Mineral Deficiency Masquerading as Staphylococcal Scalded Skin Syndrome

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Disclosure

2018 AAP Red Stick Potpourri

The listed authors have documented that she (or he) has nothing to disclose.

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Case

- 8 week old African American male admitted with a 9 day history of worsening facial and genitourinary rash. Followed by pediatrician and dermatology prior to admission and prescribed nystatin, cephalexin, and prednisone with no improvement.
- ED Course:
  - CBC: WBC – within normal limits, except for a platelet count of 649
  - CMP: within normal limits, except for an alkaline phosphatase level of 74
  - Blood culture

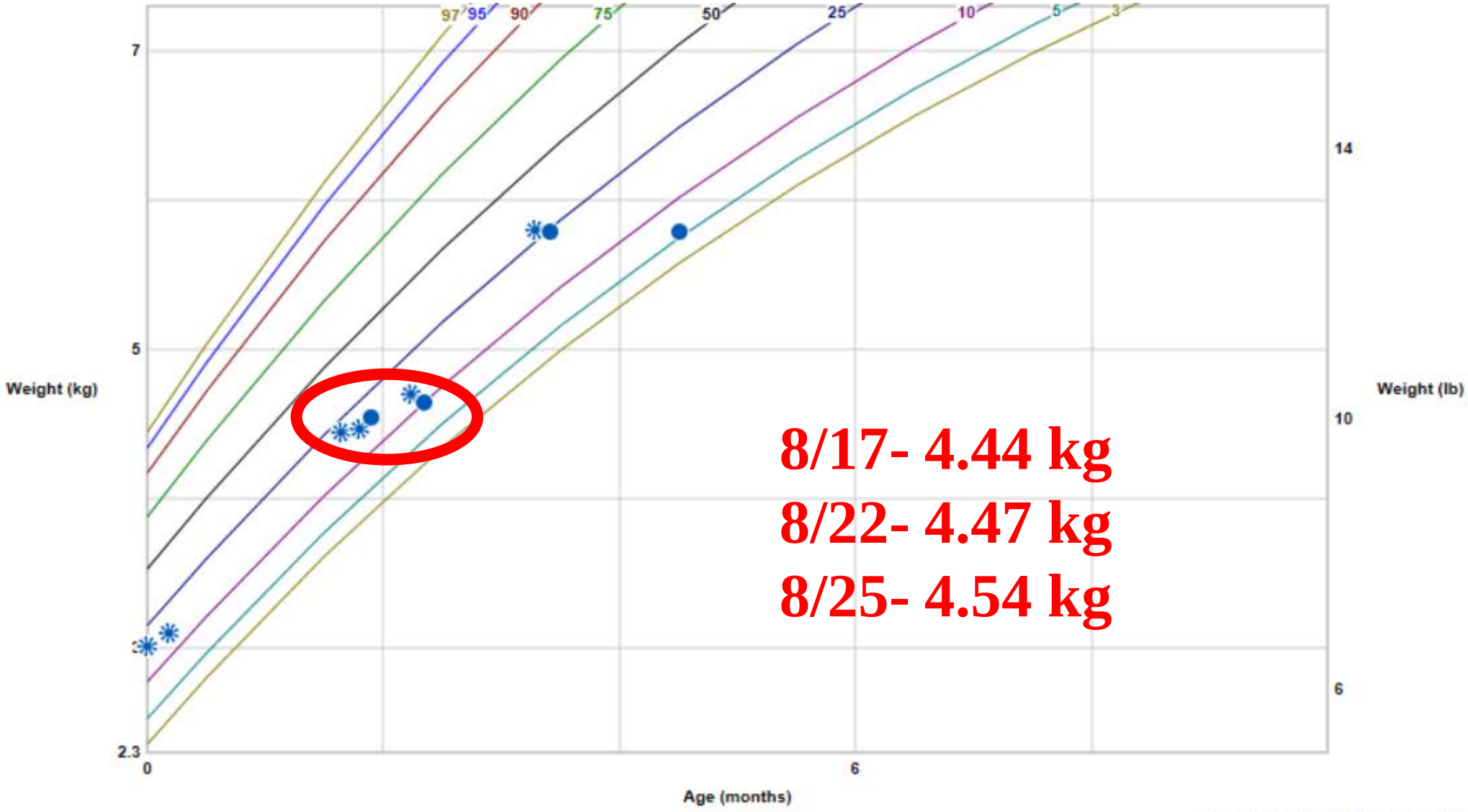
Additional History

- **Past medical history:** Full term at 39 weeks, no NICU stay, no previous hospitalizations
- **Medications:** none
- **Social:** Lives with mom, dad, brother and sister. Does not attend daycare. No known sick contacts. No new foods, no travel.
- **Family history:** Mom, sister, and brother with asthma. Sister with eczema.
- **Immunizations:** Hep B. 2 month vaccines NOT given yet.
- **Diet:** Exclusively breastfed with nursing and expressed breast milk. PO intake normal but taking longer to drink.
- **ROS:** Positive- decreased energy, alopecia, slightly fussy.
  Negative -fever, diarrhea.

Physical Exam

- Vital Signs:
  - **Temp** 97.5 °F (36.4 °C)
  - **BP** 98/64
  - **Pulse** 130
  - **RR** 34
  - **SpO2** Min: 97 %
  - **HT:** 55.9 cm (22")
  - **WT: 4.54 kg (10 lb 0.1 oz), 17th percentile**

