

Cousot has also met with a family in which the mother and four children were attacked. The disease occurs in youth, and the tendency to the attacks diminishes with age.

The clinical picture is very much alike in all the recorded cases. The paralysis involves, as a rule, the arms and legs. It comes on when the patients are in full health, and without any apparent cause, often during sleep. Sometimes it begins with weakness in the limbs, a sensation of weariness and sleepiness, not often with sensory symptoms. The paralysis is usually complete within the first twenty-four hours, beginning in the legs, to which in rare instances it is confined. The muscles of the neck are sometimes involved, and occasionally those of the tongue and pharynx. The cerebral nerves and the special senses are, as a rule, uninvolved. The attacks are afebrile, sometimes with low temperatures and slow pulse. The deep reflexes are reduced, sometimes abolished, and the skin reflexes may be feeble. One of the most remarkable features is the extraordinary reduction or complete abolition of the faradic excitability, both of muscles and of nerves.

Improvement begins sometimes in the course of a few hours or after a day or two, and the paralysis disappears completely, and the patient is perfectly well. As mentioned, the attacks may recur every few weeks, in some instances even daily; more commonly, an interval of one or two weeks elapses between the attacks. Goldflam suggests that the paralysis is due to an auto-intoxication, and that the poisonous material acts upon the nerve-endings in the muscles. He has made experiments with the urine of a case which showed that during the attacks the toxic properties of this secretion were materially increased. From the recurring, periodic character of the attacks they have been supposed to be due to malaria, but of this there is no evidence.

II. ASTASIA; ABASIA.

These terms, indicating respectively inability to stand and inability to walk, have been applied by Charcot and Blocq to diseased conditions characterized by loss of the power of standing or of walking with retention of muscular power, coördination, and sensation. Blocq's definition is as follows: "A morbid state in which the impossibility of standing erect and walking normally is in contrast with the integrity of sensation, of muscular strength, and of the coördination of the other movements of the lower extremities." The condition forms a symptom group, not a morbid entity, and is probably a functional neurosis. Knapp in a recent paper analyzes the 50 cases reported in the literature. Twenty-five of these were in men, 25 in women. In 21 cases hysteria was present; in 3, chorea; in 2, epilepsy; and in 4, intention psychoses. As a rule, the patients, though able to move the feet and legs perfectly when in bed, are either unable to walk properly or cannot stand at all. The disturbances have been very varied,

and different forms have been recognized. The commonest, according to Knapp's analysis of the recorded cases, is the paralytic, in which the legs give out as the patient attempts to walk and "bend under him as if made of cotton." "There is no rigidity, no spasm, no incoördination. In bed, sitting, or even while suspended, the muscular strength is found to be good." Other cases are associated with spasm or ataxia; thus there may be movements which stiffen the legs and give to the gait a somewhat spastic character. In other instances there are sudden flexions of the legs, or even of the arms, or a saltatory, spring-like spasm. In a majority of the cases it is a manifestation of a neurosis allied to hysteria.

The cases, as a rule, recover, particularly in young persons. Relapses are not uncommon. The rest treatment and static electricity should be employed.

V. VASO-MOTOR AND TROPHIC DISORDERS.

I. RAYNAUD'S DISEASE.

Definition.—A vascular disorder, probably dependent upon vaso-motor influences, characterized by three grades of intensity: (a) Local syncope, (b) local asphyxia, and (c) local or symmetrical gangrene.

Local Syncope.—This condition is seen most frequently in the extremities, producing the condition known as dead fingers or dead toes. It is analogous to that produced by great cold. The entire hand may be affected with the fingers; more commonly only one or more of the fingers. This feature of the disease rarely occurs alone, but is generally associated with local asphyxia. The common sequence is as follows: On exposure to slight cold or in consequence of some emotional disturbance the fingers become white and cold, or both fingers and toes are affected. The pallor may continue for an indefinite time, though usually not more than an hour or so; then gradually a reaction follows and the fingers get burning hot and red. This does not necessarily occur in all the fingers together; one finger may be as white as marble, while the adjacent ones are of a deep red or plum color.

