An infant born at 35 weeks' gestation, is transferred to your care. Striking physical examination findings include shiny, tight, thickened skin that is membranelike, with emerging cracks and moist fissures. The facial skin is taut and has pulled the mouth into an O shape, and the eyelids are everted. The hands and feet are edematous and have contractures (Figure 1).

Figure 1: Collodion baby with bilaterally everted eyelids (ectropion) and contracted lip (eclabion). Membrane is cracked in places, revealing moist erythematous skin.

Of the following, the MOST appropriate initial management for this newborn's skin is:

1. application of keratolytic agents
2. application of topical emollients
3. comfort care only, because this condition is lethal
4. manual débridement of the membrane
5. no therapy indicated, because spontaneous resolution expected

You selected 5, the correct answer is 2.
The infant in this vignette presents with features characteristic of the collodion baby. At birth, an oiled-parchment-like membrane of thick hyperkeratotic epidermis encases the infant, with underlying yellowish erythematous skin. Taut facial skin everts the eyelids (ectropion) and fixates the lip in an O shape or fish-mouth configuration (eclabium). Similarly, the nose is flattened, the pinnae are malformed, and hair may be absent or perforate the membrane (Figures 1 and 2).

Figure 1: Collodion baby with bilaterally everted eyelids (ectropion) and contracted lip (eclabium). Membrane is cracked in places, revealing moist erythematous skin.

Circumferential constriction results in peripheral edema and contractures of the hands and feet.

The term collodion baby describes a phenotype common to several disorders of cornification. Nearly two thirds of collodion babies have either classic lamellar ichthyosis or
nonbullose congenital ichthyosiform erythroderma. Less common disorders presenting with a collodion membrane include autosomal dominant lamellar ichthyosis, trichothiodystrophy, recessive X-linked ichthyosis, neonatal Gaucher's disease, neutral lipid storage disease, and Sjögren-Larsson syndrome (Table).

Table. Disorders Presenting With a Collodion Membrane

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Incidence</th>
<th>Inheritance</th>
<th>Mutation</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lamellar ichthyosis</td>
<td>1:300,000</td>
<td>Usually autosomal recessive</td>
<td>Transglutaminase I</td>
<td>Large, platelike scales</td>
</tr>
<tr>
<td>Nonbullous congenital ichthyosiform erythroderma</td>
<td>1:300,000</td>
<td>Autosomal recessive</td>
<td>Lipoxygenases</td>
<td>Fine, white scaling, overlying erythema</td>
</tr>
<tr>
<td>Recessive X-linked ichthyosis</td>
<td>1:6,000 males</td>
<td>X-linked recessive</td>
<td>Steroid sulfatase</td>
<td>Only 17% present at birth; accumulation of cholesterol sulfate may be measured in plasma</td>
</tr>
<tr>
<td>Neutral lipid storage disease (Dorfman-Chanarin syndrome)</td>
<td>Rare, less than 100 described cases</td>
<td>Autosomal recessive</td>
<td>Disruption of recycling of triacylglycerol to diacylglycerol</td>
<td>65% of patients with ichthyosis; hepatomegaly in 46%</td>
</tr>
<tr>
<td>Trichothiodystrophy</td>
<td>Rare</td>
<td>Autosomal recessive</td>
<td>Unknown; decreased DNA repair levels</td>
<td>Lamellar ichthyosis phenotype; decreased sulfur content in hair</td>
</tr>
<tr>
<td>Gaucher's disease</td>
<td>1:100,000</td>
<td>Autosomal recessive</td>
<td>Lysosomal b-glucocerebrosidase</td>
<td>Hepatosplenomegaly and neurologic symptoms</td>
</tr>
<tr>
<td>Sjögren-Larsson syndrome</td>
<td>&lt;1:100,000</td>
<td>Autosomal recessive</td>
<td>Fatty aldehyde dehydrogenase</td>
<td>Only rarely presents as colloidion baby; ichthyosis, mental deficiency, and spasticity</td>
</tr>
</tbody>
</table>
Harlequin ichthyosis, a rare (fewer than 100 described cases) and generally lethal form of congenital skin thickening, does not present with a collodion membrane.

Postnatally, the collodion membrane splits to reveal moist fissures and eventually peels off, though only to re-form in most cases. By several months after birth, the characteristic clinical features of the underlying skin disorder emerge and the diagnosis can be confirmed with skin biopsy. The membrane sheds, with apparently normal skin, in 5% of cases.

The molecular basis for many of the congenital ichthyosiform disorders is known and prenatal diagnosis with analysis of fetal DNA may be possible. In lamellar ichthyosis, mutations in the transglutaminase-1 gene on chromosome 14q11 account for 50% of the cases.

Fluid and electrolyte balance, thermoregulation, nutritional support, and infection prevention are key elements of the initial treatment of the collodion baby. Although markedly thickened, the stratum corneum is a poor barrier and allows excessive transcutaneous fluid and electrolyte loss to occur. The collodion baby has increased metabolic requirements and is at risk for hypernatremic dehydration. The eclabium interferes with the ability to suckle, and intravenous nutritional support is often initially needed. The risk of systemic infection is high and antibiotic coverage should be initiated at first suspicion. Temperature instability is managed with a humidified isolette, with care to avoid hyperpyrexia as well. Initially, pain may be considerable and the use of narcotic agents is indicated.

