inversus totalis was confirmed by barium meal and electrocardiographic examination. Sputum examination revealed a mixture of *Micrococcus catarrhalis* and streptococci. A blood count was normal, and the blood Wassermann reaction and Kahn test were negative. A plain x-ray film of the chest was suggestive of bilateral bronchiectasis, and the bronchogram was interpreted as follows: "The stem of the posterior bronchus of the right upper lobe shows dilatation, but the rest of the division is normal. The right lower lobe is grossly contracted and lingula is bronchiectatic. The inferior division shows cylindrical bronchiectasis, but the superior division is filled only in the proximal part. The left upper lobe is normal, but the left middle and lower lobes show a mild degree of cylindrical bronchiectasis."

The patient was seen by Mr. H. J. M. Stratton, who reports: "Nasal mucosa is pale and watery, with polypoidal fringes on both middle turbinates consistent with vasomotor rhinitis. Pharynx is healthy except for post-nasal thick mucus. X-ray film of sinuses shows infection of antra and absence of frontal and sphenoidal sinuses and ethmoids. X-ray film of mastoids shows no development of mastoid air cells."

Discussion

Small frontal sinuses, or their entire absence, have long been regarded as a part of Kartagener's triad, but emphasis has recently been laid on other associated congenital abnormalities of the nasal sinuses. These features are well illustrated by my two cases. The first case shows the triad, whilst the second shows congenital abnormality of the paranasal sinuses to an extent which has been seen in only one previous case (Andrews, 1949). That case was later reviewed by Banham (1950), who points out that, as at birth the maxillary and mastoid antra are well-formed cavities and the frontal and sphenoidal sinuses and ethmoid air cells are underdeveloped, it would be interesting to know if this lack of development of the upper respiratory tract was always present in these cases, especially as previous writers have merely recorded absent or small frontal sinuses.

Coincidental occurrence of bronchiectasis, situs inversus, and congenital anomalies of the nasal sinuses is regarded as evidence of maldevelopment. In discussing the incidence of situs inversus totalis Adams and Churchill (1937) have shown that two types of people are affected by this maldevelopment—normal persons known as mutants, and those in whom the abnormality is associated with other congenital defects, known as monsters. They therefore classify sufferers from the syndrome as minor monstrosities. Both the above patients, besides showing the triad, were of small stature, with short limbs and small hands and feet, and were also of low mental grade, and I think can rightly be called minor monsters.

Another interesting feature of the syndrome lies in the possibility that a congenital defect of the bronchial wall is a primary cause of bronchiectasis. Infection of the upper respiratory tract and bronchial obstruction are usually regarded as important factors in the causation of bronchiectasis, but not all cases of bronchiectasis show evidence of both or even one of them. Moreover, Olsen (1943) has reported four cases of dextrocardia with bronchiectasis without sinus infection, and the very existence of this syndrome supports the theory that the weakness in the walls of the bronchioles is the primary factor in affecting their dilatation, perhaps aided by chronic infection. Farber (1943), in discussing the aetiology of cystic disease of the pancreas, postulated that there may be a constitutional defect of secretion in various glands throughout the body, and it is therefore suggested by Cohen *et al.* (1950) that bronchiectasis may be caused by inspissated tracheo-bronchial secretion, producing atelectasis and infection. Both my patients had chronic sinus infection, which must therefore be regarded as an important factor in the aetiology of bronchiectasis; but in cases without a history of upper

respiratory infection or bronchial obstruction congenital weakness in the walls of the bronchioles must play a leading part in their dilatation.

Summary

Two cases of Kartagener's syndrome in members of one family have been observed for the first time in this country. They are here reported, and comments are offered on the extent of maldevelopment and on the aetiology of bronchiectasis.

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Medical Memoranda

Intolerance to Hexamethonium Bromide

Recent studies of the action of the hexamethonium and pentamethonium group of drugs on the gastro-intestinal tract and blood pressure will probably lead to their widespread use. It is therefore important that any idiosyncrasy or untoward action should be known, and with this object in view the following case is recorded.

