



DIPARTIMENTO DI FARMACIA E BIOTECNOLOGIE

## Life & Chemical Sciences Seminars

# MITOCHONDRIAL COMPLEX III RIESKE Fe-S PROCESSING AND ASSEMBLY

**Dott.ssa Erika Fernandez-Vizarra**

*MRC-Mitochondrial Biology Unit, University of Cambridge, UK*

**Mercoledì 21 marzo ore 14:00** – presso Aula 1 via Belmeloro 6  
(ospite Prof. A.M. Porcelli)

### Abstract

Regulation of the mitochondrial respiratory chain biogenesis is a matter of great interest because of its implications for mitochondrial disease. One of the mitochondrial disease genes recently discovered associated to encephalopathy and mitochondrial complex III (cIII) deficiency is TTC19. Our study of TTC19-deficient models, has led us to propose a post-assembly quality control role or 'husbandry' function for this factor that is linked to Rieske Fe-S protein (UQCRFS1). UQCRFS1 is the last incorporated cIII subunit, and its presence is essential for enzymatic activity. During UQCRFS1 assembly, the precursor is cleaved and its N-terminal mitochondrial targeting peptide (MTS) remains bound to the complex, between the two core subunits (UQCRC1 and UQCRC2). Due to this fact, the 78-amino acids of the N-terminus were considered to be the 11th mammalian cIII structural subunit. However, our data indicate the production of several different size UQCRFS1 N-terminal fragments upon import and assembly. In addition, in the absence of TTC19 there is a prominent accumulation of these UQCRFS1-derived N-terminal fragments that proved to be detrimental for cIII function. These findings open the possibility to discuss some of the pre-conceived ideas around UQCRFS1 processing and assembly and its importance for the regulation of cIII activity and biogenesis.

### Biosketch

I got my degree in Chemical Sciences from the University of Zaragoza, Spain in 1998. After that I joined Prof. Julio Montoya's laboratory in the Department of Biochemistry and Molecular and Cellular Biology of the University of Zaragoza as a graduate student to work on mitochondrial biogenesis under the supervision of Dr. Patricio Fernandez-Silva. In April 2005 I obtained my PhD degree. In May 2005 I joined Dr. Massimo Zeviani's laboratory at the "Fondazione I.R.C.C.S Istituto Neurologico Carlo Besta" in Milan, Italy as a post-doctoral researcher. In 2006 I was awarded a Marie Curie Intra-European Fellowship to develop the "Mitochondrial Respiratory Chain Assembly" project in Dr. Zeviani's research group. In November 2008 I went back to Spain thanks to a "Juan de la Cierva" reintegration contract to join Prof. Jose Antonio Enriquez's group in the Department of Biochemistry and Molecular and Cellular Biology of the University of Zaragoza. In February 2010 I moved to the Translational Research Unit of the "Instituto Aragonés de Ciencias de la Salud" thanks to a "Miguel Servet" contract to incorporate researchers to the Spanish National Healthcare System. I was there until April 2013 when I joined Prof. Massimo Zeviani's group at the MRC-Mitochondrial Biology Unit in Cambridge, UK, where I am presently working

*Commissione Ricerca e Attività Correlate*