pus was seen here also. The right base was dry and no signs of inflammation were observed in this area.

Sputum examination revealed a mixture of staphylococci, Friedländer's bacillus, pneumococci, and streptococci. No tubercle bacilli were found by direct or concentration methods.

In view of the association of upper respiratory infection with recorded cases of Kartagener's syndrome and the fact that the patient had been somewhat deaf all his life, particularly in the right ear, expert opinion was obtained and routine radiographs were done.

Mr. Banham reported: "Long history of deafness. No past history of otorrhoea; no nasal symptoms except nasal catarrh. Hearing: right, whisper at 2-3 ft.; left, whisper at 10-12 ft.; right, air conduction better than bone conduction with loss of bone conduction hearing = nerve deafness; left, bone conduction better than air conduction and Weber to the left = middle-ear deafness. Both tympanic membranes intact, but posterior quadrants hang in folds and balloon on inflation of Eustachian tubes. Radiographs show very small frontal sinuses and poorly developed ethmoid labyrinths. Both antra very small and poorly developed. Sphenoidal sinuses absent. No air cells in either mastoid. Nose: no evidence of sinus infection."

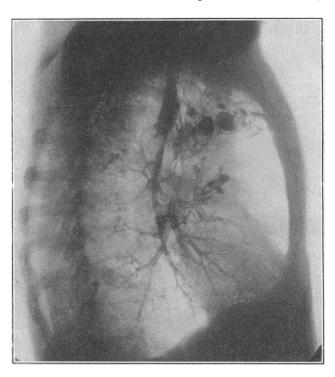
During a period of three weeks his sputum diminished in amount from 7 oz. to 2 oz. (200-56 ml.) daily as a result of postural drainage and the administration of 35 g. of sulphadiazine. His vital capacity at the end of this period was 2 litres. He was seen by three thoracic surgeons, all of whom advised against lobectomy in view of the extensive bronchiectasis and associated bronchitis.



Psychologically he was depressed and introspective. He refused to return to work and blamed the thoracic surgeons for their unenterprising attitude to his case.

### Comment

The ballooning of the ear drums in this case was assumed to be a compensatory phenomenon related to the absence of mastoid air cells. It was thought that the extreme lack of development of the sinuses had protected them from infection, the only formed cavities being in fact those of the antra. The bronchiectasis and cystic condition of the lungs are believed to be similar in aetiology and to have followed direct on his attack of pneumonia in infancy,



there being no history of cough in the first three years of life. The high incidence of bronchiectasis, however, in cases of transposition raises the possibility of congenital abnormality of the walls of the bronchioles as a factor in these and possibly some other cases. The important factors in the aetiology of bronchiectasis have in the past been considered to be upper respiratory infection and bronchial obstruction. Only about 80% of cases of bronchiectasis show evidence of upper respiratory infection, however, and in some there is no history of bronchial obstruction. It is noteworthy that four of Olsen's cases of dextrocardia had bronchiectasis without sinus infection, and that in four others sinus infection was present without bronchiectasis. Similarly, two of the cases described by Adams and Churchill had had cough from birth with no factors in the history to suggest bronchial obstruction, though all at the time of examination had upper respiratory infection.

The possibility which is suggested by cases such as this is that in cases of bronchiectasis with no history of upper respiratory infection, and in others with no history of bronchial obstruction, congenital defects in the wall of the bronchioles should be regarded as an aetiological factor.

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