Daniel Cornelius Danielssen — A great leprologist of Norway

R. MELSOM (*)

Termos indice: Biografia.
Key words: Biography.

Daniel Cornelius Danielssen (fig. 1) was born in Bergen, Norway, the 4th of July 1815. His father was a watchmaker, and the economy of the family was not too good. The intelligent boy, Daniel, therefore had to start working at the age of 13 years in a drugstore in Bergen. At 17, however, he got tuberculous coxitis and was ill in bed for 1 1/2 years. During that time he started to study for admission to the University and, being well enough, he continued his studies in Oslo. In the fall of 1835 he began his medical studies, and took the degree with the very best grades. Later on he specialized in the diseases of the skin for one year.

In 1839 Danielssen returned to Bergen to work at the crowded St. Jórgen Hospital for Hansenians. The condition of this hospital was incredibly bad — no nurses, the patients had to help themselves. They cooked their own food, and many of them had open chronic wounds which they tended to themselves. The ventilation of the hospital was very deficient, and the stench almost unbearable. Parasitical skin diseases in these patients were very common, and Danielssen described in 1847 "scabies crustosa", later called the "Norwegian scabies".

Danielssen central position in Norwegian medicine, however, stems from his valuable work on Hansen's disease, which resulted in a concise clinical description. At that time the clinical description of Hansen's disease was very deficient, discriminating only diffusely from other diseases. Undoubtedly, Hansen's disease has existed from the oldest time; it is mentioned in the third book of Moses in the thirteenth chapter. Hippocrate did not mention the disease, but it is referred to in later Greek medical literature. The Greek called it "Elephantiasis" — not because the disease in any way can be associated with this animal, but since war elephants were more terrifying than anything else to the Greek.

Hansen's disease occurs in a great variety of forms, which have given rise to many errors in older as well as in more recent literature. On the one hand the different stages of the disease have been regarded as different illnesses. On the other hand coincidentally occuring exanthems have been

(*) Lillestrom, Norway

taken as outbreaks of Hansen's disease, and a heterogeneous mixture of other diseases have erroneously been diag- nosed as Hansen's disease. Skin chan- ges by stasis in the peripheral lymph system have also been labeled "Ele- phantiasis". In order to distinguish this illness from Hansen's disease, it has been called "Elephantiasis arabum".

The Arabs did use many terms for ffansen's disease, but the Greeks trans- lated them all into Greek by Lepra (as they did with the Zaarath of the Bible).

Fig. 1 — Daniel Cornelius Danieissen

In the time of Danielssen the terms "Lepra" and "Elephantiasis graecorum" were used indiscriminately. The terms include in addition to Hansen's disease, a series of other grave disease like scurvy, pellagra, skin tuberculosis, psoriasis and so on.

Thanks to Wilhelm Boeck and Danielssen this wilderness has been cleared up. Together they published in 1847 the pictorial atlas and the book "Om Spédalskhed" or "Elephanthiasis graecorum". This great work was translated into French in 1848 (fig. 2) and immediately made Bergen a center of Hansenology. In 1855 the work was awarded a prize on which a still existing legacy was founded. From the preface of the book we learn that in 1837 a Royal Commission was appoint- ed to study skin diseases, and that the Norwegian Parliament gave a grant to an army doctor to go abroad studying the same.

A similar grant was given to Wilhelm Boeck who studied the occurrence of Hansen's disease in other parts of Europe and how the illness was treated. At the same time Danielssen was commissioned to continue his investigations of the inmates of St. Jorgen Hospital. Likewise, the necessary means to publish their joint work were granted. In this publication the available ancient literature is comment. ed upon and discussed. The authors give an excellent and detailed description of the manifold symptoms of this disease. They distinguish between the two main types: "Lepra tuberculosa" and "Lepra anesthetiea". Furthermore they argued convincingly that ancient Elephantiasis and medieval Lepra were identical.

