

remedy for the treatment of various forms of skin disease. He described it as the remedy *par excellence* for acne. He advised fifteen drops twice a day internally, and a ten-per-cent. lotion to be painted on the affected parts at night; each day the lesion was to be bathed with hot water and ichthyol soap. In eczema and other forms of skin disease he advised it to be used more cautiously and in weaker solutions. In all these conditions its internal administration is also advised, as it is supposed to possess valuable alterative properties, which assist its local action. Ichthyol has come into very extensive use for all forms of skin diseases, and for the removal of the product of inflammatory processes, in chronic rheumatism and in cases of enlarged glands. In skin diseases it is applied as an ointment of the strength of five to ten or fifteen per cent. A varnish of ichthyol is a favorite method of using the drug. It is made of ichthyol 10 parts, starch 40 parts, solution of albumin 1 part, and water to 100 parts. The starch is mixed with the water, the ichthyol is then well rubbed up with it, and lastly the albumin is added. For inflammatory deposits stronger preparations are employed; ointments containing fifty per cent. may be used, or a mixture of ichthyol and glycerin is applied and the part is covered with lint and bandaged. Ichthyol plasters and ichthyol wadding may also be obtained.

The remedy has not been much used in general surgery, but in gynecology it is a favorite application in all uterine and vaginal inflammations. It may be applied in a glycerin solution, or in its pure state on tampons, or medicated pessaries may be introduced after antiseptic and astringent douches have been used. When gonorrhoea is a factor the drug is thought to be especially beneficial. It has also been employed in inflamed states of the prostate gland, a ten-per-cent. solution being injected into the rectum.

In a paper presented at the Fourth International Medical Congress, Unna dwelt upon the value of ichthyol in the treatment of leprosy. His conclusions were: (1) *Lepra tuberosa*, even of universal extension and of more than one year's standing, can be cured; (2) the cure can be effected in a comparatively short time by the energetic internal and external employment of reducing remedies; (3) among these may be specially recommended ichthyol, chrysarobin, pyrogallol, and resorcin; the only one of which that can be administered internally, for a length of time, without danger, is ichthyol. He advised the application of a fifty-per-cent. ointment, and also the internal administration of fifteen to twenty-four minims (1.0 to 1.6) during the day.

Internally ichthyol is administered in pill form in capsules, the dose being up to twenty minims (1.3) daily for an adult. The dose for a child of two years is given as two minims (0.13) daily. *Beaumont Small.*

ICHTHYOSIS.—**DEFINITION.**—Ichthyosis is a congenital cutaneous deformity in which the skin is dry and rough and the epidermis thickened and scaly.

ETIOLOGY.—Almost all observers agree in regarding ichthyosis as a congenital disease, and the arguments in favor of this view are: That the disease commences early in life and endures throughout life, is only temporarily benefited by treatment, and immediately recurs on the cessation of treatment; that the disease is not inflammatory, as infective and toxic diseases are, but acts more like an anatomical deformity; that it is often a family disease, two or more brothers or sisters being frequently similarly affected, and even when the immediate family is free, more distant relatives may be found afflicted.

It is hardly necessary to draw attention to how characteristic this skipping over individuals is in hereditary affections, one or two members of one branch of a family and one or two of some other branch being picked out, the rest escaping. Nevertheless it must be admitted as remarkable how frequently cases of severe ichthyosis are reported in which the parents are said to be perfectly healthy, and this has been noted even when several of the children have ichthyotic skins.

An objection has been raised to calling ichthyosis a congenital disease, because it usually does not appear until the second or third year of life or even later. This, however, is not a valid objection, because ichthyosis agrees exactly in this particular with other congenital deformities. *Nævi*, for example, although certainly congenital, frequently do not appear until a considerable



Fig. 2786.—Ichthyosis. (From A. Neisser's "Stereoscopischer medizinischer Atlas.")

time after birth; not, indeed, until the gradual development of the affected individual or the growth of the lesion itself brings the blemish to view.

