PG Textbook of PEDIATRICS

Volume 3
Systemic Disorders and Social Pediatrics

Piyush Gupta
PSN Menon
Siddarth Ramji
Rakesh Lodha

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SYSTEMIC DISORDERS AND SOCIAL PEDIATRICS

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Section 48 COMMON SKIN PROBLEMS

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Chapter 48.1
Skin of the Newborn: Physiological and Pathological Changes

Keshavmurthy A Adya, Arun C Inamadar

During the neonatal period, the anatomical and physiological maturation of the skin is still in progress. Neonatal skin differs from that of children and adults with respect to both structural integrity as well as functional capacity. The neonatal skin, in general, is thinner with fewer intercellular adhesions, has fewer melanosomes, hair follicles and lesser eccrine secretions. **Table 1** outlines the structural development of the skin at neonatal stage with the corresponding functional attributes and their clinical significance. In preterm neonates, these attributes are further accentuated.

PHYSIOLOGICAL SKIN CHANGES

Majority of the physiological skin changes seen during neonatal period are attributable to the structural and functional immaturity of the skin and/or immaturity of the internal adaptive mechanisms that render the neonates respond differently to the external environmental factors. Hence, such cutaneous changes are essentially transient and resolve completely without treatment. Also, some of the maternal hormones that enter fetal circulation may produce transient skin changes in their newborns as described below.

Appearance of neonatal skin At birth, the skin of the *term neonate* is covered with vernix caseosa, a white gelatinous material that imparts an alkaline pH to the skin. It is believed to have many roles in the intrauterine development as well as protective effect on the maturing newborn skin against environmental factors. The vernix flakes off within a few hours of birth and the skin pH gradually declines to reach the normal acidic values by 4th week. The newborn skin may be covered with fine, short and minimally pigmented lanugo hairs especially on the back, forehead and shoulders. The skin of *preterm neonates* appears rather translucent covered with more obvious and thicker lanugo hairs, which would otherwise be shed off about 4 weeks prior to term to be replaced by a second coat of shorter lanugo hairs seen in term newborns. The *small-for-date babies* have a wrinkled skin due to lack of subcutaneous fat and the skin (and vernix) at birth may be yellow-green due to staining by meconium. The finger nails are often long. The *postmature neonates* appear longer but otherwise have a similar appearance to that of the small-for-date babies. The vernix is often absent.

Table 1 Structural and functional status of neonatal skin and their clinical significance

Skin parameter	*Development in neonates*	*Functional aspects*	*Clinical significance*
Epidermal thickness	Completely developed	Although the epidermal thickness is similar to that of adults, systemic absorption of topically applied substances is increased due to greater surface area to body mass ratio, presence of occlusive conditions such as waterproof nappies, and high ambient temperatures and/or humidity	Increased risk of systemic toxicity from topically applied agents
Intercellular adhesions	Near complete in term but fewer in preterm neonates	Less efficient epidermal barrier function	Increased skin fragility, increased tendency to blistering and increased TEWL
Dermal thickness and subcutaneous fat	Fewer collagen and elastic fibers. Less subcutaneous fat	Increased heat loss	Increased risk of hyperthermia, particularly in preterm neonates in whom the autonomic thermoregulatory mechanisms are also underdeveloped
Melanosomes	Fewer in term and much lesser in preterm neonates	Decreased natural protection from UV radiation	Increased photosensitivity
Eccrine glands	Completely developed anatomically but functionally immature, more so in preterm neonates	Decreased response to thermal stress. Total anhidrosis in preterm neonates	Increased risk of hyperthermia
Hair follicles	Fewer terminal hairs in term and persistent lanugo hairs in preterm neonates		
Sebaceous glands	Hyperplasia due to stimulation by transplacentally transferred maternal androgens	Increased sebum secretion that normalizes by the next 4–6 weeks	Neonatal acne, seborrheic dermatitis and neonatal cephalic pustulosis are all attributable to this increased sebaceous gland activity in neonates

Abbreviations: TEWL, transepidermal water loss; UV, ultraviolet.

Hyperpigmentation In the dark-skinned newborns, a rather conspicuous hyperpigmentation predominantly involving the perineal [typically the scrotum **(Fig. 1)** and vulva] and periungal (base of the fingernails) areas may develop attributable possibly to stimulation by melanocyte-stimulating hormone in utero. Other less commonly involved areas include helix, nipple and areola, axillae and lower abdomen. The condition is transient and resolves spontaneously over sometime. Other infrequently described patterns of transient cutaneous hyperpigmentation of newborns include the *transient linear hyperpigmentation of newborn* and the *transient reticulated pigmentation of the new born*. The former involves the flexures of the limbs and abdomen and possibly represent an incomplete migration of melanocytes in the epidermis of the deepest part of cutaneous folds while the latter has been reported to involve the back and knees presumably as a result of post-traumatic hyperpigmentation in utero.

TRANSIENT SELF-RESOLVING CUTANEOUS AILMENTS

Vascular Phenomena

Erythema neonatorum Many of the healthy term infants develop a generalized striking erythema within a few hours that fades within a day or two. It is possibly attributed to higher levels of hemoglobin in the first week of life. This has to be differentiated from erythema toxicum neonatorum described below.

