

Reporting Summary

Nature Portfolio wishes to improve the reproducibility of the work that we publish. This form provides structure for consistency and transparency in reporting. For further information on Nature Portfolio policies, see our [Editorial Policies](#) and the [Editorial Policy Checklist](#).

Statistics

For all statistical analyses, confirm that the following items are present in the figure legend, table legend, main text, or Methods section.

n/a Confirmed

- | | | |
|-------------------------------------|-------------------------------------|--|
| ☐ | ☒ | The exact sample size (n) for each experimental group/condition, given as a discrete number and unit of measurement |
| ☐ | ☒ | A statement on whether measurements were taken from distinct samples or whether the same sample was measured repeatedly |
| ☐ | ☒ | The statistical test(s) used AND whether they are one- or two-sided
<i>Only common tests should be described solely by name; describe more complex techniques in the Methods section.</i> |
| ☒ | ☐ | A description of all covariates tested |
| ☐ | ☒ | A description of any assumptions or corrections, such as tests of normality and adjustment for multiple comparisons |
| ☐ | ☒ | A full description of the statistical parameters including central tendency (e.g. means) or other basic estimates (e.g. regression coefficient) AND variation (e.g. standard deviation) or associated estimates of uncertainty (e.g. confidence intervals) |
| ☐ | ☒ | For null hypothesis testing, the test statistic (e.g. F , t , r) with confidence intervals, effect sizes, degrees of freedom and P value noted
<i>Give P values as exact values whenever suitable.</i> |
| ☒ | ☐ | For Bayesian analysis, information on the choice of priors and Markov chain Monte Carlo settings |
| ☒ | ☐ | For hierarchical and complex designs, identification of the appropriate level for tests and full reporting of outcomes |
| ☒ | ☐ | Estimates of effect sizes (e.g. Cohen's d , Pearson's r), indicating how they were calculated |

Our web collection on [statistics for biologists](#) contains articles on many of the points above.

Software and code

Policy information about [availability of computer code](#)

Data collection Confocal imaging data: Leica TCS SP8 MP; Andor Dragonfly200
HE staining imaging data: 3D Histech

Data analysis GraphPad Prism 8.2.1; Image J 1.54

For manuscripts utilizing custom algorithms or software that are central to the research but not yet described in published literature, software must be made available to editors and reviewers. We strongly encourage code deposition in a community repository (e.g. GitHub). See the Nature Portfolio [guidelines for submitting code & software](#) for further information.

Data

Policy information about [availability of data](#)

All manuscripts must include a [data availability statement](#). This statement should provide the following information, where applicable:

- Accession codes, unique identifiers, or web links for publicly available datasets
- A description of any restrictions on data availability
- For clinical datasets or third party data, please ensure that the statement adheres to our [policy](#)

All relevant data supporting the key findings of this study are available within the article, its Supplementary Information, or from the corresponding authors upon request. Source data are provided in the Source Data file. Source data are provided with this paper.

Research involving human participants, their data, or biological material

Policy information about studies with [human participants or human data](#). See also policy information about [sex, gender \(identity/presentation\), and sexual orientation](#) and [race, ethnicity and racism](#).

Reporting on sex and gender N/A

Reporting on race, ethnicity, or other socially relevant groupings N/A

Population characteristics N/A

Recruitment N/A

Ethics oversight N/A

Note that full information on the approval of the study protocol must also be provided in the manuscript.

Field-specific reporting

Please select the one below that is the best fit for your research. If you are not sure, read the appropriate sections before making your selection.

☒ Life sciences ☐ Behavioural & social sciences ☐ Ecological, evolutionary & environmental sciences

For a reference copy of the document with all sections, see [nature.com/documents/nr-reporting-summary-flat.pdf](https://www.nature.com/documents/nr-reporting-summary-flat.pdf)

Life sciences study design

All studies must disclose on these points even when the disclosure is negative.

Sample size The percentage of ciliated cells was calculated as the number of cells with cilia divided by the total number of cells in randomly selected fields. At least 100 cells were scored per experiment. For animal experiments, a minimum of three animals per group were used for statistical analysis.

Data exclusions No data exclusion in this study.

