

# PIDD Patients and Sinusitis

*Understanding the Diagnoses and Treatments*

By Jennifer Kester

To alleviate sinusitis, PIDD patients need to get to the root of its cause, as well as take some additional precautionary measures.

**S**inusitis causes symptoms of acute facial and head pain. Unfortunately, a number of patients see a doctor only to find that their chronic sinusitis is a symptom of primary immune deficiency disease (PIDD) — a diagnosis that offers little solace to those suffering from chronic sinusitis pain. Though each case is different, knowledge about a patient's particular form of sinusitis can empower them, and help to alleviate the recurring bouts of sinus infection that brought them to the doctor in the first place.



## What Causes Sinusitis?

A number of contributing factors are likely behind sinus trouble, but five in particular are worth bearing in mind.

**Gastroesophageal reflux disease (GERD).** GERD is an often-overlooked factor that can cause sinusitis. GERD arises due to acid reflux from the stomach. “Normally, the lower end of the esophagus, as it attaches to the stomach, should be ‘squeezed shut’ by the action of the diaphragm, as [acid] passes through the diaphragm,” says Dr. Terry Harville, medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences. In patients with GERD, acid readily passes into the esophagus. While sitting or standing upright, the acid may not travel very high, but once lying down, it can pass into the oral pharynx. From there, it can readily get into the eustachian tube (the tube connecting the middle ear with the upper part of the pharynx near the nasal passages), especially in younger children, and result in ear infections. The acid can pool on top of the adenoids in the back of the throat and initiate inflammation and infection, which then disrupts the ability of the uvula to properly close off the nose. Then, acid can reflux into the nose and sinuses, or the inflammation and infection from the adenoids can pass into the nose and sinuses. In worse cases, the acid also can flow into the trachea, resulting in lung problems.

To deal with GERD, sometimes aggressive therapy is necessary. Aggressive therapy involves the use of high doses of antacid medications. For example, Harville says, a child taking histamine blocker ranitidine would use 8 to 10 mg per kg divided twice a day, rather than 4 mg per kg, which pushes the dose to a slightly higher level. For an adult who takes a medicine once a day, the prescription may be upped to using it twice daily, and secondary medications may be given to promote gastrointestinal motility. If aggressive therapy doesn’t help GERD, surgery may be the next step. The anti-reflux surgery is known as Nissen fundoplication or just fundoplication. “In essence, it is using the stomach to wrap up around the esophagus, creating a valve to prevent stomach contents from passing back into the esophagus,” Harville says. (See additional information about GERD in this issue’s article,

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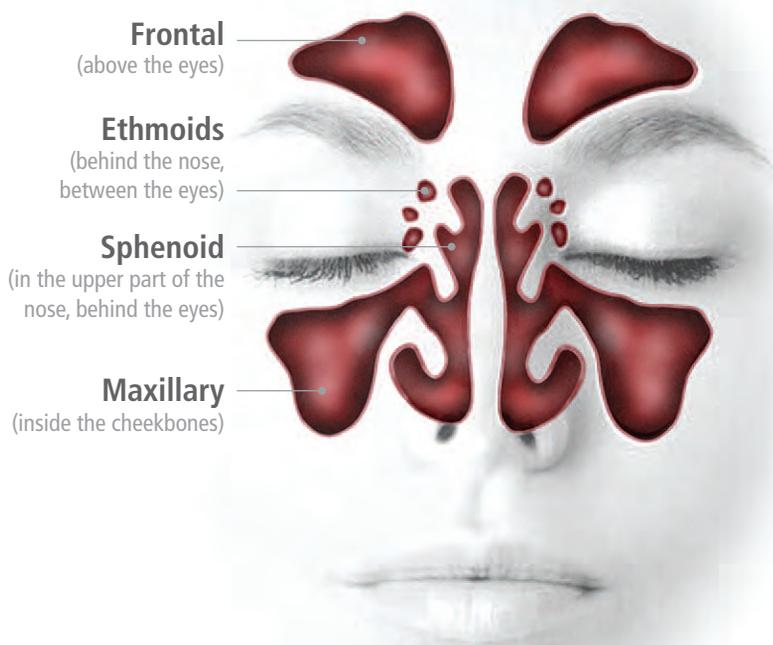
GERD and Coughing: What PIDD Patients Need to Know, on page 22.)

**Structural defect.** Structural defect is another condition that can result in sinusitis. Anatomic problems that prevent the drainage of fluids cause recurring infections. Typical defects include polyps, a growth on a mucous membrane; a deviated septum; abnormally small sinus passages; and enlarged turbinates, which are the thin plates on the walls of the nasal chambers. According to Dr. Alice Lee Kuntz, an otolaryngologist (more commonly known as an ENT—ear, nose and throat specialist) at Bellevue ENT Inc., Bellevue, Wash., larger structural defects such as a deviated septum, enlarged turbinates and large polyps can be detected during a visual exam. But, she says, imaging studies such as a CT scan are needed to take a closer look for narrow passageways, chronic infection, inflammation and smaller polyps.

