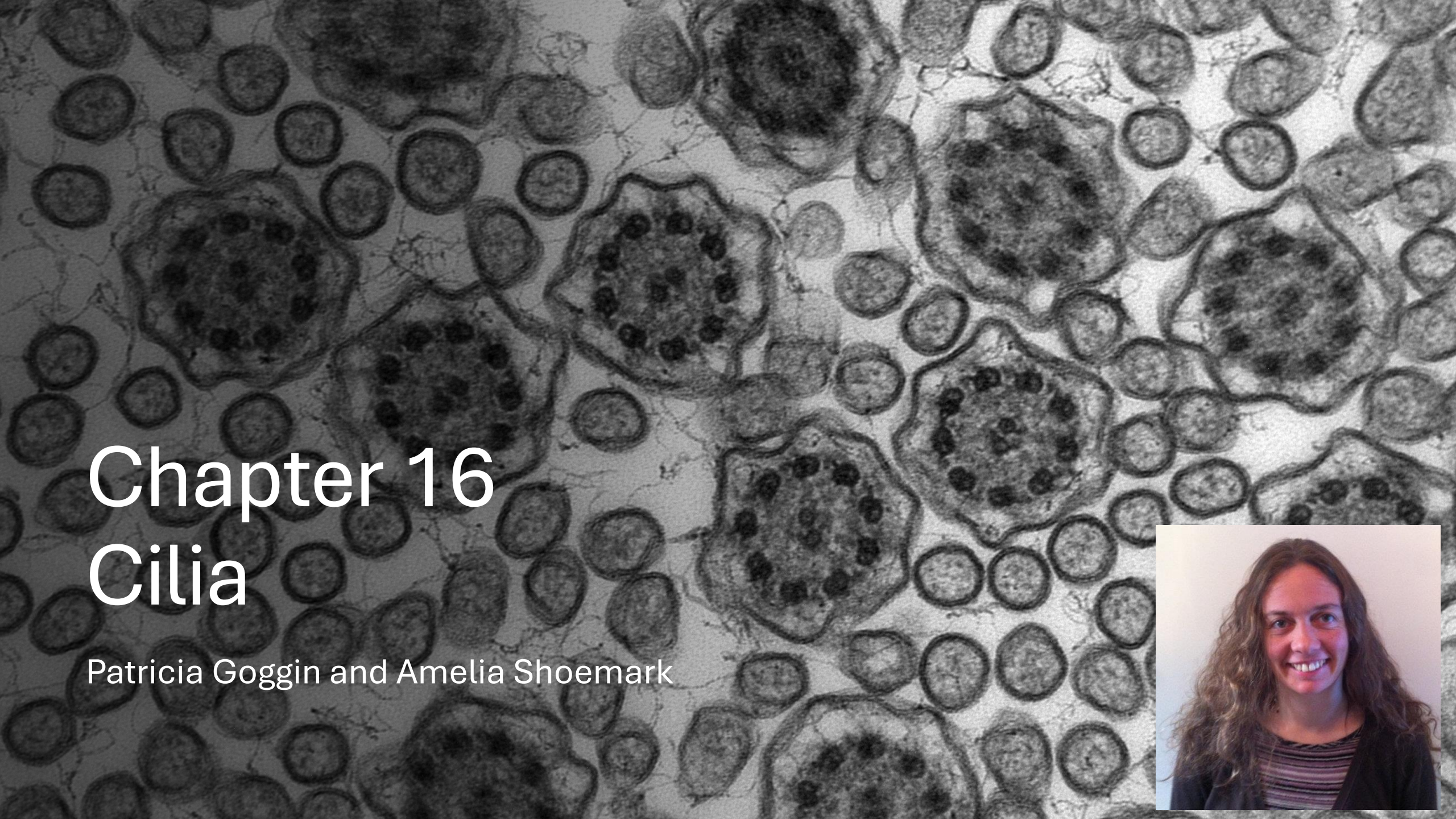


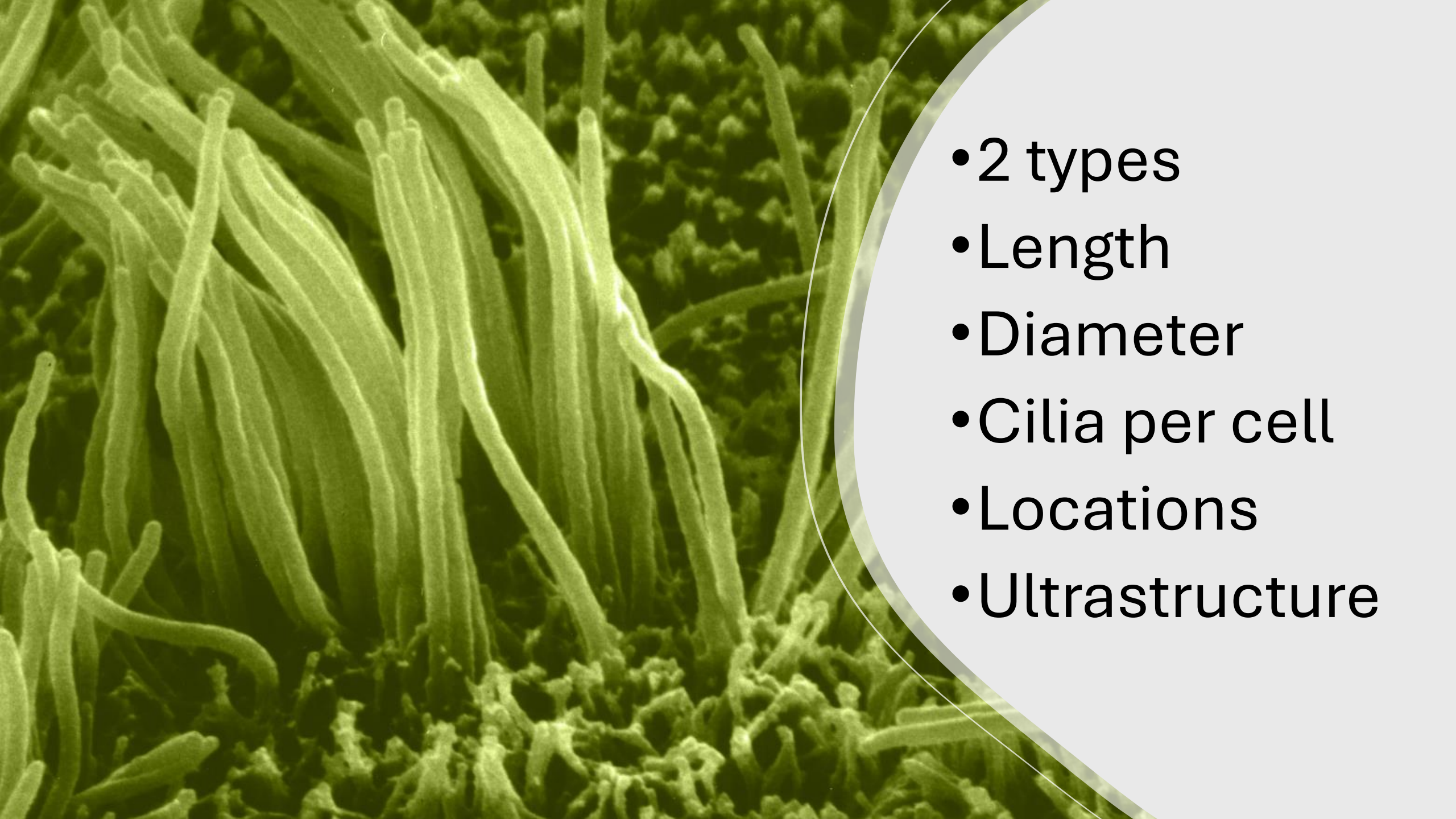
The background of the slide is a grayscale electron micrograph showing numerous cross-sections of cilia. These sections are arranged in a regular, repeating pattern, with each section displaying a characteristic 9x3 arrangement of microtubules. The microtubules appear as dark, circular structures within a lighter, textured matrix.

