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CONGENITAL DYSKERATOSES AND THEIR MORBID ASSOCIATIONS

By Dr. E. Lenglet

A recent article by Méneau which appeared in the February issue of the *Annales*, "Ichthyosis fetalis and its relation to ichthyosis vulgaris", has led me to write the following pages. The concept that dominates the present article fundamentally differs from the one developed by said author. While Méneau combines and merges, returning to the chaos of the early days of ichthyosis, we differentiate. From the body of information reported by observers in all countries, we believe we can discern sufficient reason for these distinctions, and no great effort is required to achieve this. In France and abroad, many authors are aware of the banality, deficiency, and inaccuracy of applying the term ichthyosis to morbid varieties which are differentiated by their most striking clinical characteristics and their progression.

This interpretation of the facts has led authors both here and abroad to find new designations for the cases they consider to be new, not that similar cases had not already been seen, but because, as often happens, they had been studied with too much imprecision for their distinctive characteristics, even when highly distinctive, to be perceived. A large number of these observers voluntarily exclude the word ichthyosis from the new designations they propose, although this term is often found in the attempts to classify these morbid varieties. From this approach of excluding the term ichthyosis there results a certain difficulty for the reader who wants to find similar cases; therefore it seemed to us when examining Méneau's bibliography that the most useful cases had escaped him. It appears to us that his work can be taken from a more general point of view, examining the details of the cases, and although we do not pretend to have done something new and original, it seems useful to us to examine certain cases of dyskeratosis of congenital origin in a different light.
We will justify this attempt by citing the cases or work published by Brocq, Brauns, Glawtsche, Metscherska, Sangster, Nielsen, Schourp, Hallopeau and Jeanselme, Risch, E. Besnier and Doyon, Thibierge, Nikolsky, etc., who were unfortunately not listed in Méneau's work; not because we claim to have ourselves cited all authors who merit it, but because the cases published by the authors listed here are, by the detailed accuracy of the clinical picture, able to shed light more clearly on the pathology of congenital dyskeratoses.

Before proceeding further, we would also like to comment that we only wish to establish a little order to the disparate cases identified and compared in many works, in spite of the efforts that have been made to differentiate and separate them by many careful observers, including those listed above.

GENERAL CONSIDERATIONS AND SUBDIVISIONS. Currently, it is not yet possible to consider establishing a classification of developmental abnormalities based on their pathogenesis. Although some anatomical facts relating to the development of the skin during the embryonic and fetal period are glimpsed today, most of them still escape us and we can only compensate for such imprecise knowledge by hypotheses, reasoning for man from what is known in animals.

As for the morbid influences causing a deviation from the normal skin development processes, we can only surmise them, not having any precise knowledge of these processes.

However, by combining embryological, anatomopathological, and clinical data, some order can be established in the enormous series of congenital dermatoses, not only in specifying the most important of the morbid varieties, but also in recognizing their reciprocal relationships.

When only considering the anatomical development of the embryonic and fetal epidermis, two main facts appear to dominate development and provide the key to most of the transformations: one is the separation of the homologous layer of the epitrichium in animals; the other is the formation of the permanent epithelial layer underlying the epitrichial layer.

Fact one. It is accepted that the embryonic epidermis has two distinct layers, with the superficial layer representing the epitrichial layer in animals and ordinarily possessing
a certain autonomy during its development from initial proliferation to desquamation. It seems able to lose some of this autonomy by excessively adhering to the lower layer, not desquamating normally, and continuing to survive with the bottom layer of the epithelium. Many deformities result. Here are a few examples: • If the epithelial layer continues to survive and retain its integrity by barely adhering to the lower layer, the child comes into the world with a complete sheathing reminiscent of the sheathing found on top of the hair in Bonnet's goat. This is the most perfect case: the lower layer has an almost normal structure, it develops as if the superficial layer did not exist. • If the epithelial layer continues to survive but loses part of its individuality, it acts as an abnormal element in the development of the lower layer, and its own survival and special characteristics become opportunities for various lesions in the permanent epithelium. No doubt one should include true ichthyosis fetalis here, as well as atypical ichthyosis fetalis, with these terms having a more limited significance for us than is normally given them, as we will see.

Fact two. Normal development of the superficial layer, its appearance in the second month, and its disappearance in the sixth, in no way imply perfect development of the permanent lower layer. The developmental irregularities that may arise will vary under the influence of numerous factors; but no matter how many of these factors there may be, which is a matter unknown to us, a rough classification of the lesions in the permanent epithelium remains possible because these lesions ordinarily reproduce fairly clearly defined types which the modern clinic tends to regard, if not as morbid entities, at least as important varieties. Within these clinical forms, which appear to fall exclusively within the permanent epithelial lesions, it seems we must include agenesis and dysgenesis of the skin appendages, perhaps ichthyosis vulgaris, certainly the acanthokeratolysis which dominates in congenital bullous dermatoses, whether pure or with other symptoms.

In nature these cases are often less individualized, because there is not always a clear separation between the diverse forms. Although transitional cases exist as they do everywhere else, the same morbid types recur at a high enough frequency to merit grouping them and regarding them as landmarks and references around which the transitional cases evolve.
Far from a series of congenital dermatoses forming a chain where all links are equal, there is an enormous disproportion between the importance of the morbid varieties and types when considered in comparison to the transitional cases. This is why we will attempt to study the largest groups, without overlooking the fact that these groups leave room for cases so complex that some are kept at an equal distance from multiple primary varieties, contributing to the characteristics of each of them.

In this article, we will clearly see how these general cases, these sets of symptoms which dominate the clinical history of developmental abnormalities in the skin, when all is said and done, boil down to a few terms which are found in almost all the varieties, and these common terms are fairly narrowly organized so that one can recognize an order in them and the morbid types constituted by essential symptoms associated in a well-defined manner, and by related symptoms which are almost immaterial because they are so overshadowed by the primary ones.
Before proceeding any further, we will indicate the main terms that are found clinically in all these diseases and which constitute, alone or in association, the morbid entities and varieties.

1. Agenesis, dysgenesis of the skin appendages;
2. Functional disorders (?) of the sebaceous glands or sudoriferous glands;
3. Palmoplantar keratoderma;
4. Generalized exfoliation, lamellar exfoliation of the newborn, comparable to the so-called sebaceous ichthyosis;
5. Hyperkeratosis of type ichthyosis fetalis;
6. Hyperkeratosis of type ichthyosis vulgaris;
7. Congenital ichthyosiform erythroderma;
8. Skin atrophy and deep atrophy;
9. Acanthokeratolysis, congenital nodular lesions;
10. Epidermolysis bullosa simplex.

