comidamed™

comida TYRo A formula

A practical guide for the introduction and use of **comida-TYRo A formula**, an amino acid based phenylalanine and tyrosine free* formula suitable from birth to 12 months of age.

* No added phenylalanine and tyrosine.

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Enhancing Lives Together

Important information

PURPOSE

This practical guide is for the use of **comida-TYRo A formula** in the dietary management of an infant with Tyrosinaemia.

INTENDED USERS

This practical guide is:

- for use by **healthcare professionals** working with infants diagnosed with Tyrosinaemia.
- **not** for use by parents/caregivers with Tyrosinaemia 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 Tyrosinaemia.

PRODUCT INFORMATION

comida-TYRo 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-TYRo 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 Tyrosinaemia. Information contained within the guide is based on the most recent scientific evidence available on the management of Tyrosinaemia as of date of publication.

Introducing and adjusting **comida-TYRo 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-TYRo 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-TYRo 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-TYRo A formula** must only be consumed by infants with proven Tyrosinaemia.
- **comida-TYRo A formula** must be used in conjunction with breast milk or infant formula to provide the phenylalanine, tyrosine, fluid and general nutritional requirements of the infant in quantities as advised by a 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.

COLLABORATORS

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Symbols and Abbreviations

Symbol	Abbreviation	Definition
	HM	Human milk
	BF	Breastfeed
	HCP	Healthcare Professional
	HT1	Tyrosinaemia type 1
	phe	Phenylalanine
	tyr	Tyrosine
	Phe and Tyr	Phenylalanine plus tyrosine
	Phe and tyr-free formula	Phenylalanine and tyrosine-free formula (**comida-TYRo A formula**)
	SIF	Standard infant formula

Contents

Features of comida-TYRo A formula

comida TYRo A formula

comida-TYRo A formula is a phenylalanine and tyrosine 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-TYRo A formula** provides all the essential nutrients required by an infant, with the exception of tyrosine and phenylalanine.
- **comida-TYRo A formula** is for the dietary management of Tyrosinaemia, suitable from birth to 12 months of age.

* No added phenylalanine and tyrosine.

1.0 Overview of feeding an infant with HT1

Tyrosine is a non-essential amino acid. It is found as part of dietary protein but is also synthesised from the precursor amino acid Phenylalanine, by the body. The enzyme fumarylacetoacetate hydrolase (FAH) breaks down and processes Tyrosine, however a deficiency of the enzyme causes a build-up of toxic plasma and urine metabolites, known as Tyrosinaemia type 1 (HT1).

Newly diagnosed infants with HT1 are prescribed a tyr and phe-free infant formula (for example, **comida-TYRo A formula**) as soon as diagnosis is confirmed (phe is a precursor for tyr).

Feeding an infant with HT1 is a balance between providing tyr and phe-free formula, alongside small but adequate amounts of tyr and phe from HM/SIF. The goal is to maintain levels within the target therapeutic ranges for tyr (200-600 µmol/L)[1, 2] and phe levels (20-80 µmol/L)[1, 2]. This balance can be achieved with HM or SIF as the source of tyr and phe. Families should be supported to make an appropriate choice for themselves and their infant between HM or SIF, with the help of healthcare professionals if necessary.

Breastfed infants

Healthcare providers should support a family's decision to BF or provide HM as the source of intact protein for an infant with HT1. **comida-TYRo A formula**, combined with breastfeeding or bottle-fed HM can maintain satisfactory blood tyr control. However, there must be an adequate supply of HM available and blood tyr levels closely monitored. HM provides a range of nutritional benefits, including a higher concentration of long chain polyunsaturated fatty acids and a lower concentration of phe and tyr. The phe and tyr concentration of mature HM is 48 mg Phe + 55 mg tyr/100 ml[3]; in comparison to approximately 48-60 mg Phe and 35-61mg tyr /100 ml in SIF (phe and tyr ranges, quoted from 3 common SIF brands in Canada, accessed October 2024[4]).

Breastfeeding an infant with HT1 is based on the principle of giving a measured volume of **comida-TYRo A formula**, to offset the infant's appetite for breastfeeds. Feeding a measured amount of **comida-TYRo A formula** alongside each breastfeed, or alternating measured feeds of **comida-TYRo A formula** with breastfeeds, results in a reduction in the total amount of HM consumed, consequently leading to a decrease in total phe and tyr intake.

