Prolonged Survival in an Adult with Cystic Fibrosis*

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Cystic fibrosis in patients over 40 is rare. We report a 52-year-old woman in whom cystic fibrosis was confirmed by sweat analysis. This patient represents the oldest cystic fibrosis patient (with confirmatory sweat chlorides) ever described. We conclude that any patient with the appropriate clinical presentation, regardless of age, should be investigated for cystic fibrosis.

An increasing number of children with cystic fibrosis are living into adolescence and adulthood.1-3 Patients with mild symptoms of cystic fibrosis may not be diagnosed until adolescence or even later in life.4 The diagnosis of cystic fibrosis should be considered in any adult patient with the appropriate clinical presentation, regardless of age, as shown by the following report.

CASE REPORT

A 51-year-old white woman was referred for evaluation of bronchiectasis. Her first significant illness was pneumonia at age eight from which she recovered promptly. Growth and development were normal during adolescence although she was told, when a teenager, that her chest roentgenogram was abnormal but "not TB." Her first chronic symptom was a persistent cough with mucoid sputum production which developed in her late teens. She had five successful uncomplicated pregnancies while in her 20's. None of her children has had chronic pulmonary or gastrointestinal disease. Recurrent bronchitis and pneumonia, complicated by pneumothorax on three occasions, have led to more than 20 hospitalizations over the past 25 years. Bronchography confirmed the presence of bronchiectasis in her late 20's. Dyspnea has limited her activities as a housewife for the past 15 years and she has required supplemental oxygen during exertion for the past year. She reports three to four, soft greasy stools a day.

Physical examination revealed a dyspneic woman with a respiratory rate of 36. Her oral temperature was 37.7° C; weight, 56.7 kg; and height, 163 cm. Coarse inspiratory and expiratory rhonchi were heard throughout the chest. Marked clubbing was present. Complete blood count, urinalysis, chemistry profile including liver enzymes, and electrocardiogram were normal. The chest roentgenogram revealed extensive bronchiectasis with scarring and retraction (Fig 1). Sputum culture grew mucoid Pseudomonas aeruginosa. Her FEV1 was 0.7 liter and FVC was 1.3 liters. Arterial blood gas levels while breathing room air were PaO2 42 and pH 7.44. Positive sweat chlorides were confirmed at a cystic fibrosis center (Dr. W. J. Warwick, University of Minnesota). Four sweat tests were done with pilocarpine iontophoresis (Gibson-Cooke technique) and were positive: 96, 101, 101 and 102 millimoles of chloride per liter of sweat with adequate quantities of sweat on all four tests. Fecal fat analysis after a minimum intake of 100 gm of fat per day averaged 120 gm/24 hours and serum carotene was undetectable. After treatment with parenteral antibiotics, vigorous pulmonary toilet and pancreatic enzyme supplementation, she was symptomatically improved.

DISCUSSION

The diagnosis of cystic fibrosis is firmly established in our patient. She fulfills the diagnostic criteria with elevated sweat chlorides, chronic pulmonary disease and clinical exocrine pancreatic insufficiency.5 Pansinusitis and the presence of the mucoid variant of Pseudomonas

Figure 1. Chest roentgenogram demonstrates scarring and retraction with bulla formation predominantly in the upper lung fields.

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CHEST, 77: 2, FEBRUARY, 1980
Pentazocine in her sputum complete the clinical picture.

An increasing proportion of patients with cystic fibrosis are over 20 years of age and the number of adults with cystic fibrosis is expected to increase in the future. This increased prevalence in adults appears to represent prolonged survival from childhood rather than a delay in manifestations of the disease until later in life. Few adults with cystic fibrosis have been living beyond 40 years of age. A previously reported Australian patient with fibrocytic disease of the pancreas has survived into his 80's. The patient, however, had normal sweat chlorides when studied further. This patient, the oldest documented case of cystic fibrosis ever reported, is presented to demonstrate that in the appropriate clinical setting, the clinician should suspect the diagnosis of cystic fibrosis regardless of the age of the patient.

ACKNOWLEDGMENTS: The authors wish to thank Harley Racer, M.D. for referring his patient, Warren J. Warwick, M.D. for the sweat chloride analysis and E. C. Rosenow, III, M.D. for review of the manuscript.

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Pentazocine Abuse
Report of a Case with Pulmonary Arterial Cellulose Granulomas and Pulmonary Hypertension

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Chest, 77: 2, February, 1980

We report a patient who developed pulmonary hypertension following repeated intravenous injection of dissolved pentazocine tablets. Through analysis of lung biopsy material, this was shown to be due to embolization of the cellulose filler in the tablet and the tissue reaction it produced. Administration of prednisone appeared to improve the patient's clinical state.

Pentazocine, a potent and allegedly nonaddicting analgesic was introduced in 1966. Its nonaddicting properties were soon brought into dispute with reports of abuse of oral and parenteral forms revealing definite physical dependency. Subsequent reports of abuse have demonstrated fetal addiction and the preference of addicts to pentazocine over more difficult to obtain drugs. In our experience with patients on long-term hemodialysis, we have seen several cases of addiction where patients have procured and administered parenteral pentazocine to themselves. Here we report a patient with diffuse pulmonary vascular occlusion and pulmonary hypertension secondary to the injection of crushed pentazocine tablets.

Case Report

The patient was a 25-year-old man on hemodialysis since September, 1973, as treatment for chronic renal failure secondary to congenital bladder neck obstruction. In September, 1978, the patient was placed on 15 mg of pentazocine intravenously up to three times weekly for chronic headaches following dialysis. His pain was severe, radiating up the back of his neck over the top of his head to the supraorbital area. This was associated with nausea and vomiting often lasting days. Following an unsuccessful search for organic etiologies, the patient was placed on oral pentazocine (50 mg) to be taken as needed. He noticed a need to take his medication at progressively shorter intervals to maintain relief until he was obtaining no relief from 400 mg daily. The patient had seen the use of parenteral pentazocine at the dialysis unit, and in October, 1978, he crushed and dissolved a 50 mg tablet in 3 ml of water and injected it intravenously. He noticed prompt relief and an increased sensation of euphoria. His daily injected dose increased to five crushed tablets and by November he felt "hooked." Prior to this, no drug had been the object of his abuse, other than marijuana. He maintained his accessibility to pentazocine by forging prescriptions and removing syringes from the dialysis unit.

During November, the patient developed chest pain and shortness of breath immediately following each injection. Physical exercise of any type exacerbated severe crushing left-sided chest pain and he became progressively weak and anorexic. At the dialysis unit he presented frequently in early pulmonary edema, chronically coughing whitish sputum, sometimes with flecks of blood. In January, 1977, immediately following the secret injection of one crushed pentazocine tablet, the patient suffered severe left precordial pain resulting in shortness of breath, hemoptysis and syncope. He was admitted to the hospital with blood pressure of 80/40 mm Hg, pulse rate 116, respirations labored at a rate of 32. Physical examination showed sparse bilateral basal rales, split second heart sound with a prominent pulmonic component, S1 gallop, murmurs of tricuspid insufficiency, hepatomegaly, and no peripheral edema. He was cachectic. Laboratory results revealed sodium level, 140 mEq/L, potassium 6.0