



comida PKU^A formula

A practical guide for the introduction and use of **comida-PKU A formula**,
a phenylalanine-free* formula suitable from
birth to 12 months of age



*No added phenylalanine.



Enhancing Lives Together



Important information

PURPOSE

This practical guide is for the use of **comida-PKU A formula** in the dietary management of an infant with Phenylketonuria (PKU).

INTENDED USERS

This practical guide is:

- for use by **healthcare professionals** working with infants diagnosed with PKU.
- **not** for use by parents/caregivers of patients with PKU or patients themselves.
- for **general information** only and must not be used as a substitute for professional medical advice.

TARGET POPULATION

This practical guide is for use in infants with diagnosed/proven PKU.

PRODUCT INFORMATION

comida-PKU A formula is a food for special medical purposes.

Any product information contained in this practical guide, although accurate at the time of publication, is subject to change. The most current product information may be obtained by referring to product labels and **www.comidamed.com**. Please refer to these sources for information regarding allergens.

DISCLAIMER

The information contained in the practical guide is for general information purposes only and **does not** constitute medical advice. The practical guide is not a substitute for medical advice or care provided by a licensed and qualified healthcare professional and Vitaflo does not, in the absence of negligence on vitaflo's part, accept any liability arising from reliance on information contained in this guide and or the incorrect use of **comida-PKU A formula** product.

This practical guide should be read in conjunction with local, national and international guidelines and best practice for the dietary management of Phenylketonuria. Information contained within the guide is based on the most recent scientific evidence available on the management of / use of Phenylketonuria as of date of publication.

COLLABORATORS

Vitaflo dietitians in collaboration with: Suzanne Hollander, MS, RD, LDN, Genetics and Genomics, Boston Children's Hospital. Boston, MA.

Introducing and adjusting **comida-PKU A formula** is dependent on the individual patient. Practical examples are given in this guide; however, it is the responsibility of the managing healthcare professional to use clinical judgement to introduce and adjust **comida-PKU A formula** in the most appropriate way for individual patients and it may not always be appropriate to use the practical guide.

IMPORTANT NOTICE

- **comida-PKU A formula** must only be used under medical supervision.
- Suitable from birth to 12 months of age.
- Not suitable for use as a sole source of nutrition.
- **comida-PKU A formula** must only be consumed by infants with proven Phenylketonuria.
- **comida-PKU A formula** must be used in conjunction with breast milk or infant formula to provide the phenylalanine, fluid and general nutritional requirements of the infant in quantities as advised by managing metabolic healthcare professional.
- For enteral use only.

This practical guide does not establish or specify particular standards of medical care for the treatment of any conditions referred to in this practical guide.

Vitaflo International Limited **does not** recommend or endorse any specific tests, procedures, opinions, clinicians or other information that may be included or referenced in this practical guide.

Symbols and Abbreviations

Symbol	Abbreviation	Definition
	HM	Human milk
	BF	Breastfeed
	HCP	Healthcare Professional
	phe	Phenylalanine
	phe-free formula	phenylalanine-free formula (comida-PKU A formula)
	SIF	Standard infant formula

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Features of comida-PKU A formula

comida **PKU**^A formula

comida-PKU A formula is a phenylalanine free* amino acid based powdered formula containing essential and non-essential amino acids, carbohydrate, fat, vitamins, minerals, trace elements, arachidonic acid (ARA) and docosahexaenoic acid (DHA).

- comida-PKU A formula provides all the essential nutrients required by an infant, except phenylalanine.
- comida-PKU A formula is for use in the dietary management of phenylketonuria (PKU) and for hyperphenylalaninemia (HPA) suitable from birth to 12 months of age.



*No added phenylalanine.

Overview of feeding an infant with PKU

Newly diagnosed infants with PKU are prescribed a phe-free formula (**comida-PKU A formula**) immediately after diagnosis is confirmed. Feeding an infant with PKU is a balance between providing phe-free formula alongside adequate amounts of phe from HM/SIF with the goal of keeping phe levels in target therapeutic range (120–360 µmol/L)^{1,2}. This balance can be achieved with HM or SIF as the source of phe, and a family should be supported in making the appropriate choice for their family and their infant, with relevant healthcare professionals' input as appropriate.

comida-PKU A formula may be used from diagnosis in combination with HM/SIF or as a sole source of nutrition for a brief period known as a "washout" in order to achieve a rapid reduction in blood phe levels.

