

Medicine in the Tropics

Leprosy

Edited by
Robert C. Hastings, M.D., Ph.D.

Foreword by Jacinto Convit, M.D.

Leprosy

EDITED BY

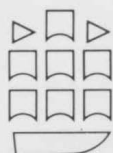
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Medicine in the Tropics Series

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Medicine in the Tropics Series

Dedication

This book is dedicated to Sarah Badiyah Sakaan, born 23 April 1983, in the hopes that her children, and her children's generation all over the world, will be free from leprosy.

Foreword

It is a privilege for me to write the Foreword to this important book, and I express my appreciation to Professor Robert C. Hastings for having given me this responsibility.

The last two decades have been very productive in valuable scientific research in the field of leprology. As a consequence, a great necessity has arisen for a comprehensive and authoritative survey of the accumulated knowledge in this complex and interesting field of medical pathology.

The collection of this knowledge and its presentation in a clear and critical manner, accessible to a broad public ranging from those interested in an introduction to the field to experienced investigators, has not been an easy task. The result of this effort is clearly successful, made possible by the fact that the contributors are all distinguished scientists who have personally experienced and actively participated in the

research movement in leprology. For these reasons, all who labor in the field of leprosy owe a debt of gratitude to the Editor and to the contributors to this volume.

This book is an example of the impact that the basic scientific disciplines have produced in the study of leprosy: a disease dominated by prejudice and negative attitudes which inspired terror and revulsion has been transformed into a model for study and teaching. The current research effort is due in large measure to the active support which the Special Program for Research and Training in Tropical Diseases of the UNDP/World Bank/WHO has provided, both in economic terms and in awakening an interest in leprosy in the international community.

Jacinto Convit
Caracas, 1985

Preface

More than twenty years have gone by since the publication of the second edition of *Leprosy in Theory and Practice*, edited by Cochrane and Davey, by the Williams & Wilkins Co. and John Wright & Sons, Ltd. in 1964. In that time a great many excellent shorter textbooks on leprosy have appeared. Many relatively comprehensive chapters on leprosy have been published in more general medical textbooks. There has been a steady flow of new information including excellent review articles on leprosy in both general and leprosy journals. In addition to these sources of information, as vital as they are, there is a need for an up-to-date textbook on leprosy — a textbook of sufficient length to present a comprehensive picture of the wealth of traditional knowledge in leprosy, and a textbook in sufficient depth to place the vast amount of new research information which has become available in the last twenty years in the context of this traditional knowledge. This book is intended to meet this need. It is intended for two audiences — those engaged or who are preparing to engage in a career in leprosy work, and those who are engaged primarily in other disciplines who need a comprehensive source of background information on the disease.

In 1985 the challenges of leprosy demand, more than ever before, the commitments and contributions of both leprologists and non-leprologists. Increasingly the problems confronting us in leprosy are the same problems confronting others working in infectious diseases, cancer, autoimmune diseases, immunodeficiency diseases, microbiology, clinical and basic immunology, biochemistry, pharmacology, and a host of other disciplines. Increasingly the basis of these problems is a lack of basic understanding of mechanisms of either health

or disease and how to apply what is becoming known about these basic mechanisms to the patient with disease and to the population at risk of contracting it. Non-leprologists have vital contributions to make to leprosy work. Career leprologists have equally vital contributions to make to those embarking on research in leprosy, and research into other diseases for which leprosy may well be a clear model. There is a need to communicate what is traditionally known by leprologists to both a newer generation of leprologists and to a newer generation of leprosy researchers whose primary interests may not be leprosy. There is an equal need to communicate relevant newer research findings to leprologists in a context which will lead to improved patient care. We hope this book will serve both these needs.

It has been a great honor to have been asked by the publishers to serve as editor of this book. The international group of contributors are the world's leading authorities in both the traditional knowledge of leprosy and in the newer advances which impact on our understanding of the disease. Despite extremely demanding schedules and a variety of other responsibilities, each contributor obviously devoted many hours of meticulous preparation to his chapter, masterfully prepared his manuscript, and accomplished what is sometimes the most difficult task — delivered it on schedule. I would like to express my deepest appreciation for the enormous talent, experience, and hard work of the contributors in making this book possible, and for the privilege of working with them.

