The Centre for Blood Research presents

CBR SEMINAR SERIES

Wednesday, February 28, 2024
1:00PM - 2:00PM PT

Life Sciences Centre
1003 (LSC3) & Zoom

"Being at the wrong place at the wrong time - when mitochondrial proteins cannot reach their destination."

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A fundamental process in maintaining mitochondrial functions is the import of proteins into the organelle. This process is typically efficient, however, perturbations in this process can occur under conditions such as oxidative stress, aging, and in diseases such as neurodegenerative diseases. Although the mechanisms governing protein import into mitochondria are well understood, little is known about how defects in this process are sensed and repaired by the cell.

We discovered that protein import defects lead to incomplete translocation of mitochondrial proteins and clogging of the mitochondrial entry channels. We found that cells have evolved to cope with this damage by activating the mitochondrial compromised protein import response (mitoCPR), a transcriptional response, that restores mitochondrial functions during protein import stress. Our research revealed mechanistic details of how mitochondria clogging is prevented and repaired under stress. Moreover, we uncovered a new role for mitochondrial targeting peptides as messengers of mitochondrial dysfunction. This work broadens our understanding of mitochondrial homeostasis and holds the potential for new therapeutic strategies targeting mitochondrial dysfunction in human diseases.

Many thanks to generous support from: