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Neonatal Skin Disorders: A Review of Selected Dermatologic Abnormalities

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Abstract

The skin serves many purposes, acting as a barrier to infection, protecting internal organs, contributing to temperature regulation, storing insulating fats, excreting electrolytes and water, and providing tactile sensory input. This article focuses on a review of normal skin structure and function and selected neonatal skin disorders. The disorders reviewed are Staphylococcal scalded skin syndrome, epidermolysis bullosa, and the ichthyoses. The basis for each skin disorder is presented. Nursing management and skin care are incorporated into the review of each selected disorder.

THE SKIN is a major body organ for the newborn and critical for the infant's health and well being. Although the skin of an adult is only 3% of its body weight, the skin of a newborn may be up to 13% of its body weight, especially in the preterm neonate. At birth, assessment of the skin is used, in conjunction with other findings, to determine physical maturity. The skin serves many purposes. The skin is a barrier to infection, protects the internal organs, contributes to temperature regulation, excretes electrolytes and water, and provides tactile sensory input. Alterations in the infant's skin can lead to multisystem problems. This article reviews skin structure, function, and physiology and selected neonatal dermatologic disorders. This information will be used as a basis for describing nursing management of these specific disorders.

EMBRYOLOGIC DEVELOPMENT OF SKIN

The skin is made up of two morphologically different layers (epithelial and mesenchymal structures), which are derived from two different germ layers. The epithelial structures are the epidermis, pilose-baceous-apocrine unit, eccrine unit, and nails. These structures are derived from the ectoderm. The ectoderm also generates the hair, teeth, and the organs for the sensations of smell, taste, hearing, vision, and touch. Collagen, reticular and elastic fibers, blood vessels, muscles, and fat are termed mesenchymal structures. These originate from the mesoderm.

The embryologic development of the skin is summarized in the box.

SKIN STRUCTURE

Human skin is composed of three layers: the epidermis, the dermis, and the subcutaneous tissue. Two types of cells make up the epidermis: keratinocytes and dendritic cells. Keratinocytes possess intercellular bridges and stainable
amounts of cytoplasm. As the keratinocytes differentiate, they are arranged into five compact layers: the basal cell layer or stratum basalis, also known as the stratum germinativum; the squamous cell layer, also known as the prickle-cell layer or stratum spinosum; the granular layer or stratum granulosum; the horny layer or stratum corneum; and the clear layer or stratum lucidum (Fig 2). The terms stratum malpighii and rete malpighii are applied to the basal, squamous, and granular cells. These lower three layers have nucleated, viable epidermis.

The functional superficial layer of the epidermis is the stratum corneum. This layer is responsible for barrier properties of the skin and is primarily composed of closely packed dead cells that are continually being exfoliated by contact. In utero, these exfoliated cells form part of the vernix caseosa that covers and protects fetal skin. The basal cell layer constantly replaces these cells. Full-cell migration from the basal (bottom) layer takes place.

The cells of the squamous cell layer are polygonal in shape and gradually flatten during immigration to the surface. The cells of the granular layer are flat and filled with granules. These cells have lost all their intercellular components, such as nuclei, Golgi apparatus, ribosomes, and mitochondria. The cells of the horny layer grow in orderly vertical stacks. The cell membranes of each cell are firmly attached to adjacent cells. The clear layer is a thin, homogeneous, and slightly eosinophilic-like area. This layer is best seen where the horny layer is thick, such as the soles of the feet.

The basal cell layer is made up of two different types of cells: the keratinocytes and the melanocytes. The keratinocytes are at the bottom of the epidermis. These cells are columnar in shape with their long axes perpendicular to the surface. The keratinocytes are attached to the dermis by intercellular bridges known as desmosomes. Keratin is produced by these keratinocytes by 17 weeks' gestation. Gradually, the keratin-forming cells migrate to the outer layer. These cells become flattened and dehydrated during the migration where they form the tough impermeable membrane of the stratum corneum. Melanocytes begin producing pigment before birth and distribute it to the epidermal cells. Melanin is the main skin pigment and is produced by melanin cells in the lower levels of the epidermis. The cells of the basal layer are the only cells of the epidermis capable of cell division.

The dermis is beneath the epidermis and consists of a dense vascular bed, connective tissue, and lymphatic channels. The function of the dermis is to support the epidermis. The connective tissue is composed of collagen and elastin fibers. The dense vascular bed of blood vessels nourish the skin cells. Nerves provide the sensation of heat, touch, pressure, and pain from the skin to the brain. Epidermal ridges extend from the developing dermis resulting from the proliferation of cells within the basal cell layer. These ridges are established by the 17th week of gestation and produce the ridges and grooves on the surface of the palms and fingers and the soles of the feet and toes. The presence of abnormal chromosomes affects the development of the ridge patterns. For example, infants with Down syndrome exhibit distinctive hand and feet patterns that are of diagnostic value.

