

Case of thickening and deep fissures of the skin in an infant at birth / by Alexander Keiller. Notes of cases of intra-uterine cutaneous disease analogous to that described in the preceding article, with remarks on the pathological nature of the affection / by J.Y. Simpson.

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Keiller, Alexander, 1811-1892.

Simpson, James Young, 1811-1870. Notes of cases of intra-uterine cutaneous disease.

Publication/Creation

[Place of publication not identified] : [publisher not identified], [1843]

Persistent URL

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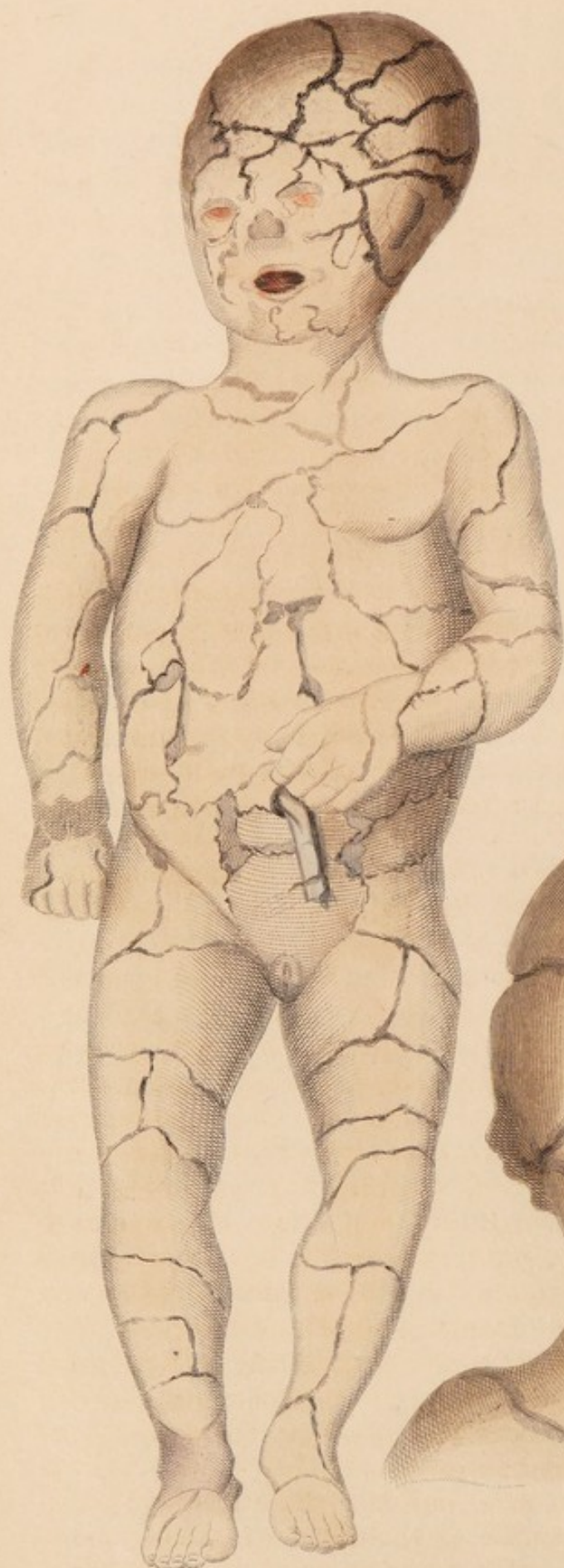
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D.^R KEILLERS CASE.
OF
INTRA-CUTANEOUS DISEASE



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CASE OF THICKENING, AND DEEP FISSURES OF
THE SKIN, IN AN INFANT AT BIRTH.

BY ALEXANDER KEILLER, M.D.

(COMMUNICATED BY PROFESSOR SIMPSON.)

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(Extracted from the Lond. and Edin. Monthly Journal of Medical Science for August 1843.)  
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32 UNION STREET, DUNDEE.

