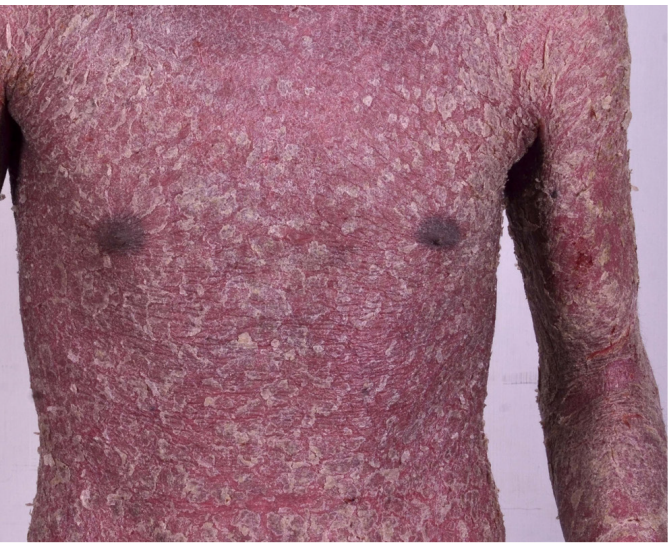
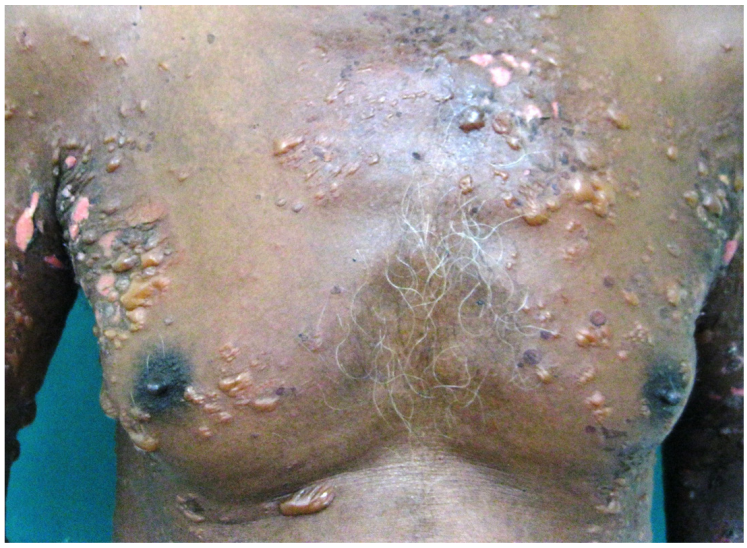
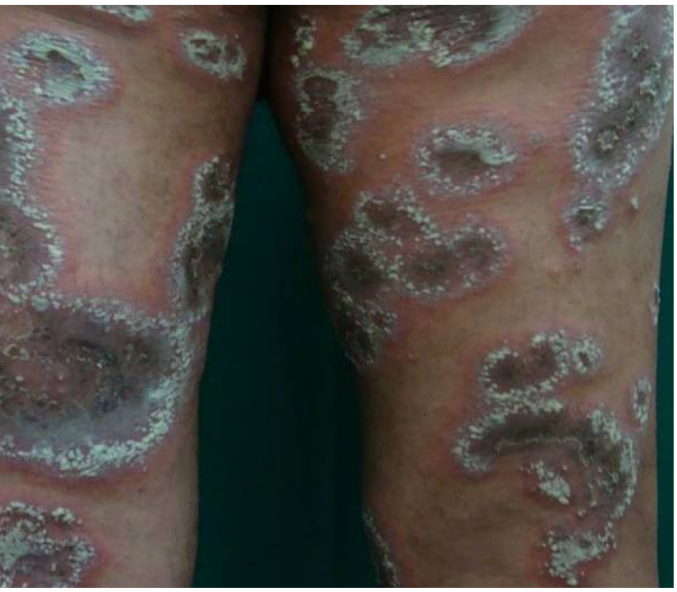
Dermatological Emergencies







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INTRODUCTION

Neonatal erythroderma is a rare condition presenting at birth or during the first month of life. The causes of neonatal erythroderma are manifold; most of these disorders are unique to this age group, some others are commonly seen in older children and adults.

Erythroderma is defined as "any inflammatory condition involving more than 90% body surface area" [1]. In neonates widespread erythema is almost always present; in addition there may be scaling, vesiculation, and eczematization alone or in various combinations. This often leads to use of the term *red scaly baby* for erythrodermic neonates. It can be a manifestation of various primary cutaneous disorders or a cutaneous reaction pattern to systemic illnesses.

The common causes of neonatal erythroderma are infections, inflammatory disorders, and congenital ichthyoses [2]. Some disorders of immunodeficiency and inborn errors of metabolism may also present with erythroderma in this age group.

Neonatal erythroderma is a diagnostic and therapeutic challenge for clinicians. Disruption of the cutaneous barrier predisposes these children to infections, dehydration, and metabolic disturbances. Early accurate diagnosis is crucial for survival of the baby as some of the underlying causes are treatable. An integrated approach by neonatologists and dermatologists is imperative for the management of neonatal erythroderma.

EPIDEMIOLOGY

The true incidence of neonatal erythroderma in various populations is not known, indicating its rarity. In an Indian study the incidence of childhood erythroderma has been quoted as 0.11% ($n = 17$) of whom three (18%) were neonates [3]. In a French study including 51 cases of neonatal and infantile erythroderma, 16 (32%) had onset at birth; of them 15 neonates had congenital ichthyosis and one had Omenn syndrome [4].

CLASSIFICATION AND CLINICAL FEATURES

Various causes of erythroderma in the neonatal period are listed in Table 18.1 and presented schematically in Figure 18.1 [2,5–8]. Important clinical features of individual disorders have been highlighted subsequently.

Infections

STAPHYLOCOCCAL SCALDED SKIN SYNDROME

Staphylococcal scalded skin syndrome (SSSS) is caused by staphylococcal exotoxins A and B (ETA and ETB) that target epidermal cadherin desmoglein-1 at the granular cell layer [8]. This results in a split below the stratum granulosum causing widespread flaccid bulla formation, followed by erosions and scaling [8,9].