Local Asphyxia.—Chilblains form the mildest grade of this condition. It usually follows the local syncope, but it may come on independently. The fingers and toes are oftenest affected, next in order the ears; more rarely portions of the skin on the arms and legs. During an attack the fingers alone, sometimes the hands, also swell and become intensely congested. In the most extreme grade the fingers are perfectly livid, and the capillary circulation is almost stagnant. The swelling causes stiffness and usually pain, not acute, but due to the tension and distention of the skin. Sometimes there is marked anæsthesia. Attacks of this sort

may recur for years, and be brought on by the slightest exposure to cold or in consequence of disturbances, either mental or, in some instances, gastric. Apart from this unpleasant symptom the general health may be very good. The attacks may recur only at long intervals or during the winter time.

Local or Symmetrical Gangrene.—The mildest grade of this condition follows the local asphyxia, in the chronic cases of which small necrotic areas are sometimes seen at the tips of the fingers. Sometimes the pads of the fingers and of the toes are quite cicatricial from repeated slight losses of this kind. So also when the ears are affected there may be superficial loss of substance at the edge. The severer cases, which terminate in extensive gangrene, are fortunately rare.

In an attack the local asphyxia persists in the fingers. The terminal phalanges, or perhaps only one finger, become black, cold, and insensible. The skin begins to necrose and superficial gangrenous blebs appear. Gradually a line of demarkation shows itself and a portion of one or more of the fingers sloughs away. The resulting loss of substance is much less than the appearance of the hand or foot would indicate, and a condition which looks as if the patient would lose all the fingers or half of a foot may result perhaps in only a slight superficial loss in the phalanges. In severer cases the greater portion of a finger or the tip of the nose may be lost. Occasionally the disease is not confined to the extremities, but affects symmetrical patches on the limbs or trunk, and may pass on to rapid gangrene. These severe types of cases occur particularly in young children, and death may result within three or four days. The attacks are usually very painful, and the motion of the part is much impaired. In some cases numbness and tingling persist for a long time.

There are remarkable concomitant symptoms in Raynaud's disease to which a good deal of attention has been paid of late years. Hæmoglobinuria may develop during an attack, or may take the place of an outbreak. In such instances the affection is usually brought on by cold weather. In a case reported by H. M. Thomas from my clinic, Raynaud's disease occurred for three successive winters and always in association with hæmoglobinuria. The attacks were sometimes preceded by a chill. Several cases of the kind are found in Barlow's appendix to his translation of Raynaud's paper for the New Sydenham Society. The onset with a chill, as in the case just mentioned, has doubtless given rise to the idea that the disease is in some way associated with ague. Cerebral symptoms, particularly mental torpor and transient loss of consciousness, have also been noticed in some cases. The case just mentioned with hæmoglobinuria had epilepsy with the attacks. Exposure on a cold day would bring on an epileptic seizure with the local asphyxia and bloody urine. Occasionally joint affections develop, particularly ankylosis and thickening of the phalangeal articulations. Southey has reported a case in which mania developed, and Barlow an instance in which the woman had delusions. Peripheral neuritis has been found in several cases.

The *pathology* of this remarkable disease is still obscure. Raynaud suggested that the local syncope was produced by contraction of the vessels, which seems likely. The asphyxia is dependent upon dilatation of the capillaries and small veins, probably with the persistence of some degree of spasm of the smaller arteries. There are two totally different forms of congestion, which may be shown in adjacent fingers; one may be swollen, of a vivid red color, extremely hot, the capillaries and all the vessels fully distended, and the anæmia produced by pressure may be instantaneously obliterated; the adjacent finger may be equally swollen, absolutely cyanotic, stone cold, and the anæmia produced by pressure takes a long time to disappear. In the latter case the arterioles are probably still in a condition of spasm.

Treatment.—In many cases the attacks recur for years uninfluenced by treatment. Mild attacks require no treatment. In the severer forms of local asphyxia, if in the feet, the patient should be kept in bed with the legs elevated. The toes should be wrapped in cotton-wool. The pain is often very intense and may require morphia. Carefully applied, systematic massage of the extremities is sometimes of benefit. Galvanism may be tried. Barlow advises immersing the affected limb in salt water and placing one electrode over the spine and the other in the water.

II. ANGIO-NEUROTIC OEDEMA.

Definition.—An affection characterized by the occurrence of local oedematous swellings, more or less limited in extent, and of transient duration. Severe colic is sometimes associated with the outbreak. There is a marked hereditary disposition in the disease. The affection has been specially studied by Quinke, Jamieson, J. E. Graham, and Matas.

