It’s not just skin deep!

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Conflicts of Interest

- Aldeyra
- Amicus
- Amryt
- Castle Creek
- Pfizer
- Promius
- Regeneron
- Up to Date
- Valeant

NONE WILL APPLY FOR THIS PRESENTATION
Atopic Dermatitis

- Systemic and Behavioral Concerns
  - Child
  - Parent and Family

- Evolving therapies
Atopic Dermatitis/Comorbidities

- Allergic conditions
- Mental health disorders
- Speech disorders
- Hypertension, Peripheral arterial disease
- Some have found no such correlation with cardiovascular disease risk factors when controlling for age, gender, alcohol, smoking

Atopic Dermatitis

- High financial and emotional burden of AD on family and children

- Correlation of personal cost and emotional impact in pts with Medicaid but not with commercial insurance
  - no correlation between annual income and emotional impact

- Larger impact from access issues, social stress, poor support system (“toxic stress”)

J Pediatr 2016;169:284
Peanuts and Eczema… not so nutty!

• LEAP (Learning Early About Peanut Allergy) study

• Panel produced recommendations re application of feeding (peanut) guidelines for infants with Severe, Mild-Moderate, No eczema (or food allergy)

Pediatrics 2017;139:e20164293
# Peanut Allergy Guidelines

**TABLE 1 Summary of Addendum Guidelines**

<table>
<thead>
<tr>
<th>Infant Criteria</th>
<th>Recommendations</th>
<th>Earliest Age of Peanut Introduction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Guideline 1. Severe eczema, egg allergy, or both</td>
<td>Strongly consider evaluation by sIgE or SPT and, if necessary, an oral food challenge. Based on test results, introduce peanut-containing foods.</td>
<td>4–6 mo</td>
</tr>
<tr>
<td>Guideline 2. Mild to moderate eczema</td>
<td>Introduce peanut-containing foods.</td>
<td>Around 6 mo</td>
</tr>
<tr>
<td>Guideline 3. No eczema or any food allergy</td>
<td>Introduce peanut-containing foods.</td>
<td>Age appropriate and in accordance with family preferences and cultural practices</td>
</tr>
</tbody>
</table>

*Pediatrics 2017;139:e20164293*
Atopic Dermatitis... newer therapies (in addition to “grease of choice”)

- **More targeted**
  - topicals
    * began with TCIs
    * PDE4 inhibitor
  - systemics
    * anti-IL4rAb (IL4, 13): dupilumab-↓inflammation, itch, ↑QOL
    * anti-IL31r: nemolizumab-↓itch

- Overall impression of need to investigate, approach variety of immune mechanisms involved... including more consistent engagement/support of the patients

NEJM 2016;375:2335
NEJM 2017;376:878
Case History

- Healthy 9 y/o girl
Alopecia Areata

- Non-scarring hair loss; approx 0.1-0.2% of population
- Inflammatory
- Localized, Total scalp, Total body
- High association with psychological concerns
- Many treatment options
Alopecia Areata - Treatments

- Observation
- Topical or Intraleosomal steroids
- Minoxidil
- Topical immunotherapy
- Methotrexate
- Others... including newer immune modifiers
Janus kinase (JAK) Inhibitors

- Impact pathways involved in messaging from cell membrane to nucleus
  - Cytokines: IFNs, Interleukins...

- Roles in immune and hematopoietic functions
  - Autoimmune disorders, Rejection, Malignancies

- Psoriasis, Atopic dermatitis, Vitiligo, Alopecia Areata

- Tofacitinib, Ruxolitinib, Baricitinib, [Oclacitinib=dogs for management of autoimmune subep blistering disease, atopic dermatitis, itch]

J Am Acad Dermatol 2017;76:736
J Am Acad Dermatol 2017;76:29
JAMA Dermatol 2016;152:490
Janus kinase (JAK) Inhibitors

- **Oclacitinib Cost:**
  - $1.50 - $2.00/pill

- **Tofacitinib Cost:**
  - $3880 (with coupon)
  - $64/pill
Longitudinal Melanonychia

