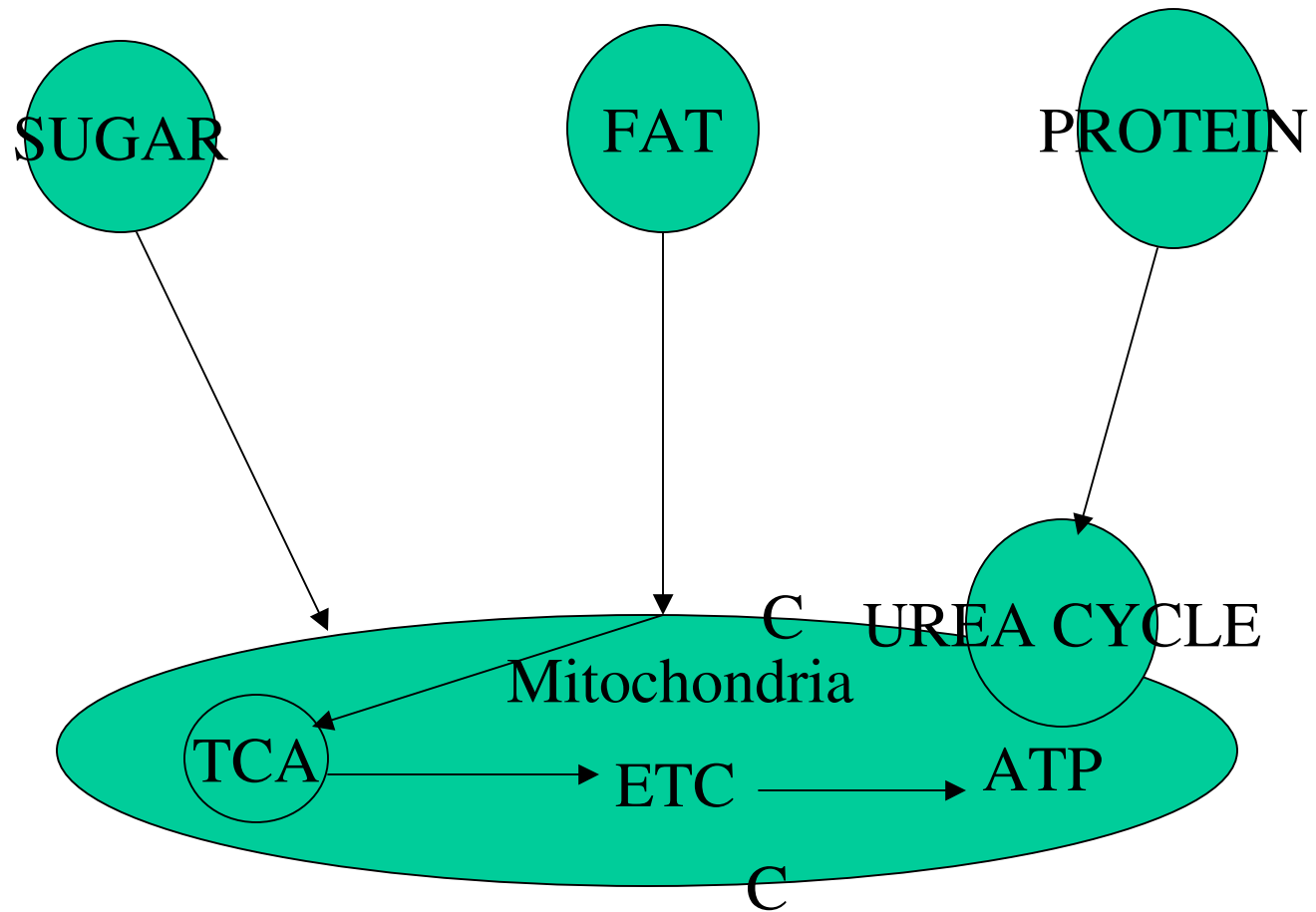


Supplements and Co-factors in the Treatment of Metabolic Disorders

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Food Groups of Metabolic Disease



Current Therapy for Metabolic Disease

Avoid Catabolism

- Newborn Weight Loss
- Febrile Illnesses, trauma, surgery
- Provide Calories from “non-offending” foods groups

Current Therapy of Metabolic Disease

Replace “created” deficiencies

- Calcium and other macro/micronutrients
- Carnitine
- Essential Fatty acids

Current Therapy of Metabolic Disease

Provide elements not released or able to be used from apoenzyme

- Biotin - Biotinidase def
- Cobalamin

Current Therapy of Metabolic Disease

Remove Toxic build up

- Carnitine in OA's & FOD's
- Glycine - Isovaleric acidemia
- Sodium Benzoate and Dextromethorphan - Nonketotic Hyperglycinemia
- Sodium Phenylbutyrate - Urea Cycle disorders