The major component of the subcutaneous layer is fatty connective tissue. This layer functions as a heat insulator, shock absorber, and fuel reservoir. Subcutaneous fat first appears around the 14th week of gestation. Fat storage achieves significant accretion near term.

Sebaceous glands and sweat glands are located in the dermis and the subcutaneous layer. Even though sebaceous glands are well developed and functional at delivery, these glands have minimal function until puberty. Sweat glands are affected by environmental temperature. In term infants, sweat gland maturation occurs about 5 days of postnatal age. However in the premature infant, this maturation occurs between 21 and 33 days after birth. Adult function does not occur until the second to third year of life.

The remainder of this article focuses on a review of neonatal skin disorders. The disorders reviewed are Staphylococcal scalded skin syndrome, epidermolysis bullosa, and the ichthyoses. The basis for each skin disorder will be presented. Nursing management and skin care will be incorporated into the review of each selected disorder.

**NEONATAL SKIN DISORDERS**
Staphylococcal Scalded Skin Syndrome

Staphylococcal scalded skin syndrome (SSSS) is an uncommon skin infection, caused by *Staphylococcus aureus*, that is usually seen in full-term neonates. Clinical presentation begins with a tender, diffusely erythematous rash that quickly develops into clusters of bullous eruptions, or fluid-filled blisters, that enlarge and then progress to generalized epidermal sloughing. Outbreaks of SSSS have frequently been traced to asymptomatic, colonized health care workers in well-baby nurseries or delivery rooms. Several studies have reported that from 10% to 30% of healthy adults are colonized with *S. aureus*, with higher rates for health care workers.

A relatively immature renal function and subsequent reduced ability to clear bacterial exotoxins are thought to be the major predisposing factors to SSSS in the neonatal period. *S. aureus*, growing on the skin of affected infants, produces exfoliative (epidermastic) endotoxins, which are then absorbed, causing infection. The initial sites of *S. aureus* colonization are commonly the anterior nares, nasopharynx, conjunctiva, and the umbilicus of affected infants. In the neonate, SSSS lesions are generally found in the perineum or periumbilical areas. Infection begins with a generalized erythema, skin tenderness, and fever, followed by the formation of large fluid-filled bullae. Bullae form by 12–14 hours of initial symptoms, then rupture, leaving large areas of red, moist, denuded, “scalded” skin. The severity of the sloughing will vary, depending on the delivery of toxins to the skin. An infant's entire epidermis may be sloughed during the course of an SSSS infection. No residual scarring or permanent skin damage has ever been reported or should be expected to occur.

During a SSSS infection, the attachment between the stratum corneum and the underlying epidermis becomes weak with intraepidermal stripping through the granular layer of the skin. Skin biopsy, if done, reveals a cleavage between the stratum spinosum and the stratum granulosum layers of the skin. Very few, if any, inflammatory cells are present, and no epidermal necrosis typically occurs. The toxins most commonly responsible for the changes in skin integrity seen in SSSS are exfoliative toxins that are produced mostly by phage group type II strains of *S. aureus*.

Two forms of exfoliative toxin have been identified. These are exfoliative toxin A (ETA) and exfoliative toxin B (ETB). ETA is chromosomally encoded, whereas ETB is plasma encoded. Most cases of SSSS in neonates are associated with ETA. These two exfoliative toxins have been described as acting like superantigens by stimulating large numbers of T cells to release lymphokines, such as interleukin 2, into circulation. The toxins are known to act specifically in the zona granulosa of the epidermis. To date, however, there has been little progress made in understanding these toxins. Their role in SSSS was first reported back in 1970; still their specific mechanism of action remains unknown.

Although SSSS was first recognized in 1870, there has been little progress over the years in management or prevention, and the mortality rate remains at 3%. Infants with SSSS are at risk for temperature instability, extensive fluid losses, secondary infections, and sepsis. SSSS is easily and quickly treated with the appropriate systemic antibiotics. Because the bullae seldom contain *S. aureus*, cultures of the nares, conjunctiva, and skin surrounding the blisters is necessary to identify the causative organism and to obtain appropriate sensitivities. Treatment with betalactamase–resistant penicillin is usually effective. Supportive measures to reduce fluid and electrolyte losses and to ensure comfort are also important treatment approaches. Application of topical antibiotic ointments is usually not necessary. Keeping open areas clean and providing appropriate wound care may prevent secondary infections and promote healing in SSSS. To avoid friction on denuded areas of skin from clothing or bedding, infants may be more comfortable if cared for in an incubator. Use of an incubator would also help to decrease insensible water losses through the excoriated skin. Parents should be supported through the sloughing stage of SSSS. Nursery staff must adhere to strict aseptic techniques with gloves worn for wound care; the importance of appropriate handwashing cannot be overemphasized. The use of hexachlorophene products for handwashing has been reported to be effective in eradicating *S. aureus* in the nursery. Others have suggested that all nursery staff should be cultured and treated if found to be carriers of *S. aureus*. 5–7,10,11

