

Preface

He was one of three frail siblings with recurrent diarrhoea, weight loss and a feeble apparition. A boy who, during childhood and adolescence, was not able to compete with other children of his own age and who was unable to take walks due to respiratory symptoms. He suffered from recurrent respiratory infections, haemoptysis and fevers and got frequent abdominal pain and diarrhoea after eating, in particular fatty pork meals. A man who, before the age of 35, was not able to climb the stairs due to dyspnoea and weakness and who spent most of his latter years spent bedridden with characteristic tiredness, dyspnoea and difficulties with sputum expectoration, until his death in 1849 at the age of 39 yrs. He was a composer and a pianist and his name was Frederic Chopin.

It seems highly likely that Chopin suffered and died from cystic fibrosis CF and not tuberculosis as has been claimed. CF is a disease that was unknown during the nineteenth century. Although it was already asserted in German folklore that "a child who tastes salty on being kissed has a poor prognosis" CF became recognised as a disease in the first half of the 1930s, when it was distinguished from celiac disease. In 1945 mucus secretion abnormalities were described and the term mucoviscidosis was suggested. Further development included characterisation of high sweat levels of sodium and chloride and in the 1960s it was established that CF was an autosomal, recessive, inherited disease. Approximately 20 yrs after this the gene responsible for the disease was located on chromosome seven.

When CF was first described the disease was found to be affecting children who died as a result of it within the first few years of their life. However, it became clear that the CF phenotype differs between individuals and this may, in parallel with improved treatment, explain why certain affected individuals appear to have a more beneficial prognosis with prolonged survival. The knowledge regarding CF has increased tremendously during the previous decades. It is now clear that the disease is caused by a mutation in the gene coding for CF transmembrane conductance regulator, an ion channel responsible for chlorine transport in epithelial cells. The treatment has improved substantially and life-time expectancy has increased from approximately 6 months to 30 yrs. Treatment of airway infections and obstructions, nutritional repletion, anti-inflammatory therapy and lung transplantation have contributed to improve survival outcomes with the possibility of gene therapy soon becoming a probable option.

In the present issue of the European Respiratory Monograph current knowledge regarding CF has been acquired by authors who are true specialists in the field. Most aspects of CF have been covered and this Monograph will be an inestimable source of information for pulmonary physicians and scientists within the field.

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