Acrocyanosis Cyanosis involving the perioral region and extremities (especially palms and soles) may occur in some of the term neonates which in absence of cyanosis of the central parts, is regarded as normal within the first 2 days. It becomes more obvious with reduction in surrounding temperatures, crying, or breath holding spells and disappears on re-warming. This physiological acrocyanosis appears to be due to an increased tone of peripheral arterioles, which in turn creates vasospasm, secondary dilatation, and pooling of blood in the venous plexuses.

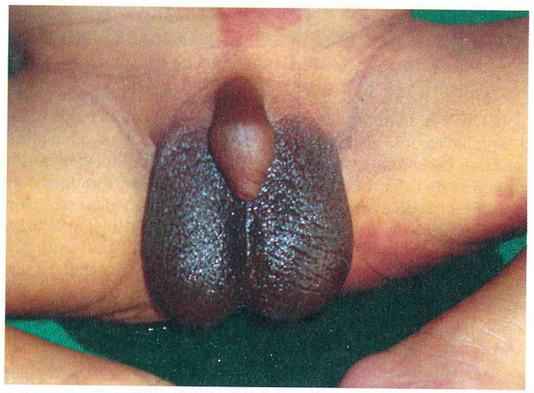

Figure 1 Physiologic hyperpigmentation of scrotum

Cutis marmorata (CM) A characteristic marbling of the skin in the form of reticulate bluish pattern **(Fig. 2)** on exposure to cold is a common occurrence in healthy neonates that readily disappears on rewarming. The trunk and extremities are involved symmetrically. The mechanism of development of CM is same as in acrocyanosis described above. Generally, CM disappears by the end of neonatal period but may, in some instances, persist for weeks to months without any clinical relevance. However, persistent CM beyond the neonatal period or the one which recurs may be a marker of Down syndrome, Edward syndrome, hypothyroidism and several others. A distinct negative pattern of mottling known as *cutis marmorata alba* may be seen in some cases due to constriction of deep vasculature.

Cutis marmorata telangiectatica congenita (CMTC), although resembles physiological CM, is a distinct entity in which the vascular mottling has a deep violaceous color, remains persistent even after rewarming and is associated with other cutaneous changes like atrophy and ulceration. Exact etiopathogenesis of CMTC is unclear but its significance lies in the fact that it forms a part of various syndromes (e.g. Adams-Oliver syndrome, phacomatosis pigmentovascularis type IV, etc.) and is also associated with various congenital anomalies (most common being limb hypoplasia).

Harlequin color change Harlequin color change is seen in about 10% of healthy newborns in whom an intense erythema on the dependent portion and pallor over the nondependent part with a clear demarcation between them develops when the baby is placed on one side. This change resolves immediately on placing the baby in supine position. It is of no pathological significance and possibly reflects the immaturity of the hypothalamic control over peripheral vascular tone. However, persistence of harlequin color change beyond 4th week may be indicative of hypoxia due to cardiovascular anomalies. It is also seen in neonates who have received general anesthesia or prostaglandin E2 infusion.

Manifestations of Circulating Maternal Sex Hormones

Neonatal acne Neonatal acne is not very uncommon and occurs within the first 2 weeks in most of the neonates. It may be congenital in about 20%. Neonatal acne is more common in male babies. The cheeks are the predominant sites affected, and closed comedones/papulopustular lesions **(Fig. 3)** rather than open comedones/cysts are the most common lesions. Stimulation of sebaceous glands by maternal androgens or transient fetal adrenal and gonadal androgen production is believed to be the cause for neonatal acne.

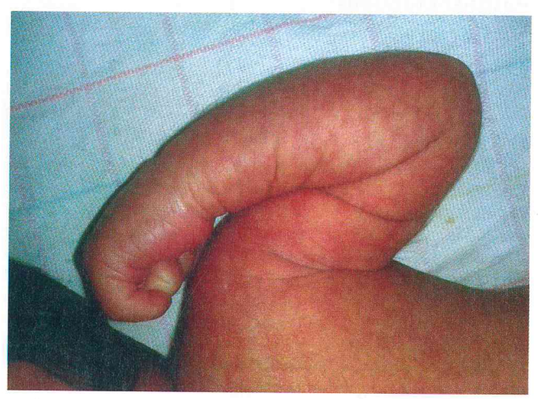

Figure 2 Cutis marmorata
(*Source:* Dr Ahamadrasool Shirasangi, Consultant Pediatrician, Bagalkot, India)

Figure 3 Neonatal acne
ırce: Dr Ahamadrasool Shirasangi, Consultant Pediatrician, Bagalkot, India)

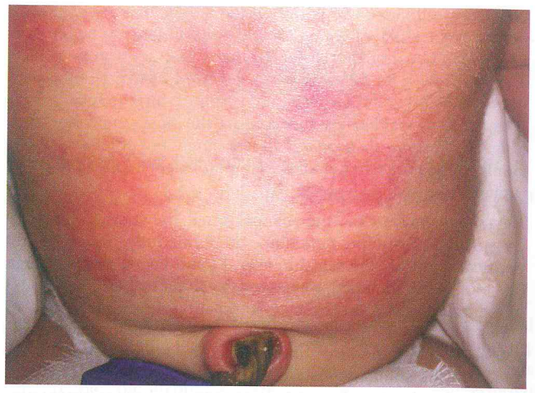

Figure 4 Erythema toxicum neonatorum

thermore, the testicular androgens stimulate primed adrenal ıds which may partially explain the male preponderance. The ons resolve spontaneously in the next 3 months without any ıtment.

aceous hyperplasia Circulating maternal androgens, or ısient overproduction of fetal androgens exert a trophic effect the sebaceous glands manifesting clinically as grouped tiny owish-white follicular papules without surrounding erythema centrated over the nose, cheeks and upper lips. They are seen in o 50% of term neonates and involute within the first few weeks tpartum.