Breast fed infants

Healthcare providers should support a family's decision to BF or provide HM as the intact protein source for an infant with PKU.

comida-PKU A formula combined with bottle-fed HM or feeding at the breast is able to maintain satisfactory blood phe control provided there is adequate HM available for the infant and blood phe levels can be closely monitored³. HM offers nutritional benefits including higher long chain polyunsaturated fatty acid concentrations and a lower phe content, 47 mg/100 ml in HM³ compared with approximately 56 mg/100 ml in SIF³.

Breastfeeding an infant with PKU is based on the principle of giving a measured volume of **comida-PKU A formula** to offset the infant's appetite for breastfeeds. Feeding a measured amount of **comida-PKU A formula** alongside each breastfeed, or alternating measured feeds of **comida-PKU A formula** with breastfeeds, decreases the total amount of HM consumed and therefore decreases total phe intake. Infants can still feed on demand, varying the quantity of feeds from day to day provided that the prescribed quantity of **comida-PKU A formula** is given throughout the day³. Successful PKU management with breastfeeding is achieved via close monitoring of blood phe levels and adjustment of the prescribed volume of **comida-PKU A formula** by the metabolic healthcare professional in order to maintain blood phe control. See sections 1.1–1.4 for more details.

SIF fed infant

There are various options for feeding an infant with SIF and **comida-PKU A formula**. With the help of healthcare providers, including the metabolic healthcare professional, families may choose a SIF supplemented with DHA and ARA that is best for their infant and family circumstances.

SIF and **comida-PKU A formula** may be given in separate or mixed bottles, with a variety of feeding approaches capable of optimizing blood phe control and catering to a family's individual circumstances.

See Sections 1.1 – 1.4 for more details.

Blood Phe Level Monitoring

Blood phe levels are used to determine whether the volume of **comida-PKU A formula** and HM/SIF should be adjusted. Expect to adjust the feeding plan weekly, especially during the first two months of life. Blood phe levels should be checked at least weekly throughout infancy^{4,5}. The quantity of phe tolerated by infants will vary and be guided by blood phe levels. Individual phe tolerance will vary significantly throughout infancy with changes in growth and development. It is vital to investigate the possible causes for changes in phe level before adjusting the feeding plan. Consider waiting for two consecutive blood phe levels to indicate the need for a feeding plan adjustment, unless the blood phe level is very low or very high.

Progression

comida-PKU A formula and HM/SIF may continue to provide 100% of the infant's nutrition requirements until the age of 6 months, or when the infant is developmentally ready for the introduction of solids.

IMPORTANT NOTICE

Breastfeeding can continue for as long as the mother and infant wish, provided that growth and blood phe levels are satisfactory. SIF should be introduced if there is inadequate HM to provide enough phe or liquid nutrition volume for age in combination with **comida-PKU A formula**. If the mother wishes to wean the infant from HM, then a gradual approach is recommended, if possible.

Overview of feeding an infant with PKU

PRINCIPLES OF INITIATING NUTRITION MANAGEMENT

If phe levels > 360 $\mu\text{mol/L}$, implement use of **comida-PKU A formula**.

Depending on diagnostic phe level and clinical circumstances, the metabolic team may consider the following strategies for initiating **comida-PKU A formula**:

- Temporarily stop HM/SIF and use **comida-PKU A formula** as sole nutrition source for ≤ 48 hours.
- Introduce **comida-PKU A formula** in combination with HM/SIF.

Once phe levels approach the target therapeutic range, if HM/SIF has been stopped, reintroduce to tolerance.

Intact protein intake is adjusted based on individual blood phe levels.

For breastfed infants, **comida-PKU A formula** volumes will be adjusted with the intended effect of modifying the infant's appetite for HM.

Continue to give a combination of **comida-PKU A formula** + HM/SIF to provide 100% of the infant's requirements and to achieve optimal growth and phe levels.

HM/SIF and **comida-PKU A formula** should provide 100% of the infants' nutrition needs until approximately the age of 6 months when solids are introduced or when the infant is developmentally ready for the introduction of solids³.

Monitor blood phe levels weekly or more frequently as needed to achieve stability in blood phe treatment range (120-360 $\mu\text{mol/L}$)²⁻³.