These morbid types exist in pure form or in association; when associated they primarily assume the following combinations, which we indicate by using the numbers designating each syndrome to avoid repeating the same words over and over.

The letters placed in front of a number designate a morbid variety; one can verify the accuracy of these associations by referring to the published data.


The figure above provides a clearer understanding of these figures.

CLINICAL AND CRITICAL STUDY. Abnormalities in the development of skin appendages. Although the developmental abnormalities which cause the dysgenesis of the skin appendages translate into a symptomatology which varies little, from case to case there is a fairly unique individuality in their evolution, and they depend on essentially distinct disorders.

These dystrophies of the skin appendages appear with an almost complete absence or a simple functional insufficiency of their matrices. They are generalized or local, sometimes affecting only the scalp, sometimes the scalp and the hair on the rest of the body, or the hair and nails or nails alone, but rarely the teeth.
Often isolated, as with monilethrix in keratosis pilaris, in certain congenital alopecias they are also frequently associated with lesions affecting all of the skin covering. As we will see, they make an appearance in the diverse varieties of dyskeratosis we will examine below.

Like these dyskeratoses, they appear influenced by the development of the epitrichial layer, but the special development of the skin appendages assures a more complete individuality than the individuality of the common tegument, and thus we fairly often see them develop on their own, without any examination as we can perform it allowing us to perceive abnormalities in the common epidermis at the same time as these dysgeneses.

These cases where the lesions of the skin appendages appear in isolation are of the least value to the point of view of our study, and therefore we will content ourselves with summarizing them with a general picture. By reviewing the medical literature, we can see that this picture is only a simple ordering of the cases published. Cases where they are in association will be discussed further on. Because of the rarity of isolated nail lesions we are leaving them out here, as we have not found sufficient reason to attempt their classification based on the published cases.

I. Certain lesions are due to *malformations of the skin appendages.*

- Generalized, almost absolute agenesis.

The result is complete or partial alopecia with absence of pilosebaceous and sudoriferous orifices.

- Incomplete agenesis, dysgenesis.
  1. Pure forms: monilethrix; keratosis pilaris;
  2. Associated forms: the same, with para-ichthyotic and ichthyotic keratoderma phenomena, with nail lesions, epidermolysis bullosa, ichthyosiform erythroderma.

II. The papillae seem to be present, but are torpid;

- *The lanugo is not shed at birth, the lanugo persists.*

This phenomenon produces Bonnet's hypotrichosis lanuginosa, Unna's trichostasis primitiva.

- *The lanugo is shed,* but the hair is replaced with abnormal hair (monilethrix), or is not replaced, which is then *definitive alopecia.*
We do not wish to detour into a discussion of the pathogenesis of this last state, but let us comment that it is possibly explained by trophoneurotic alterations which are themselves dependent on alterations in the endocrine glands (thymus, adrenal, thyroid, etc.).

We have, in this first rough classification attempt, voluntarily omitted the exceptional cases where the epitrichial layer persists and covers a well-differentiated deep epithelial layer, which is an exceptional case corresponding to the case of Bonnet's goat and the cases which we will have the opportunity to discuss further on.

Alopecia, pilar dysgenesis of any kind, and undoubtedly also alterations of the sudoriferous glands, therefore comprise the first term in the series of tegumental dystrophies; we will see them associated with the lesions examined below, progressively expanding the complexity of the clinical syndrome.\(^1\)

Once we move beyond the tightly systematized lesions of congenital origin such as those we have just indicated, we touch on much more complex processes, where the role of the common undifferentiated epidermis is highlighted in their development, although generally the adnexa are involved at the same time. This enormous group includes all the so-called ichthyotic lesions, local or generalized. In France, just as we separate palmoplantar keratoderma from actual ichthyosis, which other countries combine, we also separate from the ichthyosis group some new morbid entities for which the differentiation, done first by Brocq, denotes the prelude to a dismantling of the overly compact group of ichthyoses and its often completely dissimilar cases.

If we take an overall look at the congenital dermatoses described over the last few years, we note that we can barely distinguish three forms: 1) the former palmoplantar ichthyosis of foreign authors, which is our symmetrical congenital palmoplantar keratoderma, 2) ichthyosis vulgaris, 3) ichthyosis fetalis. All the clinical varieties are grouped almost randomly around these three forms, no matter how dissimilar they may be. However, when the cases are examined more closely, authors both French and foreign note that they do not agree with this simple all-inclusive grouping, and while Brocq singles out from this confusion a new type, which he calls congenital ichthyosiform erythroderma with hyperepidermotropism, the other authors publish cases which they

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\(^1\) Here are some of these cases: [list of references]
regard as attenuated varieties of ichthyosis fetalis, or even completely separate from ichthyosis by forcibly giving them new qualifying descriptors.

_Lamellar exfoliation of the newborn._ A first group of cases consists of ones which should not be confused with physiological desquamation in newborns, even when very accentuated.

These cases correspond to the observations of Grass and Török, Bowen, Brauns, Carini, Hallopeau and Watelet Sherwell, etc.¹ By simply reading the titles given by these authors to their observations, it would not occur to the reader, except for those concerning Grass, Török and Brauns, that there is any question of it being something other than ichthyosis fetalis in a mild form. However, when one reads the cases, one is struck by the enormous distance separating them from ichthyosis fetalis and ichthyosis vulgaris. A summary of the symptomatology of all these cases will make this clear.

The children present at birth with dry, shiny skin, light yellowish-brown in color, as if lightly coated with collodion. Shortly after birth (four hours in the Brauns case, fifteen minutes in the Hallopeau case, immediately after for Grass and Török), the epidermis superficially cracks and no fissures or ulcerations appear. The phenomenon is easily explained: the child is in a superficial epithelial sac, independent or almost completely independent of the true epithelium which remains unaltered, while the superficial sac splits, detaches in large strips and falls off most of the body. This superficial covering is compared by all authors to a very thin film of collodion applied to the entire body. Within a few days the desquamation of this lamina is complete in the typical cases, and the anomaly ends with the desquamation.

This is the most simple and most perfect case, but it is not the only one possible; the observations also report: temporary ectropion noted by Hallopeau and Watelet, Sherwell; the apparent scleroderma or atrophy noted by Hallopeau; phenomena so subtle that they are insufficient to bring to mind ichthyosis fetalis, no more than atrophy of the fingers does in other cases which we will examine further on.

This does not mean, however, that nature has violated its normal practice and created a morbid type so distinct that it cannot be complicated by more severe symptoms, that the special development which causes the formation of the collodion surface layer

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¹ [list of references]
cannot be accompanied by processes leading in the direction of ichthyosis and establishing, as exist everywhere, intermediate stages between this morbid type, ichthyosis vulgaris, and diffuse malignant keratosis. It only means that there is a very singular morbid type which can evolve without any ichthyotic transformation of the skin.