**Epidermolysis Bullosa**

Epidermolysis bullosa (EB) is a diverse group of inherited disorders in which the skin and mucous membranes are characterized by blistering caused by extremely, abnormally fragile skin. For infants with severe forms of EB, even the slightest touch may cause widespread and painful blistering of the skin, which can be fatal. For others with less severe forms of EB, there may be an occasional blister with little effect on their lifestyle. The exact incidence of EB is unknown; however, it is estimated that more than 100,000 Americans, mostly children, suffer from EB. It is estimated that one of every 50,000 infants is born with EB. The fatality rate is high.
Classification of EB.

The classification of EB is based on clinical characteristics, the pattern of inheritance, and the ultrastructure level of blistering within the skin as visualized with electron microscopy. Fig 3 demonstrates the normal ultrastructure of the cutaneous basement membrane zone and the level of split of the major forms of EB. Located below the basal cell plasma membrane is the lamina lucida, an electron-lucent zone containing bullous pemphigoid antigen and laminin. Just below the lamina lucida is the lamina densa, an electron dense line, which contains type IV collagen. Below this is the dermis, which contains the anchoring fibrils, comprised mainly of type VII collagen and which ensure cohesion between the epidermis and dermis.

Determining the histologic features, such as the location of type IV collagen, bullous pemphigoid antigen, and laminin on the floor or roof of a blister defines the three main groups or forms of EB. The three forms are simplex—within the basal cells of the epidermis; junctional—within the lamina lucida of the basement membrane zone; and dystrophic—beneath the lamina densa of the basement membrane (Table 1). A positive family history may be present. However, many families will have a negative history with infants with a dominant form caused by a new mutation or infants with recessive types who are born to unaffected parents carrying the gene. Table 2 summarizes the types of EB recognized by the Subcommittee on Diagnosis and Classification of the National EB Registry.

Genetic counseling should always be recommended to families with EB. Genetic counselors can explain the risks and alternatives, answer questions, and offer support and guidance. Each of the forms of EB is different, and the type of EB in the family determines the medical problems that EB will cause and the risk to future generations.

Prenatal Diagnosis.

Prenatal diagnosis of EB is possible for all major forms of EB. Until 1995, the prenatal diagnosis was done on skin specimens obtained by fetoscopy from the fetus at risk between 18 to 20 weeks' gestation. However, DNA-based prenatal diagnosis is now possible for recessive dystrophic and junctional EB. This method is more accurate than direct examination of fetal skin, is less invasive, and can be done during the first trimester of pregnancy.

Extremes of EB.

EB simplex is the most superficial type of EB. Blistering occurs in the epidermis usually at the level of the basal layer of the epidermis and therefore does not scar. The molecular defects of EB simplex have been localized to the keratin filaments, K14, which is located on chromosome 17 and K5, which is located on chromosome 12. The keratins are expressed mainly in the basal cells of the epidermis. The usual extent of the blistering is mild. Mucosa involvement is uncommon. Most infants with EB simplex will enjoy a normal life span, with recurrent blistering being an annoying problem.

Generalized EB simplex of Koebner variant is an inherited autosomal dominant trait. This form of EB is present at birth or in early infancy. Blisters arise over pressure points, such as the elbows and knees. During infancy, mucous membrane involvement will occur. Nails may be lost but usually grow back. The prognosis is good and blistering may decrease with age.

Another form of EB simplex is the Dowing-Meara variant. This autosomal dominant variation of EB simplex is characterized by extensive, generalized, grouped blistering during the early years of life, including the neonatal time period.