- **comida-PKU A formula** and phe source should be given together at each feed, or at alternating feeds to ensure nutrient availability throughout the day.
- If the prescribed volume of **comida-PKU A formula** is not taken, it may cause a rise in blood phe levels.
- Blood phe levels are used to determine whether **comida-PKU A formula**, SIF/HM prescription should be adjusted.
- The infant should be weighed at each clinic visit.

Nutrition prescription using comida-PKU A formula at diagnosis

THE AIM IS TO ACHIEVE A RAPID REDUCTION IN BLOOD PHE LEVELS

If temporarily stopping intact protein and using **comida-PKU A formula** alone to lower very high blood phe levels.

Step 1 - Introduction of comida-PKU A formula

- Use **comida-PKU A formula** as a sole source of nutrition for 24-48 hours or until blood phe level approaches target range.
- **comida-PKU A formula** should be offered on demand to the infant.

Feeding plan and considerations:

- Mix desired amount of **comida-PKU A formula** per feed. A 1-2 week old infant will typically take 45-90 ml (1.5-3 fl oz) per feed.
- Encourage family to track number of wet and soiled diapers to ensure intake adequacy.
- Feeding frequency may need to be higher for infants who have not yet regained their birth weight. Family should consult with their pediatrician or metabolic healthcare professional.
- Encourage breastfeeding mothers to express breast milk when the infant feeds to establish and protect breast milk supply during the washout period.
- Maternal-infant skin-to-skin contact and/or a small number of short duration breastfeeds, may be continued to help promote breast milk supply and bonding during the period when breastfeeding is stopped.