Undoubtedly ichthyosis depends in a great measure on defective action of the sweat and sebaceous glands. Three of its features point decidedly in this direction, viz.: the dryness and lack of unctuousness of the skin; the improvement of the symptoms during the summer months when the sweat glands are most active; and the tendency to the localization of the disease in those parts where the sweat and sebaceous glands are least active, as on the extensor surfaces of the limbs, leaving the articular folds, such as the bend of the elbows, the axillary, inguinal, and popliteal folds, unaffected.

Although ichthyosis is an hereditary and congenital affection, yet it probably may be accentuated by diseases which are not in their true sense hereditary. Syphilis is not in the true sense of the term an hereditary disease. What we call hereditary syphilis is an "infective disease" caught congenitally. The way that syphilis, for instance, may possibly accentuate ichthyosis is shown by two cases reported by Gaston and Emery. The father of these children had a slight ichthyosis and also had syphilis. The children acquired syphilis congenitally, and became after birth markedly ichthyotic. Besides ichthyosis, however, the children had microsphygmia, and the authors entertained the opinion that the small pulse was owing to a generalized, congenital, syphilitic arteritis, and that this affection of the arteries so interfered

with the nutrition of the fetus as to accentuate the hereditary tendency to ichthyosis.

Recently Edward Fournier has collected from the literature nine cases, and has added one of his own, in which patients having hereditary syphilis also had ichthyosis. The author clearly states, however, that he does not believe ichthyosis to be a symptom of hereditary syphilis, but explains the coincidence by the theory that hereditary syphilis is at times one of the causes of degeneration in the individual, and that ichthyosis is a symptom of degeneration.

Interesting, particularly in its connection with treatment, is the relationship between defective development of the thyroid gland and ichthyosis. As one of the symptoms of cachexia strumipriva is a rough dry skin; a mistake in diagnosis might easily occur, or defective action of the thyroid gland might accentuate an already existing ichthyosis.

SYMPTOMS.—As ichthyosis is a congenital deformity there are necessarily all grades of it, from almost unappreciable manifestations up to disgusting monstrosities. Dryness of the skin and desquamation are, however, never absent in ichthyosis, and in fact in the most attenuated form of the disease, in xeroderma, extreme dryness of the skin, a very fine desquamation, and obstruction with epithelial cells of the pilo-sebaceous orifices are the only symptoms present. It is this last symptom, the accumulation of epithelial cells about the points of emergence of the hairs, that gives those having ichthyosis their "goose-flesh" appearance. This same permanent "goose-flesh" appearance constitutes the disease called *keratosis pilaris*, and it is because the same appearance is produced by the same kind of plugging of the same orifices, and also because the favorite situation of both *keratosis pilaris* and ichthyosis is on the extensor surfaces of the upper arms and thighs, that some authors regard *keratosis pilaris*, when it exists alone, as constituting the very lightest form of ichthyosis, even lighter than xeroderma.

Of the two symptoms, dryness and desquamation, Thibierge considers dryness the more essential, even thinking dryness without desquamation often indicative of ichthyosis; therefore he would classify congenital permanent anidrosis as a form of ichthyosis, and this for the following reasons: In both diseases the skin is unusually dry, both are congenital, both occur in families as hereditary diseases, and some members of the same family may have anidrosis while others have ichthyosis. He thinks that many cases of ichthyosis which are supposed to develop during adolescence or later, are simply cases in which anidrosis was at first the sole symptom and which remained comparatively unnoticed, the more disagreeable symptom of scalliness supervening later.

