Major defects in neonatal skin have serious consequences for the health of the baby, but fortunately these are very rare. Early diagnosis of the genodermatoses can be life-saving. Birthmarks, also known as naevi, are usually first noted in neonates, and can be very alarming for parents. A variety of infections can occur because of relative immune compromise in the neonate. However, most skin changes at this stage reflect adaptation to new and sometimes hazardous external conditions of heat, humidity and cold.

**CAFÉ-AU-LAIT MACULE**

This congenital, homogeneous tan-brown macule or patch is well circumscribed and varies in size from a few millimetres to several centimetres (Fig. 1), with an average of 1 - 3 cm in diameter. It is found anywhere except on the scalp, eyebrows, palms and soles. Although usually sporadic and of no significance, it can be associated with genodermatoses such as neurofibromatosis type 1.

**CANDIDIASIS**

**Congenital candidiasis**

Infection with *Candida albicans* in the first weeks of life can be congenital, following infection from candidal chorioamnionitis due to ascending infection from the mother’s genital tract. This affects the lungs and skin, causing pneumonia and a maculopapular erythematous rash, which progresses to vesicles and pustules, sparing the mouth and napkin area. It is a serious disease with significant morbidity.

**Neonatal candidiasis**

Neonatal candidiasis occurs after the first week and is acquired during delivery. It presents as oral candidiasis (thrush), with or without napkin involvement. The skin on the buttocks and genitalia is fiery red with a fringed irregular edge, and the rash can spread to other flexures. Pustules are present and rupture easily, leaving shallow erosions. The classic feature of satellite papules and pustules at the edge of the area is present (Fig. 2). Treatment is with oral anticandidal...
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The collodion baby is susceptible to hypothermia, infection and dehydration, and should be nursed in an incubator with high humidity.

Mastocytosis is a collective term for a spectrum of disorders characterised by the abnormal proliferation of mast cells.

agents, e.g. nystatin suspension (Mycostatin, Nystacid suspension) 1 ml qid or miconazole gel (Daktarin oral gel), together with anticandidal ointments like nystatin (Mycostatin, Nystacid ointment), applied qid.

COLLODION BABY

At birth the neonate is bright red and covered in a thick translucent, shiny membrane resembling collodion. The lips protrude, and there is ectropion and flattening of the nose and ears. In a few days the membrane dries and cracks, resulting in bleeding fissures (Fig. 3). The membrane peels off and the skin normalises over weeks or months. This can be a sporadic occurrence, but can also be the presenting sign of a variety of congenital ichthyoses, especially lamellar ichthyosis and non-bullous congenital ichthyosiform erythroderma. In these cases the typical morphology will gradually appear as the collodion disappears. The collodion baby is susceptible to hypothermia, infection and dehydration, and should be nursed in an incubator with high humidity. Emollients like emulsifying ointment or liquid paraffin are applied, and sepsis is aggressively treated. Specialist referral is usually appropriate to identify a possible chronic condition.

CUTIS MARMORATA

This is a marble-like, bluish-purple appearance of the skin of the trunk and extremities (Fig. 4). It is a physiological change that invariably disappears in a few weeks, but can be aggravated by exposure to cold. Persistence of the changes warrants examination for chromosomal abnormalities or hypothyroidism.

ERYTHEMA TOXICUM

Occurring in 20 - 60% of term infants, erythema toxicum is less common in premature or small neonates. Lesions usually develop within 48 hours of birth and comprise blotty erythematous macules with superimposed papules or pustules 1 - 2 mm in diameter. The face, scalp and anterior chest are principally affected, and the eruption waxes and wanes for a few days and then disappears. The neonate is well and no treatment is required. In florid cases a pus swab to exclude Staphylococcus aureus infection is prudent.

Congenital epidermolysis bullosa is a group of inherited skin disorders presenting with fragile skin, which blisters at the slightest shearing. The first blisters are from the birth process and early handling after birth, and are much less obvious with caesarean section than with vaginal delivery (Fig. 5). Mucous membranes and nails can also be involved. There are 3 main groups, each with dominant and recessive types and a wide spectrum of severity. Typical features of specific types are only apparent later in infancy, and it is difficult to prognosticate in neonates. The work-up includes biopsy of apparently unaffected skin for light and electron microscopy to elucidate the level of the split in the skin. Management involves avoidance of all frictional stress, together with non-adherent dressings like Mepitel or Jelonet for blisters. Attention to nutrition and secondary bacterial infection is important, and the family should be referred for genetic counselling. Some types of epidermolysis bullosa improve with age, while others are progressive or fatal.
**IMPETIGO**

The neonate is particularly susceptible to bullous impetigo, caused by certain strains of *S. aureus*. The infection is usually acquired during delivery, but the lesions appear after 2 weeks. Flaccid blisters with clear fluid rupture to leave superficial erosions and crust (Fig. 6). The perineum, peri-umbilical and neck areas are common sites. Ordinary impetigo contagiosa with pustules and thick honey-coloured crusts can also occur in neonates. A pus swab for MC+S confirms the diagnosis. Both types of impetigo should be treated with systemic antibiotics, as there is a risk of septicaemia and complications. Suitable agents include cloxacillin, co-amoxiclav and cephalosporins.