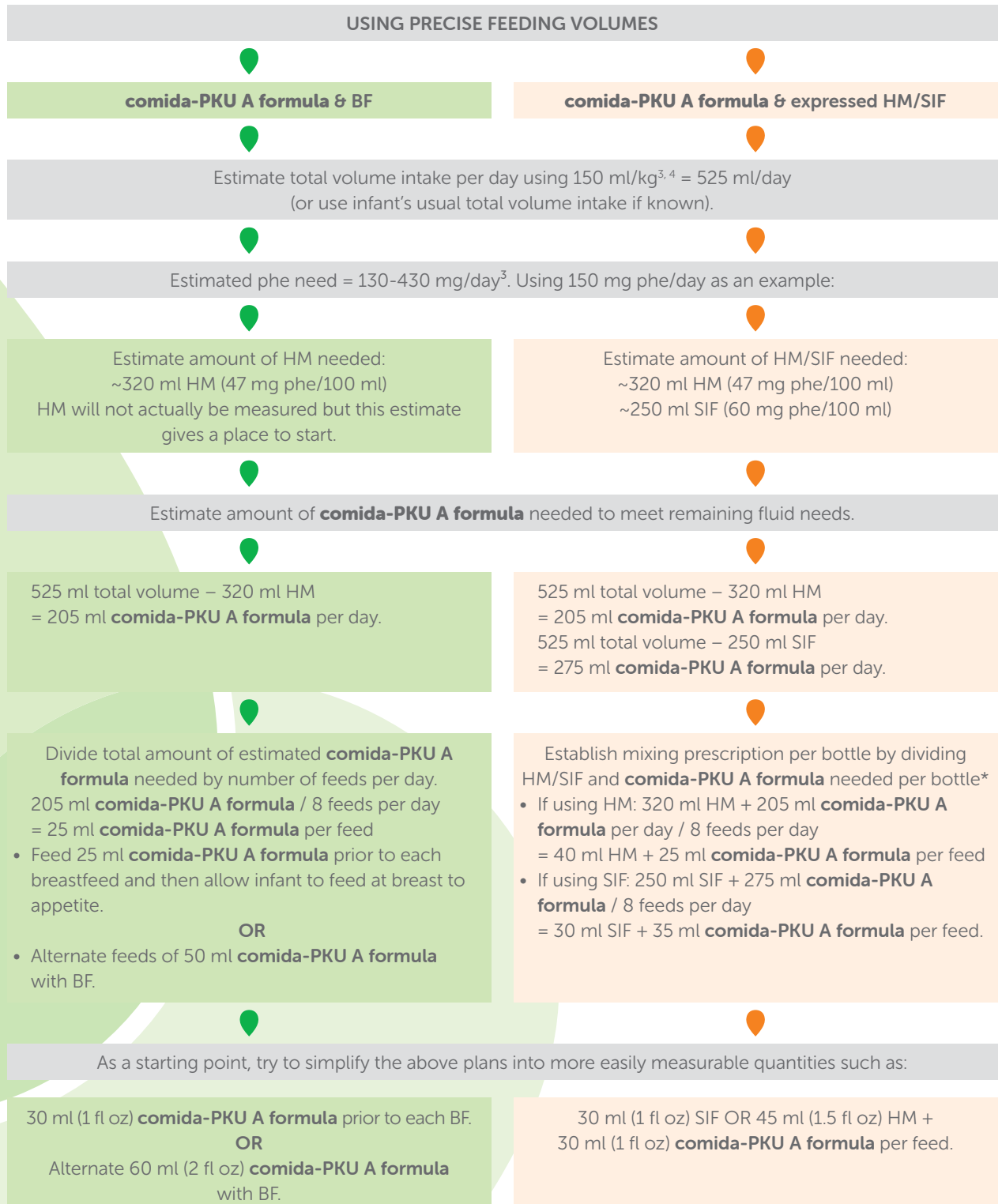
Step 2 - Reintroduction of HM/SIF

- When blood levels are in the therapeutic target range.

Example feeding plans

If a washout period is not required or reintroducing intact protein source, the following feeding plans illustrate 2 methods for calculating the feeding plan whether breastfeeding or bottle feeding with expressed HM or SIF.

Example using a 6-day old infant diagnosed with PKU, weight 3.5 kg.

