# At a Glance
# Hereditary Tyrosinemia Type 1 (HT-1)

**Deficient enzyme**: Fumarylacetoacetate Hydrolase (FAH)

**Toxic Metabolite**: Succinylacetone and succinylacetoacetate

**Clinical presentation, if untreated**: failure to thrive, rickets, hepatic failure, renal failure, neurologic comorbidities

**Restricted Amino Acids:** Phenylalanine and Tyrosine

***Goal Treatment Range**:

Plasma Phenylalanine: 20-80 µmol/L
Plasma Tyrosine: 200-600 µmol/L

*Chinsky JM, et al. Diagnosis and treatment of tyrosinemia type 1: a US and Canadian consensus group review and recommendations. Genetics in Medicine, Aug 2017.

**Nutrient Needs by Age***

| Age | Phenylalanine plus Tyrosine mg/kg/d | Total Protein g/kg/d | Energy kcal/kg/d |
|---|---|---|---|
| **0-3 mo** | 65 - 155 | 3.0 - 3.5 | 120 (95 - 145) |
| **3-6 mo** | 55 - 135 | 3.0 - 3.5 | 120 (95 - 145) |
| **6-9 mo** | 50 - 120 | 2.5 - 3.0 | 110 (80 - 135) |
| **9-12 mo** | 40 - 105 | 2.5 - 3.0 | 105 (80-135) |
| **1-4 yrs** | 380 – 800 mg/d | >/= 30 g/d | 1300 (900-1800) |

*Acosta PB. Nutrition Support Protocols: The Ross Metabolic Formula System. Abbot Laboratories, 2001

## Starting a HT-1 Diet

1. Determine goals for Phenylalanine (mg) plus Tyrosine (mg), Total Protein (g), and Energy (kcal)
   -use 50 mg Phe = 1 g protein to calculate milligrams of phe from protein
2. Calculate amount of whole protein source (breast milk, infant formula, food) needed to meet phe + tyr goal.
3. Calculate amount of medical food needed in addition to the whole protein source to meet total protein goal.
4. Calculate energy intake from whole protein and medical food sources to ensure total calorie needs are met.

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## Medical Food Therapy

| | Abbott<br>abbottnutrition.com | Cambrooke<br>Cambrooke.com | Mead Johnson<br>hcp.meadjonson.com | Nutricia<br>NutriciaMetabolics.com | Vitaflo<br>www.vitafloUSA.com |
|---|---|---|---|---|---|
| **Infant (0-1 yr)** | Tyrex®-1 | | Tyros 1 | TYR Anamix® Early Years | |
| **Toddler & Young Children** | Tyrex®-1<br>Tyrex®-2 | *Tylactin® Complete 15 Bar<br>Tylactin® RTD 15<br>Tylactin® RESTORE 10<br>Tylactin® RESTORE Powder 5<br>Tylactin® BUILD 20 | Tyros 1<br>Tyros 2 | TYR Anamix® Early Years<br>TYR Anamix® Next<br>*TYR Lophlex® GMP Mix-In | TYR gel™<br>TYR express® 15<br>TYR cooler® 15<br>*TYR sphere™ 20 |
| **Older Children & Adults** | Tyrex®-2 | *Tylactin® Complete 15 Bar<br>Tylactin® RTD 15<br>Tylactin® RESTORE 10<br>Tylactin® RESTORE Powder 5<br>Tylactin® BUILD 20 | Tyros 2 | TYR Anamix® Next<br>*TYR Lophlex® GMP Mix-In<br>TYR Lophlex® LQ | TYR express® 15, 20<br>TYR cooler® 15<br>*TYR sphere™ 20 |

*Product contains GMP

**Medical Therapy***

Nitisinone (NTBC)

Orfadin® www.orfadin.com

NITYR™ www.cyclepharma.com

-Starting dose: 1 mg/kg/d, increase to 2 mg/kg/d for those in acute severe liver failure

-Goal blood NTBC concentration- 30-70 μmol/L

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**Laboratory Monitoring***

| | | | |
|---|---|---|---|
| Plasma succinylacetone[1,4,7] | Serum AFP concentration[1,5,7] | BUN/Creat[8] | ALT/AST[2,6] |
| Plasma Amino Acids[1,4,7] | PT/PTT[1,6] | Calcium[8] | CBC[2,6] |
| Blood NTBC concentration[3,4,7] | Bicarbonate[8] | Phosphorous[8] | |

[1] At initiation of treatment, then monthly for the first year of life
[2] At initiation of treatment, then every 3 months for the first year of life
[3] Monthly for the first year of life
[4] Every 3 months from age 1 year thru 5 years
[5] Every 6 months from age 1 year thru 5 years
[6] Annually after 1 year of age
[7] Every 6 months after age 5 years
[8] At initiation then annually

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