CASE REPORT

A box-spring maker aged 53 had suffered from a duodenal ulcer for 25 years, otherwise his health had been good except for an attack of Bright's disease at the age of 7.

On January 28, 1951, he paid a routine visit to his doctor, who prescribed hexamethonium bromide for the relief of his ulcer pain, there being no history suggestive of pyloric stenosis. On the first day of treatment he took 0.25 g. before breakfast and lunch, and the following day increased each dose to 0.5 g., a total dose of 1.5 g. of the drug being given over two days. That afternoon, while watching a football match, he felt faint, vomited, and collapsed. After two to three minutes he recovered consciousness and felt very weak, but managed to walk home. He went to bed after further vomiting and slept. The next morning he felt better, but on arriving at work he again felt faint and collapsed: he lay down for a while and was finally taken home and put to bed. He continued to vomit throughout the day, and towards evening developed severe cramp-like pain in the calf muscles, which he described as "causing his legs to draw up." His doctor was called and he was admitted to hospital the following day.

On admission he was conscious and able to give a history, very lethargic and pale, but not cyanosed. His temperature was 98° F. (36.7° C.), pulse 94 and regular, blood pressure 65/49 (lying). It was impossible to take the blood pressure sitting up, as that position caused the patient to faint. No other abnormalities were found in the cardiovascular, respiratory, or alimentary system. Both pupils were dilated and did not react to light or accommodation; the central nervous system was otherwise normal.

It was considered that his condition was due to hexamethonium bromide. He was given 5 min. (0.3 ml.) of

1/1,000 adrenaline hydrochloride by intravenous injection and his blood pressure immediately rose to 125/65. However, within an hour it had dropped to 75/40; he was given an intramuscular injection of ephedrine hydrochloride, $\frac{1}{4}$ gr. (50 mg.), and it rose to 105/60. During the next day he was maintained on ephedrine hydrochloride 1 gr. (65 mg.) by mouth, and his condition was satisfactory, his blood pressure remaining about 115/75, although on sitting up it dropped to 70/60. From an absence of urine on the previous day his output rose to 66 oz. (1,870 ml.). From then on he continued to improve, although his pupils remained fixed and dilated for a further four days, and he remained on ephedrine by mouth for another week.

During his stay in hospital, routine investigation showed that his urine contained 6 parts per 1,000 of albumin but no casts or red cells. His blood urea was 142 mg. per 100 ml. and his blood bromide 30 mg. per 100 ml. It was thought at first that these changes might have been due to continued low blood pressure, but three weeks later his urine still contained albumin, his blood urea was 87 mg. per 100 ml., and a urea-clearance test showed a standard clearance of only 15% of normal. It is therefore more likely that his kidneys had already been damaged by his attack of Bright's disease. An electrocardiogram was normal. A barium meal revealed a considerable degree of pyloric stenosis, which was secondary to chronic duodenal ulceration as it did not show the characteristic picture produced by this drug.

COMMENT

It seems probable that this case represents true intolerance to hexamethonium bromide. A comparatively small oral dose produced a very marked effect which was prolonged over several days.

It is possible that the pre-existing renal damage might have interfered with the elimination of the drug, though without exact knowledge of its fate in the human body this would merely be a guess. Furthermore, it would seem unlikely, as hexamethonium bromide has been used in other cases with just as severe interference with renal function without producing such an alarming result.

This case does, however, suggest that small doses, probably no more than 120 mg. by mouth, should be used when beginning treatment with hexamethonium bromide, that the patient should be within easy reach of a bed, and that a careful watch should be kept on his blood pressure.

Our thanks are due to Dr. A. H. Douthwaite for his permission to publish this case and for his help and encouragement.

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Phaeochromocytoma Presenting as a Cardiac and Abdominal Catastrophe

In view of a report of a phaeochromocytoma presenting as an abdominal emergency (Gilliland and Daniel, 1951), it is of interest to record this very similar case.

