Emory BH4 & PKU Research Study  
SINGH RESEARCH GROUP  
404-778-8522  
www.genetics.emory.edu/NUTRITION/BH4andPKU/

SINGH RESEARCH GROUP  
Emory BH4 & PKU Research Opportunity

Participant ID ____________________________

Next visit date ___________________________

This log book should be used during the study period:

Study date________________ to Study date________________

Month/Day/Year Month/Day/Year

Date log book sent/given _____________________
Tracking your Kuvan™ (BH4) Prescription

**Instructions:** Anytime there is a change in your Kuvan™ prescription, either with the *mg* dose or with the number of pills you swallow, please write it here.

<table>
<thead>
<tr>
<th>Date when new dose or pill # was prescribed</th>
<th>Number of pills/day</th>
<th>New dose (mg Kuvan™/day)</th>
<th>How do you usually take this medicine? (ie: swallow whole, mix with apple juice)</th>
</tr>
</thead>
<tbody>
<tr>
<td>EXAMPLE: Nov.14, 2008</td>
<td>12</td>
<td>1200mg/day</td>
<td>Crush pills and mix with water</td>
</tr>
</tbody>
</table>

**NOTES:** 1 pill=100 milligrams (mg)
Tracking your PHE/Protein prescription and your Medical Food (Formula) Prescription

**Instructions for tracking PHE/protein prescription:** Anytime the dietitian or doctor changes your prescribed PHE or Exchanges or Protein allowance, please write it here.

<table>
<thead>
<tr>
<th>Recommended daily PHE (mg/day)</th>
<th>Protein equivalents (g/day)</th>
<th>PHE Exchanges (# of exchanges/day)</th>
<th>Date change prescribed</th>
</tr>
</thead>
<tbody>
<tr>
<td>EXAMPLE: 400mg PHE/day</td>
<td>8 g protein/day</td>
<td></td>
<td>Dec.12, 2008</td>
</tr>
</tbody>
</table>

**Instructions for tracking Medical Food (Formula) Prescription:** Anytime the dietitian or doctor changes your prescription medical food or formula, please write it here.

<table>
<thead>
<tr>
<th>Brand name or Type of Medical Food</th>
<th>grams/day</th>
<th>Scoops/day</th>
<th>Date MF change prescribed</th>
</tr>
</thead>
<tbody>
<tr>
<td>EXAMPLE: PhenylFree</td>
<td>100g/day</td>
<td>5 ½ scoops/day</td>
<td>Feb.13, 2009</td>
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</tbody>
</table>
Dear BH4 & PKU Study Participant,

Thank you for choosing to take part in the BH4 (Kuvan™) and PKU clinical trial. We are eager to start this research opportunity with you.

**Study Purpose**

For this clinical research trial, we are asking you to allow us to follow your health status for a 12 month period of time. This is so that we can understand the nutrition and health of people with PKU before and after starting Kuvan™ (BH4) treatment.

**Study and Standard Care Visit Schedule**

Because we want this research opportunity to be a convenient part of your life during the next 12 months, we have kept the study visits and study tests to as few as possible for this study’s research goals. Your visits will follow the timeline listed below:

- **Baseline**: 0 months
- **First study follow-up**: 4 weeks after starting Kuvan™ to determine responsiveness
- **Second study follow-up**: 4 months after starting Kuvan™
- **Third study follow-up** (Middle): 8 months after starting Kuvan™
- **Final study follow-up** (End): 12 months after starting Kuvan™

- **Standard care follow-ups for responders**: 8 weeks and 12 weeks after starting Kuvan™, for switching to regular foods and adjusting Medical Food (formula)

All study participants will need to send us weekly filter papers and 3-day diet records during weeks 1-6 after you start taking Kuvan™.

Responders to Kuvan™ can expect to have at least 7 visits during the one year study period. As a responder, you will also need to provide weekly filter papers and diet records from weeks 8-16. From months 5-12, we will need for you to send us filter papers and 3-day diet records once a month.

Nonresponders to Kuvan™ can expect to have at least 5 visits during the one year study period. After week 8, you will only need to send us filter papers and 3-day diet records once a month until the 12 month end of the study.

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The visits will take place either at the Emory Genetics Clinic, Emory University Hospital CIS (formerly known as the GCRC: General Clinical Research Center), or at a clinic closer to your home. If you are a child in the study and have agreed to a DEXA scan, you may also have an appointment at the Children’s Hospital of Atlanta (CHOA) at Egleston at the beginning and end of the study. If you are an adult in the study and have agreed to a DEXA scan, you will have an additional appointment at Emory University Radiology Clinic at the beginning and end of the study.

Regular Communication
At least every three months either Mary Jane Kennedy (research coordinator) or Teresa Douglas (graduate research assistant) will be checking in with you by phone or email, though we may check in with you more frequently as needed. We will ask you to:

1. Update us on any changes in medications and if you have felt sick at any time
2. Update us on any changes with your medical provider or long-term health condition
3. Update us on any changes in your eating and exercise routine

Issues related to Phe control, formula, and diet will primarily be handled by Dr. Rani Singh and the clinical dietitians at Emory Genetics Clinic.

BH4 & PKU Study Log Book
This Log Book has your study visit instructions and other important documents for you to use during this study. The pages you need to complete are on blue paper.

BH4 & PKU Study Container for PKU participants
We are sending you a BH4 &PKU Study bag to store your study Log Book, study forms, filter papers, and other items. Please keep all your study materials in this bag. We will provide you with refills of study forms and supplies as needed during the study period. If you use up your Log Book or study forms before your next clinic visit, we will send you more.
We look forward to working with you on this exciting project. Please call or email us with any questions or concerns you may have.

Sincerely,

Rani H. Singh, PhD, RD, LD
Principal Investigator
rsingh@emory.edu

and

Teresa Douglas, MS
Research Assistant
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CONTACT INFORMATION

To enroll in this research opportunity or schedule a study visit

Mary Jane Kennedy, RN
Study Coordinator
404-778-8522
mjkenne@emory.edu

To ask other study-related questions, please contact Mary Jane Kennedy FIRST. Otherwise, contact:

Teresa Douglas, MS
Graduate Research Assistant
404-778-8629
tddougl@emory.edu

For more information concerning the research and research-related risks or injuries, you may call:

Rani H. Singh, PhD, RD, LD
Principle Investigator
404-778-8566
rsingh@emory.edu

To call any of the above phone numbers toll-free, dial 1-800-366-1502, press 0, and ask to be connected to one of the extensions listed below:

- Mary Jane Kennedy ext. 88522
- Teresa Douglas ext. 88629
- Dr. Rani H. Singh ext. 88566
Other Contact Information

Fax: 404-778-8562

Address: Emory Genetics Metabolic Nutrition Program
2165 North Decatur Road
Decatur, GA 30033-5307

Study Website: www.genetics.emory.edu/BH4andPKU/singh

If you have any questions regarding your rights as a study participant, you may contact:

Colleen K. Dilorio, PhD
Director
Emory University Institutional Review Board
404-712-9750
sputney@emory.edu
STUDY DETAILS

Study Overview
The purpose of the BH4 & PKU research opportunity (aka Kuvan™ study) is to learn how the compound BH4, prescribed to PKU patients as Kuvan™, affects the diet, health, and biology of individuals with PKU.

