Neonatal Dermatology:
Bumps, Rashes and Birthmarks

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Objectives

Physiologic / transient skin findings in the newborn

Blisters and pustules
  • Benign and transient
  • Infectious

Birthmarks
  • Pigmented
  • Vascular
  • Spinal dysraphism
Physiologic / Transient
Physiologic Phenomena

Cutis Marmorata

Acrocyanosis

• Vasomotor instability
• Resolves with re-warming
Physiologic Phenomena

Milia

- 50% newborns
- Small, smooth white papules
- Birth-infancy
- Face most common
  - Nipple
  - Penis
Physiologic Phenomena

Sebaceous Gland Hyperplasia

- White-yellow papules
- No erythema
- Face
  - Nose / upper lip
- Hormonal stimulation
- Less common in preterm
Physiologic Phenomena

Desquamation

- Full term
  - 24-48 hrs
- Post-term
  - At birth
  - More significant
- Pre-term
  - 2-3 weeks of life
Physiologic / Transient

- Cutis marmorata
- Acrocyanosis
- Milia
- Sebaceous gland hyperplasia
- Desquamation
Blisters & Pustules

Transient

Infectious
Blisters & Pustules

**Transient**
- Erythema toxicum neonatorum
- Transient neonatal pustular melanosis
- Miliaria
- Neonatal cephalic pustulosis

**Healthy**
- No fever
- Feeding well
- Growth / development
Erythema Toxicum Neonatorum

- Rare in preterm or <2500 gr
- Etiology unknown

50% term newborns

- Start 24-48 hrs
- Wax/wane 7 days

Blisters & Pustules
Transient
Erythema Toxicum Neonatorum

Clinical

- Wheals, papules, pustules, macules
- Face, trunk, proximal extremities, buttocks
- Never on palms and soles
- “flea-bitten” appearance
Erythema Toxicum Neonatorum

Healthy

- No further work up

Other tests

- Peripheral eosinophilia
- Eosinophils on Wright stain
Blisters & Pustules
Transient

Transient Neonatal Pustular Melanosis

5% full term black

<1% white pts

Etiology unknown
Transient Neonatal Pustular Melanosis

Clinical

- Pustule → hyperpigmented macule
- Characteristic collarette of scale
- Forehead, chin, behind ears, neck, back
- May involve palms and soles
Transient Neonatal Pustular Melanosis

Healthy

• No further work up

Other tests

• Neutrophils on Wright stain
• Negative bacterial culture
# Blisters & Pustules

## Transient

<table>
<thead>
<tr>
<th>ETN</th>
<th>TNPM</th>
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<tbody>
<tr>
<td>• 24-48 hrs.</td>
<td>• At birth</td>
</tr>
<tr>
<td>• Erythema</td>
<td>• No erythema</td>
</tr>
<tr>
<td>• Palms and soles never involved</td>
<td>• Palms and soles may be involved</td>
</tr>
<tr>
<td>• Resolve without sequelae</td>
<td>• Resolve with hyperpigmentation</td>
</tr>
<tr>
<td>• Eosinophils</td>
<td>• Neutrophils</td>
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</table>
Miliaria

- Obstruction of sweat duct
- 15% neonates
- Warm weather, fever
- Small, non-inflammatory vesicles
- Erythematous vesicles / pustules
- Face, neck and trunk
Blisters & Pustules
Transient

Miliaria crystallina
- Small clear vesicles
- No redness

Miliaria rubra
- “heat rash”
- Red papules / vesicles
Blisters & Pustules
Transient

Miliaria

Location

- Trunk / face / neck
- Warm environment
- Healthy
Neonatal Cephalic Pustulosis

Aka neonatal acne, baby acne

20% newborns

Starts 2nd-3rd week

Malassezia yeast
Neonatal Cephalic Pustulosis

Clinical

- Pustules & red papules
- Forehead, cheeks, chin, neck, upper trunk
- NO comedones (black heads)
Neonatal Cephalic Pustulosis

- Resolves within weeks – few months
- Treatment with 2% ketoconazole cream
Blisters & Pustules

Infectious

• Bacterial
• Viral
• Candida
• Scabies
Blisters & Pustules
Infectious / Bacterial

Bacterial

Staphylococcus aureus

Localized skin infection
Systemic infection
Localized bacterial infection → Impetigo
Blisters & Pustules
Infectious / Bacterial

Bullous Impetigo

- Coagulase + Staph aureus
- Localized SSS
- Pustules / large vesicles
- Neck / abdomen / diaper
- +/- systemic symptoms
Blisters & Pustules
Infectious / Bacterial

