Medical Memoranda

Addison's Disease in Identical Twins

Brit. med. J., 1968, **1**, 559

Addison's disease in identical twins has been reported only once in the literature (Smith *et al.*, 1963). This paper presents another example.

CASE 1

A 46-year-old man was admitted to hospital in October 1967 with a six-week history of early morning vomiting, loss of 10 lb. (4.5 kg.) in weight, lassitude, and "attacks of weakness."

On examination he was seen to be thin, with slight pigmentation of the skin and lips, but not of the buccal mucosa. Blood pressure was 85/60 mm. Hg and the heart sounds were faint.

Investigations.—Urine normal on routine analysis; E.S.R. (Westergren) 22 mm. in first hour; Hb, W.B.C., M.C.H.C., and film all normal; serum sodium and chloride levels repeatedly below normal—for example, 126 and 88 mEq/l. respectively; serum potassium, urea, calcium, phosphate, cholesterol, and liver function tests all normal; random blood sugar 55 and 71 mg./100 ml.; *x*-ray examination of chest showed a narrow heart shadow; plain *x*-ray film of abdomen was negative; barium-meal examination normal; E.C.G. low voltage; 24-hour urinary 17-ketosteroids 6.6 mg./day and 17-hydroxycorticosteroids 6.8 mg./day in a normal urinary output; these levels are low and showed no response to standard intravenous A.C.T.H. infusion test; plasma cortisol levels (11.5 μg./100 ml. at 9 p.m. and 13.5 μg./100 ml. at 9 a.m.) showed no diurnal rhythm, and no response to Synacthen (10 μg./100 ml. was the post-Synacthen level).

The patient made a rapid clinical recovery on treatment with cortisone acetate 12.5 mg. twice daily and fludrocortisone 0.1 mg. twice daily. At follow-up four weeks after initiation of therapy he was asymptomatic, with a blood pressure of 115/80 mm. Hg, and had gained weight. Detailed questioning revealed that his identical twin brother had died very suddenly in 1955 in the same hospital. This twin brother is the subject of the following case report.

CASE 2

In January 1955 a 33-year-old man was admitted to hospital as an emergency case, having suddenly collapsed at home. For some months he had complained of lethargy.

On examination he was conscious, with generalized muscle weakness but no neurological deficit; blood pressure and pulse were unrecordable; he failed to respond to resuscitative measures and died soon after admission.

Positive findings at necropsy were faint pigmentation of the skin, a very small heart (170 g.), and macroscopic absence of adrenal tissue. However, microscopical examination of suprarenal fat tissue showed a gross degree of atrophy of the suprarenal cortex on both sides. Death was thought to be due to primary suprarenal cortical atrophy.

COMMENT

Adrenal hypofunction was proved in both cases—in Case 1 biochemically, in Case 2 histologically. In neither case was there clinical or laboratory evidence of secondary adrenal failure, for example, due to tuberculosis, candidiasis, histoplasmosis, or a reticulosis. It seems reasonable to accept both as proved cases of primary Addison's disease.

The patients were always accepted by their relatives and family doctors as identical twins; chromosome studies are impossible because one twin is dead.

The familial occurrence of Addison's disease was noted by Wilkins (1965), who recorded 41 cases in 15 families, and confirmed preponderance in males. There is evidence of a hereditary basis in Addison's disease associated with polyglandular disease (cf. Mershon and Dietrich, 1966). In children Addison's disease of the familial type is often associated with hypoparathyroidism, with or without candidiasis. In 71 cases of Addison's disease Blizzard and Kyle (1963) found 36 with antibodies to adrenal tissue.

All these reports suggest a genetic basis in some forms of Addison's disease, but more examples are necessary.

I am grateful to Dr. S. G. McAlpine and Mrs. S. Carson for assistance with these case reports, and would like to thank the staff at the Steroid Laboratory, Glasgow Royal Infirmary, for performing the adrenal function tests.

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REFERENCES

Blizzard, R. M., and Kyle, M. (1963). *J. clin. Invest.*, **42**, 1653.
Mershon, J. C., and Dietrich, J. G. (1966). *Ann. intern. Med.*, **65**, 252.
Smith, M. E., Gough, J., and Galpin, O. P. (1963). *Brit. med. J.*, **2**, 1316.
Wilkins, L. (1965) *Diagnosis and Treatment of Endocrine Disorders in Childhood and Adolescence*, 3rd ed., p. 373. Springfield, Illinois.

Air Embolism and Babinski Reflex

Brit. med. J., 1968, **1**, 559–560

This paper describes a case of air embolism which was treated in a hyperbaric recompression chamber. Alterations of a well-defined neurological sign (Babinski reflex) could be related to changes in the relative volume and/or pressure of the bubble causing the sign.

There are two major conditions under which free inert gas may appear in body tissues. (1) Decompression sickness—too rapid decompression in divers and flyers may set free inert gas which was earlier dissolved in the tissues, including blood. (2) Traumatic air embolism, occurring, for instance, during lung rupture, open heart surgery, or transfusion accidents.

The *absolute* amounts of free gas necessary to cause different kinds of symptoms in decompression sickness and air embolism are at present unknown. Even when known volumes of air are injected into the circulation for experimental purposes the results may differ greatly, depending on where the gas happens to lodge. However, once signs and symptoms are produced the *relative* changes in pressure and volume of the free gas with which the signs and symptoms vary become accessible for study. The gas volume may be influenced to a known extent by changing the atmospheric pressure in a recompression chamber. Though observations in human subjects are more accurate than in animals, the risks involved seriously limit experimentation. Most signs and symptoms are gradual in onset and disappearance—for example, unconsciousness, paralysis, sensory disturbances, bends. Subjective factors in both the patient and the examiner can further bias the accuracy of the