Apart from the two general characteristics of dryness and a tendency to adhere and form masses, the superficial epithelium in ichthyosis shows many variations. It is sometimes powdery, and the cheeks for example may look as if dusted with white face powder; this constitutes a variety of the *pityriasis simplex* of some authors. This form of ichthyosis when more marked shows a diffuse granular coating, chalky and white, which is particularly apt to show itself on the outer surfaces of the upper and lower extremities. Instead of the granular powdery condition there may be scaling, and the scales may be clear gray or grayish-black, thick or thin, lustreless, hard or soft, but as above stated they frequently show a tendency to adhere with their flat surfaces tightly to one another, sometimes forming thick, uniform, lamellated, corneous deposits. When the epidermis accumulates in thick horny masses as it does in ichthyosis hystrix, pigment may develop in it, which may be a dirty green, a bright green, dark green, or greenish-black. The green may be so bright as to be startling, even seeming to have been artificially applied. The edges of the scales may be separated from one another by little furrows, or they may overlap like fish scales. Sometimes the surface of the skin looks like the finely crumpled tissue paper called *crêpe paper* used for

lamp shades, where the ridges run in every direction, crossing one another and dividing the surface into a vast number of quadrilateral areas. The wrinkled epidermis in such cases looks as if too large for the cutis. Sometimes instead of there being ridges there are fine shallow cracks, passing through the superficial layers only. These cracks fill with dirt and, just as under the finger nails, this dirt assumes a dirty greenish-gray color, marking the skin off into lozenge-shaped areas by a dirty greenish-gray network. The lines may crack down deeper than indicated above, and the superficial epidermis may be broken and upcurved along the edges of the enclosed lozenge-shaped areas resembling the bark of some trees. The surfaces of these lozenge-shaped areas may glitter and reflect light like the faceted skin of some reptiles. Sometimes the surface looks unnaturally smooth and polished, with lines running across it at all angles, as if the surface had been varnished or had colloid poured over it, and had cracked in the drying.

Frequently the skin does not crack, but has running across it a vast number of criss-cross furrows dividing the surface in the same way into quadrilateral or irregular areas. These furrows are really the natural markings of the skin exaggerated, and are frequently chalky white and powdery; in such skins if the finger nail be drawn sharply across the surface it will leave a chalky white, crumbly, permanent mark.

Sometimes the papillary layer of the skin is hypertrophied, and when this occurs, and corneous masses or spines are formed over them, it gives rise to one of the varieties of ichthyosis hystrix. Another variety of such horny spines is formed by a great accumulation of the epithelial cells projecting from the pilo-sebaceous orifices, constituting the *keratosis pilaris* above named; this occurs generally on the extensor surfaces of the arms. Ichthyosis hystrix is never universal, although it may occur over large tracts of the skin. Usually it occupies only certain regions, while the intervening skin is xerodermic. A child at present under treatment in the University Clinic has ichthyosis hystrix only on the ankles, while other regions show several different varieties of ichthyosis. This condition of ichthyosis hystrix, literally "hedgehog, fishy disease," is considered particularly undesirable in a bed-fellow.

The true skin may be atrophied, tightly stretched, and thin, and the subcutaneous fat layer may be thin and poorly developed.

The face is frequently unaffected except by a transitory powderiness, a *pityriasis*. The palms and soles are dry and powdery but not scaly, with the furrows more marked and more numerous than normal, and this condition may remain after treatment has improved the skin in every other locality.

The hair is frequently normal, of good color, lustre, quality, and quantity, but it may be dry, frail, and thinly scattered, constituting an expression of the same fault of development that gave rise to the ichthyosis. Sometimes there is nearly complete alopecia, even the eyebrows and eyelashes being almost entirely absent (Thibierge). The nails may be strong and in every way normal. They may, however, be thin, brittle, and either transversely or longitudinally ridged, or there may be an accumulation of dry, brittle, corneous substance under the nails.

These patients sometimes give off a flat, stale, penetrating, disagreeable odor, such as is met with in other cases of generalized hyperkeratosis with maceration of the epidermis, as in Darier's disease for example (Thibierge).

Usually the only subjective symptoms of ichthyosis are itching and a sensitiveness to cold; chilly feelings being experienced on the slightest lowering of the atmospheric temperature.