The authors who have seen this morbid type have clearly perceived its specificity: Grass and Török proposed giving it the name of lamellar exfoliation of the newborn (1893), Carini called it lamellar ichthyosis (1893), and Brauns used *superdesquamatio membranacea* (1897). We believe that preference should be given to the name proposed by Grass and Török, and all similar cases should be placed under the label *lamellar exfoliation of the newborn*.

So here a first type has been established, or so we believe, and the fact that it usually is assigned the appropriated title of ichthyosis in the literature does not grant it the characteristics of ichthyosis.

The pathogenesis of these cases is still obscure. Bowen, who has particularly focused on homologues of the epitrichial layer in man, is inclined to believe that the lamellar exfoliation of the newborn is dependent on an abnormal persistence of this layer. This explanation is quite reasonable, and has the advantage of linking the lamellar exfoliation to ichthyosis fetalis without merging them, because the lamellar exfoliation is the manifestation of an almost normal development of the epitrichium, while the ichthyosis fetalis is possibly explained by an aberration or non-differentiation of the epidermal cells, which should, in normal skin development, form two superimposed and distinct epithelial layers.¹

¹ Before proceeding any further but without overly dwelling on the embryology of the skin, it is helpful to say a few words on the epitrichium and its probable role. In the human embryo at the end of the second month, the ectoderm is composed almost entirely of two rows of distinct cells. The basal layer is composed of small, fairly regularly arranged, almost flat cells with small nuclei. These cells represent the permanent layer, the one in which cells will later differentiate to form the entire adult epidermis. The external layer is formed of much larger and less ordered cells which are globular or polygonal in shape. This external layer is recognizable in the second to the sixth month of intrauterine life. It has nothing in common with a layer following the normal keratinization path, as is proved by its histochemical reactions. This layer gradually becomes independent of the basal layer. The epitrichium is lifted by the growth of the hair which never passes through it, and at no point do its cells appear to tend towards a keratic evolution. The adherence of the epitrichium, its thickness, its relationship with the persisting layer vary from one point to another in unknown proportions, but it is probable that topographical and anatomical reasons facilitate, in some cases, its adherence and its persistence in certain regions more than in others. From what we have just written there is only a single step to a morbid deduction, which we dare to make: in the first two months of intrauterine life the temporary epithelial and definitive epithelial formations exist in the ectoderm, but they
**Ichthyosis fetalis.** The second type that can logically be established from what we have just said is ichthyosis fetalis, a diffuse malignant keratosis. The symptomatology is sufficiently well-known for it to be unnecessary to dwell on it here, but let us indicate the main traits:

The child is covered at birth with a horny shell, soon split by numerous deep fissures into a series of areas separated by rhagades extending to the dermis. It is evident that there is nothing reminiscent of the normal epidermis found in the previous case. This thick and rigid shell extends over the entire body, causing retraction of the eyelids and mouth, hiding the nose and ears, with flexion contractures of the limbs to the trunk and the limb segments to each other, and encasing the fingers, in short resulting in such deformity that the child is no longer human in form.

**Transitional cases.** Isn't this a completely different picture from the one described earlier and the case of Hallopeau and Watelet, regarded by almost all authors as an attenuated form of ichthyosis fetalis; on the contrary, isn't this much closer to what we could call an aggravated form of lamellar exfoliation? In the case of Hallopeau and Watelet, there existed conspicuous discoloration of the fingertips which resembled dead fingers, in addition to the collodion-like desquamation. The limbs were half bent and could not be straightened because of the resistance of epidermis. The ectropion was very obvious at first, but disappeared as the desquamation decreased. We therefore find in this case several phenomena which bring to mind very attenuated ichthyosis fetalis, but these symptoms of flexion of the limbs, ectropion, white and half-flexed fingers, are not special to ichthyosis fetalis; we will see that they exist to varying degrees, sometimes quite accentuated, in cases where the general characteristics of the lesions are quite far from those of ichthyosis fetalis or even ichthyosis vulgaris. This is true in certain congenital keratodermas and in certain cases of congenital ichthyosiform erythroderma which we will cover later on. Similar cases are therefore links between the various diseases, but the predominance of certain symptoms requires that they be combined with one group over another, in this particular case lamellar exfoliation of the newborn.

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are still taking shape; what will happen if the ectoderm is impacted at this stage of its development? The conclusion is too easy to reach, and is still completely hypothetical; it is better to wait and allow the facts to speak in the future.
One degree further and we approach the clinical picture and development of ichthyosis fetalis: the desquamation occurs slowly, the collodion-like strips, instead of detaching easily, adhere tightly with their inner surface, and fissures occur in the flexural folds. The nails are involved in the formation disorders, but after two or three months, sometimes more, all gradually returns to normal. Sherwell's case is an example. We can therefore progressively advance in imperceptible degrees from lamellar desquamation to ichthyosis fetalis, but one cannot conclude from this that these two terms designate the same disease at two different stages of development, but only that morbid forms can always be reduced to one or more fundamental types, which must be conserved and clearly defined. Without these landmarks the morbid types become intermingled.

*Palmoplantar keratodermas. Their associations.* - We saw a new symptom enter the picture in a previous observation: the palmar lesion and the digital lesion. These lesions on the extremities are fairly common and are of sufficient importance for us to examine their relationship to the group of ichthyosiform lesions before continuing on.

If we content ourselves with simply noting the cases where a palmoplantar lesion is found in congenital dermatoses, without attempting an interpretation, we see that it can: 1) evolve on its own; 2) be associated with lesions of varied extent and type.

1) When the *palmoplantar lesion evolves alone*, it constitutes the large group in other countries entitled palmoplantar ichthyosis, congenital hereditary symmetrical palmoplantar keratodermas as they are called by E. Besnier. Little variety exists in this first group, and it can be outlined as follows: disease beginning very soon after birth, sometimes from the age of one month to three and five years old, preceded in this case by an erythrodermic phase which often passes by unnoticed, accompanied by hyperhidrosis with or without perceptible lesions involving the sudoriferous glands, resulting after this first stage in the appearance of smooth or rough keratoderma, sometimes accompanied by sclerodermic and atrophic fingers, frequently invading the dorsal surface of the fingers to a variable extent but almost never deforming the metacarpophalangeal joints. These fundamental symptoms have a certain reciprocal independence, the erythema and the hyperhidrosis by themselves possibly representing the vestiges of a keratoderma which will not evolve in the direction of hyperkeratosis. The hyperkeratotic lesions are variable in appearance, but the most interesting type is definitely the one which begins with
hyperkeratosis of the sudoriferous orifices. This type appears elsewhere, as we will see, more often in association rather than in isolation. Aside from these cases where the hyperkeratosis begins with erythema and hyperhidrosis, the most common, others exist of much greater theoretical interest: those where the keratoderma is preceded by bullae. These bullae, as we said above, are one of the favorite manifestations of congenital dermatoses. They seem to result from an abnormal friability in the binding between epidermal cells, an acanthokeratolysis as it is called by Nikolski [sic]. This friability is found in a number of congenital states, and we will revisit it. The bullous pre-keratotic process is found in the cases of Bennet, Alpar, and Du Castel. This isolated hyperkeratosis has characteristics identical to those that define the keratosis associated with all varieties of abnormalities in congenital development. We will therefore content ourselves with pointing out the special associations observed in the second group.