Chapter 16

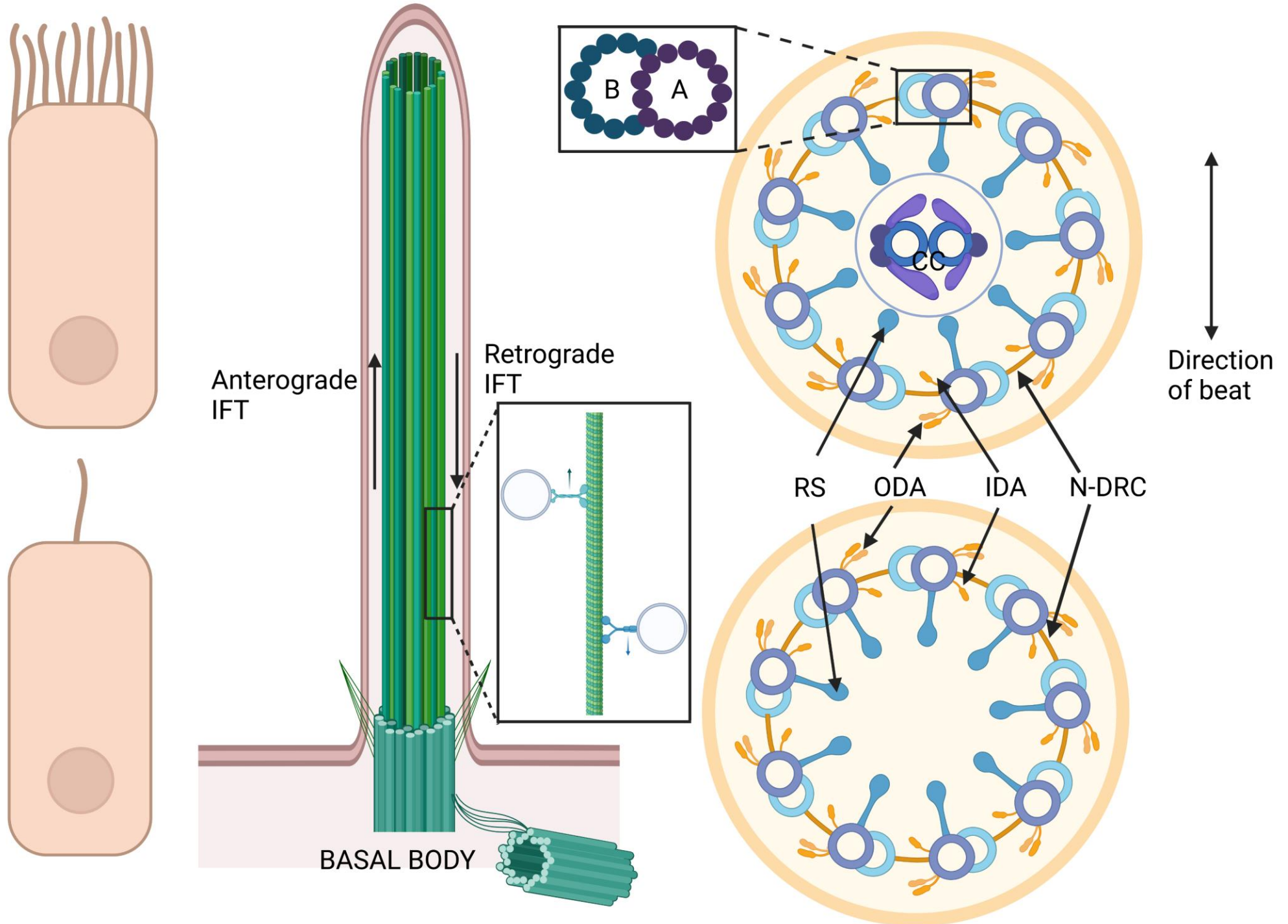
Cilia

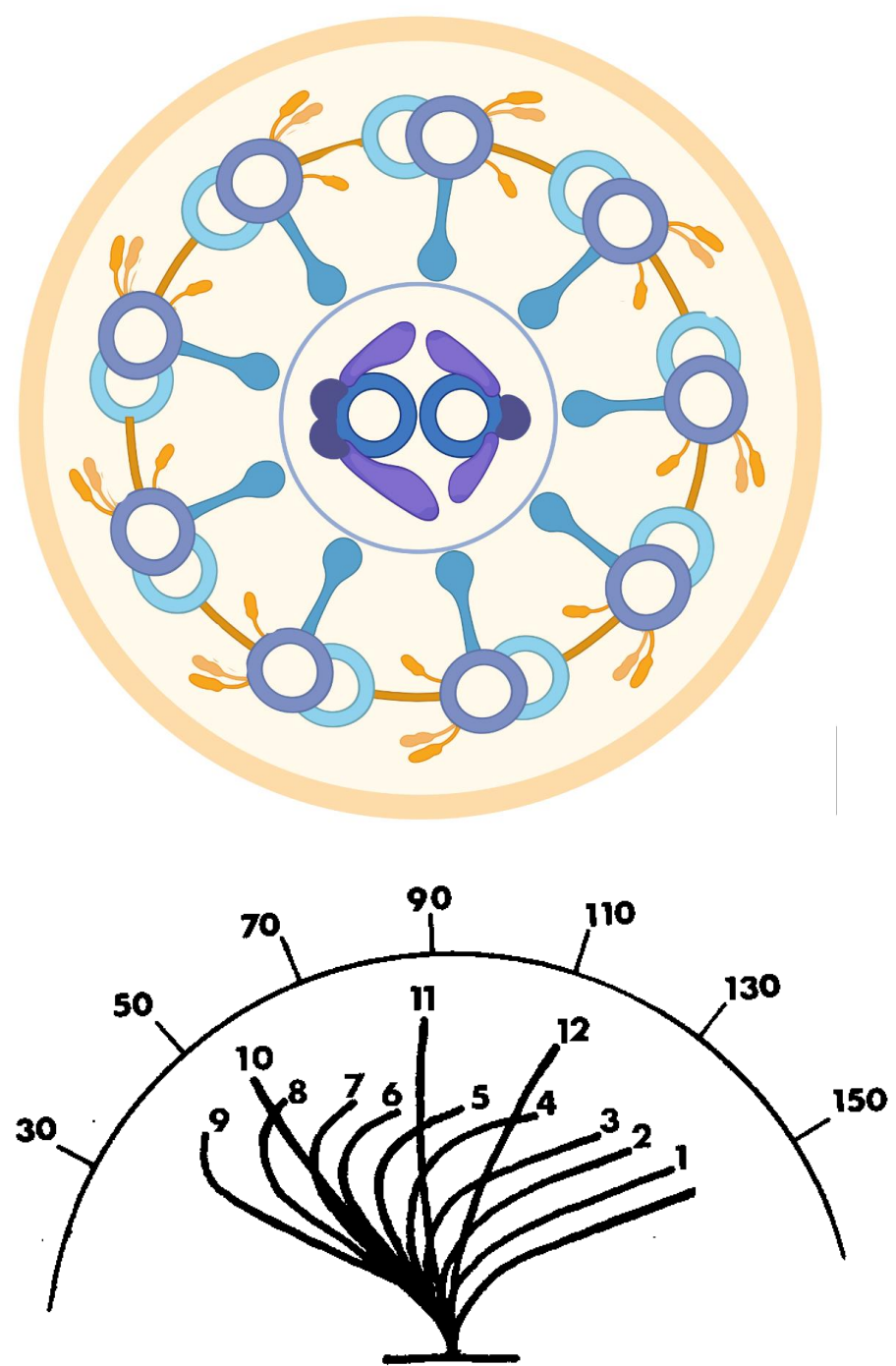
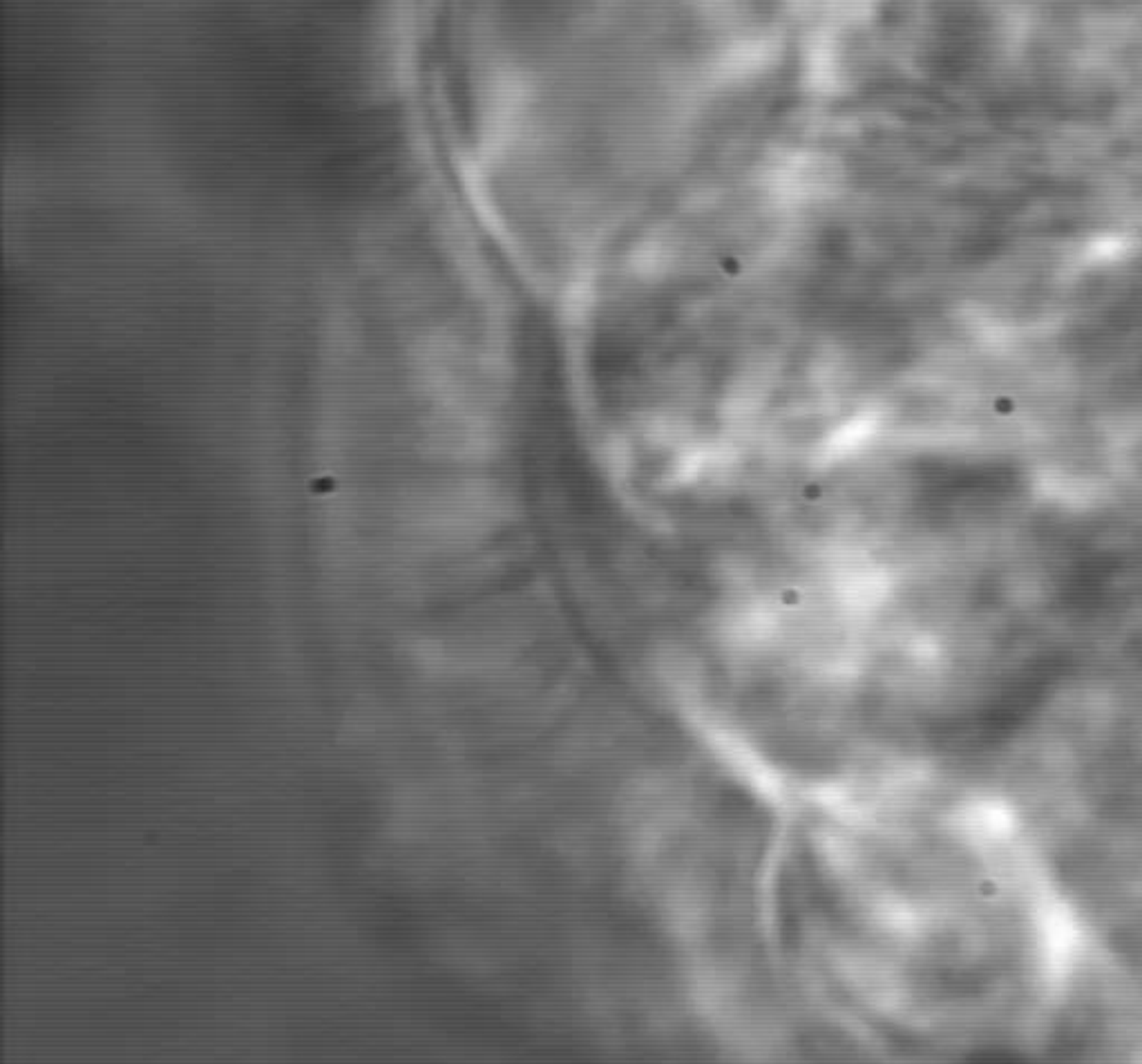
Patricia Goggin and Amelia Shoemark