The most common form of EB simplex is the Weber-Cockayne (autosomal dominant) variant. This form is one of the most easily recognized forms of EB. Blisters occur mainly on the hands and feet. This form does not usually become apparent during the neonatal period. As infants become older, foot blisters may make it difficult to bear weight. Consultation with a podiatrist is useful for identification of shoes that even
out the pressure on the sole yet ensure a snug fit. Occasionally, these infants will develop blisters in the mouth, but these generally heal quickly and seldom become a problem. 13,14

Junctional EB is an autosomal recessive form of EB. This group is characterized by blistering formation through the lamina lucida at a point between the bullous pemphigoid antigen and laminin. Molecular defects have been recognized within several proteins that make up the anchoring filaments, such as kalinin, epiligrin, nicein, and BM 600. The most serious form of junctional EB is called junctional EB of Herlitz. This type is also known as EB gravis or letalis EB because many of these infants are born with extensive blistering and may not survive beyond infancy as a result of sepsis and dehydration. Blistering is readily noted at birth. The most serious complication is gastric outlet obstruction. Infants with this form of junctional EB exhibit a large spectrum of severity. Blistering and erosions occur in the scalp, in the perioral area, and over pressure points on the body. The hands and feet are typically spared. As a result, digital fusion does not occur. Nails, however, are affected and may be lost permanently. In addition, there is defective dentition and laryngeal involvement manifested as hoarseness or stridor. These infants grow poorly. The analysis of the skin reveals that the anchoring filaments responsible for attachment of the basal cells to the basement membrane are reduced in number and abnormally structured because of the defect in the protein, kalinin. 13,14

Dystrophic EB occurs in both recessive and dominant forms. Both groups are characterized by blister formation below the lamina densa of the basement membrane. The anchoring fibrils that link the lower part of the basement membrane to the papillary dermis are comprised of Type VII collagen. This group of EB is clinically identified by milia and scarring at the sites of healed blisters. All forms are present at birth.

Most infants with a recessive dystrophic EB form have extensive blistering from birth. The most severe recessive dystrophic EB type is the Hallopeau-Siemens. Infants with this form of EB may have extensive denuded lesions at birth and during the neonatal period. Over time, the blistering formation and healing results in mitten like scars that encase the fingers and toes by late childhood. Within these mitten deformities, individual phalanges are present. Surgical separation of the fingers can be attempted. However, rescarring occurs within a few years of the surgery. Exracutaneous involvement is common and may be severe. Blistering of the mouth and esophagus results in esophageal strictures and malnutrition because of the restriction of oral intake. The blistering formations are subepidermal. There is a decrease or complete absence of anchoring fibrils associated with markedly degeneration of Type VII collagen in the papillary portion of the dermis. In addition, fibroblast cultures reveal an excess in abnormal collagenase. 13,14

Dominant dystrophic EB is less common and milder form than recessive dystrophic EB. The majority of the localized forms of dystrophic EB are autosomal dominantly inherited. Blistering may occur in only specific areas and may become less pronounced with increasing age in the localized forms. The two generalized forms of dominant dystrophic EB are Cockayne-Touraine and Paisini. Both of these forms are significantly less severe than the recessive dystrophic EB. The blistering is subepidermal and healing is with scarring, but the process is limited, usually to the hands, feet, and skin over bony areas or can be generalized especially with Paisini. Mitten deformities of the hands and feet do not occur; however, nails may be lost. 13,14

Diagnosis.

The distribution of the blisters is diagnostic for the type of EB. Early blisters occur at points of friction. The fluid within the blister will be clear or hemorrhagic but generally not purulent. Skin biopsy for immunofluorescent mapping and ultrastructural study is vital. The sample for immunofluorescence should be obtained from normal skin or perilesional skin. The ideal specimen for electron microscopy is a new spontaneous or induced blister. After the diagnosis of simplex, junctional, or dystrophic EB is made, further classification can be difficult in the neonatal time period. Thus, a careful in-depth family history for blistering should be obtained. Without a relevant family history, distinguishing features may take years to develop.

Nursing Management.

The primary goal of therapy is supportive. Wound care is a major challenge facing the newborn and the family on a daily basis. A program of cutaneous hygiene is necessary to minimize the risk of scarring and infection. Any dressing change must minimize further damage or trauma to the skin and should occur after adequate pain control medication has been administered. Dressings should be soaked off, never peeled or forcibly removed. Gloves should be lubricated to minimize friction with the skin. Infants should be lifted by placing one hand under the buttocks, while the other hand supports the back. After bathing, the skin should be gently pat dried with a soft towel or blow-dried with a hair
As scarring occurs, contractures may form. The abnormal increase in elastic skin fiber facilitates this occurrence. Gentle passive range of motion by the caregivers may decrease the contracture formation.

Another problem is nutrition because the high almost continual sloughing of epithelium tissue results in large fluid, protein, and electrolyte losses. Feedings should be done slowly and carefully to avoid aspiration and trauma. For newborns with mucous membrane involvement, tools used to feed a newborn with cleft palates can be useful and less traumatic. Also, the addition of extra holes into the nipple may allow for easier feeding. Gavage feedings are strongly discouraged. Iron deficiency is a common complication in infants with severe disease. Aggressive nutritional supplementation is routine.