Replication Data reproducibility was confirmed by at least three independent experiments. All the reported experiments were reproducible

Randomization Mice were randomly assigned to each experimental group in all experiments.

Blinding The investigators were blinded to group allocation during data collection and analysis.

Reporting for specific materials, systems and methods

We require information from authors about some types of materials, experimental systems and methods used in many studies. Here, indicate whether each material, system or method listed is relevant to your study. If you are not sure if a list item applies to your research, read the appropriate section before selecting a response.

Materials & experimental systems

n/a Involved in the study

☐ ☒ Antibodies

☐ ☒ Eukaryotic cell lines

☒ ☐ Palaeontology and archaeology

☐ ☒ Animals and other organisms

☒ ☐ Clinical data

☒ ☐ Dual use research of concern

☒ ☐ Plants

Methods

n/a Involved in the study

☒ ☐ ChIP-seq

☒ ☐ Flow cytometry

☒ ☐ MRI-based neuroimaging

Antibodies

Antibodies used

Antibodies against acetylated α -tubulin (T7451; IF 1:1000) and γ -tubulin (T6557; IF 1:1000) were obtained from Sigma-Aldrich. Antibodies against NPHP3 (22026-1-AP; IF 1:500), NPHP1 (25971-1-AP; IF 1:500), RPGRIP1L (29778-1-AP; IF 1:500), CEP164 (22227-1-AP; IF 1:500), centrin-1 (12794-1-AP; IF 1:500), CP110 (12780-1-AP; IF 1:500), MKS3 (13975-1-AP; IF 1:500), ninein (13007-1-AP; IF 1:500), IFT88 (13967-1-AP; IF 1:500), and ARL13B (66739-1-Ig; IF 1:1000) were from Proteintech. Antibodies against GFP (ab290; IB 1:1000), β -actin (AB0033; IB 1:5000), and Flag (AB0008; IB 1:1000) were from Abways. The antibody against γ -tubulin (ab11317; IF 1:500) was purchased from Abcam. The antibody against Flag (AWA06018; IF 1:500) was from Abiowell. Antibodies against CFAP100 were from OriGene Technologies (TA334871; IB 1:1000) or generated in rats (IF 1:500) using His-tagged C-terminal fragments of recombinant CFAP100 purified from E. coli. The antibody against GT335 (AG-20B-0020-C100; IF 1:500) was from AdipoGen.

Antibodies against CEP290 (sc-390462; IF 1:500) and PCM1 (sc-398365; IF 1:500) were from Santa Cruz Biotechnology. Horseradish peroxidase-conjugated secondary antibodies were from Amersham Biosciences. Alexa Fluor 488-, 568-, and 647-conjugated secondary antibodies were from Invitrogen.

Validation

Primary antibodies against CFAP100 was validated using knockdown and/or knockout samples in the main and supplementary figures. Other primary antibodies are commercially available and validation results can be found at manufacturer's websites.

Eukaryotic cell lines

Policy information about [cell lines and Sex and Gender in Research](#)

Cell line source(s)

RPE1, HEK293T, IMCD3, NIH3T3 and HeLa cells were described previously (Ref. 16, 48).

Authentication

Cell lines were authenticated by STR profiling.

Mycoplasma contamination

RPE1, HEK293T, IMCD3, NIH3T3 and HeLa cells were tested and found to be mycoplasma-free.

Commonly misidentified lines (See [ICLAC](#) register)

No commonly used misidentified cell lines were used.

Animals and other research organisms

Policy information about [studies involving animals](#); [ARRIVE guidelines](#) recommended for reporting animal research, and [Sex and Gender in Research](#)

Laboratory animals

Cfap100-knockout mice in the C57BL/6 background were used.

Wild animals

No wild animals were used.

Reporting on sex

Sex was not considered in study design.

Field-collected samples

No field-collected samples were used.

Ethics oversight

All relevant ethical regulations for animal testing and research have been complied with. All animal procedures were approved by the Animal Care and Use Committee of Shandong Normal University

Note that full information on the approval of the study protocol must also be provided in the manuscript.

Plants

Seed stocks

N/a

Novel plant genotypes

N/a

Authentication

N/a