Symptoms.—The oedema appears suddenly and is usually circumscribed. It may appear in the face; the eyelid is a common situation; or it may involve the lips or cheek. The backs of the hands, the legs, or the throat may be attacked. Usually the condition is transient, associated perhaps with slight gastro-intestinal distress, and the affection is of little moment. There may be a remarkable periodicity in the outbreak of the oedema. In Matas's case this periodicity was very striking; the attack came on every day at eleven or twelve o'clock. The disease may be hereditary through many generations. In the family whose history I reported, five generations had been affected, including twenty-two members. The swellings appear in various parts; only rarely are they constant in one locality. The hands, face, and genitalia are the parts most frequently affected. Itching, heat, redness, or, in some instances, urticaria may precede the outbreak. Sudden oedema of the larynx may prove fatal. Two members of the family just referred to died of this complication. In one member of this family, whom I saw repeatedly in attacks, the swell-

ings came on in different parts; for example, the under lip would be swollen to such a degree that the mouth could not be opened. The hands enlarge suddenly, so that the fingers cannot be bent. The attacks recur every three or four weeks. Accompanying them are usually gastrointestinal attacks, severe colic, pain, nausea, and sometimes vomiting. The colic is of great intensity and usually requires morphia. Arthritis apparently does not occur.

The disease has affinities with urticaria, the giant form of which is probably the same disease. There is a form of severe purpura, often with urticarial manifestations, which is also associated with marked gastrointestinal crises. Quinke regards the condition as a vaso-motor neurosis, under the influence of which the permeability of the vessels is suddenly increased.

The *treatment* is very unsatisfactory. In the cases associated with anæmia and general nervousness, tonics, particularly large doses of strychnia, do good; but too often the disease resists all treatment.

III. FACIAL HEMI-ATROPHY.

An affection characterized by progressive wasting of the bones and soft tissues of one side of the face. The atrophy begins, as a rule, in childhood, but in a few cases has not come on until middle age. It begins diffusely, but in some instances has started at one spot on the skin and has gradually spread, involving at first the subcutaneous tissues, then the muscles and the bones, more particularly the upper jaw. The wasting is sharply limited at the middle line, and the appearance of the patient is very remarkable, the face looking as if made up of two halves from different persons. There is usually change in the color of the skin and the hair falls. Owing to the wasting of the alveolar processes the teeth become loose and ultimately fall out. The wasting involves the tissues of the orbit, and the eye on the affected side is sunken. In a majority of the cases the atrophy has been confined to one side of the face, but there are instances on record in which the disease was bilateral, and a few cases in which there were areas of atrophy on the back and on the arm of the same side. The disease is rare. Sachs has collected 97 cases from the literature.

Two autopsies have been made. In Mendel's case there was the terminal stage of an interstitial neuritis in all the branches of the trigeminus, from its origin to the periphery, most marked in the superior maxillary branch.

In Homén's case, which came on rapidly and scarcely belongs to the typical form of the disease, a tumor was found pressing upon the Gasserian ganglion and the trigeminus nerve.

The disease is recognized at a glance. The facial asymmetry asso-

ciated with congenital wryneck must not be confounded with progressive facial hemi-atrophy. The precise nature of the disease is still doubtful.

IV. ACROMEGALIA.

Definition.—A dystrophy characterized by abnormal processes of growth, chiefly in the bones of the face and extremities.

The term was introduced by Marie, and signifies large extremities.

Etiology.—Nothing definite is known concerning the cause of the disease. It occurs rather more frequently in women. Of the 38 cases analyzed in the monograph of Souza-Leite, 16 were in men and 22 in women. The disease usually begins about the twenty-fifth year, though in some instances as late as the fortieth. Rheumatism, syphilis, and the specific fevers have preceded the development of the disease, but probably have no special connection with it. In this country five or six cases have been reported, two by J. E. Graham, of Toronto.

