Pediatric Dermatology.

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SKIN OF THE NEWBORN

- Skin is a dynamic complex organ, which performs several vital functions.
- It forms a physical barrier between the organism and the environment.
- It provides UV protection, prevents invasion of pathogens, and regulates body temperature and sensory perception.

Romanelli M, 2015

SKIN OF THE NEWBORN

- The development of the skin barrier increases with gestational age, and the epidermal maturation is complete at 34 weeks of age.
- Functional and structural skin maturation starts at the moment of delivery and ends in the first year of life.
- In full-term newborns, this process begins immediately after birth, while in preterm newborns by 2–3 weeks after birth, the skin is comparable to a full-term newborn's skin.

Romanelli M, 2015; Lee LW, 2019

SKIN OF THE NEWBORN

- The capacity to sweat is correlated with gestational age and there is a tendency to total anhidrosis in the preterm newborn in the first days after birth.
- Infants of less than 36 weeks of age start to sweat after thermal stimulation during the second week of life, but the intensity of the sweat response depends on gestational age and thermoregulation is initially low.

Romanelli M, 2015; Verbov J, 2000

BENIGN TRANSIENT SKIN LESIONS

BENIGN TRANSIENT SKIN LESIONS

VERNIX CASEOSA

- This is seen in the term and mildly preterm infant at birth.
- It is cheesy-white in appearance with material derived from fetal sebaceous glands and may be either widespread or localized to skin folds..

Verbov J, 2000

BENIGN TRANSIENT SKIN LESIONS

DESQUAMATION

- In normal term neonates superficial skin peeling may be seen over the hands and feet or with a more widespread distribution in the first days of life.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

ACROCYANOSIS

- Peripheral vasoconstriction is common and rapidly improves when the infant is warmed.

Verbov J, 2000

BENIGN TRANSIENT SKIN LESIONS

CUTIS MARMORATA

- This reticulate vascular pattern is a physiological response to chilling on the trunk and limbs in normal infants.
- The dilatation of capillaries and small venules disappears with warming.

Romanelli M, 2015

BENIGN TRANSIENT SKIN LESIONS

ERYTHEMA TOXICUM NEONATORUM

- This self-limiting condition of unknown cause is the most common eruption in the neonate.
- It usually appears on the second or third day of life or at some other time in the first 2 weeks.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

ERYTHEMA TOXICUM NEONATORUM

- The lesions may be scanty or extensive, they are maculopapular or pustular.
- Erythema surrounds the 1–3 mm pustules.
- It is seen particularly in term infants and is rare in premature babies.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

ERYTHEMA TOXICUM NEONATORUM

- Lesions show many eosinophils and there may be blood eosinophilia.
- Individual lesions may persist for hours or days.
- No therapy is required. There is no sexual or racial predisposition.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

TRANSIENT NEONATAL PUSTULOSIS

- This condition was originally described as transient neonatal pustular melanosis.
- Small superficial sterile vesicular and pustular lesions appear over face, limbs and back.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

TRANSIENT NEONATAL PUSTULOSIS

- Lesions are often present at birth and may continue to appear for many weeks.
- Pustules subside leaving either a brownish crust or a scaling edge.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

TRANSIENT NEONATAL PUSTULOSIS

- Post inflammatory hyperpigmentation may be seen in black babies.
- Smears of fluid from lesions show neutrophils rather than eosinophils with negative bacterial culture.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

TRANSIENT NEONATAL PUSTULOSIS

- The condition is more common in black babies.
- The cause is unknown.
- There is no systemic upset.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

SEBACEOUS GLAND HYPERPLASIA

- Multiple yellow papules appear over nose, cheeks and upper lips, which resolve in a few weeks.
- They are a manifestation of maternal androgen stimulation.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

MILIA

- These small white 1–2 mm follicular cysts (milk spots) over the face result from retention of keratin and sebaceous material.
- The condition clears in the first month of life.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

EPITHELIAL (EPSTEIN'S) PEARLS

- Multiple white 1–2 mm cysts may be seen within the mouth around the junction of the soft and hard palate in the midline in most neonates.
- They disappear spontaneously.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

