

Does PKU Camp Education Improve Plasma Phenylalanine Levels?

Ashley C. Taylor, MHS, PA-C
Physician Associate of Pediatrics
Program in Human Genetics

This research is supported by a grant
from the Heartland Genetics and
Newborn Screening Collaborative
(HRSA U22MC03962). This study was
approved by the OUHSC IRB #15102

Collaborators



Cavett Kids Foundation

Investigators

- OUHSC
 - Ashley Taylor MHS, PA-C
 - Ashley Ethriedge, MS, RD/LD
 - Mary Monks, RN
- University of Arkansas
 - Stephen Kahler, MD
 - Brandi Nichols, RD/LD

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 8-18yrs 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.

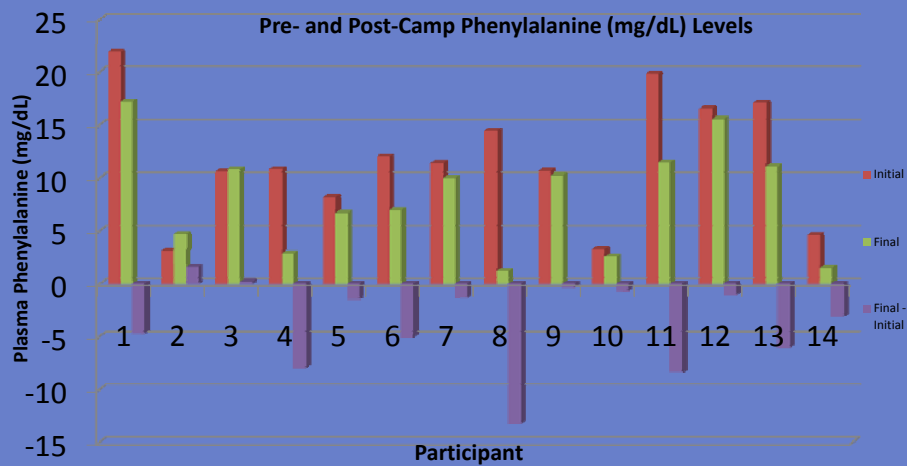
Education Sessions

- 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 Ethridge, 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



Short term results

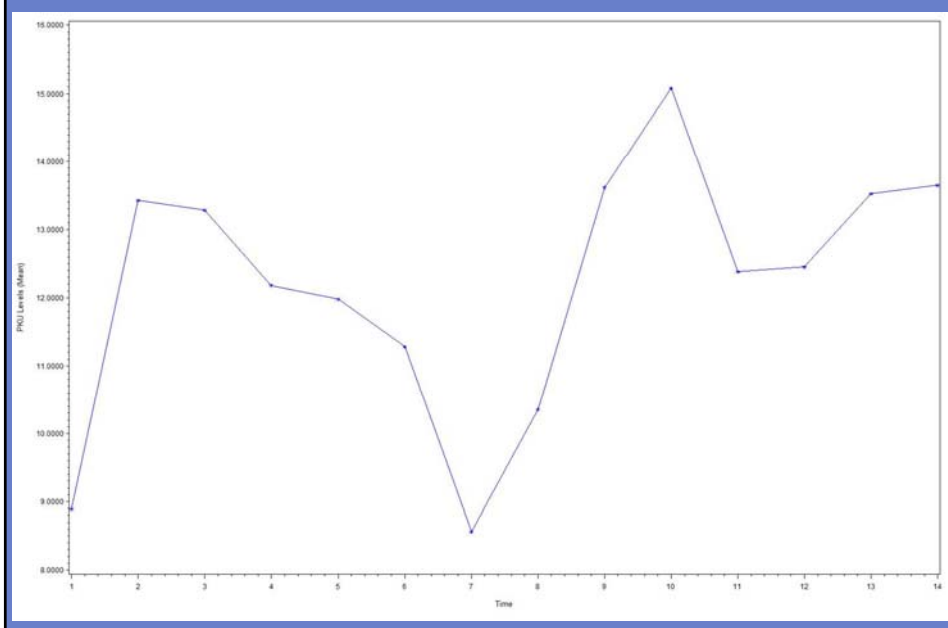
Mean Difference, F-I (mg/dL)	p-value	Power
-4 ± 4	0.005	0.872766

- 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



Long Term results

- No significant trend in plasma phenylalanine levels over time.

Before Camp

Time Period	Mean PKU
2	13.42 ^a
3	13.28 ^a
4	12.18 ^a
5	11.98 ^a
6	11.28 ^{ab}
1	8.89 ^{ab}
7	8.56 ^b

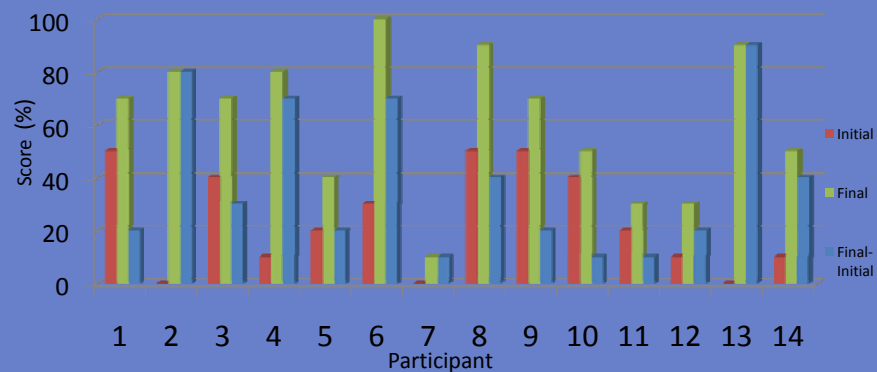
After Camp

Time Period	Mean PKU
10	15.08 ^a
14	13.65 ^a
9	13.62 ^a
13	13.52 ^a
12	12.45 ^a
11	12.38 ^a
8	10.36 ^{ab}
7	8.56 ^b

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



Exam Results

Mean Difference, F - I (%)	p-value	CI _{.95}
38 ± 28	0.0002	22, 54

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

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?

