RICKETS: NOT A DISEASE OF THE PAST

LA-AAP 2017 Annual Meeting
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LSUHSC-NO Pediatrics
Disclosure Statement

- Speaker: Wilfreda Lindsey, MD
- Dr. Lindsey has documented that she has no actual or potential conflict of interest in relation to this program/presentation.
Chief Complaint

- 7 mo male presents with seizure-like activity
HPI / ROS

- 6 days ago brief seizure like activity
  - general tonic/clonic under 1 min

- EMS → OSH demonstrating low calcium

- Repeated outpt at PCP and still low

- Sent to chnola to be evaluated for hypocalcemia

- No fever
- No rash
- No URI symptoms
- No N/V/D/C
- PO intake wnl
- UOP wnl
- +Daycare
- No recent travel
Histories

- **PMHx**
  - Reflux

- **PSHx**
  - None

- **Birth Hx**
  - FT SVD
  - Pregnancy complicated by HTN, No NICU stay
  - 7 lbs 8 oz
  - Newborn screen wnl per mom

- **FHx**
  - No history of endocrine disorders

- Immunizations UTD
- NKDA

- **Diet**
  - Breast fed
  - 2 jars baby food / day

- Development wnl
Physical Exam

- Vitals: T-98.6 F  P-128  RR-44  BP-137/87 crying  O2-98% RA
  - Wt-8.7kg 50th%  Ht-66cm 10th%  HC-44cm 50th%
- Gen: awake & alert, crying but consolable by mom
- Heent: non dysmorphic facies, mmmp, no rhinorrhea, OP clear, no LAD
- CV: rrr, no mrg, 2+ fem & brachial pulses
- Resp: ctab no wr
- Abd: +BS NT/ND, no HSM
- Ext: no c/c/e
- Neuro: alert and crying, symmetric facies, strength wnl, able to sit on own, DTR 3+ patellar & tricep bl, no clonus, no focal deficits
Labs / Imaging

iCal 2.54
Vitamin D 25 Hydroxy 4 ng/ml

Urine:
  Calcium < 5
  Ca/Cr < 0.14

PTH 570 pg/ml

Xray R Hand: Changes consistent with partially compensated rickets of the distal radius and ulna w/ fraying.

CXR: No significant reticular notching within rib cage

5.1/ 5.4 / 6.9 / 3.5 / 0.4 / 90/ 978 / 33 / 1.6
Ca/Phos/TP/Alb / Tbili/Ast/AP / Alt / Mg

5.2  106  5  98
5  21  0.2
Vitamin D Deficiency → Rickets

- Rickets
  - Disease of growing bone failing to mineralize
  - Prior to bone epiphyses closure

- USA
  - 26% infants breast fed until 6 mo
  - CDC: 5 / 1,000,000 children age 6mo-5yr

- Increased world wide
Vitamin D Metabolism

1. Sunlight exposure leads to the conversion of 7-dehydrocholesterol in skin to D₃.
2. D₃ is converted to 25 hydroxy D₃ in the liver.
3. 25 hydroxy D₃ is further converted to 1,25 hydroxy D₃ in the kidneys, which increases calcium absorption in the intestines.
4. 1,25 hydroxy D₃ increases bone mineralization and maintains calcium balance in the body.

Dietary and supplement sources of vitamin D₃, D₂ include:
- Milk
- Eggs
- Fatty fish
- Fortified cereals
Role of Vitamin D
Risk Factors

- Exclusive Breast feeding
  - Without supplementation

- Dark skin pigmentation
  - Decrease in UV light absorption

- Decreased exposure to sunlight
  - Increased risk in Fall / Winter

- Malabsorption
  - Celiac Disease
  - Cystic Fibrosis
  - IBD
Clinical Manifestations of Hypocalcemia

- Hypocalcemia
  - $Ca^{2+} < 8.5 \text{ mg/dL}$
  - $iCa < 4.5 \text{ mg/dL}$

- Painful muscle spasms
- Generalized seizures
- Vomiting
- Prolonged QT EKG

- Chvostek sign
  - Tap anterior to ear lobe below cheek bone

- Trousseau sign
  - Carpal muscle spasm when inflating BP cuff above systolic pressure
Clinical Manifestation Rickets