SSSS can occur in neonates either as a community or hospital-acquired infection. Though it is more common in older children, neonates are susceptible to this infection because of lack of immunity against the exotoxins and renal immaturity leading to poor clearance of the toxins [9].

Preterm neonates (born before 36 weeks of gestation), very low birth weight (VLBW, less than 1500 g at birth) and extremely low birth weight (ELBW, less than 1000 g at birth) babies may rarely get the infection [8,10–13]. These babies may develop recurrent episodes of SSSS even after full antibiotic therapy [12].

Unlike other severe neonatal infections where the source of organisms is the birth canal, *Staphylococcus aureus* is

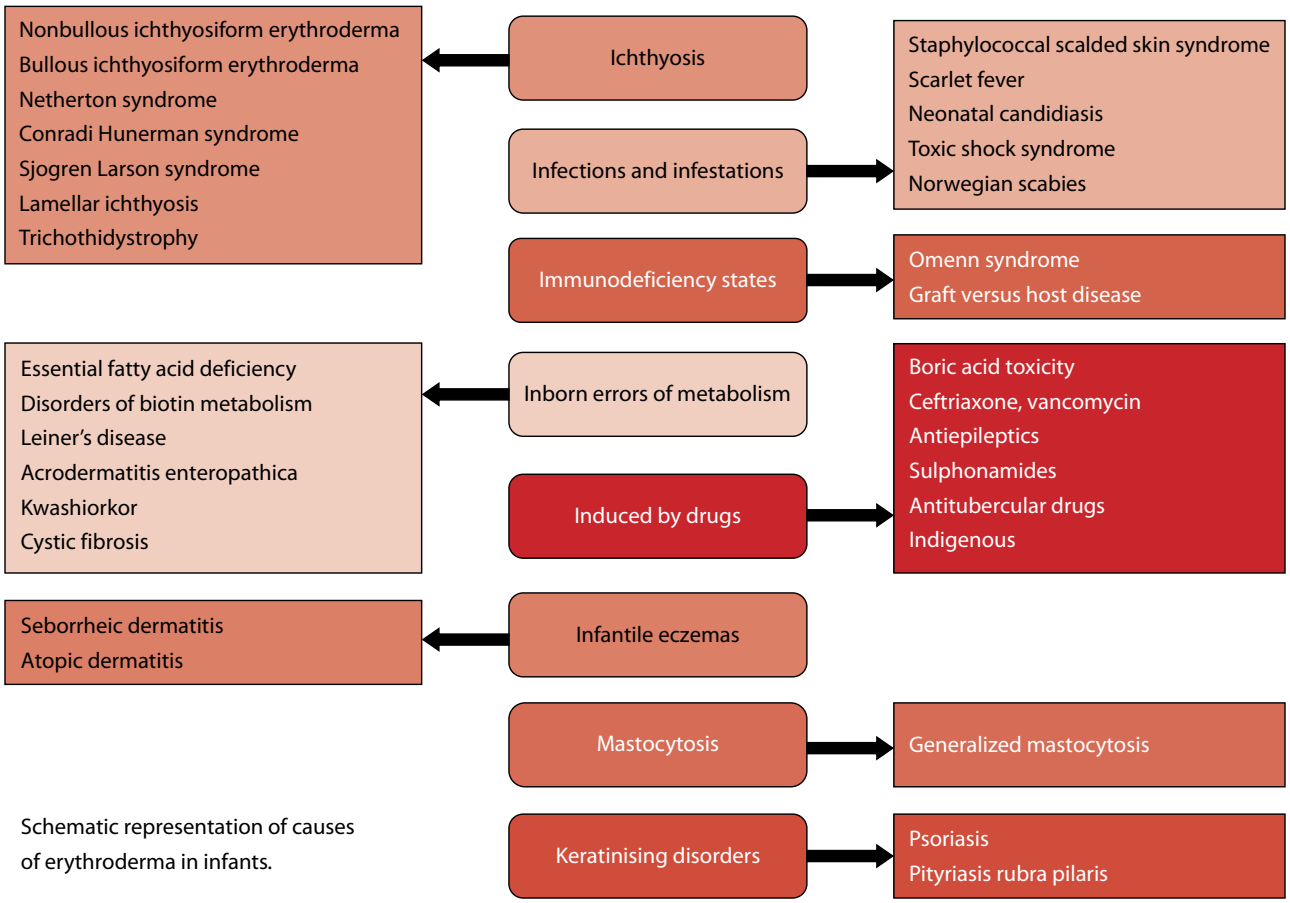


Figure 18.1 Causes of erythroderma in neonates (birth to 1 month) and infants (1 month to 1 year). (Remember the mnemonic: 6 I's in Mini Kids as the main causes of erythroderma in infants.)

Table 18.1 Causes of neonatal erythroderma

Infection	Staphylococcal scalded skin syndrome, neonatal toxic shock syndrome–like exanthematous disease, congenital cutaneous candidiasis
Nonsyndromic congenital ichthyosis	Bullous epidermolytic ichthyosis, nonbullous ichthyosiform erythroderma, lamellar ichthyosis, harlequin ichthyosis
Syndromic congenital ichthyosis	Netherton syndrome, Sjögren-Larsson syndrome, Conradi-Hünermann-Happle syndrome, Chanarin-Dorfman syndrome, keratitis-ichthyosis-deafness syndrome
Ectodermal dysplasia	Anhidrotic/hypohidrotic ectodermal dysplasia
Infiltrative disorder	Langerhans cell histiocytosis, diffuse cutaneous mastocytosis
Immunodeficiency disorders	Severe combined immunodeficiency, Omenn syndrome
Inborn errors of metabolism	Neutral lipid storage disease, multiple carboxylase deficiency, urea cycle defects, neonatal biotinidase deficiency
Cutaneous adverse drug reactions	Reported drugs: Ceftriaxone, vancomycin

Source: Dhar S et al. *Indian J Dermatol* 2012;57:475–8; Hoeger PH, Harper JI. *Arch Dis Child* 1998;79:186–91; Fraitag S, Bodemer C. *Curr Opin Pediatr* 2010;22:438–44; Bedocs LA et al. *Neo Reviews* 2011;12:e325–33; Davidson J et al. *AJP Rep* 2017;7(2):e134–7.

usually acquired from health-care givers [14]. The initial localization of the organism is at the umbilical stump or conjunctiva or at the genitalia following a very early ritual circumcision [14]. Approximately 30% of neonates acquire *S. aureus* infection by the first 6 days of life, and symptoms of SSSS may arise by the 3rd–16th days of life [15]. Usually the cases are sporadic, but outbreaks of SSSS have been reported in neonatal intensive care units due to handling of the babies by infected or asymptomatic carriers of *S. aureus*. SSSS occurring within 24 hours of birth (congenital SSSS)

as part of maternal puerperal infection has been reported [14]. When caused by the methicillin-resistant strain of *S. aureus* (MRSA), it may be life threatening in neonates [9,16].