- Rare in white children
- ?New onset or long-standing without change
  - broad distally
  - distal nail dystrophy
- Importance of pigmentation @ proximal nail fold?
- In children, most are benign nevi or lentigines
Longitudinal Melanonychia

- 40 children (< 16 yrs) seen over 3 year period
- Final diagnoses:
  - Nevus – 19
  - Lentigo – 12
  - “Melanonychia” (melanin w/o incr. Melanocytes) – 9
- No melanoma

JAAD 1999;41:17-22
What do you see?
Mastocytosis/Urticaria Pigmentosa
Mastocytosis

- Solitary mastocytoma
- Maculopapular cutaneous mastocytosis
  - Monomorphomorphic
  - Polymorphomorphic
- Diffuse cutaneous mastocytosis

J Allergy Clin Immunol 2016;137:35
Mastocytosis

- Systemic complaints; pruritus, flushing, bronchospasm, gi, bone pain
- Diagnosis usually clinical (Darier’s sign)
- Urinary, plasma histamine
- **Serum tryptase levels**
- ?Bone films, scans
- Bone marrow... ONLY if with ↑ tryptase AND organomegaly

J Allergy Clin Immunol. 2015 December ; 136(6): 1673
Mastocytosis/Management

- ... as indicated
- Avoid precipitating causes
- Antihistamines
- Topical steroids (solitary lesions)
- PUVA
- Epipen
Case History

- 10 y/o admitted for evaluation of facial eruption
- Medication started; minimal response
- Dermatology consultation
Airborne Contact Dermatitis

- Often sudden onset

- “Splash” distribution... not as symmetric as with photocontact

- Afebrile

- +/- pruritus
... one more time
Case History

Two boys with acute onset of fever and skin tenderness
Staphylococcal Scalded Skin Syndrome

- Acute onset of diffuse, often tender erythema
- Spares mucous membranes
- Desmoglein 1 is cleaved by Staph toxins A/B
  - present in granular layer of keratinized epithelium
  - not present in mucous membranes

Bukowski, et al 2010
Compared with ...
Stevens-Johnson Syndrome

- Skin findings of erythema multiforme + two or more mucous membranes
  - Drug
  - Infection; Mycoplasma (atypical)
- Eyes; can be problem later
- Pulmonary; obliterative bronchiolitis... later
- Fluid/Electrolytes
- Wound care
- ? Steroid ? IVIG

Pediatrics 2011;128:723
J Paed Child Health 2011;47:392
Biological Classification of Vascular Birthmarks

- Hemangiomas - vascular lesions marked by endothelial hyperplasia (i.e. enlarge by proliferation) - Glut-1 positive, all stages

- Malformations - lesions with normal endothelial turnover (i.e. true structural anomalies)
Vascular Birthmarks

- 400,000 Babies born in US with vascular anomaly every year
- 8% to 10% of children globally have a vascular anomaly
“Bummer of a birthmark, Hal.”
Seeing Red

- “Hemangioma” used incorrectly 71.3%
  -57.4% skin/soft.tissue (vs skeletal/visceral)

- Incorrect tx with incorrect dx term
  -20%
  -Vs NO incorrect tx with correct dx

Plast Reconstr Surg 2011;127:347
Seeing Red
Incorrect use of Hemangioma

- Peds 60%
- IM 61.4%
- Surg 68.9%
- OB/Gyn 70%
- Path 69%
- Radiol 83.8%
- Derm/Oto/Pedi Surg/Plastic Surg 32.4%

Plast Reconstr Surg 2011;127:347
Seeing Red

- Greater misdiagnosis of malformations than of vascular tumors
- Consider referral of any lesion with unclear dx
  - Complicated IHs (ulcer, L/S, perineal, vital structure, aesthetic concern)
  - CM if lg, multiple, aesthetic concern

J Pediatr Surg 2011;46:1784
Segmental Hemangioma

- Large
- Region or territory of skin
  -(?)Segments c/w risk; e.g.
  Frontotemporal and CNS
- Often plaque-like
- Higher risk complications and
  structural anomalies