:iature puberty A constellation of transient clinical phenomena ılting from the influence of maternal and placental hormones he fetus develop in the neonatal period collectively grouped *iniature puberty*. Female genitalia appear well-developed with ınspicuous clitoral hypertrophy, mucoid vaginal discharge or ı frank withdrawal bleeding beginning on 3rd or 4th day lasting 2–3 days. The male genitalia similarly appear well-developed. ıst hypertrophy is seen in both the sexes at birth which subsides he end of 4th week. In some babies, the breasts may engorge ıe first 2–3 days with lactation of the so-called *witch's milk*. No rvention is needed.

:tular Eruptions

ular eruptions in the newborn period include a host of ditions that include benign transient eruptions (describe w), infectious conditions and others. The transient eruptions, ıgh themselves do not bear much clinical significance and lve spontaneously, it is important, however, to differentiate ı from other causes as described in **Table 2**.

hema toxicum neonatorum (Toxic erythema of newborn) hema toxicum neonatorum (ETN) is a common self-resolving ılopustular eruption seen in healthy newborns. The incidence TN increases with the gestational age as it is seen in more half of term neonates and is rare in preterm neonates. It is acterized by erythematous macules, follicular papules and le pustules **(Fig. 4)**. The onset, in most of the cases, is between 1 and day 4 and the rash resolves within 4 days without any ment. The trunk is the most favored area; although, the face extremities may also be involved while the palms and soles are ed. There is no gender or racial predilection. A characteristic ıre is the presence of peripheral eosinophilia as well as ominant eosinophilic infiltrate in the follicular infundibulum. xact etiology of ETN is unknown. While some believe it to be a

Table 2 Pustular eruptions in newborns

Transient conditions	Erythema toxicum neonatorum Transient neonatal pustular melanosis Infantile acropustulosis Eosinophilic pustular folliculitis Miliaria pustulosa
Infectious conditions	*Bacterial:* Staphylococcal infections Streptococcal infections *Pseudomonas* infection Neonatal listeriosis Congenital syphilis *Viral:* Neonatal herpes simplex Varicella zoster infection *Fungal:* Congenital candidiasis Neonatal cephalic pustulosis Pityriosporum folliculitis
Neoplastic disorders	Langerhans cell histiocytosis Urticaria pigmentosa
Inherited disorders	Acrodermatitis enteropathica Incontinentia pigmenti
Others	Pustular psoriasis Scabies

response of neonatal skin to mechanical or thermal stimuli others consider it as an immune response to microbial colonization of the hair follicles. It is also proposed to be a form of graft-versus-host reaction caused by materno-fetal transfer of lymphocytes during or before delivery that react with some of the baby's antigens. An association with dyspepsia that leads to absorption of enterotoxins with subsequent development of rash (hence the name *toxic erythema*) has also been advocated.

Transient neonatal pustular melanosis (TNPM) Transient neonatal pustular melanosis is characterized by superficial noninflammatory vesicopustules that easily rupture to form crusts surrounded by a collarette of scale. These scaly lesions are later replaced by postinflammatory hyperpigmentation that resolves gradually over several weeks. The exact etiopathogenesis is unclear and it has been thought to be a variant of ETN. In some cases, the presentation may be with hyperpigmentation suggesting that the previous stages occurred in utero. TNPM differs from ETN in being congenital, affecting the chin, neck and shins more commonly than the trunk, and the lesions showing predominant neutrophilic infiltrate. The incidence is more common in term infants of African descent without any gender preference. As with ETN, no treatment is required.

Infantile acropustulosis (IA) An uncommon eruption, usually occurring between 3 and 6 months of age but can be seen at birth or neonatal period characterized by recurrent crops of intensely pruritic, acrally located (including palms and soles) papulopustular lesions is termed as infantile acropustulosis. The lesions last for 7–14 days and recur in 3–4 weeks. With time, the intensity and frequency of the rash decrease with eventual spontaneous resolution by 3–4 years. The exact etiology is not known but IA is thought to a post-scabetic hypersensitivity reaction. Indeed, infantile scabies is a differential diagnosis for IA and mineral oil examination must be carried out in these cases. However, the characteristic burrows of scabies are not present. A smear prepared from the pus reveals predominantly neutrophils. Potent topical steroids in mild-to-moderate cases and systemic dapsone in recalcitrant cases prove successful.

Eosinophilic pustular folliculitis of infancy (Ofuji disease) Ofuji disease is a rare disorder affecting almost exclusively the boys. It is characterized by recurrent crops of papulopustular lesions commonly involving the face and scalp and occasionally the trunk and extremities which evolve through a crusting phase to heal without any residua. The presentation may be at birth or in the first few weeks with spontaneous resolution occurring in a few months. Peripheral eosinophilia and numerous eosinophils in the pustular contents are helpful in the diagnosis. Treatment is mainly symptomatic.

Others

Superficial cutaneous desquamation Superficial desquamation is seen in more than 75% of the term neonates that begins at 24–36 hours of age. It initially begins at the ankles and may remain localized or become widespread. Traditionally, this type of desquamation was believed to be rare in preterm and very obvious in post-term babies. However, Rivers et al. (JAAD, 1990) observed that this phenomenon occurs with equal prevalence at all gestational ages. Superficial desquamation is always much more severe in small-for-date babies irrespective of their gestational age. Severe and excessive desquamation at birth or in the early neonatal period may be indicative of congenital ichthyosis and X-linked hypohidrotic ectodermal dysplasia.

