Cystic Fibrosis: Nutrition Guidelines and Quality Improvement

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Outline

- Introduction to Quality Improvement in CF
- Factors in Improving Nutrition Outcomes
- Cystic Fibrosis: Symptoms and Conditions, Signs of Malabsorption, Use of Pancreatic Enzymes
- Nutritional Guidelines
- Current Quality Improvement Project
Take Home Points

Nutrition

• CF patients have many obstacles to overcome in order to meet their energy needs

• CF patients have tremendous caloric requirements

• When evaluating the BMI for a CF patient, providers must use CF guidelines not those guidelines based on a healthy population

• It takes a CF TEAM to work successfully with the patient and family
Take Home Points

Quality Improvement

• Every clinician and practitioner should engage in QI work

• Identifying ways we can improve our systems, processes, and the care we provide should not be thought of as a punitive process

• Instead we should seek these opportunities to make the changes needed to provide the best care that we can provide to each patient
Origins of CF Quality Improvement

- Institute of Medicine’s “Crossing the Quality Chasm: A New Health Care System for the 21st Century” published in 2001

- Six aims for improvement in healthcare
  - Safe
  - Effective
  - Patient-centered
  - Timely
  - Efficient
  - Equitable
Basis of CF Quality Improvement

The CF Foundation has developed “Seven Worthy Goals” to improve the quality of care for CF patients and their families.
CFF Seven Worthy Goals:

#1 – People with CF, their families and CF health care professionals make up the CF care team.

The care experience of people with CF will be informed (to the extent that they wish), transparent, individualized and respectful — allowing for choice in all matters related to their person, circumstances and relationships in CF care.
Interdisciplinary Family-Centered Care

- MD
- RN
- RD
- MSW
- RRT
- Other
- Family
CFF Seven Worthy Goals:

- **#2** – Children, adolescents, and adults will have normal growth and nutrition.

- **#3** – All patients will receive appropriate therapies for maintaining lung function and decrease infection.

- **#4** – People with CF, their families and CF health care professionals will be well informed and active partners in reducing acquisition of respiratory pathogens, particularly *P. aeruginosa* and *B. cepacia* complex.
CFF Seven Worthy Goals:

- **#5** – People with CF will be screened and managed aggressively for complications of the disease, particularly CF-related diabetes.

- **#6** – Severely affected people with CF and their families will be well supported by their CF health care professionals when facing decisions about transplantation and end-of-life care.

- **#7** – People with CF and their families will have access to appropriate therapies, treatments and supports regardless of race, age, education or ability to pay.
Goals of Quality Improvement

• Create a **standardization** of care by identifying “best practices” and strategies to implement these practices.
• Develop **tools** through our QI initiatives to support these “best practices.”
• **Implement** these tools to make decisions on each individual patient’s needs.
• Build stronger **partnerships** between people with CF, families and their care center.
Nutrition Algorithm

Based on goals #2 and #3 of the CFF Worthy Goals

- #2 – Children, adolescents, and adults will have normal growth and nutrition and

- #3 – All patients will receive appropriate therapies for maintaining lung function and decrease infection.

We developed a **Nutritional Algorithm** as one of our QI initiatives.
Our Nutritional Algorithm allows us to systematically assess the nutritional care and status of each patient and develop a care plan that incorporates best practices.

The whole team uses the Nutritional Algorithm. It is especially helpful to the physicians, since three different physicians see our CF patients in our clinic. We want to make sure we are all on the same page to improving outcomes and the quality of our care.
### Nutrition Algorithm

<table>
<thead>
<tr>
<th>Question</th>
<th>Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>Should O2 be considered?</td>
<td>Yes: Finger stick, Noninvasive referral</td>
</tr>
<tr>
<td></td>
<td>No: Finger stick, Noninvasive referral, Invasive referral</td>
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<tr>
<td></td>
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<tr>
<td>Is there evidence of GI reflux?</td>
<td>Yes: PPI, Upper GI, pH probe, GI referral</td>
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<tr>
<td></td>
<td>No</td>
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<td></td>
<td></td>
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<tr>
<td>Is there evidence of other GI disease?</td>
<td>Yes: Abdominal CT, Abdominal US, Liver function tests</td>
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<tr>
<td></td>
<td>No</td>
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<td></td>
<td></td>
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<tr>
<td>Are there psychosocial or psychosomatic issues related to poor adherence</td>
<td>Yes: Social worker, psychologist referral</td>
</tr>
<tr>
<td></td>
<td>No: Recruit tube feedings</td>
</tr>
</tbody>
</table>

#### Ongoing reassessment

BMI formula: weight (kg) / (height (m))^2
Example: Weight = 60 kg, Height = 1.65 m (1.65 m^2)
Calculation: 60 / 1.65^2 = 18.4