The shedding of the membrane occurs over the first month, and can be facilitated by the liberal application of topical emollients, which also retard water loss and soften the stratum corneum (Figure 3).

Figure 3: Collodion baby slathered in emollient.

Manual débridement of scales should be avoided. The poor skin barrier of the collodion baby increases the risk of systemic toxicity with topical keratolytic agents such as salicylic acid and tretinoin, and their use in the newborn is discouraged. However, topical keratolytic agents, oral retinoids, and topical
Steroids are important in the long-term treatment of older infants and children. Over time, the ectropion and eclabium resolve, but artificial tears may be indicated to avoid corneal injury.

The long-term prognosis for the collodion baby is influenced by the underlying skin disorder. In the newborn period, with attention to thermoregulation, fluid management, and early treatment of infection, survival is the rule. Classic lamellar ichthyosis persists throughout the affected individual's lifetime as a severe and unremitting scaling skin disorder. Heat intolerance, due to obstructed sweat ducts by plates of scale, and cutaneous infections are common morbidities. Nonbullous congenital ichthyosiform erythroderma is similarly lifelong, but manifests with much milder symptoms.

References:


American Board of Pediatrics Content Specification(s):

Know how to diagnose and manage ichthyosis
A 15-day-old male infant who had been born at 28 weeks’ gestation developed several painful red macules and a hemorrhagic bulla on his right thigh. The macules varied in size from 1.5 to 3.0 cm in diameter. Within 12 hours, the macules vesiculated, some with hemorrhage, and ulcerated (Figures 1 and 2).

The skin surrounding the ulcers was bright pink. The ulcers then developed a necrotic, black, depressed appearance and the centers became encrusted. Culture specimens of the blood, urine, cerebrospinal fluid, and vesicles were obtained and broad-spectrum antibiotics were initiated. A biopsy of the ulcer showed a vasculitis with few inflammatory cells. Edema, hemorrhage, and necrosis were found surrounding the veins and some arteries. Bacteria were seen in the perivascular tissue and vessel walls. As the skin lesions evolved, the infant also developed shock requiring vigorous fluid resuscitation, mechanical ventilation, and vasopressor support.

Of the following, the organism MOST often associated with the skin lesions described for this infant is:

- Candida albicans
Pseudomonas aeruginosa
Staphylococcus aureus
Streptococcus agalactiae
Treponema pallidum

You selected 3, the correct answer is 2.

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The skin of the newborn infant serves as a physical and immunologic barrier to invasion by infectious organisms. During and following birth, the infant is quickly colonized by organisms found on the cervix, vagina, and skin of caregivers. The initial colonization occurs on the umbilicus, genitalia, groin, and abdomen, and spreads rapidly to other surfaces and mucous membranes. The organisms most often found during initial colonization are *Staphylococcus epidermidis*, diphtheroids, and gram-negative enteric organisms. Subsequently, organisms present in the environment, such as *Staphylococcus aureus*, group B streptococci, and other gram-negative microbes, also appear.

The intact skin protects the infant from microbial invasion. The presence of vernix caseosa (shed epithelial cells, sebum, and hair) at birth and stratum corneum physically block movement of microbes into the deeper layers of the skin and blood vessels. Breach of skin barriers, as seen during phlebotomy or placement of intravascular catheters, facilitates microbial penetration.

The skin immune system is composed of cellular components (keratinocytes, antigen-presenting cells, monocytes, macrophages, granulocytes, mast cells, lymphocytes, and endothelial cells) and humeral components (antimicrobial peptides, complement proteins, immunoglobulins, cytokines, and prostaglandins). Langerhans cells are antigen-presenting cells unique to the skin. Microorganisms bind to Langerhans cells and are presented to cellular immune components for processing and killing.

Microbial infections of the skin may be manifest as a maculopapular rash, vesicles, pustules, bullae, abscesses, cellulitis, impetigo, erythema multiforme, and petechiae or purpura. Lesions characteristic of a single organism are unusual because each of the aforementioned manifestations of skin infections can be caused by several different microorganisms. An exception to this generality are the ecthyma gangrenosum lesions found in the infant in the vignette.

Ecthyma gangrenosum occurs in 2% to 6% of infections caused by *Pseudomonas aeruginosa*. Although *Pseudomonas* is the predominant cause for ecthyma gangrenosum, other organisms such as group B streptococci, *Aeromonas hydrophilia*, *Enterobacter* species, *Escherichia coli*, *Proteus* species, *Pseudomonas cepacia*, *Serratia marcescens*, *Xanthomonas maltophilia*, *Aspergillus* species, mucorales, and *Candida albicans* have been infrequently associated with this lesion.