2) One can schematically classify the associated palmoplantar keratodermas as follows:

1) Formes frustes of palmoplantar keratoderma with lesions in the skin folds.
2) Palmoplantar keratoderma with lesions in the skin folds.
3) Palmoplantar keratoderma associated with alterations to the skin appendages which do not constitute a special morbid type.
4) Palmoplantar keratoderma associated with congenital lesions constituting a defined morbid type.

1) The first group: formes frustes of palmoplantar keratoderma with lesions in the skin folds, is the counterpart of the forme fruste we reported earlier. It justifies what we have written in a prior work: that the keratoderma cannot by itself be a value used for classification. An almost schematic type is provided by a case of Du Castel and Baudoin. In this familial case, the lesions reside exclusively in the creases of the elbows and armpits, groin, popliteal fossa, hands, and feet. These were essentially formed by hyperhidrosis.

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1 [references for Bennett, Alpar, and Du Castel]
2 [reference for Du Castel and Beaudouin]
2) *The true palmoplantar keratodermas with lesions in the skin folds* are represented by the types similar to Meleda disease and by Meleda disease itself. Neumann, Ehlers\(^3\) note this thickening of the skin in the folds, and the latter author considers it an important symptom. In Neumann's observations one notes the existence of hyperkeratotic plaques on the elbows and knees, the skin appendages already affected; the type is not completely pure.

This type is comparable to the cases where the lamellar desquamation of the newborn is accompanied by fissures in the skin folds and scleroderma of the extremities, but especially the cases where the ichthyosiform lesions predominate in the skin folds and exist on the palms as is seen in certain of Brocq's congenital ichthyosiform erythrodermas.

3) In the cases where the *palmoplantar keratodermas are accompanied by lesions in the skin appendages without constituting a defined morbid type*, the picture may be of considerable complexity. The nails are altered in some of the patients in the family studied by Dubreuilh and Guélain\(^1\). Hebra published a case with thickening of the facial skin and dilation of the sebaceous follicles. This case provides the transition to ours, which is the most complex of those published. In our case all the above lesions are found combined: severe palmoplantar lesions, hyperkeratosis of the skin folds, raised nails due to nail bed hyperkeratosis, agenesis of all hair on the body, complete alopecia, and considerable dilation of the sebaceous orifices on the face and scalp. Erythroderma. Palmoplantar hyperhidrosis.

Considering these noteworthy facts, it seems that in the morbid group we have just described one sees a special morbid system evolving on its own, where the keratoderma, with the skin appendages and skin fold lesions, is the predominant symptom. However, if one reflects on the considerable role that the sudoriferous glands seem to play in this morbid complex, it would seem that we are in the presence of a very special group which is completely opposite of the ichthyosis vulgaris group, uniting with the group of congenital ichthyosiform erythrodermas and thus contributing to distinguishing the congenital ichthyosiform erythrodermas from ichthyosis vulgaris.

\(^3\) [references for Neumann and Ehlers]
\(^1\) [references for Debreuilh and Guélain, Hebra, Lenglet]
This union is even more complete when one studies the fourth of the groups we have defined, the *palmoplantar keratodermas associated with congenital lesions constituting a defined morbid type*. Note the existence of the palmoplantar keratoderma with the ichthyosiform lesions: lamellar exfoliation, attenuated forms of ichthyosis fetalis; but especially with Brocq's congenital ichthyosiform erythrodermas, which we will revisit later. Lastly, complicated forms of hereditary traumatic pemphigus or epidermolysis bullosa are sometimes associated with palmoplantar lesions which manifest as atrophy, thickening, retraction of the entire palmar surface, at the same time as the bullous lesions develop around the major joints; there are nail lesions and hyperhidrosis, even congenital alopecia. New connections between keratoderma, bullous ichthyosis, congenital ichthyosiform erythroderma, and epidermolysis bullosa. A new opportunity to remove from these groups the localized ichthyosis vulgaris, because of the contrasting symptoms.

Don't all these cases sufficiently indicate the absolute necessity of establishing landmarks in order not to blindly skim across all the disease classifications for congenital dermatoses? In fact, all the symptoms we have so briefly reviewed may combine in diverse ways to pose a puzzling variety for the clinician; the alopecia, nail dystrophies, palmoplantar hyperkeratosis, hyperkeratosis of the skin folds, sebaceous dilation, hyperhidrosis, periarticular retraction, bullae, and skin atrophy form the foundations for all these similar dermatoses, and it is the eclectic evaluation of the dominant symptoms which will enable the classification of ambiguous cases.

*Brocq's congenital ichthyosiform erythroderma.* We are naturally led by the above to mention complex dermatoses, of which there are a few scattered cases published, ordinarily called ichthyosis and regarded as varieties of ichthyosis simplex although they only have a distant relationship to these two entities. These dermatological varieties constitute a new class defined by Brocq\(^1\) based on his personal observations and on other observations collected from various sources. In the footsteps of our mentor, we have taken up a new study of these observations, and we will provide a very brief summary of what he saw and what we think we observed in our later study.

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1 [reference for Brocq]
This class of congenital ichthyosiform erythrodermas is constituted essentially of erythrodermic dermatoses, an essential aspect, unvaryingly found on the face, in the skin folds in the form of hyperkeratosis; occasionally accompanied by bullous lesions and palmoplantar keratoderma, with these two lesions evolving separately or in association. In addition to these major symptoms, there are others of less value because they appear to be less fixed: exaggerated growth of the hair and nails, palmar hyperhidrosis, alopecia, skin retraction, localized atrophy, and dilation of the sebaceous orifices.

Although we find symptoms here that are analogous to those we described earlier, the grouping and the preponderance of some of them render any doubt impossible when faced with such well-characterized cases of this magnitude.

Before describing the complex forms of this group, let us establish a brief outline modeled on Brocq's observations.