In neonates the initial focal infection may go unrecognized, and the baby develops diffuse, bright, blanchable erythema simulating acute sunburn [14]. It is followed by wrinkling of the skin on periorificial areas of the face and flaccid thin-walled, large, clear bullae and/or erosions appear all over the body. Nikolsky sign is positive. The erosions give rise to glistening, wet areas surrounded by rolled-up, thin scales [14].

NEONATAL TOXIC SHOCK SYNDROME–LIKE EXANTHEMATOUS DISEASE

Neonatal toxic shock syndrome–like exanthematous disease (NTED) is induced by toxic shock syndrome-1 toxin (TSST-1), mostly produced by MRSA [17]. TSST-1 acts as a bacterial superantigen and induces the immune response; the symptomatology is collectively designated as NTED [17].

Fever, chills, bright macular erythema that may desquamate later, and thrombocytopenia are the features of NTED [17]. It is a mild disease in neonates as compared to toxic shock syndrome in older children and adults. Rapid recovery is usual in this age group [17,18].

CONGENITAL CUTANEOUS CANDIDIASIS

This is a rare infective cause of neonatal erythroderma. Generalized cutaneous candidiasis may be present in newborns when there is chorioamnionitis due to ascending infection in the mother [19]. Babies of mothers with a foreign body in the uterus (specifically, a high cervical suture or an intrauterine device) are susceptible to developing congenital cutaneous candidiasis (CCC) [19].

Skin lesions are present in the initial 24 hours of life as widespread or patchy erythema (Figure 18.2) [19]. Thereafter, generalized erythematous papulopustular lesions develop; palms and soles are typically involved, and the oral cavity and diaper area are relatively spared. Subsequently, there are vesicles and bullae formation followed by erosions and desquamation. Paronychia and nail involvement may be present [19]. In term neonates, the condition is self-limiting and aggressive intervention is not indicated.

Preterm neonates (VLBW and ELBW) are prone to develop severe manifestations of CCC because of immature keratinization [5,19,20]. They may have the classical presentation as previously discussed. More commonly, there is diffuse erythema, erosions, and scaling or multiple erosions present at birth. There may be a stillborn baby. There is risk of candidemia, pneumonitis, and meningitis. Death is a common sequel in preterm babies with CCC.

Congenital ichthyosis

A baby born with collodion membrane is termed as a *collodion baby*. In natural course, the collodion membrane is shed, gradually giving rise to a glazed erythematous and scaly body surface resulting in neonatal erythroderma (Figure 18.3) [21]. Subsequently, these babies may develop a form of congenital ichthyosis, most commonly congenital ichthyosiform erythroderma (CIE), recessive X-linked ichthyosis (RXLI), or lamellar ichthyosis (LI) [21]. Hence, all of these variants of congenital ichthyosis may give rise to neonatal erythroderma. In 10%–20% of neonates born with collodion membrane there may be complete resolution by the third month of life, known as *self-healing collodion baby* [21].

Bullous epidermolytic ichthyosis (EI) or "bullous congenital ichthyosiform erythroderma" is an uncommon disorder where neonates are born with generalized flaccid bullae and erosions resulting from friction during passage through the birth canal [21]. Superficial EI or "ichthyosis bullosa of Siemens" is a milder variant starting at the neonatal period [21]. Lesions start as short-lasting bullae that rupture shortly with annular peeling and collarette-like border (mauserung) [21].

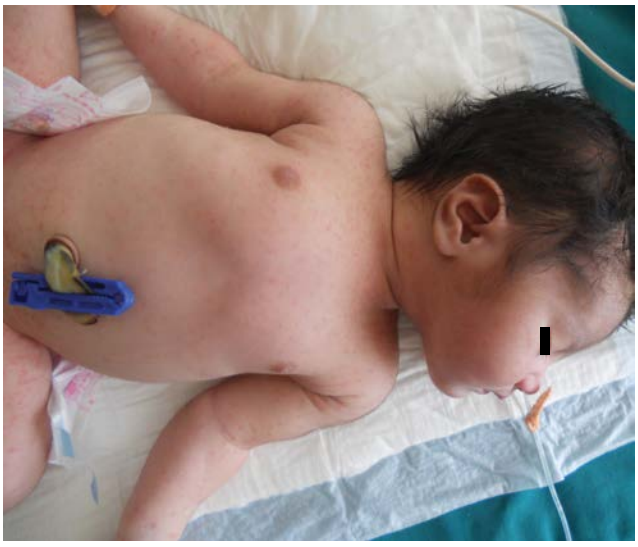


Figure 18.2 Patchy erythema in a case of congenital mucocutaneous candidiasis.

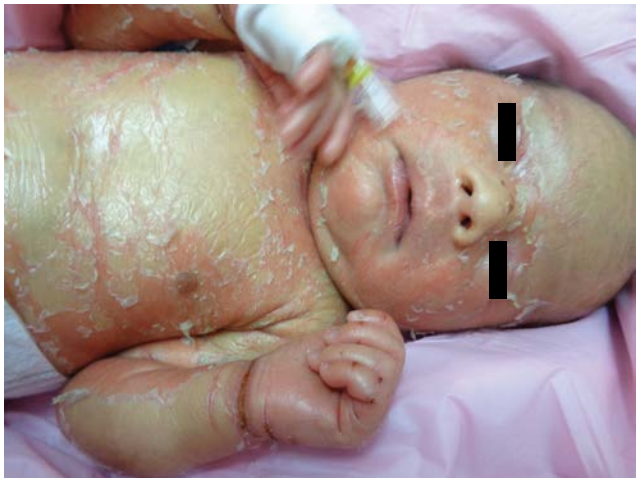


Figure 18.3 Shedding collodion membrane in a neonate. The baby developed clinical features of congenital ichthyosiform erythroderma afterward.