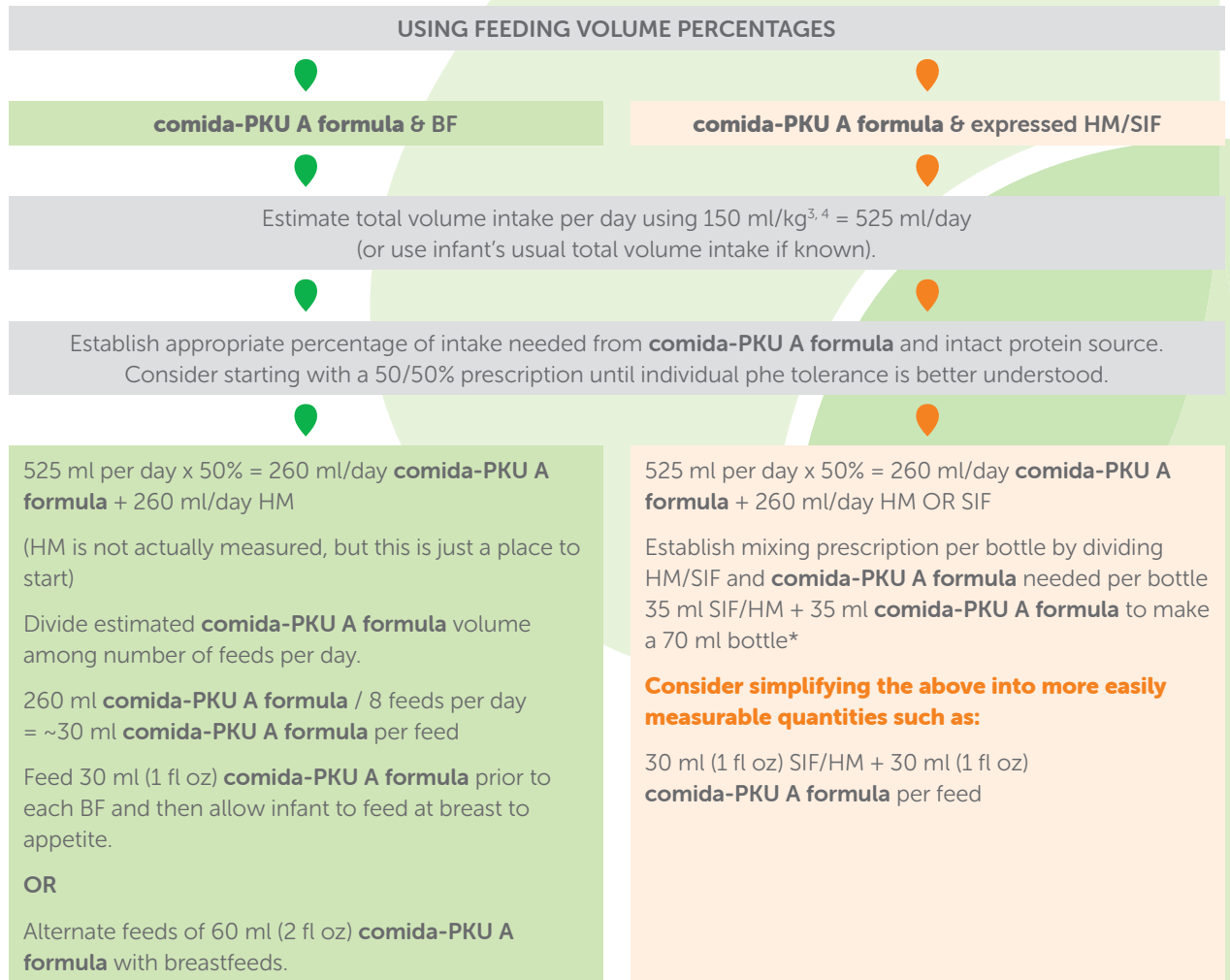


* Infants should be fed to appetite when hungry and total liquid volume per day should not be limited to establish phe level control. If the estimated volume intake does not satisfy the infant, then additional feeds of **comida-PKU A formula** alone can be given or in some cases additional **comida-PKU A formula** + HM/SIF may be indicated.

Example feeding plans

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Check list for blood PHE monitoring

Many factors can affect blood phe levels. Always check for causes of high or low blood phe level before making a change to the nutrition prescription.

Considerations for high blood PHE levels:

POSSIBLE CAUSE	ACTION
Excess intake of intact protein (HM/SIF)	• Confirm feeding preparation and provision is consistent with prescription. • Review mixing and measuring of feeds/formula. • Adjust prescription of HM/SIF and comida-PKU A formula to meet infant's phe tolerance.
Inadequate intake of comida-PKU A formula	• Ensure that adequate comida-PKU A formula supply is available. • Address symptoms that may affect tolerance such as colic, constipation, or reflux by seeking appropriate medical advice. • Determine presence of short-term symptoms affecting intake such as illness, pain, teething, or vaccination. • Monitor weight and increase comida-PKU A formula prescription as needed.
Catabolism or slow weight gain	• Monitor weights frequently to better understand growth trajectory. • Rule out illness or infection and encourage appropriate medical treatment. • Encourage optimal total volume intake and adjust feeding intervals and frequency as needed to achieve goals. • Cross-check phe and calorie intake to ensure infant is meeting requirements.
Change in blood monitoring routine	Encourage consistent timing of blood phe level within family's and infant's specific circumstances.

Check list for blood PHE monitoring

Many factors can affect blood phe levels. Always check for causes of high or low blood phe level before making a change to the nutrition prescription.

Considerations for low blood PHE levels:

POSSIBLE CAUSE	ACTION
Inadequate intake of intact protein (HM/SIF) or excessive intake of comida-PKU A formula	• Confirm feeding preparation and provision is consistent with prescription. • Review mixing and measuring of feeds/formula. • Ensure adequate HM available if applicable, supplement SIF as needed. • Adjust prescription of HM/SIF and comida-PKU A formula to meet infant's phe tolerance.
Anabolism or rapid growth phase	• Monitor weight frequently to better understand growth trajectory. • Increase phe source if blood phe level is very low; consider continuing prescription and repeating level if blood phe level is in an acceptable low range.
Change in blood monitoring routine	• Encourage consistent timing of blood phe level within family's and infant's specific circumstances.

MONITORING TIPS

For all infants, frequent monitoring of phe levels is key, but try not to make changes to a feeding plan too frequently.

Consider:

Many factors affect phe levels; review all causes in this section.

- Monitor phe level trends
 - Unless phe level is very low or very high consider:
 - Continuation of the current plan
- Monitoring 2 consecutive phe levels before adjusting prescription.
- In general, do not make more than 1 change to the plan in 1 week.