A patient usually presents examples of a number of the conditions enumerated above; for instance, the hair may be good, but the scalp is usually in a condition that cannot be distinguished from dry seborrhoea. The outer surfaces of the arms and thighs and the skin of the trunk

may be dry and scaly and criss-crossed with furrows, and studded with dry papules that look like "goose flesh." The buttocks and upper portions of the thighs may be covered with "crêpe-paper" skin. The roughness of the skin increases down the extensor surfaces of the thighs until over the front of the knees one finds immense cornuous masses. Below the knees the skin may be tightly stretched over the tibiae, smooth, shiny, and with markings on its surface dividing it off into small quadrilateral areas.

COURSE AND DURATION.—Usually ichthyosis does not become appreciable until the second or third year of life, although cases have been reported in which the malady was apparent at birth or shortly after. The first symptom, according to Peukert, is the appearance of keratosis pilaris that shows itself on the extensor surfaces of the extremities in earliest childhood in these individuals. It is simply this early appearance of the keratosis pilaris and its greater development later in life that distinguish it at all from the ordinary form of keratosis pilaris. Ordinary keratosis pilaris of moderate development is so common later in life, especially at adolescence when the lanugo hairs begin to sprout more vigorously, that it is practically a normal condition. Ichthyosis continues to grow more marked from two or three years of age until adult life, when it may remain stationary or subject only to the mutations of the seasons, becoming severe in winter and milder in summer, till the natural dryness and harshness of the skin of old age again accentuate it. In summer, in fact, the plates, scales, and fluffy epidermis may all fall off as a "moulting," leaving the skin almost normal. Sickness and a lowered condition of health may also increase the dryness and harshness of the skin, and there is reason to believe that inactivity or absence of the thyroid gland can contribute decidedly to the severity of the symptoms. The symptoms of ichthyosis may disappear completely after one of the eruptive fevers, measles for instance (Joseph), only to reappear in a short time, however. This same amelioration may also occur during pregnancy.

Quinquaud has reported a case in which the patient got an ichthyotic skin following a generalized eczema of two years' duration. He says that such acquired ichthyotic states may after a time disappear. It is doubtful, however, if such ichthyotic states as described by Quinquaud should be included in the disease ichthyosis at all.

Unna and Tommasoli believe that ichthyosis is an acquired disease that has its time of onset, development, and disappearance, but this view finds very little support among other observers.

UNUSUAL SYMPTOMS AND COURSE.—The flexures of the articulations, particularly the axillae, the bends of the elbows, the groins, the scrotal thigh folds, and the poplitea are usually unaffected. In fact, Georges Thibierge would make the freedom of those localities a sharp diagnostic feature. Recently, however, a number of cases have been reported in which one or several of these localities have been affected, and there happen just now to be in the University Clinic two patients in whom the axillae, the groins, the scrotal thigh and vulvar thigh folds respectively, and the poplitea are affected, as shown by the presence of pityriasis. In both patients there is an eczema of the upper extremities that involves the bends of the elbows, making it impossible to say whether these are ichthyotic or not. The appearance and stubborn character of the eczema, however, would lead one to infer that the condition is also present there. Joseph has reported a case in which only the flexures were affected. Usually ichthyotic patients do not sweat, but Peukert thinks the dryness of the skin is in some cases not due to lack of perspiration, but to an increase of evaporation from the more extensive cutaneous surface of these people. In some instances there is vicarious sweating of certain regions as of the face for example (Goldzieher), or of other regions not implicated in the ichthyosis, as the axillae, or the palms and soles. This sweating may be abundant. The mucous membranes are rarely affected in ichthyosis, unless the susceptibility to catching "colds" and to bron-

