Nutrition for Children with Special Health Care Needs

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Objectives
• Define CSHCN and describe common nutritional risk factors and concerns in the population
• Discuss the nutrition screening, assessment and treatment strategies for CSHCN of various etiologies
• Describe available nutrition resources for CSHCN

CSHCN
• Children with special health care needs are those
  – who have or are at risk for chronic physical, developmental, behavioral, or emotional conditions and
  – who also require health and related services of a type or amount beyond that required by children generally.

Prevalence of CSHCN
• Survey data suggests that
  – 12% of US children <18 years old have a chronic physical, developmental, behavioral, or emotional condition and
  – have used health related services beyond those required by children generally.

Common Nutrition Problems for CSHCN
• Altered growth
• Inadequate energy and nutrient intake
• Feeding problems
• Medication nutrient interactions
• Need for enteral feedings
• Chronic constipation or diarrhea
• Use of alternative therapy

Categories of CSHCN
• Neurodevelopmental
  – Autism
• Environmental
  – Fetal Alcohol Syndrome
• Chromosomal
  – Down Syndrome
• Metabolic Disorder
  – Phenylketonuria (PKU)
Assessment Guidelines for CSHCN

- Growth
- Biochemical
- Clinical
- Dietary

Growth

- Set realistic growth expectations
- Monitor growth over time
- Specialized growth charts
  - Small sample size
  - May not reflect ideal growth rates
  - Use in conjunction with CDC charts

Medication-Nutrient Interactions (Biochemical)

- Medications can affect the intake, absorption, metabolism and excretion of nutrients
- Medication side effects

Many CSHCN have feeding problems

- A feeding problem is
  - The inability or refusal to eat certain foods because of:
    - Neuromuscular dysfunction,
    - Obstructive lesion,
    - Psychosocial factors,

Common Feeding Problems (Clinical)

- Weak suck
- Poor lip closure
- Tongue thrust
- Drooling
- Bite reflex
- Hypersensitivity around the face and mouth
- Hyperactive gag reflex
- Swallowing difficulties
- Behavioral component

Nutrient Intake (Dietary)

- Energy needs maybe increased or decreased
- Energy needs estimated
  - Kcal/kg
  - Weight history
  - Diet history
  - Previous feeding history
- Protein, Fat, Vitamins and Minerals use DRI’s for age
Autism Spectrum Disorders (ASD)

- Occurs in 1 in 166 births
- More males than females
- ~70% have mental retardation; ~20% have seizures
- Multi-gene etiology

Diagnosis of ASD

- Autism
- Pervasive Developmental Disorder, Not otherwise Specified (PDD-NOS)
- Asperger’s Syndrome

Signature Behavioral Features

- Impaired social interaction
- Delayed and/or limited communication
- Restricted or stereotypic behaviors

Behaviors that Impact Nutritional Status

- Difficulty with transitions
- Impaired communication skills
- Social interaction difficulties
- Easily over-stimulated
- Short attention span
- Limited range of interests
- Need for routine

Feeding Concerns

- Difficulty in transitioning to textures
- Increased sensory sensitivity
- Restricted intake due to color, texture, temperature, odor
- Refusal of “new” foods
- Difficulty with changes in mealtime routines
- Refusal of vitamin/mineral supplements

ASD and Nutritional Status

- Growth parameters tend to be typical and within normal limits
- Energy and protein needs usually met
- Vitamins/minerals at risk if very “picky”
- Elimination diets result in marginal or deficient intake
- Typical intake: crackers, breads, cereals, chicken nuggets, hot dogs, little/no fruits and vegetables; preference for dry, crunchy textures
Intervention Techniques that May Work

• Small changes
• Keep mealtimes constant
• Calm, quiet surroundings at meals
• Offer small servings of a few foods
• Offer new foods with foods already liked
• Repeat food exposures

Intervention Strategies that Probably Won’t Work

• Bribing a child to eat a food
• Forcing a child to “take a bite” or “have at least one taste”

Alternative Therapies

• Gluten-free and casein-free diet
• Megavitamins, i.e. Super NuThera – Mega B6, magnesium,
  • Others

GFCF Diets: The Dilemma

Anecdotal vs Research

• Anecdotal reports of improved behaviors/symptoms
  – Eye contact, sociability, language
  – Bowel movements, sleep patterns
• Research limitations
  – No double-blind, placebo-controlled studies
  – Dietary intake not clarified/measured
  – Small sample size

Evaluating Alternative Therapies

• Is the product or therapy safe for children?
• How will the therapy be evaluated?
  – What will be monitored…behavior?
  – Which change related to which therapy?
• Nutritional deficiencies?
• Expense of special products/supplements?