Sucking blisters Vigorous sucking *in utero* leads to development of flaccid blisters at the affected site and is a common occurrence. Most commonly the radial forearm, wrists and fingers are involved either unilaterally or bilaterally. Presentation at birth may be with intact blisters or with erosions and callosities which subside spontaneously within a few days.

Hair changes During the 5th month of intrauterine life, there is a synchronous shedding of the entire scalp hair that regrow and enter telogen phase in a wave from front to back beginning at about 12 weeks before term. After shedding of these telogen hairs from the frontal and parietal areas, the follicles again enter the anagen phase in a similar pattern from front to back. However, the follicles in the occipital region do not enter telogen until term and, therefore, a rather conspicuous alopecia may appear at this site due to shedding of these telogen hairs in the early neonatal period.

In some babies, there is an unusually diffuse hair loss during the neonatal period (telogen effluvium of the newborn) and by the end of 6 months, most babies have regrown all the hair. At this stage, hairline often extends to the lateral ends of the eyebrows, but the terminal hairs comprising this extension are gradually converted to vellus hairs during the remainder of the first year, causing the hairline to recede to its characteristic childhood position.

Milia Milia are characterized by multiple tiny whitish-yellow papules that represent miniature epidermal cysts derived from pilosebaceous follicles **(Fig. 5)**. These are seen in 30–50% of the neonates and get extruded by themselves in a few weeks. Milia are also seen involving the hard palate (Epstein's pearls) and alveolar margins (Bohn cysts) in majority of newborns which behave in the same manner as cutaneous milia. Large, extensive milia that are persistent and involve atypical sites may be indicative of orofacial-digital syndrome type I, Marie-Unna type congenital hypotrichosis or the X-linked Bazex-Dupré-Christol syndrome.

Miliaria Miliaria results due to blockage of eccrine ducts and is a frequently encountered transient ailment affecting about 15% of the newborns. It is commonly associated with hot humid surroundings and friction, and resolves spontaneously on cooling. Depending on the level of obstruction of the eccrine duct, three clinical forms are described—(1) miliaria crystallina (block at subcorneal level), (2) miliaria rubra (block in the malpighian layer), and (3) miliaria profunda (block at the dermoepidermal junction). Miliaria crystallina is characterized by noninflammatory clear flaccid vesicles on the forehead, neck and other occluded areas which are commonly seen on 6th or 7th day of life. Miliaria rubra appears as erythematous papules due to associated inflammation **(Fig. 6)** as a result of seepage of the ductal contents into the dermis. These lesions typically appear between 11th and 15th day and are often associated with discomfort. Sometimes the inflammation is intense enough to produce pustular lesions (miliaria pustulosa). Miliaria profunda is characterized by larger fleshy papules or nodules involving the trunk. In type I pseudohypoaldosteronism, miliaria pustulosa (pustular miliaria rubra) is considered to be a specific finding.

PATHOLOGICAL SKIN CHANGES IN NEONATES

Seborrheic Dermatitis

The infantile seborrheic dermatitis, distinct from the adult form, begins commonly in the first month of life and clears by the age of 6 months. Excessive sebum production and *Malassezia* species' colonization are implicated in the pathogenesis but not proven unequivocally. The congenital form manifests as thick yellowish or whitish greasy adherent plaque mostly involving the vertex and

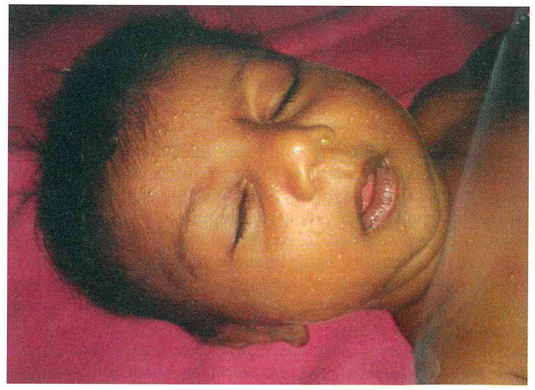

Figure 5 Milia
(*Source:* Dr Ahamadrasool Shirasangi, Consultant Pediatrician, Bagalkot, India)

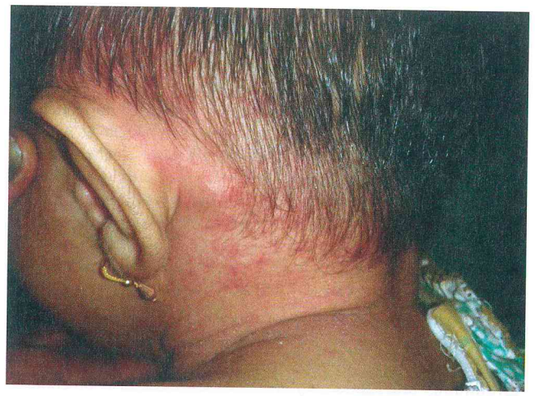

Figure 6 Miliaria rubra
(*Source:* Dr Ahamadrasool Shirasangi, Consultant Pediatrician, Bagalkot, India)