Congenital ichthyosiform erythroderma as viewed by our mentor is essentially characterized by erythroderma, the invasion predominating in the skin folds, the exaggerated rapidity of the growth of the horny skin appendages, the lesions on the face, the bullous lesions; already the latter are no longer a functional symptom, as they are inconsistent. The erythroderma extends to all the skin, appearing most often after birth; we do not find it reported in all cases, and even those which are the most conclusive sometimes make no mention of it. There is no doubt that this is a simple lapse in the observation, because the erythroderma is permanent; the proof of this omission lies in two essential cases reported independently by two authors; in the two cases, the erythroderma was unnoticed by the first two clinicians but was seen and reported by two consecutive observers and was even used by them to distinguish these cases from ichthyosis. These two observers reached the same conclusions although they did not know each other: Brocq after E. Besnier in case II in Brocq's paper cited above, and Metcherski after Glawische in a case which we will revisit. It is interesting to note that the vast majority of the authors who observed the erythroderma no longer entitled their case ichthyosis, instead attempting to find a more appropriate label. Rasch called it erythodermia exfoliativa universalis congenita familiaris; Alpar adopted exfoliatio epidermidis neonatorum; we know Brocq's label; Metcherski reused Hebra's name, hyperkeratosis striata erythematosa, etc. Here there is more than a vague suspicion of the
specificity of these morbid forms. The erythroderma is not severe, which explains why attention was not paid to it, and it is often hidden by the hyperkeratosis of the entire tegument, but it becomes quite clear if one considers the surfaces exposed by the shedding of the strips or squares of hyperkeratotic epidermis. It appears even more frequently on the face, resulting in a fairly unique coppery appearance which was noted several times by the observers. This erythroderma is reminiscent of pityriasis rubra pilaris.

*The invasion of the skin folds* is an almost invariable phenomenon, noted by Thibierge¹; by M. Joseph²; by Hallopeau and Jeanselme³; by Glawische and Metcherski⁴; and in another case by Thibierge⁵. Other examples exist, and one will certainly be struck by the fact that all these cases are referred to as "ichthyosis". There is nothing surprising in this because the first classification and subdivision efforts were made by Brocq; above we cited several other names applied to diseases of the same order. In the clear cases, the keratosis of the skin folds presents as parallel crests several millimeters thick in the flexion creases, of a blackish horny substance which only seems to have been thus divided by the necessity of flexing the limb. These horny crests are often pressed together, and are divided by narrow perpendicular furrows into cones and cubes, in papillae so tall that they result in an extraordinary villous appearance in the armpits and the creases of the elbow and popliteal fossa. These horny cones barely adhere to the underlying surface, and can be detached without causing pain to the patient; they soften without difficulty as their keratinization is incomplete. Below them the surface of the dermis appears red or bright pink, showing through the completely conserved layers of the deep epidermis which neither bleeds nor seeps, at least in the clearly formed case. The black hyperkeratosis may be much less systematic with very irregular development, and may be only a local exaggeration of the dyskeratosis of the entire tegument. This thickening in the skin folds affects the wrists, elbows, armpits, neck, popliteal fossa, and more rarely the groin.

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¹ [reference for Thibierge]
² [reference for Joseph]
³ [reference for Hallopeau and Jeanselme]
⁴ [reference for Glawische]
⁵ [reference for Thibierge]
Exaggerated growth of the superficial epidermis sometimes accompanies the growth of the hair and nails, but this phenomenon is still not well known, as Vidal and Brocq were the first authors to report it and additional cases will be needed to contribute more information on this point.

We can only write that we are entering into a multitude of contingent symptoms of these dermatoses; one would be tempted not to attribute to these symptoms the importance they deserve, if one thought that their contingency and their variability implied a lack of value clinically. But the reverse is true: these symptoms have a specific significance, because they are ordinary symptoms of congenital dermatoses that assume the appearance of neither ichthyosis fetalis, nor lamellar exfoliation of the newborn, and even less ichthyosis vulgaris.

We already know most of these symptoms, but it is important to mention them here, as they will allow us to outline further on a few target varieties in this considerable ensemble of Brocq’s ichthyosiform erythrodermas and para-ichthyoses.

Frequently associated with the erythroderma and skin fold lesions is palmoplantar keratoderma, sometimes separated into cones and cubes packed closely together, sometimes smooth and uniform, with a seeping yellowish white epidermis that appears macerated, reminiscent of Meleda disease. The same is true in one of the Thibierge cases cited earlier, in the case of Max Joseph, of Sangster\(^1\), of Nielsen\(^2\), etc. We have a confusion of choices, as there are many others, too, which we will reserve for other comments. Similarly to the essential congenital keratoderma which we mentioned earlier, this asymptomatic keratoderma is accompanied by hyperhidrosis, and is even dominated in a special variety by lesion of the sudoriferous glands as we will see later, objectively evaluated in the observations of Giovannini\(^3\), Thibierge, Glawische, and Metscherski.

But the syndrome in the extremities continues further and atrophy, hypotrophy, sclerodermic appearance are found here in association with erythroderma, lesions of the skin folds, palmoplantar keratosis, often dominated by the latter. In the Joseph case mentioned above, disseminated and verrucous lesions affect the skin folds and the palms, with very pronounced atrophy of the hands which the author attributes to compression

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\(^1\) [reference for Sangster]
\(^2\) [reference for Nielsen]
\(^3\) [reference for Giovannini]
from the hyperkeratosis. In the very complex Rona case\textsuperscript{4}, the initial erythroderma is followed by hyperkeratosis, and on the hands the hyperkeratosis is complicated by very considerably atrophy. Affected skin folds, palmoplantar keratoderma, considerable atrophy of the hands which are small, red, and shortened in the Schourp case\textsuperscript{1}. Same condition in the Hallopeau and Jeanselme patient\textsuperscript{2}: generalized lesions, invasion of the skin folds, ectropion, "on the fingertips the skin is attached to the bones as is seen in scleroderma... the hypotrophy of the hands is striking, their transverse diameters are only 10 centimeters instead of 12." This interferes with the movement of the fingers. We should remark that this relative disability, due to retraction or insufficient development of the periarticular tissues, especially the skin, is frequent in these congenital erythrodermas with hyperepidermotrophy; in one of our own observations we wrote: "The movements of the limbs are limited by the impossibility of completely extending the folds in the major joints; the tests which we conducted caused the child to cry out in immediate pain."

The symptomatology of the syndrome does not stop there. Clear and even predominant lesions of the epidermal adnexa are in some cases added to the erythroderma, periarticular hyperkeratosis, and palmoplantar keratoderma. These lesions affect the horny appendages of the skin or the glands. Rasch\textsuperscript{3} noted dilation of the sebaceous glands on the face; his case falls within the Brocq classification because of the generalized desquamation occurring in dark red erythrodermic skin, accompanied by dyskeratosis and thickening of the joint folds.