Infants can still feed on demand, varying the quantity of feeds from day to day provided that the prescribed quantity of **comida-TYRo A formula** is given throughout the day. Successful HT1 management with breastfeeding is achieved via close monitoring of blood tyr levels and adjustment of the prescribed volume of **comida-TYRo A formula** by the metabolic healthcare professional in order to maintain blood tyr control.

See sections 1.1-1.4 for more details. In practice, we start by alternating between breastfeeding and formula feeding. We then adjust the amount of formula according to the rate of lactation.

SIF fed infants

There are several options for feeding an infant with SIF and **comida-TYRo A formula**. With the help of the metabolic healthcare professional, families can choose a SIF supplemented with DHA and ARA, that is best suited to their child and family circumstances.

Powdered SIF is preferred (rather than liquid ready-to-feed SIF) to avoid losses/wastage due to storage. A suitable nutritional recipe is calculated by the metabolic healthcare professional and given to the parents. It will be adjusted as needed according to tyr blood levels. It is recommended to give a calculated recipe of SIF plus **comida-TYRo A formula**, catering for individual and family circumstances.

See sections 1.1-1.4 for more details.

Blood tyr Level Monitoring

Blood tyr levels are used to determine whether the volume of **comida-TYRo A formula**, and HM/SIF should be adjusted. The current recommendations (interpreted as minimum recommendations) are to assess blood tyr levels on a monthly basis from the time of diagnosis until the age of five[1, 2]. However, general practice is to perform blood spot checks to assess blood tyr levels weekly and to adjust the feeding plan weekly, especially in the first two months of life. The amount of phe and tyr tolerated by infants varies and is determined by blood tyr levels.

Individual tolerance to phe and tyr varies significantly throughout childhood, depending on growth and development, and varies from one individual to another. It is essential to investigate the possible causes of variations in tyr levels before adapting the feeding plan. It is advisable to wait until two consecutive tyr blood levels indicate the need to adapt the feeding plan, unless the tyr blood level is very low or very high.

It's also important to assess phe levels, as phe is an essential amino acid and a precursor of tyr. If phe levels are lower than 20 µmol/L, it may be appropriate to increase intact protein intake or supplement phe[1, 2, 4]. Once stability has been achieved for tyr levels, it is appropriate to reduce the frequency of dried blood spots (according to the individual) to eventually.

1.0 Overview of feeding an infant with HT1

Progression

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

IMPORTANT NOTICE

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

1.0 Overview of feeding an infant with HT1

PRINCIPLES OF INITIATING NUTRITION MANAGEMENT

If tyr levels > 600 µmol/L, implement use of **comida-TYRo A formula**.

Depending on diagnostic tyr levels and clinical circumstances, the metabolic team may consider the following strategies for initiating **comida-TYRo A formula**:

- Stop HM/SIF temporarily and use **comida-TYRo A formula** as the sole nutrition source for </= 48 hours.
- Introduce **comida-TYRo A formula** in combination with HM/SIF.

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

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

For breastfed infants, **comida-TYRo 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-TYRo A formula** + HM/SIF to provide 100% of the infant's requirements and to achieve optimal growth and tyr levels.

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

Monitor blood phe and tyr levels weekly, to achieve stability: blood tyr ranges: 200 - 600 µmol/L and phe 20-80 µmol/L. Once stable, healthcare professionals may consider reducing the frequency of blood monitoring as appropriate. This will vary according to the local resources and policy.

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

1.1 Nutrition prescription using comida-TYRo A formula at diagnosis

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

If temporarily stopping intact protein and using comida-TYRo A formula alone to lower very high blood tyr levels, a two-step method is used:

Step 1 - Introduction of comida-TYRo A formula

- If tyr level at diagnosis is very high, use **comida-TYRo A formula** as a sole source of nutrition for 24-48 hours ('washout period') or until blood tyr level approaches target range. If newborn screening active in your area, then this washout period is not always necessary.
- **comida-TYRo A formula** should be offered on demand to the infant.

Feeding plan and considerations:

- Mix desired amount of **comida-TYRo 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 feeds per day (record) and 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 managing 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, (if approved by metabolic healthcare provider), 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.
- See example feeding plans over the following pages.

1.2 Example feeding plans

If a washout period is not required or reintroducing intact protein source, the following feeding plans illustrate different 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 HT1, weight 3.5 kg.

1.2.1 If breastfeeding:

As we don't know how much human milk is consumed, start by alternating feeds:

Breastfeed then **comida-TYRo A formula** from a bottle (see 1.2.2 for guidance on volume). Let the infant take as much as they want at each feed (more information to follow).

