LEPROSY.—(Synonyms: Gr., λέπρα (from root meaning scaly); Lat., *lepra* (in classical Lat., *lepræ*), *elephantiasis Græcorum, satyriasis, leontiasis, lepra Arabum*; Fr., *lèpre*; Ital., *lebbra*; Ger., *Aussatz*; Norweg., *spedalskhed*.

DEFINITION.—Leprosy is a chronic, contagious, and infectious disease, produced by the bacillus lepræ; characterized by the formation of new growths in the skin, peripheral nerves, and internal viscera, producing various deformities and mutilations of the human economy, and usually ending fatally.

SYMPTOMS.—Clinically, leprosy occurs in two quite distinct forms. These were termed by Danielssen and Boeck [1] the "nodular" and the "anæsthetic." The terms "lepra tuberosa" and "lepra maculo-anæsthetica," as adopted by Hansen and Looft,[2] seem more acceptable, as they more nearly express the condition. While, as a rule, each affection runs its special course and is marked by symptoms so entirely different from those belonging to the other as to appear as a distinct disease, yet some cases exhibit symptoms which are common to both forms, and in these the relationship is evident. They have a common etiological factor, the bacillus lepræ, which, however, is in a different anatomical location, and varies as to numbers in the two forms. As in the case of other contagious and infectious diseases, the clinical history may be divided into stages.

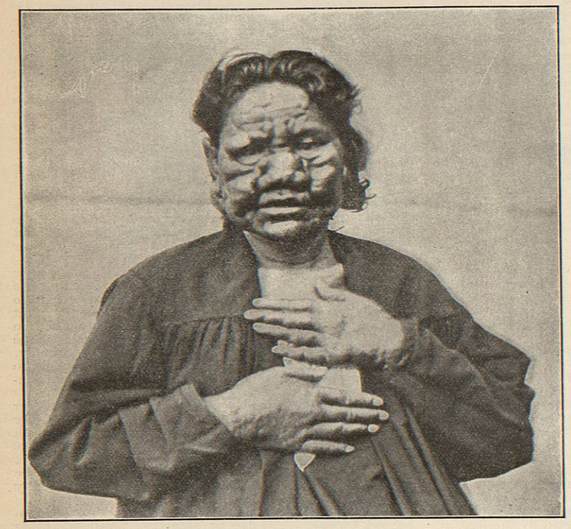

FIG. 3188.—Tubercular Leprosy. (Dr. James Nevins Hyde's photograph of a Leper in the Sandwich Islands.)

The *period of incubation* has been carefully studied by competent observers, and cannot be said to have a definite length. It is estimated to extend over a period of from a few weeks to many years. Before the eruptive and characteristic stage develops, various prodromal symptoms such as might precede any infectious disease occur. Among these may be mentioned fever, chilliness, malaise, headache, mental depression, drowsiness, pains in the limbs, and various anomalies of the motor and sensory apparatus, especially in the limbs. Those preceding the maculo-anæsthetic form are more variable, and are chiefly symptoms which naturally arise from nerve involvement, such as pruritus, formication, pain, hyperæsthesia, etc. These may be slight or very severe, and may last for only a few weeks or perhaps for many years.

Lepra Tuberosa.—In the skin this type of leprosy appears first as a macular eruption, of varying persistence, which may come and go, but in which finally the skin becomes infiltrated with characteristic tubercles. Their commonest location is on the forehead, cheeks, nose, chin, and ears, the forearms and the thighs. Unna [3] says that the lobe of the ear is a favorite and early site. The macules are well defined, and are round, oval, or irregular in shape; in size they range from 1 to 10 cm. or more in diameter. Their color, depending on the race of the patient and the age of the lesion, varies from a light-red to a purplish or bronze shade. They may appear slightly elevated or infiltrated, or quite smooth and shiny, and somewhat hyperæsthetic. Sooner or later these patches become permanent and infiltrated with tubercles which are pea-sized, yellowish or reddish-brown, and which enlarge more or less rapidly, some of them becoming as large as a walnut or larger. The development of these is not limited to the site of the macules, for some appear on apparently normal skin, and their efflorescence is preceded by febrile symptoms, more or less severe; nor do they appear simultaneously, but rather in successive groups, each new efflorescence being preceded by febrile symptoms, and perhaps epistaxis. Although the eruption may occur on any part of the cutaneous surface, it is uncommon on the palms and soles, and is rarely found on the scalp and glans penis. In typical cases of tubercular leprosy, the face presents a characteristic appearance, so much so that the disease has been termed "leontiasis" (or lion-face) from the fancied resemblance which the distorted features suggest. The eyebrows are practically always the seat of nodules in greater or less abundance. The brow appears thickened, and when the nodules are well formed the hairs are lost. The facial appearance of a leper who has the tubercular form of the disease, is well described by Thin,[4] as follows: "The thickened skin of the forehead, studded with unequal tubercular masses, and marked horizontal furrows; the tumid, greasy cheeks, uneven with tubercles; the everted lips; the nose thickened, widened, flattened, and crushed like a negro's; and the projecting nodular ears, present an appearance which distinguishes leprosy from all other diseases, and which requires to be seen once to be always recognized." At times, by confluence of individual tubercles, large plaques are formed. (*Lépromes en nappe*, Leloir, which are dark in color and which desquamate slightly.) They are inert, lasting sometimes for years unchanged. They may be the seat of pruritus, or the sensation may become less acute; the hair falls in the involved regions and they may finally ulcerate. These large plaques usually occur on the limbs and, according to Danielssen and Boeck,[1] indicate an unusually chronic case. The tubercles may undergo ulceration, and discharge a yellowish-brown, viscid fluid, which may form crusts. Some of these ulcers soon heal, especially under appropriate treatment, while others extend deeply, becoming gangrenous and destroying much tissue. Bones are laid bare; tendons, ligaments, joints, and even whole members, such as fingers and toes, are destroyed. Usually at this extreme stage symptoms of the maculo-anæsthetic type are also present. During the course of the disease, the glands of the axilla, groin, neck, and throat become enlarged. In the latter situation, this adenopathy may interfere with breathing and swallowing. Eventually, the glands soften and break down, forming fistulous tracts discharging large quantities of material. At times, without apparent reason, the disease remains stationary for long periods, then suffers an exacerbation. Intercurrent diseases, such as variola, pleurisy, and pneumonia, may cause it to disappear temporarily.

Lepra occurring in children retards the physical growth and the development of the sexual organs, arresting the functions of the latter. Menstruation may be delayed or entirely inhibited. When the disease occurs after puberty, the menopause may be prematurely brought about, and the procreative faculty lessened or lost. Bracken,[10] in a report of cases in Minnesota, states that twenty-one out of thirty-four patients were married, to twenty of whom seventy-eight children were born. Alopecia, especially of the eyebrows and eyelashes, usually occurs, but the scalp is rarely attacked. Occasionally the nutrition of the nails is disturbed, as evidenced by thinning, thickening, or other deformity. The secretions of the sebaceous and coil glands are early increased, but later diminished or entirely lost in the affected area. Comparatively early in the disease, small, flattish tubercles form on the conjunctiva and cornea, extending to and involving the iris, and gradually filling the anterior chamber. The eyeball swells and the lids cannot be closed. There is pain, and the lachrymal secretion is increased. Later, the mass softens and contracts, the secretion lessens, pain stops, and the lids can again be closed (Danielssen and Boeck [1]). Other and more chronic processes occur in the eye during the course of the disease. According to Hillis,[5] throat symptoms occur during the febrile attack. The same author states further that patches having raised crescentic edges, situated at the back of the pharynx and on the roof of the mouth, the back of the throat and the uvula, which are uniformly red and congested, are pathognomonic of leprosy. Later, the epiglottis, vocal cords, and other structures in the larynx become studded with tubercles, as does also the nasal septum; and when ulceration occurs the cartilage and bony framework of the nose are destroyed, producing the characteristic deformity. Morrow believes that the earliest manifestations of leprosy in most cases are located in the mucous membrane of the pharynx and upper air passages, as shown by alteration in the voice, rhinitis, increased nasal and salivary secretions, and sometimes epistaxis.

Danielssen and Boeck [1] have described an acute form of leprosy similar to acute tuberculosis. It is manifested by a continuous fever of about twelve days' duration, when, with a sudden efflorescence, raised, shiny, bluish spots appear over nearly the whole body. These rapidly increase in volume and hardness, become confluent, and progress as far in a few weeks as does the ordinary form in years. With the appearance of the eruption, the constitutional symptoms abate, and after the tubercles have softened the affection becomes chronic. In any case in which the cutaneous exanthem fails to appear, the patient usually dies of pneumonia, pleurisy, or meningitis in the course of a few days.