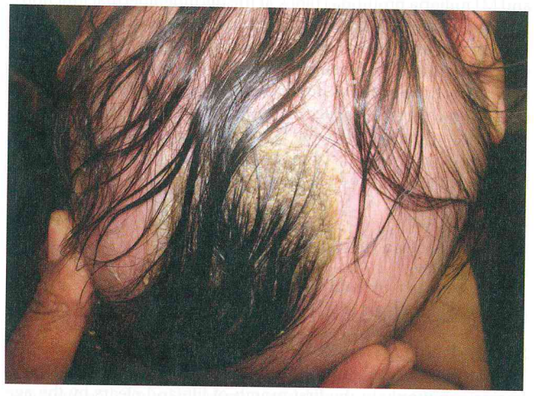

Figure 7 Infantile seborrheic dermatitis presenting as *cradle cap*

frontal regions [cradle cap **(Fig. 7)**]. The acquired form develops on previously clear scalp and is associated with involvement of body folds like neck, axillae, groins where the lesions are more inflamed and macerated. At this stage, infantile seborrheic dermatitis is difficult to differentiate from atopic dermatitis and inverse psoriasis. However, the prognosis of infantile seborrheic dermatitis is excellent with resolution occurring in a few weeks even without treatment and mild topical steroids are helpful for faster clearance.

Psoriasis

Psoriasis in the neonatal period is, in general, uncommon. The common forms of psoriasis at this age are the napkin psoriasis (presenting as diaper dermatitis) and much rarer pustular psoriasis. Although rare, congenital psoriasis is also described. Sometimes infantile psoriasis may involve the scalp and face when a differentiation from infantile seborrheic dermatitis is possible only by biopsy. Koebner's phenomenon is common and nail changes are seen in 10% of the cases. Pustular psoriasis may present at birth or in the few weeks after birth. Presentation is often with fever and sheets of small pustules. Annular or circinate forms may also be seen. Associated findings include geographic tongue, sterile osteomyelitis, and very rarely pulmonary involvement with the capillary leak syndrome.

Napkin Dermatitis

An inflammatory rash involving the diaper area is commonly encountered between 3rd and 12th week, which is considered to be an irritant contact dermatitis because of prolonged contact with feces and urine. It is exceedingly rare in the absence of napkin wearing and other associated factors like maceration by water, friction, frequent urination, diarrhea, pancreatic proteases and lipases in feces, ureases produced by fecal bacteria, use of broad spectrum antibiotics and developmental anomalies of urinary tract. The most common presentation is with confluent erythema of the convexities in the closest contact with the napkin (buttocks, genitalia, lower abdomen, pubic area and upper thighs) with sparing of deeper parts of the groin flexures. Sometimes the eruption may be confined to the margins of the napkin area (tidemark dermatitis) or present as an erosive form with small vesicles and erosions which develop into shallow, round ulcers (Jacquet's dermatitis). Several distinctive variants have been observed. Secondary infection by *C. albicans* is common when the rash is surrounded by satellite pustules. Diaper dermatitis may be an early feature of atopic dermatitis or infantile seborrheic dermatitis.

Treatment includes use of good quality napkins with a topsheet impregnated with an emollient, frequent changing of napkins, and use of mild topical steroids or anti *Candida* antifungals as indicated.

Subcutaneous Fat Necrosis

Subcutaneous fat necrosis (SCFN) presents as asymptomatic or tender erythematous and indurated plaques or nodules due to focal areas of fat necrosis appearing in the first month of life with characteristic histological features. The condition is rare and self-limiting and is seen with predispositions like maternal diabetes, perinatal asphyxia, hypothermia, hypoxia, neonatal infections and other causes. The neonatal fat undergoes crystallization more readily in presence of hypothermia leading to fat cell damage and granulomatous reaction. The cheeks, back, buttocks and arms are the frequently involved sites. Skin biopsy shows an infiltrate of lymphocytes, histiocytes and fibroblasts, with necrosis of fat and multinucleated giant cells. Doubly refractile crystals can be seen under polarized light. Calcium deposits and eosinophilic granules are also common.

Cold Panniculitis

Painful red, indurated nodules developing on exposure to cold climate occurs in newborns attributable to thermally induced damage and necrosis of subcutaneous fat as in SCFN. Induration resolves over a period of a week or so, often with postinflammatory hyperpigmentation.

Sclerema Neonatorum

Sclerema neonatorum is a condition characterized by a rapidly spreading nonedematous skin hardening seen most commonly in preterm neonates who are severely ill with poor perfusion. Hypothermia, metabolic acidosis, sepsis, cardiovascular disease, pulmonary hemorrhage, central nervous system abnormalities, and glucose or electrolyte imbalances have been implicated in the pathogenesis. Neonatal sepsis, however, is the most commonly encountered predisposing factor. Thickening initially begins over the buttocks and thighs which rapidly spreads to involve the entire body except for the palms and soles. Reduced chest expansion and flexion deformities of joints may be seen. The scleredermatous skin shows higher concentration of saturated fatty acids which solidify easily in presence of hypothermia and impaired perfusion. Treatment is mainly aimed at removal of underlying cause at the earliest. However, the prognosis of sclerema neonatorum is usually guarded.