Fetal Alcohol Spectrum Disorders

• FAS
  – Permanent birth defect caused by maternal alcohol consumption during pregnancy
  – 0.5-2.0 cases/1000 live births
• ARND
  – Neurocognitive and behavioral problems without facial features of FAS
FAS Diagnostic Features

• Growth Deficiency
  – Prenatal and/or postnatal
  – Height and/or weight below 10%
• Facial Anomalies
  – Short palpebral fissure lengths
  – Smooth philtrum
  – Thin upper lip

CNS Abnormalities

• Microcephaly
• Abnormal MRI findings
• Global developmental delay
  – Cognition, language, memory, executive function, sensory-motor
  – Range from mental retardation to low normal range

Nutrition Concerns

• Pregnancy
  – Alcohol abstinence for preconceptual and pregnant women
  – Impairs placental transfer of essential nutrients
  – Increases the risk of prematurity or SGA

Lactation

• Alcohol disseminates into blood and breast milk
• Questionable effect on baby’s developing brain
• May change the taste of breast milk and cause decreased intake and growth

Growth

• FAS children often have growth deficiency- ht and/or wt less than 10%
• Distinguish between FTT and FAS

Feeding Difficulties

• Weak Suck
  – Length of feedings, sign of CNS abnormalities
• Distractibility
  – Inability to focus and signal hunger
  – Decrease stimuli (swaddle, dim lights, separate room)
• Sensory Issues
  – Weakness in oral motor planning and swallowing coordination
• Behavior
**Down Syndrome**

- 1 in 800 births; increased incidence with maternal age
- Genetic disorder
- Health concerns
  - Developmental delay
  - Hypotonia
  - Short stature
  - Increased risk for overweight
  - Potential for congestive heart disease, hearing/vision defects, hypothyroidism, premature aging, seizures, constipation, dental hypoplasia

**Down Syndrome Assessment/Intervention**

- Growth
  - Use DS growth charts with CDC charts
- Nutrition and Feeding
  - Height and activity level to determine energy needs; kcal/cm height for 5-11 year olds (boys 16.1 cm/ht; girls 14.3 cm/ht)

**Down Syndrome Nutrition Assessment (Continued)**

- Evaluate progression with textures and self-feeding skills (refer to feeding team?)
- Constipation (due to hypotonia and inactivity)-prevention and intervention
- Physical Activity- prevention of overweight and support of wellness
- Supplement use (Nu TriVene-D, Piracetam, herbs and other alternative therapies

**Metabolic Disorder**

Genetic disorders, such as inborn errors of metabolism, e.g.,
- phenylketonuria
- maple syrup urine disease
- urea cycle disorders

**Metabolic Disorder: Implications for Nourishment**

Affected metabolic pathways are involved in producing energy or building body tissue
- Requires ensuring individual is adequately fed
- Requires restriction of specific nutrients and supplementation of others
- Requires careful monitoring of all nutrients
- Can expect typical growth
  - if intercurrent illness does not interfere

**Nutrition Support for PKU**

- Correct substrate imbalance
  - restrict phenylalanine intake to normalize plasma concentration
  - $\text{Phenylalanine} \xrightarrow{\text{hydroxylase}} \text{Tyrosine}$

- Supply product of reaction
  - Supplement tyrosine to maintain normal plasma tyrosine levels
  - $\text{Phenylalanine} \xrightarrow{\text{phenylalanine hydrolase}} \text{Tyrosine}$
Goals of Nutrition Support: Phenylketonuria [PKU]

- Maintain plasma phenylalanine (phe) between 1-6 mg/dl
  - without PKU phe = 1.0 mg/dl
- Maintain plasma tyrosine (tyr)
  - between 0.9-1.8 mg/dl
  - normal = 0.9-1.8 mg/dl

Monitoring Adequacy of Treatment

- Measure plasma amino acids
  - maintain in treatment range
- Monitor nutrient intake
  - restrict phenylalanine, supplement tyrosine, adequate protein, energy, nutrients to support growth and ensure good health
- Monitor growth increments
  - typical growth expected
- Monitor cognitive development
  - typical achievement expected

Goals of Lifetime of PKU Management

- To maintain metabolic balance while providing adequate nutrients and energy for normal physical and intellectual growth

Tools of Management

- Specialized formula provides
  - 85% of energy and protein needs
  - 100% of vitamin and mineral needs
  - tyrosine supplements
  - no phenylalanine
- Phenylalanine to meet requirement from infant formula or foods

Tools of Management: Low protein food products

- Phe content of regular products:
  - rice = 250 mg/c
  - pasta = 325 mg/c
  - bread = 105 mg/sl
  - cheerios = 93 mg/1/2 cup
  - saltines = 65 mg/5 crax
  - potatoes = 145 mg/c

- Phe content of low protein products:
  - LP rice = 10 mg/cup
  - LP pasta = 5 mg/cup
  - LP bread = 10 mg/sl
  - LP cheerios = 2 mg/1 c
  - LP saltines = 5 mg/5 crax

