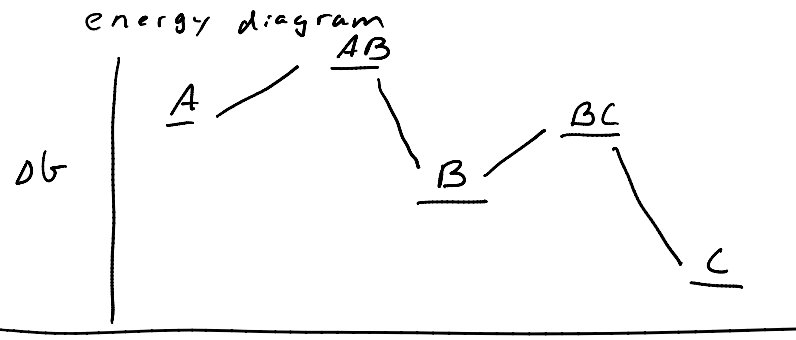
Biochemistry 7

- iClicker 19A
- Metabolism
  - PKU and why it is recessive
  - Pathways
- iClicker 19B

- Due in Lab next week
  - Pre-Lab 8
  - GFP Lab Report
  -
- Register your iClicker
-

Summary

A -> B -> C



Metabolism = Sum of all enzymes & reactions in the body

ex. protein metabolism

(phenylketonuria PKU – genetic disease)

autosomal rec.

protein in food —digestive enzymes→ amino acids → processed in liver

(break peptide bonds do not break down to

extra a.a. → a.a. to be used

(do not break down to individual atoms)

a.a. → urine

a.a. → blood → cells

cells use amino acids to build new protein

adult male ~ 400g/day of protein

eats ~150g/day of protein

reuses ~250 g/day → turnover

Phenylalanine (phe) = amino acid
- humans can't make phe from other amino acids

- normal protein diet 2g phe/day
- your body only needs 0.5g phe/day to make proteins
- 1.5g phe must be degraded per day
  - done in liver by enzymatic pathways

degradation begins with phenylalanine hydroxylase (PAH)
↳ enzyme

$$\text{phe} + O_2 + \text{stuff} \xrightarrow[\text{enzyme}]{PAH} \text{tyrosine} + H_2O \xrightarrow[\text{enzymes}]{\text{other}} \cdots \rightarrow \text{urine}$$

Notes:
- not enough phe in your diet → death
- too much phe → brain damage

∴ phe levels must be carefully regulated

PKU - inherited intolerance of phe (auto. rec. disease)

allele	contribution to phenotype	PAH enzyme encoded by the allele
D	normal (dom)	functional PAH
d	PKU (rec)	non-functional PAH

why is PKU recessive?

genotype	PAH enzyme	phenotype
DD	functional - 100%	normal
d/d	non-functional - 100%	PKU → can't degrade

dd non-functional - 100% PKU → can't degrade phe ∴ phe builds up → brain damage

Dd → functional 50% normal
→ non-functional 50%

why? - 50% normal PAH is enough to degrade phe in a normal diet
- enzymes are very effective catalysts
- "doing something (breaking down phe) is dominant"

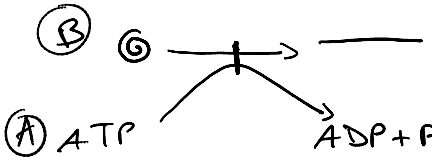
Energy - enzymes can speedup a reaction if it is ΔG−
what if a cell needs to do a ΔG+ reaction?
→ add chemical energy (not heat) from a reaction with big ΔG−

Reaction Coupling

snap bracelet example: - Kinking the snap bracelet is Ea
- unrolling the snap bracelet is ΔG+

energy in cells comes from ATP

$ATP \rightarrow ADP + P_i \longrightarrow \Delta G-$ Huge



ΔG+
ΔG−

both reactions happen together
energy from A drives B