Some of the syndromic ichthyoses may present as collodion baby at birth. These include ichthyosis follicularis, alopecia, and photophobia (IFAP) syndrome; Netherton syndrome (NS); and Sjögren-Larsson syndrome (SLS) [21]. Gradually these children develop the classical features of individual syndrome.

NS and Conradi-Hünermann-Happle syndrome (CHHS) present as neonatal erythroderma [21]. In NS, the classical feature is bright orange-red scaly skin and sparse hair (scalp, eyebrow, eyelashes) at birth (Figures 18.4 and 18.5); during infancy the baby develops the classical double-edged polycyclic scales (ichthyosis linearis circumflexa). In CHHS, the scales are often aligned in swirls and whorls in Blaschkoid pattern [5–7,21].

Patients with keratitis ichthyosis deafness (KID) syndrome have transient erythroderma at or soon after birth [21]. Chanarin-Dorfman syndrome presents with erythroderma at birth due to generalized ichthyosis [21].

Ectodermal dysplasia

ANHIDROTIC/HYPOHIDROTIC ECTODERMAL DYSPLASIA

Patients with this type of ectodermal dysplasia are sometimes born with collodion membrane. It is shed by a few days after birth giving rise to transient erythroderma. The neonate may have the classical midfacial hypoplasia and periorbital wrinkling, which point to the diagnosis. Evolution of the classical manifestations of the disorder occurs through infancy and childhood.

Congenital psoriasis

Psoriasis at the neonatal age is usually localized involving the diaper area and flexures. However, congenital psoriasis, presenting as neonatal erythroderma, has rarely been reported (Figure 18.6) [2,5–7,22].

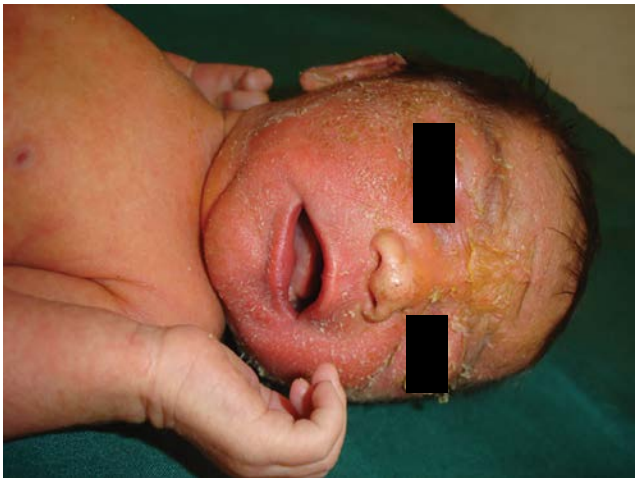


Figure 18.4 A baby with Netherton syndrome at birth. Note the orange-red erythema and double-edged polycyclic scales on the forehead.

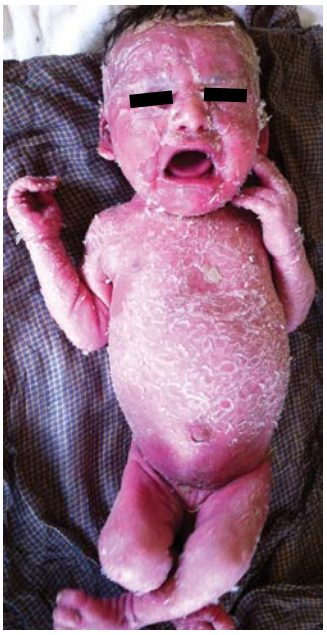


Figure 18.5 The neonate with Netherton syndrome on the seventh day of life with diffuse scaling.

Eczema

ATOPIC DERMATITIS

Atopic dermatitis (AD) may occasionally present as localized lesions in a typical facial distribution in neonates. AD presenting as neonatal erythroderma is extremely rare [2,5–7].

SEBORRHEIC DERMATITIS

The usual presentation of seborrheic dermatitis (SD) in neonates is localized on the scalp (cradle cap) and perineum (diaper dermatitis). Erythroderma resulting from SD may occur in early infancy. However, in a large series of patients with infantile SD, the disease onset was recorded even during the neonatal period [2,5–7,23]. The difference between atopic dermatitis and seborrheic dermatitis is presented in Table 18.2.

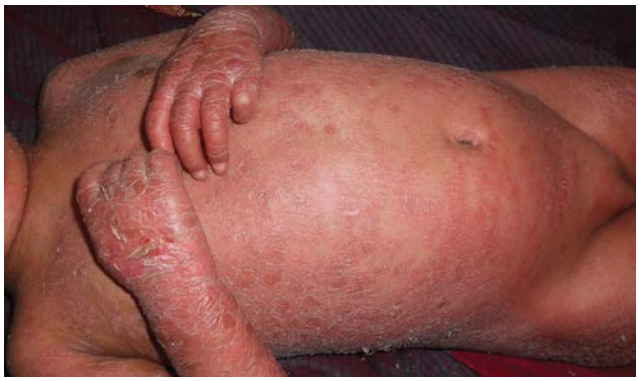


Figure 18.6 Extensive scaling on trunk and upper limbs in a case of congenital psoriasis.

Table 18.2 Differentiating features between atopic and seborrheic dermatitis

Atopic dermatitis	Seborrheic dermatitis
Erythroderma rare in neonatal period	Presents in first few months of life
Dermatitis characterized by vesicles and exudation	Inflammatory, yellowish scaling on the scalp
Itching only after 3 months of age	Erythematous patches are well demarcated and less pruritic
Sparing of napkin area and axilla	Diarrhea, failure to thrive
Positive family history of atopy	Involvement of neck, axilla, and groin

Proliferative disorders

LANGERHANS CELL HISTIOCYTOSIS

Langerhans cell histiocytosis (LCH) is a rare proliferative disorder that may present at birth or in the early neonatal period with diffuse involvement of the skin [24]. Cutaneous involvement may be part of single-system skin-only LCH (SS-LCH), also known as congenital self-healing reticulohistiocytosis (CSHRH) or Hashimoto-Pritzker disease [24]. Otherwise it may be part of multisystem LCH (MS-LCH), also known as Letterer-Siwe disease [24].