Study visits: Schedule and procedures
The BH4 & PKU study has 5 research visits total, but 7 anticipated visits for responders who will require standard care dietary adjustment. The Baseline (0 month) and End-of-study (12 month) visits will be the busiest and most time consuming. You will be asked to spend the night at the Emory University Research Center for these two visits.

NOTE: When you stay overnight at the Emory Research Center, you will have several options for dinner. We will contact you prior to the visit to ask which meal you would prefer.

Schedule of study visits & standard care clinic visits

<table>
<thead>
<tr>
<th>TYPE OF VISIT</th>
<th>WHEN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Research Visit: Screening &amp; Baseline</td>
<td>Upon appointment</td>
</tr>
<tr>
<td>Research Visit: Follow up</td>
<td>4 weeks after starting Kuvan™</td>
</tr>
<tr>
<td>Standard Care Visit (Responders): Follow up</td>
<td>2 months after starting Kuvan™</td>
</tr>
<tr>
<td>Standard Care Visit (Responders): Follow up</td>
<td>3 months after starting Kuvan™</td>
</tr>
<tr>
<td>Research Visit: Follow up</td>
<td>4 months after starting Kuvan™</td>
</tr>
<tr>
<td>Research Visit: Follow up</td>
<td>8 months after starting Kuvan™</td>
</tr>
<tr>
<td>Research Visit: End-of-study Follow up</td>
<td>12 months after starting Kuvan™</td>
</tr>
</tbody>
</table>

*Other standard care visits may be scheduled during this time by the dietitian or doctor so that your nutrition and health can be monitored properly.
How to prepare for the first (Baseline) and final (12 month) research visits

1. The baseline and final study visits will include an overnight stay at Emory University’s Research Center. You will first come to the Emory Genetics Clinic on North Decatur Road before going to the Research Center.
2. You do not have to be fasting when you arrive for baseline and final study visits.
3. Urine samples will be collected during the overnight stays at the Research Center.
4. For both visits, you will complete a 3-day diet record before you arrive. Record any nutrition supplements you take on your diet record!
5. Bring any medications you are currently taking, and supplements you are currently taking, with you to each visit
6. Bring your medical food (formula) and your Kuvan™ to each visit
7. Bring any mixing ingredients for your medical food or Kuvan™ to the overnight visits!!
8. Bring movies to watch for the overnight visits
9. Bring your insurance card with you to each visit
10. Bring your log book to the final visit
11. You will be asked to complete the medical update form IF there have been changes to at least one of the following:
   • Your health care provider, your diet, your medicines, your exercise routine, your personal health

Special instructions for all other research visits

1. You will need to be fasting (NPO) after midnight
2. Bring your log book
3. You will be asked to complete the medical update form IF there have been changes to at least one of the following:
   • Your health care provider, your diet, your medicines, your exercise routine, your personal health
4. Complete an overnight urine sample the night before the study visit
5. Complete a 3-day diet record before you arrive. Record any nutrition supplements you take on your diet record!
6. Bring any new medications you are taking, and new supplements you are taking, with you to each visit
7. Bring your insurance card with you to each visit
For **Standard Care** follow-up appointments at the Emory Genetics Clinic:

1. You will need to be fasting (NPO) after midnight for your plasma amino acid test
2. Complete a 3-day diet record before you arrive. **Record any nutrition supplements you take on your diet record!**
3. Bring any new medications you are taking, and new supplements you are taking, with you to each visit
4. **Bring your insurance card with you to each visit**

**STUDY VISIT LOCATION**
The study visits will usually be conducted at the Emory University Research Center at Emory University Hospital or at the Emory University Genetics Clinic. We will contact you prior to an appointment and discuss with you which of these two places will be the location of your visit.

For the **first and last study visits** we encourage all subjects to schedule an overnight stay at the Emory Research Center.

**STUDY MAILINGS**

**Mail-outs to subjects:** Study bag, extra supplies (log book, medical update form, blood spot filter papers, urine containers, etc.), notices about the study, appointment reminders, lab results

**Return mail:** Diet records, medical update forms, caregiver questionnaires, blood spot filter papers

**REGULAR TELEPHONE AND EMAIL COMMUNICATION**

At least once every 3 months during the study Mary Jane Kennedy or Teresa Douglas will contact you by phone or email to check on how you are doing. During these communications, we will go over the study logbook as well as any other health related updates. We will ask if you have any questions for us. You may receive more frequent contacts from us as needed. Issues regarding Phe control, formula, and your diet will be handled primarily by Dr. Rani Singh and the assigned dietitians at Emory Genetics Clinic.
What to expect at study visits

Visit 1: Screening & Baseline measures
- Consent form
- Height, weight, and head circumference
- Blood pressure and vital signs
- Physical
- Fasting blood samples
- Filter paper blood spots
- Demographic and medical history forms
- Provide overnight urine sample
- Indirect Calorimetry
- DEXA
- Fill out questionnaires:
  - Quality of Life (QOL)
  - Kuvan™ Customer Satisfaction Survey (CSS)

Research related follow-up visits
- Provide medical update form (IF applicable)
- Height and weight
- Head circumference for pediatric patients only
- Blood pressure
- Provide overnight urine sample
- Fasting blood samples
- Fill out QOL and CSS questionnaires

Final End-of-study Follow up
- Provide medical update form (IF applicable)
- Height, weight, and head circumference
- Blood pressure and vital signs
- Physical
- Provide overnight urine sample
- Fasting blood sample
- Indirect Calorimetry
- DEXA
- Fill out QOL and CSS questionnaires

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Study Bag Contents

1. Log Book
   - Contact Information
   - Medication log
   - Illness log
   - Appointment Calendar

2. Diet Records
   - Instructions
   - 3-day diet records
   - Ruler
   - Measuring cups and spoons

3. Forms
   - Medical update forms
   - Self-addressed envelopes

4. Filter Paper Cards
   - Instructions
   - Filter paper cards
   - Lancets
   - Self-addressed envelopes

5. Urine collection
   - Instructions and procedure for overnight urine collection
   - Toilet-hat
   - Silver cooler bag
   - Labeled Plastic container for pouring overnight urine collection