MILIARIA

- Multiple small pinhead-size clear vesicles 1–2 mm in diameter appear particularly over neck and chest in an over-swaddled infant.
- There is no erythema.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

MILIARIA

- Lesions are usually seen not so much at birth as at 2–3 weeks of age.
- It indicates sweat retention as a result of blockage of the intraepidermal portion of eccrine sweat ducts.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

MILIARIA

- Avoidance of excessive heating and humidity and looser lightweight cotton clothes aid rapid resolution.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

LANUGO HAIR

- Lanugo hair is usually shed in utero at 7 or 8 month's gestation.
- Pre-term infants may show profuse downy lanugo hairs on face, limbs and trunk that will be shed in the first few months of life.

Verbov J, 2000; Hardaway CA 2019

BENIGN TRANSIENT SKIN LESIONS

LANUGO HAIR

- Infants of less than 26 weeks' gestation are born with transparent, friable skin without lanugo hair.

Verbov J, 2000; Hardaway CA 2019

PIGMENTARY ABNORMALITIES

DEVELOPMENTAL ABNORMALITIES

MONGOLIAN SPOT

- Blue to black in color flat patches.
- They are seen in about 90% of black infants, 70% of oriental and Hispanic infants and in about 5–10% of white Caucasian infants.
- They are seen particularly over the lower back but can be more widespread and can occur over limbs and face.
- They do disappear with the time but this may take many years..

Verbov J, 2000, Kirkorian A, 2021

DEVELOPMENTAL ABNORMALITIES

CAFE´-AU-LAIT MACULES

- These well-defined macules are seen in healthy children often as single lesions.
- Multiple lesions greater than 0.5 cm in diameter should lead to suspicion of neurofibromatosis type 1 (NF-1) and six or more are presumptive evidence of NF-1.

Verbov J, 2000, Kirkorian A, 2021

DEVELOPMENTAL ABNORMALITIES

HYPOMELANOTIC MACULES

- White spots or hypopigmented macules occur in 0.2–0.3% of all neonates.
- They are usually of no clinical significance, and many disappear with age. However, it should be remembered that most babies with tuberous sclerosis have such macules which may be the earliest sign of the autosomal dominant disorder.

Verbov J, 2000, Kirkorian A, 2021

DEVELOPMENTAL ABNORMALITIES

CONGENITAL MELANOCYTIC NEVI

- Congenital melanocytic nevi are pigmented lesions that are usually present a birth. They are generally benign, but a small percentage (especially the larger ones) can potentially transform into malignant melanoma.

Verbov J, 2000, Kirkorian A, 2021

DEVELOPMENTAL ABNORMALITIES

CONGENITAL MELANOCYTIC NEVI

- They are classified according to their estimated adult size as small (less than 1.5 cm), medium (1.5 to 20 cm), large (20 to 40 cm), and giant (>40 cm). This classification is essential as the size of nevi is one of the main risk factors of melanoma development and neurological involvement.

Romanelli M, 2015

DEVELOPMENTAL ABNORMALITIES

CONGENITAL MELANOCYTIC NEVI

- Small congenital nevi occur in 1 in 100 births
- Medium congenital nevi occur in 1 in 1000 births
- Giant congenital melanocytic nevi are much rarer 1 in 20,000 live births

Verbov J, 2000, Kirkorian A, 2021

DEVELOPMENTAL ABNORMALITIES

CONGENITAL MELANOCYTIC NEVI

- At birth, they usually present as pigmented macules or slightly raised oval papules or plaques, which can be initially mistaken for café-au-lait macules.
- They usually show color darkening over time and tend to end up having a well-defined edge.

Romanelli M, 2015

DEVELOPMENTAL ABNORMALITIES

CONGENITAL MELANOCYTIC NEVI

- They can display a wide variety of colors ranging from light brown to black.
- Hypertrichosis is a common finding and is usually accompanied by perifollicular hypopigmentation.

