

decade. In other families alpha₁-antitrypsin deficiency has been accompanied by cirrhosis in infancy or childhood, without clinical pulmonary disease. A 13-year-old girl has recently been studied, who has had recurrent pulmonary infections and progressive dyspnea since age 18 months. Chest x-rays show bullous emphysema and pulmonary function tests reveal severe airway obstructive disease with a low diffusing capacity. Lung biopsy done 5 years ago show emphysema and smooth muscle hypertrophy of the small bronchioles. Immunoglobulins, sweat electrolytes and liver function studies have been normal. Biochemical and electroimmunodiffusion studies of serum reveal a deficient level of alpha₁-antitrypsin and biochemical studies show a deficiency of inhibition of elastase. Family studies indicate that the deficiency is inherited in an autosomal recessive manner. The emphysema associated with hereditary alpha₁-antitrypsin deficiency may become symptomatic in early childhood and should be considered in the diagnosis of obstructive pulmonary disease in infants and children.

- 166 *Abnormal Erythrocyte Sodium Transport in Cystic Fibrosis (CF)*. ALLEN LAPEY and JERRY D. GARDNER, NIH, Bethesda, MD (introduced by Paul A. di Sant'Agnese).

To further explore the recently reported erythrocyte cation transport defect in cystic fibrosis (CF) patients and their parents [BALFE *et al.*, *Science*, 00: . . ., 1968], we have measured sodium (Na) content and the major components of Na outflux in erythrocytes from 21 normal young adults, 22 CF patients (ages 7-27), and 20 obligate heterozygotes.

Na content of red cells from patients and heterozygotes was normal. Of the various components of Na outflux measured, there was no difference between heterozygotes and their normal male or female counterparts.

Of the major components of Na outflux, that portion sensitive to ouabain (0+) was normal in all CF groups. Abnormalities of Na outflux in CF were primarily due to that portion which was insensitive to ouabain and sensitive to ethacrynic acid (0-/E+). 0-/E+ fractional outflux was decreased in CF males of all ages (0.027±0.02/h) relative to male controls (0.051±0.02). 0-/E+ was decreased in 7 CF females over the age of 16 (0.012±0.005) compared to female controls (0.023±0.01). However, in the 4 CF females under 16, 0-/E+ outflux (0.057±0.003) was greater than that of older CF females.

The normal data from heterozygotes indicate that erythrocyte Na outflux cannot be used as a 'genetic marker' for CF. These data document altered cation transport in non-exocrine tissue from males and older females with CF. Furthermore, this abnormality does not simply reflect a general alteration of erythrocyte cation transport, but is localized to a single specific component of Na outflux, which has a characteristic requirement for metabolic substrates as well as distinctive kinetic parameters. Studies of this transport system in other tissue from patients with CF may permit a characterization of the disease at the molecular level.

- 167 *The Distribution of Fluid Intake from Mist Tent Therapy*. NORMAN ASPIN, SAMUEL K. BAU, HENRY LEVISON and DONALD E. WOOD, Dept. of Paed., Hosp. for Sick Children, and Div. of Nucl. Med., Toronto Gen. Hosp., Toronto (introduced by Andrew Sass-Kortsak).

Mist tent therapy is widely used in the treatment of cystic fibrosis to increase deposition of fluid in the lower airways of the lung. To measure the time course and extent of this fluid uptake we have introduced a solution of radioactive ^{99m}Tc into the fluid reservoir of an ultrasonic nebulizer. Subjects breathed in the mist tent for periods up to 5 h following which the distribution of inhaled radioactivity was measured with a whole body counter and a rectilinear scanner. The results show that less than 5% of the radioactive aerosol entering the tent is inhaled by the subject. Of the inhaled aerosol 90% is initially trapped by the nasopharynx and later much of this radioactivity appears in the stomach. It is difficult to detect radioactivity in the lung. Similar results have been obtained in ten subjects using three different ultrasonic nebulizers. This work indicates that very little fluid from a mist tent reaches the terminal airways of the lung. (Supported by Canadian Cystic Fibrosis Foundation.)

- 168 *Re-evaluation of Mist Therapy in Children with Cystic Fibrosis Using Maximum Expiratory Flow-volume Curves*. ETSURO K. MOTOMYAMA, LEWIS E. GIBSON, CHARLENE J. ZIGAS and CHARLES D. COOK, Yale Univ. Sch. of Med., Depts. of Ped. and Anesth., New Haven, Conn.

The measurement of maximum expiratory flow rates (\dot{V}_{max}) on MEFV curves is a simple yet sensitive method for detecting peripheral airway obstruction. In order to re-examine the efficacy of mist therapy (MT) in cystic fibrosis, MEFV curves as well as vital capacity (VC) and timed vital capacity (FEV_{1.0}) were measured in 16 patients every two weeks for a period of 4 to 5 months. In half of the patients, all of whom had been in mist tents at night for at least 6 months, the studies were done during an initial 8 to 12 week period out of mist and then a similar period in mist; in the other half the test conditions were reversed. The results were expressed as percent of predicted values.

	\dot{V}_{max} (25% VC)	\dot{V}_{max} (50% VC)
In mist (% pred. ±S.E.)	43.3±6.2	60.3±6.9
Out of mist	45.9±6.7	63.9±6.4
Significance	n.s.	n.s.
	VC	FEV _{1.0}
In mist (% pred. ±S.E.)	86.6±4.5	71.0±4.5
Out of mist	89.9±5.3	74.5±4.7
Significance	n.s.	p < 0.025

During the period without MT, 4 patients improved and 2 worsened as indicated by changes in all 4 parameters studied. As a group, FEV_{1.0} was significantly (p < 0.025) better when they were without MT. VC and \dot{V}_{max} at 25% VC and 50% VC were also closer to normal without MT but the difference was not significant. Thus, these studies failed to show any beneficial effect of mist therapy in cystic fibrosis. (Supported by PHS HD00989, NCFRF and the State of Conn.)

- 169 *Physiological Mechanisms Underlying Periodic Breathing in Low Birth Weight Infants*. HENRIQUE RIGATTO, JUNE BRADY, WARREN TICKNOR and FE DUMPTRE, Dept. of Ped. and Cardiovascular Res. Inst., Univ. of California, San Francisco, CA.

Twenty babies (b.w. 1-2 kg) were studied 106 times in the first 34 days of life. In 11 babies breathing periodically and 9 breathing regularly, we compared minute