![Fig. 6. Neonatal bullous impetigo.](image)

**HERPES SIMPLEX**

Neonatal herpes simplex infection is a serious disease with a high mortality. Infection is acquired by different routes, most commonly from contact with the mother’s active genital lesions of herpes simplex virus type 2. Lesions occur 2 - 20 days later, and consist of grouped vesicles on the scalp, face or trunk, with greasy scales and yellow-brown papules. Skin biopsy confirms the diagnosis, and areas of purpura within the rash are a clue. Bone, lymph node, liver and lung involvement can occur. Multisystem disease is grave and requires systemic chemotherapy. Other forms present as scattered or solitary brownish-red nodules. These types are usually self-limiting.

![Fig. 7. Neonatal herpes simplex.](image)

**LANGERHANS CELL HISTIOCYTOSIS**

Langerhans cell histiocytosis is a rare reactive condition. The most characteristic presentation is a seborrhoeic dermatitis-like eruption on the scalp, neck, face and trunk, with greasy scales and yellow-brown papules. Skin biopsy confirms the diagnosis, and areas of purpura within the rash are a clue. Bone, lymph node, liver and lung involvement can occur. Multisystem disease is grave and requires systemic chemotherapy. Other forms present as scattered or solitary brownish-red nodules. These types are usually self-limiting.

![Fig. 8. Mastocytosis.](image)

**MASTOCYTOSIS**

Mastocytosis is a collective term for a spectrum of disorders characterised by the abnormal proliferation of mast cells. Urticaria pigmentosa is the most common type and presents with generalised small, pruritic, brown-red macules, papules or nodules (Fig. 8). The lesions resemble small melanocytic naevi, and become inflamed on stroking. Diagnosis is confirmed on skin biopsy, and treatment consists of the administration of antihistamines. The condition usually resolves spontaneously.

**MELANOCYTIC NAEVI**

Melanocytic naevi are very common at birth, presenting as dark-brown macules, papules or nodules at a single site. The initial size is highly variable, and small lesions are commonest. They grow in proportion to the rest of the body, reaching their final size in later adolescence. Congenital melanocytic naevi are generally larger and more hairy than their acquired counterparts, which occur later in infancy or childhood. Rarely congenital melanocytic naevi are huge (Fig. 9) and associated with CNS melanocytosis. This can cause epilepsy and hydrocephalus. The malignant potential of small and medium-sized congenital melanocytic naevi is not clear, but probably extremely low. Nevertheless many authorities recommend elective excision of such lesions before adulthood.

![Fig. 9. Giant congenital melanocytic naevus.](image)

**MILIARIA**

Miliaria is caused by obstruction of the sweat duct. Superficial obstruction causes miliaria crystallina, presenting
as a myriad of tiny dewdrop-like vesicles on the head, neck and trunk. This is usually caused by overdressing and overheating, or from resolution of a high fever. No treatment is necessary apart from avoiding excessive clothing. Deeper obstruction causes milia rubra that presents as itchy red papules or pustules on the forehead, upper trunk and arms. This so-called ‘heat rash’ is invariably caused by overdressing or an extremely humid environment. Cooling by fan or immersion in tepid water can be helpful.

**SEBORRHEIC DERMATITIS**

Present at or shortly after birth, the most common presentation is cradle cap with greasy scales on the scalp (Fig. 10). This can resolve without further problems, or it can spread to the forehead, eyebrows, perioral region, retroauricular creases and neck, causing erythematous macules and papules with greasy overlying scale. Weeping can occur, together with secondary impetigo or candidiasis. Well-circumscribed moist erythema can occur in the axillae, elbow and knee creases and the groins. Later a dry scaling supervenes, and postinflammatory hypopigmentation is marked in darker skins. Despite the sometimes severe appearance of the rash of neonatal seborrhoeic dermatitis, there is little or no itching and the child is undisturbed. This helps to differentiate the condition later from atopic dermatitis. Seborrhoeic dermatitis is usually self-limiting, and disappears within a few months. Some cases persist a year or two. In the setting of immune compromise a steadily worsening course occurs, often ending in erythroderma. Treatment of seborrhoeic dermatitis consists of mild corticosteroid lotions, creams and ointments such as 1% hydrocortisone or diluted betamethasone valerate. Topical antifungal and antibiotic agents are useful in more severe cases. Cradle cap can be resistant to treatment and a useful application is 2% sulphur in emulsifying ointment, which is left on overnight and washed off in the morning with baby shampoo.

**VASCULAR NAEVI**

The reddish macules, patches or nodules of vascular naevi are very common birthmarks. The commonest type is the stork bite or salmon patch, more correctly known as naevus simplex. It consists of dilated dermal capillaries that are presumed to be the result of persistent fetal circulation. Common sites include the nape of the neck, the glabella and the eyelids. The lesions are pink to red in colour, blanch easily on pressure, and darken with crying or fever. Most of these naevi fade gradually by the age of 3 years. Nape lesions quite often persist into adulthood. Port-wine stains (naevus flammeus) are persistent vascular malformations that are present at birth, mostly on the face and neck (Fig. 11). They can be very large, and associated with other congenital abnormalities. With age the lesions darken and thicken, becoming purple and hyperplastic. Laser therapy has become a viable treatment. Strawberry naevi (infantile haemangioma) cause considerable distress to parents. These benign neoplasms present at birth or in the first few weeks, principally on the head and neck. Initially they can resemble flat vascular naevi, like port-wine stains or naevus simplex, but they proliferate and grow in size during the first year, becoming bright red, raised, well-circumscribed tumours. In the succeeding years there is gradual involution. Infantile haemangiomas do not require intervention unless they interfere with functions like sight, feeding and breathing. In these cases systemic corticosteroids are given in the early proliferative phase, together with referral for surgery or laser therapy if available.

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