The neonate presents with a generalized polymorphic eruption composed of papules, vesicles, crusts, telangiectasias, petechiae, and ulcers (Figure 18.7). The classical feature is seborrheic dermatitis-like greasy, yellow, crusted papules accentuated over the scalp and diaper area observed in 75%–100% cases [24]. As the name suggests CSHRH is a self-regressive disorder, whereas Letterer-Siwe disease carries a poor prognosis requiring systemic therapy [24].

DIFFUSE CUTANEOUS MASTOCYTOSIS

Diffuse cutaneous mastocytosis (DCM) may present at birth. There is generalized involvement of skin with infiltrated papules giving rise to a doughy feel [5–7]. The skin may become erythematous due to handling or cuddling, and Darier's sign may be positive. Occasionally there may be vesicle or bulla formation (Figure 18.8).

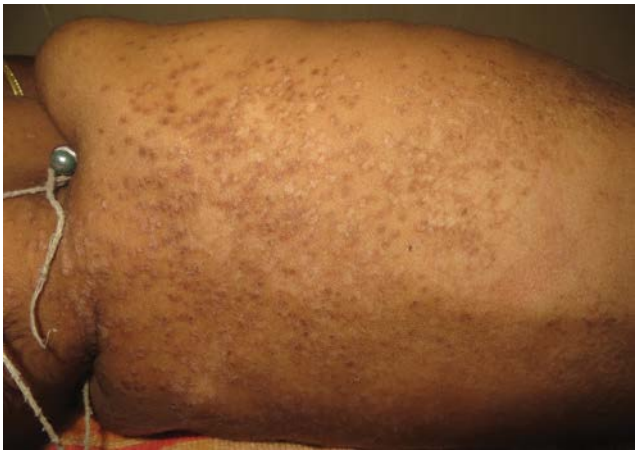


Figure 18.7 Extensive papular eruption in Langerhans cell histiocytosis.

Immunodeficiency disorders

Neonates with several congenital immunodeficiency disorders may present with erythroderma at birth or during the second to third week of life due to graft versus host disease (GVHD). The GVHD results from transplacental transfer and engraftment of maternal lymphocytes to the fetus (either self or from nonirradiated blood/blood product transfusion during pregnancy) [5,7]. This may also result due to postnatal exchange transfusion to the neonate [5]. In contrast to immunocompetent neonates where GVHD is mild and self-resolution is common, it is severe in immunodeficient babies [5].

SEVERE COMBINED IMMUNODEFICIENCY

Neonates with severe combined immunodeficiency (SCID) may present with exfoliative erythroderma due to GVHD resulting from severe T-cell immunodeficient state [7]. In severe cases there are extensive erosions simulating toxic epidermal necrolysis (TEN) [7].

OMENN SYNDROME

Omenn syndrome (OS) is considered as a variant of SCID. Neonates with OS present with erythrodermic eczematous eruptions at birth, and it is a constant feature of the disorder [25]. Often the skin has a pachydermatous appearance due to diffuse infiltration which is strongly suggestive of this

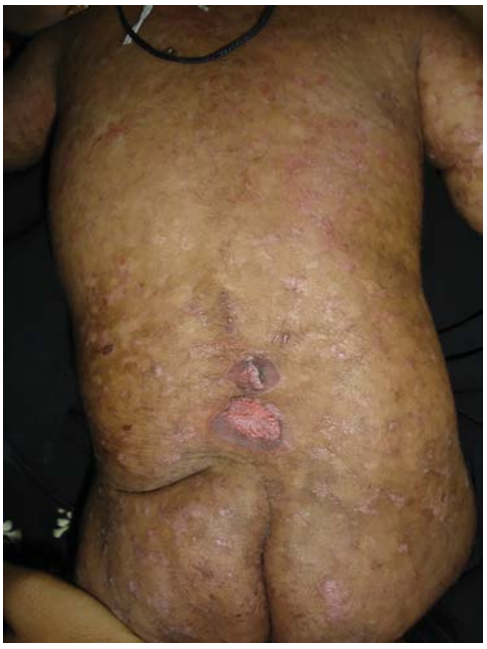


Figure 18.8 Neonate with diffuse cutaneous mastocytosis present since birth. Note the thick skin and erosions on the lower back.

condition [4,6]. There is near total lack of scalp and body hairs at birth [5,26]. Widespread, marked lymphadenopathy, hepatosplenomegaly, and ascites develop subsequently. Recurrent diarrhea, episodes of septicemia, and food intolerance are the causes of failure to thrive in neonates with OS. Life span is short, up to early infancy. The mother often gives the history of death of earlier children in the neonatal period [5].

OTHER IMMUNODEFICIENCY SYNDROMES

Widespread dermatitis is a feature of several primary immunodeficiency disorders like hyper-IgE syndrome, Wiskott-Aldrich syndrome, common variable immunodeficiency, selective IgA deficiency, and X-linked agammaglobulinemia [25]. Sometimes these patients may present with neonatal erythroderma.

Hereditary metabolic disorders

Some rare inborn errors of metabolism may present with erythroderma during early infancy and childhood and rarely during the neonatal period. These conditions are listed in Table 18.3 [27]. Severe metabolic abnormalities dominate the clinical picture in these children, manifesting as poor feeding, recurrent vomiting, dehydration, respiratory distress, lethargy, hepatosplenomegaly, severe ketoacidosis, hypotonia, seizures, and gradual deterioration of consciousness [27].

Rarely, these neonates may present with generalized eczematous rash with accentuation in acral, periorificial areas of the face, perineum, and genitalia simulating acrodermatitis enteropathica (acrodermatitis enteropathica-like eruptions) [27]. There may be erosions and scaling simulating SSSS. The skin lesions may be psoriasiform giving rise to erythroderma [27]. Thinning of hair and alopecia may be seen in these children.

These conditions are fatal with poor prognosis, and death is the eventuality in most of the cases.

Table 18.3 Inborn errors of metabolism that may present as neonatal erythroderma

Organic acidemia	Methylmalonic academia, propionic academia, maple syrup urine disease, glutaricacidemia type 1
Urea cycle defects	Carbamoyl phosphate synthetase deficiency, ornithine transcarbamoylase deficiency, citrullinemia
Neonatal biotin deficiency	Biotinidase deficiency

Source: Inamadar AC, Palit A. In: Inamadar AC, Palit A (Eds), *Advances in Pediatric Dermatology*. New Delhi: Jaypee Brothers Medical Publishers; 2011:234–48.

