

The Importance of Early Intervention and Education for Caregivers of Children with Hereditary Tyrosinemia

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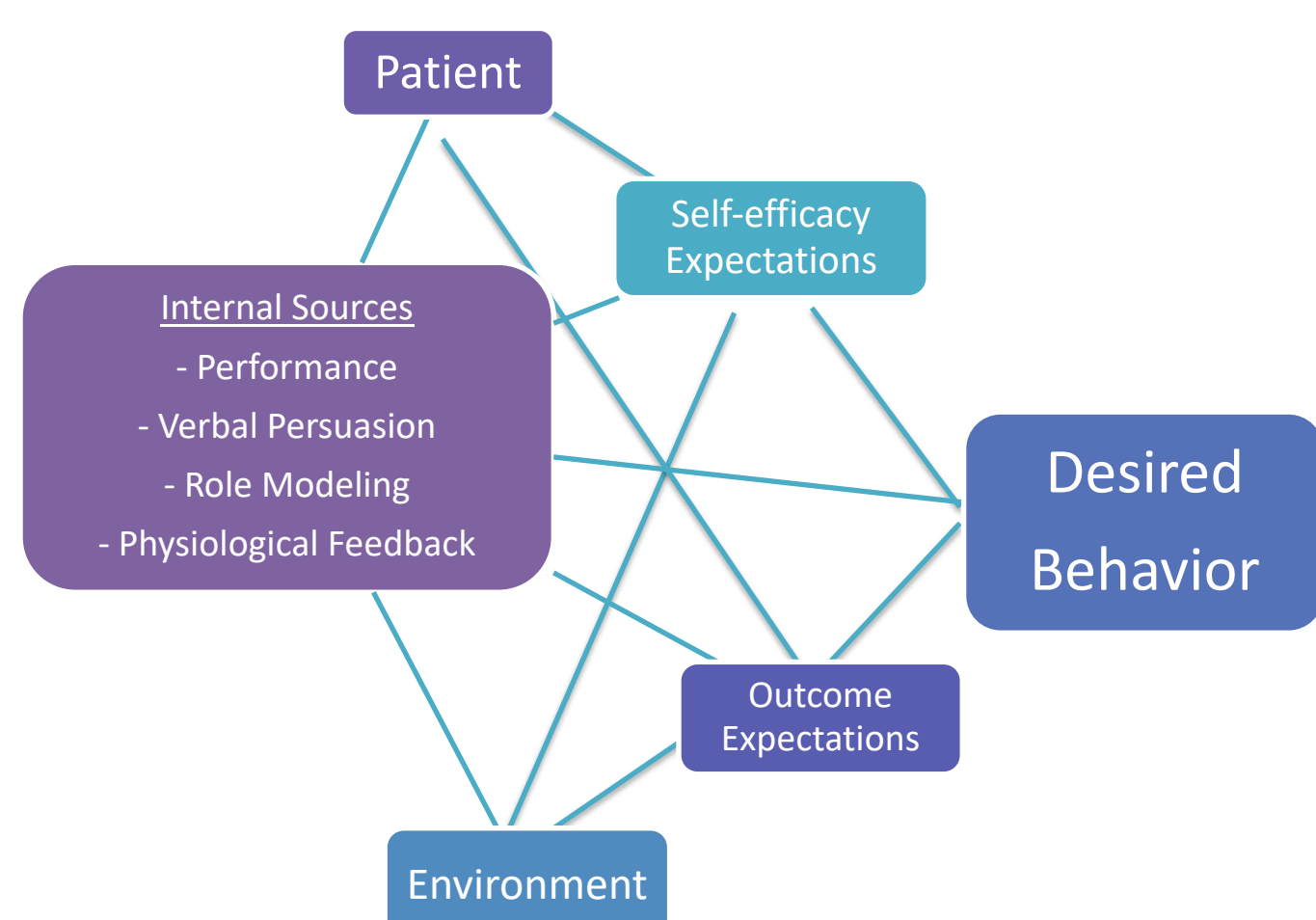
Identification of the Problem

- Health literacy is the patient's ability to apply medical information when making a decision about one's health.
- Health literacy is connected to a patient's education level.
- For patients to be involved in their care plan, health materials must be written at an appropriate reading level.
- Brochures were created with 10.7 readability score to explain Hereditary Tyrosinemia in terms of pathology, screening, and treatment.

Enzyme	Disease	Incidence	Tyrosine/Plasma (pmole/l)	Signs
TAT	HT2	Rare	370-3300	Keratosis, keratitis, corneal eruptions, mental retardation
	HT3	Very rare	355-640	Mental retardation and ataxia
HPD	Transient tyrosinemia of the newborn	30-50% of premature babies	Up to 2000	Mild symptoms or asymptomatic
FAH	HT1	Rare to frequent	400-800	Liver and kidney dysfunction

Conceptual Framework

- Self-efficacy describes a patient's evaluation of his or her abilities to organize and execute a plan of care.
- The Theory of Self-Efficacy examines the triadic relationship between patient, behavior, and environment.



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Implementation

Hereditary Tyrosinemia is a group of autosomal recessive disorders characterized by a deficiency in an enzyme catalyst (FAH, TAT or HPD) in the tyrosine catabolic pathway. The lack of functional enzymes causes an elevation in the serum levels of tyrosine and its byproducts. Due to the rarity of the disorders, patients and caregivers lack access to educational materials describing the disorders and how to manage the signs and symptoms.



Evaluation

- With simplified educational material, patients and caregivers will be able to understand the complicated disease process of Hereditary Tyrosinemia and seek the correct treatment.
- Early intervention with patients with Hereditary Tyrosinemia is key to decreasing morbidity and mortality for all non-transient forms of Tyrosinemia.

References

1. De Chesnay, M., & Anderson, B. A. (2016). *Caring for the vulnerable: Perspectives in nursing theory, practice, and research* (4th ed.). Burlington, MA: Jones & Bartlett Learning.
2. Tanguay, R. M. (Ed.). (2017). *Hereditary tyrosinemia: Pathogenesis, screening and management*. Cham, Switzerland: Springer International.
3. Smith, M. J., & Liehr, P.R. (2014). *Middle range theory for nursing* (3rd ed.). New York, NY: Springer.