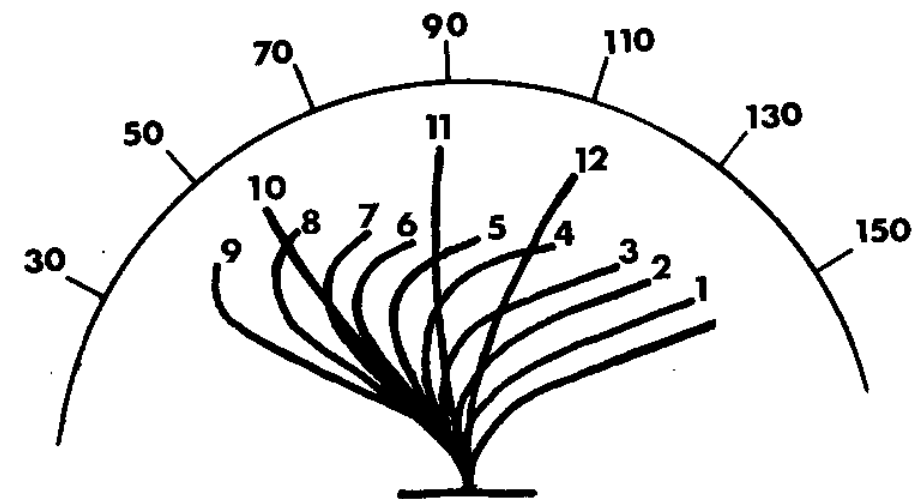
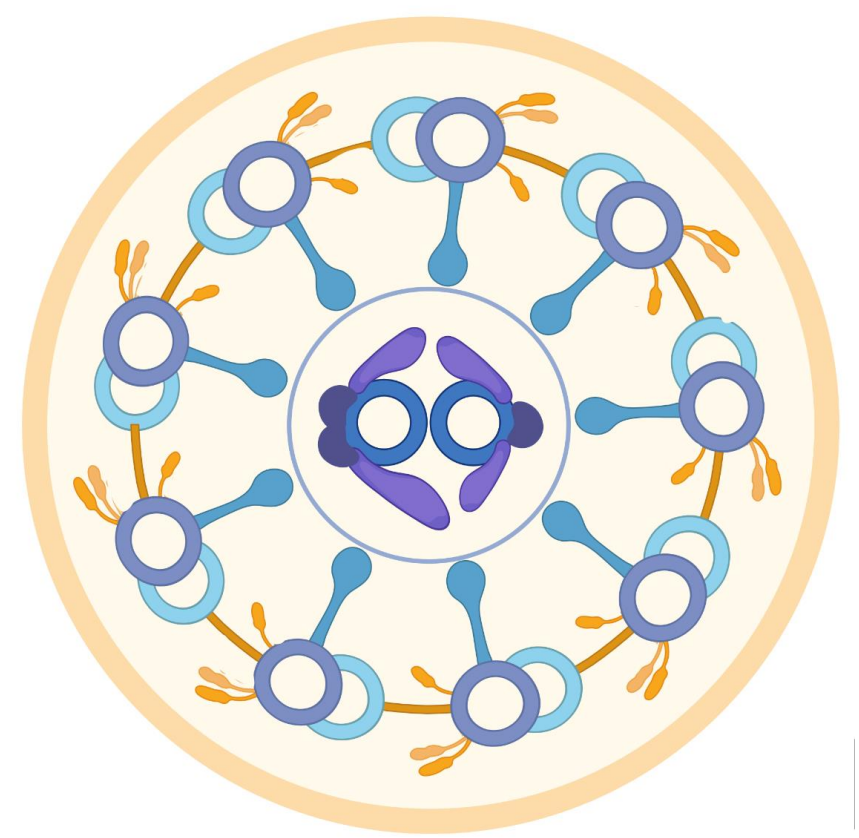
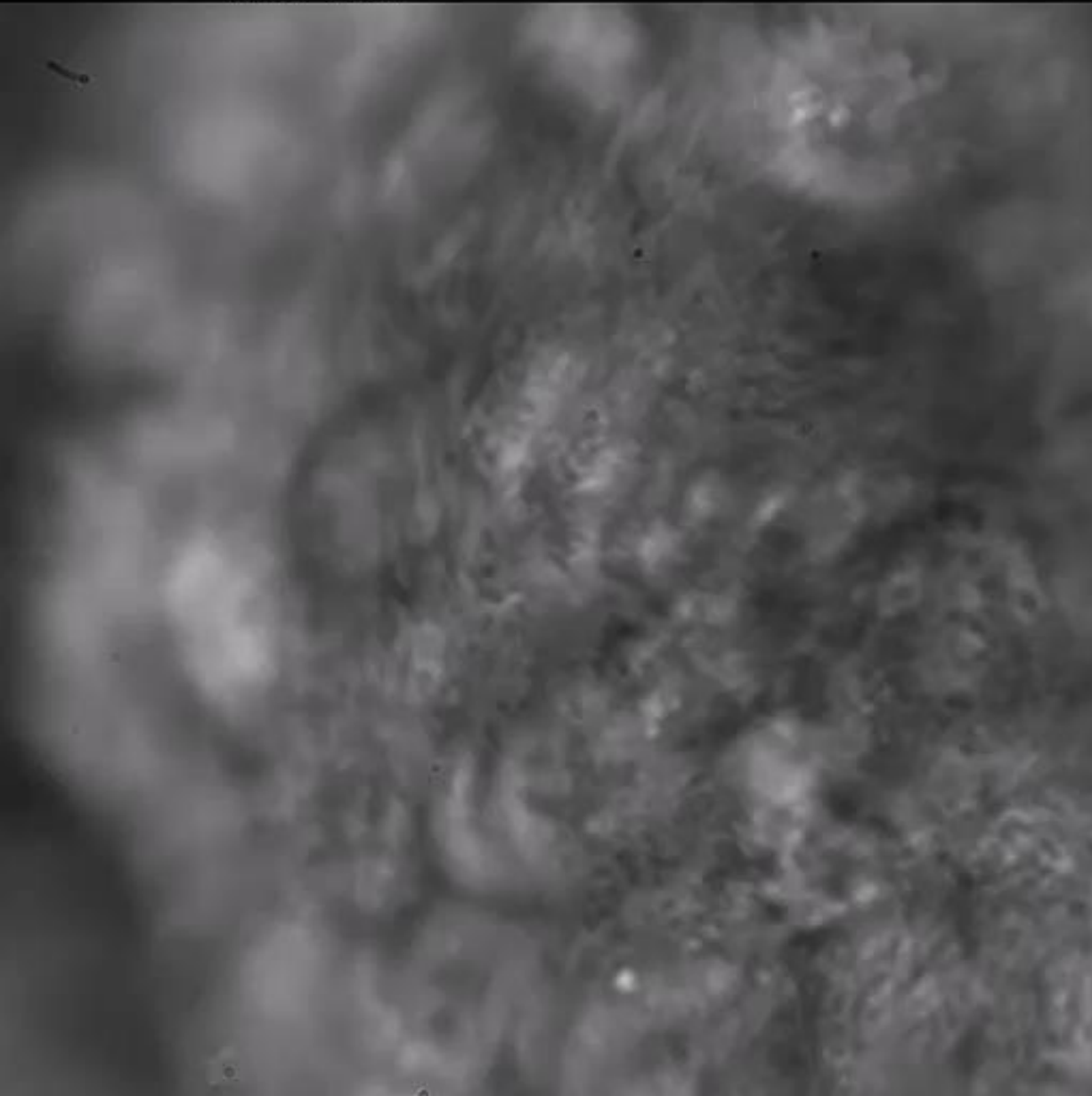