The physical condition which is the ultimate lot of the unfortunate victim of tubercular leprosy cannot be equalled in any other disease. Leloir [6] graphically describes it thus: "If the patient does not die of some intermittent disease or special complication, the unhappy leper becomes a terrible object to look upon. His deformed, leonine face is covered with tubercles, ulcers, cicatrices, and crusts. His sunken, disfigured nose is reduced to a stump. His respiration is wheezing and difficult. A sanious, stinking fluid, which thickens into crusts, pours from his nostrils. The nasal mucous membrane is completely covered with ulcerations. A part of the cartilaginous and bony framework is carious. The mouth, throat, and larynx are mutilated, deformed, and covered with ulcerated tubercles. The patient breathes with the greatest difficulty, and is threatened with frequent fits of suffocation, which interrupt his sleep. He has lost his voice; his eyes are destroyed; and not only his sight, but his senses of smell and taste have completely gone. Of the five senses, hearing alone is usually preserved. Owing to the thickened and pachydermic state of the skin of the limbs, which gives to them the appearance of elephantiasis, and to the presence of ulcerating tubercles, crusts, and cicatrices, the sense of touch is abolished. Usually at this time the peripheral nerves are involved, so that the symptoms of both the tuberous and the anæsthetic type of leprosy are present. The patient suffers excruciating pains in the limbs, and even in the face, while the ravages of the disease in his legs render walking difficult and even impossible. From fistulous openings in the hypertrophied inguinal and cervical glands pus flows abundantly. In certain cases the abdomen is increased in size on account of involvement of the liver, spleen, and mesenteric glands. With these visceral lesions, the appetite is irregular or lost. There are pains in the stomach, diarrhœa, bronchial and pulmonary lesions, intermittent febrile attacks, and a hectic state. The peculiar smell, recalling that of the dissecting-room, mixed with the odor of goose's feathers or of a fresh corpse, was recognized but badly described by authors in the Middle Ages, who compared it to that of a male goat. This is the complexus of symptoms which the patient presents, unless some fatal complication has come to his relief. In this light one can understand how, in the ancient poem of Job, leprosy is called 'the eldest daughter of Death.' Nevertheless, in spite of his condition, the unhappy leper, although in great prostration, commonly preserves his intelligence unaffected to the end. I have been struck," continues Leloir, "with the calm stoicism with which the Norwegian lepers supported their misfortune, and with the indifference or even gayety of the lepers in Italy and other countries, and with the care which they gave to their toilet. I have never seen a leper ask for death, and I do not know of an instance of suicide among these patients, who observe with the greatest resignation the slow and progressive decomposition of their bodies."

Lepra Maculo-anæsthetica.—In this variety, the bacilli are located chiefly in the neuroglia of the peripheral nerves, and consequently the symptoms exhibited in the part supplied by the affected nerves are those which would naturally follow their irritation, compression, or degeneration. Chief among these are the development of spots or macules, bullæ, muscular atrophy, anæsthesia, motor paralysis, and finally mutilation by loss of parts. There is no regular sequence observed in the evolution of these symptoms. Usually, however, the maculæ and bullæ are among the earliest manifestations, but their appearance may be delayed for years.

The course of this form of the disease is exceedingly chronic, its average duration being estimated at about eighteen years. The appearance of the spots is usually preceded by anomalies of sensation, such as formication, a sensation of burning or stinging, or pruritus. The size of the lesions varies from that of a fifty-cent piece to that of the palm or larger. By peripheral extension and coalescence, large, irregular areas, having a curved contour, may be produced. At first they are reddish in color, changing with age to yellowish or brown, or even darker shades, when they tend to become slightly elevated and to desquamate. Their centres become depigmented and anæsthetic, while the border may be hyperpigmented and hyperæsthetic. Their commonest seats are usually considered to be the back, shoulders, face, arms (especially about the elbows), the nates, and around the knees. When the spots are fully developed, they may cover very large areas of the body surface. In the anæsthetic portion, and at times extending beyond it, the production of