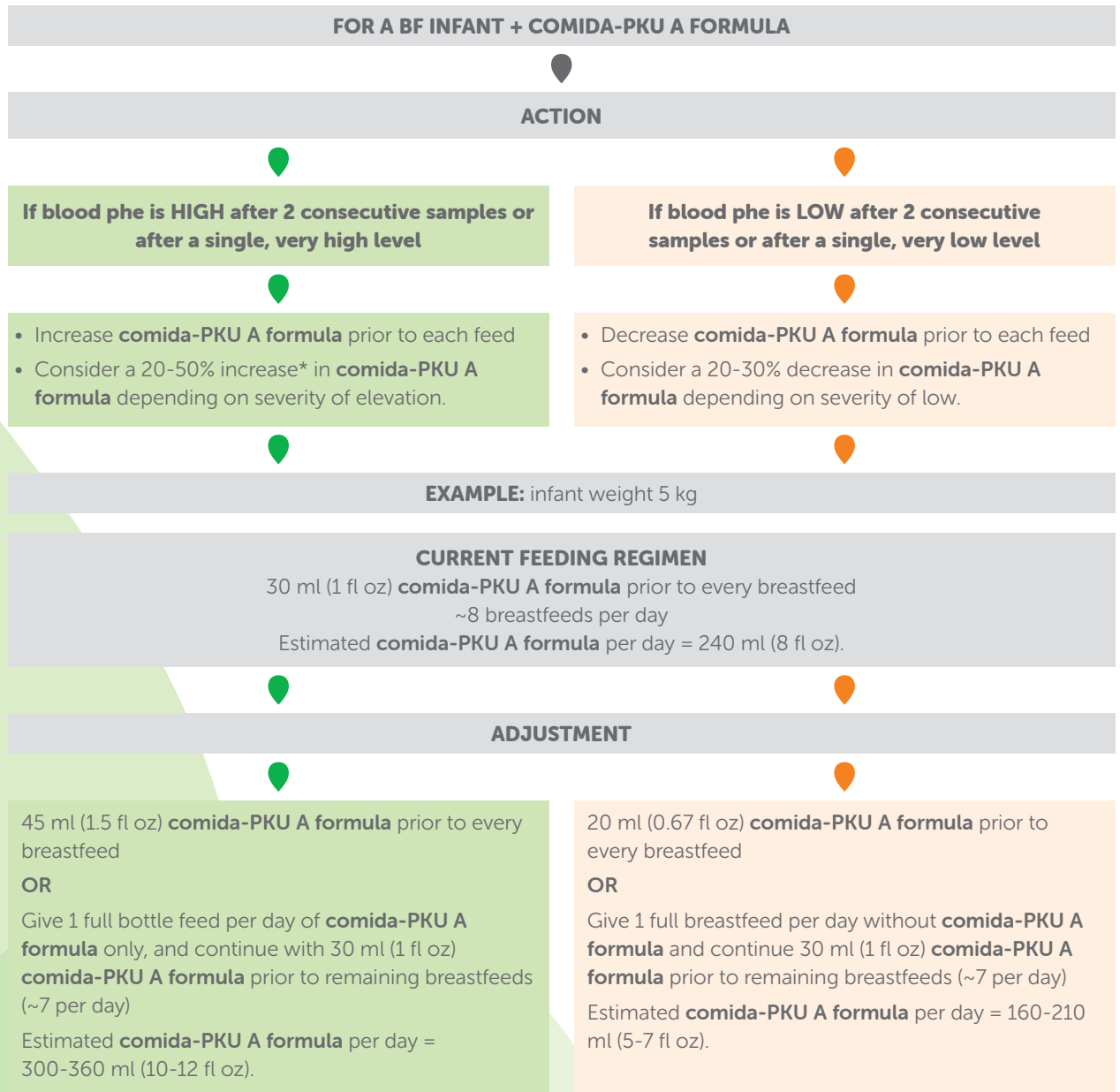
For phe levels that ARE very low or very high:

- Consider repeating blood phe level sooner than 1 week to guide interventions.

REMEMBER:

