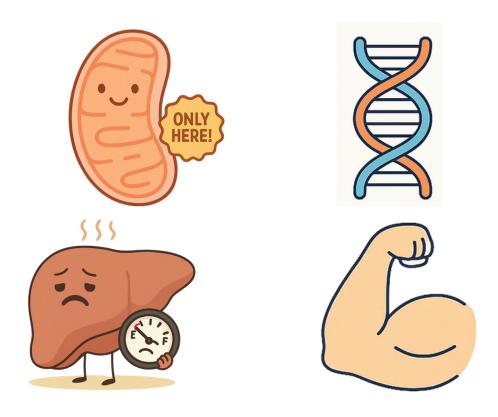


### 25 Biochemistry Fast Facts to Smash Step 1

1<sup>st</sup> Edition

(Memorize Smarter, Score Higher.)

Essential high-yield facts Step 1 students can't afford to miss.



**Helping Elevate Your Clinical Mastery** 

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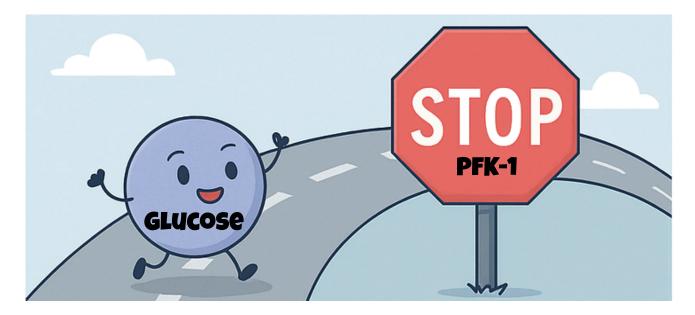


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# Fast Fact # 1: Rate-Limiting Step of Glycolysis

The step you MUST know cold for Step 1.



Phosphofructokinase-1 (PFK-1) is the ratelimiting enzyme in glycolysis.

### Why it Matters:

- PFK-1 controls the pace of glycolysis no PFK-1 = no go.
- Highly regulated by ATP (inhibits) and AMP (activates).
- Major USMLE favorite: expect questions involving low energy states (AMP upregulates).

#### **Pro Tip:**

If a question mentions energy control and glycolysis, think: PFK-1 is the boss!

### **Quick Memory Hook:**

Glucose can sprint... but PFK-1 decides when to hit the brakes.

# Fast Fact #2: Pyruvate Kinase Deficiency

The key cause of hemolytic anemia is tied to energy failure.



Pyruvate kinase deficiency leads to hemolytic anemia.

### Why it Matters:

- Without pyruvate kinase, RBCs can't maintain Na+/K+ pumps, leading to hemolysis.
- No ATP → cell death.

#### **Pro Tip:**

Look for signs of newborn jaundice, anemia, and reticulocytosis.

### **Quick Memory Hook:**

RBCs without fuel = pop and drop.

# Fast Fact #3: Only Gluconeogenesis Enzyme in Mitochondria

Know where pyruvate carboxylase lives — Step 1 loves this.



Pyruvate carboxylase is the only gluconeogenesis enzyme in mitochondria.

### Why it Matters:

- Converts pyruvate to oxaloacetate inside mitochondria.
- First step in gluconeogenesis.

#### **Pro Tip:**

Always connect pyruvate carboxylase to mitochondria — easy points!

### **Quick Memory Hook:**

Mitochondria: the official headquarters of pyruvate carboxylase.

# Fast Fact #4: Hereditary Fructose Intolerance

Common cause of hypoglycemia and vomiting in infants.



Aldolase B deficiency leads to hereditary fructose intolerance.

### Why it Matters:

- Fructose-1-phosphate builds up → hypoglycemia.
- Symptoms after eating fruits or juices.

### **Pro Tip:**

Any vomiting after fruits? Think fructose metabolism issues first.

### **Quick Memory Hook:**

Fruit makes baby puke? Blame Aldolase B.

# Fast Fact #5: Classic Galactosemia

Recognize galactosemia signs early — high-yield diagnosis



GALT deficiency causes classic galactosemia.

### Why it Matters:

- Early milk ingestion → vomiting, cataracts, hepatomegaly.
- Fatal if untreated.