SIR,—From having observed in a recent number of *THE MONTHLY JOURNAL*, (p. 84,) a brief report of the case of “Monstrosity caused by Thickening of the Skin,” which was lately read before the French Academy of Medicine by M. Souty, I have been induced to submit to you the following particulars of an equally singular, and somewhat analogous case, which a short time ago occurred in my obstetric practice.

If deemed sufficiently interesting to merit professional publicity, a place in your widely circulated *Journal* will much oblige, Sir, your obedient servant,

ALEX. KEILLER, M.D.

To DR CORMACK.

On the 25th January 1841, Mrs H., then well advanced in her fourth pregnancy, was suddenly seized, while shopping, with an acute pain in the left side of the abdomen. With considerable difficulty she reached home, when my immediate attendance was requested. I found her in bed, complaining as if in the first stage of parturition. On interrogating her, however, respecting the period of her pregnancy, and the recent condition of her general health, she stated that she did not consider herself much advanced beyond the seventh month, and that during the four months preceding her present indisposition, she had suffered very severely from acute paroxysms of pain in the abdomen, the left iliac region of which she more particularly described as the peculiar seat of her long-continued and present suffering.

From her feelings having been so painfully different from those experienced during the corresponding period of her former pregnancies, she repeatedly expressed her conviction that “something very unusual was the matter,” and doubted not but that the

many unpleasant forebodings which she had for months entertained regarding the condition of her unborn offspring would yet be sufficiently accounted for. Knowing my patient to be of an extremely nervous temperament, and finding the os uteri undilated, I was induced to consider the case as one of simple puerperal neuralgia, and accordingly prescribed an anodyne draught with castor oil, which, with rest and quiet, appeared quickly to subdue the uterine irritation, and with it the previously almost intolerable abdominal pain.

On the evening of the 28th, however, the regular pains of labour commenced, and continued slowly to progress until nine A.M. of the 29th, when the birth of a male child, the singularly-conditioned subject of the accompanying sketch, was effected.

During the first stage of parturition, and previous to the discharge of the liquor amnii, the hard and globular presentation sufficiently indicated a head; on rupturing the protruding membranes, however, and bringing my fingers into immediate contact with the presenting structure, in order to ascertain its relative position, I was not a little astonished to find that, instead of the smooth, soft, yielding scalp, a rough, hard, and irregularly fissured surface presented itself, the usual guides to position—the fontanelles, sutures, and ears—being altogether undistinguishable. Although at a loss to determine what the advancing structure consisted of, its unyielding serrated-like feel led me to anticipate the existence of an anencephalous development, or some other abnormal condition of the bones of the skull. On delivery being accomplished, however, a sufficient excuse for any previous uncertainty immediately presented itself in the *hard, preternaturally-thickened and deeply rent dermoid covering, not only of the head, but of the whole body*, which seemed as if completely enveloped in an irregular coating of cartilaginous structure. So deep were the numerous fissures in the hard and much thickened scalp, that in appearance, as well as to the touch, they very much resembled the sutures, or rather the serrated edges of the bones, of a disarticulated adult skull.

The skin of the face, in like manner, consisted of ossified-looking portions, also irregularly fissured; the nose and ears were represented by mere tubercular knobs without apertures; the eyelids were altogether undeveloped; red fleshy-like cushions occupied the *eyeless orbits*; the mouth seemed large and gaping; the tongue was voluminous and protruding, and their ill-starred owner, as if to render his hideousness still more complete, was yelling most vociferously.

The skin of the trunk and extremities was equally hard and cartilaginous, though not so thick as that of the head and face; the rents, or cracks, were so extensive on the abdomen, that it appeared, in several parts, altogether void of dermoid covering.

The carpus and tarsus of the right extremities were apparently somewhat diseased.

The genitals, like the nose and ears, seemed a mere tubercular knob.

The child was otherwise well formed, and, judging from its size and apparent strength, more especially of its vocal organs, appeared perfectly "*viable*."

