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Some previous publications on the history of cystic fibrosis

1972 Cystic Fibrosis. A Comprehensive Bibliography of the Medical Literature 1813-1972. US Department of Health Education and Welfare. National Institutes of Health. Compiled and edited by Douglas S Holsclaw and Anne Lloyd Topham. Details of over 5000 references on CF published between 1813 and 1972. This must have been a massive undertaking in the pre-computer and internet era. The entries are in alphabetical order of first author and there are also useful separate subject and author indices. By 2009 there were over 30,000 references to cystic fibrosis on the medical data base "Medline".

1990 Kulczycki LL. Five decades of cystic fibrosis (1938-1988). Acta Universitatis Carolinae Medica 1990; 36:7-12.

A brief outline of recent history as seen by Lucas Kulczycki who worked closely with Harry Shwachman in Boston from 1955 to 1970 until he moved to Georgetown. For many years Lucas Kulczycki was a leading figure in the CF world and published widely on the subject.



Figure 49: Lucas Kulczycki

1992 Super M. Milestones in cystic fibrosis. In; Warner JO, editor. Cystic Fibrosis. British Medical Bulletin 1992; 4:717-737.

Maurice Super's account of the history of CF has a strong emphasis towards the genetic advances as he was both a paediatrician and also a geneticist in Manchester.

1993 Mearns MB. Cystic Fibrosis: the First 50 Years. A review of the clinical problems and their management. In: Dodge JA, Brock DJH, Widdicombe JH, editors. Cystic Fibrosis - Current topics. Vol I. Chichester: John Wiley and Sons, 1993: 217-250.

An account of the treatment of cystic fibrosis by Margaret Mearns a UK paediatrician of great experience and one of the few UK paediatrician who was closely involved with CF for many years from the Fifties, first working with Winifred Young and then as Consultant Paediatrician in charge of the Cystic Fibrosis Clinic at the Queen Elizabeth Hospital for Children, Hackney, London.

1998 Welsh MJ, Ramsey BW. Research on cystic fibrosis: a journey from the Heart House. Am J Respir Crit Care Med 1998; 157 Suppl: S148-S154.

A review of research in CF by Michael Welsh, one of the leading US researchers in the field, tracing the "pathway of discovery leading to understanding and cure of a genetic disease". The stages can be summarised as follows - first the clinical identification of the condition, then description of the physiological/biochemical defect, identification of the gene and mutations, elucidation of the function of the gene product, understanding how mutations cause dysfunction of the gene product, explanation how mutations cause disease and the development of therapies.

1999 Quinton PM. Physiological basis of cystic fibrosis: a historical perspective. Physiological Reviews 1999; 79: S3-S22.

An interesting and very clear review by one of the pioneers of CF research particularly putting into perspective the various phases of understanding of the basic defect; in particular. discussing the two apparently distinct faces of CF – that of a mucus abnormality and one with defects in electrolyte transport. Quinton notes that CF "has advanced from innumerable speculations about its cause to a precise definition of causative mutations accompanied by accurate quantitative descriptions of their physiological effects".



The History of Cystic Fibrosis

2001 Cystic Fibrosis in the 20th Century. People Events and Progress. Doershuk CF (Ed). A M Publishing Ltd, Cleveland, Ohio. 2001

A very interesting multi-author account of the developments in CF with a strong emphasis on the North American scene edited by Dr Carl Doershuk. Carl Doershuk joined Leroy Matthews at the CF clinic in Cleveland in 1960, initially as a Senior Resident, within 3 years of Matthews starting his "comprehensive and prophylactic (preventive) treatment programme" in 1957.

There are chapters by many of the leading figures in CF in North America, such as Paul di Sant'Agnese, Paul Quinton and many others, looking back with their personal views on the developments in many areas.

Dr Doershuk kindly gave permission to use any of the illustrations in his book for which I am most grateful.