6. Other items
   - Instructions for DEXA and Indirect Calorimetry
   - Instructions for how to prepare for your study visits
   - Directions to the Emory Research Center and to the Emory Genetics Clinic
APPOINTMENT CALENDAR

MONTH______________, YEAR______________

<table>
<thead>
<tr>
<th>Sun</th>
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Visit #_______ Visit #_______
Day_______________ Day_______________
Time_______________ Time_______________
Location_______________ Location_______________

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Time_________________       Time_________________
Location_________________    Location_________________
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# MEDICATION LOG

Anytime you need to take a medication different from your routine medicines please record it in this log

<table>
<thead>
<tr>
<th>Medication Name</th>
<th>Dose</th>
<th>How many doses per day?</th>
<th>Start Date MM/DD/YY</th>
<th>Stop Date MM/DD/YY</th>
<th>Reason for taking medication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Example: regular strength ibuprofen</td>
<td>2 tablets</td>
<td>1</td>
<td>Monday, 06/11/08</td>
<td>Monday, 06/11/08</td>
<td>Headache</td>
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Example: regular strength ibuprofen 2 tablets 1 Monday, 06/11/08 Monday, 06/11/08 Headache
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Example: regular strength ibuprofen 2 tablets 1 Monday, 06/11/08 Monday, 06/11/08 Headache
MEDICATION LOG

Anytime you need to take a medication different from your routine medicines
please record it in this log

<table>
<thead>
<tr>
<th>Medication Name</th>
<th>Dose</th>
<th>How many doses per day?</th>
<th>Start Date MM/DD/YY</th>
<th>Stop Date MM/DD/YY</th>
<th>Reason for taking medication</th>
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</thead>
<tbody>
<tr>
<td>Example: regular strength ibuprofin</td>
<td>2 tablets</td>
<td>1</td>
<td>Monday, 06/11/08</td>
<td>Monday, 06/11/08</td>
<td>Headache</td>
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</tbody>
</table>
ILLNESS LOG

If you feel sick or come down ill, even if it is brief or minor, please record it in this log

<table>
<thead>
<tr>
<th>Illness</th>
<th>Start Date MM/DD/YY</th>
<th>Stop Date MM/DD/YY</th>
<th>Medication taken?</th>
<th>Life-threatening?</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Example: Headache</td>
<td>Monday, 06/11/08</td>
<td>Monday, 06/11/08</td>
<td>Yes</td>
<td>Yes</td>
<td>Lots of construction noise nearby</td>
</tr>
</tbody>
</table>
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<td>Example:</td>
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<td>Monday, 06/11/08</td>
<td>Yes</td>
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<th>Comments</th>
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<td>Example: Headache</td>
<td>Monday, 06/11/08</td>
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<td>☑ Yes</td>
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Emory BH4 & PKU Research Study
SINGH RESEARCH GROUP
404-778-8522
www.genetics.emory.edu/NUTRITION/BH4andPKU/
# KUVAN™/BH4 Calendar

**Directions**
- If you miss a daily dose, write a zero (0) on the day you missed
- If you take only part of your daily dose, write the number of capsules you actually took on that day
- If you take your entire daily dose, leave the day blank

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MONTH________________, YEAR________________

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Emory BH4 & PKU Research Study
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FILTER PAPER BLOOD TEST PROCEDURE

**Important:** Blood tests are analyzed every Thursday and results are reported on Fridays. Please place the filter paper into the mailbox by Sunday so that it arrives by Wednesday. All tests that arrive after Wednesday afternoon will not be reported until the following Friday.

*Blood spots should be taken at least 1.5-2 hours after a meal*

1. Fill out filter paper card with patient’s name, date of birth, collection date.
2. Fill out parts of pages 1 and 3 of the Emory Genetics Lab Test Requisition Form where highlighted.
3. Do not touch the filter paper before or after applying blood spot.
4. Wash hands with soap and warm water and dry thoroughly.
5. Clean fingertip to be pricked with a 70% alcohol pad and air dry. 
   *Tip: you can rub the fingertip to increase blood flow before pricking.*
6. Prick side of fingertip near fingernail with a sterile lancet. Lancets can be found in any drug store. Discard the lancet after use.
7. Allow drop of blood to form. Wipe first drop of blood away with gauze or cotton ball. Allow second drop to form.
8. Apply large drop of blood to filter paper. One drop will fill circle from front to back. Do not layer additional drops of blood, and apply blood to only one side of the filter paper.
9. Fill two circles on the card.
10. Re-stick finger if blood does not flow freely. Follow steps 5 to 8.
11. Allow to dry for 4 hours on a flat, clean, nonabsorbent surface that is away from heat and sunlight. Please send the filter paper and test requisition form completed in the provided envelope to: *Emory Genetics Metabolic Nutrition Program, Attn: Teresa Douglas, 2165 N. Decatur Road, Decatur GA 30033.*
Three-Day Food Record Instructions

- We will ask you to keep a 3-day food record:
  - Three days before each study visit, and
  - Three days before taking a blood sample for each filter paper card

- **Please record everything you actually eat and drink for three days in a row**

- Write down any nutritional supplements you take, such as calcium supplements or a multivitamin.

- Record each day on a separate record sheet

- Weigh and/or measure your food before you eat it
  - Subtract any amount that you did not eat and record your actual intake on the food record

- To estimate portion sizes, use the instructions in this packet, measuring spoons and cups, a ruler, and a scale

- Record what you eat and drink as soon as you can to reduce the chance of forgetting

- Write only one food or drink item per line

- Estimate the amount of phenylalanine (Phe) in milligrams or exchanges for the serving of food or drink that you actually ate (15mg of Phe equals 1 Phe exchange)

- **Double-check your food record.** Did you remember to write down:
  - All meals, snacks, nibbling, and beverages including cocktails?
  - Recipes?
  - Ingredients used in mixed dishes, sandwiches, etc?
  - Seasonings, spices, dressings, or condiments added to foods?
  - Whether weights are for cooked or raw portions?
  - How the food was prepared (uncooked vs. cooked)
    - Specify how the food was cooked: boiled, baked, broiled, fried, grilled, steamed, toasted, poached
  - If the food is a specialty low protein item?
  - Whether there was any ice in any beverages?