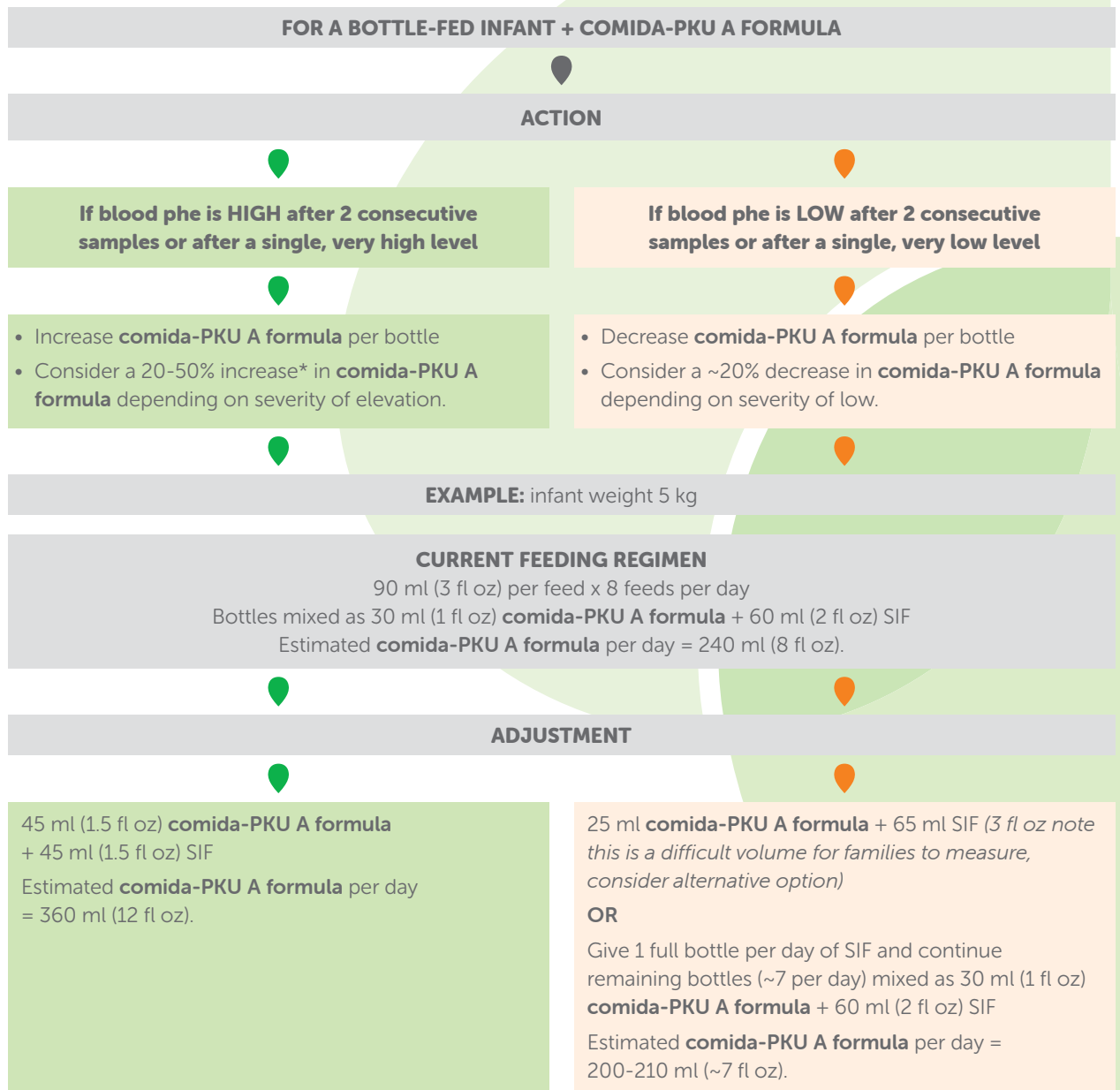
Nutrition prescription adjustments made during illness or infection will be temporary and should be closely monitored.

Fine-tuning the nutrition prescription



* Although a 50% increase seems high, it often coincides with a simultaneous increase in total infant feeding volume per day.

Fine-tuning the nutrition prescription



* If the infant is still hungry after the feed, there are different approaches that may be used to achieve satiety for the infant while maintaining blood phe control (infants should always be fed to appetite, and total feeding volume should not be restricted to achieve phe level control):

- 1) Offer additional **comida-PKU A formula** to achieve satiety.
- 2) In some cases offer a mixed bottle of **comida-PKU A formula** + HM/SIF if indicated by the metabolic healthcare professional. In some cases additional **comida-PKU A formula** + HM/SIF may indicated.

Practical feeding strategies to use with caregivers



Be prepared with feeding supplies. It is important for HCPs to know what supplies the family/caregivers have on-hand and to ensure they can get proper supplies when needed.