As is probably the case with many if not most textbooks, there are dozens of individuals whose work has been vital to the production of this book. I would like to thank my colleagues at Carville and



elsewhere for their advice on a number of decisions made in connection with the book. My secretary, Renee Painter, dispatched all reminders, schedules, and correspondence with her customary superb efficiency. My thanks go to my wife, Jenny, and my son, Jeff, who have tolerated my absences without complaint. My thanks go to Dr John R. Trautman, Jr and Dr Waldemar F. Kirchheimer both for permitting me to undertake this work, and for introducing me into the clinical and laboratory

aspects of leprosy work. On behalf of the contributors, the publishers, and the dozens of other people who have worked long and hard to make this book a reality, I hope that it will serve its ultimate purpose — to hasten the day when leprosy can be eradicated from this earth and, in the interim, to help its readers care for its victims.

Robert C. Hastings
Carville, 1985

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Contents



1. The history of leprosy <i>Stanley G. Browne</i>	1	9. Treatment <i>Robert R. Jacobson</i>	193
2. The epidemiology of leprosy <i>S. K. Noordeen</i>	15	10. Eye complications of leprosy <i>Margaret E. Brand and T. J. ffytche</i>	223
3. The microbiology of leprosy <i>R. J. W. Rees</i>	31	11. Ear, nose and throat involvement in leprosy <i>R. P. E. Barton</i>	243
4. The immunology of leprosy <i>Morten Harboe</i>	53	12. Control programs in leprosy <i>Michel F. Lechat</i>	253
5. Classifications of leprosy <i>Dharmendra</i>	88	13. Experimental leprosy <i>Charles C. Shepard</i>	269
6. The pathology of leprosy <i>Dennis S. Ridley and Charles K. Job</i>	100	14. Rehabilitation in leprosy <i>Paul W. Brand and Ernest P. Fritschi</i>	287
7. Clinical leprosy <i>Roy E. Pfaltzgraff and Anthony Bryceson</i>	134	Index	320
8. Differential diagnosis <i>D. L. Leiker</i>	177		

The history of leprosy



INTRODUCTION

Although leprosy is often referred to as 'the oldest disease known to man', the origins of which are lost in the mists of antiquity, several lines of evidence throw doubt on such assertions. Signs of osteoarthritis, tuberculosis and infections certainly exist in bone, but no bones or representations in stone or pottery bearing unmistakable signs of leprosy have come to light from antiquity, and the imprecisions and uncertainties of terms in ancient texts that may have been translated as leprosy at some time or other are now widely recognized.

EARLY EVIDENCE

No objective evidence of leprosy predates the findings of recent excavations in the Egyptian oasis of Dakhleh, which have disclosed four leprosy skulls in white representatives of the ruling class buried there in the second century BC (Dzierzykray-Rogalski, 1980). No sign of leprosy has been found in Egyptian skeletons from 6000 BC onwards or in skeletons unearthed at Lachish, dated 700–600 BC.

The earliest skeletal remains from the present era showing indubitable signs of leprosy are of two mummies, Coptic Christians, found in a burial ground in El-Bigha in Upper Egypt, which date from the sixth century AD. The hands and feet, according to Smith & Dawson (1924), show clear evidence of mutilating leprosy, and Møller-Christensen & Inkster (1965) confirmed the diagnosis of leprosy after examining the bones of the extremities and the skull. Andersen (1969) feels justified in ascribing to leprosy bony changes in a female skull from the same period.