If you have any questions about your food record, please call or email Mary Jane Kennedy (404-778-8522, mkenne@emory.edu) and she will refer you to dietitian at the Emory Genetics Clinic. If you cannot reach Mary Jane Kennedy, you may contact the research assistant Teresa Douglas (404-778-8629, tddougl@emory.edu).
A Handy Way to **Estimate** Portion Size

<table>
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<tr>
<th>Part of Hand</th>
<th>Approximate Portion Size</th>
<th>Examples of Foods</th>
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<tr>
<td>Whole Fist</td>
<td>1 cup</td>
<td>2 servings vegetables</td>
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<td>1 serving rice or pasta</td>
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<td>1 piece of fruit</td>
</tr>
<tr>
<td>Palm of Hand</td>
<td>1 oz</td>
<td>1 serving of snack chips or pretzels</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1 serving of meat product</td>
</tr>
<tr>
<td>Finger Length</td>
<td>2.5 inches</td>
<td>Diameter of 1 fruit serving or tennis ball</td>
</tr>
<tr>
<td>Whole Thumb</td>
<td>2 Tbsp, 20 ml, 1 fluid oz, 1 solid oz, 28g</td>
<td>1 serving of peanut butter or cheese</td>
</tr>
<tr>
<td>Thumb Tip</td>
<td>1 tsp, 5 ml</td>
<td>1 serving of margarine</td>
</tr>
</tbody>
</table>
### Ways to Describe Portion Size

<table>
<thead>
<tr>
<th>Illustration</th>
<th>Description</th>
<th>Examples</th>
</tr>
</thead>
</table>
| ![Measuring cups](Image) | Measuring cups (C)  
Teaspoons (t or tsp)  
Tablespoons (T or Tbsp) | Vegetables, canned or frozen fruits, pasta, rice, casseroles, all liquids such as water, beverages, soups, sauces, salad dressings, sorbet, or smoothies |
| ![Fluid Ounces](Image) | Fluid Ounces (fl oz) | All liquids such as water, beverages, soups, sauces, salad dressings, sorbet, or smoothies |
| ![Weight in grams](Image) | Weight in grams (g) or solid ounces (oz) | Any solid food such as cheese, frozen entrees, or dry medical food (formula) |
| ![Fraction of the whole](Image) | Fraction of the whole | 1/8 of a 9 inch pie or 1/4 of a 6 inch cantaloupe |
| ![Diameter](Image) | Diameter (D) | Any sphere such as a 3 inch diameter apple, roll, or tomato |
| ![Diameter and Thickness](Image) | Diameter and Thickness | Any cylinder or disk. Such as a pancake, cracker, cookie, or low protein burger patty |
| ![Length and Height and Width](Image) | Length and Height and Width | Any rectangle or square, such as a piece of cake or bread |
| ![Length, Height, and Width of an Arc](Image) | Length, Height, and Width of an Arc | Any wedge, such as a slice of pie or low protein pizza |
Completing your 3-Day Diet Record

★Example★

1. Start Date: ________________________  End Date: ________________________
   Name: ____________________________
   Date of Birth: ____________________
   Height: __________________________
   Weight: __________________________

2. Reporting Kuvan™ intake on your 3-Day Diet Record

   How do you take your Kuvan™?
   - Mixed with water
   - Mixed with apple Juice
   - I swallow pills whole
   - Other: ____________________________________________
   - NA: I do not take Kuvan™

   KUVAN™ Prescription: 20 mg/kg, 6 pills/day

3. Reporting Medicines and supplements on your 3-Day Diet Record

<table>
<thead>
<tr>
<th>Other Medications or supplements (vitamin, mineral, herbal, etc.)</th>
<th>Dose (μg or mg)</th>
<th>When and How Often</th>
</tr>
</thead>
<tbody>
<tr>
<td>Advil</td>
<td>200mg</td>
<td>1X this morning</td>
</tr>
<tr>
<td>Publix brand Calcium with vitamin D</td>
<td>500mg</td>
<td>2X daily</td>
</tr>
</tbody>
</table>

4. Reporting Formula on your 3-Day Diet Record

<table>
<thead>
<tr>
<th>Formula Prescription: (Please include the name and amount of each ingredient used to make your Formula.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ingredient</td>
</tr>
<tr>
<td>Phenex-2 in water with Hershey strawberry syrup</td>
</tr>
</tbody>
</table>

   Volume Mixed: ______ oz.  Volume Consumed: ______ oz.  # of servings: ____
5. Reporting Food (including recipes and restaurant meals) on your 3-Day Diet Record ★Example★

**DAY 1**

* Meal: **B**=Breakfast; **L**=Lunch; **D**=Dinner; **S**=Snack

** Exchanges: 1 exchange = 15 mg Phe

<table>
<thead>
<tr>
<th>* Meal</th>
<th>Food/Beverage Item</th>
<th>Amount of Food Eaten</th>
<th>Phe (mg)</th>
<th>** Exchanges</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>B</td>
<td>Coffee at I-Hop</td>
<td>1 cups</td>
<td>32</td>
<td>2</td>
<td>At I-Hop restaurant</td>
</tr>
<tr>
<td>B</td>
<td>sugar</td>
<td>2 packets</td>
<td>0</td>
<td>0</td>
<td>At I-Hop restaurant</td>
</tr>
<tr>
<td>B</td>
<td>Green apple, Granny Smith</td>
<td>1 medium</td>
<td>7</td>
<td>0.5</td>
<td>Ate at home</td>
</tr>
<tr>
<td></td>
<td>Phenex-2 with strawberry syrup</td>
<td>19oz</td>
<td>0</td>
<td>0</td>
<td>Drank all day</td>
</tr>
<tr>
<td>L</td>
<td>Lays baked potato chips</td>
<td>1 oz</td>
<td>81</td>
<td>5.5</td>
<td></td>
</tr>
<tr>
<td>L</td>
<td>Low protein spaghetti (Loprofin)</td>
<td>1 cup cooked</td>
<td>15</td>
<td>1</td>
<td>Boiled</td>
</tr>
<tr>
<td>L</td>
<td>Promise margarine</td>
<td>1Tbsp</td>
<td>6</td>
<td>0.5</td>
<td>Cooked with Spaghetti in water</td>
</tr>
<tr>
<td>L</td>
<td>Prego regular spaghetti sauce</td>
<td>½ cup</td>
<td>48</td>
<td>4</td>
<td>Ate over spaghetti</td>
</tr>
<tr>
<td>L</td>
<td>1 bottle Coca-Cola</td>
<td>20 fluid oz.</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>S</td>
<td>Fresh Strawberries</td>
<td>½ cup</td>
<td>13</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>S</td>
<td>Unsweetened Tea</td>
<td>20 fluid oz</td>
<td>0</td>
<td>1</td>
<td>Brewed</td>
</tr>
<tr>
<td>D</td>
<td>Low-Pro LBP Burger Bun</td>
<td>1 roll</td>
<td>9</td>
<td>1</td>
<td>Camden</td>
</tr>
<tr>
<td>D</td>
<td>Romaine lettuce</td>
<td>1 outer leaf</td>
<td>19</td>
<td>1.5</td>
<td></td>
</tr>
<tr>
<td>D</td>
<td>Red Tomato</td>
<td>1 slice</td>
<td>5</td>
<td>0.5</td>
<td>Fresh uncooked roma</td>
</tr>
<tr>
<td>D</td>
<td>Ice Water</td>
<td>20 oz</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
</tbody>
</table>

Day 1 Total: __235__mg __18.5__ Ex.
INSTRUCTIONS: OVERNIGHT URINE SPECIMEN

You will need:

- Collection container with label
- A pen, pencil or marker
- Clean toilet-hat
- Silver cooler-bag with ice or cool packs
- Watch or clock.

