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Talk Title: **Chibby Family Members and Their Regulators in Ciliogenesis and Ciliopathies**

Abstract:
Cilia are microtubule-based protrusions from the cell surface and are surrounded by a ciliary membrane with distinct protein and lipid compositions. Cilia perform important biological functions, ranging from signal transduction to fluid movement. Dysfunctional cilia are associated with ciliopathies, such as intellectual disability, polycystic kidney disease, respiratory infection, infertility, and obesity. Although >1,500 proteins exist in cilia, their physiological functions especially in mammalian systems are poorly understood. My laboratory investigates the roles of Chibby (Cby) family members and their interacting proteins in ciliogenesis and organogenesis using mouse models. Many of these factors are mutated in ciliopathy patients. We demonstrated that Cby1 localizes to the base of cilia and plays a critical role in ciliogenesis, and that Cby1-knockout mice show ciliopathy phenotypes, including respiratory infection and kidney and pancreatic cysts. We found that Cby1 forms a complex with BAR domain-containing ciBAR1 and ciBAR2 proteins and binds to lipid membranes, suggesting that the Cby1/ciBAR complex may regulate ciliogenesis through membrane remodeling and shaping. More recently, we have established new knockout mouse models for ciBAR1, ciBAR2, and a Cby family member, Cby1-Like (Cby1L). Their ciliopathy phenotypes and the molecular functions of ciBAR and Cby1L will be discussed.