Sample Menu ~1 year old child

<table>
<thead>
<tr>
<th>Food</th>
<th>Pro</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/4 c cheerios</td>
<td>.8</td>
</tr>
<tr>
<td>1/2 banana</td>
<td>.6</td>
</tr>
<tr>
<td>1/2 c milk</td>
<td>4</td>
</tr>
<tr>
<td>2 graham crax</td>
<td>2</td>
</tr>
<tr>
<td>1/2 c milk</td>
<td>4</td>
</tr>
<tr>
<td>1/4 tuna sandwich</td>
<td>8</td>
</tr>
<tr>
<td>1/2 peach</td>
<td>.6</td>
</tr>
<tr>
<td>2 saltines</td>
<td>.6</td>
</tr>
<tr>
<td>1/2 c juice</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>32.5</td>
</tr>
</tbody>
</table>

Phe req. = 250 mg
Sample Menu
~1 year old child

- To meet 250 mg phe (5 gm protein)
  - minus milk, tuna, cottage chez = 32.5 - 27 = 5.5
- Is this adequate protein to support growth?
- Is this an adequate energy intake to support growth?
- What about adequacy of nutrients?

Typical Food Pattern for Child with PKU

- 1 year old child
  - Weight & length 50thile
- Energy needs:
  - 1000-1300 kcal
- Protein needs:
  - 20 gm/day
- Phenylalanine needs:
  - 250 mg phe/day
- Formula prescription:
  - Phenyl-Free- 125 gm
  - Similac powder- 50 gm
  - water to 40 oz
- Food
  - 25 mg phe, 200+ kcals, 0.5 gm pro

Effect of single amino acid deficiency on growth

- Initial referral from mom (Fri, 4:45 PM)
- History from mom
  - 3 mo old infant with PKU
  - Normal BW- 8 # 4 oz.
  - Current wt. - 9 # 5 oz.
  - Blood phe levels - she didn't know, told they were low
- No referral from previous clinic

Any ‘red flags’ here?

Initial Growth Data

- Feeding hx
  - Currently Lofenalac
  - Several different formulas tried bec. of poor growth
  - Intake - 16 oz/day
- Initial visit
  - Physical exam
    - Infant cranky, no subcutaneous fat, yellow skin, overtly malnourished
  - Plan
    - Full strength infant formula
    - Monitor blood phe levels and intake
    - Complete, repeat NBS

- Initial plan
  - Infant's blood drawn ASAP (Sat AM) to document Dx
  - Offer infant 1:1 Lofenalac + standard infant formula to begin to assure adequate nourishment
  - Clinic visit (Mon AM)
    - Blood draw to document AA levels
    - Physical exam
    - Nutrition assessment
    - Document history, etc

Initial plan

- Physical exam
  - Infant cranky, no subcutaneous fat, yellow skin, overtly malnourished
- Plan
  - Full strength infant formula
  - Monitor blood phe levels and intake
  - Complete, repeat NBS

- Initial phe level = 'off the charts' - per mom
  - F/U levels = normal/<normal - per mom
  - At initial visit: phe = 0.2 mg/dl; tyr = 1.6 mg/dl
  - After 2 weeks: phe = 2.0 mg/dl; tyr = 2.1 mg/dl
  - After 1 mo; phe = 3.3 mg/dl; tyr = 1.3 mg/dl
### Nutrient intake

<table>
<thead>
<tr>
<th>Age</th>
<th>At 3 mo</th>
<th>At 4 mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>Formula</td>
<td>Lofenalac</td>
<td>PF + Enf</td>
</tr>
<tr>
<td>Amount</td>
<td>16 oz</td>
<td>24-30 oz</td>
</tr>
<tr>
<td>Protein</td>
<td>10.5 g</td>
<td>12.9 g</td>
</tr>
<tr>
<td>Energy</td>
<td>320 kcal</td>
<td>540 kcal</td>
</tr>
<tr>
<td>Phe</td>
<td>56 mg</td>
<td>352 mg</td>
</tr>
<tr>
<td>Tyr</td>
<td>560 mg</td>
<td>900 mg</td>
</tr>
</tbody>
</table>

### Effect of single amino acid deficiency on growth

### Issues

- Understanding the disorder
  - principles of treatment
  - principles of MNT
- Monitoring of treatment
- Coordination of care

### Resources

- University of Washington PKU Clinic website
  - http://depts.washington.edu/pku
- National PKU News website
  - http://pkunews.org
- Really Living with PKU video

### Challenges Related to Nutrition and CSHCN

- Feeding is an integral part of parenting
- Energy and nutrient requirements are individual
- Reliable dietary intake is difficult to obtain
- The use of supplements and undocumented nutrition therapies

**SUMMARY:** Individualize nutrition care