Staphylococcal Scalded Skin Syndrome

- SSSS
- Same strain that causes bullous impetigo

Infection source
- Conjunctivae
- Nasopharynx
- Omphalitis

Toxin
- Exfoliative

Skin
- Blistering
- Desquamation
Blisters & Pustules
Infectious / Bacterial

Staphylococcal Scalded Skin Syndrome

• Ill-appearing
• Generalized redness
• Flaccid blisters and desquamation
Blisters & Pustules
Infectious / Bacterial

Staphylococcal Scalded Skin Syndrome

• Secondary infection
• Fluid / electrolyte imbalance
• Temperature instability
• Mortality of 4%
Blisters & Pustules
Infectious / Bacterial

Bullous impetigo
- Localized skin infx
- Skin Cx positive

SSS
- Extracutaneous infx
- Skin Cx negative
Blisters & Pustules
Infectious

Viral

• Herpes Simplex Virus
• Varicella Zoster Virus
• Cytomegalovirus
Blisters & Pustules
Infectious / Viral

Neonatal Herpes Simplex

- 70% cases HSV-2
- Maternal primary genital infection
  - Highest risk (50%)
- Recurrent maternal infection
  - 5% risk
- Perinatal
Blisters & Pustules
Infectious / Viral

3 syndromes

- Skin/Eyes/Mouth
- Disseminated
- CNS infection

Significant morbidity / mortality
Blisters & Pustules
Infectious

Bullous impetigo
- Localized
- Larger
- Culture +

HSV
- Clustered
- Smaller
- Culture -
- Viral PCR +
Blisters & Pustules
Infectious / Candida

Candida in newborn

Congenital candidiasis

Intrauterine infection

Neonatal candidiasis

Delivery / postnatal

Thrush Diaper rash
Congenital Candidiasis

*Candida albicans*

At birth or 1st week

Premature / VLBW @ higher risk

Papules → pustules and scaling

Anywhere on body (palms and soles)
Blisters & Pustules
Infectious

Candidiasis
• Palms / soles
• Not transient
• Culture +

ETN
• No palms / soles
• Transient
• Culture -
Blisters & Pustules
Infectious / Scabies

Scabies

*Sarcoptes scabiei*

Hypersensitivity reaction

After 3-4 weeks

Infants > newborns
Blisters & Pustules
Infectious / Scabies

Scabies

- Pustules / vesicles / redness / scaling
- Medial feet, wrists, palms, soles
- Irritability, poor feeding, itching
- Younger pts may not have pruritus
Blisters & Pustules

Transient
- Erythema toxicum neonatorum
- Transient neonatal pustular melanosis
- Miliaria
- Benign cephalic pustulosis

Infectious
- Bacterial
- Viral
- Fungal
- Parasites
Birthmarks
Types of birthmark

**Pigment**
- Melanocytic nevi
- Café-au-lait macules

**Vascular**
- Nevus simplex
- Port wine stains
- Hemangiomas
Birthmarks of Pigmentation

Congenital Melanocytic Nevus

Café-au-lait Macules
Congenital Melanocytic Nevus

- Melanocytic nevi present at birth or evident during the first year of life
- 1-6% of neonates

Classified by size (as an adult)

- Small (<1.5 cm)
- Medium (1.5-20 cm)
- Large / Giant (>20 cm)
- 1 in 20,000 births

Classification by size (predicted as adult)

Large CMN in neonate

- > 9 cm on scalp
- > 6 cm on body
Why are they important?

Risk of melanoma

- Depends on the size of the nevus

**Small / Medium**
- <1%
- After second decade

**Large / Giant**
- 5%
- 50% in the first 5 years of life
Large / Giant CMN

- Complex
- Multidisciplinary approach

Risks

- Melanoma
- Neurocutaneous melanosis
- Spinal dysraphism
- Other tumors
What to do?

Small / Medium CMN

- Evaluate for concerning features
- Periodic evaluation
  - Mainly at/after puberty
- Baseline photographs
- Parent education
  - Self-exams
  - Educate on concerning features
  - Sun protection
What to do?

• Complex
• Multidisciplinary
• Risk of melanoma
  • Periodic evaluation first decade
• Neurocutaneous melanocytosis

Large / Giant
Birthmarks of Pigmentation

Congenital Melanocytic Nevus

Café-au-lait Macules
Café-au-lait Macules

- Common birthmark
- At birth or first few years of life (< 6 yo)
- African Americans > Caucasians
Café-au-lait Macules

Clinical:

- Well circumscribed
- Evenly pigmented
- Variable size 1-2 mm to >20 cm
Why are they important?