Written records

The earliest written records describing true leprosy, which were most probably preceded (perhaps by centuries) by orally transmitted traditions, come from India, in recensions brought together in about 600 BC. These are surprisingly full and accurate, and testify to a high degree of observation and diagnostic skill (Dharmendra, 1967; Lowe, 1942). The disease is called *Kushta*; it is differentiated from vitiligo, and affects the skin of the trunk and extremities. Then there is a vague reference to slaves from Darfur, in the Sudan, coming to Egypt at the time of Rameses II and bringing a chronic non-irritating skin disease with them. Africa may thus vie with India for the undeniable distinction of being the cradle of leprosy. The possibility of a multifocal origin of a disease that is so widespread at present and that has been so widespread in the past cannot of course be excluded a priori.

From India, true leprosy spread to China in about 500 BC and thence to Japan. Good clinical descriptions of true leprosy come from China, dated c. 190 BC: they mention cutaneous lesions and anesthesia, hoarseness and eye damage. As for causation, they mention overcrowding, promiscuity, lack of hygiene and dirt.

False leads

Other allusions to conditions that have been thought at some time or other to be leprosy are far from convincing. References to Chons' swellings, urededu and tumors of the god Xensu in the Ebers' papyrus, dated about 1350 BC, are far too vague to bear the weight of the precise identification with

leprosy sometimes placed upon them (Feeny, 1964). Similarly, the resemblance between the features depicted on a pottery grain storage jar unearthed in the Amenophis III temple at Bethshan in Palestine, dated from about 1450 BC, and the changes due to advanced lepromatous leprosy (Yoeli, 1953, 1968) is probably fortuitous and imaginary. The distinguished French medical historian Zambaco (1914) thought that he could detect leprosy lesions in the dry skin of certain Egyptian mummies, but to most commentators his arguments are unconvincing.

In the nature of things, it is not to be expected that incontrovertible evidence of the origin of leprosy — or, for that matter, of other cosmopolitan diseases — should be forthcoming.

The influence of the Bible on Western attitudes

In the Western world, many of the assumptions concerning the history of leprosy derive from Christian tradition, which is itself based upon translations of Old Testament texts uncritically accepted by succeeding generations. The underlying connotation of Hebrew *tsara'ath* in the Mosaic code (Leviticus 13 and 14) seems to be ritual uncleanness associated with changes in the color of a surface — i.e. of human skin, of cloth or leather, or of the damp walls of dwellings. There is much uncertainty concerning the precise meaning of the clinical terms used in the original text, and no positive evidence for the existence of true leprosy in the lands of the Fertile Crescent in Old Testament times can be deduced from these writings.

Despite the historical contacts between the Empires of the Medes and Persians and the Indian subcontinent, there are no records of leprosy having been imported at that time into ancient Persia or neighboring countries.

TRUE LEPROSY IN THE WEST

The earliest written records of a disease closely resembling — and probably identical to — modern leprosy in the lands of the West are from

Greece, and date from about the turn of the third century BC (Andersen, 1969). It is more than likely that the soldiers of Alexander the Great, returning from the Indian campaign about 327–326 BC, unwittingly brought back with them *Mycobacterium leprae* together with the booty of silks and spices from the fabulous Orient. Andersen has collected data from various Greek sources that make this suggestion very plausible. The observant and knowledgeable Greek physicians were at this time confronted with a disease showing novel features, a 'new disease' in fact. Had they seen it before, it is tolerably certain that they would have recognized it. 'The original records have been lost', writes Andersen (1969), 'but Straton, a disciple of the Alexandrian physician Eristratos (c. 300–250 BC) is quoted by Rufus of Ephesus (AD 98–117) as giving an accurate description of low-resistant leprosy.' This new disease was called elephantiasis. Later, the word *Graecorum* was added, to distinguish it from *elephantiasis Arabum* (which we know today as bancroftian filariasis). To confuse the issue still further, *lepra Arabum* was the equivalent of *elephantiasis Graecorum*, which we today call leprosy. Among the Greeks, popular descriptive appellations were leontiasis and satyriasis: Aristotle (384–322 BC) mentions the latter (Browne, 1975).

It is true that Hippocrates (c. 460–377 BC) used the term *lepra*, but his descriptions indicate a blotchy condition of the skin like summer prurigo or pityriasis simplex. He never hints at the neurological manifestations of leprosy.