sweat is entirely suppressed, and the hairs become white. At this stage the disease may remain apparently quiescent for a long period of time, the only symptom indicative of interference with the general health being neuralgic pains. Danielssen, quoted by Thin, states that he has seen spots remain unchanged for from eighteen to twenty years, during which time the patient suffered so little that he required no medical attendance. He adds that he has seen the anæsthetic spots on mucous membranes accompanied by redness and thickening, which disappeared without ulceration. The formation of bullæ is characteristic of this form of leprosy. They may be the initial symptom, in which case they are usually smaller and more numerous than in other affections, and are either hyperæsthetic or are normally sensitive. The older are often single, large, and may be anæsthetic. They appear suddenly, and, if short-lived (which is the rule), have serous contents, the latter becoming purulent when the lesions persist. They heal after rupture, leaving a pale scar, or, by infection, develop into deep-seated ulcers. When the nerve trunks are the seat of severe neuritis, they become much thickened, and can be readily felt by the finger of the examiner. The fusiform enlargement of the ulnar nerve behind the olecranon process at the elbow is characteristic. Other nerves especially involved may be the tibial, peroneal, and, less often, the radial, median, brachial, and cervical. Atrophic changes are noticed first in the interosseous muscles of the hands, the thenar and hypothenar being the next involved; and the atrophy extends thence up to, and involving, the muscles of the forearm. In a lesser degree, the same process occurs in the lower extremities. This atrophy in the hands produces the "lepra claw," in which the proximal phalanges are extended, while the middle and distal are more or less flexed. All the muscles of the face may atrophy, leaving it expressionless. By paralysis, the face may be drawn to one side; the lower lip, by involvement of the orbicularis oris muscle, may droop so as to make it difficult to close the mouth, thus allowing the saliva to flow over the chin. By the involvement of the orbicularis palpebrarum muscle, the lower eyelid may droop, making the closure of the eye impossible, thus rendering it liable to external injury, which produces at times ulceration and destruction of the globe. The perforating ulcer of the foot which often appears in people who go barefooted, is a trophic affection, added in its development by pressure, and is usually located under the heel or ball of the foot. It may be deep and lead to necrosis of the bone, and is exceedingly difficult to heal. The phalanges of the feet and hands are attacked with necrosis, and complete exfoliation of the bones occurs. The fingers and toes may thus disappear, leaving only small, soft processes, upon which a sort of nail remains, to identify the member. Hansen and Looft[2] regard this necrosis as being largely due to external injury to a part of low vitality. To this stage of the disease "lepra mutilans" was the term formerly applied. Owing to a lack of unguent from its glands, the skin becomes dry, fissured, and the seat of ulceration.

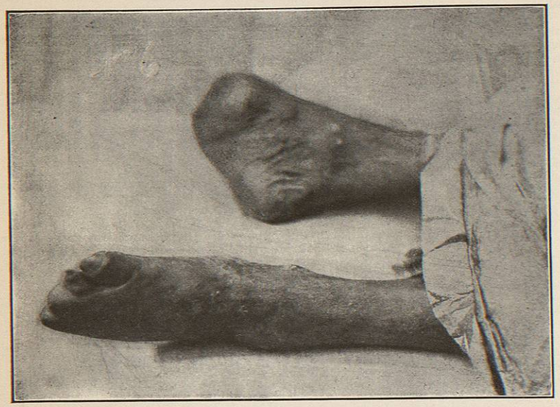

FIG. 3189.—Anæsthetic Leprosy with Mutilating Results. (Dr. James Nevins Hyde's photograph of a Leper in the Sandwich Islands.)

Several constitutional symptoms which are not primarily leprous often occur in the course of the disease. These are mainly gastro-intestinal disturbances. The kidneys are liable to amyloid degeneration, which is one of the common causes of death. Finally, in time the disease may become arrested and only its sequelæ remain, such as anæsthesia, paralyzed and atrophic muscles, and mutilated members, these patients attaining a good age.