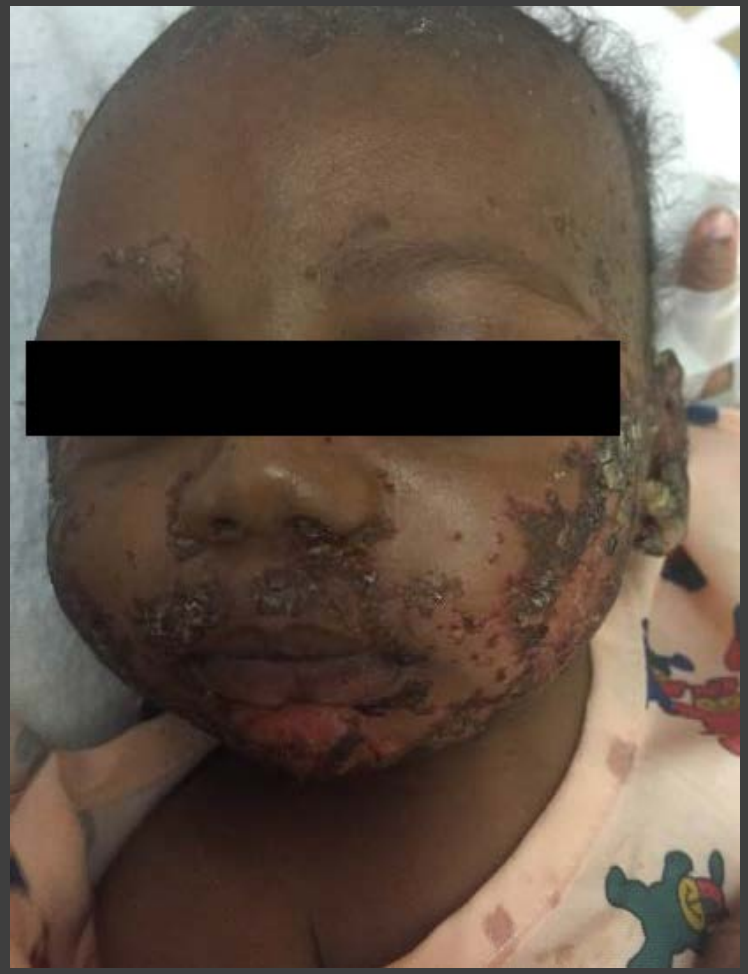
CDC Weight Chart

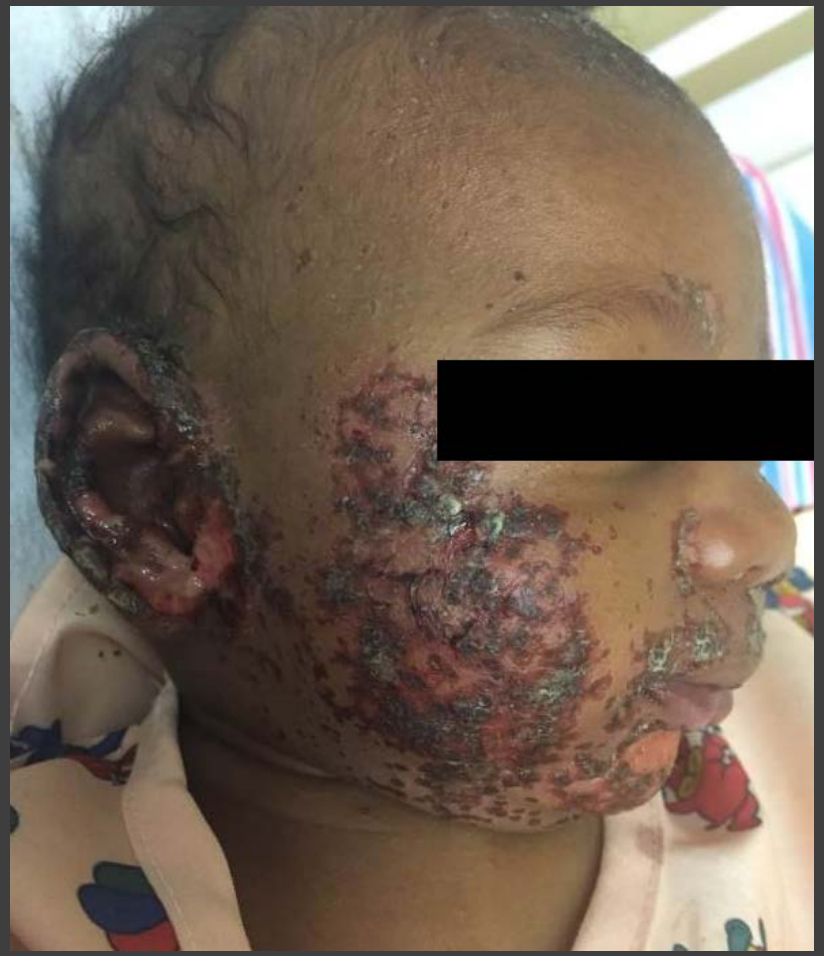


Source: Centers for Disease Control and Prevention (CDC), 2000

Physical Exam

- Constitutional: well-developed, no distress
- Skin: warm, moist, CR < 3 secs, no purpura, **large areas of crusting, denuded skin over bilateral cheeks, bilateral ears, within ear canals, bilateral nares, scalp, neck, and perianal region; spots of bleeding and ulcerations also noted**
- HEENT: no nasal discharge, dry mucous membranes, no pharyngeal erythema/lesions, **alopecia to scalp and eyebrows**
- Cardiovascular: Normal rate and regular rhythm, no murmurs
- Pulmonary/Chest: CTA b/l, no wheezes
- Abdominal: Soft, normal bowel sounds present, non-distended
- Genitourinary: Penis circumcised, **erythematous and denuded lesion on the underside of tip of penis**
- Neurological: normal muscle tone, no irritability, soft and flat fontanelles, photophobia





First Day of Admission

Differential

- Staphylococcal scalded skin syndrome
- Severe impetigo
- Seborrheic dermatitis
- Atopic dermatitis
- Underlying immunologic or nutritional deficiency

Severe Impetigo Vs Staphylococcal Scalded Skin Syndrome

Day 1

IV Clindamycin and Vancomycin

Zinc plasma levels

Bactroban ointment, Hydrocortisone cream, and Aquaphor for wound care

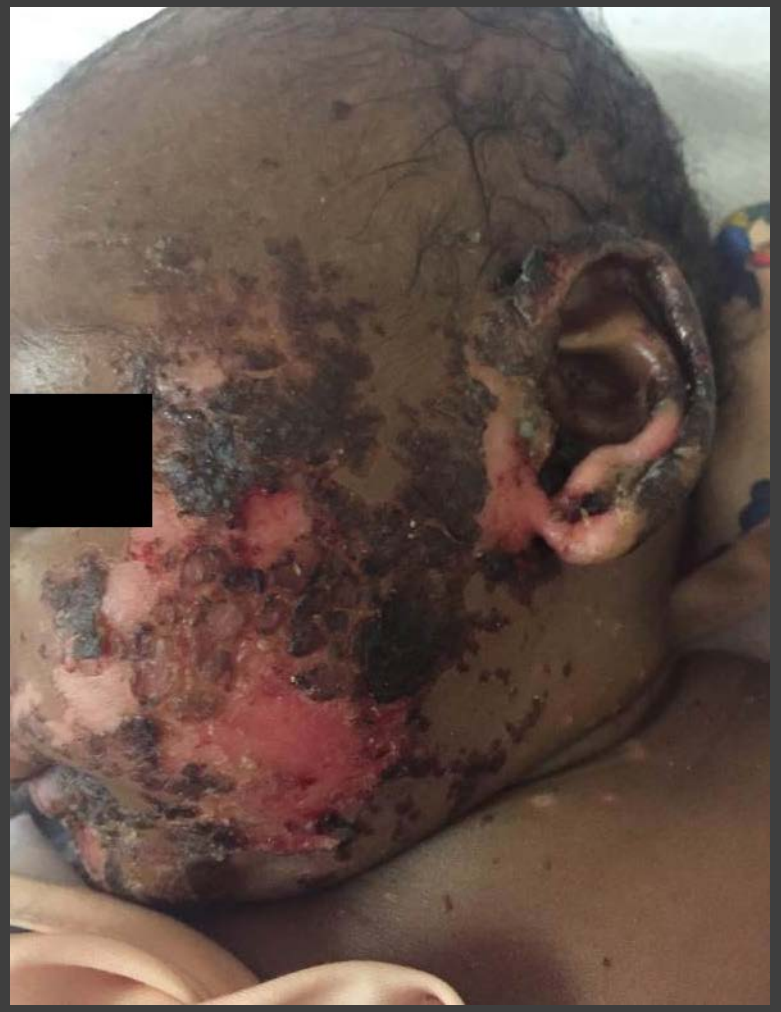
Day 2

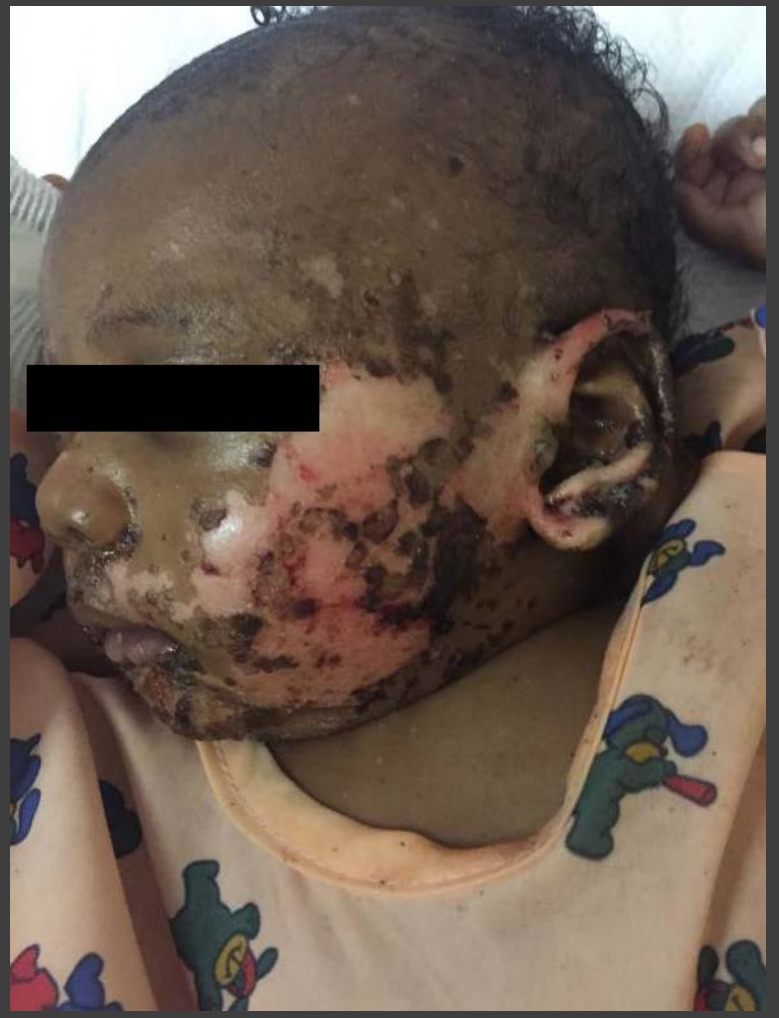
Skin Culture Positive
MSSA, Acinetobacter baumanni, Klebsiella pneumoniae

