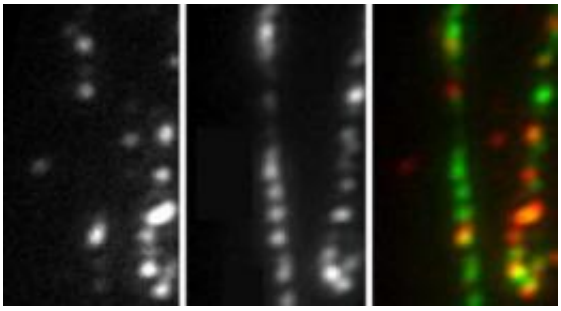
BBS proteins shown to run an export business that protects cilia

December 28 2009



The BBSome (red) removes signaling proteins from flagella by linking them to a subset of IFT particles (green).

A protein complex mutated in human disease removes excess signaling molecules to prevent them from damaging cilia, say researchers from UMass Medical School. The study will be published in the December 28 issue of the *Journal of Cell Biology*.

Defective cilia cause a range of diseases including Bardet-Biedl syndrome (BBS), a rare, multi-tissue disorder linked to mutations in 12 different proteins. Seven of these form a complex called the BBSome, but the function of this protein assembly in cilia and flagella is unclear.

In worms, the complex glues together the intraflagellar transport (IFT) machinery that assembles and maintains cilia by hauling cargo back and forth along the organelle's microtubules. But most mammalian cell types

can still form cilia in the absence of BBS proteins, suggesting that the BBSome isn't essential for IFT.

Lechtreck et al. turned to the green alga Chlamydomonas, and found that BBS proteins were only present on a subset of IFT particles in each of the alga's two flagella. Strains lacking components of the BBSome showed normal rates of IFT and proper flagellar structure, but couldn't steer away from bright light like wild-type cells could. Mutant flagella accumulated several signaling-related proteins, which the researchers think may disrupt the alga's response to light.

The researchers speculate that a similar buildup of disruptive proteins causes cilia dysfunction in BBS patients; the BBSome may remove excess signaling proteins from flagella by linking them to a subset of IFT particles undergoing retrograde transport out of the cilia. Author Karl Lechtreck says that the next step is to fluorescently tag the signaling proteins and compare their movements to BBS and IFT proteins.

More information: Lechtreck, K.-F., et al. 2009. J. Cell Biol. doi:10.1083/jcb.200909183

Provided by Rockefeller University