

KEY WORDS glycogen

Separate the two glycogen banks, count gluconeogenesis costs, and read a storage-disease vignette.

Liver ~100 g EXPORTS (has glucose-6-phosphatase); muscle ~400 g KEEPS (lacks it). Insulin stores both; glucagon opens only the liver.

Liver bank 100 g $\approx$ 400 kcal vs brain $\sim$ 480 kcal/day – about one day, then gluconeogenesis (cost 4 ATP + 2 GTP + 2 NADH per glucose). ✓

2.3 Gluconeogenesis from 2 pyruvate: count the high-energy phosphates spent. [3]

Predict the fuel sequence hour by hour. Compute how long liver glycogen alone covers the brain. State the three gluconeogenic substrates and their origins. Explain why prolonged fasting spares muscle protein. [8]

Localise the defect. Explain the absent lactate. Distinguish from a PFK-1 deficiency using the same test. State the long-term exercise advice implied by the mechanism. [7]

Solutions

2.1

- Muscle lacks glucose-6-phosphatase, so G-6-P cannot become free glucose for export.

2.2

- Glucagon; liver only (no muscle receptors).

2.3

- $4 \text{ ATP} + 2 \text{ GTP} = 6 \text{ high-energy phosphates (plus 2 NADH)}$.

3.1

- Hours 0–4 absorbed fuels; 4–12 hepatic glycogenolysis; 12–24 gluconeogenesis dominant; day 3+ ketones cover $\sim 2/3$ brain needs.
- $100 \text{ g} \div \sim 5 \text{ g/h} \approx \text{up to 24 h}$; strictly, glycogen alone ≈ 1 day of brain use (400 vs 480 kcal).
- Substrates: alanine (muscle), glycerol (fat), lactate (Cori).
- Ketones substitute for glucose $\rightarrow$ less gluconeogenesis $\rightarrow$ protein sparing.

3.2

- Muscle glycogen phosphorylase.
- No glycogen $\rightarrow$ no extra G-6-P flux $\rightarrow$ no pyruvate surge $\rightarrow$ no lactate.
- PFK-1 deficiency: glycogen BREAKS down but flux dies at the gate – also low lactate, but with phosphate intermediates accumulating.
- Advice: aerobic, low-intensity exercise (fat/fatty acids still usable); avoid sprints.