THE NEED OF ESTABLISHING A PROGRAM TO PROVIDE FOR THE TREATMENT AND CARE OF CYSTIC FIBROSIS PATIENTS OVER THE AGE OF TWENTY-ONE

REPORT OF THE STATE DEPARTMENT OF HEALTH

Senate Document No. 4

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COMMONWEALTH OF VIRGINIA

MACK I. SHANHOLTZ, M. D.
COMMISSIONER

DEPARTMENT OF HEALTH
RICHMOND, VA. 23219

September 7, 1973

The Honorable Linwood Holton
Governor of Virginia

Members of the General Assembly
of Virginia

Gentlemen:

The State Department of Health is pleased to present a report on the need of establishing a program to provide for the treatment and care of cystic fibrosis patients over the age of twenty-one, as directed by Senate Joint Resolution 116* of the 1973 General Assembly.

An item to cover the recommended appropriation is being included in the Health Department's 1974-76 biennial budget request.

Sincerely,

Mack I. Shanholtz, M. D.
State Health Commissioner

* - Appendix A

Cystic fibrosis is a generalized disorder of the exocrine, or outward secreting, glands of the body. These glands over produce abnormal viscid secretions causing accumulation of secretions, blockage of ducts, and dilatation of the secretory organs themselves.

Principal complications are caused by the abnormal secretion in the lung and the ducts of the pancreas and liver.

In the lungs the heavy secretions of thick mucous increasingly block the tubes. Swelling and inflammation of the linings of the tubes ensue causing a decrease in air exchange.

The patient has difficulty in breathing. Even breathing at rest may require considerable effort, especially to expire air. Rapid, labored breathing occurs, as more air is trapped in the lungs and small areas of collapse occur.

As a result of the serious mechanical problem posed by this blockage, infection invariably ensues. Infection further increases the lung obstruction. The patient develops a chronic cough, with paroxysms resembling whooping cough. The bronchial tubes enlarge and lose their resiliency (bronchiectasis), clubbing of fingers and toes develops, indicating chronic oxygen insufficiency.

In the pancreas the abnormal secretions plug the pancreatic ducts preventing vital digestive enzymes from reaching the intestines. Malabsorption, vitamin deficiencies, and malnutrition ensue.

Cystic fibrosis is now known as the most frequent lethal genetic disease among children with an incidence of between 1:1,000 to 1:1,5000 live births. At present birth rates 57 to 86 new cases of cystic fibrosis can be expected in Virginia per year. The State Health Department covers 30 to 35 new cases per year. Although the disorder is present at birth the destructive process may not give signs and symptoms until varying stages of childhood.

TREATMENT

Treatment for children with cystic fibrosis must be individualized. Treatment is directed primarily toward maintenance of pulmonary hygiene. Aerosol inhalation helps liquify the viscid secretions so that they can be expectorated. Sleeping in a mist tent and intermittent positive pressure inhalation are often necessary. Physical therapy techniques for postural drainage help remove the deeper secretions from the lung. Dietary control with enzyme and vitamin replacement plus good nutrition is necessary for digestive and nutritional problems.

In the early sixties subspecialties developed around the country for the treatment of cystic fibrosis involving a comprehensive team including a pediatrician, radiologist, allergist, otolaryngologist, pulmonary function technician, physical therapist, and inhalation therapist. This is a chronic crippling disease requiring continuous care, expensive drugs, expert medical attention, and public health nurse supervision of home care.

PROGNOSIS

Up until the past twenty years the prognosis was hopeless and in the natural course of the disease all children died at a very early age. Before 1945, the majority of patients with cystic fibrosis died at an average age of about one year.

Now, however, because of increased knowledge of the pathology and physiology of the disease, new developments in treatment modalities (enzymes, antibiotics, nebulization, etc.) and the discovery of a definitive test for identification, the outlook for patients with cystic fibrosis has improved considerably.

The future holds many promising developments. Research has recently isolated a CF factor in the blood of children with cystic fibrosis which will lead to more research into the cause of cystic fibrosis so that the disease itself can be attacked rather than the effects of the disease.

Once the present "sweat test" is refined and simplified, the control of the disease will be in testing all newborns with immediate treatment for positive cases. Studies have shown that if diagnosis is made under three months of age, before destruction sets in, and if treatment is initiated at that time and maintained, the cystic fibrosis patients have an 80% chance of reaching age twenty years.

STATE HEALTH DEPARTMENT PROGRAM

In 1966 the General Assembly appropriated funds for the State Health Department to establish a Cystic Fibrosis Program through its Bureau of Crippled Children. The first treatment center was established in cooperation with the University of Virginia. Subsequent appropriations enabled the Bureau of Crippled Children to establish the Cystic Fibrosis Centers in Richmond and Norfolk and to expand the Charlottesville Program through utilization of a field team from Charlottesville to Fairfax and Wytheville, thus providing specialty facilities to all the populated areas in Virginia.