DIAGNOSIS

Clinical clues

Diagnosis of a case of neonatal erythroderma is mostly clinical. However, misdiagnosis of the underlying condition is frequent because of the common clinical presentation of erythroderma. Some conditions present initially with bullae and erosions (wet disorders), whereas scaling is the primary finding in others (dry disorders). Neonates with wet disorders like SSSS, CCC, and EI often become scaly following rupture of bullae and subsequent drying up of the erosions. Flowchart 18.1 presents the diagnostic clues to various underlying causes of neonatal erythroderma based on the cutaneous features at presentation [2,5–7].

Often systemic involvements may be associated with erythroderma and point to the diagnosis of underlying disorder. Flowchart 18.2 presents the systemic clues to the underlying causes of erythroderma [2,5–7].

Investigations

Laboratory investigations are supportive to clinical diagnosis in neonatal erythroderma.

BEDSIDE LABORATORY PROCEDURES

A few simple bedside laboratory procedures may be helpful in the diagnosis of some cases of neonatal erythroderma. These are presented in Table 18.4 [2,5–7].

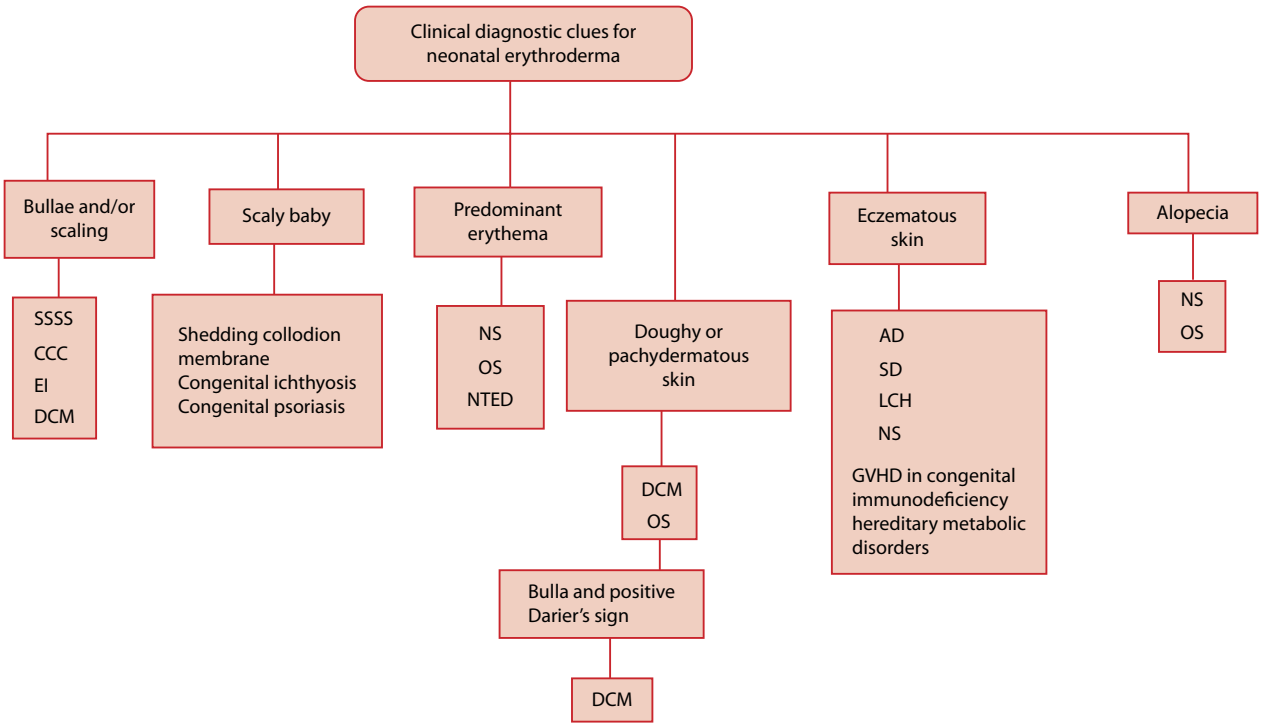
SKIN BIOPSY AND HISTOPATHOLOGY

Histopathology of skin is not diagnostic in all cases of neonatal erythroderma. However, early skin biopsy and histopathological examination are imperative to detect suggestive findings if there are any [2,5–7].

BIOCHEMICAL AND HEMATOLOGICAL INVESTIGATIONS

Biochemical and hematological investigations have a supportive role in the diagnosis of neonatal erythroderma. Table 18.5 presents the list of these investigations helpful in etiological diagnosis [2,5–7]. Complete hemogram is imperative in all cases where systemic involvement is suspected as well as in neonatal infections. Serum electrolytes should be estimated when NS is a diagnostic possibility to detect hypernatremia at an early stage [2,5]. Estimation of the serum IgE level is helpful in conditions with eczematous skin lesions. If hereditary metabolic disorders are suspected, a panel of plasma amino acid level and urinary organic acid levels is to be estimated [6,7]. However, these tests may not be available in routine neonatal setup and can be performed at specialized centers.

Neonates with erythroderma due to conditions like nonsyndromic congenital ichthyosis and psoriasis do not require any biochemical and hematological investigation at the outset. However, the clinical course of a case of neonatal erythroderma may be complicated at any point of patient

