Does PKU Camp Education Improve Plasma Phenylalanine Levels?

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Collaborators

- OUHSC
- Ashley Taylor MHS, PA-C
- Ashley Ethriedge, MS, RD/LD
- Mary Monks, RN

- University of Arkansas
- Stephen Kahler, MD
- Brandi Nichols, RD/LD

Investigators
Many Others to Thank

- Kevin Winters, BS
- Andrea Wierenga, PhD
- Jae Lindsey Chaloner, LCGC
- Michael Anderson, PhD
- Ashley Davis LGC
- Chunyan Wang
- Danny Cavett
- Jenny Rodgers
- Sarah Foster
- Andrew Spaziani
- Renee Bethel, RD/LD
- Matt Grim

Project Aims

- Establish a Phenylketonuria (PKU) camp in the Heartland region.
- Education for patients with PKU.
- Evaluation of the effectiveness of education sessions at improving plasma phenylalanine levels in both the short and long term.
Background

• Medical nutrition therapy for patients with PKU includes ‘diet for life’ for successful long term control of PKU.
• Education is essential for PKU patients to understand their strict diet and therapeutic regimens.
• PKU camp is a way to provide education in a fun and interactive way.
  • Daily education sessions
  • Children with PKU were able to network with each other at camp along with children with other chronic disorders.
  • Shared life experiences.

Camp Cavett

• Danny Cavett founded the Cavett Kids Foundation in 1997
• Largest camp at Lake Texoma (OK and TX border)
• Over 160 children participate with different illnesses including cancer, heart disease, kidney disease, GI disorders, and lung disorders.
• Staffed by physicians, nurses, dietitians, other health care providers.
  • Over 500 recurring volunteers
• Cabins provided
• Activities such as water sports, paintball, rock climbing, ropes course, and beauty salon.
Target Population and Recruitment

- Children and Adolescents between 8-18 yrs old from OK and AR
  - 14 total participants
  - 1 participant from MO
- Brochures given in clinic.
- Discussed during clinic visits via physicians, PA, and dietitians.
- Emails to patients on the PKU mailing list.
- Advertised in PKU quarterly newsletter.
- Announcement to medical directors at Tulsa and Missouri clinics.

Travel

- Bus from AR
- Bus from OUHSC
- Travel allowance (gift cards) given to families who brought their children to camp.
Medical Nutrition Management

- Menus with PKU food options.
- Dietitians counted amount of phenylalanine (phe) consumed by PKU campers and kept track of phe allowances.
- PKU formula and Kuvan brought from home.
  - Ensured routine at camp was similar to home routine.
  - PKU food prepared and served by volunteers.

Education Sessions

- Session 1 – Kevin Winters, 1st year medical student
  - Information on completing filter papers for plasma phenylalanine and tyrosine analysis
- Session 2 – Jae Lindsay Chaloner, CGC
  - Genetics of PKU
  - Science project – DNA extraction from banana
- Session 3 – Ashley Ethriedge, RD/LD
  - Food preparation activity
  - Practiced counting amount of phe in foods
- Session 4 – Ashley Taylor, MHS, PA-C
  - Question and answer session
  - Group Discussion
Short Term Results

- Blood spots for plasma phenylalanine levels were collected on all 14 test subjects at the beginning and end of camp.
- Blood spots analyzed at the OUHSC biochemical laboratory.
  - Amino acids were determined from dried blood on filter paper by ion-exchange chromatography.
Short term results

<table>
<thead>
<tr>
<th>Mean Difference, F-I (mg/dL)</th>
<th>p-value</th>
<th>Power</th>
</tr>
</thead>
<tbody>
<tr>
<td>-4 ± 4</td>
<td>0.005</td>
<td>0.872766</td>
</tr>
</tbody>
</table>

- Out of 14 test subjects, 12 showed a decrease in plasma phenylalanine levels.
- The mean difference was -4 ± 4 mg/dL.
- A paired t-test indicated a p-value of 0.005, which suggests statistical significance by conventional standards.
- 3 subjects had phenylalanine concentrations in the desired treatment range (2-6 mg/dL) before treatment, while 5 subjects were below 6 mg/dL after treatment.
- 8 subjects had phenylalanine concentrations of 10.0 mg/dL or less after treatment compared to 4 subjects before treatment. These findings suggest clinically significant results as well.

Long Term Results

- A review of the laboratory results in the patients’ clinic chart was conducted to determine their plasma phenylalanine levels 6 months before and 6 months after camp.
Long Term Results

- No significant trend in plasma phenylalanine levels over time.

<table>
<thead>
<tr>
<th>Time Period</th>
<th>Mean PKU Before Camp</th>
<th>Mean PKU After Camp</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>13.42</td>
<td>15.08</td>
</tr>
<tr>
<td>3</td>
<td>13.28</td>
<td>13.65</td>
</tr>
<tr>
<td>4</td>
<td>12.18</td>
<td>12.38</td>
</tr>
<tr>
<td>5</td>
<td>31.98</td>
<td>12.52</td>
</tr>
<tr>
<td>6</td>
<td>11.28</td>
<td>13.52</td>
</tr>
<tr>
<td>7</td>
<td>8.89</td>
<td>10.36</td>
</tr>
<tr>
<td>8</td>
<td>8.50</td>
<td>7.36</td>
</tr>
</tbody>
</table>
Pre- and Post- Education Exams

- The 14 campers with PKU were given a 20 question pre-and post-test on day 1 and day 4 of camp.
- Education is crucial for improved compliance.

Pre- and Post- Camp Exam Scores

![Bar chart showing pre- and post-exam scores for 14 participants.]
Exam Results

- All 14 participants showed an increase in exam scores.
- Improvement in exam scores indicate an increase in knowledge about PKU.

<table>
<thead>
<tr>
<th>Mean Difference, F – I (%)</th>
<th>p-value</th>
<th>CI 95</th>
</tr>
</thead>
<tbody>
<tr>
<td>38 ± 28</td>
<td>0.0002</td>
<td></td>
</tr>
</tbody>
</table>

Overall Results

- Short term results and Exam Scores results showed significant improvement.
- Long term results showed no significant improvement over time.
Discussion

- Positive feedback from participants and families.
  - Families expressed interest in attending future camps.
- Education session improved participant knowledge.
- Networked with other children with PKU and children with other chronic disorders.
- Since children with chronic disorders other than PKU attended camp, regular foods were available.
  - Simulated more of a real life experience that a child with PKU would normally experience.

Challenges

- Recruitment was short of our goal of 20 participants.
  - 15 enrolled and 1 didn’t attend that enrolled
- Became more aggressive with recruitment
  - Contacted every child within the 8-18 year old age range by phone.
  - Reached out to other states such as MO. Gained one extra participant.
- Only about 4 participant plasma phenylalanine measurements per month were collected during the chart review for long term results.
Ideas for Next PKU Camp

• Parents are a huge part of patient compliance, especially the younger patients.
  • Next time, there could be an orientation for the parents with an education session for the parents.
  • Quarterly follow-up education sessions for both the patient and his/her parents. Lab measurements could be done on the patients at these education sessions.
• Include more states in the Heartland region
  • Possible have both a North and a South camp in the Heartland region.

Questions?