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

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Until every child is well™

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?

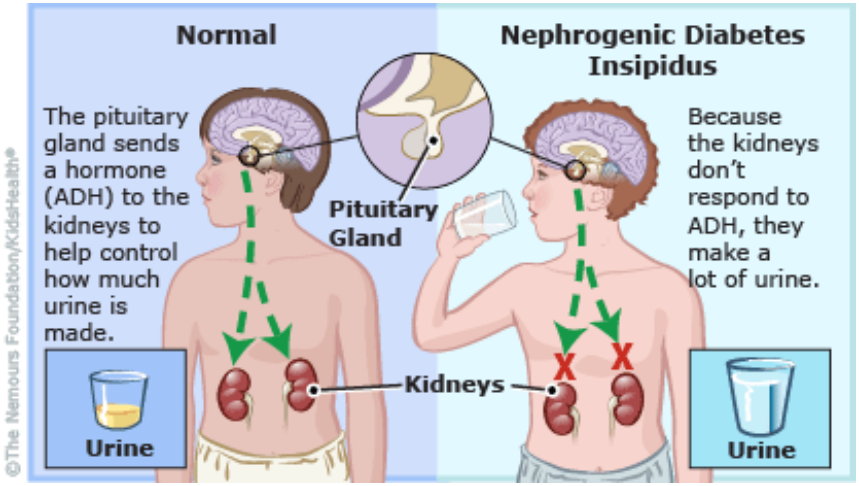
Answer

- 180-200 Liters per day
  equal to 563 cans of Coca-Cola

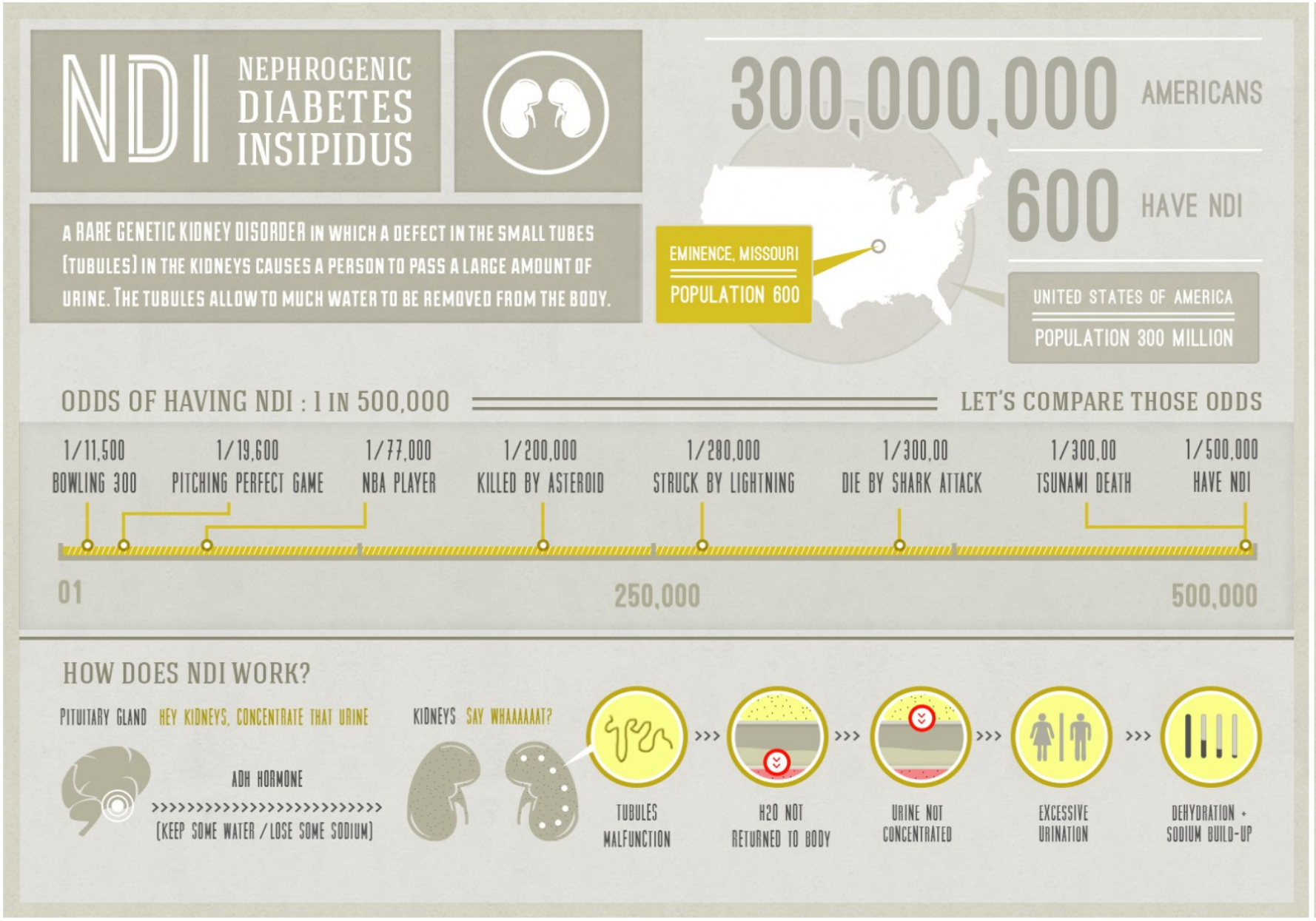
Nephrogenic Diabetes Insipidus (NDI)