- 2 types
- Length
- Diameter
- Cilia per cell
- Locations
- Ultrastructure

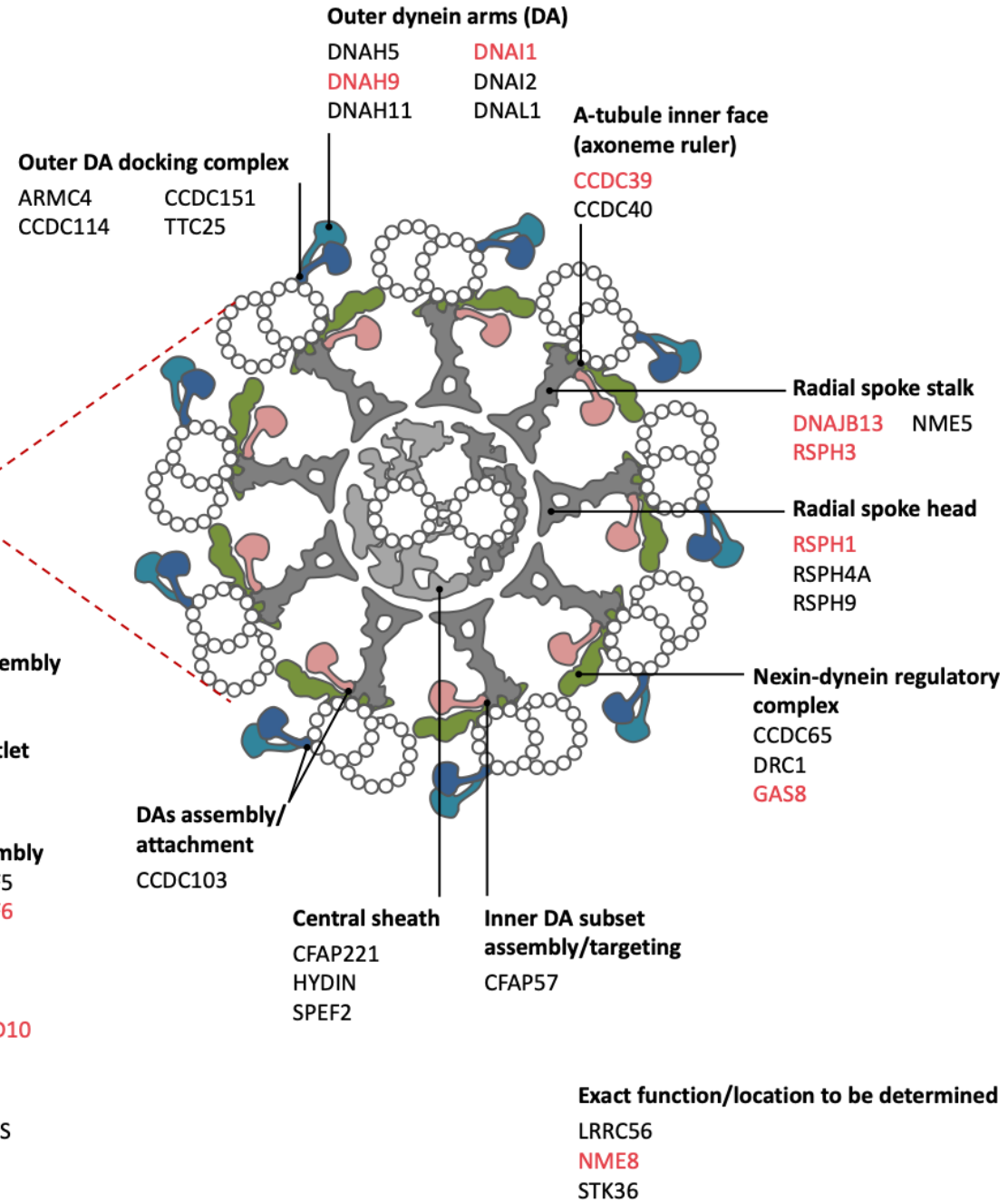
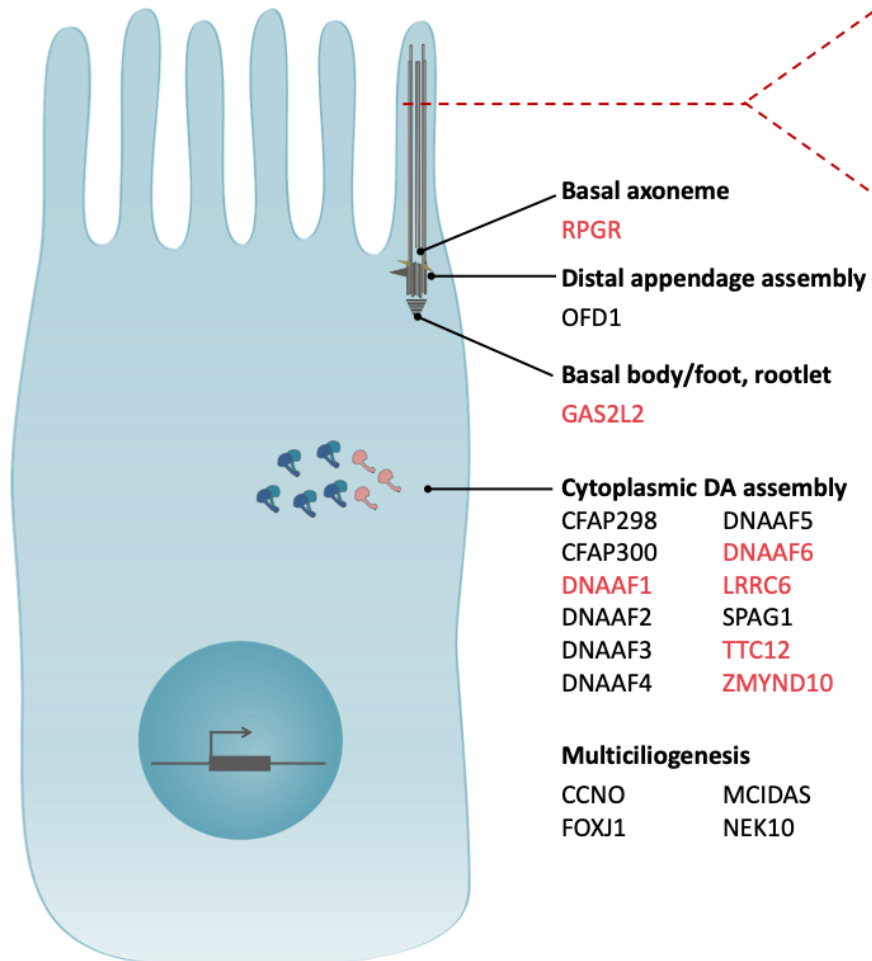