The Alpar infant had extremely red skin from the first days after birth, the scalp is covered with seborrheic crusts, the hair is thin. The Glawtsche case is notable because it is so clear: all the skin is affected, but lesions predominate in the armpits, popliteal fossa, elbow creases, buttock creases, with digital atrophy, palm retractions, erythroderma noted only by Metcherski, hyperkeratosis of the sudoriferous orifices which is so obvious, Pospelow observed that from the location of the horn one can infer the presence of a sudoriferous gland, which is verified histologically. Analogous phenomena are seen in

\textsuperscript{4} [reference for Rona]
\textsuperscript{1} [reference for Schourp]
\textsuperscript{2} [reference for Hallopeau and Jeanselme]
\textsuperscript{3} [reference for Rasch]
the observations of Thibierge\textsuperscript{4}, but here new symptoms appear, with the lesion gaining in complexity: the child never had hair, the nails resemble the pith of reeds, the mucous membranes are affected; the rest of the case coincides with the others, and the palmoplantar lesions are probably triggered by the alteration in the sudoriferous glands. This is not an isolated case, as Giovannini\textsuperscript{1} reported one that is quite comparable, coinciding almost exactly with the case which we published and discussed earlier concerning palmoplantar keratoderma associated with lesions of the skin appendages.

To those who have followed this overly concise report thus far, it would seem that we have traveled a circle where each degree is an alteration resembling the points to each side but differing sufficiently that the link is not immediately apparent, and one must travel the entire circle before seeing the general direction and orientation of the path followed.

\textit{Congenital bullous lesions} - But more remains to be said: we can repeat here what was written above concerning bullous lesions in palmoplantar keratoderma. These bullae, a preferred symptom of some congenital dermatoses, are undoubtedly dependent on an unusual friability of the intercellular bridges, which is itself steered by a phenomenon in the epidermal development. However, their appearance indicates a very special kind of transformation undergone by the epidermis, its significance justifying the distinction made by Brocq between the congenital ichthyosiform erythrodermas of a \textit{bullous variety}, and those of a \textit{non-bullous variety}.

The appearance of bullae complicates but does not change the general process described above. In the observations of E. Besnier and A. Doyon\textsuperscript{2}, there were locations on the child's body at birth where the epidermis was raised as if a vesicant had been used; this child was later seen by Brocq, and was one of first primary cases of congenital ichthyosiform erythroderma. At that point the bullous eruptions had greatly decreased in intensity, but the patient had all the characteristics of ichthyosiform erythroderma. E. Besnier had already noted how much this case differed from true ichthyosis, as he wrote: "At first appearance this seems to be the most pronounced form of true ichthyosis that one can imagine. But even before having heard the account just reported, the intensity of

\textsuperscript{4} [reference for Thibierge]
\textsuperscript{1} [reference for Giovannini]
\textsuperscript{2} [reference for Besnier and Doyon]
the exfoliation which is lamellar in many points, the complete encasing of the hands and fingers in a sheath of layered lamellae which can be torn off fairly easily; the invasion of the face, scalp, and all flexion creases, eliminate the initial impression. At some points on the instep and wrist, there is slight seeping when squamae are removed, and the rubbing of a shoe or cuff results in bullous swelling." We should briefly stress this traumatic etiology of the bullae. It is in the same child that Brocq later noted erythroderma, and rapid growth of the nails and hair. After reporting the symptoms and noting the decrease in the bullae, our mentor concluded: "In sum, the locations of the areas primarily affected in this disease are the complete inverse of the locations in true ichthyosis vulgaris." By all its other symptoms this case is the counterpart of those we have already reported: palmoplantar keratoderma exists here. The same comments apply to the Nikolsky case, where the author reported inconsistent flares of fever with the appearance of the bullae; we have noted the same in one of the Brocq patients. Nikolsky\textsuperscript{1}, much struck by the bullous processes, proposes for his case the title \textit{acanthokeratolysis universalis congenita}.

These two cases of hyperkeratosis complicated by bullous lesions should recall those of Du Castel and Alpar, where the palmoplantar keratoderma is the consequence of a bullous process appearing almost immediately after birth.

These lead us to cases which are noteworthy because of the distinctiveness of the symptoms, although the keratoderma does not appear: one such is case no. 1 in the Brocq paper, which we have supplemented in our work on congenital dermatoses. This case is one of the most complete and specific of the disease of concern.

\textit{The bullae} of the congenital ichthyosiform erythroderma appear primarily at the points where the skin is subject to trauma, particularly near the joints and on the neck, where movement facilitates the detachment and production of these elements. They are located fully within the epidermis, with little or no distension: they are only a simple detachment of two strata in the malpighian layer, between which some serosity accumulates. They dry out shortly after, then others form next to them or in the same area; any trauma avulses the hyperkeratotic layer above them, uncovering sections of the red or pink basal layers of the epidermis which do not seep if the avulsion does not take

\textsuperscript{1}[reference for Nikolsky]
place immediately after the formation of the bullae. This reddish pink surface is soon covered by hyperkeratosis and the original appearance returns. At certain periods the bullous process seems to be exaggerated, sometimes with a slight fever, and during the days that follow this bullous and febrile occurrence the hyperkeratosis drops extensively, only to recur soon after. During its existence the bullae seem to decrease in intensity and frequency, and through phases of remission and exacerbation the patient gradually is completely cured or there is at least a considerable improvement.

Is it necessary to stress further the special characteristics of this new morbid type? Its relationship with the various congenital syndromes, palmoplantar keratoderma, skin fold lesions, retraction, scleroderma or atrophy of the extremities, hyperhidrosis, dilation of the sebaceous glands, disorders in the development of the skin appendages, hyper- and hypotrophy of the hair and nails, and bullous lesions, relate it to the entire category of these special lesions as examined above. The erythroderma, facial lesions, and exaggeration of the skin fold lesions, special characteristics of this barely-adhering keratosis frequently complicated by bullous lesions, define a special, well-determined group.

_Transitional cases._ Between this group and similar groups there are transitions indicating the connections and we will indicate these as we go, merely summarizing them here in a few words: these transitions closely link the congenital ichthyosiform erythrodermas: 1) to dissociated syndromes of congenital lesions; 2) to pure palmoplantar keratodermas; 3) to pure hypotrophies, atrophies of the tegument which are of congenital origin; 4) to congenital bullous dermatoses: epidermolysis bullosa and hereditary traumatic pemphigus.

As for the other varieties of congenital dermatoses, they are poles apart from these cases, at least in the clinical picture, as we will see below; this is why we do not regard them as having distinct connections with it.