Danielssen was then aware of the two main types of Hansen's disease. Later on the Norwegian adopted the term Lepra tuberosa for what is to day called "lepromatous lepra", the term Lepra mixture for the borderline cases, and Lepra makulo-anesthetica for the tuberculoid type of lepra. In the book both macroscopic and microscopic observations made at the autopsies at the St. Jiirgen Hospital were thoroughly reported — furthermore detailed descriptions of the illness and a family register of Hansenians.

Boeck and Danielssen struggled with the problem of the causes of Hansen's disease. Even Moses might have been aware of the possibility that this disease was contagious. This view, common in the Middle Ages, had its spokesmen at the time of Danielssen and Boeck. Other causes were mentioned: malnutrition, bad housings and sanitary conditions,' small animals (as scabies), divine punishment, and heredity. The authors were inclined to believe in the last theory. They were, however, fully aware that Hansen's disease apart from clustering in certain families, also did occur without any observable possibility of being inherited. This they tried to explain by the hypothesis that Hansen's disease could leave one or more generations unaffected.

Danielssen took a great interest in the blood system. At the St. argen Hospital he performed thoroughly blood analysis at the standards of the time, the value of which is hard to estimate to day. The authors concluded that Hansen's disease was caused by a specific hereditary blood dyscrasia. The book also provides a complete his- torical survey of the occurrence of the disease in Norway.

The disease had many names in old Norwegian, and different names in different parts of Norway. Most common was Spedalskhet, derived from the word Hospital. In some places it was called "the Hereditary disease".

TRAITÉ

DE LA

SPÉDALSKHED

OU

ÉLÉPHANTIASIS DES GRECS,

PAR

D.-C. DANIELSSEN,
Médecin en chef des hôpitaux de spédalsques, à Bergen.

WILHELM BOECK,
Professeur de la Faculté de Médecine, à Christiania.

Ouvrage publié aux frais du Gouvernement Norwégien.

TRADUIT DU NORWÉGIEN, SOUS LES YEUX DE M. D. DANIELSSEN,

PAR

L.-A. COSSON (DE NOGARET).

Avec un atlas de 24 planches coloriées.

PARIS,
CHEZ J.-B. BAILLIÈRE,
LIBRAIRE DE L'ACADÉMIE ROYALE DE MÉDECINE,
RUE DE L'ÉCOLE DE MÉDECINE, N. 17.
A Londres, chez H. Baillière, 19, Regent-Street.

Fig. 2 — Title-page of the French translation "Traits de Spedalsktied" (Book 2780 of the Library of the Institute of Health. Secretariat of Health, S. Paulo, Brazil)

THE VIKINGS

We know that the disease occurred among the Vikings — it is mentioned both in the laws of Gulating and those of Frostating. In the Middle Ages the number of patients probably increased in Norway and in Western Europe as a whole. Several hospitals for Hansenians were built during that time, in Denmark 30, in Norway 3.

It has been claimed that the Crusades provided the link through which Hansen's disease was transmitted from the Near East to Western Europe. This can hardly be so, considering that infection rarely hits at adults. The disease did almost completely disappear in West Europe in Modern Time and so did the hospitals for Hansenians.

In Finland, Iceland and Scandinavia it persisted in the outskirts, particularly on the West coast of Norway — illustrated by this quotation of Boeck and Danielssen:

"In a report dated Nordfjord, April 12th, 1670 made by the local judge S6fren Christens on the request by the common people it says: So great are the misery and distress among us, that most of us have to eat the bark of the trees, from which many have perished and some have caught the loathsome leprosy due to some indigestable food. Where this disease occurs, we have collected money to bring the lepers into hospitals. This has been done for years, and close to 60 persons have been hospitalized. For this purpose this poverty-stricken bailiwick has spent 1400 rigsdaler. But, alas, the illness has again increased by around 50 lepers who live among us in their pitiful con- dition. We are not able to get these into hospital unless each of them brings 36 rigsdaler in addition to the 4 shillings that every man in this bailiwick annually must pay to the hospital".