Neonatal Purpura Fulminans

Purpura fulminans is a potentially life-threatening, progressive condition characterized by hemorrhagic necrosis of skin due to cutaneous vascular thrombosis. In the neonatal period, it almost always is indicative of congenital homozygous or compound heterozygous deficiency of protein C or S. The condition manifests within a few hours to 5 days of birth characterized by noninflammatory retiform purpura which later coalesce to form large hemorrhagic bullae and necrotic eschar. The pressure points like the buttocks and lower extremities are involved initially and in severe cases mucosal surfaces and internal organs may also be affected. In addition, the neonates are at increased risk of cerebral and retinal vessel thromboses. Unlike with acute infectious purpura fulminans, patients with congenital protein C or S deficiency are often hemodynamically stable and afebrile at presentation unless the condition is precipitated by an infection. Treatment must be prompt and timely, and involves supportive measures with administration of protein C and S either with fresh frozen plasma or protein C/S concentrate followed by oral long-term oral anticoagulants.

Blueberry Muffin Baby

Blueberry muffin baby refers to a newborn that is born or presents in a few days with firm, nonblanchable, bluish-magenta colored papulonodules involving preferentially the head and neck region. These lesions represent persistent dermal erythropoiesis which, in the prevaccination era was commonly a feature of congenital rubella. However, many other vertically transmitted infections present with such lesions, so do many of the hematological and neoplastic disorders **(Table 3)**. The lesions evolve into dark purple to brownish macules and fade away spontaneously within 2–6 weeks. Babies with multiple vascular disorders, such as hemangiopericytoma, hemangioma, blue rubber bleb nevus syndrome and glomangioma may be mistaken for *blueberry muffin* baby. Investigations must include complete blood analysis, TORCH screening, viral cultures, and a Coombs test. A skin biopsy is not always indicated but is helpful if an underlying neoplastic disease is in question. Specific treatment is guided by the underlying disease.

Table 3 Causes of blueberry muffin baby

Extramedullary (Dermal) hematopoiesis	*Congenital infections:* Toxoplasmosis Rubella Cytomegalovirus Herpes simplex Coxsackievirus Parvovirus *Hematological disorders:* Hemolytic disorders (ABO/Rh incompatibility, hereditary spherocytosis, etc.) Twin-twin transfusion Erythroblastosis fetalis
Neoplastic disorders	Transitory myeloproliferative disease Neuroblastoma Langerhans cell histiocytosis Congenital leukemia Congenital alveolar cell rhabdomyosarcoma
Other disorders	Hemangiomatosis Multifocal lymphangioendotheliomatosis Glomuvenous malformations

Infections

Infections in neonatal period can be those that are transmitted vertically, acquired during passage through an infected birth canal, or those that are acquired from external environment after birth.

Staphylococcal scalded skin syndrome Staphylococcal scalded skin syndrome (SSSS) is an inflammatory toxin-mediated generalized desquamating disorder caused by *Staphylococcus aureus*, most commonly by the phage group 2 types 71 and 55 that produce *Epidermolytic toxin A* and/or *B* which target *Desmoglein 1*, an intercellular adhesion molecule leading to acantholysis. SSSS is common in neonates, attributable to the inefficient metabolism and renal excretion of the toxin which is produced at foci of infections like the umbilicus, breast, and conjunctiva. The disease is characterized by erythema and tenderness of the skin which later develops flaccid bullae and exfoliation. SSSS in older children is discussed in the Chapter 48.3. Specific treatment of SSSS involves administration of a penicillinase resistant penicillin analog like flucloxacillin or methicillin, or with an appropriate cephalosporin or sodium fusidate with the route of administration guided by the severity of the disease.

Ecthyma gangrenosum Ecthyma gangrenosum is a potentially fatal illness caused by *Pseudomonas aeruginosa* which is common in the hospital environment and infections are encouraged by widespread use of broad spectrum antibiotics. In the neonatal period, ecthyma gangrenosum is seen in the setting of prematurity or immunodeficiencies. Although, classically ecthyma gangrenosum is a cutaneous manifestation of pseudomonal septicemia, in premature neonates the lesions may be seen at the site of inoculation without bacteremia. The lesions are characterized by tense hemorrhagic vesicles or bullae with a typical violaceous hue that rupture to form ulcers with central necrotic eschar. Anogenital area and extremities are frequently involved sites. Infection is potentially dangerous when it occurs in the setting of septicemia. Treatment is with intravenous antipseudomonal penicillins or third generation cephalosporins. Even with early treatment, mortality is generally high.

Neonatal herpes simplex Majority of neonatal herpes simplex virus (HSV) infection are caused by HSV 1 and 2 that is acquired through contact with an infected birth canal. Intrauterine infection can also occur either through transplacental transmission or ascending infection. HSV infection in newborns is a potentially serious disease with possible mortality. The lesions appear between day 2 and 20 involving the skin and/or mucousa, unless intrauterine infection has occurred when lesions are seen at birth. Isolated or grouped vesicles on an erythematous base are the most common skin lesions that commonly involve the face and scalp. Mucosal lesions present as erosions on the tongue, palate, gingivae and buccal mucosa. Systemic involvement in the form of meningitis may occur when the mortality and long-term complications are more likely even with appropriate antiviral therapy. Early diagnosis and acyclovir therapy prevents dissemination of the infection when it is confined to the skin.