It is important that you drink what is the normal amount of fluids for you prior to the urine sample. This means, do not drink more or less fluids than you usually would during a normal day.

You will have a plastic container designated for each clinic visit. Use the labeled container that matches the next day’s visit. For example, if you are coming in for your third study visit, use the container that is labeled “Visit # 3”.

All collection containers will have a label on them like the one pictured below:

Subject ID___XYZ000____

STUDY VISIT # 1
Date of study visit__June 4,2008__
Date of collection__June 3,2008__
Collection Start Time___8:00____PM
Collection End Time___8:00____AM

Subject ID, and Study Visit # will already be printed on the label. On the night you begin your urine collection, please write in the Date of Collection. You may also need to fill in the date of your study visit. The date of your study visit should be one day after the Date of Collection.
Please read the instructions listed below. Follow these instructions during the overnight urine collection. Please remember to write your Start Time and End Time on the label according to the instructions.

1. You will collect urine anytime you use the restroom starting from 8pm the night before your appointment until 8am of the morning you will be arriving at the clinic. This should equal a 12 hour time period.

2. 8pm should be the start-time you write onto the label. 8am should be the end-time you write onto the label.
   *NOTE: If circumstances require you to change your start-time and end-time, please record that on the label and inform us when you provide us with your specimen.*

3. Place the red urine collection container into the silver-cold pack along with a couple cold packs or a layer of ice.

4. Place the clean toilet-hat between the toilets bowl top and lid to hold it in place. Make sure the curved part of the hat is lined up with the front edge of the toilet bowl.

5. Anytime you urinate during the 12 hour time period, urinate in the toilet-hat. *If it is convenient,* you can instead urinate directly into the red container.

6. The toilet-hat has a pour spout which makes it easy to pour the urine into the red collection container. Remove the white cap off the red container and pour the urine in without splashing or spilling.
   *NOTE: It is best to do this immediately after urinating. If you get up during the night but do not pour the urine into the red container immediately, then do so as soon as possible the next morning.*

7. Stop collecting urine into the container once the 12 hour collection period has ended.

8. Write the End Time on the label of the red collection container.

9. Cap the red container and close it tightly.

10. Place the red urine container into the silver cold-bag with fresh ice or cold packs.

11. Your overnight urine sample is now complete and ready to bring to us at Emory.
DEXA

- DEXA (also known as DXA) is a short form of the word Dual Energy X-ray Absorptiometry.

- DEXA scans are used for 2 main purposes
  1. To determine bone density, which means how strong and healthy your bones are
  2. To determine the amounts of fat tissue and lean tissue (muscle) in your body

- The amount of radiation that one DEXA scan exposes you to is equal to 1/10th of the radiation you would get from a chest X-ray

- DEXA is an open air scan, painless and very quick. A DEXA scan only takes 15-20 minutes.

- During a DEXA scan, you need to lie very still on the table until the procedure is finished.

- Parents are allowed to be in the room with their children. A PDF information form is available for parents from the Children’s Healthcare of Atlanta (CHOA) website: http://www.choa.org/Menus/Documents/OurServices/radiology_dexa.pdf

- We will give you the results of your DEXA scan as soon as they are available.

- For this study, we are asking research volunteers to have 2 DEXA scans. One scan will be at baseline, before you start taking Kuvan™. The second scan will be 1 year later during the very last research visit.

- Children will most likely have their DEXA scan at CHOA-Egleston on Clifton Road. Adults will have their DEXA scan at Emory University Hospital Radiology clinic (Building C) on Clifton Road.

- If you think you might be pregnant at any time during the research study, please inform Mary Jane Kennedy or Dr. Rani Singh. We would need to make sure you do not have a DEXA scan while you are pregnant. Also, if you are at the scanning place and think you might be pregnant, please tell the radiology technician.
Indirect Calorimetry

- Indirect Calorimetry measures the amount of calories (energy) your body uses while you are resting.

- Emory University has an Indirect Calorimetry machine (pictured on this page) located at the Emory CIS (formerly known as the GCRC—General Clinical Research Center) located at Emory University Hospital. All Indirect Calorimetry will take place here.

- Indirect Calorimetry involves having a plastic shaped dome placed over your face for 20 minutes. This is so the oxygen you breathe in and the carbon dioxide you breathe out can be measured.

- The procedure is open air, painless, and quick.

- Parents are allowed to be in the room with their children.

- For this study, we are asking research volunteers to have 2 indirect calorimetry tests done. One test will be at baseline, before you start taking Kuvan™. The second test will be 1 year later during the very last research visit.

- We will give you the results as soon as they are available
Directions to the Emory Research Center

Address
Emory Clinical Interaction Site (CIS), formerly the General Clinical Research Center (GCRC)
Emory University Hospital, Suite GG-23
1364 Clifton Road, NE
Atlanta, GA 30322

Parking
Parking for the Emory University Hospital is available in front of the Emergency Department and across Clifton road in the visitor parking tower. We will provide you with a voucher to pay for your parking.

Directions
From the Northeast (I-85):
Take I-85 South to Clairmont Road (Exit #91). Turn left on Clairmont Road and follow the signs towards Decatur. Continue on Clairmont Road for 3.8 miles to North Decatur Road. Turn right on North Decatur Road. Proceed one mile to Clifton Road. Turn Right on Clifton Road. The hospital and ambulance entrance are located 0.2 miles on the left. Clinic Buildings A and B, as well as Visitor Parking, are located 0.2 miles on the right.

From the North (GA 400):
Take GA 400 South to Sidney Marcus Boulevard exit. Turn left on Sidney Marcus Boulevard. Turn left on Buford Highway. Turn right on Lenox Road. Get on I-85 North. Take I-85 North to Clairmont Road (Exit #91). Turn right on Clairmont Road and follow the signs towards Decatur. Continue on Clairmont Road for 3.8 miles to North Decatur Road. Turn right on North Decatur Road. Proceed one mile to Clifton Road. Turn right on Clifton Road. The hospital and ambulance entrance are located 0.2 miles on the left. Clinic Buildings A and B, as well as Visitor Parking, are located 0.2 miles on the right.

