Passing the Baton of Medical and Nutritional Management of Inborn Errors of Metabolism

Matthew Rasberry, RD, LD, CNSC
Outpatient Metabolic Dietitian
Division of Genetics and Metabolism
Children’s National Medical Center
What’s an Inborn Error of Metabolism (IEM)?

[Diagram showing the process of metabolism with genes, enzymes, cofactors, and energy conversion.]
“Passing the Baton”

Clinic → Parent → Child
Talk Outline

1) Clinic to Parent/Child – “Getting the most out of your clinic”

2) Parent to Child – “Preparing your child for world – from infant to young adult”
Part 1:
Clinic to Parent/Child – “Getting the most out of your clinic”
Compliance vs. Adherence

**Compliance** ("to conform") suggests patient obeys the physicians’ instructions to conform to the medical goals

- Non-complaint = not following “doctors orders”

**Adherence** ("to hold closely") implies the patient is independent, intelligent and autonomous people who take more active and voluntary roles in defining and pursuing goals for their medical treatment

- Non-adherent = there’s a reason why the treatment goals are not held closely

Diabetes Care 1999; 22(4): 635-639
The Paradox of Dietary Management

• With better outcomes and treatments; treatment goals are advancing to achieve the “best-possible” neuropsychological outcomes with a higher quality of life
  • Complex, difficult diets with high expectations ...
    • Can negatively influence quality of life?
    • May not be as executable?
    • Perfect with the diet = Best outcome?

• Have an open discussion
  • Transparency on both sides
  • Discuss what's working and not working
  • Set goals as a team

Adherence Issues with IEMs

• Adherence is LIFELONG
  • It’s RELENTLESS

• Food is social, food is life
  • Dietary management is very invasive

• Expectations are high, management is complex
  • Stress, anxiety, fear

• What are your experiences?
General Strategies to Improve Adherence

1. Education – Practical and Theoretical
2. Improved communication between patient and clinician
3. Simplification of the regimen when possible
4. Everyone involved (Family, Friends, Teachers, etc.)
5. Positive attitudes
6. Self-reliance and self-efficacy
7. Acknowledgement of success
8. Increased access to health care professionals

Understanding the Science

- Carbohydrates
- Protein
- Fats

4 hr 12 hr 16 hr 24 hr
Perspectives on Metabolic Nutrition Education

• Metabolic clinician reported nutrition education usage

Perspectives on Metabolic Nutrition Education

- Metabolic clinician perceived effectiveness of education.
Perspectives on Metabolic Nutrition Education

• Parents and clinician both view one-on-one counseling as invaluable
  • Patients (kids) viewed parents as the most effective education tool
  • The clinicians perceived a decline in the parents used as education tool with increasing age

• The least perceived effective education (handouts) is used very often and the more effective educations are not used often (group clinics/classes)
  • 91% of families reported they would be interested group clinic/classes

Practical Recommendations

• Ask questions (Keep a diary/notebook). Come prepared.
• Ask why? Understand it.
• Read the clinic notes
  • Understand what is going on (ask for clarifications), correct mistakes/fill in the missing gaps, know the labs
• Let us know what is working and not working for you
• Keep a calendar/date book
• Connect with another family (ask your clinic)
Practical Recommendations

• Request group clinic/cooking classes (start your own if you have to)
• Advocate for yourself. Ask for help. Ask for “refresher”.
• Bring all carers to appointments at least sometimes
• Set ACHEIVABLE and MEASURABLE goals with the clinic

• What’s been helpful for you when interacting with your clinic?
Part 2:
Parent to Child – “Preparing your child for world – from infant to young adult”
Increasing Fruit and Vegetable Intake

• Why do kids not like fruits and vegetables?

• Why does salt, sugar and fat taste so good?

• Why are Fruits and Vegetables so important for IEMs?
The Benefits of Fruits and Vegetables

• Why should we eat fruits and vegetables?
  • The diet already limits the variety, we don’t want to fruits and vegetables to not be an option
    • Fruits and vegetables have great color, texture, flavor, etc.
  • Vitamin supplements are great for correcting deficiencies and being therapeutic in some cases, but they do not replicate whole foods
    • i.e. Vitamin E and Cancer compared to whole fruits/vegetables
  • Different food groups provide different nutrients – variety ensure you get everything
  • Fruits and vegetables have fiber (soluble and insoluble) and fluid
  • Naturally low protein, low fat and low calorie (healthy weight)
Low Fat "My Plate" (for fat restricted diets)
Increasing Fruits and Vegetables

• Cochrane Review – of all the studied interventions to increase fruits and vegetables over the long term – nothing really works that well...
  • Most successful intervention = Repeated exposure with non-food reward (tangible or social) administered by the parent
  • Sticker Charts
    – 50 stickers = $5.00 for the toy store
    – 25 stickers = pick the movie for movie night

Cochrane Database of Systematic Review 2012; 11: CD008552
Fruits and Vegetable Tricks