Verbov J, 2000, Kirkorian A, 2021

DEVELOPMENTAL ABNORMALITIES

CONGENITAL MELANOCYTIC NEVI

- Neurocutaneous melanosis is a disorder where CMNs are associated with melanocyte proliferation in the CNS.
- Those with larger CMN and satellite lesions are especially at risk.

Romanelli M, 2015

DEVELOPMENTAL ABNORMALITIES

CONGENITAL MELANOCYTIC NEVI

- They may be asymptomatic, or they may have neurological symptoms, including seizures, cranial nerve dysfunction, or signs and symptoms of increased intracranial pressure.
- Symptomatic patients usually have an ominous prognosis.

Verbov J, 2000, Kirkorian A, 2021

DEVELOPMENTAL ABNORMALITIES

CONGENITAL MELANOCYTIC NEVI

- Dermoscopy can provide the physician with very useful information in distinguishing CMN from other cutaneous lesions and in diagnosing early malignant transformation.
- Whenever there is clinical suspicion of melanoma, an urgent biopsy with histopathological examination must be performed.

Romanelli M, 2015

DEVELOPMENTAL ABNORMALITIES

CONGENITAL MELANOCYTIC NEVI

- Patients with large or giant CMN, especially when satellitosis is present, should undergo prompt evaluation (within the first 4 to 6 months of life) with gadolinium-enhanced magnetic resonance imaging (MRI) to rule out neurological involvement.
- MRI can detect melanocytic deposits.
- The temporal lobe (especially the amygdala) is the most commonly affected brain region.

Verbov J, 2000, Kirkorian A, 2021

VASCULAR ABNORMALITIES

VASCULAR ABNORMALITIES

NAEVUS SIMPLEX

- Naevus simplex is a common, benign capillary vascular malformation.
- It presents at birth as a pink or red patch and is most often observed on the nape of the neck, eyelid, or glabella.

Verbov J, 2000

VASCULAR ABNORMALITIES

NAEVUS SIMPLEX

Naevus simplex is also called:

- Salmon patch
- Stork bite (when on the nape of the neck)
- Angel's kiss (when on the glabella and eyelids)
- Naevus flammeus simplex.

Verbov J, 2000

VASCULAR ABNORMALITIES

NAEVUS SIMPLEX

- Naevus simplex affects infants of all races.
- it occurs in approximately 40% of Caucasian infants and less often in darker-skinned infants.
- It affects male and female infants equally.

Verbov J, 2000

VASCULAR ABNORMALITIES

NAEVUS SIMPLEX

- Naevus simplex is present at birth as either single or multiple, flat, pink or red patches, typically occurring with an indistinct, irregular border and blanching on compression.
- Naevus simplex becomes more red with crying, fevers, breath holding, straining, vigorous physical activity, and changes in ambient temperaturę.
- They tend to become less prominent as the child gets older.

Verbov J, 2000

VASCULAR ABNORMALITIES

NAEVUS SIMPLEX

- The skin lesions usually occur at midline and can be bilateral and symmetrical in presentation.
- They are most commonly found on the nape of the neck, eyelids, and glabella, but they may also involve the forehead, scalp, nose, lips, and back.
- Naevus simplex is not painful or itchy.

Verbov J, 2000

VASCULAR ABNORMALITIES

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Romanelli M, 2015

VASCULAR ABNORMALITIES

- **NAEVUS FLAMMEUS**

- Naevus flammeus is a capillary vascular malformation found in 0.3% of newborn infants.
- The lesions present as pink, red or purple patches and can be localised, extensive or multiple.
- Unlike naevus simplex, naevus flammeus tends to be unilateral, often persisting, darkening and thickening with increasing age

Verbov J, 2000

VASCULAR ABNORMALITIES

NAEVUS FLAMMEUS

- Most port-wine stains occur as an isolated lesion but they can be part of a more widespread vascular disorder.
- An association of facial port-wine stain with congenital glaucoma should be appreciated, particularly as the glaucoma is usually asymptomatic early in life.

Verbov J, 2000

VASCULAR ABNORMALITIES

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- An association of facial port-wine stain with congenital glaucoma should be appreciated, particularly as the glaucoma is usually asymptomatic early in life.