Current Therapy of Metabolic Disease

Go around metabolic block or create a block we can manage

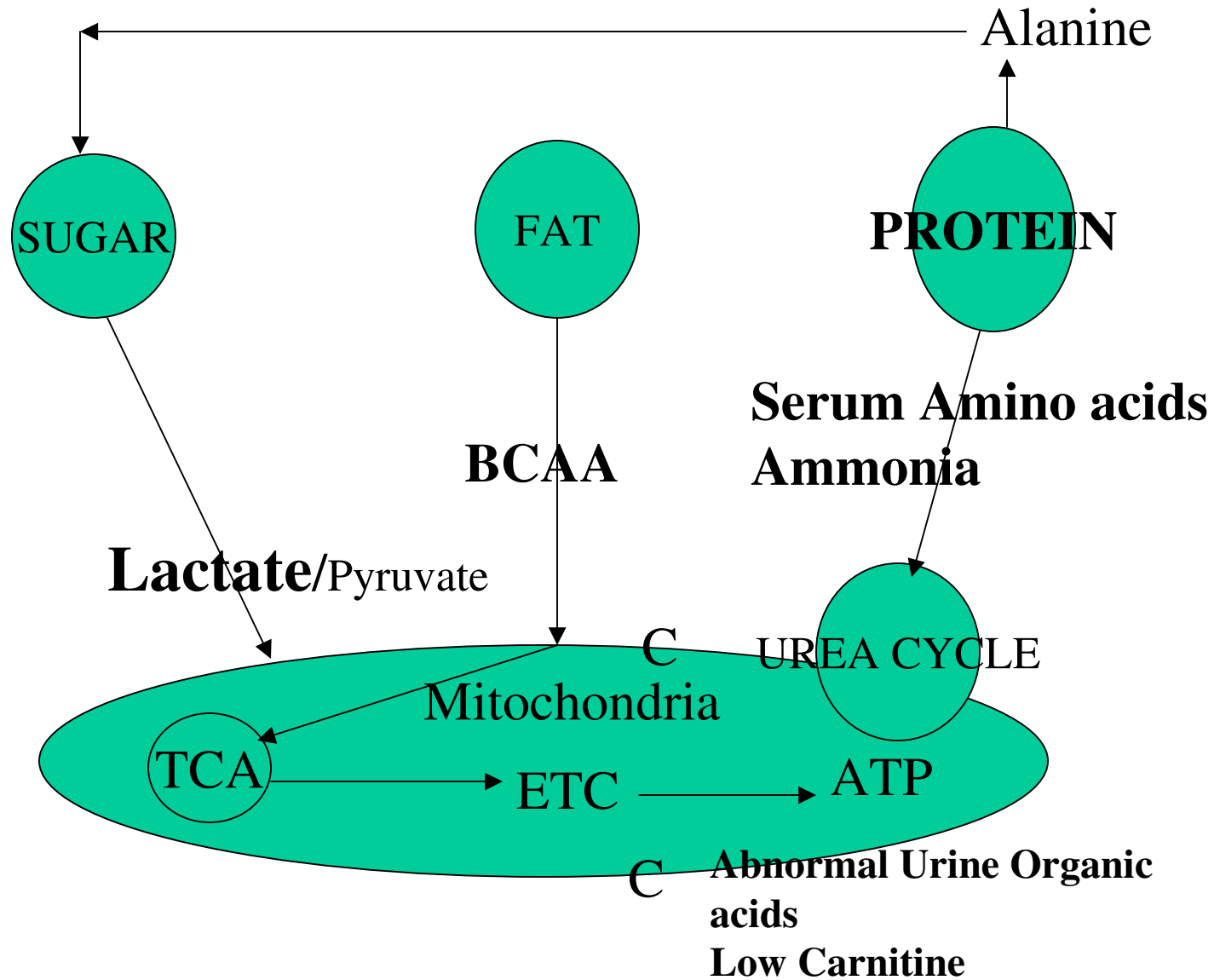
- MCT Oil - Long chain FOD's
- Orfadin (NTBC) - Tyrosinemia

