Abetalipoproteinemia and Related Disorders Foundation (ABL + Foundation)

Template Letter of Medical Necessity for VITAMIN K Coverage

Patients with abetalipoproteinemia and other familial hypobetalipoproteinemia disorders possess gene mutations required to package lipids into particles for absorption and transportation throughout the body. Fat-soluble vitamins, including vitamins E, A and K are also dependent on these pathways for absorption. Consequently, patients develop severe deficiencies of these vitamins and the clinical outcomes can be catastrophic, including debilitating neurologic impairment, blindness and life-threatening bleeding.

Vitamin K is a cofactor in the production of several active clotting factors by the liver that are essential for proper hemostasis. **Vitamin K deficiency** may result in severe and lethal bleeding. Of particular concern, internal bleeding including intracranial hemorrhage, may occur spontaneously or from relatively minor injuries. High-dose (5-35mg/week) oral supplementation with vitamin K preparations is required in patients with abetalipoproteinemia and other hypobetalipoproteinemia disorders to prevent lethal complications (1, 2).

For patients with abetalipoproteinemia and other hypobetalipoproteinemia disorders, vitamin K is not a micronutrient supplement, instead an essential therapeutic treatment.

As the ABLRDF Medical Advisory Panel, our members include established researchers and medical professionals who care for patients with these disorders. We attest that a high dose of vitamin K taken lifelong is medically necessary to prevent devastating complications in patients with abetalipoproteinemia and other hypobetalipoproteinemia disorders.

Sincerely,

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1. Zamel R, Khan R, Pollex RL, and Hegele RA. Abetalipoproteinemia: two case reports and literature review. *Orphanet J Rare Dis.* 2008;3:19.
2. Bredefeld C, Hussain MM, Averna M, Black DD, Brin MF, Burnett JR, et al. Guidance for the diagnosis and treatment of hypolipidemia disorders. *J Clin Lipidol.* 2022.