Parents and caregivers should be given anticipatory guidance about protective measures, wound care, and nutrition. The Dystrophic Epidermolysis Bullosa Research Association of America (DebRA) is a national not-for-profit organization that provides services and support for people with EB and their families. DebRA can be reached at (212) 513–4090, http://www.debra.org, or via email at staff@debra.org.

Ichthyosis.

The ichthyoses are a heterogenous group of disorders that share one common characteristic: scaly skin. There are differences ranging from focal to generalized scaling, from spiny to flat, or small to huge scales, and from blistering to redness beneath the scales. The term ichthyosis has been in use for well over 200 years. The word root is from the ancient Greek word *ichthy*, which means fish. There are four major types of ichthyosis: (1) X-linked ichthyosis, (2) lamellar ichthyosis, and (3) epidermolytic hyperkeratosis, which presents at birth, and (4) ichthyosis vulgaris, which commonly occurs around the third month of life (Table 3). Other well-known terms that are commonly used to describe an infant with ichthyosis include collodion baby and harlequin fetus. These descriptive terms inadequately identify the type of ichthyosis that may be present. 5,19

Ichthyoses are basically abnormalities in the formation and desquamation of the keratinocytes in the epidermis. Keratinocytes, in the basal layer, produce keratins, the principal structural proteins of the outer stratum corneum. The basal layer divides approximately every 19 days. Once the cells leave the basal layer, their transit time to the granular layer is approximately 14 days, followed by another 14 days for transit from the granular layer to desquamation. As the cells migrate upward, they produce keratins, lose their cellular components, flatten out, and eventually are shed. A disturbance in any of these processes will result in defective desquamation. Retention or hyperproliferation of squamous cells form the basis for the ichthyoses (Fig 4). With retention ichthyoses (ichthyosis vulgaris, recessive X-linked ichthyosis, and lamellar ichthyosis), epidermal homeostasis is normal, but the desquamation process is defective, resulting in visible scales being shed in clumps. In the hyperproliferative ichthyosis (bullous congenital ichthyosiform erythroderma), both epidermal homeostasis and the desquamation process are defective. 18–20
Ichthyosis Vulgaris.

Ichthyosis vulgaris (IV) is the most common form of the ichthyoses. Literally meaning fish-like scaly skin, IV occurs in approximately 1:250 to 1:500 infants. IV is an autosomal dominant inherited condition. Clinically, IV may be expressed along a continuum, with generalized dry skin at one end and full-blown lizard-like scaling with erythema and itching at the other. This variability may be seen within the same family. In IV, an infant's skin looks and feels normal at birth but gradually becomes dry and begins to feel rough. The scales are fine and irregularly shaped, often appearing white. The trunk and face are generally spared. Most scaling appears on the extremities, primarily on the calves. Diaper and flexor areas are usually not affected, most likely owing to the moist nature of these areas. Palms and soles may show hyperlinearity and hyperkeratosis. Fissures are common on the fingers, especially during dry weather. Hair, teeth, and nails are normal. Keratosis pilaris, spiny, flesh-colored to red, perifollicular papules, may be present on the cheeks, neck, buttocks, or thighs. As with any genetically inherited condition, IV is controllable, but it is not curable. IV varies with environmental temperature and humidity. Symptoms may lessen during summer months or when living in warm, moist climates. Diagnostically, skin biopsy reveals a mildly thickened stratum corneum and a diminished or absent granular layer.

Recessive X-linked Ichthyosis.

During the 1960s, recessive X-linked ichthyosis (RXLI) was identified as a distinct form of the ichthyoses, separate from ichthyosis vulgaris. As the name implies, this uncommon form of ichthyosis is a sex-linked condition. This form is caused by a deficiency of the enzyme steroid sulfatase, which is carried on the X chromosome. Ninety percent of cases are caused by a deletion of the steroid sulfatase gene. The incidence is approximately 1:2,000 to 1:6,000 in male infants. These infants display scaling at birth in 17% of cases, with another 84% developing scaling within the first 3 months of age. There are no reported incidents of the onset of RXLI after the first year of life.