-or-

Take GA 400 South to I-285. Take I-285 East for approximately 5.6 miles. Take I-85 South to Clairmont Road (Exit #91). Turn left on Clairmont Road and follow the signs towards Decatur. Continue on Clairmont Road for 3.8 miles to North Decatur Road. Turn right on North Decatur Road. Proceed one mile to Clifton Road. Turn right on Clifton Road. The hospital and ambulance entrance are located 0.2 miles on the left. Clinic Buildings A and B, as well as Visitor Parking, are located 0.2 miles on the right.
**Directions to the Emory Research Center, Continued**

**From the South (I-75):**
Take I-75 North to Freedom Parkway/Carter Center/International Boulevard (Exit #248-C). Bear right off the exit and continue straight for approximately 2.1 miles following the signs to the Carter Center and Moreland Avenue. Turn left on Moreland Avenue. Continue north on Moreland Avenue (Moreland Avenue becomes Briarcliff Road) for approximately 1.7 miles. Turn right on North Decatur Road and travel approximately 1.1 miles, going through Emory Village. Turn left on Clifton Road. The hospital and ambulance entrance are located 0.2 miles on the left. Clinic Buildings A and B, as well as Visitor Parking, are located 0.2 miles on the right.

**From the East (US 78):**
Take 285 to Stone Mountain Freeway/US-78, follow signs towards Decatur. Proceed on Scott Blvd towards North Decatur Road. Turn Right on North Decatur Road. Proceed approximately two miles on North Decatur Road. Turn right on Clifton Road. The hospital and ambulance entrance are located 0.2 miles on the left. Clinic Buildings A and B, as well as Visitor Parking, are located 0.2 miles on the right.

**From the East (I-20):**
Take I-20 West to Moreland Avenue (Exit 60). Turn right (north) on Moreland Avenue. Continue on Moreland Avenue (Moreland Avenue becomes Briarcliff Road) for approximately 3.4 miles. Turn right on North Decatur Road and travel approximately 1.1 miles, going through Emory Village. Turn left on Clifton Road. The hospital and ambulance entrance are located 0.2 miles on the left. Clinic Buildings A and B, as well as Visitor Parking, are located 0.2 miles on the right.

**From the Northwest (I-75):**
Take I-75S to I-85N. Take I-85 to Clairmont Road (Exit #91). Turn right and follow the signs towards Decatur. Stay on Clairmont Road for 3.8 miles to North Decatur Road. Turn right on North Decatur Road. Proceed one mile to Clifton Road. Turn right on Clifton Road. The hospital and ambulance entrance are located 0.2 miles on the left. Clinic Buildings A and B, as well as Visitor Parking, are located 0.2 miles on the right.

**From the West (I-20):**
Take I-20 East to Moreland Avenue (Exit 60). Turn right (north) on Moreland Avenue. Continue on Moreland Avenue (Moreland Avenue becomes Briarcliff Road) for approximately 3.4 miles. Turn right on North Decatur Road and travel approximately 1.1 miles, going through Emory Village. Turn left on Clifton Road. The hospital and ambulance entrance are located 0.2 miles on the left. Clinic Buildings A and B, as well as Visitor Parking, are located 0.2 miles on the right.

Emory BH4 & PKU Research Study
SINGH RESEARCH GROUP
404-778-8522
www.genetics.emory.edu/NUTRITION/BH4andPKU/
MAPS OF CLINIC SITES

Location of Emory University Hospital
1364 Clifton Road
Atlanta, GA 30322

Location of Emory Genetics Clinic
2165 N. Decatur Rd.
Decatur, GA 30033
Directions to Emory Genetics

Address
Emory Genetics Clinic Building
2165 North Decatur Rd.
Decatur, GA 30033

Parking
Free parking is available in front of the building.

Directions

From Interstate 20 East
Take exit 60-B, the Moreland Avenue North exit. Turn right on Moreland and follow it approximately 3 miles. Moreland will change to Briarcliff Road once it crosses Ponce de Leon Avenue. Continue straight on Briarcliff approximately 2 miles to North Decatur Road. Turn right and follow North Decatur one mile to the Oxford Road intersection. Stay on North Decatur for 2 miles until you reach the intersection of North Decatur and Clairmont Rd. See below ***

From Interstate 20 West
Take exit 60-B, the Moreland Avenue North exit. Turn right on Moreland and follow it approximately 3 miles. Moreland will change to Briarcliff Road once it crosses Ponce de Leon Avenue. Continue straight on Briarcliff approximately 2 miles to North Decatur Road. Turn right and follow North Decatur one mile to the Oxford Road intersection. Stay on North Decatur for 2 miles until you reach the intersection of North Decatur and Clairmont Rd. ***

From Interstate 75 North or South
Take exit 248-C, the Freedom Parkway exit. Cross Boulevard; continue on Freedom Parkway; veer left at split; continue until it ends at Ponce de Leon Avenue; then turn right. Off Ponce, turn left on Briarcliff Road. Go approximately 2 miles to North Decatur Road. Turn right and follow North Decatur Road one mile to the Oxford Road intersection. Stay on North Decatur for 2 miles until you reach the intersection of North Decatur and Clairmont Rd. ***

From Interstate 85 North and Hartsfield Atlanta International Airport
The airport is located in the southwest section of the city, approximately 25 minutes from the Emory University campus. Visitors driving from the airport should take I-85 North. To Emory from Interstate 85 North, take exit 248-C, the Freedom Parkway exit. Cross Boulevard; continue on Freedom Parkway; veer left at split; continue until it ends at Ponce de Leon Avenue; then turn right. Off Ponce, turn left on Briarcliff Road. Go approximately 2 miles to North Decatur
Road. Turn right and follow North Decatur Road one mile to the Oxford Road intersection. Stay on North Decatur for 2 miles until you reach the intersection of North Decatur and Clairmont Rd. ***

***Continue straight on North Decatur through the intersection. We are located just past the Publix on the right.

**From Interstate 85 South**
Take exit 91, the Clairmont Road exit. Turn left (east) on Clairmont and follow it approximately 3 miles to North Decatur Road. Turn left on North Decatur. We are just past the Publix on the right.
NOTES
Local Attractions

- **At Emory**
  Michael C. Carlos Museum
  571 South Kilgo Circle
  Atlanta, GA 30322
  Phone: 404-727-4282
  [www.carlos.emory.edu/](http://www.carlos.emory.edu/)

  Emory Visual Arts Gallery
  700 Peavine Creek Drive
  Atlanta, GA 30322
  Phone: 404-727-6315
  [www.visualarts.emory.edu/](http://www.visualarts.emory.edu/)