D/C Vancomycin
IV Clindamycin, Ketoconazole, and Ampicillin-Sulbactam

Day 3

T&B Cells, Quantitative Ig





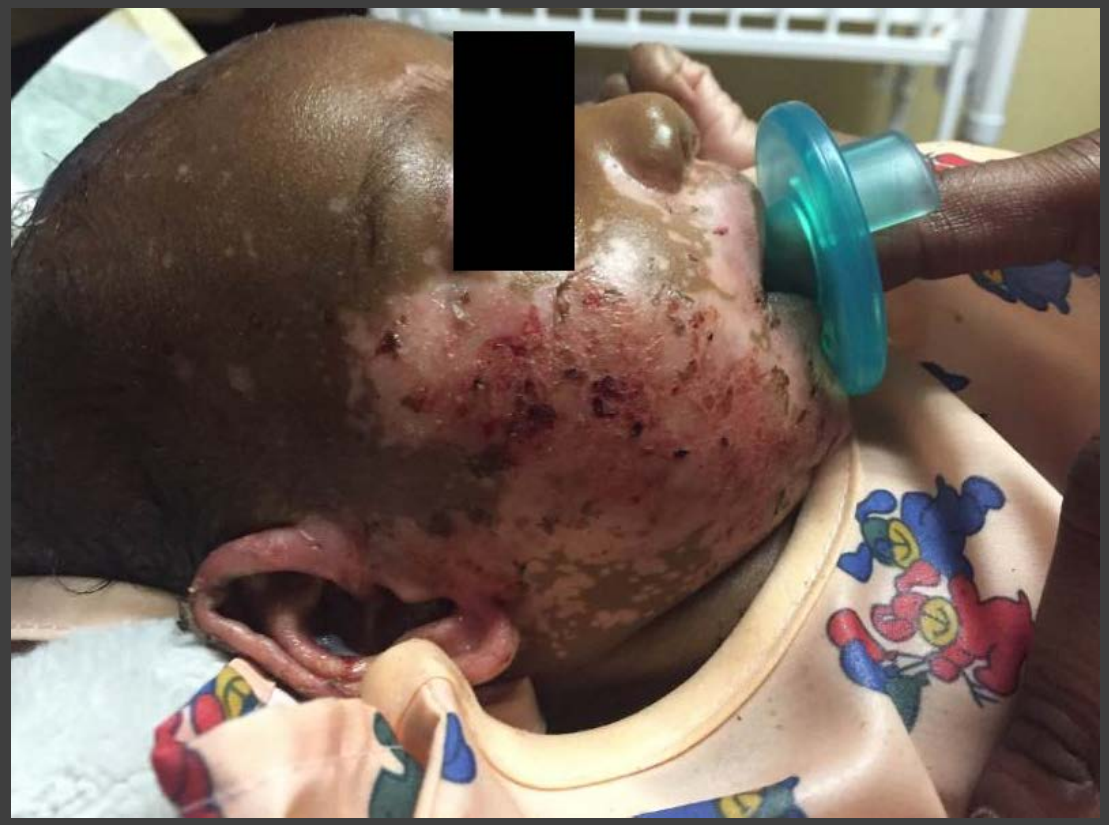
3 to 5 Days of Antibiotic Therapy

Staphylococcal Scalded Skin Syndrome with Suspected Underlying Nutritional Deficiency