<table>
<thead>
<tr>
<th>Question</th>
<th>Action</th>
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<tbody>
<tr>
<td>metastatic disease</td>
<td>Consider chemotherapy, radiation, surgery</td>
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<td></td>
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<tr>
<td>Inadequate oral intake (adequate)</td>
<td>Consider tube feedings</td>
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<tr>
<td>Clinical food diary or recall</td>
<td>Consider tube feedings</td>
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<tr>
<td>Adequate intake</td>
<td>Consider tube feedings</td>
</tr>
</tbody>
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Nutritional Outcomes

Improving nutritional outcomes is a complex process

Our CF Team created a “Fishbone” diagram as part of the Dartmouth Institute’s Learning and Leadership Collaboration to show these complexities
What is Cystic Fibrosis?

An inherited chronic disease that affects multiple organ systems.

A defective gene causes the body to produce unusually thick, sticky mucus that:

- Clogs the lungs and leads to life-threatening lung infections
- Obstructs the pancreas and prevents the small ducts from transporting natural enzymes to the intestine where food is broken down and nutrients are absorbed

The effects of CF on the pulmonary and gastrointestinal (GI) systems most directly affect nutritional care.
Nutritionist - Goals of Treatment

- Minimize fat in the stool
- Promote optimal lung function

In order to:
- Achieve normal growth and development in children
- Maintain good health in children and adults, which includes preventing vitamin deficiencies
Pulmonary Symptoms and Conditions

- Chronic Cough
- Increased Work of Breathing
- Repeated Bronchial Infections
- Nasal Polyps

Energy Needs
GI Symptoms and Conditions

- Meconium Ileus
- Rectal prolapse
- CF Related Diabetes (CFRD)
- Malabsorption
Signs of Malabsorption

- Abdominal pain or cramps
- Frequent gas
- Feeling bloated or full
- Voracious appetite
- Poor weight gain or growth
- Decreased fat soluble vitamin levels
- Low albumin
Treatment of Malabsorption

- 85-90% of patients are pancreatic insufficient
- Infants may be pancreatic sufficient, but within first year become pancreatic insufficient

GOAL of TREATMENT

Minimize fat in the stool to promote normal growth and development in children and good health in children and adults
Pancreatic Enzymes (PERT)

- Capsules containing small beads
- Each bead is “enterically” coated
  - Protects it from stomach acids
  - Dissolves in small intestine where nutrients absorbed
- Long term high doses of enzymes
  - May be associated with colonic strictures and fibrosing colonopathy
- Inadequate enzymes
  - May result in distal intestinal obstructive syndrome (DIOS)
Pancreatic Enzymes

- Should be taken immediately before meals, snacks, and formula including tube feeds
- Work for about one hour after taking them
- Should never be crushed or chewed

- For infants and small children:
  - Capsules may be opened and beads mixed with soft acidic foods (like applesauce) to be given by spoon
  - Helps to give breast milk or formula after beads are given
Pancreatic Enzymes

- Dosages vary depending on the individual’s degree of fat malabsorption and the amount of fat eaten.

- Dosages are calculated based on units of lipase/capsule and the patient’s weight. We prescribe for a specific number of units of lipase/per kg of the patients body weight/meal.

- Prescriptions are then written for the number of capsules to be taken before each meal or snack.
Enzyme Dosing Guidelines

- **Infants**
  - 2000-4000 Units lipase/120ml breast milk/formula or
  - 1000 Units lipase/kg/meal

- **Children**
  - 1000 Units lipase/kg/meal
  - 500 Units lipase/kg/snack

- **Older Children and Adults**
  - Start with 500 Units lipase/kg/meal
  - Desired range – 1000-2000/Units lipase/kg/meal
  - Maximum dose
    - 2500 Units lipase/kg/meal
    - 10,000 Units lipase/kg/day
## Current PERT Products

<table>
<thead>
<tr>
<th>Brand</th>
<th>Lipase</th>
<th>Protease</th>
<th>Amylase</th>
<th>Notes</th>
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</thead>
<tbody>
<tr>
<td><strong>Creon ®</strong></td>
<td>3,000</td>
<td>9,500</td>
<td>15,000</td>
<td>G-tube admin</td>
</tr>
<tr>
<td></td>
<td>6,000</td>
<td>19,000</td>
<td>30,000</td>
<td></td>
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<td></td>
<td>12,000</td>
<td>38,000</td>
<td>60,000</td>
<td></td>
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<td></td>
<td>24,000</td>
<td>76,000</td>
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<td><strong>Zenpep®</strong></td>
<td>3,000</td>
<td>10,000</td>
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<td>25,000</td>
<td>85,000</td>
<td>136,000</td>
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<td><strong>Pancreaze®</strong></td>
<td>4,200</td>
<td>10,000</td>
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<td></td>
<td>16,800</td>
<td>40,000</td>
<td>70,000</td>
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<tr>
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<td>21,000</td>
<td>37,000</td>
<td>61,000</td>
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</table>
Nutrition and Growth

The Body Mass Index (BMI) is a statistical measure of body weight based on a person's weight and height. Current CFF guidelines use this measure to assess nutritional risk for children over 2 years old and for adults.