In the first full year of the program, 1967, the State Cystic Fibrosis Registry listed 52 cases from all over the state. With additional new cases over the next five years the registry grew to 160 cases.

Last year, 1972, the State Health Department treated 138 cystic fibrosis children at a cost to the state of $38,697 for treatment, equipment, and follow-up, and $7,152 for hospitalization. Private patients' hospitalization insurance and patients' fees were utilized where appropriate and in accordance with the family's ability to pay.

Federal funds utilized by the Bureau of Crippled Children, State Health Department, are limited by Federal law to those under twenty-one years of age and, therefore, the current program makes no provision for caring for the increasing number of patients with cystic fibrosis surviving past the age of twenty-one years.

PROBLEM

Today approximately 20% of all patients with cystic fibrosis are over fifteen years of age and the number of patients in their teens and twenties is steadily increasing. The Cystic Fibrosis Foundation has located ten young adults over twenty-one years residing in Virginia and the Health Department is presently following thirteen children in their late teens.

Until very recently cystic fibrosis has been a childhood disease and, therefore, the comprehensive treatment has been in the field of pediatrics. The internal medicine specialists and general practitioners have had very limited experience in the care and treatment of cystic fibrosis.

It should be emphasized that treatment must be continuous. The daily procedures for thinning sputum and mucous (nebulization), cleaning mucous out of the lungs (physical therapy), prophylaxis against constant threat of infection (antibiotics), enzyme replacement and careful monitoring are life saving and continous throughout life.

RECOMMENDATIONS

The State Health Department has through its Bureau of Crippled Children, the structured program and experienced personnel (the clinical director, physical therapist, nebulization equipment, pulmonary function and

laboratory capabilities, etc.) all concentrated for the treatment of cystic fibrosis. Therefore, it would seem prudent to extend this program to those over the age of twenty-one years. At present this could be accomplished with a relatively modest additional state expenditure.

It is recommended to the General Assembly that the Code of Virginia be amended to permit the Board of Health to provide health services for adults over age twenty-one years suffering from cystic fibrosis and to appropriate funds therefor. (Appendix B).

An appropriation in the amount of $10,000 per year would permit follow-up and treatment of cystic fibrosis after age twenty-one years (x-rays, antibiotics, nebulizers, tents, solutions, physical therapy, hospitalization, etc.).

SENATE JOINT RESOLUTION NO. 116

Directing the Department of Health to conduct a study on the needs of establishing a program to provide treatment and care for cystic fibrosis patients over the age of twenty-one.

Offered January 24, 1973

Patrons—Messrs. Edmunds, Canada and Hirst

Referred to the Committee on Rules

Whereas, cystic fibrosis is a serious hereditary disease that shortens the lives of its victims and greatly restricts their enjoyment of life; and

Whereas, cystic fibrosis patients must sleep in mist tents every night of their lives, must receive daily physical therapy treatment and constantly take medication; and

Whereas, the Commonwealth, through its Bureau of Crippled Children, is providing the cost of this expensive medical treatment for almost all cystic fibrosis patients under the age of twenty-one; and

Whereas, the present life expectancy of a cystic fibrosis child is fourteen years, but recent developments in medical research have enabled many such patients to live to early adulthood; and

Whereas, it seems proper that laws and policies should be changed so that the Commonwealth can insure proper medical treatment for all cystic fibrosis patients who are not able to afford it regardless of their age; now, therefore, be it

Resolved by the Senate of Virginia, the House of Delegates concurring, That the Department of Health is directed to conduct a study on the need of establishing a program to provide for the treatment and care of cystic fibrosis patients over the age of twenty-one and to determine the cost of conducting such program.

All agencies of the Commonwealth and of its political subdivisions shall assist the Department in this study upon the Department's request.

The Department shall complete its study and make its recommendations, including recommendations for appropriations, to the Governor and General Assembly not later than November one, nineteen hundred seventy-three.

A BILL

To amend the Code of Virginia by adding a section numbered 32-8.3 to permit the Board of Health to provide health services for young adults over age twenty-one (21) years suffering from cystic fibrosis; and to appropriate funds therefore.

Be it enacted by the General Assembly of Virginia:

1. That the Code of Virginia be amended by adding a section numbered 32-8.3 as follows:

The Board may provide home and clinic health services for young adults over age 21 years suffering from cystic fibrosis, through cooperative agreements with medical facilities or other appropriate means. Charges from persons receiving care or treatment under this section shall be determined by the Board. Funds received in payment for such services are hereby appropriated to the Board for the purpose of carrying out the provisions of this section.

2. To carry out the purposes of this act, there is hereby appropriated from the general fund of the State treasury the sum of twenty thousand dollars for the 1974-76 biennium.