Parents should keep a meal/feed diary, noting the time of each meal (breastfeeding or bottle-feeding) and the precise amount consumed when **comida-TYRo A formula** milk is given.

1.2.2 If human milk is given by bottle-feed:

Estimate amount of HM needed, consider:
(*calculations in blue)

- Phe requirements and tyr requirements[1, 2], stated as a combined requirement figure for **0-3 months = 65-155 mg/kg/day**

  Using an average assumed tolerance 80 mg/kg/day = 280 mg phe and tyr/day (80 x 3.5 = 280 mg/day)

- **HM = 103 mg phe + tyr/100 ml = 270 ml HM/day (280/103 x 100 = 270 ml HM)**

1.2 Example feeding plans

USING FEEDING VOLUME PERCENTAGES

comida-TYRo A formula & BF	**comida-TYRo A formula** & expressed HM/SIF
Estimate total volume intake per day using 150 ml/kg[1, 5] = 525 ml/day (or use infant's usual total volume intake if known).	
Estimated phe and tyr need = 280 mg phe and tyr/day:	
Estimate amount of HM needed: ~270 ml HM (103 mg phe and tyr /100 ml) HM will not actually be measured but this estimate gives a place to start.	Estimate amount of HM needed: ~270 ml HM (103 mg phe and tyr /100 ml)
Estimate amount of **comida-TYRo A formula** needed to meet remaining fluid and nutritional needs.	
525 ml total volume – 270 ml HM = 255 ml **comida-TYRo A formula** per day.	525 ml total volume – 270 ml HM = 255 ml **comida-TYRo A formula** per day
Divide total amount of estimated **comida-TYRo A formula** needed by number of feeds per day. 255 ml **comida-TYRo A formula** / 8 feeds per day = 30 ml **comida-TYRo A formula** per feed • Alternate feed: 60 ml **comida-TYRo A formula** before breastfeeding every other feed (can be more convenient for mother and infant) OR • Alternate 120 ml (4 fl oz) **comida-TYRo A formula** with BF (this may be an option for infants which tend to take larger volumes and feed less often)..	Establish mixing prescription per bottle by dividing HM and **comida-TYRo A formula** needed per bottle* • If using HM: 270 ml HM + 255 ml **comida-TYRo A formula** per day / 8 feeds per day = 35 ml HM + 30 ml **comida-TYRo A formula** per feed
As a starting point, try to simplify the above plans into more easily measurable quantities such as:	
OR – A further option if mother prefers: • Feed 30 ml **comida-TYRo A formula** prior to each breastfeed and then allow infant to feed at breast to appetite.	Adjust amounts accordingly to make easily measurable amounts to make a total 60 ml (2 fl oz) bottles if metabolic healthcare professional feels comfortable allowing the change in phe and tyr prescription.

* These figures are provided only as a guide. Appetite can change from day to day and from feed to feed - sometimes infants drink less, sometimes more - so some variation in intake is to be expected. Infants should never be force fed. Equally, feed volume should not be limited to establish tyr level formula control. If the estimated volume intake does not satisfy the infant, then additional feeds, either of **comida-TYRo A formula** alone or mixed **comida-TYRo A formula** + HM/SIF may be given, depending on what is clinically appropriate. It may be appropriate to provide infants with slightly more than their theoretical requirements at each bottle-feed - this will allow a hungrier infant to continue feeding uninterrupted, as there won't be a need to prepare an additional bottle. Regular communication with families, combined with ongoing monitoring of blood biochemistry is key.

1.2 Example feeding plans

1.2.3 If parents choose to use SIF: Aim: to meet fluid, energy, phe and tyr requirements (Note - this is one example of practice to determine a feeding regime and local clinical practice may vary);

- Estimate total volume intake per day using 150 ml/kg[4] = 525 ml/day (or use infant's usual total volume intake if known).
- Energy requirement = **using 100 kcal/kg/day[2]** to start = 350 kcal/day **(95-145 kcal/kg/day).**
- Phe requirements and tyr requirements[1, 2], stated as a combined requirement figure for 0-3 months[5] = **using an assumed average[2] tolerance 80 mg/kg/day (65-155 mg/kg/day).**

Nutrient figures to consider:

Nutrient	SIF* (per 100 g)	HM (per 100 ml)	comida-TYRo A formula (per 100g)
Energy (kcals)	510	-	486
Protein (g)	11	varies	
Protein equivalent (g)	-	-	14,3
Phe & Tyr (mg)	890	103	-

Using SIF in combination with comida-TYRo A formula in a 3.5 kg newborn infants.
(calculations are shown in blue text in brackets).