CASE REPORT

A housewife aged 47 was admitted to hospital as a case of coronary thrombosis. She had a 12-hours history of sudden severe substernal and epigastric pain lasting half an hour, nausea, vomiting, sweating, and collapse. The attack had begun at 11 p.m. while strolling by the seashore. A local doctor recorded a blood pressure of 220/140 and a pulse of 160, gave her morphine, and asked for her to be admitted. The patient gave a past history of diabetes mel-

litus for over three years, stabilized at a Birmingham hospital at 20 units P.Z.I. daily, urine tests being done twice a year. She had had her insulin the day of the attack, but not on the morning of her admission. She had also had a similar but less severe attack of pain of a few minutes' duration three months previously.

On admission she was in an obviously shocked condition; pale and grey about the lips, cold, nauseated, and sweating profusely. Her temperature was 101.8° F. (38.8° C.), pulse 160, respirations 26, and blood pressure 60/?. The heart sounds were distant, with a soft apical systolic murmur. There was no abdominal tenderness or palpable tumour. The lungs and nervous system appeared normal. The urine was brick-red to Benedict's test and showed quantities of acetone, 1+ of albumin, and 8 g. of chloride per litre. Her haemoglobin was 113% and W.B.C. 12,500. She was put on anticoagulants and soluble insulin twice a day, with glucose feeds.

By the second day her blood pressure had risen to 140/80. She complained of encircling abdominal pains and appeared as shocked as on admission. Her abdomen was soft but tender in the left hypochondrium. The morning specimen of urine showed green to Benedict's test and no acetone.

On the third day she was found to have generalized extreme abdominal tenderness, some distension with gas, absence of bowel sounds, and a mobile mass the size of an orange to the left of the umbilicus. She said she had passed only a little flatus and no faeces since admission. A loud apical systolic and softer diastolic murmur had developed. Electrocardiograms showed no definite evidence of a coronary thrombosis but diffuse myocardial damage. X-ray films of the chest and abdomen were negative. Her haemoglobin was now 88% and W.B.C. 6,500. Her blood pressure was still 140/80, her pulse settled to about 120, and she remained slightly febrile.

A surgical opinion was obtained, but in view of her critical state and the likelihood of a coronary thrombosis no interference was recommended.

For the next ten days her general condition remained practically unchanged; the abdominal pain passed off but the distension remained; occasionally bowel sounds were audible and a little flatus was passed. Enemata and flatus tubes had little effect. The mass moved to the right of the umbilicus and became more palpable. The diastolic murmur vanished and the character of the systolic murmur changed. Meanwhile difficulty was experienced in stabilizing both her diabetes and her blood prothrombin time. She could take only small quantities of fluids by mouth, even through Ryle's tube, without vomiting, and finally an intravenous drip was set up.

Her blood pressure was 125/80 on the ninth day, rose to 200/110 on the tenth day, and fell to 180/80 by the twelfth day. She sweated a great deal during this phase. On the twelfth day she passed three motions and the distension disappeared, but next day she complained of severe pain in the left chest, had an epistaxis, became dyspnoeic, and died after a short period of Cheyne-Stokes respiration.

Pathology.—At necropsy (Dr. J. A. V. Morgan) all organs were found to be normal except the lungs, which were congested and oedematous, and the left suprarenal, which "was replaced by a soft encapsulated haemorrhagic tumour 8.5 cm. in diameter with a cystic centre. There had been a recent haemorrhage into the capsule of the tumour and the compressed tissues surrounding it. The left kidney was compressed and grossly distorted by the growth." Histology showed mainly necrotic matter with areas of cells compatible with those of a phaeochromocytoma.

COMMENT

As with the case of Gilliland and Daniel, there was much conjecture over the diagnosis of this case. The substernal pain and fall in blood pressure made a coronary thrombosis