

POSTER PRESENTATION

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SYSCILIA, “A systems biology approach to dissect cilia function and its disruption in human genetic disease”

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Primary cilia are basically signaling hubs, harboring amongst others the noncanonical WNT, Hedgehog, and PDGF signaling systems, and their disruption leads to striking developmental defects. Some ciliopathy-associated proteins have recently been revealed to be physically or functionally associated in several distinct groupings, with limited connections to other crucial biological processes. Early proteomics studies have also suggested a discrete repertoire of about 1000 proteins within the organelle (i.e. <5% of the proteome) that are still in need of organisation into pathways and networks. Small, relatively isolated systems are often targeted by systems biology approaches under the assumption that a limited set of molecules and interactions will be more tractable for modelling systems. Cilia are thus ideal organelles for systems biology as they can be regarded as semi-closed systems being both largely spatially and biologically separated from many other cellular structures and processes.

Scientific and technical objectives

Our overall objectives are to establish a paradigm for studying and modelling complex eukaryotic systems, to understand how system perturbation contributes to the modulation of clinical phenotypes, and to provide a better understanding of ciliary processes in biology and their associated diseases. Our objectives focus on all critical components of the systems biology process, namely:

- assay development and application
- data generation, handling and integration
- model building and testing followed by refinement.

We also exploit the models to find new insights into biological mechanism and human disease, and to develop approaches for ciliotherapies.

<http://syscilia.org/>

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