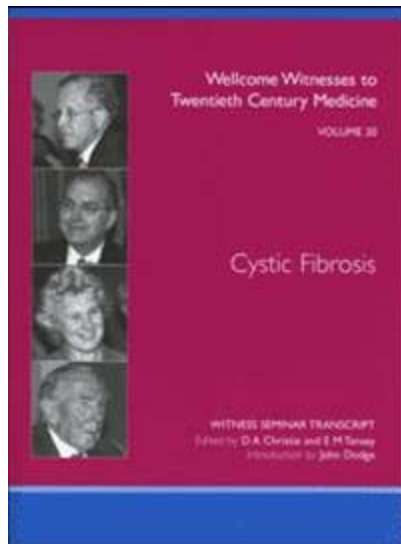


Figure 50: Wellcome Witness Seminar held in 2002



Figure 51: Jim Littlewood receiving the Rossi Medal from Prof. Gerd Doring in 2004.

2004 Wellcome Witnesses to Twentieth Century Medicine. Volume 20. Cystic Fibrosis. Christie DA, Tansey EM. (Eds). Wellcome Trust Centre for the History of Medicine at University College. London.

The transcript of a Witness Seminar held by the Wellcome Trust Centre for the History of Medicine at UCL, London, on 11 June 2002. Introduced and co-chaired by Professor John Walker-Smith retired Paediatric Gastroenterologist and Dr Jim Littlewood.

Some 40 people who had been involved with either the science or clinical aspects of CF spent the afternoon reminiscing on their experiences. In particular Dr Archie Norman and Sir John Batten both contributed as did Dr Phillip Farrell, who was visiting from the USA at the time.

2004 Littlewood JM. "Looking back over 40 years and what the future holds". The Joseph Levy Memorial Lecture of the Cystic Fibrosis Trust and the Ettore Rossi Medal Lecture of the European Cystic Fibrosis Society.

In 2004 Jim Littlewood (figure 51) was awarded the Joseph Levy Memorial Lectureship by CF Worldwide in 2004 and also the Ettore Rossi Medal (figure 52) by the European Cystic Fibrosis Society in 2004.

The Levy Lecture was given at the Opening Ceremony of the 27th European Cystic Fibrosis Conference 2004 in Birmingham. A full transcript of the Joseph Levy lecture with references can be downloaded from the UK CF Trust website www.cftrust.org.uk

2007 Littlewood JM. Chapter 1. "History". In: Hodson ME, Geddes DM, Bush A. editors. Cystic Fibrosis. Third edition. London: Hodder Arnold Health Sciences, 2007:3-20.

An detailed account of the history of cystic fibrosis up to the discovery of the gene in 1989.

2008 Fanos JH. "We kept our promises": An oral history of Harry Shwachman, MD. Am J Med Genet Part A 146A:284-293.

A fascinating account of an interview with Harry Shwachman and his wife in 1984 - some 2 years before his death.

Additional historical articles by Dr Ronald Busch

Dr Med. Roland Busch (Medizinisches Zentrum Mitte Rostock, E.-Hilzheimer-Straße 22-29, Rostock 1, DDR-2500, GDR) has written extensively on the history of cystic fibrosis and reviewed many reports from the Middle Ages onwards of



Figure 52: Ettore Rossi Medal of the European Cystic Fibrosis Society.

children many of whom may have had the condition.

1971 Busch R. Zur Erfassung der Mukoviscidose beim Neugeborenen. (Comprehension of mucoviscidosis in the newborn). *Kinderärztliche Praxis* 1971; 39:268-271. (German).

A short article dealing mainly with newborn screening which in 1971 was by examination of the meconium for excess albumin. There is mention of the suggestion of Schutt & Isles (1968, above) to use the Albustix test for detecting the excessive albumin in a solution of meconium.

1978 Busch R. The history of cystic fibrosis. *NIH Lib. Trans.* 53; 316-381.

1978 Busch R: On the history of cystic mucoviscidosis. *Deutsche Gesundhs.* 1978; 33:316-320.

This article is in German and contains many references. The English summary: Up to now the casuistry on a newborn that had died of a meconium ileus, its pancreas showing histological typical alterations, published by Karl Landsteiner in 1905 was considered to be the oldest scientific publication. From the literature published before Landsteiner preponderantly cases of gastrointestinal or mixed form of mucoviscidosis may be traced back to the first half of the 19th century. A postmortem record from Vienna in 1838 signed by Rokitansky contains the oldest scientific exactly described presentation in the form of a perforation of the small intestine accompanied by a meconium peritonitis. Besides that by means of an ancient superstition the mucoviscidosis in the folklore area may be traced back till the 17th century.