*Pseudomonas* infection often occurs in patients with suppressed immune systems. The organism may be spread hematogenously or by direct inoculation of the skin. As described for the infant in the vignette, the lesion usually begins as a painful red or purpuric macule that centrally vesiculates or becomes pustular. Bullae may also form. Surrounding tissues are pink or violaceous. The lesion quickly ulcerates, develops raised edges, and becomes necrotic in the center. A black, crusted eschar covers an erythematous base that microscopically is characterized by a vasculitis, especially the veins. The ecthyma gangrenosum lesions are caused by production of enzymes and proteases (especially elastase but also gelatinase, collagenase, lecinthinase, neutral and alkaline protease, cytoxin, and phospholipase C), endotoxin, and exotoxins A and S. The lesions are characteristically devoid of inflammatory
cells. *Pseudomonas* organisms may be found in the adventitia and media of the dermal veins. Culture of the base of these lesions, not the exudate or eschar, is necessary to determine the microbial source of infection. The presence of ecthyma gangrenosum generally indicates treatment with anti-*Pseudomonas* antibiotic agents.

Skin lesions associated with *C. albicans* vary in presentation and include diffuse erythema with or without pustules (especially congenital and systemic candidiasis), diffuse maculopapular lesions, cutaneous abscesses at insertion sites of intravascular lines, confluent red and scaly plaques with adjacent red satellite lesions in skin creases of the groin, axilla, and neck, and rarely, as ecchymoses and necrosis (ecthyma gangrenosum). Crusting of lesions and desquamation may follow these presenting lesions. All body surfaces may be involved including the palms, soles, and face in the diffuse skin disorders (ie, congenital and systemic forms) caused by *C. albicans*.

Of note, congenital and local forms of candidiasis can be treated with topical agents alone (nystatin, gentian violet, naftifine, terbinafine). Recurrent lesions may require oral fluconazole or itraconazole. If systemically ill, intravenous treatment with amphotericin B for 14 to 21 days may be needed. Intravenous fluconazole is also effective for *C. albicans* but other *Candida* species are often resistant; amphotericin B is the usual drug of choice for infections caused by non-*C. albicans* species. 5-Fluorocytosine, a pyrimidine antimetabolite that penetrates the blood brain barrier, acts synergistically with amphotericin B against fungal pathogens such as *Cryptococcus* and *Candida* species.

*Staphylococcus aureus* is a frequent cause for skin infections. Superficial infections (such as impetigo and bacterial folliculitis), cutaneous and subcutaneous abscesses (such as breast and scalp abscesses), nonnecrotizing subcutaneous infections (such as funisitis and omphalitis), necrotizing subcutaneous infections (such as necrotizing fasciitis), and toxin-mediated disorders (staphylococcal scalded skin syndrome) may be caused by *S. aureus*. Ecthyma gangrenosum is rarely caused by *S. aureus* and, when associated, it most often presents late in the course of necrotizing fasciitis.

Infection caused by *Streptococcus agalactiae*, or group B streptococcus, is rarely accompanied by skin manifestations. Cellulitis is the most frequently encountered skin disorder, occurs more often with late-onset disease, and has a predilection for facial and submandibular skin. Impetigo, cutaneous and subcutaneous abscesses, erythema nodosum–like lesions, necrotizing fasciitis, and purpura fulminans have been reported with group B streptococcus. Ecthyma gangrenosum is rarely caused by group B streptococcus.

*Treponema pallidum*, the spirochete that causes syphilis, is responsible for a wide variety of skin manifestations in congenitally infected infants. At birth, fewer than half of all infected infants have skin lesions. Lesions include papulosquamous plaques, erythematous macules, hemorrhagic vesicles and bullae (pemphigus syphiliticus), annular lesions, and polymorphous rashes. The palms, plantar surfaces of the feet, perioral skin, and anogenital region are common sites for lesions to be found. Snuffles (syphilitic rhinitis) may be the first sign of congenital syphilis and begins as a clear nasal discharge that becomes profuse, chronic, and hemorrhagic. Papules and plaques at the mucocutaneous junctions of the nose, mouth, and anus are highly infectious (condyloma lata) and, if chronic, lead to rhagades. Bullae resembling the early lesions that develop into ecthyma gangrenosum are infrequently found in infants with congenital syphilis. The bullae form on an erythematous base and after rupturing, maceration occurs, and a crusty exudate appears.

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References:


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**American Board of Pediatrics Content Specification(s):**

Understand the causes and differential diagnosis of infections of the skin and mucous membranes

Understand the clinical and laboratory features of infections of the skin and mucous membranes

Understand the clinical manifestations of group B streptococcal infections

Understand the clinical manifestation and diagnostic criteria of neonatal infections with *Staphylococcus aureus* and *Staphylococcus epidermidis*

Understand the clinical manifestations and diagnostic criteria of perinatal infections with *Treponema pallidum*

Understand the clinical manifestations of neonatal infections with *Candida*, coccidioidomycosis, cryptococcus, histoplasmosis, and *Malassezia*

Recognize the cutaneous and laboratory manifestations of congenital syphilis

Recognize the cutaneous and laboratory manifestations of severe candidiasis

Know the treatment of severe candidiasis

Know the etiology and cutaneous manifestations of nonpurpuric skin lesions