We have just indicated the existence of transitional cases between these congenital erythrodermas and the _pure congenital atrophies_ of the tegument and even of all the tissues. There is no need to emphasize this, as we have seen the association of the atrophy, the sclerodermic manifestation, and the hypotrophy in all the varieties of congenital dermatoses just examined. Also, there are cases where these dermatoses
evolve without atrophic manifestations, while there are others where the atrophy constitutes the entire morbid picture. These are the extremes in the series; these cases which we have termed landmarks are the pure types which should serve as focal points, as failing to recognize their individuality would result in confusion. We have examples of these pure local or generalized atrophies, in the cases of Audry and Dulous¹, Tændlau², and Behrend³.

As for the transitional cases leading towards congenital bullous diseases, the cases are numerous and puzzling. There are, as we know, two main forms of epidermolysis bullosa or hereditary traumatic pemphigus. The first is a pure form, characterized essentially by the evolution of the bullae around the joints or in the regions of the skin exposed to trauma. These bullae are frequently hemorrhagic, occurring on a slightly erythematous foundation, evolving within a few days to heal without leaving scars. Hyperhidrosis is common; it persists in the regions affected the most often as a special puckering of the skin with regional pigmentation, but there are never scars or mucosal and ungual lesions in this simple form.

Once we begin to study the complicated form, we no longer see so clearly the boundaries of hereditary traumatic pemphigus, Brocq-Duhring polymorphous dermatitis, or Brocq's congenital ichthyosiform erythroderma with bullous outbreaks.

In fact, we find in the literature cases labeled bullous dermatolysis, where each case is an ensemble, a kind of syndrome grouping a wide variety of congenital dermatosis symptoms: generalized bullous eruptions with successive flare-ups, absence of all nails, alopecia, atrophy of the teguments, dermo-aponeurotic retraction, pityriasic desquamation, ichthyosis, keratoderma, palmar hyperhidrosis. It is probable that the existence of bullae has led many to regard these cases as belonging to the epidermolysis bullosa group, while they could just as rationally be connected to congenital ichthyosiform erythroderma. However, one must recognize that certain cases are impossible to place in either class, such as the Wende case⁴, where a child became alopecic three months after birth, and in whom all skin appendages were absent; the child

¹ [reference for Audry and Dulous]
² [reference for Tændlau]
³ [reference for Behrend]
⁴ [reference for Wende]
had palmoplantar keratoderma, bullae, and lesions on the buccal mucosa. There is also the case we published with M. Mantoux which cannot be placed in any specific class.

Even so, it is still true that aside from the simple form of epidermolysis bullosa, one must recognize the existence of a complicated form which may have numerous links with the other congenital bullous dermatoses: this form is essentially characterized by bullae which leave scars and epidermal cysts, by nail alterations, by palmoplantar hyperhidrosis, and by lesions on the buccal mucosa, but this pure type is rarely encountered.

All these congenital bullous conditions have one obvious connection: the friability of the bridges between the epidermal cells, or acanthokeratolysis as Nikolsky calls it.

*Ichthyosis vulgaris.* This is the part we have left aside until now. We can say that the morbid types we have just reviewed are, by the sites most affected, inverted ichthyoses. In fact, ichthyosis occupies the entire tegument aside from the joint folds, palms, and soles. The palmar hyperhidrosis which is so frequent in the previous forms is replaced by absolute dryness in ichthyosis vulgaris. Just as there are primary lesions where hyperhidrosis is the main sign, as in the case of Du Castel and Baudouin, there are also primitive ichthyoses where the only symptom is the dryness of the tegument. Ichthyosis does not affect the face; while congenital ichthyosiform erythrodermas, lamellar exfoliation of the newborn, and even more the diffuse malignant keratoma, affect it constantly. The hair is absent in ichthyosis in the regions where hair normally reaches a certain level of development, while the extremely hyperkeratotic regions are traversed by numerous fine body hairs in Brocq's ichthyosiform erythrodermas, as we have related. Mucosal lesions do not appear to exist in ichthyosis, or at least they are not recorded in the cases of true ichthyosis vulgaris; it is not rare, however, to find them in our opposing dermatoses.

Does this mean that there can't be ambiguous cases, or transitional cases as we and our mentor call them, which establish the transition between ichthyosis vulgaris and the inverted ichthyosis class of hyperkeratoses? Certainly not, but it is certain that these transitional cases are less common in these two types of lesions than everywhere else.

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[1] reference for Lenglet and Mantoux
It was therefore of some surprise to us when we read Dr. Méneau's article, where the conclusions are as follows:

"To summarize, most of the authors who have written of ichthyosis fetalis have only included under this label the cases of monstrous deformities incompatible with survival, or only permitting a survival that is short-lived. The cases capable of longer survival have been confused with the vulgaris form…"

"In our opinion, there is no discontinuity between the attenuated forms of ichthyosis fetalis and the severe forms of ichthyosis vulgaris, which are links in the same chain. One should regard these as differences based on the level of intensity of the lesions, with no possible demarcation between the types, where the extremes consist of fatal ichthyosis fetalis at one end of the chain, and the other end by the simple forms of ichthyosis vulgaris which develop later on; between these extremes are found the transitional cases, those cases of ichthyosis developed in the womb which present an exaggerated form of the same lesions as ichthyosis vulgaris and the ichthyosis developed in the first months of intrauterine life, the hereditary ichthyosis of the authors."

Before we begin a brief critique of Méneau's opinions, let us remember that he adopts in his publication an unvarying stance in opposition to what guides us in the writing of this article: Méneau wants to establish an analogy between all forms described by the authors that is sufficient to justify combining all varieties; we, on the other hand, want to prove what has been clearly noted by some of the observers we have cited: that in this enormous set of diverse facts, there is a certain number of fundamental clinical types interconnected by transitional cases, as all diseases of the organs are interconnected in medical pathology.

In doing so, we have the advantage of having interpreted specific cases, retaining wherever possible the character their authors wanted to give them. This is how we were able to establish a certain number of types in the crowd of congenital dyskeratoses, the primary ones being the Grass-Török lamellar desquamation of the newborn, true ichthyosis fetalis, Brocq's congenital ichthyosiform erythroderma, palmoplantar keratoderma and its associations, the pure circumscribed or generalized atrophic lesions, the bullous lesion types complicated with one of the previous morbid forms, and ichthyosis vulgaris.
To Méneau, all these types are immaterial; the names of some of them do not even appear in his memorandum.