Arch Dermatol 2002;138:1567
Arch Dermatol 2004;140:591
Pediatrics 2006;117:698
PHACE(S) Syndrome
PHACE Syndrome - Update

- Workshop: Milwaukee, WI, June 2014
- New comorbidities:
  - Headache, Language/Speech defects
  - Endocrine: Thyroid dysfxn, Hypopituitarism
  - Dental: enamel defects w intraoral IH
- Discussion of risk stratification for stroke
- Expanded dx criteria to include segmental IH of neck, upper trunk, trunk + prox UE

J Pediatr 2016;178:24
Case
Case

- Full term uncomplicated pregnancy
- Consulted regarding skin findings

What do you see?
... what would you do?
Case

- MR and MRA head and neck (normal)
- ECHO (normal)
- Presented to ED at approximately 1 month with stridor
- Admitted by pediatric service for bronchiolitis
- Improved with medical therapy
- Re-admitted twice over first 2-3 months
Case

- Evaluation for PHACE syndrome
- Distribution suggestive of risk for airway hemangiomma
- Aggressive medical therapy
  - Propranolol
  - Low dose prednisolone
Propranolol Guidelines

- Use for infantile hemangioma now well known
- Ongoing discussion re: monitoring of the use of propranolol
- Such monitoring suggested; ? Necessary
- 105 infants (1-5 months) tx’d
  - hospitalized for induction... followed during maintenance
  - 2/105 (each) during induction/maintenance with <5th % SBP/DBP

Pediatr Dermatol 2015;32:802
Propranolol Guidelines

- No pt with clinical hypotension, bradycardia, or other side effects
- Authors suggest BP monitoring not necessary during tx of HIs
- What we do:
  - BP, HR at baseline and again at 7-10 days
  - Have never found any significant drop
  - One child with lethargy... could not associate with any clinical or laboratory change
Vascular Birthmarks – Other therapies

- Sclerotherapy; VM, LM
- Embolization; AVM, ? IH
- Rapamycin*; LM, KHE, ? IH (JID2011;131:2467) - mTor pathway

*Pediatr Blood Cancer 2011;57:1018; Pediatrics 2016;137:1

- Laser; PDL, YAG
- Excision - which IH??

Pediatr Blood Cancer 2011;57:1018
Seeing Red… Vascular Birthmarks, A Collaboration

- **Multidisciplinary** management is key !!!
  - Derm, Plastic surgery, Interventional radiology, Hematology, Genetics, Psychosocial, others

- Early referral !!!

- Research opportunities

- ? Better clinical outcomes
Is the Healthcare Environment Changing?

- Practices recommended by:
  - Cheerfulness of practice
  - Staff’s ability to work together
  - Physician’s overall care provision

- J Pediatr 2016:169:4
Changing Healthcare Models

- Engaging the patient/family
- Virtual care
  - Home monitoring
  - Telemedicine
- Home care
- “Horizontal” care models... Integrated Practice Units (IPUs)... Community integration
- Transitional care
Primary Care Model of Care

- **Patient aligned care team (PACT)**
- Providers, Nurses, Assistants, Office administrators
- Relationships built around patient needs
- True collaboration with all team members
- Coordination... not functioning in “traditional” roles

Uncertainty

- Teaching, historically... and protocols stress the certainty of medical practice

- Challenge of accepting uncertainty can impact patient interactions... testing, for instance

- Need to communicate uncertainty to better engage patients

- Conversation useful to engage patients/families in decisions re: care

NEJM 2016;375:1713
NEJM 2016;375:1918
**Transitional Care…**

What to do with our patients as they become young adults?

- Physicians having problem “separating” after years of affiliation
- Engaging of patients 13-21 yrs with long-term health conditions
- Importance of informing of condition
- Assumption of responsibility of care
- Care coordination
- Need to establish guidelines for transition (see gottransition.org)

Front Pediatr 2016;4:125
Increased Patient/Physician Communication...

Get on the floor with your patient

• Our (physicians) talking (w/pts) is motivator to make earlier and better care decisions

• Relationships are built around the patient’s needs

NEJM 2016;375:1918
Increased Patient/Physician Communication...

*Get on the floor with your patient*