Fetal varicella syndrome Varicella contracted for the first time during pregnancy has a 25% chance of getting transmitted to the fetus and when the infection occurs in early pregnancy, spontaneous abortion occurs or in about 2% or the child is born with a variety of congenital anomalies collectively termed as fetal varicella syndrome. These including hypoplastic limbs (unilateral and involving the lower extremity), cutaneous scarring (often dermatomal), ocular anomalies (chorioretinits, microphthalmia, Horner syndrome) and CNS anomalies (seizures, mental retardation, encephalitis, dorsal radiculitis). Pregnant women exposed to varicella zoster virus for the first time should receive

varicella zoster immunoglobulin (VZIG) which appears only to modify clinical varicella but does not prevent fetal infection. When a nonimmune pregnant woman develops varicella 4 days on either sides of delivery, the infection may be transmitted to the newborn that carries a mortality rate of up to 30%. Such cases should be managed by VZIG.

Candidiasis Congenital candidiasis is a rare condition which reflects maternal *Candida* chorioamnionitis resulting from the ascending infection from the genital tract. Congenital candidiasis is characterized by presence of widespread discrete erythematous macules and papules at birth that progress to vesiculopustular lesions in the next few days. Palmoplantar pustules are regarded as hallmarks of congenital cutaneous candidiasis. The lesions of congenital candidiasis are confined to skin in majority of the cases. However, in preterm neonates mucosal lesions may coexist and in very low birthweight babies such lesions may disseminate systemically. Appropriate anticandidal antifungal therapy is enough for patients with cutaneous lesions only. Amphotericin B is the drug of choice for systemic candidiasis.

Neonatal candidiasis is acquired from an infected birth canal and presents as oral candidiasis, with or without napkin candidiasis presenting as a moist *beefy-red* plaque often with satellite pustules. Neonatal candidiasis is effectively managed by topical antifungals. These have been discussed in Section 33.

Congenital Nevi and Hamartomas

Nevi and hamartomas in the neonatal period are mostly congenital and may occur as isolated lesions or be associated with other anomalies or as features of a syndrome.

Verrucous epidermal nevi These keratinocytic hamartomas present at birth affecting both the sexes equally. Initial appearance is that of a slightly pigmented streak or plaque which becomes darker and verrucous with age. In most of the cases the lesions are linear following the lines of Blaschko; although, a systematized form affecting larger body surface area bilaterally is described. Treatment depends upon the type of the lesion and extent of involvement. Ablative procedures with cryotherapy, carbon dioxide or neodymium-doped yttrium aluminum garnet (Nd:YAG) lasers are probably most effective treatment options than topical retinoids or keratolytics for linear lesions. For systematized or extensive lesions, systemic retinoids are helpful but recurrence is common with discontinuation of treatment.

Nevus sebaceous Nevus sebaceous is an epidermal hamartoma comprised of sebaceous glands presenting at birth with equal gender predilection. Head and neck regions are most commonly involved with a greater tendency to affect the scalp. In the neonatal period, it appears as a circumscribed area of alopecia with a velvety texture and yellowish to fleshy color that remains unaltered until adolescence when it grows and becomes lobulated due to androgen stimulation. Histopathological findings in infancy and childhood are similar to those of verrucous epidermal nevus with sparse immature sebaceous glands. However, presence of cords and buds of poorly differentiated epithelial cells representing primordial pilosebaceous follicles are diagnostic features at this stage. Surgical excision is the treatment of choice.

Congenital smooth muscle hamartoma Congenital smooth muscle hamartoma originates from smooth muscle fibers of the erector pili muscle. It is often congenital, but an acquired form is also seen. It is usually a solitary lesion occurring on the trunk or proximal extremities with predilection for the lumbosacral area. Clinically, it presents as a skin colored or slightly pigmented patch or plaque with hypertrichosis which becomes more obvious and slightly elevated on stroking (the *pseudo-Darier* sign). Histopathologically it is composed of proliferating smooth muscle bundles of the erector pili muscle in the reticular dermis. Surgical excision is curative.

Congenital melanocytic nevi Congenital melanocytic nevi (CMN) are present at birth and are characterized by well-defined deeply pigmented macules or patches that proportionately grow with the child. Depending on the size, CMN are classified into small (< 1.5 cm), medium (1.5–19.9 cm), large (≥ 20 cm) and giant. Large and giant CMN are, associated with malignant potential. Large and giant CMN preferentially involve the lower back (bathing trunk nevus). With age, they develop surface rugosities, hypertrichosis and the color intensifies as well. Satellite lesions also develop. Treatment of small CMN may not be necessary. Management of giant CMN is difficult and involves surgical approaches like serial excision with tissue expansion techniques and skin grafting.

Dermal melanocytoses Dermal melanocytoses refer to the dermal melanocytic nevi which develop as a result of arrest in the migration of melanocytes in their journey from the neural crest to the skin. These include Mongolian spots, nevus of Ota and nevus of Ito.

Mongolian spots They present as ill-defined bluish or slate-gray macules and patches at birth. They appear as a single patch or as multiple lesions commonly involving the lumbosacral region. Extrasacral sites include buttocks, shoulders or extremities **(Fig. 8)**. The color deepens for a period after birth peaking at 2 years of age and then disappears gradually by the first decade. Mongolian spots are also associated with inherited syndromes like phacomatosis pigmentovascularis (types II, IV, and V) and Sjögren-Larsson syndrome. Large and extensive lesions are associated with GM1 gangliosidosis type 1, Hunter and Hurler syndromes, and trisomy 20 mosaicism. Such lesion do not regress until about the second decade and Q-switched ruby laser or Q-switched Nd:YAG lasers have been used successfully in these cases.