### **Pro Tip:**

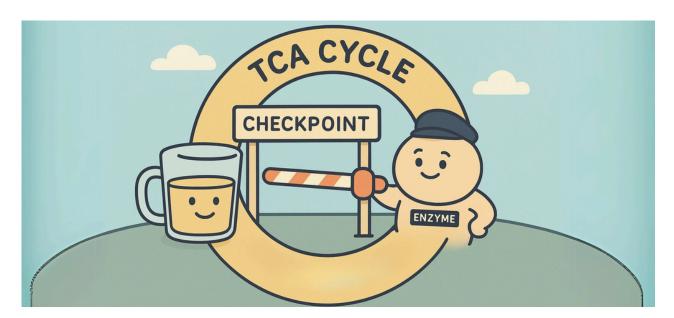
Check for cataracts and liver dysfunction after feeding in neonates.

### **Quick Memory Hook:**

Galactose turns milk sweet... and dangerous without GALT.

# Fast Fact #6: Rate-Limiting Step of TCA Cycle

Know the control point for cellular respiration.



Isocitrate dehydrogenase is the rate-limiting enzyme of the TCA cycle.

### Why it Matters:

- Regulates pace of TCA cycle energy production.
- Activated by ADP, inhibited by ATP and NADH.

### **Pro Tip:**

If ATP is high, TCA slows; if ADP is high, TCA races!

### **Quick Memory Hook:**

Isocitrate = checkpoint traffic cop of the TCA!

### Fast Fact #7:

### Pathway Requiring Thiamine (Vitamin B1)

Memorize the B1 pathways! Step 1 high-yield.



Thiamine (Vitamin B1) is required for the pyruvate dehydrogenase complex.

### Why it Matters:

- Needed for decarboxylation reactions.
- Deficiency leads to lactic acidosis.

#### **Pro Tip:**

Always associate B1 with pyruvate dehydrogenase and  $\alpha$ -ketoglutarate dehydrogenase.

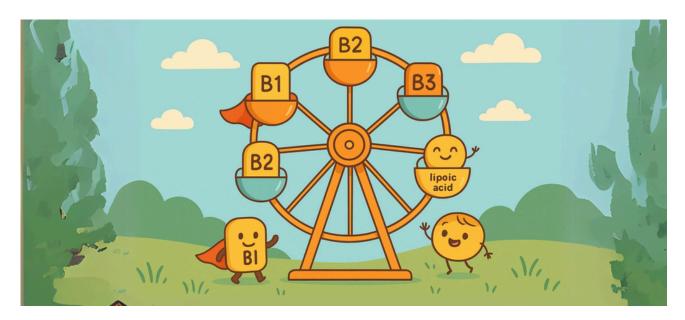
### **Quick Memory Hook:**

B1: The cape-wearing superhero of metabolism.

### Fast Fact #8:

### Cofactors for α-Ketoglutarate Dehydrogenase

TCA cycle essentials you need to know cold.



α-Ketoglutarate dehydrogenase requires B1, B2, B3, B5, and lipoic acid.

#### Why it Matters:

- Major Step 1 trap: missing any one cofactor derails TCA cycle.
- Especially important in thiamine deficiency (Wernicke-Korsakoff).

### **Pro Tip:**

Remember: Cofactors spin the TCA wheel smoothly!

### **Quick Memory Hook:**

No passengers, no spin — cofactors make TCA turn.

# Fast Fact #9: Main Enzyme Deficiency in PKU

The classic inborn error — Step 1 favorite.



Phenylalanine hydroxylase deficiency causes classic PKU.

### Why it Matters:

- Accumulation of phenylalanine leads to this high-yield triad: intellectual disability, musty odor, and eczema.
- Treatment: low-phenylalanine diet.

### **Pro Tip:**

Musty odor + developmental delay? Always think PKU!

### **Quick Memory Hook:**

Broken phenylalanine bridge leads to brain fog.

### Fast Fact #10:

### **Enzyme Deficiency in Tay-Sachs Disease**

Classic presentation: cherry-red macula with progressive neurodegeneration and **no**hepatosplenomegaly



Hexosaminidase A deficiency causes Tay-Sachs disease.

### Why it Matters:

- Accumulation of GM2 ganglioside.
- Neurodegeneration, developmental delay, exaggerated startle reflex.

### **Pro Tip:**

Cherry-red macula + no hepatosplenomegaly = Tay-Sachs!