  Lullwater Conservation Park
  1463 Clifton Road NE
  Atlanta GA 30329

- **In Atlanta**
  Fernbank Museum of Natural History
  767 Clifton Road NE
  Atlanta GA 30307
  Phone: 404-929-6300
  [www.fernbankmuseum.org](http://www.fernbankmuseum.org)

  Fernbank Science Center and Planetarium
  156 Heaton Park Drive
  Atlanta GA 30307
  Phone: 678-874-7102
  [http://fsc.fernbank.edu/](http://fsc.fernbank.edu/)

  Atlanta Zoo
  800 Cherokee Ave, SE
  Atlanta GA 30315
  Phone: 404-624-WILD
  [www.zooatlanta.org](http://www.zooatlanta.org)

  World of Coca-Cola
  121 Baker Street NW
  Atlanta, GA 30313
  Phone: 1-800-676-COKE
  [www.woccatlanta.com](http://www.woccatlanta.com)

  Atlanta Botanical Garden
  1345 Piedmont Ave., NE
  Atlanta GA 30309
  Phone: 404-876-5859
  [www.atlantabotanicalgarden.org](http://www.atlantabotanicalgarden.org)

  High Museum of Art
  1280 Peachtree Street, NE
  Atlanta, GA 30309
  Phone: 404-733-HIGH
  [www.high.org](http://www.high.org)

  Center for Puppetry Arts
  1404 Spring Street NW at 18th
  Atlanta GA 30309
  Phone: 404-873-3391
  [www.puppet.org](http://www.puppet.org)

  Georgia Aquarium
  225 Baker Street NW
  Atlanta GA 30313
  Phone: 404-581-4000
  [www.georgiaaquarium.org](http://www.georgiaaquarium.org)

  Atlantic Station
  I-75 and 17th street
  Atlanta GA 30363
  Phone: 404-876-2616
  [www.atlanticstation.com](http://www.atlanticstation.com)

  Lenox Square mall & shopping center
  3393 PeachTree Road
  Atlanta GA 30326
  Phone: 404-233-6767
  [www.lenoxsquare.com](http://www.lenoxsquare.com)
Near Atlanta

Six Flags over Georgia
275 Riverside Parkway
Austell, GA 30168
Phone: 770-739-7400
www.sixflags.com/overGeorgia/

Six Flags White Water
250 Cobb Parkway North
Marietta, GA 30062
Phone: 770-590-4067
www.sixflags.com/whiteWater/

Stone Mountain Park
U.S. Highway 78 East, Exit 8
Stone Mountain, GA 30087
Phone: 404-498-5690
www.stonemountainpark.com

Mall of Georgia
3333 Buford Drive
Buford GA 30519
Phone: 678-482-8788
www.mallofgeorgia.com

Kangaroo Conservation Center
222 Baily Waters Road
Dawsonville GA 30534
Phone: 706-265-6100
www.kangaroocenter.com

PHE Friendly Places to Eat

The Real Chow Baby
(Build your own stir fry)
1016 Howell Mill Road, Suite A
Atlanta, GA 30318
www.therealchowbaby.com

Hot Stix (Build your own stir fry)
Lindbergh City Center
570 Main Street
Atlanta, GA 30324
www.hotstixstirfry.com

Doc Green’s (Build your own salad)
782 Ponce De Leon Avenue
Atlanta, GA 30306
www.docgreens.com

Soup Garden (Salad & Soup Bar)
2945 Cobb Parkway
Atlanta GA, 30339
Phone: (770) 955-5726

Café Sunflower (vegan/vegetarian cuisine)
2140 Peachtree Road
Atlanta, GA 30309
www.cafesunflower.com

Lettuce Souprise You (Salad & Soup Bar)
2470 Briarcliff Road, NE
Atlanta, GA 30329
www.lettucesoupriseyourestaurant.com
STUDY FAQ
(Frequently Asked Questions)

1. I have PKU and would like to enroll in this study. How do I know if I am eligible?
   A. Your are eligible if the following applies:
      • You are at least of 5 years of age
      • You have been diagnosed with Phenylketonuria
      • You intend to try Kuvan™ therapy
      • You have not taken BH4 or Kuvan™ for at least 8 weeks.
      • You are not pregnant
      • You can provide informed consent

2. How long will you be recruiting study volunteers?
   A. For 6 months from the official date in October when recruitment begins

3. How long will this study be?
   A. PKU participants will be followed for 12 months from the point in time that they begin Kuvan™

4. How many clinic visits are required during this study?
   A. Five research study visits. More standard care clinic visits may be included if recommended by an Emory Genetics Physician or Dietician. For more details, see page 11 in this log book.

5. What will happen at each research clinic visit?
   A. Each research clinic visit will involve a blood draw and you will be asked to provide an overnight urine sample, a three day diet record, and be asked to fill out forms and questionnaires. There may be other procedures depending on which visit it is. For more details, see page 14 in this log book.
6. Where will the study visits take place?
   A. The first and final visits will be at the Emory Research Center at Emory University Hospital. We encourage overnight admission for these two visits. The other follow-up visits will most likely occur at the Emory Genetics Clinic on North Decatur Rd, Decatur GA.
   Your DEXA scan will most likely be at Emory Hospital, but children may have their DEXA scans at Children’s Healthcare of Atlanta at Egleston (across the street from Emory Hospital). See pages 11-13 of the log book.

7. What is Indirect Calorimetry?
   A. Indirect Calorimetry measures how much energy your body uses while at rest by measuring the amount of oxygen and carbon dioxide in your breath. A machine will be used to measure your energy expenditure. The procedure is brief, painless, and non-invasive.

8. What is DEXA?
   A. DEXA is a type of scan that uses weak X-rays to measure bone density as well as the amount of fat and muscle in the body. It is open air (no tube), and the procedure is brief, painless, and non-invasive.

9. What specific tests are you running on my blood and urine?
   A. In urine samples we are looking at the neurotransmitter levels of serotonin and catecholamines such as dopamine, epinephrine, and norepinephrine. In the blood samples, we are looking at levels of neurotransmitters, vitamins and minerals, protein status, blood PHE and Tyrosine, blood lipids, and liver enzymes.

10. Will I be able to look at my personal blood and urine lab results?
    A. We will be able to provide you with all of your blood lab results as soon as they are available. The only exception is blood and urine concentrations of serotonin and catecholamines. Because serotonin and catecholamine levels are being analyzed in a research lab, rather than a clinical laboratory, we may be unable to provide you the individual results.

