Correspondence

Newly diagnosed cystic fibrosis in middle and later life

SIR,—In their study Drs B Hunt and DM Geddes (Thorax January 1985;40:23-6) make the diagnosis of cystic fibrosis (CF) on dubious grounds. CF is a generalised disease of exocrine glands. The diagnosis requires demonstration of exocrine gland abnormalities uniquely characteristic of the disease.

The sweat of children with CF is uniquely characteristic. A sweat Na or Cl concentration of greater than 60 mEq/l is diagnostic if there is also evidence of pulmonary disease or pancreatic disease or a family history of CF. Other causes of a positive result in the sweat test are usually easy to rule out.

In adults the sweat test is less helpful. Na or Cl concentrations above 60 mEq/l are common in normal people. A concentration below 50 mEq/l does, however, rule out CF. Pulmonary changes in CF are primarily the result of chronic infection and are not, by themselves, diagnostic.

Drs Hunt and Geddes put faith in the diagnostic significance of suppression of sweat Na concentration by fludrocortisone, as described by Hodson et al. This paper presents a graph where two CF patients have no drop in sweat Na concentration after the steroid, while two control subjects show a marked suppression. The data on all patients, unfortunately, are much less impressive. The average suppression in controls is 26.3%, while that in CF patients is 8.7%. The average Na concentration, before steroid, in the controls was 43.2 mEq/l and that in the CF patients 103.0 mEq/l. The authors did not calculate the average drop in Na concentration. This is 11.4 mEq/l for the controls and 9.0 mEq/l for the CF patients. The difference between these numbers is not significant and is not of diagnostic importance.

The diagnosis of CF in an adult can be established. All exocrine glands are involved. Almost every CF male has total obstruction of the vas deferens. Eighty five per cent to 90% of all patients have virtually total obstruction of the pancreatic ducts. With a positive sweat test and pulmonary disease, the demonstration of either azoospermia or absent pancreatic enzymes confirms the diagnosis. Even the difficult case of a woman without malabsorption may be diagnosed. When the CF pancreas is only partially obstructed the pancreatic juice is still abnormal. The output of enzymes may be normal, but the outputs of water and bicarbonate are markedly, and diagnostically, reduced. A duodenal drainage can establish the diagnosis.

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Serum concentrations of vitamin D metabolites in untreated tuberculosis

SIR,—We have read with interest the study carried out by Dr PD O Davies and colleagues (March 1985;40:187-90) on Indian patients with untreated tuberculosis, abnormal calcium metabolism and low serum 25-hydroxycalciferol (25-OH Vit D) levels. These authors also underline the risk of iatrogenic vitamin D deficiency and osteomalacia occurring particularly in this population treated with antituberculous drugs. They therefore proposed prevention by supplementation with 1,25-dihydroxy calciferol (1,25-(OH)2 vit D).

Nevertheless, high serum calcium concentrations have been found in 25% or more of patients with pulmonary tuberculosis where it is associated with vitamin D supplementation. A study on an anephric patient reports hypercalcaemia during clinically active tuberculosis; it was associated with an increase in serum 1,25 (OH)2 vit D levels, which suggested an extrarenal 1α-hydroxylation of 25-(OH) vit D. We have recently been able to demonstrate in a 26 year old Haitian woman with active pulmonary tuberculosis and hypercalcaemia that this conversion could actually occur in vitro in alveolar macrophages recovered from bronchoalveolar lavage fluid. Our method was similar to the one described by Adams et al in patients with hypercalcaemia during sarcoidosis.
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Thorax 1985 40: 639
doi: 10.1136/thx.40.8.639

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