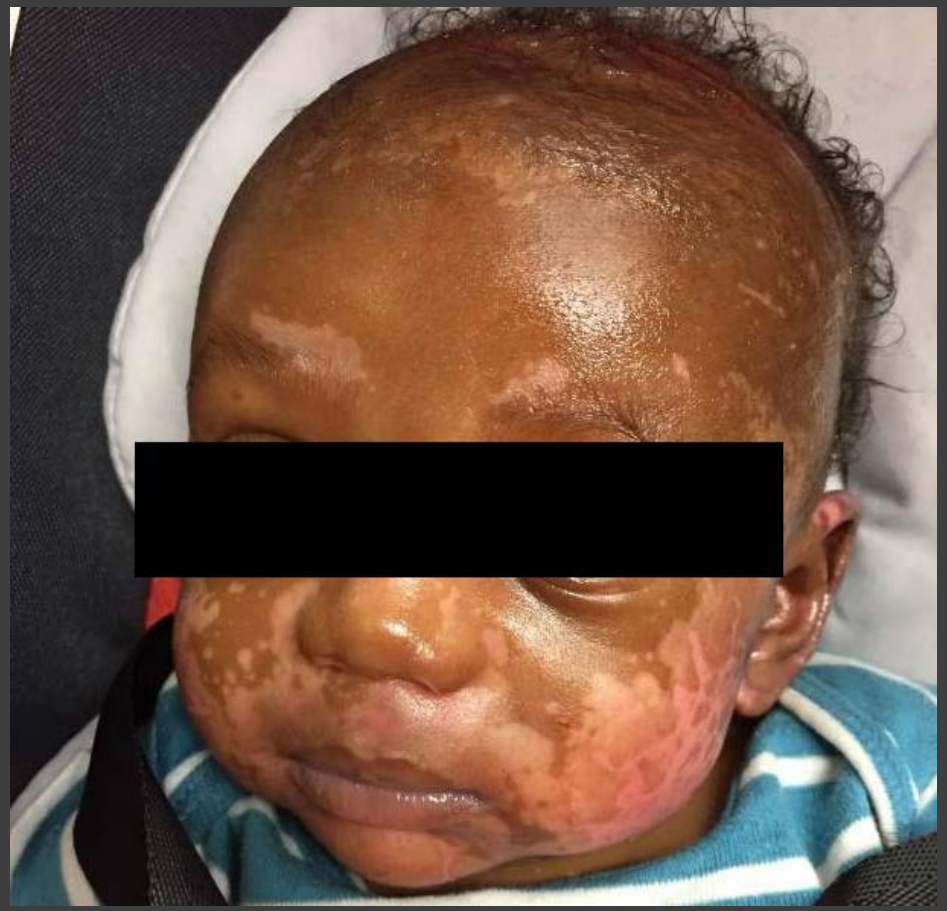
Day 4	Day 5	Day 6
IV Clindamycin, Ketoconazole, and Ampicillin-Sulbactam Blood culture negative T&B cells, Ig levels normal		**Plasma Zinc Level 0.10 mcg/mL** (0.6-1.20 mcg/mL) PO Zinc 3 mg/kg daily **Acrodermatitis Enteropathica**

Acrodermatitis Enteropathica with Secondary Bacterial Infection

Day 7	Day 8	Day 9
	Continued antibiotics for secondary infection Counseled family on zinc therapy	**Discharged** Clindamycin, Amoxicillin-Clavulanic acid, Fluconazole, and Zinc 3 mg/g daily



Day of Discharge



10 Days after Zinc Therapy Started

Acrodermatitis Enteropathica (AE)

- Estimated to affect **1 in 500,000**
- No predilection for gender or ethnicity
- **Classic Triad**

  Perioral and acral dermatitis, alopecia, diarrhea
- **Associated Symptoms**

  Secondary skin infections, failure to thrive, irritability, photophobia, poor wound healing, anemia, neuropsychiatric manifestations, delayed sexual maturation, hypogonadism

Acrodermatitis Enteropathica (AE)

- **Acquired zinc deficiency -** Those with an inadequate diet of zinc or have increased zinc requirements. (ex: Malnutrition, prematurity, total parenteral nutrition, burns, congenital malabsorption syndromes such as CF)

- **Genetic zinc deficiency**

1. Classical AE - Autosomal recessive mutation of SLC39A4 (Zip 4) gene on chromosome 8q24.3 causing decreased intestinal absorption of zinc in infant