We have a great number of reservations concerning his way of thinking. It is not accurate to state, as he does in the first sentence of his conclusions, that "the cases (of ichthyosis fetalis) likely to survive longer have been confused with the vulgaris form." What are these cases confused with the vulgaris form? We have noted quite the opposite, that the authors who have published over the last ten or fifteen years have all focused on finding dominant symptoms in the morbid clinical pictures to justify a new and more accurate understanding of these dermatoses. As for the forms more similar to ichthyosis fetalis, we have seen Hallopeau and Watelet entitle their case ichthyosis fetalis; Sherwell and Ballantyne\(^1\) label their cases congenital ichthyosis and not ichthyosis vulgaris. Authors who describe lamellar desquamation of the newborn without any awareness of the other cases, focus on the need to avoid confusing such cases with ichthyosis vulgaris, and each of them attempts to find a name to differentiate it: Grass and Török call it lamellar desquamation, Brauns uses superdesquamatio membranacea; Carinin discusses the diagnosis of true ichthyosis and ichthyosis fetalis, excluding these dermatoses to admit that this is a variety of the so-called sebaceous ichthyosis, a title which was unacceptable to him and he proposed replacing it with the name lamellar ichthyosis. Brauns does not even believe he needs to discuss the question of ichthyosis vulgaris. In the case reported by Bowen, he establishes a relationship with congenital ichthyosis. We believe we can therefore state, contrary to Méneau, that the contemporary authors at least no longer confuse these cases with ichthyosis vulgaris, some of them refusing to regard it even as an attenuated form of ichthyosis fetalis, and they logically consider it a special variety of lesions resulting from the development of the ectoderm.

Méneau also writes: "the symptomatology does not confer any absolute and special character to one of the two ichthyosis, preventing identification of the morbid process." This is not what the vast majority of current authors believes. Omitting the didactic opinions that the necessities of teaching require adapting to given frameworks, one only needs to look at the facts to see what they say.

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\(^1\) [Ballantine. Congenital ichthyosis, *Arch. f. Pediat.*, 1894, V XI. The other names cited in these pages correspond to the bibliographical references provided above.]
They can be organized into two classes: in one we will place those that observers have labeled ichthyosis with various adjectives; in the other, we will place comparable or coinciding cases which their authors wanted to designate with other names.

First category of cases. Thibierge published three; he separates them from ichthyosis vulgaris and includes them with abnormal varieties of ichthyosis fetalis. Later, Brocq goes even further, taking two of these cases and separating them from ichthyosis fetalis to classify them in his new group of congenital ichthyosiform erythrodermas. Méneau is not at all hindered by this, and attributes them in defiance of Thibierge and Brocq to ichthyosis vulgaris. The third case coincides with the Glawtsche case, which we have mentioned, and is regarded by us as Glawtsche, Mecher斯基 and Pospelow regard their identical case: these two cases are separated from the ichthyosis class by their authors: 1) because the lesions occur in an erythematous setting; 2) because the armpits, face, elbow creases, and popliteal fossa are almost exclusively what is affected. Méneau includes them with ichthyosis in general. The very comparable Giovannini case undergoes the same treatment, in spite of the erythroderma, palmoplantar keratoderma, nail lesions, alopecia, and hyperkeratotic lesion of the sudoriferous glands. Where is the ichthyosis vulgaris here?

Second category of cases. Here we mean the cases that are related to the ones entitled ichthyosis by their authors. Méneau completely omits those cases where the authors did not believe they should use the term ichthyosis, although they are completely analogous to the above. Therefore he remains silent on the comparable cases of Rasch and Sangster, one of which is entitled keratolysis exfoliativa while the other is called erythrodermia exfoliativa congenita familiaris, both of them seeming to fall within the group of congenital ichthyosiform erythrodermas. Thus Brocq's congenital ichthyosiform erythrodermas are completely ignored, although one of the cases is established on symptoms so similar to ichthyosis that E. Besnier first believed it was "the most accentuated form of black ichthyosis one could imagine."

It would not be difficult to pursue this exposé further with numerous supporting cases of the same order. Instead, we will simply repeat a sentence we wrote in the conclusion of our thesis: "it would be useless to attempt to group them (these processes) and classify them except that certain components of the morbid system reappear in a very
large number of varieties of congenital dermatoses, and by their symptomatic significance these primary lesions obliterate the secondary symptoms grouped around them. This is how the terms generally allowed today have gradually been formed through the subdivision of congenital lesions, and it is for this reason also that we have been able to find terms that permit the creation of new groups and attempt their classification.” Méneau, however, has specifically searched for reasons to combine everything and differentiate nothing, and has easily found them through a failure to recognize that the laws in pathology are such that one can only establish a morbid type by choosing the cardinal symptoms and grouping the secondary, always variable symptoms around them. Instead, he gave all symptoms the same priority; by this very simple process, we could establish a single morbid state for all human diseases.

After saying this, it is hardly necessary to dwell on the second sentence in Méneau's conclusions cited above. In fact, we have just seen that the cases he regards as links between ichthyosis vulgaris and ichthyosis fetalis cannot be related to either of these dermatoses; they constitute distinct morbid species in which two new forms can be distinguished: Brocq's congenital ichthyosiform erythroderma and the lamellar exfoliation of the newborn. It is therefore impossible for us to side with the opinion of Méneau, and although it is true to say that there exist transitional cases between ichthyosis vulgaris and ichthyosis fetalis, these must be found in other observations. In reality, there are many fewer relations between these two latter dermatoses than there are between ichthyosis fetalis, lamellar exfoliation, and congenital ichthyosiform erythrodermas, whether or not they are associated with other congenital syndromes, and this relationship is explained by the point in the embryonic and fetal development where these later types begin to appear, which in most cases is immediately or almost immediately after birth.

We therefore conclude from what we have just said that it is possible to distinguish independent varieties in the group of congenital dyskeratosis; that each of the varieties is characterized by a group of special symptoms which should serve as the basis for the classification; and that all these varieties are recognized by the majority of authors although there is no agreement on the name that should be attributed to them.
Based on current scientific knowledge, special classifications for the symptomatology of these clinical pictures should be accepted, corresponding to: 1) isolated lesions on the skin appendages; 2) lamellar exfoliation of the newborn; 3) ichthyosis fetalis; 4) symmetrical hereditary palmoplantar keratodermas; 5) Brocq's congenital ichthyosiform erythrodermas; 6) ichthyosis vulgaris; 7) congenital bullous lesions; 8) partial or generalized atrophies.

One should also remember that these morbid species easily associate with each other, but even when grouped they are still recognizable through meticulous analysis, and then constitute complex morbid clinical pictures, as are so often seen in dermatology. From an association of symptoms and syndromes, one should not conclude that there is a confusion of the types; nature does not move in huge jumps. A study of the facts leads us to believe that there exists a chain of continuous links, but these links are arranged into groups; the chain is not equally developed at all points, and the links that constitute the groups are not each of the same nature.