That Danielssen regularly performed dissections at St. Jfirgen Hospital, provoked strong resentment among the patients. This work was, however, legalized by a Royal Commission. Danielssen was early aware of small brown bodies in the tissue from the Hansenians, which he believed to be characteristic of Hansen's disease. In 1859 Virchow visited Bergen, and Danielssen discussed his findings with him. Virchow believed, however, these brown bodies to be clumps of degenerated fat cells, but Danielssen had indeed seen the pithognomical nodules characteristic of Hansen's disease.

It has been claimed that patients suffering from other serious skin diseases were hospitalized among the Hansenians during the Middle Ages. Hansen's disease, however, presents such marked and unequivocal clinical features that it can be diagnosed early by nearly anybody in an environment where the disease is not uncommon. This seems to contradict the confusion in older literature. Dr. Vilhelm Miller Christensen, the Danish Professor of medical history, has performed extensive excavations a a forgotten churchyard for Hansenians in Sjelland. He has demonstrated very characteristic deformities of the skeletons of the Hansenians, these changes appear in almost all the skeletons he so far collected.

In 1842 the Norwegian Parliament decided to found two new hospitals for Hansenians in Bergen. One of these, Lungegarden Hospital, should on the request of Danielssen, become a general research institute for skin diseases

and not only for Hansen's disease. The other was "Pleiestiftelsen for Spedalske". The first one was finished in 1849 — a couple of years later it burned down, but was soon reconstructed in brick on a new site, and Pleiestiftelsen was built on the old one. Danielssen was appointed chief medical doctor at Lungegarden Hospital, supervising doctor at Pleiestiftelsen.

The old Hospitals for the Hansenians in Molde and Trondheim were modernized and extended. At the end of the 1860-ies these construction programs were fulfilled, and hospitals beds for 800 patients were then provided. In 1855 Health Commissions were appointed by a Royal Decree in districts where Hansen's disease did occur. The commission should register and supervise these patients, and in • addition report to the chief medical officer for Hansen's disease the number of patients and their family conditions. From 1856 we have thus had a reliable statistics of the Hansenians in Norway.

HANSEN, NEISSER, LOOFT AND LIE

In 1868 the young Armauer Hansen arrived in Bergen to work with Danielssen as his assistant doctor. At that time there was already a well established and renowned scientific milieu in Bergen.

Five years later Armauer Hansen made the discovery which forever has connected his name with the ancient disease. The discovery of the bacillus as the cause of Hansen's disease was so revolutionary that it provoked strong resistance and heated discussions. Even Danielssen, Hansen's father-in-law and good friend, stubbornly maintained the theory of heredity. Armauer Hansen published his discovery in the Norwegian Journal of Medical Science, but his results were only scantily known outside Scandinavia.

In 1879 the young Neisser, who had already discovered the gonococcus, visited Bergen. Armauer Hansen and Neisser now jointly tried to dye the Hansen's disease bacillus with the new anilin dyes, but their efforts failed. Neisser left Bergen well supplied with materials from Hansenians, and back in Germany he succeeded in dying the bacillus. He published his nice preparations in German journals. In his first publications, however, he clearly stated that the discovery must be wholly credited to Armauer Hansen.

Important scientific results were obtained at the Hansenians Hospitals in Bergen in the later part of the 19th. century. Danielssen continued his work with the pictorial atlas of "lepra maculo-anesthetica" and another one of other skin diseases. Carl Looft demonstrated in 1891 the Hansenian bacillus in the tuberculoid type (the smooth type), a very neat and painstaking work as the bacilli can be observed only in the active edges of the patches, and only in limited numbers.

H. P. lie was appointed as a doctor in 1893 at the hospitals in Bergen, where Danielssen was the chief doctor. H. P. Lie and Lyder Borten published in 1899 "Die Lepra des Auges" ; in 1904 H. P. Lie finished his paper "Lepra im Riickenmark und die Peripheren Nerven" where he proved that the bacilli of the Hansen's disease occurred in the peripheral nerves, but not in the central nerve system.

Many have tried to cultivate this bacillus in vitro, but still without success. Just as many experiments have been done to transmit the disease

to laboratory animals, and occasionally to human beings. Danielssen tried to inoculate himself 3 times the last time with pleural exudate from a Hansenian. He got necrosis on the inoculations site. Danielssen had tuberculosis infected. Armauer Hansen was married to the daughter of Danielssen, but she died from tuberculosis after one year of marriage.