11. Can I have a friend, relative, or parent with me during any of the tests or procedures if I am uncomfortable?
    A. Yes. If you are uncomfortable with a procedure, someone may accompany you, preferably a parent or other close relative.
12. I will be hungry after my blood tests are done. Will there be snacks available?
   A. We will have snacks available for you once the fasting procedures are complete. If
      you want to be sure you get to snack on something you like, then it is best to bring
      your own snacks with you. You can eat them after the blood draws and indirect
      calorimetry are over.

13. What if I forget to fast before a study visit?
   A. We can still collect information on your height, weight, blood pressure, and quality
      of life as well as do your DEXA scan. We can also still collect your overnight urine
      sample. However, we would need to set up a second appointment very soon to
      collect a fasting blood sample, or to do indirect calorimetry.

14. Who will be managing my diet during the study period?
   A. The clinical dietitians at the Department of Human Genetics will work with Dr. Singh
      to manage your diet appropriately during the study.

15. How do I fill out a 3-day diet record?
   A. Instructions for filling out a 3-day diet record begin on page 35 of this log book. We
      have also provided instructions on the research study website. You can also contact
      Mary Jane Kennedy if you have any questions about how to fill out your diet record
      prior to a visit. She can get you in touch with a clinic dietitian if necessary.

16. If my blood PHE drops less than 30%, but more than 15% while trying Kuvan™, can I stay
    on the drug?
   A. Yes. Having a PHE response of greater than 15%, but less than 30% means you are a
      mild responder as defined by our research protocol. For as long as you are
      participating in the research study, as a mild responder you will have access to
      Kuvan™.

17. After enrolling, I discovered that I am a responder to Kuvan™, but I still want to stop
taking it. Can I still be in the study even if I voluntarily stop Kuvan™?
   A. Yes. Even if you voluntarily decide to stop Kuvan™ you can still participate in the
      study for the full 12 month time period.
18. At my home clinic, I tried Kuvan™, and though my blood PHE dropped by more than 15%, because it was less than 30% I was considered a non-responder and had to stop Kuvan™. Can I still participate in your study?
   A. Yes. If you have been off of Kuvan™ for at least 8 weeks and pass the screening criteria, you can enroll in the study. You will go through another 4 week period where your response will be tested. If your plasma PHE levels drop by at least 15% during that study period, you will be allowed to stay on Kuvan™ for the entire time you are a part of the study.

19. If I am a non-responder to Kuvan™, can I still stay in the study?
   A. Yes. For a full 12 months, we are following all the PKU subjects who try Kuvan™, even those who are non-responders and stop taking Kuvan™. However, there will be less clinic visits for nonresponders.

20. Where do I mail my blood spot filter papers or study forms?
   A. Please mail these to the Emory Genetics Clinic on North Decatur Road, C/O Teresa Douglas. See page 30 for more details.

21. Who do I contact if I find out I am pregnant during the study?
   A. Please contact either Mary Jane Kennedy or Dr. Rani Singh as soon as you find out you are pregnant. Please let us know the approximate time that you became pregnant.

22. Can I continue to be in the study if I find out I am pregnant?
   A. No. If you discover you are pregnant during the study it is important that you let Dr. Singh or Mary Jane Kennedy know. You may be asked to withdraw from the study if it is confirmed that you are pregnant. Whether you continue Kuvan™ treatment during your pregnancy will be determined by your clinic physician after you have spoken with him/her about your pregnancy.

23. I am participating in the DHA trial conducted by Emory Genetics Clinic. Can I still participate in the BH4 study?
   A. No. You will need to have completed the DHA trial before you can enroll in the BH4 study.
24. **Will participating in the study cost me anything?**  
   A. Volunteers will **not** be charged for any testing or clinic visits that are part of the research study. Those with health insurance will have their insurances billed for any testing or treatment considered to be standard care for any PKU patient trying Kuvan™. PKU subjects will **not** be expected to provide a copay or any financial reimbursement for standard care that is part of the research study. Subjects will still be responsible for any copays attached to their Kuvan™ or formula prescriptions.

25. **What standard care would I be getting if I were not participating in the research study, or if I decide to withdraw from the study?**  
   A. As a PKU patient, you would have your blood PHE levels monitored during your 4 week Kuvan™ trial period to determine if you are responding to the medicine. If your blood PHE levels decrease by at least **30%**, your protein or dietary PHE tolerance could be tested by challenging with powdered milk. Your formula and dietary protein (PHE allowance) might then be gradually adjusted to accommodate your changes in PHE tolerance while monitoring your blood PHE levels and overall diet. This is the standard care for **any** PKU patient at the Emory Genetics Clinic beginning Kuvan™.

26. **Will reimbursement for travel expenses be provided?**  
   A. After your 12 month appointment, we will reimburse you with $90 compensation for time and travel.

27. **How do I withdraw from the study if I no longer want to participate?**  
   A. At the screening visit, you will be provided with a form that you can mail to us if you decide to withdraw from the research study. This form will let us know that you no longer wish to participate in the research study. If you cannot find this form and need one, you can contact Mary Jane Kennedy or Teresa Douglas and we will provide one to you.

28. **If I withdraw from the study before my 12 month appointment, will I still be eligible to receive the $90 compensation?**  
   A. If you withdraw due to pregnancy or adverse events related to Kuvan™, you will be compensated the full $90. All other reasons for withdrawal will not guarantee compensation.
29. **What if I am confused about how to fill out one of the study forms or questionnaires?**
   A. Please speak to Mary Jane Kennedy or Teresa Douglas at your clinic visit, or by phone or email. They will be happy to help you out with any forms or questionnaires.

30. **How do I report a change in my phone number, email address, or mailing address?**
   A. Please report changes in phone number, email, or mailing address to either Mary Jane Kennedy, Research Coordinator or to the research assistant, Teresa Douglas. Contact information is located on page 9 of this book.

31. **Who do I contact with questions or concerns I may have about the research study?**
   A. For questions about your diet or diet record contact Mary Jane Kennedy and she will connect you to an assigned dietitian.
   For all other questions, contact either Mary Jane Kennedy or Teresa Douglas. If we cannot answer your question, we will refer your question to Dr. Rani Singh and get back to you. Contact information is located on page 9 of this book.

32. **Is there a website set up for this research study?**
   A. The study website is [www.genetics.emory.edu/NUTRITION/BH4andPKU/](http://www.genetics.emory.edu/NUTRITION/BH4andPKU/). The study website links from the nutrition page on the Emory Department of Human Genetics website.
   You can also link to a summary of the research study on the [http://clinicaltrials.gov](http://clinicaltrials.gov) website from the research study webpage.

33. **Where are places I can eat at, or that I can visit, once my study appointment is over?**
   A. The study webpage provides information on places to eat and things to do in or around Atlanta. There is also a list provided in this log book starting on page 50.

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*Thank you for participating in our research investigation!* 😊