Symptoms.—In a well-marked case the disease presents most characteristic features. The hands and feet are greatly enlarged, but are not deformed, and can be used freely. The hypertrophy is general, involving all the tissues, and gives a curious spade-like character to the hands. The wrists may be enlarged, but the arms are rarely affected. The feet are involved like the hands and are uniformly enlarged. The big toe may be much larger in proportion. The nails are usually broad and large. The head increases in volume, but not as much in proportion as the face, which becomes much elongated and enlarged in consequence of the increase in the size of the superior and inferior maxillary bones. The latter in particular increases greatly in size, and often projects below the upper jaw. The alveolar processes are widened and the teeth separated. The soft parts also increase in size, and the nostrils are large and broad. The eyelids are sometimes greatly thickened, and the ears enormously hypertrophied. The tongue in some instances becomes greatly enlarged. Late in the disease the spine may be affected and the back bowed—kyphosis. The bones of the thorax may slowly and progressively enlarge. With this gradual increase in size the skin of the hands and face may appear normal. Sometimes it is slightly altered in color, coarse, or flabby, but it has not the dry, harsh appearance of the skin in myxœdema. The muscles are sometimes wasted. Changes in the thyroid have been found, but are not constant. The gland has been normal in some, atrophied in others, and in a third group of cases enlarged. Erb, who has made an elaborate study of the disease, has noticed an area of dulness over the manubrium sterni, which he thought possibly due to the persistence or enlargement of the thymus. Headache is not uncommon. Menstrual disturbance may occur early, and there may be suppression. In some instances vision has been involved, owing to a gradual atrophy of

the optic nerve. The disease may persist for fifteen, twenty, or more years.

The *pathological anatomy* has been studied in a few cases. In addition to enlargement of the bones, which is a true hypertrophy, enormous enlargement of the hypophysis (pituitary body) has been found, and some have regarded the disease as associated in some way with this. Less constant have been the changes in the thymus and in the thyroid. In some instances the peripheral nerves have been involved. The most exhaustive anatomical study made as yet is that published by Arnold, of Heidelberg, on the case which was described clinically by Friedreich and Erb.

As stated, the true nature of the disease is unknown. Marie regards it as a systemic dystrophy, analogous to myxœdema and possibly due to the morbid condition of the pituitary body, just as myxœdema is associated with disease of the thyroid.

Diagnosis.—The disease must be carefully separated from the *osteitis deformans* of Paget, in which the shafts of the long bones are chiefly involved, and in the head the bones of the cranium, but not those of the face. Marie states that in Paget's disease the face is triangular with the base upward; in acromegalia it is ovoid, or egg-shaped, with the large end downward; while in myxœdema it is round and full-moon shaped. The disease must not be confounded with the instances of congenital or progressive hypertrophy of a single member, as of the leg or arm, the so-called giant growth, in which the various proportions are maintained.

Lastly, Marie has separated from acromegalia a group of cases characterized by hypertrophy of the bones of the extremities and of the shafts, producing great disability. The spine is also affected and curvature takes place. The fingers are characteristic. The terminal phalanges become bulbous, enlarged, and the nails are curved, which gives the appearance of the so-called Hippocratic finger, a very different condition indeed from the flattened terminal phalanges of acromegalia. Etiologically, Marie regards this form as associated in some way with pulmonary troubles. Thus, for instance, two of the patients had purulent pleurisy, the cases of Ewald and of Saundby had new growths in the lungs, and others presented chronic bronchitis. Marie, therefore, terms this form *osteo-arthropathie pneumique*. It is doubtful, however, as Arnold states in his exhaustive study of Friedreich's case, whether this form can really be separated from acromegalia.

The treatment does not appear to have any influence upon the progress of the disease.

Here may be mentioned a remarkable dystrophy, met with so far only in women, known as *sclerodactyle*, in which there are symmetrical involvements of the fingers, which become deformed, shortened, and atrophied. The skin becomes thickened, of a waxy color, and is sometimes pigmented. Bullæ and ulcerations have been met with in some instances, and a great deformity of the nails. The disease has usually followed exposure, and the

patients are much worse during the winter and are curiously sensitive to cold. There may be changes in the skin of the feet, but the deformity similar to that which occurs in the hand has not been noted. Some of the cases have presented in addition diffuse sclerodermatous changes of the skin of other parts. An admirable description of the disease has been given by Gordinier.*

V. SCLERODERMA.

Definition.—A condition of localized or diffuse induration of the skin.

Two forms are recognized, the localized or circumscribed, which corresponds to the keloid of Addison and to morphea, and the diffuse, in which large areas are involved.