INOCULATIONS AND IMMUNITY

As late as in 1960 the third great breakthrough in the research of Hansen's disease took place. Shepard obtained positive and reliable results from inoculating footpads of mice with Hansenian materials. By treating the infected mice with x-rays or "thymectomizing" them, the disease spread to other organs. Mouse experiments enable us thus today to control and investigate old and new medicines of Hansen's disease. These have shown that if the animals get diamino-diphenyl-sulf one even in very small concentrations along with the inoculation of the Hansenian material, the bacilli become inactivated, and the animals do not get ill. This medicine is to day the far most important one in the treatment of the Hansenians. It has, however, an everlasting effect in only 25% of the patients — the remaining 75% of them have to con- tinue this medical treatment for the rest of their lives.

In 1970 the ninebanded armadillo was found to be the better experimental animal. The armadillos are primitive mammals with a low body temperature of 32°C. They are considerable bigger than mice, and have a considerable longer lifetime (10-12 years). About 25% of the animals respond positively to inoculations — and they develop very rapidly, within half a year — a lepromatous Hansenia similar to that seen in human Hansenia with affections of the skin, the eyes, the lymphsystem, liver, spleen and so on. In the armadillos, however, much higher concentrations of bacilli are found. Great amounts of bacilli for medical research can thus be obtained from infected armadillos.

The immunological spectrum of Hansen's disease is somewhat complicated. Lymphocytes play an important role in this system. This immunity is present in varying strenght both in individuals suffering and not suffering from Hansen's disease. The immunity determines whether and how Hansen's disease will develop in a person exposed to the bacilli. We know that the illness does not develop in everybody who is exposed to contagion. Married couples very rarely transfer the illness to each other. In a family where one of the parents is an Hansenian, only a few of the children may be ill. If both parents are Hansenian, however, a greater number of their kids are sus- ceptible. In one-egged twins the disease follows exactly the same course.

One thing is clear : without lepra bacilli, no Hansenia. The transmission is not clearly understood, but droplet infection is most likely. It seems reasonable to assume that Boeck and Danielssen were right in believing that the disease is due to a completely or partly depressed immunity which may be congenital.

Daniel Cornelius Danielseen

SPORADIC CASES IN WESTERN EUROPE

Today Hansen's disease is completely controlled in Western Europe. In the developing countries the disease still represents a great medical and social problem. Single cases of Hansen's disease are sporadically imported to Western Europe, especially to former colonial powers like the Netherlands and Great Britain. In Norway a few such cases have occurred, a sailor from Tbnsberg in the early 1930s, a young Chinese girl from Indonesia, and two visiting workers late in 1973.

As the only country in the world, Norway has reliable statistics of Hansen's disease from 1856 when almost 3000 patients were reported, close to 2% of the population. In some districts where the infection was endemic, a higher percentage was reported and in some areas and within families almost 50% suffered from this illness. About one third of the patients could be hospitalized when the constructions of the hospitals were completed in 1860-1870. It goes without saying that preference were given to the most severe cases. Thus, without even knowing at the time this was a contagious disease, the most dangerous sources of contagion were isolated.

Danielssen, presuming that the disease was hereditary, was of the opinion that the Hansenians should avoid getting children. Armauer Hansen discovered the bacillus in 1873, and when this was accepted, a law providing forced hospitalization was enacted. This law has, by the way, later been repealed.

Danielssen was a highly gifted scientist with broad talents. His medical publications were numerous. He was vitally interested in the Bergen Museum of Natural History of which he became president in 1844 and remained so until his death in 1894. He obtained a doctor's degree both in medicine and in philosophy. Furthermore, he was strongly engaged into politics and for years member of the Norwegian Parliament. Following his death in 1894 his funeral ceremonies took place in Bergen Museum, and his ashes were kept there in a specially created urn beneath his bust.

Recebido para publicação em 23 de agosto de 1976