

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

Sylvie Schneider-Maunoury, Alexia Mahuzier, Hélori-Mael Gaudé, Isabelle Anselme, Flora Silbermann, Margot Leroux-Berger, Mireille Montcouquiol, Sophie Saunier, Christine Vesque

► **To cite this version:**

Sylvie Schneider-Maunoury, Alexia Mahuzier, Hélori-Mael Gaudé, Isabelle Anselme, Flora Silbermann, et al.. Dishevelled stabilisation at the cilium by RPGRIP1L is essential for planar cell polarity. First International Cilia in Development and Disease Scientific Conference, 1 (Suppl 1), pp.O21, 2012. <inserm-00752956>

HAL Id: inserm-00752956

<http://www.hal.inserm.fr/inserm-00752956>

Submitted on 16 Nov 2012

HAL is a multi-disciplinary open access archive for the deposit and dissemination of scientific research documents, whether they are published or not. The documents may come from teaching and research institutions in France or abroad, or from public or private research centers.

L'archive ouverte pluridisciplinaire **HAL**, est destinée au dépôt et à la diffusion de documents scientifiques de niveau recherche, publiés ou non, émanant des établissements d'enseignement et de recherche français ou étrangers, des laboratoires publics ou privés.

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