In the *circumscribed form* there are patches, ranging from a few centimetres in diameter to the size of the hand or larger, in which the skin has a waxy or dead-white appearance and to the touch is brawny, hard, and inelastic. Sometimes there is a preliminary hyperæmia of the skin, and subsequently there are changes in color, either areas of pigmentation or of complete atrophy of the pigment—leucoderma. The sensory changes are rarely marked. The secretion of sweat is diminished or entirely abolished. The disease is more common in women than in men, and is situated most frequently about the breasts and neck, sometimes in the course of the nerves. The patches may develop with great rapidity, and may persist for months or years; sometimes they disappear in a few weeks.

The *diffuse form*, though less common, is more serious. It develops first in the extremities or in the face, and the patient notices that the skin is unusually hard and firm, or that there is a sense of stiffness or tension in making accustomed movements. Gradually a diffuse, brawny induration develops and the skin becomes firm and hard, and so united to the subcutaneous tissues that it cannot be picked up or pinched. The skin may look natural, but more commonly is glossy, drier than normal, and unusually smooth. Of 44 cases, in 24 the first appearances were on the arms, in 7 on the legs, in 1 on both, in 10 on the face and neck, and in 2 on the trunk (Dinkler). The disease may gradually extend and involve the skin of an entire limb; in rare cases, it becomes universal, the face is expressionless, the lips cannot be moved, mastication is impossible, and it becomes extremely difficult to feed the patient. The hands become fixed, the fingers immobile, on account of the extreme induration of the skin over the joints. The disease is chronic, lasting for many months or many years. There are instances on record of its persistence for more than twenty years. Recovery may occur, or the disease may be arrested.

* American Journal of the Medical Sciences, January, 1889.

The patients are apt to succumb to pulmonary complaints or to nephritis. Rheumatic troubles have been noticed in some instances; in others, endocarditis. The pathology of the disease is unknown. It is usually regarded as a tropho-neurosis, probably dependent upon changes in the arteries of the skin leading to connective-tissue overgrowth.

The patients require to be warmly clad and to be guarded against exposure, as they are particularly sensitive to changes in the weather. Frictions with oil, and galvanism are recommended.

AINHUM.

Here a brief reference may be made to the remarkable trophic lesion described by Da Silva Lima, which is met with in negroes in Brazil, Africa, India, and occasionally in the Southern States. It is confined to the toes, usually the little toe, and begins as a furrow on the line of the digito-plantar fold. This gradually deepens, the end of the toe enlarges, and, usually without inflammation or pain, the toe falls off. The process may last some years. Cases have been reported in this country by Hornaday, Pittman, F. J. Shepherd, and Morrison.

SECTION IX.

DISEASES OF THE MUSCLES.

I. MYOSITIS.

Definition.—Inflammation of the voluntary muscles.

A primary myositis occurs as an acute or subacute affection, and is probably dependent on some unknown infectious agent. Several characteristic cases have been described of late years. The case of E. Wagner may be taken as a typical example. A tuberculous but well-built woman entered the hospital, complaining of stiffness in the shoulders and a slight oedema of the back of the hands and forearms. There was paræsthesia, the arms became swollen, the skin tense, and the muscles felt doughy. Gradually the thighs became affected. The disease lasted about three months. The post-mortem showed slight pulmonary tuberculosis; all the muscles except the glutei, the calf, and abdominal muscles were stiff and firm, but fragile, and there were serous infiltration, great proliferation of the interstitial tissue, and fatty degeneration. Similar cases have been reported by Unverricht, Hepp, and Jacoby of New York. In the case reported by Jacoby the muscles were firm, hard, and tender, and there was slight oedema of the skin. The duration of the cases is usually from one to three months, though there are instances in which it has been longer. The swelling and tenderness of the muscles, the oedema, and the pain naturally suggest trichinosis, and indeed Hepp speaks of it as a pseudo-trichinosis. The nature of the disease is unknown. Senator's case presented marked disorders of sensation, and there is a question whether the peripheral nerves are not involved with the muscles. Wagner suggests that some of these cases were examples of acute progressive muscular atrophy. The separation from trichinosis can be made only by removing a portion of the muscle. There are septic cases in which a diffuse, purulent infiltration of the muscles of different regions occurs. Instances have been reported in which this has been described as the primary affection, the condition of the muscles even passing on to gangrene.

A remarkable affection is *myositis ossificans progressiva*, in which portions of the muscles undergo a progressive calcification.