Q: Interesting data! Does the alterations in circulating amino acids and improved skeletal muscle force indicate that ELAM improves skeletal muscle energy status and size (less atrophy)?

A: Yes, the metabolic observations from TAZPOWER BTHS patients are consistent with improved skeletal muscle energy status after 12 weeks of elamipretide (ELAM) therapy. Muscle size was not evaluated in TAZPOWER, although improved mitochondrial function is expected to attenuate atrophy and promote beneficial physiological remodeling of muscle fibers over time. These benefits were previously seen with elamipretide in several different pre-clinical studies.

In rodent models of disuse atrophy, elamipretide treatment preserved force generation and muscle cross-sectional area following either ventilator- or hindlimb cast-induced atrophy1,2. These improvements were associated with significantly reduced markers of apoptotic cell death, such as caspase-3, and lower oxidative stress. Pathological skeletal muscle remodeling in cardiomyopathy was also reversed with long-term (90 days) elamipretide treatment3. In this study, the significant improvement in skeletal muscle mitochondrial energetics was accompanied by beneficial remodeling in muscles (restoration of Type 1, oxidative fibers after elamipretide treatment in the myopathic dogs).

References:
1Powers et al. Critical Care Med 2011: Reference
2Min et al. J Applied Physiol 2011: Reference
3Sabbah et al. ESC Heart Failure 2019: Reference