Different Renal Diagnosis, Different Needs: Nephrogenic Diabetes Insipidus and Polycystic Kidney Disease
Disclosures

- Yes, consultant for Genentech
Outline

- Definition of disease
- Statistics
- Characteristics and Complications of disease
- Nutrition Interventions
- Differences among NDI and PKD
Trivia Question

How many liters of blood do our kidneys filter per day?
180–200 Liters per day
*equal to 563 cans of Coca-Cola*
Nephrogenic Diabetes Insipidus (NDI)
Defining NDI

- Inability to concentrate urine leading to polyuria and polydipsia
  - More specifically, kidney does not respond to antidiuretic hormone (ADH) also known as vasopressin
Statistics

**NDI**
Nephrogenic Diabetes Insipidus

A rare genetic kidney disorder in which a defect in the small tubules in the kidneys causes a person to pass a large amount of urine. The tubules allow too much water to be removed from the body.

300,000,000 Americans

600 have NDI

Eminence, Missouri
Population 600

United States of America
Population 300 Million

**Odds of having NDI:** 1 in 500,000

- 1/11,500 bowling 300
- 1/19,600 pitching perfect game
- 1/77,000 NBA player killed by asteroid
- 1/200,000 struck by lightning
- 1/200,000 die by shark attack
- 1/300,000 tsunami death
- 1/500,000 have NDI

**How does NDI work?**

Pituitary gland: Hey kidneys, concentrate that urine

ADH hormone: (keep some water, lose some sodium)

Kidneys say: Whaaaaaat?

Tubules malfunction

H2O not returned to body

Urine not concentrated

Excessive urination

Dehydration + sodium build-up

Let's compare those odds.
Characteristics of NDI

- **Most commonly**
  - Polyuria
  - Polydipsia
  - Dehydration
  - Failure to Thrive

- **Occasionally**
  - Vomiting
  - Gagging or retching
  - Poor feeding
  - Constipation or diarrhea
  - Lethargy or irritability
  - Fever without explanation
Ins and Outs

- Metabolism 10%
- Foods 30%
- Beverages 60%

Average intake per day:
- 250 ml
- 750 ml
- 1500 ml

Total average intake: 2500 ml

Average output per day:
- 100 ml
- 200 ml
- 700 ml
- 1500 ml

Total average output: 2500 ml

- Feces 4%
- Sweat 8%
- Insensible loss via skin and lungs 28%
- Urine 60%
Complications of NDI

- Dehydration
- Hypernatremia

- Adequate nutrition
  - Competing with water (empty calories)
    - Most infants prefer water over formula
  - Frequent feedings
    - Prompted by parents (waking at night)
<table>
<thead>
<tr>
<th>Challenges</th>
<th>Goals</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnosis is difficult; symptoms are similar to those at age</td>
<td>Decrease UOP</td>
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<tr>
<td>Fluid balance</td>
<td>Optimize Intake</td>
</tr>
<tr>
<td></td>
<td>◦ Based on function</td>
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<tr>
<td></td>
<td>Promote Growth</td>
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<tr>
<td></td>
<td>Control fluid status</td>
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<td></td>
<td>Normalize feeding pattern</td>
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</table>
Reduce Sodium in Diet/Decrease Renal Solute Load
- Sodium: 1 meq/kg/d
- Protein: 2 g/kg/d
  - Sodium restriction most important since children need protein for growth

How so?
- Dilute formula by concentration but add calories via oil or Duocal
Defining Renal Solute Load

- **Renal Solute Load (RSL)**
  - solutes of endogenous or dietary origin that require excretion by the kidneys
  - Actual RSL is the PRSL – solute excretion by non-renal routes like feces and skin

- **Potential Renal Solute Load (PRSL)**
  - solutes of dietary origin that would need to be excreted in the urine if none were diverted into synthesis of new tissue and none were lost through non-renal routes
  - \( PRSL = \frac{N}{28} + Na + Cl + K + Pa \)
  - \( N = \text{dietary nitrogen in mg/28 mmol of urea} \)
Nutrition Interventions for NDI continued...

- Encourage intake of large volume of water
  - Some children may need to be woken up in the middle of the night to provide water
  - Water in between feeds (every 2 hours)

- Normalize but control feeding schedule
  - Some patients come to you feeding hourly because of thirst.
  - Indicator that they need more medications to better manage output.

- “Higher calorie intake”
  - ~150–200% of estimated needs
    - Based on experience and discussion with other renal RDs

- Placement of GT for those who cannot keep up with intake.
What to Feed?

