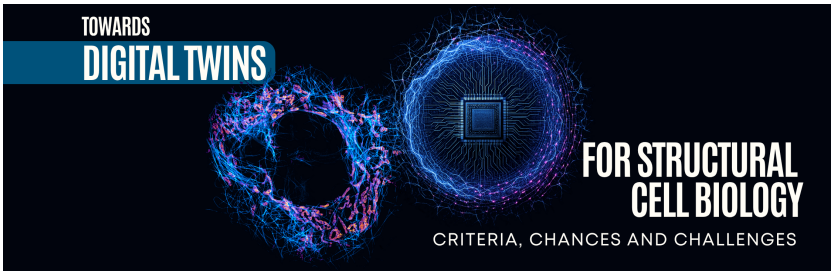

Contribution ID: **60** Type: **not specified**

A paradigm change for cardiolipin remodeling in mitochondria

Cardiolipin (CL) is the signature phospholipid of mitochondria. CL is essential for cristae formation and the functionality of the enzyme complexes involved in biological energy conversion. The mitochondrial transacylase Tafazzin catalyses the obligatory remodeling of CL, a process in which the composition of the four CL acyl chains is altered. Tafazzin dysfunction causes Barth syndrome, a severe multisystem disorder. We combined biochemistry, structure determination, and molecular simulations to identify the mechanism of tafazzin action. Our finding that Coenzyme A is a co-factor set up for direct acyl-chain transfer challenges established views. Moreover, our work provides clues as to how pathogenic mutations impact the function of Tafazzin and thus enables a molecular understanding of Barth syndrome.

Presenter: Prof. ZICKERMANN, Volker (Goethe University Medical School)

Session Classification: POSTER SESSION