It only, however, survived twelve hours.

The placenta was small, soft, and easily torn.

The umbilical cord was unusually thin.

The mother made a perfect, though slow recovery, and the agitation, consequent on what she was pleased to consider as the melancholy fulfilment of her long-entertained surmises, having gradually subsided, she soon again became pregnant, an event which seemed greatly to aggravate her previous mental despondency, and to produce other nervous symptoms, for which I was frequently called upon to prescribe.

Despite, however, the continuance and fullest operation of that, by some, foetal-staining, monster-making influence,—*a brooding, evil-anticipating, maternal imagination*,—she was in due time (on the 30th March 1842) safely delivered of a full grown healthy child, and has since enjoyed excellent health.

So far as I could judge or ascertain, both parents were altogether free from any constitutional taint; their second child, (which was a breech case, and premature,) was, however, still-born. The surviving three are healthy thriving children.

The smaller of the two accompanying sketches exhibits the appearance presented by the hard and deeply fissured scalp over the presenting part of the head. The larger shows the appearance of the face, and anterior surface of the child.

NOTES OF CASES OF INTRA-UTERINE CUTANEOUS DISEASE,
ANALOGOUS TO THAT DESCRIBED IN THE PRECEDING ARTICLE,

WITH

REMARKS ON THE PATHOLOGICAL NATURE OF THE AFFECTION.

By J. Y. SIMPSON, M.D.,

PROFESSOR OF MIDWIFERY IN THE UNIVERSITY OF EDINBURGH.

The form of cutaneous congenital disease, that has been so well and accurately described by Dr Keiller, in the preceding notice, seems to be of very rare occurrence. At Dr Keiller's suggestion, I add some short notes of a few similar cases with which I am acquainted:—

1. I saw three years ago, a foetus presenting the same disease in the Museum of Guy's Hospital, London. The only note of it to which I can refer is a pencil jotting made at the time to the following effect: "foetus born with skin-bound disease, and integuments fissured in all directions."

2. In 1792, Richter gave a very imperfect description of an instance of the disease in the appendix to an essay, entitled *Dissertatio de Infanticidio in Artis Obstetriciæ Exercitio non semper Evitabili*.

3. Another case, much more fully detailed, was published in 1802 by Hinze in his *Kleinere Schriften*.

4. An infant, showing a well marked instance of this form of disease, is preserved in the Berlin Museum. It was described in 1826 by Steinhausen in a thesis, entitled, *Dissertatio de Singulari Epidermidis Deformatate*.

Instead of giving a separate account of each of these cases individually, it may be more instructive, and will avoid repetition, if we merely state some of the principal peculiarities connected with their pathological history.

In all the cases the skin showed the same diseased appearance, with numerous fissures extending over it in every direction, and leaving between them irregular shaped compartments of thickened integumentary substance. In Steinhausen's case, the fissures were fewest on the back and extremities, and more numerous on the head, fore part of the trunk, and about the pudenda and anus. About the neck, the intervening compartments or scales were few in number, having, apparently, being rubbed off by the motions of the parts. In Hinze's case the fissures were longest and deepest at the wrists, and over the malleoli, where they extended down even to the bone, which was seen lying bare on raising the edges of the rents. Hinze did not see the child till two days after its birth, at which time the fissures extended over the whole body. He was told, however, that at birth they existed only over the face, head, and neck. Those on the head had, when he saw it, become dry and horny.

In Richter's case, the remarkable hardness of the skin is not particularly mentioned; it is simply stated to have been of a reddish yellow colour. In Hinze and Steinhausen's cases it is described as having the same horny cartilaginous feel, as in Dr Keiller's patient. Steinhausen states that the whole body appeared as if covered with scales, over-lapping each other, and varying in size in different situations. These scales, he adds, were composed of two layers, which could be easily separated from each other. The external layer was the hardest and thickest, and appeared in some parts to be divisible into several lamellæ. On removing this layer, the exposed surface showed numerous depressions and elevations, and was beset with innumerable pro-

jections like small hairs. Beneath both the two layers the skin is stated to have shown no abnormal appearance. In Hinze's case, on some parts, the segments of the skin had a smooth surface; in other situations the surface was rough, and here and there had a tuberculated appearance.

In Richter's case, as in Dr Keiller's, the eyes are said to have been wanting, and in their place fleshy masses, very tender to the touch, were seen projecting from the orbits. In Steinhausen's the eyelids presented the same appearance as the rest of the skin; the eyes themselves were, probably, sound, as the contrary is not stated. In Hinze's case there was ectropion of the upper eyelids, so that the eyes, which were healthy, appeared as if covered with a bloody skin. This would probably present precisely the same appearance as is described in Richter's case, and naturally leads to the question whether the true state of the organ may not have been overlooked by him, especially as in all other situations the disease seems to have been confined almost entirely to the cutaneous texture.

In all, the ears were almost or altogether wanting; in Richter's case, there were small foramina in their place, and in Steinhausen's merely small tubercles without any opening. The nose in Richter's case was also absent; the opening of the nostrils was present. In Steinhausen's the nose was very small, and the lips were much disfigured, though internally they had their natural appearance. Richter mentions that the tongue in his case was unusually large and cleft, and that the anus was wanting. In Steinhausen's this last mentioned opening was very small.

The extremities in Hinze's case were much deformed and swollen, and the fingers and toes, with the fore part of the foot, are described to have been so disfigured as to resemble bird's claws. Otherwise, the child appears to have been well formed. In the case described by Steinhausen the hands were much swollen, and had each five fingers; the same was the case with the feet. In Richter's case both hands and feet were malformed; the extremities terminating in rounded knobs. Those of the upper extremities had warty projections instead of fingers. The head was also malformed, and, like all the others, was very scantily provided with hairs.

In Richter's case the infant lived for three days, though it could take no nourishment without being threatened with suffocation. The child described by Hinze lived four days. During the first twenty-four hours it was remarkably lively, and took eagerly the milk and water on which it was fed, the mother not being allowed to see it. The meconium was of the natural colour. The urine is said to have been clear and inodorous. In Steinhausen's case the child lived three days and a half. In one case (Richter's) it was born about the eighth month; in the others not till the full period.

In one of these cases (Hinze's) the mother, who was 34 years of age, had enjoyed excellent health during her pregnancy, and had previously given birth to three healthy children. In another (Steinhausen's) both parents are said to have been healthy, as in Dr Keiller's case.

Nature of the Disease.—The pathological nature of the disease before us is, under our present deficiencies of accurate anatomical details, a difficult and unsatisfactory problem. One thing, however, seems manifest, viz. that the cutaneous fissures are in all probability merely a secondary and mechanical result. The integumentary parts having by previous disease lost their expansibility, and no longer increasing with the increasing dimensions of the growing body of the foetus, at last split and fissure under the stretching from within to which they are subjected. It is a much more difficult matter to determine the character of the morbid action which has produced the physical in expansibility of the integumentary coverings of the body. Hinze considered the disease to be of the nature of the lepra of the Greeks, or elephantiasis leprodes, and supposed that it depended on a syphilitic taint on the part of the parents, though of

this he seems to have had no proof whatever, and the healthy state of the parents and of their children directly contradicts any such idea. It would appear to be much more analogous to Ichthyosis than to any other skin disease that can be referred to. We have no evidence of the disease affecting, primarily at least, more than the epidermic layer, or rather the surface of the true skin which secretes it. Steinhausen expressly states, that in the case he has described, the skin under the two layers of thickened epidermis appeared to be quite healthy. Hinze's case is particularly instructive, as showing the disease in its progress, before it had reached the complete form in which it is seen in the others. The whole body appears to have first become incrustated with the inelastic morbid epidermic covering. As the growth of the fœtus proceeded, this covering was split up in various directions, and the process of fissuring was probably hastened after birth, by the exposure of the surface to the air, causing it to harden and to contract. The fissures extending to the surface of the true skin, reparative or ulcerative inflammation necessarily followed, and from the great extent of surface affected, the irritation and exhaustion thus induced, speedily proved fatal to the infant. Had it been able to survive this irritation and exhaustion for some time, till the other fissures, like those on the head, were become dry and horny, it would have afforded a condition presenting a perfect specimen of ichthyosis in its most extreme form, and affecting the whole surface of the body.

If the disease be thus considered as a species of intra-uterine ichthyosis, we may further find some explanation of the differences presented by it, and those presented by the various forms of ichthyosis that sometimes supervene in early infancy, or in more advanced life, by recollecting the very opposite conditions to which the surface of the diseased skin is subjected before and after birth. During extra-uterine life, the surface of the diseased and hypertrophied epidermis is generally kept dry, and densely fissured, by constant exposure to the air, and by the flexions and movements of every part on which it may be situated, whilst by continued friction and abrasion, it is in most situations prevented, except in extreme cases, from acquiring any very great thickness. During intra-uterine life, on the other hand, the thickening crust of epidermis is, from the absence of these last causes, allowed to accumulate, and the exposure of the diseased surface to the liquor amnii, instead of the atmospheric air, may account for the other points of distinction between this new species of ichthyosis, (*Ichthyosis Intra-uterina*) and those previously known to nosologists.

As our knowledge of the pathological anatomy and exact pathological nature of this curious disease is still so imperfect, it is greatly to be desired that some of the excellent and zealous pathologists connected with Guy's Hospital would favour the profession with the result of a minute examination of the case contained in the splendid Museum attached to that institution.

I may add, that if the affection prove to be connected in any degree, as suggested in my original note of the Guy's museum case, with the "skin-bound disease," (an opinion which, as will be seen from the preceding context, I am not, on a fuller examination of the recorded cases, now inclined to hold,) there will probably be found, in addition to anasarca, appearances showing the existence of Bright's disease of the kidney. Physicians are well aware, that the most vague and contradictory opinions have been promulgated in regard to the true pathological nature of the skin-bound disease. In the Edinburgh Lying-in Hospital, I have only seen two cases of this affection, and the examination of these before and after death, with the co-existence in them of coagulability of the urine, and of slight yellow striated deposits in the structure of the kidney, seemed strongly to support, if not to prove, the idea that this affection was merely a form of anasarca connected with the occurrence of Bright's disease in the earliest period of infancy,—an opinion which had at once occurred to the acute mind of my colleague Mr Ziegler, on his examining, during the lifetime of the little patient,

the first of the two cases to which I refer. If this opinion be corroborated by additional observations, it will be valuable in two respects:—

First, As so far resolving a pathological problem in regard to the nature of skin-bound disease, that has hitherto, in a great measure, set all explanation at defiance;¹ and,

Secondly, As showing that albuminous nephritis,—a form of malady which is so frequent and important in many respects in the pathology of adult life,—is an affection which may occur, and under some conditions very frequently does occur,² at the earliest period of infantile existence, and even during intra-uterine life.

M. Souty's Case.

Since writing the above, I find a long and detailed account of the case of M. Souty (referred to in Dr Keiller's communication) has been published in the *Bulletin de L'Académie de Médecine* for October 30, 1842. In all its essential features, this case corresponds closely with the others, and it confirms in a great measure the view I have taken of its pathology. It was the third child of healthy parents, who had previously had two healthy children. About the fourth month of pregnancy, the mother had met with a severe fright, after which she was troubled with great disgust for food, intense pain in the lower part of the belly, with constant general uneasiness, and a frequent feeling of suffocation. Six weeks before her confinement, she had frequent copious discharges from the vagina of a thick yellowish matter with a foetid smell.