There has been some speculation that Greek merchants following the long caravan routes overland to the East may have brought leprosy back with them, but no firm evidence exists. Once leprosy had established itself in Greece, however, because of the maritime connections of Greek merchants and sailors it spread slowly to the lands of the Mediterranean littoral. When Rome replaced Greece as the major military power in the region, the smouldering leprosy endemic in Italy was augmented by the returning troops of Pompey who had been fighting in the Egyptian campaign (62 BC), according to Pliny the Elder (AD 23–79).

The linking of the ceremonial defilement of the *tsara'ath* of the Mosaic code with Greek thought is usually traced to the Septuagint translation of

the Hebrew originals of the Old Testament books into Greek by a group of Alexandrian scholars in about 200 BC. Wherever the word *tsara'ath* or its cognates appeared in the Old Testament, the Greek equivalent *lepra* is offered. The Hebrew *tsara'ath* and its cognates were translated by Greek terms already in use by physicians for a scaly skin condition (e.g. summer prurigo and psoriasis), a word also applied to bark and flakes. This word was *lepra*.

In many and diverse cultures, however, leprosy itself and its victims were surrounded by deeply ingrained beliefs and popular legends. For instance, in Japanese Shintoism, the same word did duty for both leprosy and sin. Chinese influence in Southeast Asia probably accounts for the association of a repulsive skin condition and guilt. In the ancient civilizations of Asia Minor, many words designate a whole range of skin abnormalities calling for quarantine and ritual purification. The Jews therefore cannot be regarded as the originators of the 'leprosy concept', since in many ancient civilizations (China, Egypt, India) there seems to be an association between sin and skin disease. The word *lepra* is the one used by the Evangelists (Matthew, Mark and Luke) in their Gospels as the equivalent of the everyday colloquial Aramaic. In the Palestine of Our Lord's day, the word probably included the ritualistic overtones of the original Hebrew, and its association with sin, divine displeasure, and divine power and punishment. The accepted Latin medical terms for true leprosy at that time were *elephantiasis Graecorum* or *lepra Arabum*, but the Greek equivalents of neither of these appears in the New Testament. This, then, would appear to be the situation at the beginning of our era: leprosy was spreading slowly eastwards from India, and to the lands bordering the Mediterranean.

There is presumptive evidence that it was present in Palestine at the time of Our Lord. The association of a chronic skin disease with Mosaic ideas of ceremonial uncleanness, sin and punishment was widely current there. Those so suffering were relegated to the uninhabited border country (Luke 17.11); they regarded themselves, and were regarded by society, as 'unclean' (Mark 1.41). They were 'cleansed', rather than healed (Mark 1.42, Luke 17.14; but not in Luke 17.15).

'Cleansing the leper' was seen as an authentication of the Messianic mission (Luke 7.22), and the disciples were enjoined to imitate their Master (Matthew 10.8). There are no records of apostolic obedience to this command.

Lucretius, the Roman poet who lived 91–55 BC, is credited with mentioning the disease in his *De Natura Rerum* in a passage translated as follows:

High up the Nile midst Egypt's central plain
Springs the dread leprosy and there alone.

Early descriptions of true leprosy appeared under the term 'elephantiasis' in the writings of Celsus (25 BC–AD 37), although the disease was very rarely seen in Italy in his day. Fanciful accounts of clinical signs conceal evidence that he had probably observed for himself the skin changes he describes. The Roman historian Pliny the Elder (d. AD 79) amplifies the records of Celsus.

CLINICAL LEPROSY DESCRIBED

The most complete account of the clinical signs and symptoms of leprosy (called *elephas*) as it appeared in the Western World comes from the prolific pen of Aretaios, writing about the year AD 94. He mentions macules and nodules, the lion-like features of the long-standing affection, the long silent period before signs appear, the possibility of spontaneous arrest, ozena from chronic nasal obstruction, the loss of hair, and damage to the eyes. He also refers to plantar ulceration and loss of digits, and pain in peripheral nerves. He regards leprosy as a generalized infection of the whole body, probably communicable and probably transmitted via the respiratory system. His accurate and full descriptions leave no doubt that he was familiar with lepromatous leprosy. However, there are no descriptions that could apply to high-resistant, or tuberculoid leprosy.