Verbov J, 2000

VASCULAR ABNORMALITIES

INFANTILE HEMANGIOMAS

- Infantile hemangiomas (IHs) are the most common benign vascular tumors in infancy.
- Usually, IHs are not present after birth.
- Mostly IHs occur within a period of 1–2 weeks after birth.
- The growth of infantile hemangiomas is divided into three phases: the rapid proliferation phase, the plateau phase, and the involution phase.

Romanelli M, 2015

VASCULAR ABNORMALITIES

Infantile hemangiomas

- Infantile hemangiomas are the most common vascular tumors of infancy; incidence of IHs ranges from 4% to 5%, but can be up to 20% in premature infants.
- They are more common among white infants and have a two to three times higher prevalence among females.
- The incidence also increases with the low birth rate (up to 30% of infants born weighing < 1 kg are affected).

Kurzeja M, 2022

VASCULAR ABNORMALITIES

Infantile hemangiomas

- The clinical classification is based on their depth (superficial, mixed and deep IHs) and pattern of involvement (focal, multifocal, segmental, intermediate).

Kurzeja M, 2022

VASCULAR ABNORMALITIES

Infantile hemangiomas

- Superficial IHs are localized within the superficial dermis and are clinically visible as a red, firm lobulated plaque. The superficial IHs, previously termed “strawberry hemangiomas” or “capillary hemangiomas”, are the most common.
- While deep IHs are localized in the deeper part of the dermis and/or in the subcutaneous tissue visible as a skin-colored or bluish subcutaneous nodule and were previously termed “cavernous hemangiomas”.
- Mixed IHs involve both layers of the skin and exhibit clinical features of superficial and deep IH.

Kurzeja M, 2022

VASCULAR ABNORMALITIES

Infantile hemangiomas

- The focal IHs are well-defined single lesions within the one anatomic area.
- Segmental IHs clinically appear as linear lesions with a geographic distribution, covering larger anatomic regions.

Romanelli M, 2015

VASCULAR ABNORMALITIES

Infantile hemangiomas

- Segmental IHs occur more likely among female infants.
- Also segmental IHs more frequently need intensive and prolonged therapy
- Segmental IHs localized on the face or in the lumbosacral area may be associated with PHACE and LUMBAR.

Kurzeja M, 2022

VASCULAR ABNORMALITIES

Infantile hemangiomas

- Multiple IHs defined as either more than at least five and, in other publications, more than ten lesions occur in up to 20% of infants.
- They are more common among infants from multiple gestation pregnancies.
- Multiple IHs are associated with an increased risk of visceral involvement, mostly in the gastrointestinal tract and liver.

Romanelli M, 2015

VASCULAR ABNORMALITIES

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Kurzeja M, 2022

VASCULAR ABNORMALITIES

Infantile hemangiomas

- IHs are mostly self-limiting and do not require any treatment.
- However in some cases IHs may be associated with complications such as ulceration, obstruction of airways, functional impairment or disfigurement, in these cases treatment with oral propranolol is the first line of treatment.

Romanelli M, 2015

VASCULAR ABNORMALITIES

Infantile hemangiomas

- The first-line agent for IHs requiring systemic therapy is a nonselective β-blocker – propranolol. Several randomized, controlled trials confirmed the efficiency and safety of propranolol in the management of His
- In 2014, the US Food and Drug Administration and the European Medicines Agency approved propranolol for treatment of infantile hemangiomas.

Kurzeja M, 2022

VASCULAR ABNORMALITIES

Infantile hemangiomas

- An initial dose of oral propranolol 1 mg/kg per day in 2–3 doses with escalation to maximum dose 3 mg/kg per day.
- The response rate after 6 months' therapy with oral propranolol at a dose of 2–3 mg/kg/day is 96–98%.

Romanelli M, 2015

VASCULAR ABNORMALITIES

Infantile hemangiomas

- The therapy with oral propranolol should be started within the proliferative phase, although it may still lead to improvement when it is initiated after 9–12 months of age.
- The therapy lasts until 12 to 18 months of age.

Kurzeja M, 2022