Current Therapy of Metabolic Disease

Augment Enzyme function

- Riboflavin - FOD's, GAI
- Biotin - Propionic acidemia

Food Groups of Metabolic Disease



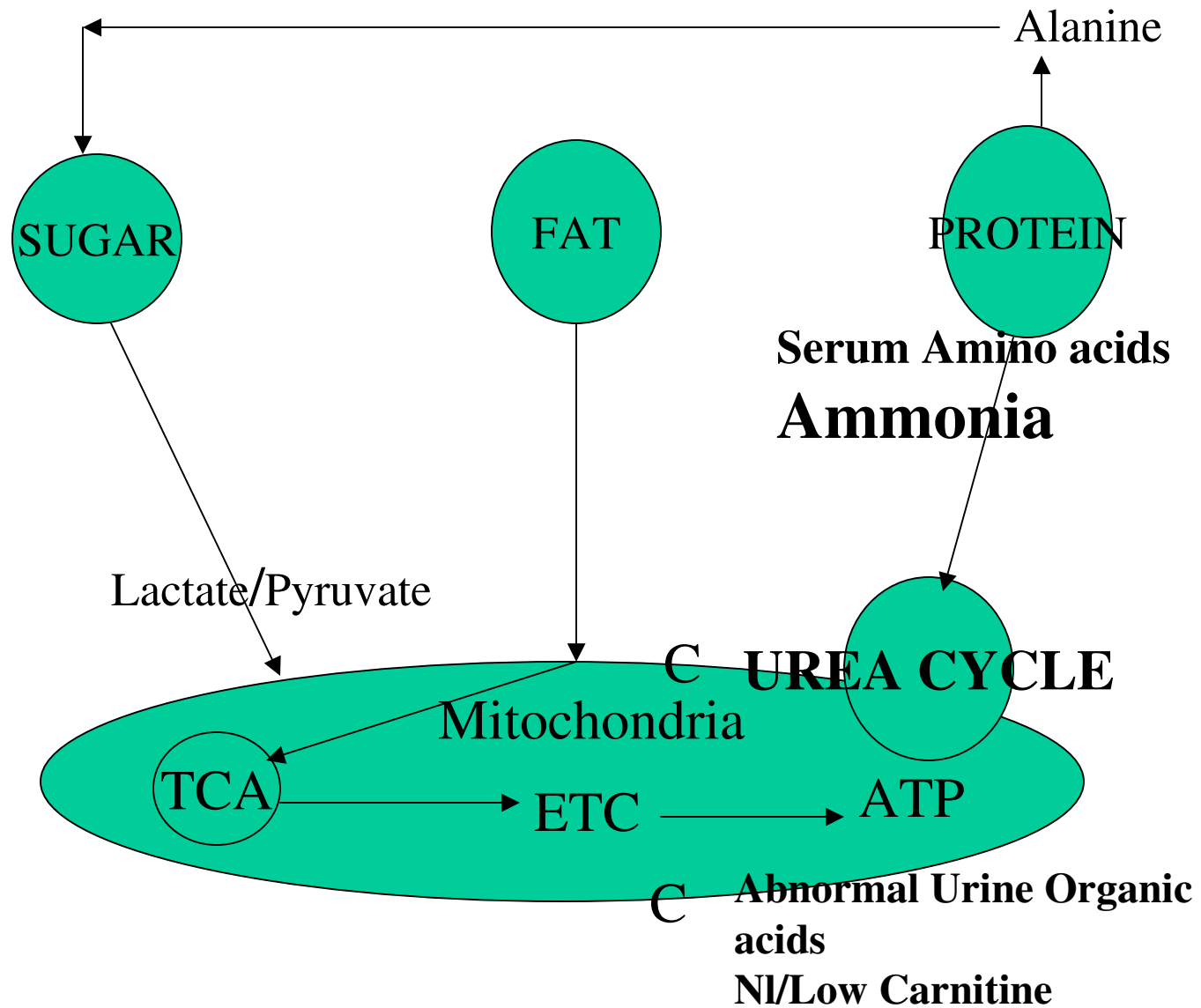
Current Treatment

- Restrict Offending Amino acids
- Supplement single Amino acids as needed
- Carnitine supplementation
- Add calories with carbohydrates and fats
- Biotin, Glycine in specific disorders
- +/- Antibiotics (Flagyl)

? Potential New Treatments

- BH4 supplementation - PKU
- Enzyme Replacement - PKU
- CoQ
- Antioxidant Vitamins
- LNAA supplementation
- More aggressive Organ Transplantation