Association with underlying syndrome

- Neurofibromatosis 1
- Neurofibromatosis 2
- Multiple familial CALM
- Legius sdr
- McCune-Albright sdr
- Ring chromosome sdr
- LEOPARD sdr
- Cowden sdr
- Banayan-Riley-Ruvalcaba
- Constitutional mismatch repair def sdr

NF-1
## Café-au-lait Macules / Epidemiology

<table>
<thead>
<tr>
<th></th>
<th>Neonates</th>
<th>School-aged</th>
</tr>
</thead>
<tbody>
<tr>
<td>Single</td>
<td>2.5%</td>
<td>22-36%</td>
</tr>
<tr>
<td>3 or more</td>
<td>1-2%</td>
<td>1-2%</td>
</tr>
</tbody>
</table>

![Nationwide Children's Hospital](Nationwide.png)

![Ohio State University](OhioState.png)
CALM / Neurofibromatosis-1

6 or more CALM

- Pre-pubertal: > 5 mm
- Post-pubertal: > 15 mm

NF CALM

- Present at birth
- Increase in number in the first 6-10 years
Neurofibromatosis-1 / Diagnostic Criteria

2 or more

- 6 café-au-lait macules
- Axillary or inguinal freckling
- ≥ 2 neurofibromas / 1 plexiform neurofibroma
- Optic pathway glioma
- ≥ 2 Lisch nodules
- Distinctive osseous lesions
- 1st degree relative with NF-1
Neurofibromatosis-1 / Newborn period

Skin
- Café-au-lait macules
- Plexiform neurofibroma

Macrocephaly

Ocular
- Buphthalmos
- Proptosis
- Glaucoma

1st degree relative with NF-1
What to do ≥ 6 CALM’s?

- Examine for other signs
- Examine parents
- Ophthalmologic evaluation
- Genetic evaluation
Vascular Birthmarks

Nevus Simplex

Port Wine Stain

Hemangiomas
Vascular Birthmarks

- Nevus Simplex
- Port Wine Stain
- Hemangiomas
Capillary Malformations

Nevus simplex

Port wine stain
Nevus Simplex

Common birthmark

Other names:

- Salmon patch
- Angel’s kiss
- Stork bite

Present in 30-40% newborns
Nevus Simplex

Locations:

- Posterior neck / scalp
- Glabella
- Forehead
- Upper eyelids
- Nose, nasolabial folds, philtrum
Nevus Simplex

Nevus Simplex / What to do?

Reassurance
- 95% facial salmon patches fade 1-2 years

Posterior neck
- May persist indefinitely

Rarely associated with other syndromes
- Large and extensive
Capillary Malformations

Nevus simplex

Port wine stain
Port Wine Stains

Nevus flammeus

Congenital capillary malformation

Incidence

• 0.3% newborns
• 1:1 M:F

Most common on the face

• May occur anywhere
• Unilateral
Port Wine Stains

Static nature

• Darkening
• Bleb formation
• Thickening
Why are PWS important?

Potential psycho-social impact

Potential syndrome associations
Syndromes with PWS

- Sturge-Weber sdr
- Klippel-Trenaunay sdr
- Parkes-Weber sdr
- Phakomatosis pigmentovascularis
- Proteus sdr
- Cobb sdr
- Bannayan-Riley-Ruvalcaba sdr
- Beckwith-Wiedemann sdr
- Von-Hippel-Lindau sdr
- Rubinstein-Taybi sdr
- Wyburn-Mason sdr
- Roberts sdr
- Coat disease
Sturge-Weber Syndrome

Triad

• PWS in the distribution of the V1 trigeminal branch
• Leptomeningeal angiomatosis
  • Seizures in the first year
• Glaucoma
When to worry with facial PWS?

Risk for Sturge-Weber sdr

- V1 distribution
- 7-28%
- Bilateral involvement
- >1 dermatome involved

What to do with facial PWS?