1979 Busch R. Zur Frühgeschichte der zystischen Pankreasfibromatose. *NTM-Schriftenr Gesch Naturwiss* 1979; 16:95-109. (German).

Only 11 of the 104 references are in English. An extensive review of the previous reports suggesting cystic fibrosis with pictures of Landsteiner, Heubner and Fanconi but interestingly no mention of Dorothy Andersen!

1983 Busch R. Mucoviscidosis (cystic fibrosis), a disease of unclear structure until recently. (German). *Gegenbaurs Morphologisches Jahrbuch* 1983; 129:459-465.

The summary is as follows - "Cystic fibrosis (mucoviscidosis) is described in a review article. The cause of this common lethal hereditary disease of white people is hitherto unknown. The early death of patients with cystic fibrosis, a genetic paradoxon, a lot of hypotheses and very high demands in treatment should challenge more scientists for research. There are added some short notes about the authors work in the history of cystic fibrosis".

1986 Busch R. Historical aspects of cystic fibrosis. *Wissenschaftlike zeitsschrift der Willempieck Universitat Rostock.* 1986; 35:84-87.

1987 Busch R. Cystic Fibrosis in the XIX century. *Archiwum Historii I Filozofii Medycyny* 1987; 50: 427- 434. (English with Polish summary)

The author investigated the medical literature of the 19th century to detect case records suggestive of the patients having had cystic fibrosis. During the century, notwithstanding microscopic techniques, there was little progress in research about the pancreas and meconium. There are useful references to and comments on the German literature dealing with this subject. A number of these reports suggested that the children may have had cystic fibrosis.

1990 Busch R. The history of cystic fibrosis. *Acta Univ Carol Med* 1990; 36:13-15. Cystic fibrosis is said to have arisen due to a gene mutation 4000 to 5000 years ago (but also see Busch 2005 below). Migration of peoples, gene mutations and new conditions in nourishment could have been the causes. Resulting from old cleaning ceremonies and preventing or treating uncanny effects in children, it was usual to lick the forehead of newborns and children crosswise. If one perceived a salty taste, the child was called bewitched or fascinated and was feared to die soon. The author identified such descriptions in 12 states of modern Europe. Medical documents are reviewed from the first case report until Carl von Rokitansky in 1838 (see below).

1991 Busch R. On the history of cystic fibrosis. *Nord Medicinhist Arsb* 1991; 95-98

“It is supposed that CF appeared about 3000 BC. Migration of peoples, gene mutations and new conditions in nourishment could have been the cause. Resulting from old cleaning ceremonies and preventing or treating uncanny effects in children, it was been usual to lick the forehead of newborn and children crosswise. If one perceived a salty taste, the child was called bewitched or fascinated and was feared to die soon. The author found describings in 12 states of modern Europe. Medical documents from first case reports are reviewed”.

It is noted that there are no suggestive reports from the United Kingdom and Northern Ireland, nor does the *Encyclopedia of Superstitions* by Radford and Radford, 1948, contain any key word on cystic fibrosis. The case of Pauw (1595 above) is mentioned also a girl of 3 years treated in 1673 by Georg Seger, a German doctor. Fever, vomiting, diarrhea and wasting for a considerable time before death; the only pathological finding at autopsy was an indurated and scirrhus pancreas.

2005 Busch R. What do we know about the history of cystic fibrosis? *Quebec Adult CF Newsletter*. 2005; 28-30.

An excellent brief account of many of the earlier references to children who may possibly have had cystic fibrosis. “Around the time when Palaeolithic man left Africa and entered eastern Mediterranean, that is about 52,000 years ago, a gene mutation, the gene mutation responsible for cystic fibrosis first appeared, likely in the Near East”(note also Busch 1990 above re the dates of the suggested mutation).

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