ENERGY AND FLUID REQUIREMENTS

Fluid requirement = 150 ml/kg/day = 525 ml/day*.
Energy requirement = 100 kcal/kg/day = 350 kcal/day.

PROVIDING THE PHE & TYR ALLOWANCE (Assuming tolerance = 80 mg/kg/day)

Phe and Tyr tolerance = 280 mg/day.

This can be provided by 31 g of SIF.
(890/100 = 8.9, 280/8.9 = 31 g SIF).

This amount of SIF provides 158 kcal.

MEETING ENERGY REQUIREMENTS

Energy that needs to be provided by **comida-TYRo A formula** = 192 kcal/day.
(350 – 158 = 192 kcal/day)

This amount of energy can be provided by 40 g of **comida-TYRo A formula**.
(486/100 = 4.86 kcal/g) - 192/4.86 =40g **comida-TYRo A formula**).

Total amount of powder formula = 31 (SIF) + 40 (**comida-TYRo A formula**) = 71 g of powder/day.

CALCULATING ADDED WATER

Added water = 525 – (71 x 0.7[†]) = 475 ml/day
* Based on an approximate displacement factor of 0.7 ml/g or 70%.

INDIVIDUAL BOTTLE RECIPES

Assuming 8 feeds per day; divide SIF and **comida-TYRo A formula** powder total daily amounts and added fluid into 8 equal portions, whilst providing instructions that are practical for the family.

EACH BOTTLE SHOULD CONTAIN: 4 g of SIF + 5 g of **comida-TYRo A formula** + 60 ml of water*

(31/8 = 4 g SIF and 40/8 = 5 g **comida-TYRo A formula**) (475/8 = 60 ml water)

This feed recipe has an energy density of approx.

20 kcal fl oz (0.67 kcal/ml)** which is typical for a standard SIF.

This feed recipe will provide 2.3 g of protein equivalent/kg/day***.

* These figures are provided only as a guide. Appetite can change from day to day and from feed to feed - sometimes infants drink less, sometimes more - so some variation in intake is to be expected. Infants should never be force fed. Equally, feed volume should not be limited to establish tyr level formula control. If the estimated volume intake does not satisfy the infant, then additional feeds, either of **comida-TYRo A formula** alone or mixed **comida-TYRo A formula** + HM/SIF may be given, depending on what is clinically appropriate. It may be appropriate to provide infants with slightly more than their theoretical requirements at each bottle-feed - this will allow a hungrier infant to continue feeding uninterrupted, as there won't be a need to prepare an additional bottle. Regular communication with families, combined with ongoing monitoring of blood biochemistry is key.

** As SIFs provide 20 kcal/ fl oz (0.67 kcal/ml), it is advisable to begin with that concentration. If growth is below the recommended level and/or if the plasma amino acid profile indicates deficiency, the concentration should be increased.

*** It is challenging to achieve a protein intake of 3.0-3.5 g/kg/day. Infants should not be forced to drink high volumes of formula to achieve this intake. Adequacy of protein intake can be assessed on an ongoing basis using parameters such as growth and biochemical control.

1.3 Check list for blood TYR monitoring

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

Considerations for high blood TYR 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-TYRo A formula** to meet infant's phe + tyr tolerance.
Inadequate intake of comida-TYRo A formula	• Ensure that adequate **comida-TYRo A formula** 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-TYRo A formula** prescription as needed.
Catabolism or slow weight gain	• Monitor weight 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 tyr and calorie intake to ensure infant is meeting requirements.
Change in blood monitoring routine	Encourage consistent timing of blood tyr level within family's and infant's specific circumstances.

1.3 Check list for blood TYR monitoring

Considerations for low blood TYR levels:

POSSIBLE CAUSE	ACTION
Inadequate intake of intact protein (HM/SIF) or excessive intake of comida-TYRo 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-TYRo A formula** to meet infant's phe + tyr tolerance.
Anabolism or rapid growth phase	• Monitor weight frequently to better understand growth trajectory. • Increase phe + tyr source if blood tyr level is very low; consider continuing prescription and repeating level if blood tyr level is in an acceptable low range.
Change in blood monitoring routine	• Encourage consistent timing of blood tyr level within family's and infant's specific circumstances.