From the foregoing description of the clinical course, with its subjective symptoms and objective lesions, it will be seen that typical cases in the two forms are very different; but there are many cases which present appearances that are puzzling even to the most experienced. In a certain percentage of cases the bacilli find favorable soil for development, either simultaneously or successively, both in the peripheral nerves and in the skin, in which case symptoms peculiar to both forms of leprosy are present. Occasionally, in the tuberous variety, the nodules disappear, and the case apparently becomes the milder or maculo-anæsthetic form. The reverse also is true, though much more rarely. As to deciding any case to be a "mixed" one, Hansen and Looft[2] say: "But since every case of nodular leprosy is accompanied by affection of the nerves and anæsthesia, and the natural termination of every case of nodular leprosy is to pass into the anæsthetic form, if only, as occasionally happens, the patient live long enough; and since the skin eruptions of the maculo-anæsthetic form are characterized, just as are those of the nodular form, by the presence of the leprosy bacillus, we regard the transformation of a case of maculo-anæsthetic into nodular leprosy only as a sign of the unity of the two forms, and we delete altogether the name of mixed leprosy; otherwise every case of nodular leprosy must, in all events, after some years of existence, properly be called mixed, for in such case anæsthesia is never absent." In the Berlin Conference, in 1897, Hansen[7] described a case which presented symptoms typical of both varieties. There were red tubercles in both eyebrows and on both cheeks. These were rather soft, and had the appearance of leprous tubercles, except that no hairs had fallen out of the brows, and some hairs were even situated on the tubercles. A cover-glass examination revealed no bacilli. In 1898 he excised a tubercle, made sections, and found the usual histological structure of lepra, but only a few bacilli. At this time anæsthetic spots appeared on the arms, and there was some anæsthesia in the hands. The firstma croscopic diagnosis was tubercular leprosy. Later, he considered it to be of the maculo-anæsthetic type. Some time later, he again saw the case, at which time the tubercle-like spots were transformed into true nodules, hard and devoid of hairs. Although he was now more inclined to consider it as belonging to the maculo-anæsthetic type on account of the spots and anæsthesia, yet the other appearances left the case doubtful in his mind.

History.—It is probable that leprosy has existed among the Jews from the earliest date of which we have record until the present time, and that they acquired the disease during their residence in Egypt. Kaposi believed that the leprosy of the Old Testament was merely vitiligo. Undoubtedly, many cases of vitiligo were then considered leprosy, but the cases in which ulceration (raw flesh) occurred evidently were not vitiligo. Job's affliction is considered by Thin and others to have been leprosy. Aristotle, 345 B.C., refers to a disease which he calls "satyria," and from his description Danielssen and Boeck think he must have referred to tubercular leprosy, and that his observations were made on the coast of Asia Minor. The best early clinical description of the disease was given by Aretæus in the first century of the Christian era. Celsus (53 B.C.–7 A.D.) states that leprosy was almost unknown in Italy; yet it seems probable that about this time it was introduced into Italy and spread from there into Northern and Western Europe. From the second to the seventh century it was prevalent in Europe. Galen, in the second century, writes of it in France, Germany, and Spain. In the eleventh and twelfth centuries it spread all over Europe, and in 1229 nineteen thousand leproseries existed, there being two thousand in France alone. It was unknown in America until after the arrival of the negroes from West Africa. In the fourteenth century it began to decline, and by the seventeenth it remained in only a few isolated localities. Early in the sixteenth century it had nearly disappeared from Italy, and somewhat later from France. At this time Denmark was also free. It remained in Scotland for some time after leaving England, the last cases occurring in Shetland, in the latter part of the eighteenth century. It persisted in Sweden until the end of the eighteenth century, while in Norway many cases still exist. During the last century it made its appearance and spread in new localities. About the middle of the century it was introduced into the Hawaiian Islands, where a serious epidemic has prevailed. Morrow[8] affirms that here, as in other newly infected districts, the disease was at first quickly fatal, and was usually of the tubercular variety; but of late years it has been less serious, and the milder or maculo-anæsthetic type is the more prevalent. In Louisiana, leprosy has been known since 1785, and has of late years increased, becoming endemic there about 1866.

