

- Description**
- Genetic disorders affecting breakdown and use of food such as: PKU, maple syrup urine disease, galactosemia, diagnosed by provider
 - Usually a gene mutation or deletion, special diets and special formula may be needed

Assess Readiness for Change: with open ended questions

- “How is Sam tolerating his special formula for his xx (i.e. PKU)?”
- “What other meal instructions did the Dr give you for Sam’s xx (i.e. PKU)?”

Discussion, Counseling and Education

If the answer is:	Try Counseling Strategies	Try Education
<p>Not ready →</p> <p>“We see the Dr again next week. He has been taking his formula.”</p> <p>Raise awareness & personalize change</p>	<ul style="list-style-type: none"> • “What questions do you have for the Dr next week?” • “What PKU specialty clinics have you attended?” 	<ul style="list-style-type: none"> • “You are eligible for WIC because Sam has xx (i.e. PKU), which affects what he can eat. Your ENPR is approved. We will schedule your next appointment with the RD. She will work with you to assure his WIC foods fit with his medical condition. What else can I help you with today?” • High risk, refer to RD
<p>Unsure →</p> <p>“They gave me instructions on how to mix his special formula but it is hard to do?”</p> <p>Raise awareness & confidence & identify barriers to change</p>	<ul style="list-style-type: none"> • “Tell me more about the trouble you are having mixing the formula?” • “On a scale of 1-10, how comfortable are you with mixing his formula since the instructions are different than the can says?” • “Excuse me for a minute; let me see if the RD can see you today.” 	<ul style="list-style-type: none"> • Treatment: <ul style="list-style-type: none"> ○ Remove problem item from diet- usually a protein or sugar in food, restriction is for life ○ May require specialty formula (and ENPR) or altered mixing instructions even in children ○ May require medication ○ Disorder may affect person’s thinking, language and behavior
<p>Ready →</p> <p>“I give him special formula. We go to the specialty clinic monthly.”</p> <p>Help plan change-support</p>	<ul style="list-style-type: none"> • “Tell me about the specialty clinic. What recommendations have they given you?” • “How are those changes going?” 	<ul style="list-style-type: none"> • “Your next appointment will be with our WIC RD. She will be approving all of your special formula requests.”

Record Plan or Goal Child attending specialty clinic at hospital. ENPR approved. High risk, scheduled with RD for next appointment.

Record Plan or Goal RD able to see client to review mixing instructions. RD wants to see again next appointment as follow-up.

Record Plan or Goal Attending PKU specialty clinic. To see WIC RD next appointment.

Education Resources

- <http://www.aafp.org/afp/20060601/1981.html> Resource- *IEM Update (see table 4 for therapies)*
- <http://www.medicinenet.com/phenylketonuria/article.htm> Resource- *PKU*
- <http://www.nlm.nih.gov/medlineplus/ency/article/007257.htm> Resource- *Newborn Screening Tests*
- <http://www.galactosemia.org/> Resource- *Galactosemia*
- http://www.rarediseases.org/search/rdbdetail_abstract.html?disname=Maple%20Syrup%20Urine%20Disease Resource- *Maple Syrup Disease*