

**Project Narrative**

Pulmonary hypertension is a deadly disease of the vessels that supply the lung that may in part be caused by a deficiency in iron-sulfur clusters – crucial metal complexes involved in a wide range of cellular functions. Frataxin, an iron-sulfur assembly protein, is mutated in a neurologic disease called Friedreich's ataxia where the presence of pulmonary hypertension has largely been neglected. This proposal aims to establish a direct molecular link between frataxin and the development of pulmonary hypertension to expand our understanding of diseases dependent upon iron-sulfur clusters, and, if successful, to improve management of Friedreich's ataxia patients and offer new drug targets for more effective treatment of pulmonary hypertension.