Mitosense LI ™

- Mitosense LI ™ is a highly specific fluorescent iron sensor that readily enters the cells and resides exclusively in the mitochondria by means of mitochondria-homing peptide sequences. It has the ability to sensitively evaluate the mitochondrial labile iron pool by virtue of an attached iron chelating residue that has high specificity and affinity for iron (**Figure 1**).
- Mitosense LI ™ preferentially accumulates in the mitochondria of cells (see example of human primary skin fibroblast in **Figure 2**).
- Loading of cells with iron reduces the fluorescence signal of Mitosense LI ™ and when the added iron is chelated by an iron chelator (e.g. deferiprone in **Figure 3**), the fluorescence is reinstated.
- Mitosense LI TM has been utilised to evaluate the level of mitochondrial labile iron of cultured fibroblasts obtained from Friedreich's ataxia (**FRDA**) patients when compared to skin fibroblasts from healthy donors. (**Figure 4**).

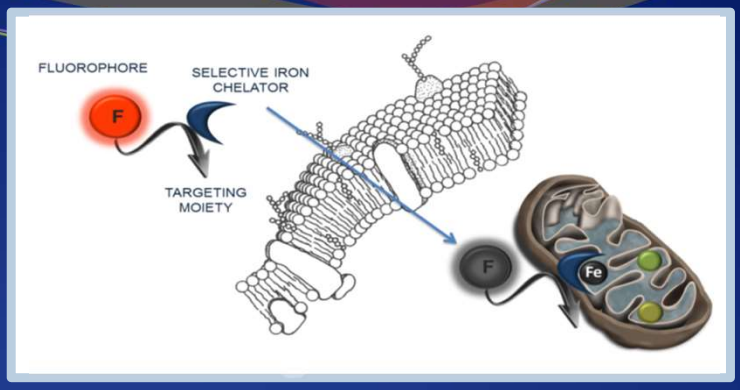

Figure 1. Mitosense LI ™ is a mitochondria-targeted fluorescent iron sensor that looses its fluorescent signal in presence of mitochondrial labile iron.

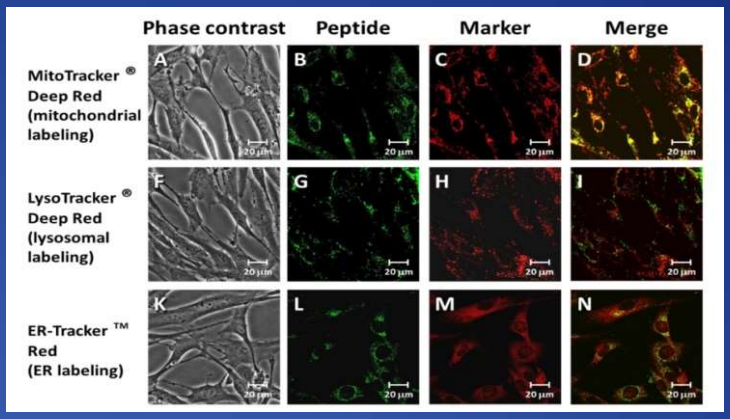

Figure 2. Subcellular distribution of Mitosense LI ™ In human skin fibroblasts with mitochondrial (A–D), lysosomal (F–I) and ER (K–N) compartments.

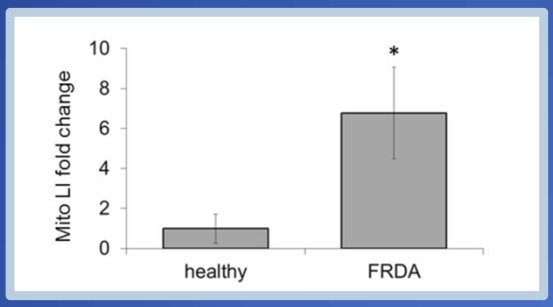

Figure 4. Mitosense LI ™ provided the first quantitative measurement of mitochondrial labile iron from Freidreich's ataxia (FRDA) fibroblasts and demonstrated that FRDA fibroblasts have significantly higher levels of mitochondrial labile iron than healthy fibroblasts.

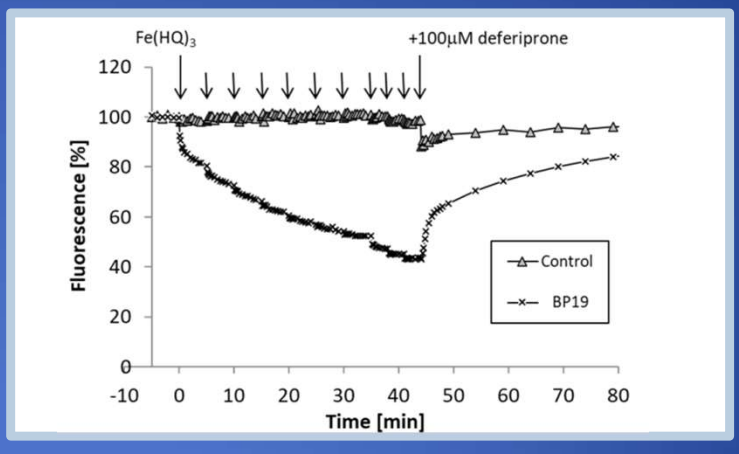

Figure 3. Fluorescence quenching and de-quenching of Mitosense LI ™ in human skin fibroblasts in response to the manipulation of cellular levels of iron. Cells incubated (or not) with the iron sensor were loaded with iron(III) in the form of iron hydroxyquinoline complex, Fe(HQ)3 and then treated with a 100 µM bolus of deferiprone. The arrows illustrate the timepoints at which additional aliquots of Fe(HQ)3 or deferiprone were added.

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