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



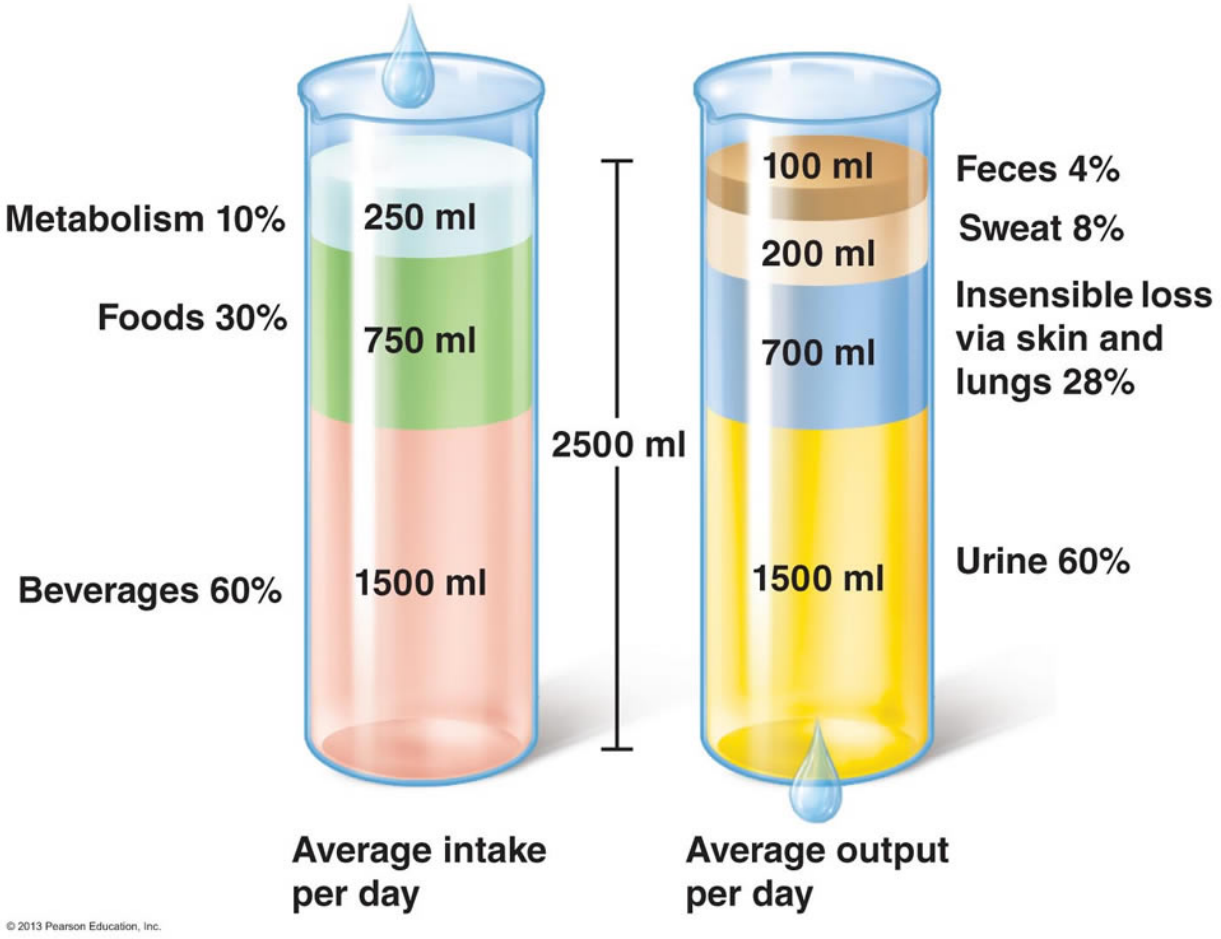
Statistics


NDI
NEPHROGENIC DIABETES INSIPIDUS
A RARE GENETIC KIDNEY DISORDER IN WHICH A DEFECT IN THE SMALL TUBES (TUBULES) IN THE KIDNEYS CAUSES A PERSON TO PASS A LARGE AMOUNT OF URINE. THE TUBULES ALLOW TO 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
LET'S COMPARE THOSE ODDS
1/11,500 BOWLING 300
1/19,600 PITCHING PERFECT GAME
1/77,000 NBA PLAYER
1/200,000 KILLED BY ASTEROID
1/280,000 STRUCK BY LIGHTNING
1/300,00 DIE BY SHARK ATTACK
1/300,00 TSUNAMI DEATH
1/500,000 HAVE NDI
01
250,000
500,000
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

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



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)

Challenges and Goals of Managing NDI

Challenges

- Diagnosis is difficult; symptoms are similar to those at age
- Fluid balance

Goals

- Decrease UOP
- Optimize Intake
  - Based on function
- Promote Growth
- Control fluid status
- Normalize feeding pattern

Nutrition Interventions for NDI

- 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= N/28 + Na + Cl + K+ Pa*
  - *N = 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 1.

Potential Renal Solute Load of Infant Foods

	PRSL*	
	mosm/L	mosm/100 kcal
Human milk	93	14
Milk-based formula	135–260	20–39
Cow milk	308	46

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

Trivia Question

- 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”

Statistics

- ~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

Summary

- 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

Similarities

- Both diseases may present with the following:
  - Polyuria
  - Polydipsia
  - dehydration
- Patients also suffer from Failure to Thrive

Differences

- Etiology of failure to thrive:
  - NDI
    - r/t fluid balance
  - ARPKD
    - r/t anatomy

Questions and Discussion