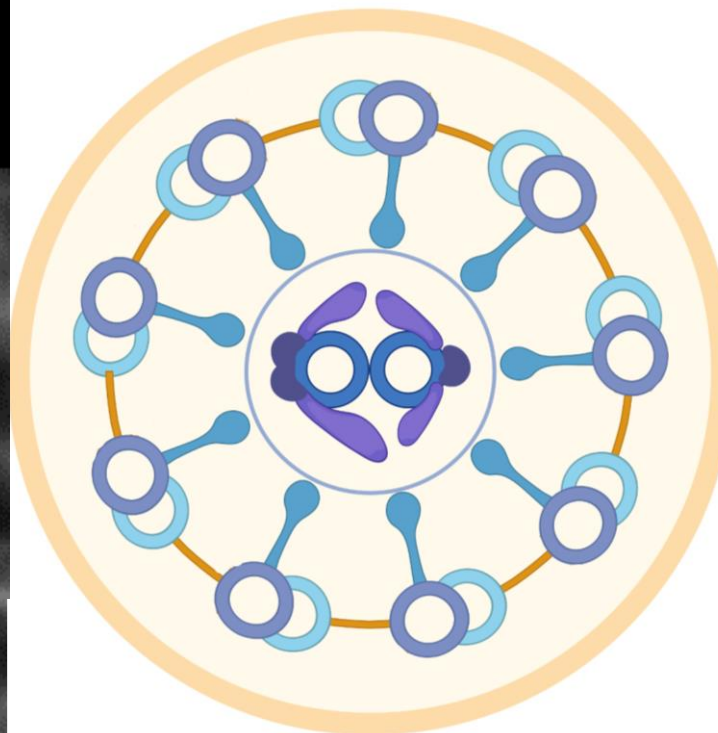
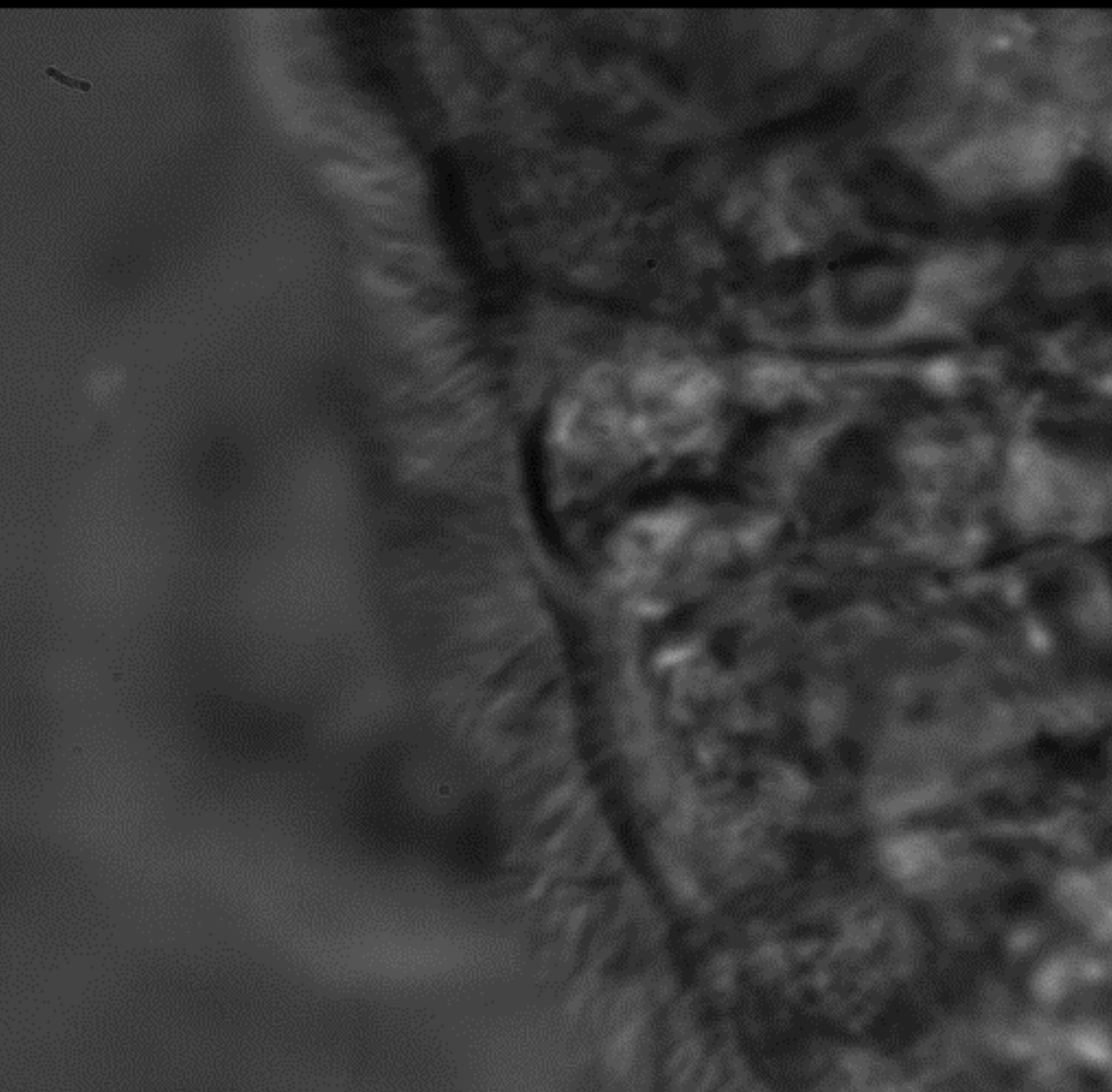
Photron FASTCAM MC2-... 500 fps
1/500 sec 512 x 512 Start
frame : 0 +0 ms Date : 2018/5/1
Time : 10:58







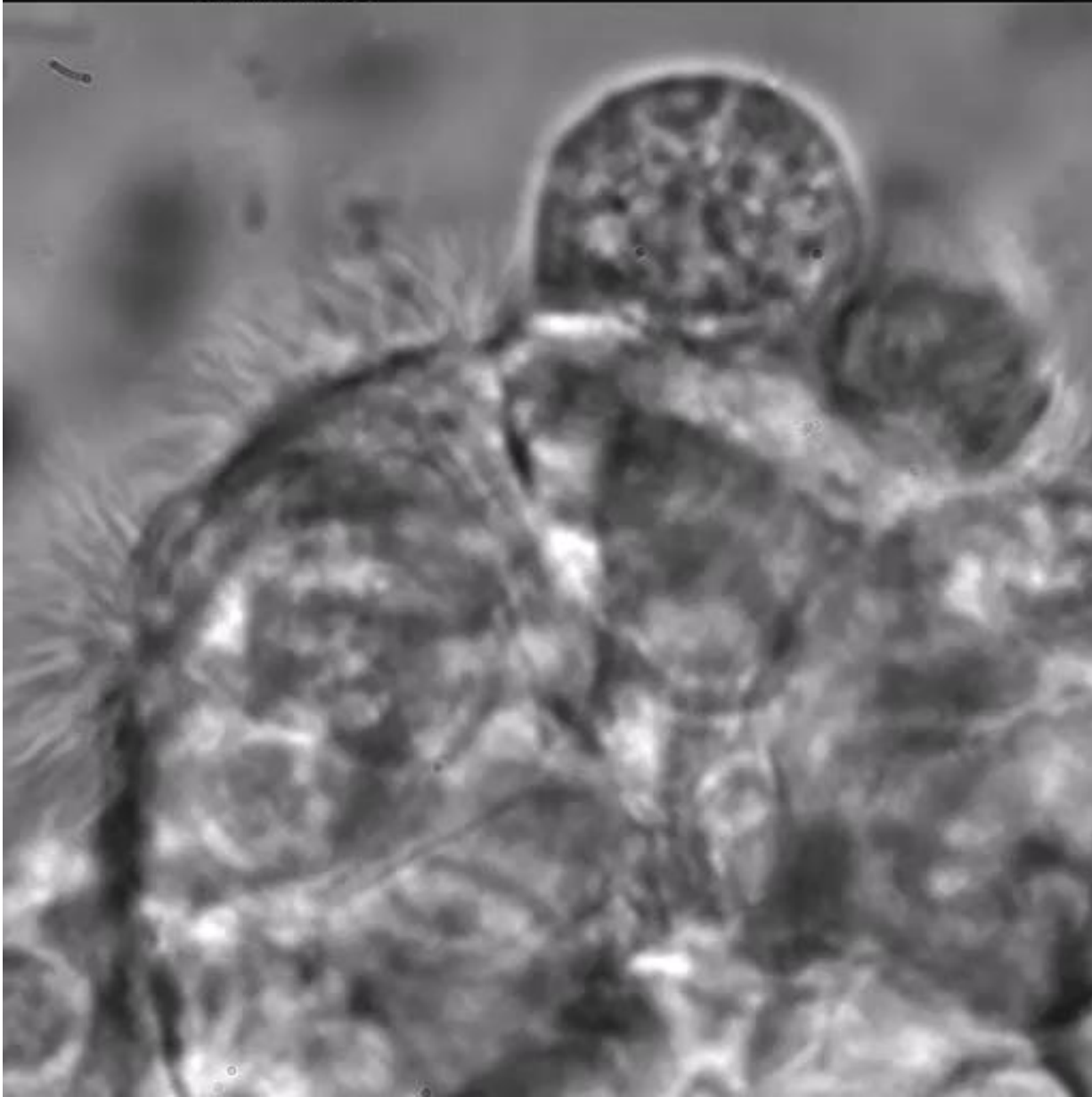
Photron FASTCAM MC2-5... 500 fps
1/500 sec 512 x 512 Start
frame : 0 +0 ms Date : 2018/10/4
Time : 12:34



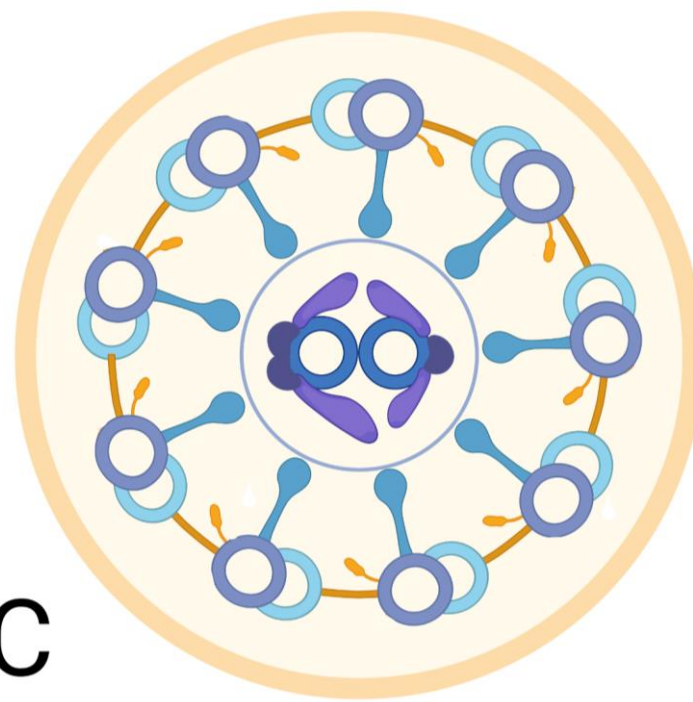
11 genes including:
DNAAF1
DNAAF2
DNAAF3
DYX1C1
HEATR2