The whole surface of the infant at birth was traversed by reddish bands, of different shades, from a violet tint to purple, and varying in length and breadth. These inclosed irregular scaly patches of skin, which at birth were white, but soon became yellow. The epidermis on these patches was somewhat rough, its appearance resembling that of morocco leather. In some parts, it was more than a millimetre thick, and approached in hardness to the skin of the heel of the adult.

The surface of the fissures was covered with a thin pellicle like serous membrane, or like the cicatrix of a recent burn or blister. In some situations, as in the neck and inner surface of the articulations, this cuticle had, from the movements of the infant, given way, exposing a raw surface that showed very distinctly the texture of the true skin, which appeared to have preserved its natural structure. Over the thorax and abdomen, the red bands were arranged pretty symmetrically; a broad band ran along the middle line in the course of the linea alba, from which transverse lines passed off on either side. On the limbs, they were also somewhat symmetrical, and they were broadest in the neighbourhood of the joints, where a considerable space intervened between the different segments of epidermis. They were far most numerous on the anterior surface of the body and on the limbs.

¹ In illustration of the variety of opinion that has been held in regard to the pathological nature of the skin-bound disease of infants, we may mention that Andry and Auvity thought that this affection was brought on by the action of cold on the surface of the body, stopping the transpiration, and leading to condensation of the fluids in the cellular tissue; Denis and others supposed the cellular tissue to be in a state of inflammation; Baron, that it was simply œdematous, from some obstruction to the circulation; while Underwood and Stutz speak of its being in a state of tonic contraction or spasm. Others have imagined it to depend on a disordered condition of some internal organ. Thus Breschet, finding in some cases he examined, that the foramen ovale was still open, attributed the disease to this circumstance, as one of its causes. From similar evidence, Hulme ascribed it to inflammation of the lungs; Paletta, to congestion of the liver; M. Leger, to unusual shortness of the intestinal canal, &c.

² Of 644 infants received into the Infirmary of the Hospice des Enfants Trouvés at Paris during the year 1834, 289, or about 45 per cent. of the whole, were affected with this disease.

A few brown hairs were present on the head. The nose scarcely projected from the face, the cartilages being apparently bound down by the thick and almost horny integument which covered them. The auricles were also in a rudimentary state, and the foramina obscured and narrowed by the thickened epidermis. The eyes had the appearance of red fleshy projections. This arose, however, from the upper eyelids being completely everted, so that the palpebral conjunctiva projected between the two tarsal cartilages, which had been stopped in their growth by the thick and horny epidermis; on raising the red tumour, the ball of the eye itself was seen to be healthy. The mouth was kept widely open from the contraction of the epidermis. The external genital organs had also the appearance of having been arrested in their development.

The extremities were well formed, but the hands and feet were much disfigured by a collection of matter under the skin, which was so much thinned and distended that the whole had the appearance of a scrofulous tumour. The matter, which was a reddish sanies, extended up to the points of the fingers, and gave them a deformed curved appearance, so as to resemble claws. Nothing abnormal was found in the interior of the thorax.

The child lived for fifty hours; it cried feebly, but deglutition and respiration were easy; it passed some meconium, but no urine. M. Souty has deposited it in the Musée Dupuytren at Paris.

In this case the disease appears to have been originally in a somewhat milder form than in any of the others, and also to have advanced farther in the process of reparation. This is seen from the degree of cicatrization of the fissures, and by the greater distance between the different segments of thickened epidermis. In all probability, however, the surface had been at one time in much the same condition as in the other cases. This case also clearly shows that the disease was originally confined to the epidermis, and that the deeper parts, as the cartilages of the ears and nose, the external genital organs, &c. were contracted and disfigured simply from being confined by the thick and unexpandible epidermic covering. The degree of development of these parts thus gives us pretty accurate information as to the period of intra-uterine life at which the disease supervened, or at least the period at which it acquired such a degree of intensity, that the skin had become so thickened as to be able to repress these subjacent parts in their growth.