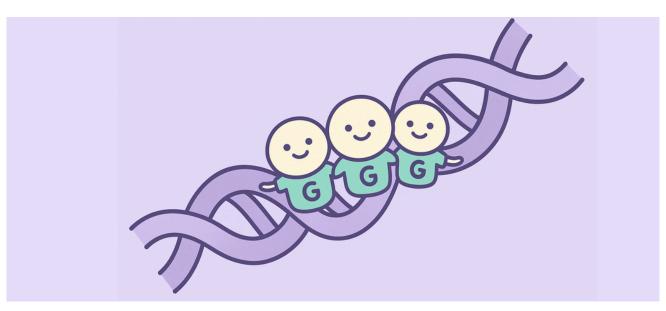
### **Quick Memory Hook:**

Tay-Sachs stacks GM2 until neurons collapse.

### Fast Fact #11:

### Key Amino Acid for Collagen Synthesis

Know your collagen components — easy Step 1 points.



Glycine is the most abundant amino acid in collagen.

### Why it Matters:

- Essential for collagen's tight triple helix formation.
- Deficiency affects structural stability (e.g., scurvy, Ehlers-Danlos).

#### **Pro Tip:**

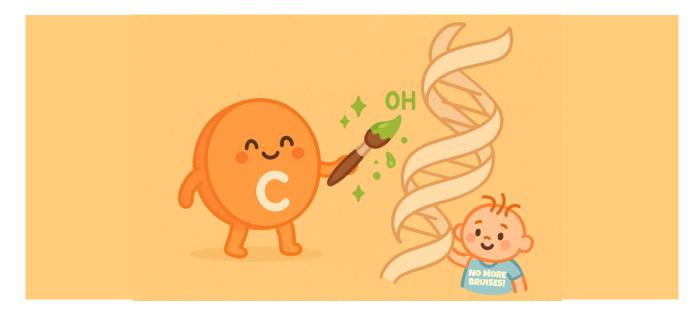
Glycine makes collagen twist tight and strong!

### **Quick Memory Hook:**

Without glycine, collagen unravels like cheap yarn.

# Fast Fact #12: Vitamin C Role in Collagen Synthesis

Vitamin C = strong connective tissues.



Vitamin C is required for hydroxylation of proline and lysine residues in collagen.

### Why it Matters:

- Hydroxylation stabilizes collagen fibers.
- Deficiency = Scurvy (easy bruising, gum bleeding).

### **Pro Tip:**

Vitamin C paints strength onto collagen!

### **Quick Memory Hook:**

No C, no sturdy sea ropes (collagen collapses).

### Fast Fact #13: Enzyme Deficient in Alkaptonuria

Easy black-pigment clue on Step 1.



Alkaptonuria is caused by homogentisate 1,2-dioxygenase deficiency.

### Why it Matters:

- Black pigment deposits in connective tissues (ochronosis).
- Urine darkens on standing.

### **Pro Tip:**

Black urine + joint pain in an adult = Alkaptonuria!

### **Quick Memory Hook:**

Spill black ink? Think Alkaptonuria sink!

# Fast Fact #14: Maple Syrup Urine Disease (MSUD)

Think syrupy mess in branched chains.



Branched-chain α-ketoacid dehydrogenase deficiency causes MSUD.

### Why it Matters:

- Accumulation of branched-chain amino acids.
- Sweet-smelling urine, lethargy, poor feeding.

### **Pro Tip:**

Sweet-smelling baby? Suspect MSUD immediately!

### **Quick Memory Hook:**

Maple syrup flows until enzymes clog the branching trees!

# Fast Fact #15: Homocystinuria

The marfanoid mimic with a deadly twist.



Cystathionine β-synthase deficiency causes homocystinuria.

### Why it Matters:

- Marfanoid habitus, lens subluxation (downward), thrombotic events.
- High risk for vascular accidents.

### **Pro Tip:**

Tall, thin, dislocated lens? Always screen for thrombosis = Homocystinuria.

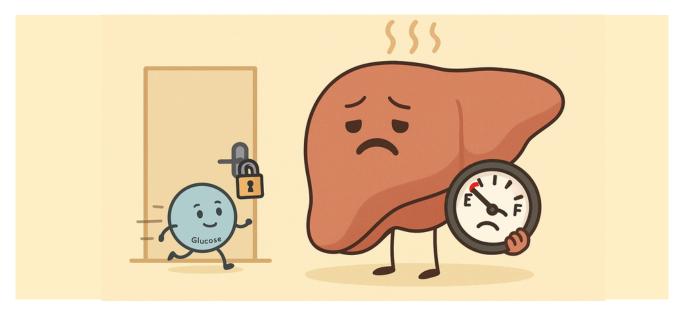
### **Quick Memory Hook:**

Broken sulfur bridge = clot bridge collapse!