MONITORING TIPS

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

Consider:

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

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

For tyr levels that ARE very low or very high:

- Consider repeating blood tyr level sooner than 1 week to guide interventions or other intervention as recommended by metabolic healthcare professional.

REMEMBER:

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

Sick day protocols are not addressed in this guide because the risk of metabolic decompensation is particularly associated with non-compliance with medication (NTBC: Nitinisone), and advice should be sought from the metabolic healthcare team.

1.4 Fine-tuning the nutrition prescription

FOR A BREASTFED INFANT + **comida-TYRo A formula**

ACTION

If blood tyr HIGH after 2 consecutive samples or after a single, very high level	**If blood tyr LOW after 2 consecutive samples or after a single, very high level**
• Increase **comida-TYRo A formula** prior to each feed or spread it out over the day. Some parents prefer to give certain feeds through breastfeeding only, such as at night. • Consider up to 20% increase in **comida-TYRo A formula** depending on depending on severity of elevation.	• Reduce the total amount of **comida-TYRo A formula** by spreading it out over the day (either before each feed or alternating with breastfeeding). Consider up to 20% decrease in **comida-TYRo A formula** depending on how severe the 'low' is.

EXAMPLE: infant weight 5 kg

CURRENT FEEDING REGIMEN
30 ml (1 fl oz) **comida-TYRo A formula** prior to every breastfeed
~8 breastfeeds per day
Estimated **comida-TYRo A formula** per day = 240 ml (8 fl oz).

ADJUSTMENT

45 ml (1.5 fl oz) **comida-TYRo A formula** prior to every breastfeed. **OR** Give 3 full bottles a day of **comida-TYRo A formula** only (approximately 120 ml/bottle), alternating with feeding (~5 a day). There will therefore be 2 consecutive feeds during the day. Estimated quantity of **comida-TYRo A formula** per day = 300-360 ml (10-12 fl oz).	20 ml (0.67 fl oz) **comida-TYRo A formula** prior to every breastfeed. **OR** Give 1 full bottle a day of **comida-TYRo A formula** alone (approximately 120 ml/bottle), followed by another 40 ml of **comida-TYRo A formula** before a feed later in the day. Other feeds will be by breastfeeding only (6 feeds). Estimated **comida-TYRo A formula** per day = 160 ml (5-6 fl oz). There are several ways of doing this. The important thing is to distribute natural proteins as evenly as possible, and to make this decision with the parents to make it easier to apply the recommendations.

1.4 Fine-tuning the nutrition prescription

FOR A BOTTLE-FED INFANT + **comida-TYRo A formula**

ACTION

If blood tyr is HIGH after 2 consecutive samples or after a single, very high level	**If blood tyr is LOW after 2 consecutive samples or after a single, very high level**
• Consider a 10- 20% increase in **comida-TYRo A formula** depending on severity of elevation.	• Consider a 10-20% decrease in **comida-TYRo A formula** depending on severity of low.

* 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 tyr control (infants should always be fed to appetite, and total feeding volume should not be restricted to achieve tyr level control): Caregivers should call the healthcare provider to adjust the amount of formula or feeding routine. The healthcare provider will then take tyr blood levels into account when making recommendations.

2.0 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 measuring demarcation for infant's recipe.

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

REVIEW FORMULA MIXING INSTRUCTIONS:

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

Confirm that family/caregivers are able to procure more **comida-TYRo 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.

2.1 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.

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

PREPARATION GUIDELINES

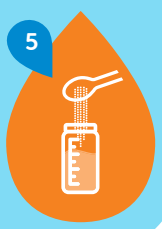
Follow the instructions exactly and feed immediately. Incorrect preparation can make your infant ill.

1. 
Wash hands well.

2. 
Sterilise bottle, teat and cap by boiling in water for five minutes. Leave covered until use.

3. 
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.

4. 
Pour exact amount of lukewarm water into the bottle.

5. 
Using the scoop provided, add the prescribed number of scoops of **comida-TYRo 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.

6. 
Shake well until the powder is fully dissolved.

7. 
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-TYRo A formula**.

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

Preparation Guidelines				
Water (ml)	comida-TYRo A formula no. scoops	comida-TYRo 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-TYRo A formula** should be stored in a cool, dry place.

OPENED: Use within 3 weeks.

Always replace container lid after use.

4.0 References

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