

Primary Ciliary Dyskinesia

Primary ciliary dyskinesia (PCD) is a genetic, incurable progressive lung disease affecting both the structure and function of the cilia which line the airways, ears and sinuses.

Clinical symptoms are generally limited to the upper and lower airway but motile cilia are also found in the reproductive tract and ventricles of the brain. Most males with PCD will be infertile and women sub-fertile.

Without appropriately functioning cilia, people with PCD are unable to maintain clear airways in their upper and lower respiratory system. A chronic productive cough, coryza and recurrent otitis media are hallmarks of PCD in infancy.

Frequent infections of the lungs, ears, throat, and sinuses are common and can lead to serious and permanent damage.

As for all causes of bronchiectasis, the treatment for PCD is focussed on airway clearance.

Kartageners Syndrome is the presence of both primary ciliary dyskinesia and situs inversus (a congenital condition where the major visceral organs are reversed or mirrored from their normal positions).

For more information:

European Respiratory guidelines for PCD:

<https://erj.ersjournals.com/content/early/2016/11/11/13993003.01090-2016.long>

Genetic Alliance Australia:

http://www.geneticalliance.org.au/conditions_detail.php?Immotile-Cilia-Syndrome-Primary-ciliary-dyskinesia-282