These Include:

Feeding bottles with necessary measurement markings (demarcation) for infant's recipe

- Appropriately staged nipples for infant feeding
- Adequate supply of **comida-PKU A formula**
- HM pump and supplies if needed to express
- Adequate supply of SIF (if using)

REVIEW FORMULA MIXING INSTRUCTIONS:

- Use **comida-PKU A formula** scoop (or gram scale) to mix **comida-PKU A formula**. Do not use scoops from other formula containers to mix **comida-PKU A formula**
- Ensure family has access to a safe water source for formula mixing
- Inquire about the most effective written method for family to receive new recipes and mixing instructions.

Review mixing at every visit

Confirm family/caregivers are able to procure more **comida-PKU A formula** and SIF (if using) when supplies are low. Consider providing written guidance on where families can obtain these formulas. Direct families to appropriate agencies and services if needed to obtain SIF at low-cost or for free.

Practical points for effective communication between HCPs and caregivers

ESTABLISH LINES OF COMMUNICATION BETWEEN HCP AND CAREGIVER/FAMILY

Phone calls, e-mail, text and/or the medical record portal may all be used. Ensure caregivers/families are comfortable using communication method.

KEEP INFORMATION SIMPLE AND PRACTICAL AND CHECK FAMILY/CAREGIVER UNDERSTANDING OF INFORMATION.

Allow time for questions and encourage questions between visits.

Consider written feeding plans.

ENCOURAGE QUESTIONS

Caregivers should be encouraged to ask questions and speak up whenever they do not understand. Effective and open lines of communication as well as appropriate teaching methods are critical for caregiver success.

EDUCATE ALL CAREGIVERS

Let primary caregiver(s) know that anyone taking care of the infant is welcome at clinic visits and education pieces may be provided for all caregivers depending on their needs.

ESTABLISH FREQUENCY OF PHE LEVELS AND CLINIC VISITS

Ensure caregivers understand what communication method will be used to report results and adjust the diet between visit.

INFORM OTHER HCPS, INCLUDING THE PRIMARY CARE PHYSICIAN, OF THE MANAGEMENT PLAN

DIRECT CAREGIVERS TO APPROPRIATE PATIENT/FAMILY SUPPORT GROUPS AND APPROPRIATE INFORMATION PLATFORMS

REMEMBER THAT DIFFERENT PATIENTS/FAMILIES SUCCEED WITH DIFFERENT COMMUNICATION METHODS;

Be as adaptable as you can within your clinic's capabilities.

Directions for preparation, use, and storage of comida-PKU A formula

PREPARATION GUIDELINES

Follow the instructions exactly and feed immediately. Incorrect preparation can make your infant ill.



Wash hands well.



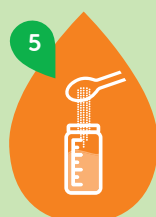
Sterilise bottle, teat and cap by boiling in water for five minutes. Leave covered until use.



Boil drinking water for five minutes. Allow to cool to lukewarm drinking temperature, i.e., water that feels warm on the wrist but not hot.



Pour exact amount of lukewarm water into the bottle.



Using the scoop provided, add the prescribed number of scoops of **comida-PKU A formula** to the water, levelling each scoop off with the back of a clean dry knife. Do not press the powder into the scoop.



Shake well until the powder is fully dissolved.



Always test the temperature before feeding by shaking a few drops onto the inside of your wrist - the feed should feel warm but not hot.

It is important for caregivers to carefully follow the instructions for the preparation, use and storage of **comida-PKU A formula**. The standard dilution of 21 kcal/fl oz or 0.68 kcal/ml is made by adding 1 level scoop of **comida-PKU A formula** (4.7 g) to 30 ml (1 fl oz) of water.

Use only the scoop provided in the can or a gram scale for greatest accuracy.

Any formula remaining in the bottle after 1 hour **should be discarded**.

- Do not reheat **comida-PKU A formula** once feeding has started.
- Do not heat **comida-PKU A formula** in a microwave as uneven heating may occur and could cause a burn.
- Do not boil **comida-PKU A formula**.
- Infants should be supervised at all times when feeding.
- Regular teeth cleaning is recommended.

Preparation Guidelines

Water (ml)	comida-PKU A formula no. scoops	comida-PKU A formula (g)	Drinking volume (ml)	Protein equivalent (g)
30	1	4,7	35	0,67
90	3	14,1	100	2
180	6	28,2	200	4

STORAGE

UNOPENED: **comida-PKU A formula** should be stored in a cool, dry place.

OPENED: Use within 3 weeks.

Always replace container lid after use.

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All information correct at the time of publication.