chitis be looked upon as the particular way they show their inherent weakness. There are atrophic forms of the disease, which Hallopeau considers simply an exaggeration of the form in which the skin is like crêpe paper. The skin is very thin and has little elasticity, so that when pinched it retains the impress of the fingers for a long time. The skin may appear too large for the patient, or on the contrary it may be too small, not permitting the closure of the eyelids, exaggerating the cutaneous folds, especially in the movements of the facial muscles, and impeding the flexion and extension of the fingers. In two of the cases so reported the skin seemed glued to the subjacent aponeurosis. Kaposi has shown a case which he called ichthyosis hystrix pterygoidea s. plumiformes, in which there were hundreds of little growths from 0.5 to 1.5 cm. long that looked like feathers. These growths consisted of a central core of connective tissue with tufts of cornified epithelial tissue depending from them. The patient was a girl six years of age and the palms and soles were thickened, cracked, and warty. This case stands alone in literature, and it is not at all certain that it should be included among the ichthyoses.

Those having ichthyosis and those that are scrofulous have in common a tendency to inflammation of the respiratory tract, and besides that, Unna has noted with great particularity in one of his cases an unusual growth of hair down the cheeks in front of the ears. An unusual growth of fine downy hair in this situation was considered by the late Professor Horner, of Zurich, a symptom of scrofula.* A little girl suffering from ichthyosis, and at present attending the University Clinic, has quite a little beard of fine long hair growing down in front of each ear.

DIAGNOSIS.—The first disease to be sharply differentiated from ichthyosis is *naevus linearis*, and this is not because there is any great similarity between the diseases, but because some writers persist in describing *naevus linearis* under the name ichthyosis hystrix. *Naevus linearis* is a warty congenital disease, in which the warts are frequently covered with horny masses, but here the similarity ceases. *Naevus linearis* occurs in lines or in localized areas, and is usually asymmetrical, and the skin between the patches or lines is absolutely normal. In ichthyosis the disease is symmetrical, and almost always the whole skin is affected, with the exception of the folds of the articulations. Another good point of differentiation lies in the fact that *naevi*, *naevus linearis* included, have generally a buff or a slate-colored pigmentation, and the pigment lies in the connective tissue of the skin, usually in the affected papillae, while if there is a black or other color in ichthyosis, it looks more like dirt in the horny layers or pigment in the epithelium, rather than like pigment in the papillae or in the true skin. In ichthyosis the palms and soles are almost always unnaturally dry with a powderiness of the surface, while in *naevus linearis* the palms and soles are normal, when not directly the seat of the *naevus*, which they rarely are. Furthermore *naevus linearis* does not attack several members of a family, but occurs irregularly here and there throughout a community; in other words it is not a family disease.

There is another disease, however, which is hereditary, which occurs in families, and which consists of an exaggerated development of the horny layer of the skin. This disease, *keratoma hereditarium palmare et plantare*, it seems desirable to keep separate from ichthyosis. This is all the more difficult to do because, besides the above points of similarity with ichthyosis, there is also in ichthyosis thickening of the horny layer of the palms and soles. In *keratoma*, however, the thickening is limited sharply to the palms and soles, and there may be increased action of the perspiratory glands of the affected surfaces. It therefore lacks the generalization of ichthyosis and also the dryness, two important features. Furthermore, in *keratoma* the thickening of the horny layer of the palms and soles is frequently much greater

* He used frequently to say "they, the scrofulous, all wear a beard."

EXPLANATION OF
PLATE XXXIII.

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DR. DOUGLASS W. MONTGOMERY'S CASE OF ICHTHYOSIS HYSTRIX.

The faint redness, the swelling, and also the violet hue in the neighboring skin seen in the plate were all due to an accompanying dry eczema.

The patches of thick green crusting were caused by epithelial masses covering the papillomatous skin, and constituted the ichthyosis hystrix. It was only on the ankles that the child had the papillomatous or hystrix variety of ichthyosis. In other situations, as on the upper parts of the legs, on the thighs, trunk, upper extremities, face and scalp, the ichthyosis was of other types.



ICHTHYOSIS HYSTRIX.