Nevus of Ota (nevus fuscoceruleus ophthalmomaxillaris) It is characterized by unilateral bluish-gray macules and patches in a dermatomal pattern affecting the skin supplied by the first and second division of trigeminal nerve. In half of the cases it is congenital and appears during the second decade of life in the rest. The lesions are persistent and may darken at puberty. Q-switched Alexandrite laser is useful for treatment. *Nevus of Ito* is similar to nevus of Ota and involves the acromioclavicular region. It occurs in the areas innervated by the posterior supraclavicular and lateral cutaneous brachial nerves.

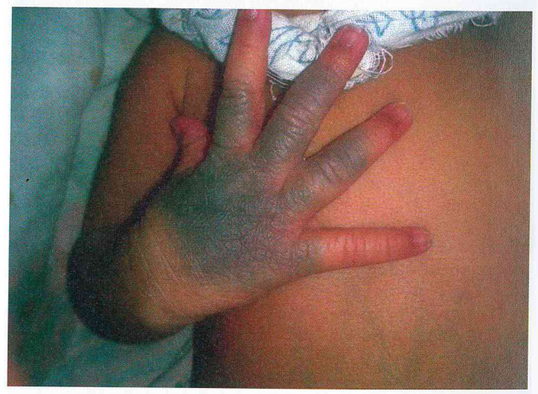

Figure 8 Aberrant Mongolian spot involving the extremity (*Source:* Dr Ahamadrasool Shirasangi, Consultant Pediatrician, Bagalkot, India)

Congenital Defects

Aplasia cutis congenital Aplasia cutis congenita is characterized by a circumscribed area of absent skin with a predilection for the midline of vertex of scalp. It may occur as an isolated defect or in association with other developmental anomalies. Possible causes for this condition include genetic factors, vascular compromise, trauma, teratogens and intrauterine infections. Clinically aplasia cutis congenita may present as erosion, a deep ulcer, a scar, or the most common membranous form in which the defect is covered by a thin, translucent membrane. A rim of long coarse hair may be present along the margin of the defect (hair collar sign), which is a relatively specific marker of an associated neural tube defect. Lesions may appear bullous due to the presence of serous fluid within the membrane. The lesions eventually flatten leaving behind an atrophic scar.

Supernumerary digits These occur as isolated anomalies and show an autosomal dominant inheritance pattern. They appear as small fleshy or warty papules most commonly on the ulnar side of the fifth digit. They are present at birth, often bilaterally. Histologically, they are composed of nerve fiber fascicle. Larger lesions may contain cartilage or a vestigial nail. Surgical excision is advised when removal is desired.

Supernumerary nipples Supernumerary nipples are the most common type of accessory mammary tissue found in 1–6% of the population with equal sex predilection. Although usually sporadic, about 10% of cases are familial. Supernumerary nipples are most commonly seen on the inframammary chest as a small, soft, pink or brown papule, either with a surrounding areola. However, they may be located anywhere along the milk line. Lesions are most often single, but they may be multiple and/or bilateral. Supernumerary nipples are found in several multiple congenital anomaly syndromes, including Simpson-Golabi-Behmel syndrome, cleft lip/palate-ectodermal dysplasia syndrome and tricho-odonto-onychial dysplasia.

Cutaneous markers of spinal dysraphism As the skin and nervous system share an ectodermal origin, coexistence of anomalies of the structures is often seen. Midline cutaneous lesions are seen in more than 80% of cases with closed spinal dysraphism serving as a valuable marker for spinal dysraphism, and, in the majority of patients, they are the finding that leads to the diagnosis. Most of the cutaneous lesions associated with spinal dysraphism are located in the lumbosacral area, reflecting the relative rarity of neural tube defects in the cervicothoracic region. Sacrococcygeal dimples, lipoma in the midline, midline localized hypertrichosis (faun tail) and also midline hemangiomas comprise such markers and their presence at birth must alert the clinicians.

MORE ON THIS TOPIC

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Chapter 48.2
Care of Skin in the Newborn

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The skin acts as a barrier against mechanical, chemical and thermal injuries, penetration of toxins and infections, and prevention of fluid and electrolyte imbalances. It also provides thermoregulation, fat storage and insulation. It provides tactile sensation and promotes immune surveillance. As the neonate is exposed to a dry, external, aerobic world from a sterile, aquatic, uterine environment, the skin undertakes important developmental changes. Also, though the function of the skin barrier is believed to begin in utero, it is actually an ongoing process for up to 12 months after birth. Hence, it is extremely essential to preserve the integrity of the skin in the neonatal age group, and hence the importance of newborn skin care.

The skin of the newborn exhibits certain differences from adult skin **(Table 1)**. Anatomically, the body surface area of the neonate is much higher (700 cm^2/kg) as compared to that of an

Table 1 Structural differences between newborn and adult skin

Skin component	*Adult*	*Full-term newborn*	*Preterm newborn*
Epidermis	Normal	Strong stratum corneum	Few, thin layers of stratum corneum
Dermis	Normal	Thin	Thin
Dermo-epidermal junction	Strong coherence between epidermis and dermis	Weak coherence	Weak coherence
Hair type	Vellus and terminal	Vellus	Lanugo
Sweat glands	Functionally mature	Immature	Immature
Sebaceous glands	Structurally and functionally mature	Mature	Mature
Penetration	Strong barrier impeding infiltration	Penetration and absorption of substances present	Immature barrier function leading to increased infiltration and absorption of substances
pH	5.5	> 6	> 6
Body surface area	x	3x	7x

x, body surface area in adults