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## Introduction

***“To write an article of any sort is, to some extent, to reveal ourselves. Hence even a medical article is, in a sense, something of an autobiography”***  
***John Chalmers Da Costa Selected Papers and Speeches, 1931.***

There follows a miscellany on various aspects of cystic fibrosis (CF) as seen by one person, a general paediatrician from Leeds in the North of England who qualified in medicine in 1956. I was originally a general paediatrician in the true sense of the term involved in all aspects of paediatrics as was usual in the Fifties, but by the nature of the childrens' problems referred to me following my appointment as a consultant general paediatrician in 1968, I became particularly involved with respiratory, gastrointestinal disorders and neonatal care. Over the next twenty years or so this eventually evolved into an increasing and ultimately a major involvement with the treatment of people with CF and the development of the Leeds Regional Cystic Fibrosis Centre.

The details of the development of the Leeds Regional Paediatric CF service, which started with a small monthly clinic at Seacroft Hospital, Leeds in 1975 and later at St James's University Hospital, have been described elsewhere (Littlewood & Kelleher, Cystic Fibrosis News, Cystic Fibrosis Trust. Dec/Jan 1988/89), as has the later development of the service for adults with CF under the leadership of Dr Steve Conway and more recently with Dr Daniel Peckham (Conway & Littlewood, Association of CF Adults Newsletter. Cystic Fibrosis Trust. Dec. 1990). The encouragement and support of the late Mr. Ron Tucker OBE, the then Director of the UK Cystic Fibrosis Trust from 1964, and his frequent advice to parents, patients and doctors in the UK to seek an opinion in Leeds, was another major factor encouraging the early development of our CF service. Also there was invaluable financial support from the UK Cystic Fibrosis Trust and the Joseph Levy Foundation which allowed us to appoint a CF Research Fellow, Dr Mike Miller (a registrar grade doctor) and CF Nurse Specialist (Mrs. Teresa Robinson) from the early Eighties; both were crucial in the development of the CF service and were among the first specialised CF appointments in the UK.

On this local note, for a service to flourish it must be perceived to be benefiting the patients. This must have been the case as many families and patients returned year after year, often from great distances, even from as far as Hong Kong, for an Annual Comprehensive Assessment by the team at St. James's University Hospital, Leeds (to where we had moved the 2 miles from Seacroft in 1980). Paediatricians in the Yorkshire Region also found the Comprehensive Assessments helpful and in 1983 their Regional Paediatric Advisory Committee recommended that the Regional Health Authority officially recognise the service as a "tertiary referral service" for cystic fibrosis – the first to be so recognised and funded in the UK. Finally, we are grateful to the numerous colleagues at Seacroft and St James's Hospitals who have contributed their skill and expertise to the care of the patients and over 30 years to the building of a CF service which now provides full care for over 500 children and adults with cystic fibrosis.

The Early Years section of this account records early reports where the patients were likely to have had cystic fibrosis. The main format considers the developments by decades, starting each decade with a short commentary followed by details of the important references of that decade with personal comments on each. Some publications on CF have had a major influence on the understanding, diagnosis, treatment or outlook of people with cystic fibrosis. For example Dorothy Andersen's 1938 paper clearly describing the condition (Andersen DH, 1938 below) and Paul di Sant'Agnesse's recognition of the abnormal sweat electrolytes (di Sant'Agnesse et al, 1953 below) clearly come into this category. The words "above" and "below" indicate the particular reference is present either earlier or later in the document and can be identified by date. Precisely which of many other important publications deserve a "bold" rating is a purely personal opinion of a non-scientist treating many hundreds of people with CF (in excess of 600) and frequently learning whilst doing the job. Some of the articles significantly influenced our treatment of people with CF and others advanced the general understanding of the condition. The content of the whole document is of necessity



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heavily biased towards publications in English and developments in the UK; these were relatively sparse in the Forties during the Second World War. It is almost certain that some important contributions in German, French and other languages have been overlooked and for this I apologise. I am grateful to my friend and erstwhile surgical colleague Mr. Archie Crompton for some of the German translations. Where summaries were available for the later papers I have endeavoured to extract the relevant message and reduce the number of statistics as, with the modern electronic databases, the reader can obtain these from the originals if necessary. The full texts of many papers from before 1960 have been obtained from library of the Royal Society of Medicine, London.

Some research, although of basic scientific interest, is of little or no immediate relevance to the treatment of people with cystic fibrosis. Unfortunately, and understandably, in the past a significant amount of CF research has fallen into this latter category, particularly when there was no clear idea as to the nature of the basic defect. Fortunately, since the early Eighties and particularly after the identification of the CF gene in 1989, much research is clearly focused on the investigation and correction of the basic defect, by either gene therapy or pharmacological means.

At times I will digress to discuss the impact of parallel developments in related fields of medicine where progress has been central to further understanding and treatment of cystic fibrosis, for such advances have been essential to the progress in CF research - for example the understanding of the role of gluten in coeliac disease, the advances in genetics and molecular biology.

The major developments in electronic communications, particularly over the past 10 years, has been timely for an undertaking such as this. The unprecedented advances in access to previous published work via the Internet, and to Medline and PubMed in particular, and the ease of transferring visual images, have presented an opportunity which I hope will add a slightly more human touch to some of the important contributions over the years. To actually see pictures of Harry Shwachman, Paul di Sant'Agnesse, Archie Norman and many others, who have made such major contributions to our knowledge of the disorder, will I hope, add to the interest and value of this account of some aspects of the story of cystic fibrosis. These developments have resulted in the decision to publish this account on the web so as to increase the access for all those interested in the story of cystic fibrosis. The excellent and widely used website developed by Dr Daniel Peckham and his colleagues in Leeds ([www.cysticfibrosismedicine.com](http://www.cysticfibrosismedicine.com)) seemed to be the ideal place to publish this book and I am most grateful for his offer to publish it.

Finally, I am in total agreement with the late Maurice Super's observation that "it is far easier to write on these milestones in retrospect, since discovery of the gene and agreement on the transport defect. For a long time CF was fertile only in the perfusion of false dawns and in controversy between workers who could not reproduce one another's work. Now we have a fine distillate from what was once very muddy wine" (Super, 1992 below). These words were written in the heady days soon after the identification of the CF gene in 1989 and before the first papers on gene therapy were published from 1993 onwards - before the many problems in correcting the basic defect were experienced. Nonetheless, the identification of the CF gene was the definite turning point in the investigation, understanding and treatment of CF after which there was steady, focused progress in correcting the basic defect by either gene replacement or pharmacological therapy.

So cystic fibrosis is truly a disorder of our times. During the lifetime of many of us the condition was first clearly described as a specific entity, the heredity aspects were recognised, the treatment was steadily improved, the gene and its product were identified and undoubtedly within the foreseeable future one or more effective treatments for the basic defect will be available.

Very many thanks to the numerous colleagues in so many disciplines who have contributed to our knowledge of CF – not least the patients and their families. These people are too numerous to mention by name and have contributed in their various ways to the expertise and experience now available in Leeds, and indeed in many CF Centres, for treating people with cystic fibrosis. Much of their experience gained during treatment of many hundreds of children and adults with CF forms the basis of the "The Leeds Method of Management" now in 2008 in its 7th version.

Finally, I would welcome comments particularly if there are factual errors, misinterpretations or significant omissions. The text will be revised annually when there will be an opportunity for alterations and additions. The ten years of the new millenium will be added in the summer of 2010 with the first revision. So please do feel free to contact me at [history@cfmedicine.com](mailto:history@cfmedicine.com)

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