Mitochondria are responsible for many essential cellular processes, including respiration, and mitochondrial dysfunction is linked to multiple human diseases including myopathies, cancer, Alzheimer’s and Parkinson’s diseases. Proper regulation of mitochondrial biogenesis is critical to preserve mitochondrial function. The MTERF protein family plays a crucial role in this process. Initially thought to solely regulate mitochondrial gene transcription, these proteins are now understood to be versatile nucleic acid binders that influence multiple key steps in biogenesis. MTERF1, the founding member, controls transcription termination. MTERF3 and MTERF4, structurally similar, play distinct but vital roles in assembling mitochondrial ribosomes, essential for protein synthesis within the organelle. Our research sheds light on the mechanisms by which MTERFs operate within mitochondria, particularly the intricate details of mitochondrial ribosome assembly, a process that remains under active investigation.

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