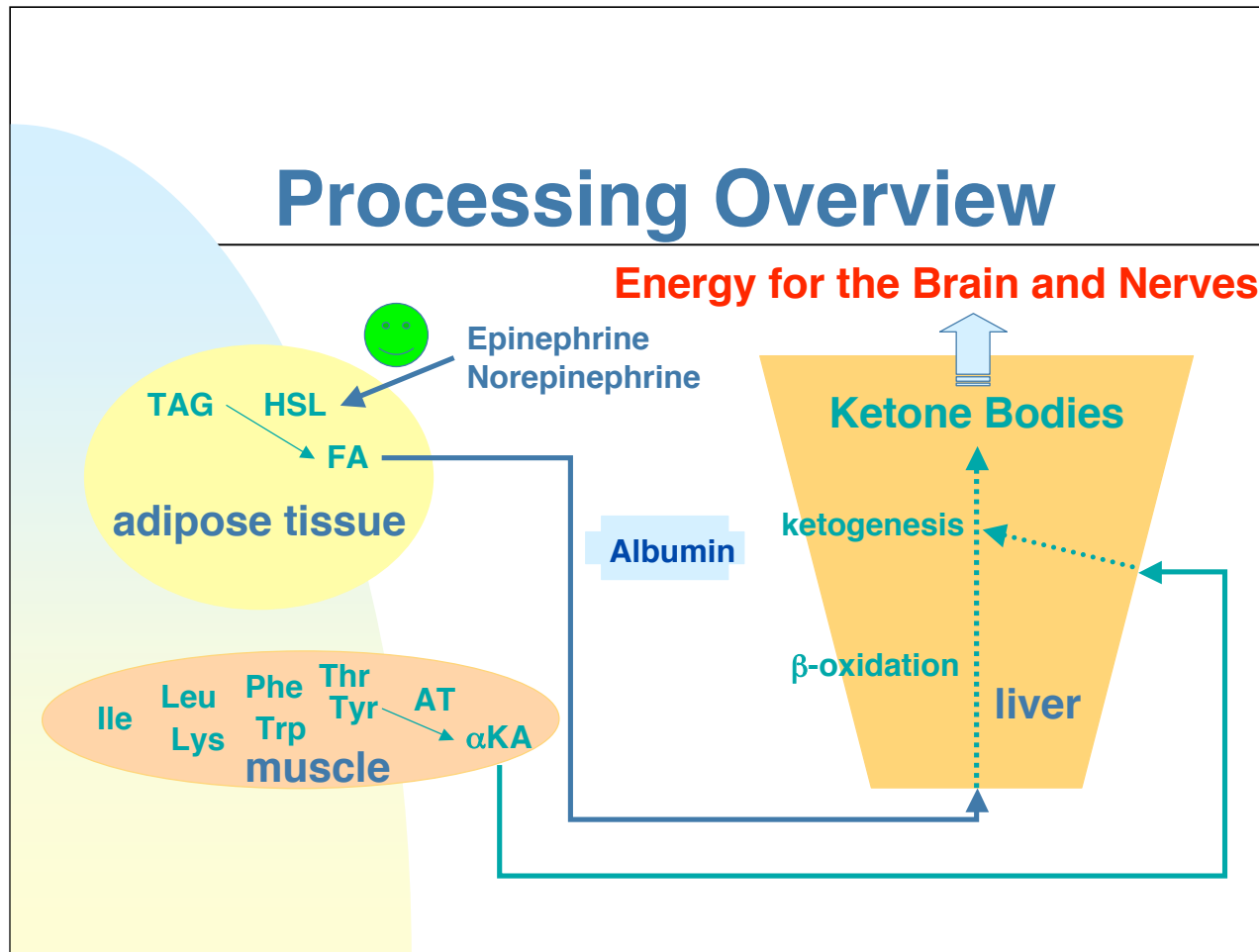


Ketone Bodies

- **Processing overview**
- **Ketogenesis and utilization**

Eric Niederhoffer
SIU-SOM



TAG: triacylglycerol

HSL: hormone sensitive lipase

FA: fatty acid

Ile: isoleucine

Leu: leucine; prominent source

Lys: lysine

Phe: phenylalanine

Trp: tryptophan

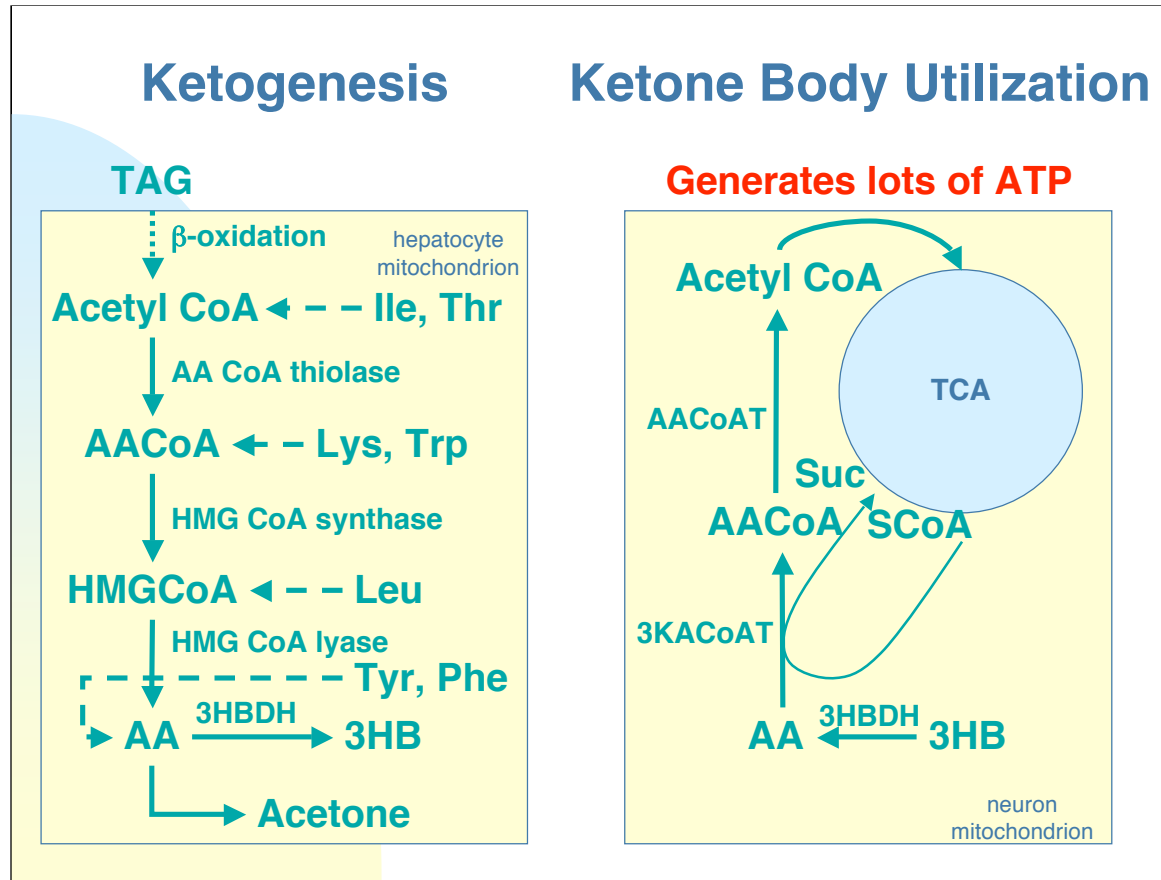
Tyr: tyrosine

AT: aminotransferase

α KA: α -ketoacid

Ketogenic amino acids: Ile, Leu, Lys, Phe, Trp, Tyr

FA are bound to serum albumin and are transported through capillaries



TAG: triacylglycerol

AACoA: acetoacetyl coenzyme A

HMGCoAS: 3-hydroxy-3-methylglutaryl coenzyme A synthase

Leu: leucine

3HBDH: 3-hydroxybutyrate dehydrogenase

Inborn errors in HMGCoA synthase and HMGCoA lyase (increased [HMGCoA])

Alcoholic ketoacidosis can result from abrupt halt in EtOH consumption leading to high 3HB:AA ratio (NADH is required). Clinical assay only looks for AA, so underestimates ketone bodies.

β-oxidation is inhibited by high [NADH]/[NAD⁺], so ketogenesis is most likely through ketogenic amino acids.

Utilization of ketone bodies spares the muscle wasting associated with gluconeogenesis from muscle amino acids.

Acetyl CoA: acetyl coenzyme A

Lys: lysine

HMGCoA: 3-hydroxy-3-methylglutaryl coenzyme A

AA: acetoacetate

3HB: 3-hydroxybutyrate

Ile: isoleucine

Trp: tryptophan

Tyr: tyrosine

Phe: phenylalanine

3KACoAT: 3-ketoacyl coenzyme A transferase

AACoAT: acetoacetyl coenzyme A thiolase

Review Questions

- **What are ketone bodies?**
- **Which amino acids are ketogenic?**
- **How are ketone bodies generated (substrates, enzymes, pathways, location)?**
- **How are ketone bodies metabolized (enzymes, pathways, location)?**
- **What is the effect of NADH levels on ketone bodies?**