Scales adhere tightly to the underlying epidermis. They appear small, are light brown in color, and polygonal in shape. Both extensor and flexor surfaces of the extremities are involved, making differentiation from IV clinically possible. Because of their moist and more humid nature, the flexor surfaces may be less affected than extensor surfaces. Scales are almost always present on the sides of the neck and the lower abdomen. Parents often report being unable to “get the baby clean.” The central face, scalp, palms, soles, hair, teeth, nails, and mucous membranes are spared. Keratosis pilaris is not seen. Cryptorchidism may be seen in as many as 25% of affected infants. Although RXLI is a lifelong condition, it is not debilitating and normal life activities should not be adversely affected. As with IV, there may be some seasonal improvement in symptoms, and warm, wet climates can often alleviate the condition. Diagnostically, a decrease or absence of steroid sulfatase can be measured by enzyme assay, serum cholesterol sulfate accumulation, or fluorescent in situ hybridization (FISH) testing. Microscopically, a thickened but otherwise normal-looking stratum corneum with a normal to thickened granular epidermal layer may be seen.

Epidermolytic Hyperkeratosis.

Epidermolytic hyperkeratosis (EH), formerly known as bullous congenital ichthyosiform erythroderma, is caused by an inborn error in keratin synthesis. As such, EH may be more appropriately identified as a keratinopathy. This rare form of the ichthyoses occurs in 1:300,000 infants. EH is an autosomal dominant disorder caused by an error in the synthesis of keratins 1 and 10. These keratins are found in the upper spinous to granular layers of the epidermis. EH may be expressed differently, depending on the penetrance or expression of the gene disorder and on which keratin is affected. For example, defects in keratin 1 synthesis will result in bullae formation on the palms and soles. In EH, bullae may be formed for one of two reasons. One, there is an abnormal keratin filament to desmosome formation that results in an abnormal structure and epidermal fragility, and two, desmosomal attachments are defective, resulting in blister formation. Clinically, infants with EH present with red scaly lesions and widespread areas of denuded skin. Blisters are superficial and, as such, do not leave scarring. Hyperkeratosis may be present but can be subtle. As the infant matures, bullae formation decreases (but does not cease entirely) and hyperkeratosis becomes more prevalent, with large, thick, dark scale formation. These scales are characteristically porcupine-quill like in nature. They involve the entire body and are especially prominent in flexor areas and on the face and scalp. If affected, the palms and soles will become thickened and waxy. The scaling in EH causes the formation of furrowed ridges of spiny skin, hence, the porcupine-quill like appearance. After the neonatal period, bullae form underneath the quills, frequently becoming infected, resulting in generalized epidermal sloughing. The underlying skin is red and temporarily normal looking—until new quills are formed. Infected areas contain both aerobic and anaerobic bacteria. These bacteria produce a
foul smelling odor that is clinically characteristic of EH. Again, and as with all the ichthyoses, EH is a lifelong condition. Life expectancy is normal, however. Blister formation and foul smelling body odors are chronic. The body odor may be the most distressing aspect of EH, as well as the most difficult symptom to control. Normal sweating mechanisms can be disrupted, leading to heat intolerance in warmer climates. Diagnostically, skin biopsy reveals a distinctive, thickened epidermis with areas of cellular breakdown. The granular layer is also markedly thickened, and irregular keratinocytes are clearly visible. 18,19

**Lamellar Ichthyosis and Congenital Ichthyosiform Erythroderma.**

Lamellar ichthyosis (LI) and congenital ichthyosiform erythroderma (CIE) are the most severe forms of ichthyosis. They are also the most recognizable, owing to the alligator-like appearance in the skin of affected infants. Also known collectively as autosomal recessive primary ichthyoses, both are autosomal recessive inherited disorders. They are rare, each occurring in approximately 1:100,000 to 1:300,000 infants. 18,19 They share an overlapping phenotype. The most severe phenotype is displayed in LI. LI is caused by mutations in the transglutaminase 1 gene. Transglutaminases are a large group of enzymes that catalyze transamidation of glutamine residues. This process is essential for desquamation, formation of hair follicles, and keratinization of the epidermis. CIE, the less severe phenotype, is thought to be caused by a defect of the lamellar granules. The lamellar granules (Odland bodies) house the transglutaminases and are responsible for organizing the enzymes and lipids that are excreted by the keratinocytes. The lipids are important in providing a barrier to water loss, and the enzymes facilitate desquamation. In CIE, lamellar granules are present but irregular. This irregularity is thought to interfere with normal enzymatic function. 18,19