NO V1 involvement

- Consider referral for laser treatment

V1 involvement

- Rule out SWS
- Ophthalmologic evaluation
- Neurology evaluation
  - Timing of imaging is controversial
  - Consider referral for laser treatment
Vascular Birthmarks

- Nevus Simplex
- Port Wine Stain
- Hemangiomas
Epidemiology

- Most common vascular tumor
- 5% infants in US

More common in:

- Females
- Caucasians
- Premature
- Low birth weight

Other risk factors

- Multiple gestation
- Advance maternal age
- Placenta previa / preeclampsia
Life Cycle

SIZE

Proliferative

Involution

1 year

AGE
Clinical classification

Depth
- Superficial
- Deep
- Mixed

Pattern
- Focal
- Multifocal
- Segmental
Clinical classification

Depth of involvement

<table>
<thead>
<tr>
<th>Previous</th>
<th>Current</th>
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<tbody>
<tr>
<td>Strawberry / Capillary / Cherry</td>
<td>Superficial</td>
</tr>
<tr>
<td>Cavernous</td>
<td>Deep</td>
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<tr>
<td></td>
<td>Mixed</td>
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</table>
Clinical Pattern

- **Focal**
- **Multifocal**
  - → potential visceral involvement
- **Segmental**
  - → potential structural anomalies
  - (PHACE, PELVIS, SACRAL, LUMBAR)
Life Cycle

AGE

SIZE

Proliferative phase

- Non linear
- 80% of growth
- By 3 months
- Accelerated phase
- 5.5 – 7.5 weeks

- Early
- Late

Proliferative

1 year
Approach
Most hemangiomas do not require treatment / intervention

Recognize

- Treatment
- Work up
When to treat?

Local complications
- Ulceration
- Infection

Functional problem
- Vision
- Feeding
- Respiratory function

Anatomic distortion
- Psychosocial impact
…further work up?

- Peri-ocular
- “Beard” hemangiomas
- Multifocal
- Segmental
  - Facial
  - Perineal
  - Lumbar
Peri-ocular Hemangiomas

**Eyelids / orbital rim**

**Amblyopia risk**
- > 1 cm diameter
- Ptosis
- Proptosis

**Ophthalmology evaluation**
Fig. 1. Location of hemangiomas, which follow a beard distribution, included in this study.
Airway

Mandibular region
- Pre-auricular
- Mandible
- Lower lip / chin
- Neck

Bilateral

Hemangiomas upper airway / subglottic
- Airway obstruction
“Beard” hemangiomas

- Airway evaluation
- Stridor
- Treatment
  - Airway obstruction
  - Systemic
Multifocal Cutaneous Hemangiomas

- Risk for extra cutaneous hemangiomas (15%)
- US abdomen

Hepatic hemangiomas

- Most are asymptomatic
- Heart failure / liver dysfunction
- Hypothyroidism

Horii et al. Ped Dermatol, 2011
Structural anomalies

Morphology
- Segmental

Location
- Facial
- Lumbo-sacral
- Perineal
Facial Segmental Hemangioma

PHACE(S) syndrome

- **P** → Posterior fossa abnormalities
- **H** → Hemangioma (facial)
- **A** → Arterial anomalies
- **C** → Cardiac defects
- **E** → Eye abnormalities
- **(S)** → Sternal cleft
Segmental Facial Hemangioma

Risk of PHACE with large facial hemangioma

- 30%

Multidisciplinary approach

- CNS imaging
- Cardiovascular work up
- Ophthalmology evaluation

Treatment if no contraindications
PELVIS / SACRAL / LUMBAR

- Spinal dysraphism
- Lipomyelomeningocele
- Anogenital anomalies
- Vesico-renal anomalies
- Bony deformities
Most hemangiomas do not require treatment / intervention
When to treat?

- **Local complications**
  - Ulceration
  - Infection

- **Functional problem**
  - Vision
  - Feeding
  - Respiratory function

- **Anatomic distortion**
  - Psychosocial impact
...further evaluation

Potential complications

• Periocular
• “beard distribution”
• Multifocal (>5)

Structural anomaly

• Segmental facial
• Segmental perineal
• Segmental lumbosacral
Spinal Dysraphism
Occult Spinal Dysraphism

Spinal dysraphism ➔ absent or incomplete fusion of bony elements at midline

Cutaneous findings @ lumbosacral area:

**High Risk**
- Lipomas
- Hypertrichosis
- Hemangioma
- Prominent sacral dimple
- Skin tags
- Aplasia cutis congenita
- Dermoid cyst / sinus

**Low Risk**
- Telangiectasias
- Port wine stain
- Melanocytic nevi
- Gluteal cleft asymmetry
Occult Spinal Dysraphism

2 or more $\rightarrow$ higher risk

Evaluation

- MRI $\rightarrow$ imaging of choice
- US
  - $< 4$ months old
  - Low risk cutaneous findings
  - Small lesions may be missed
  - Operator dependent
Occult Spinal Dysraphism

High Risk

- Lipomas
- Hypertrichosis
- Hemangioma
- Prominent sacral dimple
- Skin tags
- Aplasia cutis congenita
- Dermoid cyst / sinus
Thank you