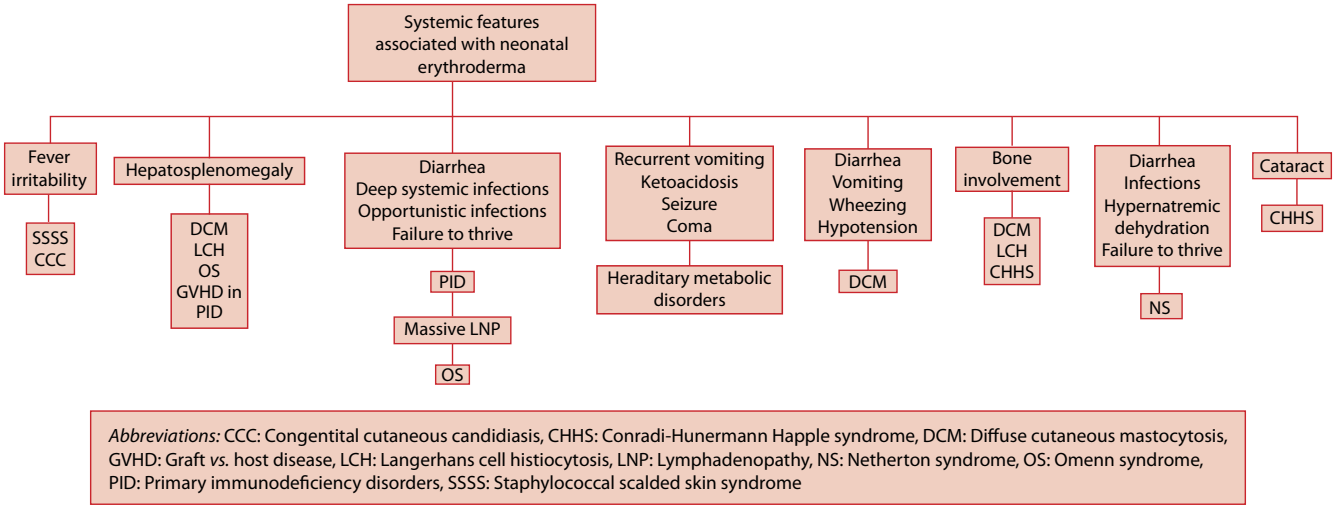


Flowchart 18.1 Clinical diagnostic clues in neonatal erythroderma. (From Dhar S et al. *Indian J Dermatol* 2012;57:475–8; Hoeger PH, Harper JI. *Arch Dis Child* 1998;79:186–91; Fraitag S, Bodemer C. *Curr Opin Pediatr* 2010;22:438–44; Bedocs LA et al. *Neo Reviews* 2011;12:e325–33.)

care due to gross cutaneous barrier dysfunction, infective complications, and metabolic disturbances. In such situations, serial monitoring of biochemical and hematological parameters is necessary. Increased total leukocyte count is indicative of sepsis.

MICROBIOLOGICAL INVESTIGATIONS

When the underlying cause of erythroderma is infection (SSSS, NTED, CCC), isolation of the organism by culture may be attempted if there is any primary focus. Umbilical stump, genitalia, maternal vaginal canal (high up), or



Flowchart 18.2 Systemic involvement in neonatal erythroderma. (From Dhar S et al. *Indian J Dermatol* 2012;57:475–8; Hoeger PH, Harper JI. *Arch Dis Child* 1998;79:186–91; Fraitag S, Bodemer C. *Curr Opin Pediatr* 2010;22:438–44; Bedocs LA et al. *Neo Reviews* 2011;12:e325–33.)

Table 18.4 Various bedside laboratory procedures helpful in the diagnosis of neonatal erythroderma

Procedure	Result	Underlying condition
Tzanck smear	Acantholytic cells	Staphylococcal scalded skin syndrome
Potassium hydroxide preparation from skin lesions, umbilical cord of newborn and placenta, and high vaginal swab of mother	Pseudohyphae and spores	Congenital cutaneous candidiasis
Light microscopy of hair (scalp/eyebrow/eyelashes)	Trichorrhexis invaginata	Netherton syndrome

Source: Dhar S et al. *Indian J Dermatol* 2012;57:475–8; Hoeger PH, Harper JI. *Arch Dis Child* 1998;79:186–91; Fraitag S, Bodemer C. *Curr Opin Pediatr* 2010;22:438–44; Bedocs LA et al. *Neo Reviews* 2011;12:e325–33.

Table 18.5 Biochemical and hematological tests helpful in the diagnosis of neonatal erythroderma

High serum IgE level and eosinophilia	Netherton syndrome, Omenn syndrome, atopic dermatitis
Neutrophilia	Staphylococcal scalded skin syndrome, neonatal toxic shock syndrome–like exanthematous disease, congenital cutaneous candidiasis
Thrombocytopenia	Langerhans cell histiocytosis, neonatal toxic shock syndrome–like exanthematous disease
Lymphopenia	Congenital immunodeficiency syndromes
Hypoproteinemia	Omenn syndrome
Low serum bicarbonate level: acidosis	Hereditary metabolic disorders
Serum electrolytes: hypernatremia	Netherton syndrome
Raised plasma amino acids, high urinary organic acids, urinary ketone bodies	Hereditary metabolic syndromes

Source: Dhar S et al. *Indian J Dermatol* 2012;57:475–8; Hoeger PH, Harper JI. *Arch Dis Child* 1998;79:186–91; Fraitag S, Bodemer C. *Curr Opin Pediatr* 2010;22:438–44; Bedocs LA et al. *Neo Reviews* 2011;12:e325–33.

placenta may bear the evidence of primary infection [14,19]. In premature neonates with CCC, blood, urine, and cerebrospinal fluid samples should be cultured to rule out invasive infection by *Candida*. In the presence of sepsis in an erythrodermic neonate, repeated culture sensitivity test from the fissures on skin or blood culture may be necessary as and when indicated.

Other disease-specific specialized investigations that can confirm the diagnosis of certain disorders are presented in Table 18.6 [2,5–7].

MANAGEMENT

Management of neonatal erythroderma is difficult and requires devoted care from dermatologists and neonatologists. The two challenges are the very young age of the patient and disruption of the cutaneous barrier.

General measures

Management in a neonatal intensive care unit is compulsory, and stringent maintenance of ambient temperature and humidity is essential. This minimizes transepidermal water loss in erythrodermic neonates, especially preterm babies. Close monitoring of fluid and electrolyte balance and maintenance of intake-output chart are necessary. Vital parameters to be monitored in an erythrodermic neonate are presented in Box 18.1 [5–7].