The influence of the great Galen (AD 130–201) on the thinking of his time and subsequently about everything to do with leprosy can only be regarded as confusing, whatever his knowledge and reputation in other branches of medicine. He refers to both *elephes* and *lepra*, but neither of these conditions corresponds precisely or clearly to true leprosy.

According to Andersen (1969), Johannes Damascenus (AD 777–857), who was responsible for the making of a medical terminology in Arabic, was the first author in the West to describe leprosy under the name *lepra*. Leprosy, then, was by this time firmly established in the world of Greece and Rome. It was a chronic and usually progressive general disease involving several organs and systems. To medical practitioners and historians in these countries, it was a disease entity, while elephantiasis (or *lepra*) was mysterious on several counts — it did not have the connotation of the old Hebrew ideas of ritualistic defilement or uncleanness.

This, then, would appear to be the situation in the early years of our era: leprosy was spreading slowly eastwards from India, and to the lands bordering the Mediterranean. The earliest reference to concern for leprosy sufferers is the founding by Christians of a hospital in Rome early in the fourth century (Mercer, 1915), and in AD 372 St. Basil established a leprosy hospital in Caesarea.

Objective criteria for establishing the existence of leprosy

In the face of all the verbal uncertainty in the written records of leprosy and the absence of objective clinical criteria accepted generally in the ancient world, it is reassuring to refer to indubitable evidence in the bony skeleton of low-resistant leprosy. By his osteopaleopathological researches, reinforced by clinical observations and radiological examinations in living sufferers from leprosy, Møller-Christensen (1953, 1961) has shown that the anterior nasal spine and the alveolar process of the maxilla are specifically eroded in low-resistant leprosy — and not by tuberculosis, syphilis or trauma. (The erosive changes in phalanges are evidence of peripheral neuropathy, and hence are not pathognomonic.) By the application of this criterion to osseous remains found in various countries, it is possible to determine the existence of leprosy in any community. Of course, sufferers from leprosy might have passed their days and ended their lives in rustic obscurity, far from urban burial grounds, and their social status may have precluded (in Egypt, for instance) expensive embalming rites. In any case, it is a fact that very few

skeletons have been discovered showing evidence of leprosy. An exception is the burial ground at Naestved in Denmark, attached to a medieval monastery and evidently catering for sick folk suffering from advanced leprosy (Møller-Christensen, 1953). This observation has proved of inestimable value in elucidating the spread of leprosy and in providing unequivocal evidence to resolve definitively doubts and suppositions that have long bedevilled medical historians. A case in point is that of Robert the Bruce, the Scots chieftain (1274–1329). On literary grounds, based mainly on a single reference by a possibly biased Franciscan monk writing in England a century after his death, the diagnosis of leprosy was at least doubtful, but a plaster cast of the skull, made when the remains were briefly exhumed in 1819 in Dunfermline Abbey, permitted a positive identification. (Rennie and Buchanan, 1978).

Examination of skeletons removed from burial grounds in England has shown that leprosy certainly existed in early Saxon times in Dorchester (Reader, 1974), and in the later Saxon period (Wells, 1962), especially in Norfolk (Wells, 1967), and in the post-Conquest era near Scarborough (Brothwell, 1958). Similar findings are documented from the continent of Europe, all of which indicate the slow spread of leprosy in the wake of soldiers, merchants and administrators. As the endemic progressed northwards and westwards in Europe, more and more people were affected.