Photron FASTCAM MC2-5... 500 fps
1/500 sec 512 x 512 Start
frame : 0 +0 ms Date : 2018/4/26
Time : 12:03



C

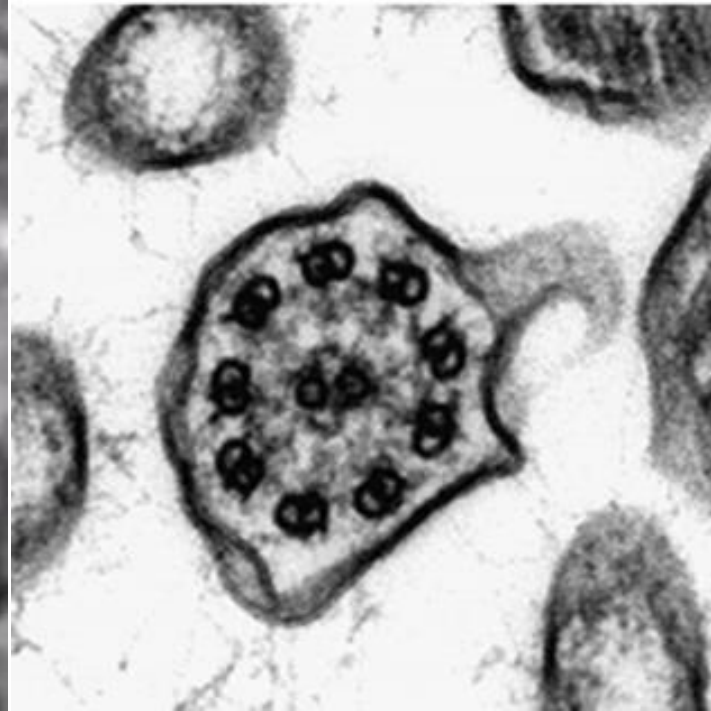


>20 genes including:

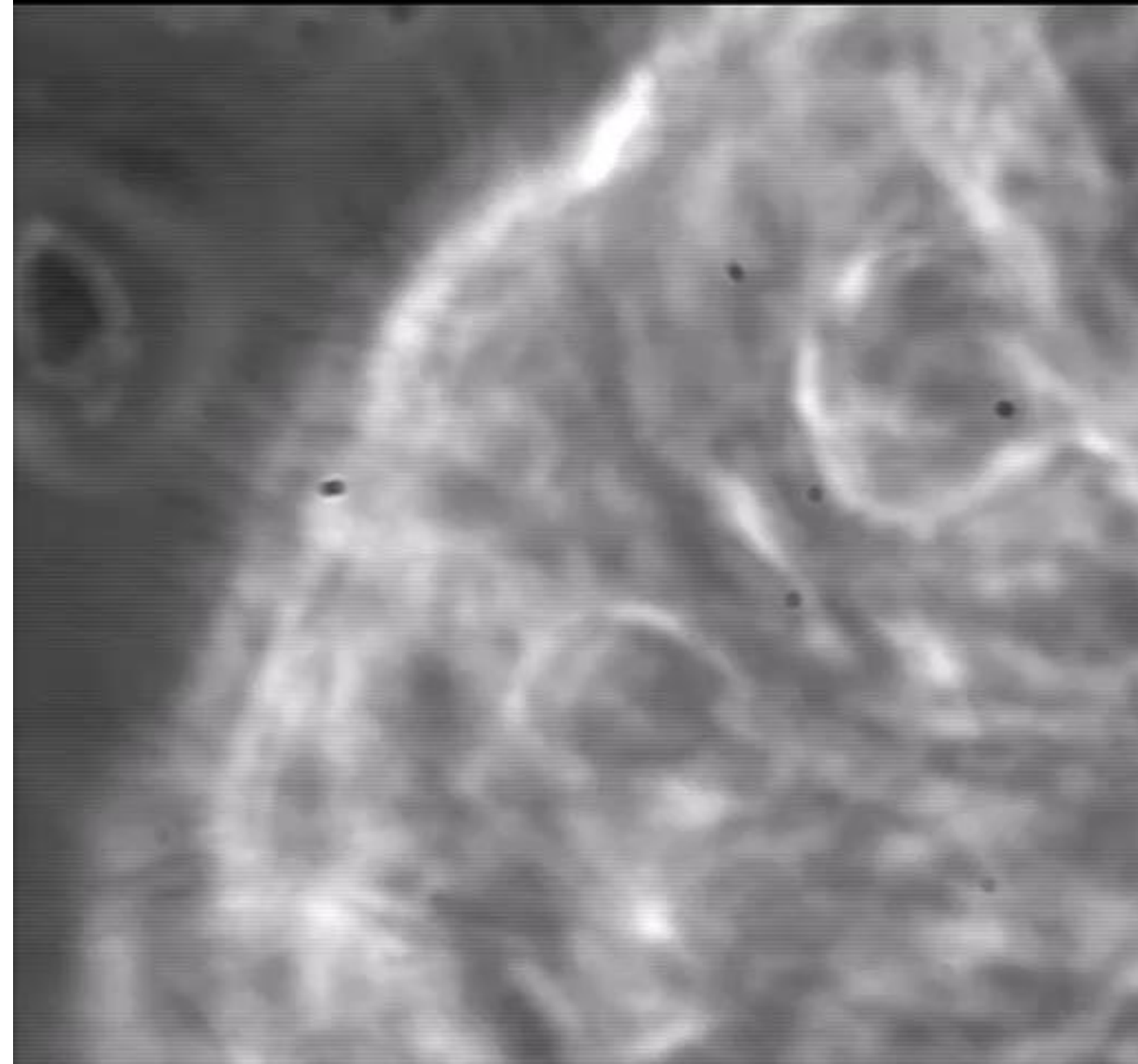
Structural proteins
SDNAH5, DNAI1,
DNAI2, NME8

Docking complexes
CCDC114, CCDC151,
ARMC4, TTC25

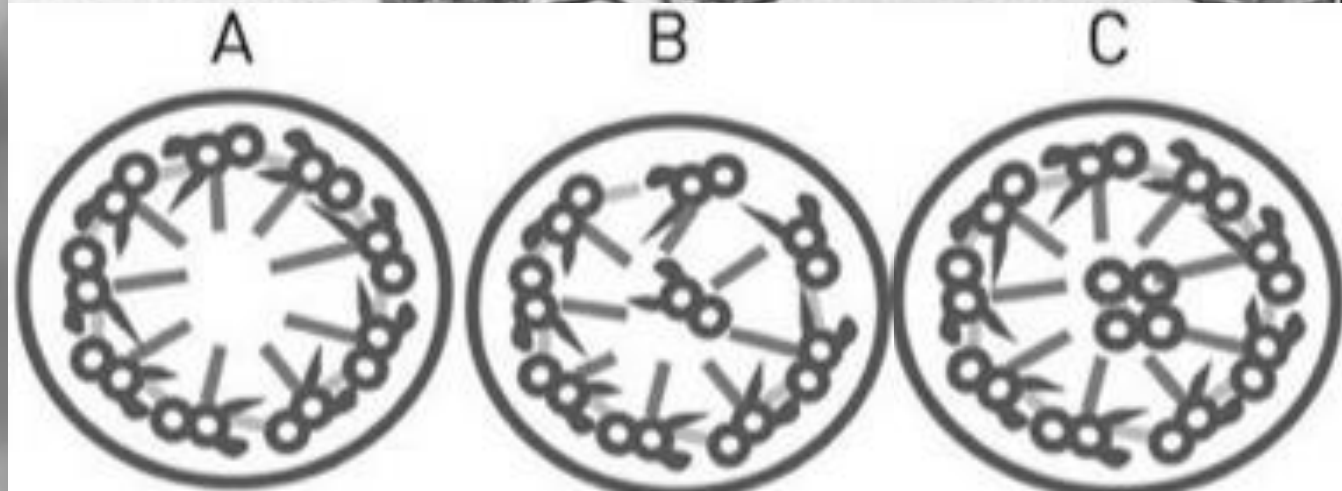
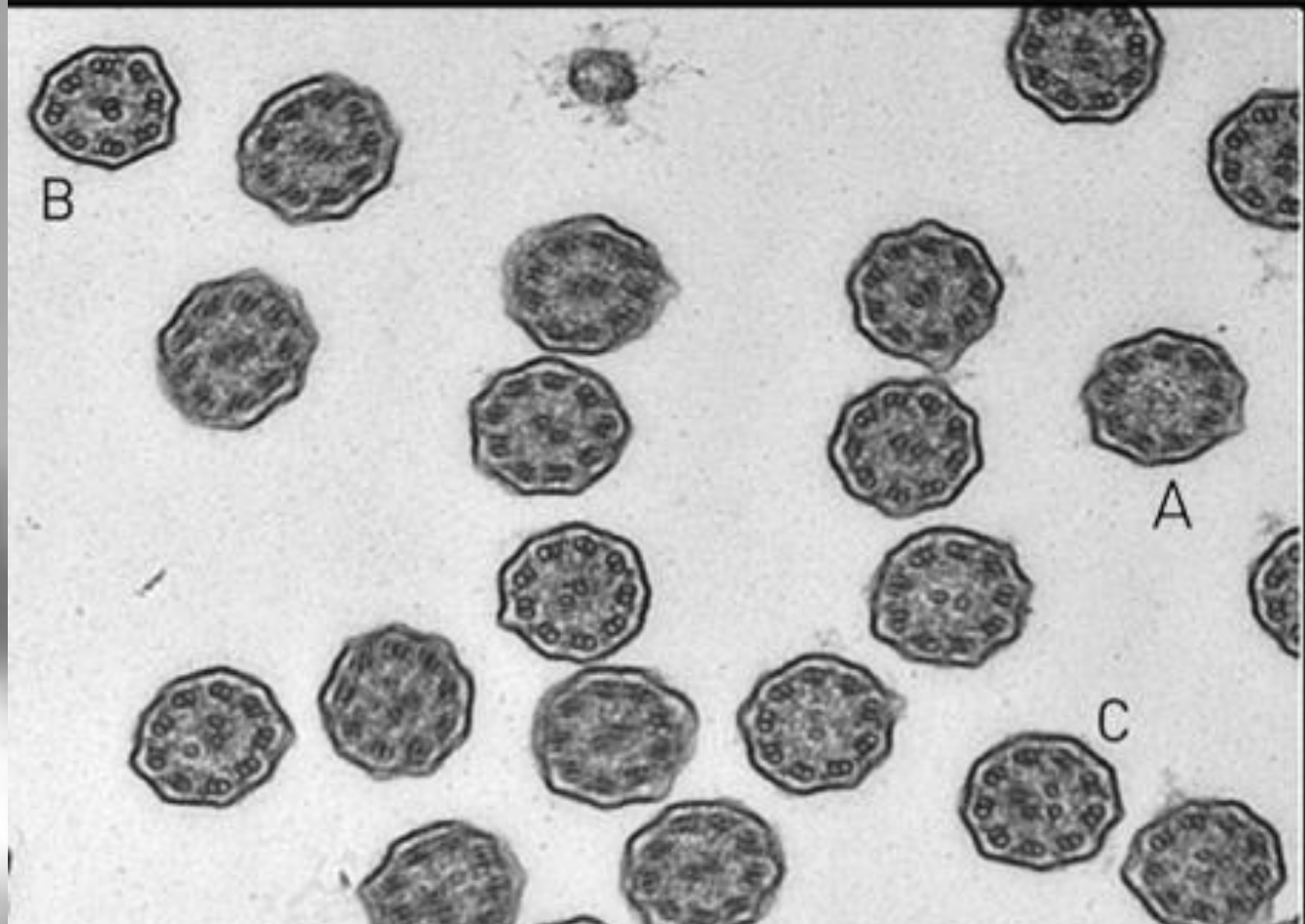
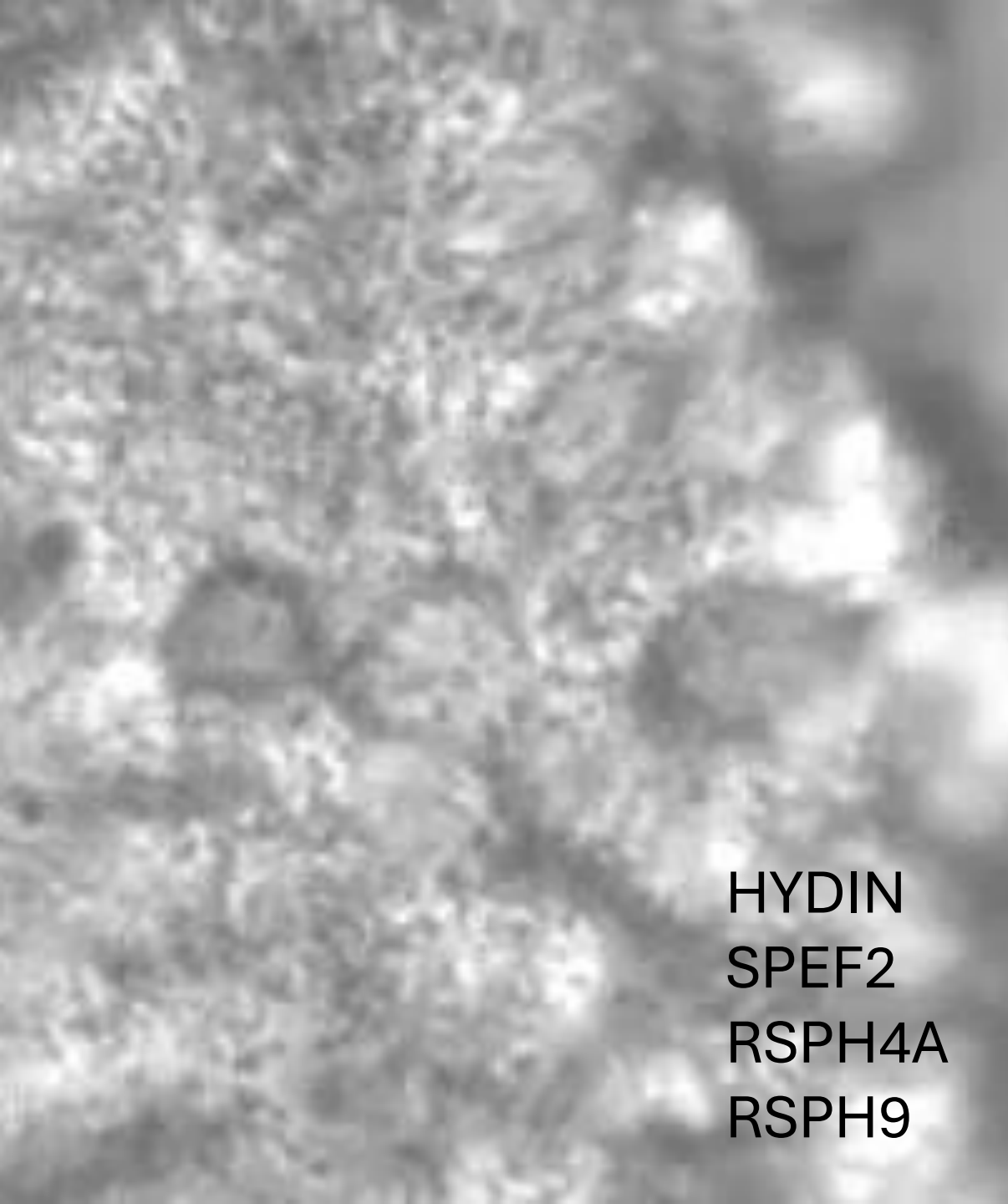
Attachment factor
CCDC103



Photron FASTCAM-PCI R2 m...
250 fps 1/250 sec 512 x 480
Start frame : 1 +00:00:00.000000sec



DNAH11
GAS2L2
CCDC65

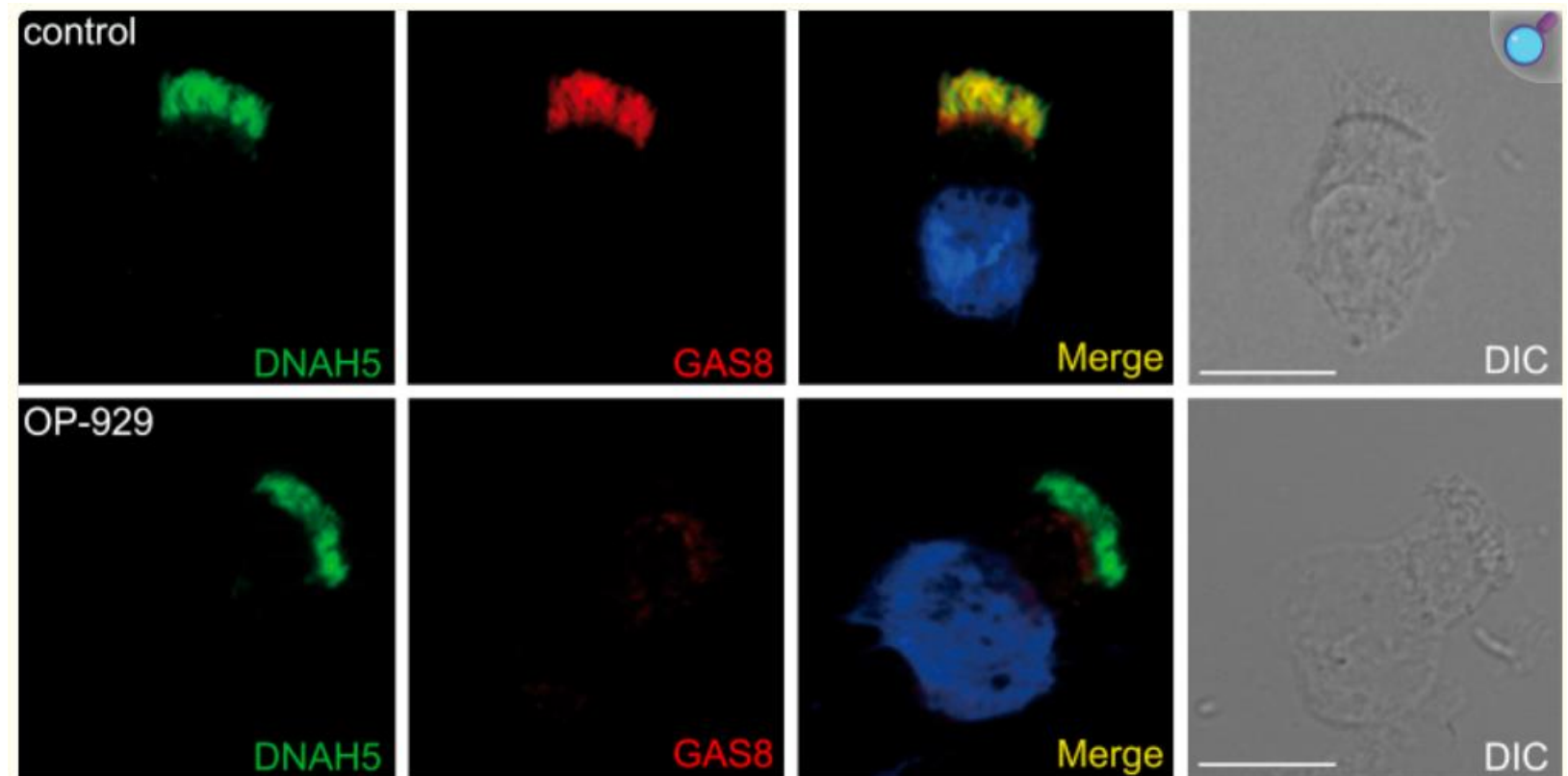


GAS8 - nexin-dynein regulatory complex protein

- Normal TEM
- Normal/near normal LM

GAS8 - nexin-dynein regulatory complex protein

- Normal TEM
- Normal/near normal LM
- IF
- Clinical features
- Genetics

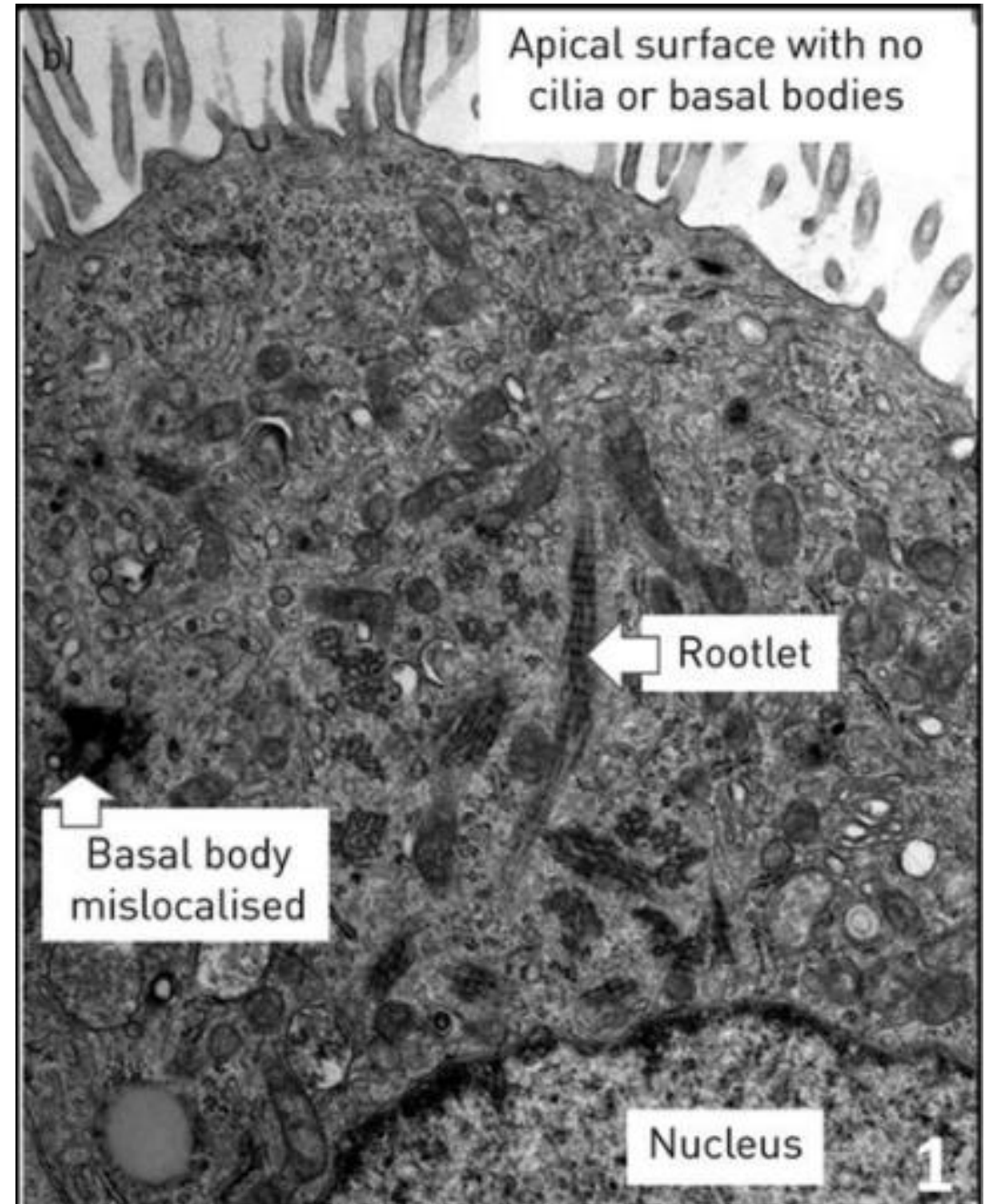


Reduced generation of multiple motile cilia

MCIDAS

CCNO

FOXJ1





CrossMark

International consensus guideline for reporting transmission electron microscopy results in the diagnosis of primary ciliary dyskinesia (BEAT PCD TEM Criteria)

Amelia Shoemark^{1,2}, Mieke Boon³, Christoph Brochhausen⁴, Zuzanna Bukowy-Bieryllo⁵, Maria M. De Santi⁶, Patricia Goggin⁷, Paul Griffin^{1,8}, Richard G. Hegele⁹, Robert A. Hirst¹⁰, Margaret W. Leigh¹¹, Alison Lupton¹², Karen MacKenney¹³, Heymut Omran¹⁴, Jean-Claude Pache¹⁵, Andreia Pinto¹⁶, Finn P. Reinholt¹⁷, Josep Schroeder⁴, Panayotis Yiallourous¹⁸ and Estelle Escudier¹⁹, representing the BEAT-PCD Network Guideline Development Group¹



@ERSpublications

This tested and validated consensus statement provides clear terminology and defect definitions for the ultrastructural diagnosis of primary ciliary dyskinesia (PCD). The guideline will facilitate standardisation of PCD research and diagnosis. <http://bit.ly/2RDkz7k>



BEAT-PCD
BETTER EXPERIMENTAL APPROACHES TO TREAT PCD

? Ciliopathy Alliance



CrossMark

TASK FORCE REPORT
ERS GUIDELINES



European Respiratory Society guidelines for the diagnosis of primary ciliary dyskinesia

Jane S. Lucas^{1,2}, Angelo Barbato³, Samuel A. Collins^{1,2}, Myrofora Goutaki^{4,5}, Laura Behan^{1,2}, Daan Caudri^{6,7}, Sharon Dell^{8,9}, Ernst Eber¹⁰, Estelle Escudier^{11,12}, Robert A. Hirst¹³, Claire Hogg¹⁴, Mark Jorissen¹⁵, Philipp Latzin⁵, Marie Legendre^{11,12}, Margaret W. Leigh¹⁶, Fabio Midulla¹⁷, Kim G. Nielsen¹⁸, Heymut Omran¹⁹, Jean-Francois Papon^{20,21}, Petr Pohunek²², Beatrice Redfern²³, David Rigau²⁴, Bernhard Rindlisbacher²⁵, Francesca Santamaria²⁶, Amelia Shoemark¹⁴, Deborah Snijders³, Thomy Tonia⁴, Andrea Titieni¹⁹, Woolf T. Walker^{1,2}, Claudius Werner¹⁹, Andrew Bush¹⁴ and Claudia E. Kuehni⁴