



Anesthetic Management of Patient with Long-Chain 3-hydroxyacyl CoA Dehydrogenase Deficiency

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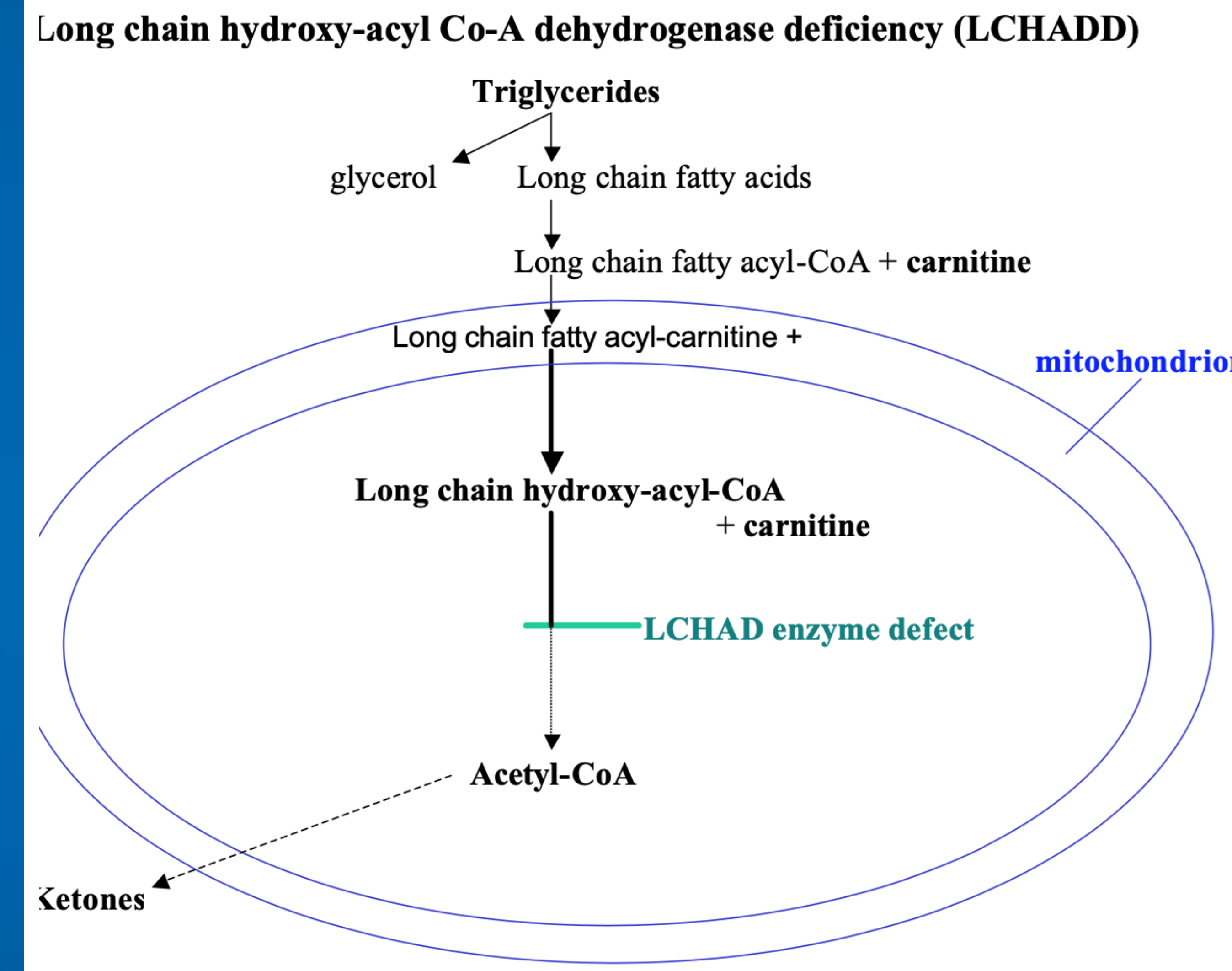


Case Description

A 16 year old female with LCHAD presented for medial patellofemoral ligament reconstruction. It was noted that patient has previously experienced difficulties with anesthesia, including high anesthetic requirements, difficult emergence, PONV, and hypoglycemia. Additionally, she was unable to receive propofol due to presence of long-chain fatty acids in the medication. It was decided to proceed with regional anesthesia, consisting of a single shot spinal, light sedation with midazolam and dexmedetomidine, and a postoperative femoral nerve block. She was monitored in the PACU and discharged safely home after 4 hours.

Discussion

LCHAD is one variant of several rare fatty acid oxidation disorders, which involves inborn errors of metabolism, categorized by length of fatty acid chain. These errors affect mitochondrial oxidation and transport of fatty acids into the mitochondria.³ Clinical presentations can vary in severity by phenotype, but common symptoms include hypoglycemia, peripheral neuropathy, myopathies, as well as cardiomyopathy and encephalopathy in severe cases.² Symptoms can be exacerbated by stress, prolonged fasting periods, instances of increased energy demands, illness and infection. This is due to an inability to use the alternative energy source of fatty acid oxidation in times of low glucose levels.¹ Surgery is a large stressful event on the body with required fasting periods putting these patients at increased risk of perioperative complications.



- The body's primary source of fuel is glucose.
- When glucose levels are low, the body uses fatty acid metabolism to produce ketones for energy
- Long chain fatty acid oxidation disorders are caused by defects in the metabolic pathway that converts fatty acids into energy, causing deficiency in mitochondrial energy production.⁴

Conclusion

Regional anesthesia can be a great option for LCHAD patients; however, patient selection is critical. Patients and guardians must be open to discussions about alternative anesthesia plans and how this will affect the overall surgical experience. The goal is to maintain a low stress environment to prevent adverse outcomes in this patient population. Luckily, the family in this case was extremely motivated to pursue options outside of general anesthesia, which also contributed to a successful outcome for the patient.

References

1. C. Peixoto, C. Dourado, H. Santos, P. Roberto, Perioperative anesthetic management of very long-chain acyl-coenzyme a dehydrogenase deficiency, *Revista Española de Anestesiología y Reanimación* (English Edition), Volume 70, Issue 4, 2023, Pages 231-234
2. Long-Chain 3-Hydroxyacyl-CoA Dehydrogenase Deficiency (LCHADD) | Syndromes: Rapid Recognition and Perioperative Implications, 2e | AccessAnesthesiology | McGraw Hill Medical (
3. Yuasa, H., Onoda, Y., Kitaura, A. *et al.* Anesthesia management in a patient with very long-chain acyl-Coenzyme A dehydrogenase deficiency. *JA Clin Rep* 6, 72 (2020).
4. Jerry Vockley, Deborah Marsden, Elizabeth McCracken, Stephanie DeWard, Amanda Barone, Kristen Hsu, Emil Kakkis, Long-term major clinical outcomes in patients with long chain fatty acid oxidation disorders before and after transition to triheptanoin treatment—A retrospective chart review, *Molecular Genetics and Metabolism*, Volume 116, Issues 1–2, 2015, Pages 53-60.
5. *Long chain hydroxy acyl-CoA dehydrogenase deficiency (LCHADD)*. New England Consortium of Metabolic Programs. (n.d.). <https://www.newenglandconsortium.org/lchadd>