ETIOLOGY.—It is now conceded practically by all observers that leprosy is a parasitic disease, and produced by the bacillus lepræ. Although attempts to reproduce the disease in the lower animals with this bacillus have thus far been unsuccessful, it can be positively affirmed that the bacillus is the active factor in the production of the disease. In ancient and mediæval times, and even up to a quite recent date, leprosy was considered contagious, but in 1867 the Royal College of Physicians declared against this theory. Hansen and Looft[2] quote statistics and give undeniable proof of the contagiousness of the disease in Norway. Thin[4] quotes quite a large number of cases in which leprosy was undoubtedly contracted by contagion. Among other examples, Morrow[8] quotes the case of Father Damien, a priest, whose personal and family history bore no taint of previous disease, but who, after close contact with lepers in the Hawaiian Islands, in a leper settlement, contracted the disease. Its development in new countries, or in new localities in the same country, can usually be traced and proven to have been produced by contagion. The contagiousness is not marked, and it requires the closest relationship between the healthy and the leprous before the disease can be contracted. It is not directly transmissible. Both ancient and modern writers until quite recently have considered it an hereditary disease; but the mere fact of its existence for a long period of time, perhaps for several generations, in the same family, would not demonstrate this to be the case, especially if one accepts the undoubted fact that it is contagious. The Lepra Commission in India, in 1893, reported that leprosy in India cannot be considered an hereditary disease, and stated that the evidence which exists is hardly sufficient to establish, to an appreciable degree, an hereditary predisposition to the disease in the offspring of leprous patients. Besnier[9] says that if any hereditary predisposition exists in lepra, it is less than that in tuberculosis. He suggests early and repeated bacteriological examination of the nose and pharynx, as these may be affected before the disease becomes obvious elsewhere. He explains the long incubation period by saying that the bacilli lie dormant, unable for the time being to germinate on account of bad soil. As to the mode of transmission from one person to another, different opinions prevail. Thin[4] says that close personal contact is usually necessary, but that the possibility of the infection being carried by an intermediate host, such as the mosquito or the acarus, is not impossible. Hansen and Looft[2] quote cases in which the infection was carried by way of clothing. Morrow[8] believes that in the majority of cases the contagion finds entrance to the body through the mucous membranes of the respiratory and gastro-intestinal tracts. As predisposing causes, bad hygiene, dirt, filth, and careless habits undoubtedly play an important rôle. Among the Norwegian peasants, where the disease prevails, Leloir, quoted by Thin, says that the greater number of these people have never bathed. Their clothing, generally made of wool, is never taken off, even for sleeping, and as it is never washed dirt naturally accumulates upon it; and, when not too worn, it is handed down from generation to generation. As a rule, several people sleep in the same bed; all eat at the same table, from the same dish, often from a common spoon, and drink from the same vessel. Hansen confirms the above, and states further that, under the civilizing influences of life in the United States, these habits are dropped, with the result that the disease no longer continues to be propagated, but dies with the death of those already infected. Neisser says that the number of lepers in any community bears an inverse ratio to the care taken to insure the isolation of infected persons. Leprosy may occur at any age, seldom before the fifth year, and most often between the thirtieth and fiftieth years. Damp cold climates, as well as moist hot ones, are most favorable for its development.

Geography.—The geographical distribution of leprosy is quite extensive, covering probably one-quarter of the globe. It occurs endemically in Northern and Eastern Africa, Egypt, Arabia, Persia, China, Japan, and India; Russia, Norway, Sweden, Italy, Greece, France, and Spain; the islands of the Pacific and Indian Oceans; it is prevalent in Central and South America, Mexico, the West Indies, Australia, the Hawaiian Islands, and New Zealand; and it is found in New Brunswick and other parts of Canada. It is also pretty well distributed in the United States, the most important centres being in Louisiana, California, and Minnesota. According to a report sent to the Senate on March 24th, 1902, and made by a commission of medical officers of the Marine Hospital service, leprosy is distributed as follows in the United States: Alabama, 1; California, 24; Florida, 24; Georgia, 1; Illinois, 5; Iowa, 1; Louisiana, 155; Maryland, 1; Massachusetts, 2; Minnesota, 20; Mississippi, 5; Missouri, 5; Montana, 1; Nevada, 1; New York, 7; North Dakota, 16; Oregon, 1; Pennsylvania, 1; South Dakota, 1; Texas, 3; and Wisconsin, 3; making a total of 278 cases.

PATHOLOGY.—Histologically, leprosy belongs to the group of granulomata, along with syphilis, tuberculosis, and certain other affections. It is a neoplasm, made up largely of partially reverted connective-tissue cells. In this new growth the specific bacillus is present in extraordinarily large numbers. This organism presents a