Collodion baby and erythroderma due to congenital ichthyoses present with eclabium, making breastfeeding and

Table 18.6 Confirmatory tests for diagnosis of certain disorders causing neonatal erythroderma

Disorder	Confirmatory test
Netherton syndrome	• LEKTI immunostaining of skin specimen • *SPINK5* mutation analysis
Severe combined immunodeficiency	• *RAG*1 and 2 mutation analysis (null mutation)
Omenn syndrome	• Lymphocyte subset profile; lack of B cells and aberrant expansion of oligoclonal T cells (TH2 subtype) • *RAG*1 and 2 mutation analysis (hypomorphic mutation)
Bullous epidermolytic ichthyosis	• Keratin (K5 and K10) mutation analysis
Hereditary metabolic disorders	• Metabolic analysis of cultured fibroblasts

Source: Dhar S et al. *Indian J Dermatol* 2012;57:475–8; Hoeger PH, Harper JI. *Arch Dis Child* 1998;79:186–91; Fraitag S, Bodemer C. *Curr Opin Pediatr* 2010;22:438–44; Bedocs LA et al. *Neo Reviews* 2011;12:e325–33.

BOX 18.1: Monitoring a neonate with erythroderma

- Pulse, blood pressure, respiratory rate
- Body temperature
- Fluid input and level of hydration
- Urine output
- Level of consciousness (alert with good reflexes or lethargic)
- Evidence of cutaneous infection (skin fissures, umbilical stump, genitalia)
- Evidence of systemic infection (pulmonary, meningeal)

Source: Hoeger PH, Harper JI. *Arch Dis Child* 1998;79:186–91; Fraitag S, Bodemer C. *Curr Opin Pediatr* 2010;22:438–44; Bedocs LA et al. *Neo Reviews* 2011;12:e325–33.

assisted feeding difficult. Neonates with hereditary metabolic disorders are usually lethargic and refuse feeding. These babies require assisted feeding through nasogastric tubes.

Ectropion is a common association in erythrodermic neonates, especially in conditions associated with scaling. Frequent lubrication of the eyes with artificial tears and protective eye pads or shields are to be used to prevent exposure keratitis.

Flexion contracture of digits and limbs are common in these children, which can be prevented by frequent gentle passive movements. In neonates with adherent collodion membrane or severe erythroderma due to congenital ichthyosis, restriction in chest movement may occur; these babies should be observed for respiratory distress. Accumulated scales in the external nares and auditory canal may result in blockage and require periodical cleaning.

Daily cleansing is necessary, especially the flexures, genitalia, and perianal areas to minimize colonization by pathogenic organisms.

Use of long adhesive tapes for fixation of various gadgets (intravenous canula, nasogastric tube) to the skin of neonates with SSSS and EI should be avoided as these babies have fragile skin; either short strips of adhesive tapes or roller bandage can be used if the situation permits.

BOX 18.2: Indications of systemic retinoid therapy in neonatal erythroderma due to congenital ichthyoses

1. Severe ectropion and eclabium causing difficulty in feeding
2. Respiratory distress due to adherent, taut collodion membrane on thoracic region
3. Flexion contractures of digits and limbs

Excessive handling, rubbing, and cuddling should be avoided in neonates with DCM. Such stimulations may induce flushing episodes and bullae formation in these babies.

Specific treatment

TOPICAL

Bland emollients like white soft paraffin should be applied all over in neonates with ichthyotic and eczematous skin conditions several times a day. In the presence of large erosions resulting from rupture of bullae, the denuded areas can be covered with liquid paraffin embedded gauze dressings.

Psoriasis and eczematous conditions like atopic and seborrheic dermatitis are mostly treated with mild topical corticosteroids like hydrocortisone. Caution should be undertaken to prevent enhanced absorption of topical corticosteroids due to the damaged cutaneous barrier. Topical tacrolimus ointment (0.03%) can be used in addition in some cases with atopic dermatitis. The latter agent should never be used in erythrodermic neonates where NS is a diagnostic possibility; a very high serum concentration of tacrolimus may be attained in these babies due to the damaged skin barrier [5–7].

SYSTEMIC

Neonatal erythroderma due to infections are treatable and curable. SSSS and NTED are treated with an IV antistaphylococcal antibiotic such as cloxacillin [5–7]. If MRSA is isolated, vancomycin is the drug of choice [5–7]. CCC is treated with systemic antifungal agents such as oral fluconazole or in severe cases with IV amphotericin B. Topical clotrimazole cream can be added to a systemic regimen.

Disabling erythroderma due to congenital ichthyosis requires treatment with systemic retinoids, either isotretinoin or acitretin (0.5–1 mg/kg). Indications of starting systemic retinoids in these patients are presented in Box 18.2.

Neonates with erythroderma due to DCM are treated with a combination of histamine receptor (H1 and H2) blockers. Oral sodium chromoglycate can also be used [6]. Potential mast-cell degranulating drugs must be avoided in these patients [6].

Superficial and deep pyogenic infections in primary immunodeficiency disorders, OS, and NS are treated with appropriate antibiotics. If blood transfusion is required for the neonates with SCID and OS, irradiated blood must always be used to prevent a chance of occurrence of GVHD [5,6]. Bone marrow transplantation is the specific therapy for OS [5,6].

CONCLUSION

It is difficult to reach a definitive diagnosis of erythroderma in neonates. Persistent collodion membrane and congenital ichthyosis are the most common causes of erythroderma present at birth. Infective causes prevail within a few days to 2 weeks of life. Hereditary metabolic disorders

KEY POINTS

- Erythroderma due to staphylococcal scalded skin syndrome in premature neonates may be recurrent in spite of complete antibiotic therapy.
- Premature neonates with erythroderma due to congenital cutaneous candidiasis require blood, urine, and cerebrospinal fluid culture to rule out invasive infection.
- Erythroderma with predominant systemic features at presentation may be indicative of rare hereditary metabolic disorders or primary immunodeficiency disorders.
- A combination of consistent presence of erythroderma, pachydermatous skin, and near total lack of scalp and body hair are the clinical pointers to the diagnosis of Omenn syndrome.
- Generalized eczematous rash rather than scaling with periorificial accentuation is a clinical pointer to rare organic acidemias, and the neonate should be screened accordingly. In a neonate with erythroderma if Netherton syndrome is a diagnostic possibility, serum electrolytes are to be estimated to rule out hypernatremia.

and primary immunodeficiency disorders are distinct by predominant systemic features. A close look at the erythrodermic neonate may provide clinical clues to the diagnosis. Histopathological examinations of skin and laboratory tests are helpful in some cases. A collaborative approach is essential for effective management of the baby. Prognosis is better in infective conditions and nonsyndromic group of ichthyosis. Preterm baby and systemic involvement are the two poor prognostic factors irrespective of the underlying cause.

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