### Fast Fact #16:

### Von Gierke Disease (Type I Glycogen Storage)

Glycogen stuck = major hypoglycemia.



Glucose-6-phosphatase deficiency causes
Von Gierke disease.

### Why it Matters:

- Severe fasting hypoglycemia, hepatomegaly, lactic acidosis.
- Can't release free glucose into bloodstream.

### **Pro Tip:**

If you see fasting hypoglycemia + big liver, think Von Gierke!

### **Quick Memory Hook:**

Liver has glucose... but no exit door without glucose-6-phosphatase.

### Fast Fact #17:

### Pompe Disease (Type II Glycogen Storage)

Big heart failure in infants = major clue.



Lysosomal α-1,4-glucosidase deficiency causes Pompe disease.

### Why it Matters:

- Cardiomegaly, hypotonia, early death.
- Glycogen accumulates in lysosomes.

### **Pro Tip:**

Pompe pumps the heart — when it fails, the heart fails too.

### **Quick Memory Hook:**

Pompe packs glycogen until the heart collapses.

### Fast Fact #18:

### McArdle Disease (Type V Glycogen Storage)

Muscle cramps = glycogen breakdown failure.



Muscle phosphorylase deficiency causes

McArdle disease.

### Why it Matters:

- Painful muscle cramps, myoglobinuria after exercise.
- Blood glucose levels typically normal.

### **Pro Tip:**

If cramps + dark urine after exercise = suspect McArdle!

### **Quick Memory Hook:**

Muscles try to run... but crash without energy fuel.

## Fast Fact #19: Most Abundant Amino Acid in Blood

Most Abundant Amino Acid in Blood



Glutamine is the most abundant amino acid in the blood.

### Why it Matters:

- Important nitrogen carrier, buffer in acid-base balance.
- Critical for rapidly dividing cells.

### **Pro Tip:**

Think of glutamine as the river taxi for nitrogen transport!

### **Quick Memory Hook:**

Glutamine keeps the bloodstream flowing strong.

### Fast Fact #20:

### **Enzyme Deficient in Lesch-Nyhan Syndrome**

Purine salvage system meltdown = an exam favorite.



Lesch-Nyhan syndrome is caused by hypoxanthine-guanine phosphoribosyltransferase (HGPRT) deficiency.

### Why it Matters:

- Hyperuricemia, self-mutilation, gout, intellectual disability.
- X-linked recessive inheritance.

#### **Pro Tip:**

If you see self-mutilation + gouty symptoms in a boy = Lesch-Nyhan!

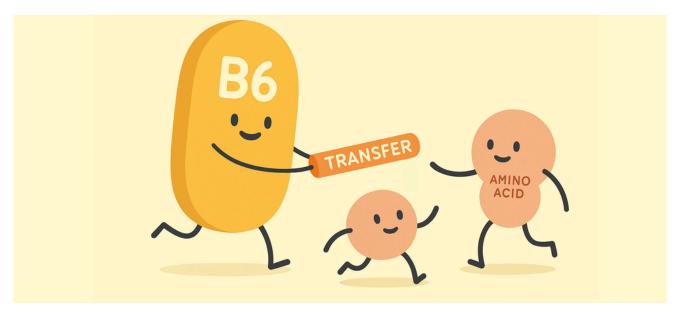
### **Quick Memory Hook:**

INo HGPRT = Purine castle collapse and chaos!

### Fast Fact #21:

### Vitamin Needed for Transamination Reactions

B6 = MVP of amino group transfers.



Pyridoxine (Vitamin B6) is needed for transamination reactions.

### Why it Matters:

- Transfers amino groups between molecules.
- Required for amino acid metabolism.

### **Pro Tip:**

No B6 = no handoff = no amino metabolism!

### **Quick Memory Hook:**

B6 passes the amino baton to keep metabolism racing.

### Fast Fact 22:

### Classic Finding in Biotin (Vitamin B7) Deficiency

Watch for hair loss clues — easy Step 1 point.