The Orders of Chivalry

The medieval Orders of Chivalry have been historically and traditionally associated with leprosy. The Order of St. Lazarus of Jerusalem paid special attention to the sick and needy beggars afflicted with leprosy. It was a condition of appointment that the early Masters of the Order were themselves actually victims of the disease, and the Knights of the Order have been noteworthy for their concern. Founded in 1050, the Order had its headquarters in a center appropriately named Burton Lazars, situated in the rural midlands of England. It became very rich and influential, attracting to itself over the years benefactions and endowments of land from individuals and distant ecclesiastical institutions that had been founded

for the benefit of leprosy sufferers. The Order of St. John of Jerusalem is still active in countries like France, and although mainly Roman Catholic, counts Protestants among its members. Perhaps the best-known and largest of these Orders of Chivalry is that of Malta, or, to give it its full title, The Sovereign Military Hospitaller Order of St. John of Jerusalem, of Rhodes and of Malta. Founded before the taking of Jerusalem in 1099, the Order has from the first been especially concerned with leprosy sufferers, initially in countries bordering the Mediterranean and more recently in Asia, Africa and South America. Today, it is responsible for the publication of *Acta Leprologica*.

The Crusades

The influence of the Crusades on the spread of leprosy has long been debated. For some, the returning crusaders introduced leprosy into Christian Europe; for others, those who had caught leprosy during their sojourn in the countries of the Ottoman Empire in which leprosy was prevalent, served to increase the dimensions of the existing endemic when they returned home. A serious theological threat was posed by the fact that Christian warriors engaged in a Crusade that had attracted episcopal blessing, had somehow contracted a disease widely regarded as divine punishment for sin. The conundrum was rather cleverly resolved by assuring the faithful (victims and relatives) that the sufferers were indeed privileged to share the sufferings of their Divine Master. Their return (from 1096 onwards) coincided in time with a wave of Christian charity that created almshouses and hospices for those afflicted by leprosy. Royalty and high-born ladies vied with each other for the privilege of kissing the ulcerated feet of 'Christ's poor' and showing them especial compassion.

The example of St. Francis of Assisi in his attitude to sufferers from leprosy generated a wave of Christian concern throughout the Western World, inspiring both high and low to care for the neglected and shunned victims of the disease, and thus helped to transform conventional attitudes towards them.

During the fourteenth century, to judge by the founding of new places of refuge for victims of leprosy, the scourge was spreading, but in England

the serious famine of 1325–6, followed by the Black Death of 1349 (which is said to have killed off a third of the population) must have been especially lethal to the undernourished and flea-ridden inmates of the lazarets. Certain it is that the leprosy endemic began to wane in England, and a century later (in 1470) a Royal Commission appointed by Edward IV reported a marked decrease of leprosy. From that time, the dimensions of the endemic problem of leprosy have progressively declined.

In France, it is thought that about 2000 lazarets and hospices for the victims of leprosy existed at the height of the endemic in that country.

The confusion between leprosy and syphilis continues to interest medical historians. The medieval reference to 'leprosy' being contracted 'by carnal intercourse with women in stews', and its hereditary nature and response to inunctions of mercury, point to syphilis rather than to leprosy. As a matter of interest, mercury is said to have been used as treatment for 'leprosy' in China as long ago as 2300 BC. The vexed question of the introduction of syphilis into Christendom by the returning shipmates after the early Columbian expedition (1492) is still not resolved.

THE SPREAD OF LEPROSY

Leprosy in Britain

In Britain, the earliest legal enactment in which leprosy is specifically mentioned is the Code of Laws promulgated in Wales during the reign of Hoel (Hywell Dda), who died in AD 950. Somewhat later, during the reign of Edgar in England, a law was passed making leprosy a valid cause for divorce. During the reign of Henry I, probably around 1100, a statutory measure entitled *De Leproso Amovendo* regulated the movements of those afflicted with leprosy. Much later, when leprosy was still much feared though less prevalent, Edward III issued (in 1346) a proclamation to the Mayor and Sheriffs of London expelling 'lepers' from the city. There are many descriptions extant of life in medieval leprosy hospices, of the rules and regulations governing admission, conduct, dress and the like. These hospices were a kind of prison, monastery and almshouse combined. In-