PKU MANAGEMENT QUESTIONNAIRE

Fill in the following information about the STUDY PARTICIPANT. Your answers to the following questions will be used for research purposes only and will be kept strictly confidential.

**Section A: Obtaining Information**

Who is completing this form?

- [ ] The study participant
- [ ] A parent/legal guardian of study participant
- [ ] Spouse of study participant
- [ ] Interviewer
- [ ] Other

Name of person completing this form: ________________________________

**Section B: Kuvan® Status**

1. When did you take your **first** dose of Kuvan® *(month and year)*?

2. Did you respond to Kuvan®?  
   - [ ] Yes  
   - [ ] No

   If “No”, when was the last time you took Kuvan® *(month/year)*:
   __________________

3. Do you currently use Kuvan®?  
   - [ ] Yes  
   - [ ] No

   If “Yes”, what is your daily dose? *(Fill in one of the spaces below)*
   - [ ] _________ mg/kg/day
   - [ ] _________ mg
   - [ ] _________ tablets

4. If you currently use, Kuvan®, have you ever missed or forgotten to take a dose?  
   - [ ] Yes  
   - [ ] No  
   - [ ] I do not take Kuvan®

   If “Yes”, how often do you **MISS** your Kuvan® dose?
   - [ ] Less than once a month
   - [ ] Once a month
   - [ ] Two or three times a month
   - [ ] Once a week
   - [ ] Two times a week
   - [ ] Every other day
   - [ ] Four or more times a week
### Section C: Diet Prescription – As prescribed by your metabolic dietitian/clinician

#### Formula Prescription

For example: Phenylfree 2 180 grams + Kool-Aid 2 Tbsp + water to make 20 fl oz

**Record formula amount as weight or as scoops**

#### Diet Prescription

For example: 20 exchanges, 300 mg Phe, or 6 grams of protein per day

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### Section D: Blood Phenylalanine Levels

In the past YEAR, which method(s) did you use to monitor your blood Phe levels?

**Mark all that apply**

- Filter paper blood spots
- Blood drawn in Clinic or Doctor’s office
- I do not monitor my blood levels
- Other: ____________________________

In the past YEAR, how often did you check your blood Phe levels?

**Mark only one**

- Once a week
- Once a month
- Once every 3 months
- Once every 6 months
- I did not check my levels in the past year
- Irregularly (please explain below):
  
  _______________________________________
- Once every other week
- Every other month
- Every 4 months
- Once a year
- I was in the Emory Kuvan® study in the past year

Irregularly (please explain below):

____________________________________

- Other (please explain below):

____________________________

In the past YEAR, my blood Phe levels have been OUTSIDE of the recommended range (2-6 mg/dL):

**Mark only one**

- Never
- Once
- Twice
- Three times
- Other:

Four or more times

- Every time I checked my levels
- My levels were not checked in the past year
- I don’t know

- Other: ____________________________

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### Section E: Diet and Formula History

<table>
<thead>
<tr>
<th>Diet-Related Questions</th>
<th>Formula-Related Questions</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>I first started my diet restriction when I was:</strong></td>
<td><strong>I first started taking my formula when I was:</strong></td>
</tr>
<tr>
<td>□ A baby</td>
<td>□ A baby</td>
</tr>
<tr>
<td>□ ______ years old</td>
<td>□ ______ years old</td>
</tr>
<tr>
<td>□ I have never restricted my diet</td>
<td>□ I have never took formula</td>
</tr>
<tr>
<td>□ I do not know</td>
<td>□ I do not know</td>
</tr>
<tr>
<td><strong>I significantly changed my diet restriction:</strong></td>
<td><strong>I significantly changed my formula intake:</strong></td>
</tr>
<tr>
<td>□ I have never changed my diet restriction</td>
<td>□ I have never changed taking my formula</td>
</tr>
<tr>
<td>□ Because I responded to Kuvan®</td>
<td>□ Because I responded to Kuvan®</td>
</tr>
<tr>
<td>□ Between ages _____ and _____ years old and I was not taking Kuvan®</td>
<td>□ Between ages _____ and _____ years old and I was not taking Kuvan®</td>
</tr>
<tr>
<td>□ Other:_____________________________</td>
<td>□ Other:_____________________________</td>
</tr>
<tr>
<td><strong>I completely stopped my diet restriction:</strong></td>
<td><strong>I completely stopped taking my formula:</strong></td>
</tr>
<tr>
<td>□ I have never stopped my diet restriction</td>
<td>□ I have never stopped taking my formula</td>
</tr>
<tr>
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</tr>
<tr>
<td>□ Other:_____________________________</td>
<td>□ Other:_____________________________</td>
</tr>
</tbody>
</table>

**On average, in the past year I ate MORE Phe than my diet prescription:**
- □ Never
- □ Less than once a week
- □ 1-2 days a week
- □ 3-4 days a week
- □ 5-6 days a week
- □ Everyday
- □ In the past year, I was not on a restricted diet and I am NOT using Kuvan®
- □ In the past year, I was not on a restricted diet because I AM using Kuvan®
- □ Other:_____________________________

**On average, in the past year I drank LESS than my entire prescription of formula:**
- □ Never
- □ Less than once a week
- □ 1-2 days a week
- □ 3-4 days a week
- □ 5-6 days a week
- □ Everyday
- □ In the past year, I did not drink formula and I am NOT using Kuvan®
- □ In the past year, I did not drink formula because I AM using Kuvan®
- □ Other:_____________________________