Clinically, infants with LI and CIE present at birth with red, scaly skin. Most are born encased in a collodion membrane (collodion baby). This membrane is taut and shiny, with varying degrees of thickness and often temporarily distorts facial features. Ears may appear crumpled, and the eyelids (ectropion) and the lips (eclabium) may be inverted. The digits may be fixed and immobile. Varying degrees of nail dystrophy may be present. The surface of the skin is shiny, yellow to brown in color, and may resemble oiled parchment paper. Peeling and fissuring begins shortly after birth. The collodion membrane is generally shed by 10 to 14 days of life, revealing reddened and erythematous skin underneath. After the collodion membrane is shed, the scaling of LI or CIE is clinically noticeable. In LI, the scales are large, dark, polygonal in shape, platelike, and tightly bound to the epidermis. The entire body surface is involved, including the face, scalp, flexor areas, palms, and the soles. The eyelids and the mouth may remain inverted, even after the collodion membrane is shed. Hair is matted and sparse. Because the thickened skin lacks elasticity, fissures may develop over joints. In CIE, there are platelike scales on the extensor areas of the lower extremities, and fine, white scales covering the face, scalp, trunk, and flexor areas. Inverted eyelids and mouth resolve with the shedding of the collodion membrane. Hair is less effected, but scarring alopecia may develop. Nails, teeth, and mucous membranes are not usually affected. 18–22

The prognosis for LI and CIE is poor. Both conditions are lifelong, with continual new scale formation. The ability to sweat may be severely impaired, causing marked heat intolerance. The cracks between the lamellar plates of the scales are at high risk of becoming infected. Nutritional requirements are high and, if not adequately met, may result in failure to thrive. Histopathologically, LI shows a markedly thickened stratum corneum. CIE shows only mild thickening. 18–22

**Management.**

The primary goal of therapy of the ichthyoses is aggressive hydration of the skin. Hydrating the skin and locking in moisture are the therapeutic goals of treatment. Adequately hydrating the stratum corneum increases pliability and comfort. Hydrating also increases desquamation by facilitating hydrolytic enzyme activity within the epidermis. Simple soaking in warm water, twice a day, followed immediately by the application of a thick, greasy moisturizer to damp skin is beneficial for most infants with ichthyosis. More frequent baths may be beneficial, as long as the moisture is sealed in with afterbath ointments. Bathing without moisturizing may worsen symptoms. The temperature of the bath has little to nothing to do with moisturizing; thus, any comfortable temperature is acceptable. Likewise, length of bathing does not effect the moisture content of the skin. The stratum corneum soaks up as much water as is possible within 10 minutes. Because soap can strip the skin of protective fats, it is not always necessary. Adequate cleansing can occur with plain water. Moisturizers that are available in jars and that need to be scooped out work best. Aquaphor, Plastibase, and petrolatum jelly are examples of recommended moisturizers. As previously noted, warm, moist climates do help, as does humidifying the home. 5,18,19

Infants who display bullae can be at high risk for fluid and electrolyte imbalances, temperature instability, and sepsis. Careful, gentle handling
is necessary to avoid pain and discomfort and to prevent further blister formation. As with EB, therapy goals for an infant with an ichthyosis that displays blisters are mainly supportive. The same treatment approaches that are used with EB can be applied to ichthyosis. Humidified incubators offer hydration and comfort. They also provide increased control of insensible water losses through the denuded areas of the infant's skin and temperature support. Pain management, including nonsteroidal anti-inflammatory agents and narcotics, may be necessary to provide comfort. Preventing and treating possible infection is a top priority. Systemic antibiotics may be necessary. The use of topical antibiotic ointments or antibacterial soaps are not generally recommended except in cases of EH. With the overgrowth of bacteria that is characteristic of EH, it may be necessary to use antibacterial soap when bathing. The addition of a small amount of bleach or betadine (povidone iodine) to the bath water may help reduce the bacteria present on the skin. The use of essential oils in the bath water may help to alleviate the persistently foul-smelling body odor. 5,18,19

**Alternative Treatment Options.**

Alpha-hydroxy acids, urea based creams, and propylene glycol can also be used for hydrating the skin. Topical alpha-hydroxy acids (10% glycolic acid lotions) may be helpful. These simple, naturally occurring acids work well in hydrating the skin by interrupting the formation of corneocytes in newly formed layers of the stratum corneum. They provide moisture to the skin at the same time that they encourage desquamation. Used twice a day, these lotions have been shown to be more effective than petrolatum-based ointments in reducing the severity of symptoms in IV, RXLI, and EH. These acid-based lotions can cause pain, and infants may not tolerate their use. Application of acid-based lotions is not currently recommended in the neonatal period. Because the neonate has a high surface area-to-body weight or volume ratio, absorption of total body applications of acid-based creams can cause systemic acidosis. 18,19,22