- Breastmilk
  - Lowest renal solute load
- Formula
  - Higher than BM
- Cow’s milk
  - Higher than formula but lower than skim
- Skim milk
  - Highest vs all (326 mosm/L and 93 mosm/100 kcal)

<table>
<thead>
<tr>
<th>Table 1.</th>
<th>Potential Renal Solute Load of Infant Foods</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>PRSL*</td>
</tr>
<tr>
<td></td>
<td>mosm/L</td>
</tr>
<tr>
<td>Human milk</td>
<td>93</td>
</tr>
<tr>
<td>Milk-based formula</td>
<td>135–260</td>
</tr>
<tr>
<td>Cow milk</td>
<td>308</td>
</tr>
</tbody>
</table>
Low renal solute load formulas

- **Standard**
  - 126–136 mosm/L
    - Gerber Good Start may be best option

- **Soy Protein Based**
  - 154–164 mosm/L
    - Prosobee

- **Semi–elemental**
  - Alimentum and Pregestimil both ~165 mosm/L

- **Elemental**

- **Specialized**
  - Similac PM 60/40 122 mosm/L
“Low renal solute load” solids

- Low sodium crackers, fruit, vegetables, and biscuits
- Make food at home
- Watch choices when out to eat
Summary

- NDI is rare
- Defined by the inability to concentrate urine leading to polyuria and polydipsia
- Complications include dehydration and poor growth
- RSL and PRSL play an important role in figuring out nutrition intervention
When was the first formula made?
Answer

1865 by Justus von Liebig
Polycystic Kidney Disease (PKD)
Defining PKD

- Genetic disorder that causes numerous cysts to grow on kidneys
- Two main types of PKD
  - Autosomal dominant
  - Autosomal recessive
    - Often called “infantile PKD”
~1 per 20,000 – 40,000 people suffer from the ARPKD

ARPKD more rare than ADPKD

More than 50% of patients with ARPKD progress to renal failure in the first decade of life

Mortality in the neonatal period can be as high as 30–50%.
Characteristics of PKD

- Oligohydramnios
- Enlarged Kidneys (with cysts)
- Hypertension
- Growth problems
Complications of PKD

- Bone disease as well as growth concerns
- Dehydration (related to polyuria)
- Feeding Problems
  - About 25% of children with ARPKD also are Failure to Thrive
- GERD
Challenges and Goals of Managing PKD

Challenges

- Anatomy (enlarged kidney)
  - Causes intra-abdominal pressure
    - Breathing concerns
    - “Feeling of fullness”
    - GI motility is slowed
      - Leading to reduced absorption

Nutrition Goals

- Optimize Intake
  - Based on function
- Promote Growth
- Control fluid status
Nutrition Interventions for PKD

- Poor growth
  - Growth hormone
- Elevated blood pressure
  - Medications
- Declining kidney function
  - Low sodium, low k, low phosphorus
- Limited ability to absorb nutrients properly
  - ? Elemental feeds
  - GJT feeds
  - Anti-reflux medications
Thoughts on Soy Protein Isolate and Omega-3 Polyunsaturated fatty acids

- **Pre-clinical studies**
  - Show that SPI and Omega 3 ameliorate PKD progression and severity as well as improve bone status

- **Soy Protein Isolate**
  - Enhanced bone mineral content and bone mineral density
    - Estrogenic isoflavones
    - Reduced acid load

- **Omega-3 Polyunsaturated fatty acids**
  - Enhanced bone mineral content and bone mineral density
  - ALA
    - Renal protective
ARPKD is rare

Defined by disorder where kidney is enlarged due to large cysts

Complications include dehydration, breathing difficulties, elevated blood pressure, feeding problems and poor growth

Optimizing nutrition for growth is very important
## Similarities and Differences among NDI and PKD

<table>
<thead>
<tr>
<th>Similarities</th>
<th>Differences</th>
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<tr>
<td>- Both diseases may present with the following:</td>
<td>- Etiology of failure to thrive:</td>
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<tr>
<td>◦ Polyuria</td>
<td>◦ NDI</td>
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<tr>
<td>◦ Polydipsia</td>
<td>• r/t fluid balance</td>
</tr>
<tr>
<td>◦ dehydration</td>
<td>◦ ARPKD</td>
</tr>
<tr>
<td>- Patients also suffer from Failure to Thrive</td>
<td>• r/t anatomy</td>
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- **NDI**
- **PKD**
Questions and Discussion