The Ciliopathies – PCD and Acquired Ciliary Dyskinesia

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The figure above shows some Cilia, found on ciliated respiratory epithelial cells. These cells line your airway, including middle ear, nose, sinuses, trachea, and bronchioles. The pink ball is a speck of pollen, and the gray specks are pollution particles.

The cilia normally sweep these particles away. When they can’t do their job normally, bad things happen:

otitis, mastoiditis, rhinitis, sinusitis, bronchitis, and pneumonia.

Here are some definitions, and some tips that might help.
When those little hair-like cilia don’t work normally, bad things happen. The medical terms are “dysmotile cilia syndrome”, “primary ciliary dyskinesia”, and acquired or “secondary ciliary dyskinesia.”

All of these conditions contribute to ear infections (otitis, mastoiditis), sinus infections (rhinosinusitis), and other respiratory infections like bronchitis, bronchiolitis, and pneumonia.

Whether “primary” or acquired, here are some tips that might help.

**Background (you might want to skip this part):**

*Cilia* are little hair-like projections on many different cell types. Their function is extremely complex. Our understanding of their function is still evolving. But we do know that normal function of cilia is crucial for normal **development** of many organs including kidney, brain, eye, and the cardiovascular system.

After development, normal cilia function is crucial for normal function of several organ systems, especially the lining of the respiratory tract – the **respiratory epithelium**. This epithelium lines the larger passages of the lower airway, and the upper airway including the ears, Eustachian tubes, nose, and sinuses. Here, cilia function to sweep a thin layer of secretions out of the airway generally toward the pharynx, or throat. These secretions, containing various microorganisms and airborne pollutants, are swallowed and the stomach acid generally kills them.
Primary Ciliary Dyskinesia (PCD):

PCD means to be born with an abnormality that prevents cilia from working normally. Although there is great excitement over recent discovery of some of the genes that are involved in “primary ciliary dyskinesia,” this is a relatively rare disease, with an incidence estimated to range from 1 in 15,000 to 1 in 60,000. Classic symptoms include chronic otitis, chronic sinusitis, and chronic bronchitis with pneumonias.

Click on this link to visit the PCD Foundation website.

Acquired Ciliary Dyskinesia:

Much more common is secondary, or acquired, ciliary dyskinesia. This is a condition where the cilia don’t function normally due to some insult – a viral infection for example, or some irritant such as tobacco smoke or other pollutants, or even the inflammation caused by allergens - things we are allergic to, like pollen. The bottom line is that healthy sinuses, ears, and lungs, all require normal muco-ciliary clearance: normally functioning cilia, and normal mucus with the proper viscosity and proper amount.

So, treat your cilia well ... be excellent to them:

So, keep ‘em safe from toxins and pollutants.

See previous blog post, “The Human Pollution Experiment,” for study on how air pollution affects our nasal and sinus cilia. Also see report, “Stop Breathing, Your Air is Killing You (and what to do about it)” for 8 steps to detoxify your air.
Also, Keep ‘em Moist:

Even without being exposed to one of the above insults, cilia may be unhappy. They are happiest in 100% humidity. If they become too dry they stop working normally. Use a humidifier at night if you live in a dry climate (see earlier post on what features to look for in a humidifier). Use a simple saline spray during the day (these are not addicting, unlike some nose sprays). See blog posts for how to use saline nasal rinses daily for happiest cilia (see earlier posts on sinus rinses, sinusitis, and rhinitis, listed below).

The Result:

If the cilia do not sweep away mucus with particulate matter including virus particles or bacteria or mold, the result can be chronic rhinitis and sinusitis – chronic rhinosinusitis, or CRS. Injury of the respiratory ciliary mechanism is just one of the ways that exposure to toxins and irritants contribute to asthma exacerbations and chronic rhinosinusitis.

Nose Pudding:

That is, if the cilia aren’t working normally, secretions containing all those microorganisms, pollutants, allergens, and general irritants, simply stagnate. They accumulate. The result is that your child always seems to have thick green “pudding” running from their nose.

Whereas some parents, and even some physicians, dismiss this as just a “snotty-nosed kid,” this is not normal. It can simply be a viral URI or “cold”. But a “cold” that lasts for more than 2 weeks is a sign of rhinosinusitis. That foul stuff is a common trigger for reactive airway, or asthma. If YOUR child has green “nose-pudding” for more than 2 weeks, if they always seem to have it, get them to a boogor doctor.
Bottom line: if your cilia don’t work normally, the result is not pretty.

What to do?

How can you use this information to help prevent these recurrences in your child? While you are waiting to see a doctor, think about the causes of unhappy cilia that we have reviewed. Do daily saline sinus rinses - this will help rinse away pollutants, allergens and other irritants, viruses, bacteria, and mold.

Check these earlier posts – they will help familiarize you with nasal and sinus anatomy and physiology, and help you reduce exposure to toxins and pollutants, and reduce triggers for allergic rhinitis, and help detoxify your air.

Resources (The boogor doctor Blog):

- Allergic Rhinitis CAN Be Controlled – Here’s How
- What is Sinusitis?
- Nasal and Sinus Anatomy and Histology
- STOP BREATHING: Your Air Is Killing You (and what to do about it)
- Saline Sinus Rinses: What Good Are They? Part 1
- Saline Sinus Rinses: What Good Are They? Part 2
- Saline Sinus Rinses: What Good Are They? Part 3
- Saline Sinus Rinses: What Good Are They? Part 4

You might find the tips in those posts helpful, but NOTE: there is NO QUICK CURE for the child with chronic nose pudding (CRS). If you are fortunate you will make your way to a pediatric boogor doctor (ENT – Ear, Nose & Throat, Otolaryngologist) who uses an integrative approach to controlling and preventing CRS for your child.

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Question for you: If you have a child with PCD, how are you optimizing their airway? Please visit my blog and let me know.

Thanks for visiting, and see you here again. I appreciate your comments and questions. Keep 'em coming. Please, "be excellent to one another."

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Stay informed, stay healthy.

Best of health and success to you and your families.

Until next time, remember ... you can pick your friends, and you can pick your nose, but you can't pick your friend's nose (unless you're a boogor doctor :~D)