Biotin deficiency causes dermatitis, alopecia, and enteritis.

### Why it Matters:

- Seen with raw egg white ingestion (avidin binds biotin).
- Also caused by long-term antibiotic use.

### **Pro Tip:**

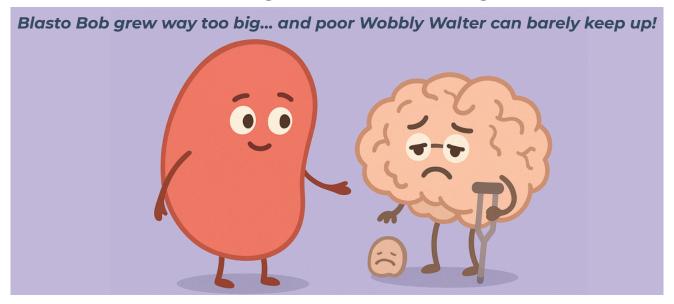
If hair falls out + stomach aches = suspect biotin deficiency!

### **Quick Memory Hook:**

No B7 = hair falls, gut stalls.

# Fast Fact #23: Vitamin Causing Megaloblastic Anemia and Neuro Symptoms

B12: Big cells + brain fog.



Vitamin B12 deficiency causes megaloblastic anemia and neurologic symptoms (e.g., paresthesia, ataxia), whereas folate deficiency causes only megaloblastic anemia without neurologic symptoms.

### Why it Matters:

- Impaired DNA synthesis → big RBCs.
- Demyelination → neuro deficits (paresthesia, ataxia).

### **Pro Tip:**

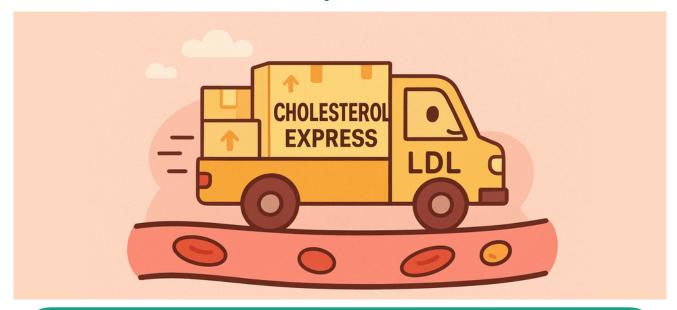
If anemia + weird neuro signs = B12 deficiency over folate!

### **Quick Memory Hook:**

Big cells, foggy brain? Blame B12 drain.

# Fast Fact #24: Primary Carrier of Cholesterol in Blood

LDL = delivery truck of doom.



tissues; oxidized LDL is taken up by macrophages, leading to foam cell formation and atherosclerotic plaque development.

### Why it Matters:

- High LDL = risk factor for atherosclerosis.
- Target of statin therapy.

#### **Pro Tip:**

LDL drops off cholesterol wherever it pleases — and wrecks arteries.

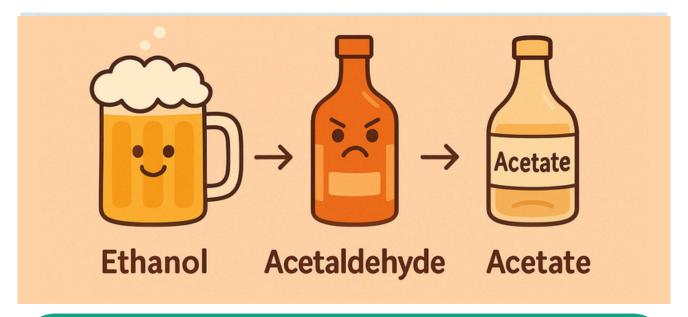
### **Quick Memory Hook:**

LDL: Little Delivery of Lipid (into artery walls)!

### Fast Fact 25:

### Basic Principle of Ethanol Metabolism

Alcohol journey: happy → toxic → tired.



Ethanol → Acetaldehyde (via alcohol dehydrogenase) → Acetate (via acetaldehyde dehydrogenase).

### Why it Matters:

- Acetaldehyde is toxic = hangover symptoms.
- Disulfiram inhibits acetaldehyde dehydrogenase (worse symptoms).

### **Pro Tip:**

If you block acetaldehyde breakdown, you get instant hangover horror.

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### **Quick Memory Hook:**

Happy to toxic to tired: the 3-act alcohol play!

### Thank You

