

ORAL PRESENTATION

Open Access

Dishevelled stabilisation at the cilium by RPGRIP1L is essential for planar cell polarity

S Schneider-Maunoury^{1*}, A Mahuzier¹, HM Gaudé², I Anselme¹, F Silbermann², M Leroux-Berger¹, M Montcouquiol³, S Saunier², C Vesque¹

From First International Cilia in Development and Disease Scientific Conference (2012)
London, UK. 16-18 May 2012

Cilia are involved in planar polarity in different systems but the mechanisms by which they influence the polarization process are unclear [1]. In order to clarify this issue, we investigated the function of the ciliary gene *Rpgrip1l* (*Ftm/NPHP8/MKSS*) in the mammalian cochlear sensory epithelium and in the zebrafish floor plate. We and others have previously shown that mutations in the human *RPGRIP1L* gene cause Meckel and Joubert type B syndromes [2]. The *Rpgrip1l* protein is localised at the ciliary transition zone and is required for transduction of the Hh/Gli pathway [3]. Our recent work has shown that *Rpgrip1l* patterns the telencephalon via the regulation of Gli3 proteolytic cleavage [4]. Here we show that in both the mammalian cochlear sensory epithelium and the zebrafish floor plate, *Rpgrip1l* is required for correct positioning of the basal body along the planar polarity axis. Our results strongly suggest that *Rpgrip1l* is essential for stabilizing the adaptor protein dishevelled at the basal body and/or cilium. Finally, we demonstrate that, in the zebrafish floor plate, the function of *Rpgrip1l* in basal body positioning is mediated by dishevelled. We propose that *Rpgrip1l* participates in a protein complex required for stabilizing dishevelled at the cilium, and that this stabilization is essential for asymmetric localization of the basal body along the planar polarity axis.

Author details

¹CNRS UMR7622, Université Pierre et Marie Curie, France. ²INSERM U983, Hôpital Necker-Enfants Malades, France. ³INSERM U862, Université Bordeaux 2, France.

Published: 16 November 2012

References

1. Wallingford, Mitchell : *Genes Dev* 2011, **25**:201-13.

* Correspondence: sylvie.schneider-maunoury@snv.jussieu.fr

¹CNRS UMR7622, Université Pierre et Marie Curie, France

Full list of author information is available at the end of the article

2. Delous, *et al: Nat Genet* 2007, **39**:875-81.
3. Vierkotten, *et al: Development* 2007, **134**:2569-77.
4. Besse, *et al: Development* 2011, **138**:2079-88.

doi:10.1186/2046-2530-1-S1-O21

Cite this article as: Schneider-Maunoury *et al.*: Dishevelled stabilisation at the cilium by RPGRIP1L is essential for planar cell polarity. *Cilia* 2012 **1**(Suppl 1):O21.

Submit your next manuscript to BioMed Central and take full advantage of:

- Convenient online submission
- Thorough peer review
- No space constraints or color figure charges
- Immediate publication on acceptance
- Inclusion in PubMed, CAS, Scopus and Google Scholar
- Research which is freely available for redistribution

Submit your manuscript at
www.biomedcentral.com/submit