• Role model the behavior – do you eat fruits and veggies?
• Conquer your fruits and sweet, mild tasting vegetables first
• Don’t intimidate
  • Starting out? – 2 carrot coins, 1 green bean
  • Don’t stack the plate with broccoli
• Get the texture right
  • Crock pot, stews, canned fruit/veggies
• Don’t be afraid to hide it
• Be creative, make it pretty

• What else?
Formula Intake

• **Challenges:** Taste, large volumes, poor appetite, frequency

• **Solutions to improve intake?**
  - High concentration, low volume vs. Diluted, high volume
  - Additives (SunnyD, Hershey Syrup, Extracts, etc.)
  - Trialing new formulas
  - Reward, positive reinforcement
  - Understanding the importance of the formula
  - G-tube

• What works for everyone else?
Tracking the diet

• Yes and No Foods
• Red Light, Yellow Light, Green Light
• Games
  • i.e. Treasure Hunt at the grocery store - find the granola bar that is 1 gram of protein, find a dessert that is 0 grams protein, etc.
• Journals, food logs, white boards (start early)
• Lesson Plans: Eat Bright; Stay Bright; Know to Grow

• How do you teach your child to track the diet?
Tracking Resources (Comprehensive)

http://ndb.nal.usda.gov/
Or Google “USDA Database”
http://www.fda.gov/food/ingredientspackaginglabeling/labelingnutrition/ucm063367.htm

Or Google “FDA Fruits and Vegetables”
Tracking Resources (Internet/Apps)

- [http://www.choosemyplate.gov/supertracker-tools/supertracker.html](http://www.choosemyplate.gov/supertracker-tools/supertracker.html)
- [https://nutritiondata.self.com](https://nutritiondata.self.com)
10 Things to Make Home Cooking Easier

1. Have a plan
   - Menu Cycle and Grocery List
   - Meals Ready to Go (freezer, simple recipes)

<table>
<thead>
<tr>
<th></th>
<th>Sun</th>
<th>Mon</th>
<th>Tues</th>
<th>Wed</th>
<th>Thurs</th>
<th>Fri</th>
<th>Sat</th>
</tr>
</thead>
<tbody>
<tr>
<td>Week 1</td>
<td>Chili</td>
<td>Pizza</td>
<td>Spaghetti</td>
<td>Tacos</td>
<td>Rice Dish</td>
<td>Sloppy Joes</td>
<td>Soup</td>
</tr>
<tr>
<td>Week 2</td>
<td>Loaded baked potato</td>
<td>Stew</td>
<td>Breakfast for Dinner</td>
<td>Salad</td>
<td>Cold Pasta Salad</td>
<td>Stir fry</td>
<td>Sandwiches</td>
</tr>
</tbody>
</table>

2. Keep it simple and kid friendly
   - Simple recipe (few ingredients)
   - Tacos, Pizza, Pasta, Rice, etc.
10 Things to Make Home Cooking Easier

3. Season the food... (special diets don’t have to be bland...)
   • Minced ginger, lemon zest, diced onions, minced garlic, fresh herbs (cilantro, parsley, basil, mint, etc.)
   • Salt/Pepper, Mrs. Dash/Herb Blends, Old Bay, Adobo, Hot Sauce, etc.

4. Go to cooking classes or start your own
   • Regular cooking class – any recipe can be modified
   • Low Protein/Fat Cooking Classes – meet and talk with other families

5. Keep it simple and kid friendly
   • Simple recipe (few ingredients)
   • Tacos, Pizza, Pasta, Pasta Salad, Rice, etc.
10 Things to Make Home Cooking Easier

6. Be efficient
   - Do you prep work on the day-off/Sunday (Tupperware)
   - Make big batches (meals for the week; freeze for later)

7. Garnish the food/make it look pretty
   - Make it look like fine dining – sprigs of parsley, stacked food
   - Make shapes
   - Colorful plates and spoons
10 Things to Make Home Cooking Easier

8. “Live to eat, not eat to live”
   • Be adventurous
   • Cooking and baking = quality family time (plan it)

9. Practice makes perfect

10. What Else?
Growing up and taking over.

- PKU Toolkit (has some tips that can apply for OA/FODs)
  - Questionnaire:
    1) I know the name of my condition?
    2) I can explain my condition and my special health needs?
    3) I know what medical insurance I have?
    4) I know what foods are appropriate and inappropriate?
    5) I know the name of my formula and how to make it?
    6) I know how to order my formula?
    7) I know the name of all the medications I take and how often I take them?
    8) I have discussed the use of drugs, tobacco and alcohol with my doctor?
    10) I know how to reach my clinic and schedule an appointment?
    11) I know who to call in an emergency?
    12) I have a support system (family, friends, teacher, coworker)?
What needs to be done to take over

- Start early, educate often (kids learn from parents more than the clinician?)
- Start involvement whenever possible
  - Making formula, tracking the diet, etc.
- Develop the support system and maintain positive attitudes
  - Nobody can do this alone
- Make sure they have a manageable plan in place
  - Theoretical and practical knowledge
  - Attention, organization skills, planning, decision making
Questions?