Food Groups of Metabolic Disease



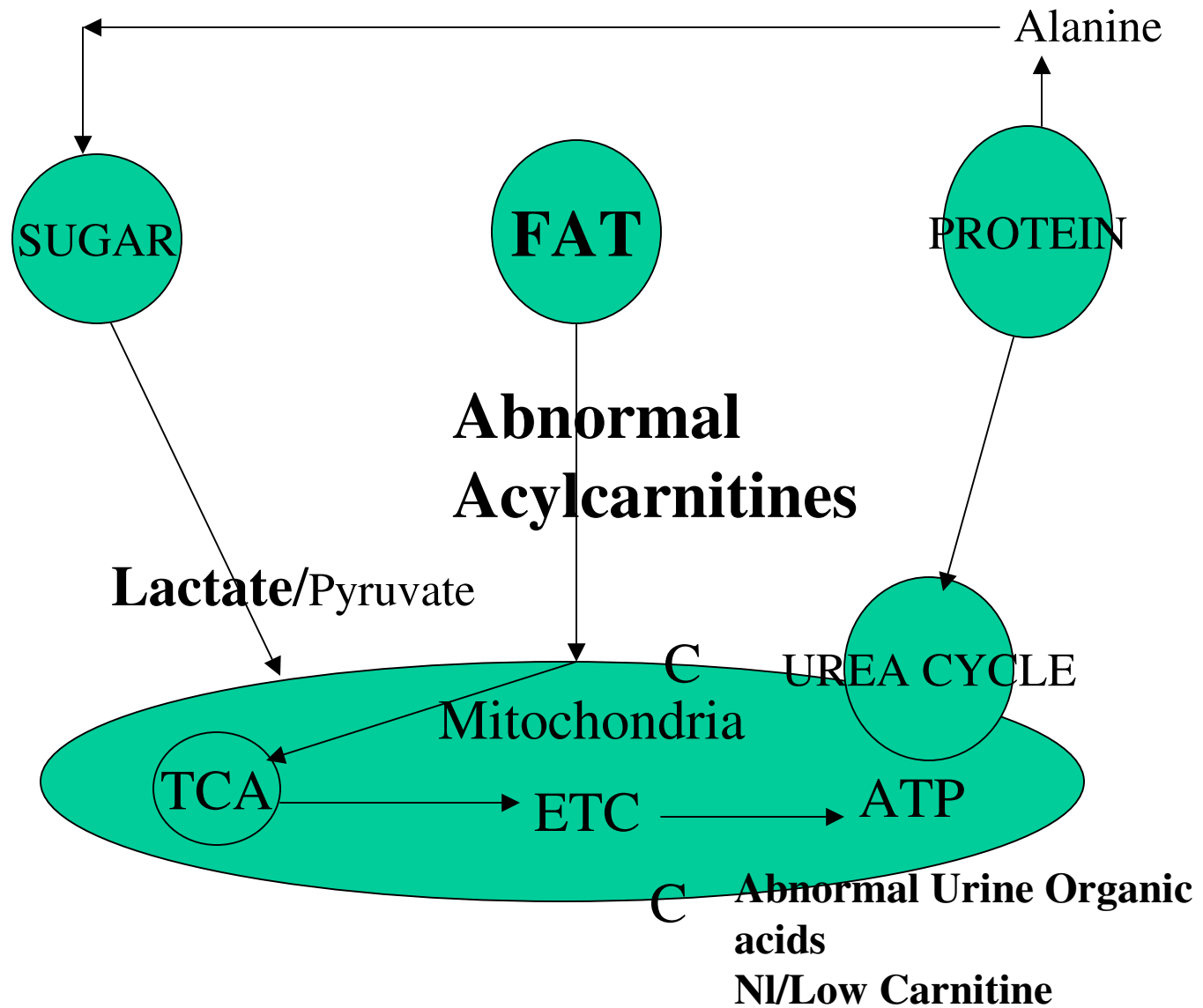
Current Treatment

- Strict, very low protein diet
- Single Amino acid supplements
- Sodium Phenylbutyrate/Acetate/Benzoate
- +/- B Vitamins
- +/- Carnitine

? Potential New Treatments

- IV Phenylbutyrate/acetate
- Gene therapy

Food Groups of Metabolic Disease



Current Treatment

- Avoid Fasting
- Low Fat diet, provide extra calories with Protein and Carbohydrates
- MCT Oil in long chain defects
- +/-Essential Fatty acid supplements
- +/- DHA
- +/- Riboflavin
- +/- Carnitine
- Avoid Toxins (Depakote etc...)

? Potential New Treatments

- Creatine
- CoQ
- Antioxidant Vitamins

Other supplements ?

- Cornstarch
- Glutamine
- Inulin (FOS fiber)
- Antibiotics
- Senna
- Periactin (Cyproheptadine)
- Zofran (Ondansetron)

QUESTIONS ?????

THANK YOU