2. Transient neonatal zinc deficiency (TNZD)– Autosomal dominant mutation of SLC30A2 (ZnT2) causing a zinc transporter defect in mother's breast milk

AE – Zinc

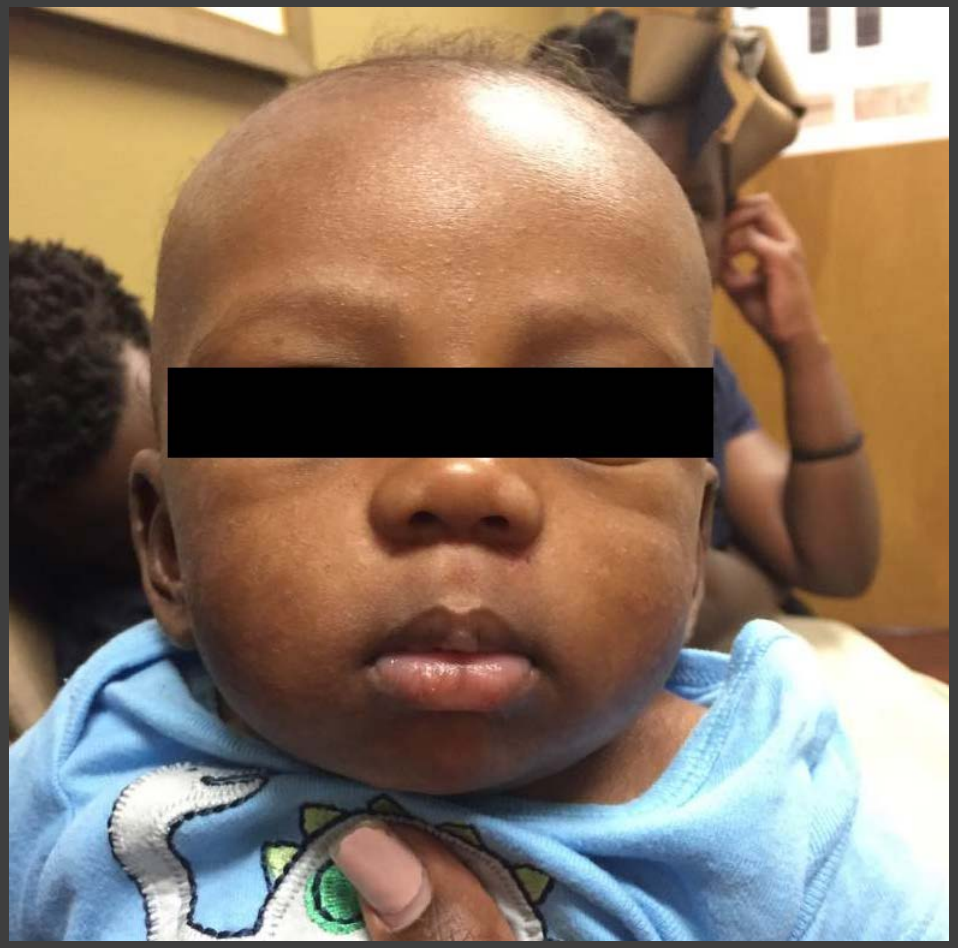
- Absorption
- Plasma circulation
- Storage
- Excretion

AE – Diagnosis

- Plasma zinc levels
- Rapid clinical response to zinc supplementation
- Alkaline phosphatase
- Skin biopsy
- Genetics

AE – Treatment

- 3 mg/kg/day of elemental zinc
- Lifelong supplement
- Measure levels every 3-6 months
- Toxicity



6 weeks later!

Discussion

- AE is a rare condition characterized by zinc deficiency which can lead to poor immune function, frequent infections, dermatitis, alopecia, diarrhea, and growth retardation.
- The incidence of the hereditary form of AE is about 1 in 500,000 births. Treatment involves lifelong oral replacement of zinc and yields a highly effective response.
- Pediatricians must have a high index of suspicion for zinc deficiency in infants presenting with severe skin infections. Measurement of the plasma zinc level is a simple and readily available test that can prevent delayed diagnosis of this easily treated condition.

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Questions and Comments