FOR CF PATIENTS:
RED < 10 % (High Risk) Urgent Intervention
YELLOW 10-49% (Mild-Moderate Risk)
GREEN > 50% (Well Nourished)

BMI Formula: \( \text{weight (kg)} / [\text{height (m)}]^2 \)
Example: Weight = 68 kg, Height = 165 cm (1.65 m)
Calculation: \( 68 \div (1.65)^2 = 24.98 \)
Nutrition and Growth

- Birth – 2 yrs old
  **GOAL:** weight/height $\geq 50^{th}$ percentile

- 2-20 yrs old
  **GOAL:** BMI $\geq 50^{th}$ percentile

- > 20 yrs old
  **GOAL:** BMI $\geq 50^{th}$ percentile

Standards used to evaluate a healthy population are **NOT** the same as for CF patients – BMI below 50% is considered an increased nutritional risk for CF patients.
Nutrition and Growth

- Nutritional status has been “associated” with lung disease.

- Higher pulmonary function tests (PFTs) correlate with increased BMI values.

- The correlation of a higher BMI with a higher PFT value levels off at a BMI of about 50% (based on age)
FEV1 Percent Predicted vs BMI Percentile, Patients 6 to 20

- Nutrition and Growth

BMI Percentile vs FEV1 Percent of Predicted for Males and Females.
Nutrition

- Generally, CF patients require 110-200% of the RDA for calories and 200% RDA for protein.

A high fat, high calorie diet is prescribed.
General Nutrition Tips

- **Infants** – breast milk or formula may need caloric supplementation
- **Children and Adults**
  - Add butter or margarine to foods
  - Add oils, cheese, heavy cream, and sour cream
  - Whole milk
  - High calorie snacks – supplements, flavored syrups on ice cream
Vitamins and Minerals

- Fat soluble vitamin needs increased (A,D,E,K)
- Require water miscible forms
  - AquADEKs®
  - Source CF®
  - Vitamax®

- Dosages
  - Infants: 1 ml daily
  - Children ages 1-4 years: 2 ml or ½ tab daily
  - Children ages 4-10: 1 tab daily
  - Children >10: 2 tabs daily

- Serum vitamin levels are assessed annually
Sodium/Salt

Sodium lost through perspiration can lead to dehydration

• Infants
  • 1/8-1/4 tsp salt added to breast milk/formula daily (Size of a packet of salt provided at fast food restaurants like McDonalds)

• Children and Adults
  • Salty foods; add salt to food – up to 1-2 tsp per day (May need more depending on climate, season, and activity levels)
# Nutrition Algorithm

<table>
<thead>
<tr>
<th>Question</th>
<th>Action</th>
<th>Date</th>
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<td>Is patient in green zone?</td>
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<td></td>
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<td>2</td>
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<td>Yes</td>
<td>3</td>
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<td>Yes</td>
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<td></td>
<td>Yes</td>
<td>5</td>
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<td>Yes</td>
<td>6</td>
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<td></td>
<td>No</td>
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<tr>
<td>Met weight goal and/or improvement from previous visit?</td>
<td>Yes</td>
<td>7</td>
</tr>
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<td></td>
<td>No</td>
<td></td>
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<tr>
<td>Evidence of pulmonary exacerbation or other medical problem? (i.e., smoking, allergies)</td>
<td>Yes</td>
<td>8</td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>9</td>
</tr>
<tr>
<td></td>
<td>No</td>
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<td>Evidence of malabsorption?</td>
<td>Yes</td>
<td>10</td>
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<tr>
<td></td>
<td>Yes</td>
<td>11</td>
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<td>Yes</td>
<td>14</td>
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<td>No</td>
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<tr>
<td>Is oral dietary intake adequate?</td>
<td>Yes</td>
<td>15</td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>16</td>
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<td>19</td>
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<tr>
<td></td>
<td>No</td>
<td></td>
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<tr>
<td>Consider appetite stimulant</td>
<td></td>
<td></td>
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<tr>
<td>Initiate tube feedings</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**On-going reassessment**

- **BMI Formula**: weight (kg) / (Height (m)^2)
- Example: Weight = 56 kg, Height = 1.65 m, BMI = 21.97
- Calculation: 56 / (1.65)^2 = 21.97

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**Nutrition Algorithm**

[Pharmacy logo]

[Sign-off]

