

Fueling our Muscles

Carbohydrates ~ Protein ~ Healthy Fats



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What happens when we

EAT





https://www.slideshare.net/RozPaws/ketogenic-diets



https://opentextbc.ca/anatomyandphysiology/chapter/24-1-overview-ofmetabolic-reactions/



Carbohydrate (сно)

Fruit, Vegetables, Grains, Beans



Carbohydrates \rightarrow glucose

- There are different types of CHO simple/complex/refined/unrefined
- When we eat CHO, our bodies break them down into simple sugars, which are absorbed in the bloodstream.
- As the sugar level rises in our bloodstream, the pancreas releases a hormone called insulin
- Insulin helps to regulate blood sugar levels by stimulating muscle, fat, and liver cells to absorb glucose

GLYCOLYSIS – breakdown of glucose \rightarrow ATP



Carbohydrates

- If we eat more CHO then our bodies require, the body stores them in the liver and muscle (glycogen) and coverts the rest to fat
- However, in GSDV, CHO metabolism is impaired
 - We cannot access MUSCLE GLYCOGEN (80% of stored CHO)
 - Muscle glycogen stores are always full, therefore excess CHO will be stored as fat

GLYCOGENOLYSIS – breakdown of glycogen to glucose \rightarrow ATP





However, it is important to remember that glucose metabolism is not completely impaired in GSDV. LIVER GLYCOGEN (AEROBIC) BLOOD GLUCOSE (AEROBIC) MUSCLE GLYCOGEN (ANAEROBIC)





Meat, Dairy, Nuts, Eggs, Fish



Protein \rightarrow amino acids

- 20 amino acids ~ 9 of which are essential & must be consumed in the diet
- Protein is a building block to build, strengthen, and repair the body and generally not used for energy
- However if we do not get enough calories from other nutrients, protein can be used for energy

GLUCONEOGENESIS – generation of glucose from non CHO substrates \rightarrow ATP



Healthy Fats

Oils, Nuts, Dairy, Meat, Olives



HEALTHY FATS \rightarrow FATTY ACIDS AND GLYCEROL

- Fats are complex molecules composed of fatty acids and glycerol
- The body needs fats for growth and energy
- Fats are also used to synthesize hormones and other substances
- Fats are the SLOWEST source of energy, but the MOST energy efficient
 - Each gram of fat supplies the body with 9 calories of energy; more than twice supplied by protein or CHO (4 calories)



HEALTHY FATS \rightarrow FATTY ACIDS AND GLYCEROL

LIPOLYSIS – Triglycerides must be first broken down into fatty acids and glycerol FATTY ACID OXIDATION OR β -OXIDATION

- Fatty acids are oxidized into acetyl CoA -> ATP
- Glycerol enters glycolysis pathway -> ATP
- KETOGENESIS Formation of ketone bodies in the liver in response to low blood glucose \rightarrow ATP

KETOLYSIS – Utilization of ketone bodies (muscle, heart, brain)



Healthy Fats \rightarrow fatty acids and glycerol





High CHO **High Protein** Low Carb Ketogenic Diet Low CHO



Composition C~F~P	High CHO 65%~20%~15%	High Protein 43%~29%~ <mark>28%</mark>	High Fat 10%~70%~20%
Overview	 -Constant day-time supply of blood glucose -Simple CHO are quickly digested -> rapid energy -Top up liver glycogen -37g sucrose 5 min before exercise 	-Repair muscle cells -Amino Acids can be used as energy	-Up-regulate fat metabolism -Improve activity tolerance* -Minimize risk of secondary conditions associated with dietary sugar* -10%/75%/15% -> .5mmol/L
Considerations	 -Risks associated with (too much) dietary sugar – obesity, heart disease, diabetes, cancer, etc. Insulin prevents fat breakdown Not practical for ADLs 	-Other health conditions	-Other health conditions -Diet compliance
Research	Nogales-Gadea et al (2015) Quinlivan et al (2014) Andersen & Vissing (2008) Lucia et al (2008) Andersen (2008)	Quinlivan et al (2014) Maclean (1998) Kushner (1990) Jensen et al (1990) Slonim & Groans (1985)	Løkken et al (2019) Reason et al (2017)* Orngreen et al (2009) Andersen et al (2009) Vorgerd & Zange (2007)
Limitations	-Small sample size	-Single case studies -Not blinded	-Anecdotal* -Clinical trials underway





Anecdotal reports of individuals with McArdle disease following a Low CHO diet to achieve improved activity tolerance



Is one MACRONUTRITIENT better able to fuel McArdle muscle?



It may depend...



- Genetics disease causing/modifying mutations
- Physical Activity (*ADLs) vs. Exercise
- Access to Care/Information

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- ☞ Lifestyle Active vs. Sedentary
- Aerobic Capacity ability of heart and lungs to get oxygen to muscles

? CONDITIONED vs DE-CONDITIONED

- Diet composition/satisfaction/compliance
- Comorbidities other health conditions
- Environment ambient temperature/wind/terrain



At present, there is not enough CLINICAL evidence to support a specific nutrition management strategy (Quinlivan et al, 2014)



AEROBIC CAPACITY

is the primary modifier for disease severity (perhaps) independent of which nutrition management strategy is followed.





Food for Thought

- Eat whole foods
- Choose healthy Fats (nuts, avocados, olive oil, eggs, cheese)
- Stick to complex/unrefined CHO (vegetables, grains, fruit)
- Eat to satiation
- Do not mix high fat with high CHO (processed foods)
- Consult your physician
- > 37g sucrose 5 min pre-exercise (bi-weekly)
 - Equal to a can of Coke or 9 ¼ teaspoons of sugar
- Stay hydrated





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