- Hypocalcemia
- Hypophosphatemia
- Elevated PTH
  - *Always with nutritional def.*
- Elevated Alk Phos
  - *Every type of Rickets*
- Bone pain
- Decrease in GR / FTT
- Widened epiphysis
- Bowed legs, rachitic rosary, craniotabes
- Dental complications
Prevention of Rickets and Vitamin D Deficiency in Infants, Children, and Adolescents
Carol L. Wagner and Frank R. Greer
Pediatrics 2008;122:1142
DOI: 10.1542/peds.2008-1862

**TABLE 2** Oral Vitamin D Preparations Currently Available in the United States (in Alphabetical Order)

<table>
<thead>
<tr>
<th>Preparationa</th>
<th>Dosage</th>
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<tbody>
<tr>
<td>Bio-D-Mulsion (Biotics Research Laboratory, Rosenberg, TX; <a href="http://www.bioticsresearch.com">www.bioticsresearch.com</a>)</td>
<td>1 drop contains 400 IU; also comes in a preparation of 2000 IU per drop; corn oil preparation</td>
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<tr>
<td>Carlson Laboratories (Arlington Heights, IL; <a href="http://www.carlsonlabs.com">www.carlsonlabs.com</a>)</td>
<td>1 gel cap contains 400 IU; also comes in 2000 IU and 4000 IU gel caps and in single-drop preparations of 400 IU, 1000 IU, and 2000 IU; safflower oil preparation</td>
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<tr>
<td>Just D (Sunlight Vitamins Inc [Distributed by UnitDrugCo, Centennial, CO]; <a href="http://www.sunlightvitamins.com">www.sunlightvitamins.com</a>)</td>
<td>1 mL contains 400 IU; corn oil preparation</td>
</tr>
<tr>
<td>Multivitamin preparations: polyvitamins (A, D, and C vitamin preparations)c</td>
<td>1 mL contains 400 IU; variable preparations that include glycine and water; may also contain propylene glycol and/or polysorbate 80</td>
</tr>
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Note that higher-dose oral preparations may be necessary for the treatment of those with rickets in the first few months of therapy or for patients with chronic diseases such as fat malabsorption (cystic fibrosis) or patients chronically taking medications that interfere with vitamin D metabolism (such as antiseizure medications).

a A study by Martinez et al. showed that newborn and older infants preferred oil-based liquid preparations to alcohol-based preparations.

b Single-drop preparation may be better tolerated in patients with oral aversion issues, but proper instruction regarding administration of these drops must be given to the parents or care provider, given the increased risk of toxicity, incorrect dosing, or accidental ingestion.

c The cost of vitamin D-only preparations may be more than multivitamin preparations and could be an issue for health clinics that dispense vitamins to infants and children. The multivitamin preparation was the only preparation available until recently; therefore, there is a comfort among practitioners in dispensing multivitamins to all age groups.
<table>
<thead>
<tr>
<th>Underlying Cause</th>
<th>Ca</th>
<th>Phos</th>
<th>Alk Phos</th>
<th>PTH</th>
<th>1,25 Vit D</th>
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<tr>
<td>Vit D def</td>
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<td>Lack of Nutrition / Sun</td>
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<td>Vit D dependent</td>
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<td>1- α hydroxylase deficiency</td>
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<td>Vit D resistant</td>
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<td>End Organ resistance</td>
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<td>X-Linked Familial hypophosphatemic</td>
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<td>Defect tubule resorption of phos</td>
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<td>Renal Disease</td>
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<td>Defect excretion phos</td>
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Back to Our Patient

- Given IV Calcium Gluconate
- Started on Vitamin D and Calcium Supplementation
  - CaCO₃ 750mg PO QID
  - Ergocalciferol 3200 units PO Qday (Vitamin D₂)
  - Calcitrol 0.25 mcg PO TID (1,25 dihydroxy Vit D₃)
- Stabilized and Improved
  - Ca²⁺ 7.2
  - iCal 4.17
Questions?