Keratolytics, such as salicylic acid (6% gel), induce desquamation and as such may be helpful in removing scaling in IV, RXLI, and LI. Urea-containing creams (10%–20%) act as humectants, increasing pliability of the stratum corneum. Propylene glycol (50%–60%) creams work well by establishing a water gradient that pulls water through the dry stratum corneum. The stratum corneum subsequently swells, closing painful fissures. These creams may be applied to the skin, then covered with another thicker moisturizer to help hold in moisture. Again, the increased skin-to-body weight ratio of the neonate makes use of these products contraindicated in the first few months of life. To avoid salicylism, it is recommended that salicylic acid products, if used at all, be kept to a maximum of twice a day. Elevated plasma urea concentrations have been reported during usage of a 10% solution with a collodion baby with LI. Plasma urea concentrations returned to normal once their use was discontinued. 18,19,22

Perhaps the most likely successful treatment for ichthyosis may come from the use of retinoids. Retinoids are a large group of vitamin A derivatives that have a variety of effects on the skin. Use of topical tretinoin (Retin-A) has resulted in some improvement in the scaling of ichthyosis by stimulating desquamation. Limitations, however, include the large amount of skin needing treatment, the cost, and the potential for systemic absorption. Synthetic retinoids for oral use include isoretinoin (Accutane). The mechanism of action is still not clearly understood. What is known, however, is that isoretinoin thins the epidermis, increases circulating lipids, and has been shown to decrease overall scaling, the thickness of platelike scales, and erythema in EH and LI. Treatment must be continued at high doses and for indefinite periods of time, making the safety of such an approach questionable at best. The side effects of long-term oral retinoid use and teratogenic risks prohibit their use at this time. 18,19,22

**Genetic Counseling and Prenatal Diagnosis.**

In all cases of ichthyosis, a diagnosis is the first step in genetic counseling. With a definitive diagnosis, accurate information may be provided to the family. Any family who has given birth to an infant with ichthyosis will want to understand their infant's prognosis and the pattern of inheritance in their family. The variability expressed among the phenotypes of the ichthyoses, and even among family members with the same condition, may be confusing to the family. A detailed family history including affected and nonaffected individuals is important. The genetic pattern of inheritance of ichthyosis and the implications for future pregnancies need to be addressed.

Despite recent advances in the area of dermatology, prenatal diagnosis of ichthyosis is still not possible. Currently, fetal skin tissue sampling is not helpful, because the skin does not keratinize until late in the third trimester, and both false–positive and false–negative skin biopsy results are common. Additionally, while in the fluid filled environment of the uterus, the fetal epidermis of many of the ichthyoses appears normal.

The sight of an infant with ichthyosis can be frightening. Parents will need to be provided with interdisciplinary emotional support, as well as
anticipatory guidance. Ichthyosis is a lifelong condition that never goes away. There is no cure. The effect of such a disorder on the family cannot be underestimated. Psychologic stresses may be as important as the physical stresses that are involved with dealing with such a chronic situation. Additional support may be offered from the Foundation for Ichthyosis and Related Skin Types (FIRST) at 1–800–595–1265, (206) 616–3179, or via email at ichreg@u.washington.edu.

This article has reviewed basic skin structure and function and the importance of the skin as a major body organ in the neonate. Because the skin offers protection from fluid and electrolyte imbalances, infection and temperature instability disruptions in neonatal skin integrity and function can have significant adverse consequences for the infant. Skin care is a critical component of neonatal care. Neonatal nurses must understand the structure and physiology of neonatal skin to promote skin integrity as well as to provide appropriate care of infants with specific dermatologic disorders, such as staphylococcal scalded skin syndrome, epidermolysis bullosa, and the ichthyoses.

**Embryonic and Fetal Development of Skin**

Weeks of gestation

3 Epidermis, which develops from surface ectoderm, consists of one layer of cells
5 Cutaneous nerves detectable in embryonic dermis
6–7 Periderm, a thin protective layer of flattened cells, is formed
11 Collagen and elastic fibers are developing in the dermis
Epidermal ridges (fingerprints) are forming
Nails begin to develop at the tips of the digits
13–16 Scalp hair patterning determined
17–20 Melanocytes migrate to the epidermal–dermal junction and begin to produce melanin
Skin covered with vernix caseosa and lanugo
21–25 Skin is wrinkled, translucent, and pink to red because blood in the capillaries is visible
26–29 Subcutaneous fat begins to be deposited and starts to smooth out the many wrinkles in the skin. Eccrine sweat glands are anatomically developed and located over the entire body. Their function is immature
30–34 Skin is pink and smooth
Fingernails reach fingertips
Lanugo is shed
35–38 Fetuses are usually round and plump with subcutaneous fat
Skin is white with pinkish hue
Toenails reach toe tips


**REFERENCES**


Key words: *collodion baby; epidermolysis bullosa; ichthyosis; infant; neonate; newborn; skin; skin care; Staphylococcal scalded skin syndrome*

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**IMAGE GALLERY**

[Fig 2](#) [Fig 3](#)