[Double-sided form] 1/2/2020
<table>
<thead>
<tr>
<th>Evidence of pulmonary exacerbation or other medical problems? (i.e., Sinusitis, allergies)</th>
<th>Yes</th>
<th>Treat</th>
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<tbody>
<tr>
<td></td>
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<tr>
<td></td>
<td>Yes</td>
<td>Referral________________________</td>
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</tr>
<tr>
<td>Evidence of malabsorption?</td>
<td>Yes</td>
<td>Discuss enzyme dosing, timing, storage, expiration</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Change enzyme dose</td>
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<tr>
<td></td>
<td></td>
<td>PPI/H2 Blocker</td>
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<tr>
<td></td>
<td></td>
<td>Consider bacterial overgrowth</td>
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<td></td>
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<td>72-hr fecal fat test</td>
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<td></td>
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<td>Laxative</td>
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<td>GI referral</td>
</tr>
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<td></td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Is oral dietary intake inadequate?</td>
<td>Yes</td>
<td>Educate on supplements; increased calories in meal/snack selections</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Consider metabolic cart</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Problem solve time management barriers</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Feeding/swallowing issues referral</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Discuss role of tube feedings</td>
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<tr>
<td></td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Should CFRD be considered?</td>
<td>Yes</td>
<td>Finger stick</td>
</tr>
<tr>
<td></td>
<td></td>
<td>OGGT</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td>Endocrine referral</td>
</tr>
</tbody>
</table>
Enzymes Not Working

- Acid blockers
  (neutralize stomach acid allowing enzymes to pass into small intestine)
- Possible non-adherence
  (with snacks or forgetting to take enzymes at all)
- Switch brand of enzyme
- Other cause of loose stools? Refer to GI
Inability to Gain Weight

- **Is food intake adequate to meet increased caloric needs?**
  Financial issues, time management, appetite, metabolism?

- **Evidence of illness or medical problem?**
  Lung infection? Exacerbation of lung symptoms? Anemia?

- **Malabsorption?**
  Enzyme availability, dosing, timing, storage, expiration? Need to change enzymes, add H2blocker/PPI, perform a fecal fat test?
Inability to Gain Weight

- **Other GI problems?**
  Gastrointestinal reflux, constipation, bacterial overgrowth

- **CFRD?**
  Undiagnosed or not compliant

- **Zinc**
  - May improve weight gain
  - 1 mg/kg of elemental zinc per day for 6 months
Tube Feedings

Patients have difficulty gaining/maintaining weight

- Usually provide ~50% of total daily caloric requirements
- Run ~ 8-10 hours overnight
- Often beneficial for concentrated formulas
- Meal enzyme dosage before and after
- Common formulas used include: Peptamen 1.5, Nutren 2.0, 2 cal HN
Cystic Fibrosis Related Diabetes - CFRD

- Start monitoring with OGTT and HgA1C tests at age 10 (HgbA1C may show a false low)
- Often diagnosed during teen years
- Different type of Diabetes
  - Symptoms of both Type 1 and Type 11
  - Pancreas does produce insulin, but not usually enough to meet CF patients elevated energy needs
CFRD

- Insulin treatment common

- Hyperglycemia is exacerbated by high doses of steroids

- Dietary advice – continue with high fat, high protein diet adjusting insulin to blood sugar – many endocrinologists not familiar with CFRD differences
Success Story

- Enzyme dose increased
- Switched to whole milk
- Started having an instant breakfast drink after school each day
- Used his smart phone alarm to remind himself to take enzymes before after-school snack
- Started having higher calorie snacks at night and weekends
- Parents bought a small refrigerator for the patient’s bedroom.
New Directions

- In August 2011, our CF Team (with Dr. Cori Daines as principal investigator) applied to the CFF for a QI grant.
- Our goal is to standardize CF adult inpatient care processes to improve inpatient outcomes based on accepted recommendations and guidelines.
- We ultimately plan to carry these changes over into the pediatric population.
Initial Goals

- Improve communication between the inpatient and outpatient CF Care Teams
- Set specific time limited goals for hospitalization to decrease the length of stay
- Improve inpatient pulmonary and nutrition outcomes
- Strengthen patient and family inpatient care involvement
References

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- Cystic Fibrosis Nutrition 101 (2011) Port CF
  - Amanda Leonard, MPH, RD, CDE – The Johns Hopkins Children’s Center
  - Terri Schindler, MS, RD – Rainbow Babies and Children’s Hospital
- CF Foundation Evidenced-Based Guidelines for Management of Infants with Cystic Fibrosis: JPediatr 2009;155(6);S73-93.
- Cystic Fibrosis-related Diabetes; Pediatric Diabetes 2—9:10 (Suppl.12): 43-50
- Evidence-Based Practice Recommendations for Nutrition-Related Management of Children and Adults with Cystic Fibrosis and Pancreatic Insufficiency: Results of a Systematic Review; Journal of the American Dietetic Association: May 2008; Vo. 108 #5; 832-839
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