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[1]:

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

Nutrient Needs by Age[2]

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)

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 abbottnutrition.com	Cambrooke Cambrooke.com	Mead Johnson hcp.meadjonson.com	Nutricia NutriciaMetabolics.com	Vitaflo www.vitafloUSA.com
Infant (0-1 yr)	Tyrex®-1		Tyros 1	TYR Anamix® Early Years	
Toddler & Young Children	Tyrex-1 Tyrex-2	*Tylactin® Complete Tylactin RTD 15 Tylactin RESTORE Citrus Tylactin RESTORE Powder Berry Tylactin BUILD 20	Tyros 1 Tyros 2	TYR Anamix Early Years TYR Anamix Next	TYR gel™ TYR express™ plus 15,20 TYR cooler® 15 *TYR sphere® 20
Older Children & Adults	Tyrex-2	*Tylactin Complete Tylactin RTD 15 Tylactin RESTORE Citrus Tylactin RESTORE Powder Berry Tylactin BUILD 20	Tyros 2	TYR Anamix Next	TYR express plus 15, 20 TYR cooler 15 *TYR sphere 20

*Product contains GMP

Medical Therapy [1]

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

Laboratory Monitoring [1]

Plasma succinylacetone [A,D,G]	Serum AFP concentration [A,E,G]	BUN/Creat [H]	ALT/AST [B,F]
Plasma Amino Acids [A,D,G]	PT/PTT [A,F]	Calcium [H]	CBC [B,F]
Blood NTBC concentration [C,D,G]	Bicarbonate [H]	Phosphorous [H]	

[A] At initiation of treatment, then monthly for the first year of life
[B] At initiation of treatment, then every 3 months for the first year of life
[C] Monthly for the first year of life
[D] Every 3 months from age 1 year thru 5 years
[E] Every 6 months from age 1 year thru 5 years
[F] Annually after 1 year of age
[G] Every 6 months after age 5 years
[H] At initiation then annually

References

1. 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.
2. Acosta PB. Nutrition Support Protocols: The Ross Metabolic Formula System. Abbot Laboratories, 2001