Surgery may be required to fix structural defects, though the proper course of action will depend on the patient’s history, as well as the nature of the defect. According to Harville, the simplest option is to flush the sinuses, draining these passages while also looking out for organisms that may be causing painful inflammation. If this doesn’t work or isn’t suitable for the patient, doctors may use surgery to create better drainage in the sinus cavities.

“More aggressive forms of surgery actually result in ‘cutting’ of the tissues and possibly bone, for example, to open the sinus openings or to create new openings,” Harville says. “This latter form of surgery may be helpful in adults, but frequently results in more problems in children. We tend to recommend against this latter surgery, because in patients with immunodeficiencies, frequently the tissue never fully heals or recovers, [and] therefore, recurrent procedures are required. In children, the lack of healing is extremely important.” Other surgeries that can be useful, if needed, include the correction of a deviated septum or the removal of enlarged turbinates.

Kuntz warns that when it comes to surgery for children, doctors have to be very selective in the scope and use of incisions to avoid leaving behind surgical wounds. The most important thing for a child who has an immune deficiency, she says, is to treat the immune deficiency as aggressively ➤



as possible before jumping into surgery. Surgery is only helpful, she adds, if there's an anatomical problem, such as a deviated septum that needs correction or enlarged turbinates that may require revision.

**Allergies.** Allergies are also a contributing factor to sinusitis. Patients should work with their doctors to check for any allergies and figure out how to avoid or treat them. If a patient suspects allergies may be exacerbating their condition, a good rule of thumb is to avoid allergens such as pollen, pet dander, mold and dust mites. Their doctor might prescribe some medications to combat these allergies, such as antihistamines, decongestants, corticosteroids and leukotriene modifiers. However, unless patients have had prior sinus surgery, which provides a clearer passageway to sinuses, Kuntz says that nasal rinses won't treat the infection itself. But even without a previous surgery, nasal rinses are still helpful for washing irritants that are known to trigger sinus infections, such as allergens, out of nasal passages.

**Trough levels.** Patients who suffer from chronic sinusitis should have their doctor regularly check their trough levels, the amount of immunoglobulin (IG) in their blood right before their infusions, to see if they are at the proper IVIG (intravenous immunoglobulin) dose. After an infusion,

these levels will spike, but IG levels then steadily decrease until the patient's next infusion.

According to Harville, higher infection rates can correlate to lower trough levels. "In growing children, the dose should be adjusted often to accommodate growth, and troughs should be used again to demonstrate sufficient dosing—as long as there are no infections." However, Harville warns that checking trough values and maintaining higher levels of IG may be deceptive. Ideally, he says, trough levels should be about 1,000 mg/dL, which is a departure from the previously accepted notion that a trough greater than 400 mg/dL was adequate. "The trough should be demonstrated to indicate that [a] sufficient amount of IVIG is being given, but should not be used to demonstrate that it is 'adequate,' since some patients require trough values of 1,000 mg/dL to control sinus disease," he says. "If chronic infections are present, the dose of IVIG should be increased, and antibiotics used to control the infections, but the trough should not be used to demonstrate 'adequacy' of treatment. Indeed, if the trough is very good, the dose and dosing interval seem reasonable, and appropriate antibiotics are being used, then one should be looking for things like GERD, fungal infection in the airways, tracheal esophageal fistula, cleft palate—especially submucous cleft—ciliary dysfunction, etc."

**Cilia disease.** Another common cause of chronic sinusitis in PIDD patients is cilia disease or dysfunction. The cilia are microscopic hairs on the nasal membranes that act like oars to drain the mucous produced naturally by the body. Some viruses, exposure to cigarette smoke and other inflammatory processes can paralyze and even destroy cilia. When this happens, the excess mucous gets trapped in the sinuses, which leads to sinus pain and headaches.

Harville says that ciliary dysfunction is evaluated by carefully scraping cells from inside the nose and examining them under a light microscope to see whether a uniform and appropriate beat pattern—that is, the hair-like cilia sway in a repetitive cycle—is present, and then examining a specimen by electron microscopy to determine if the internal structure is normal. "Typically if there is true ciliary disease, there is the condition of situs inversus, or the internal structures of the body, including heart, lungs, liver, intestines, etc., are on the reverse side," he says. "Cilia are necessary

for the normal placement of the organs during development. Therefore, the risk for ciliary disease can be obvious in this setting. Less obvious would be ciliary dyskinesia, where they fail to beat in an appropriate uniform pattern to move mucous—this is why the light microscope exam is needed.”

If ciliary disease is the culprit, IVIG, daily antibiotics, nasal flushing, nebulized antibiotics (such as Tobi) and nebulized anti-mucolytic agents (like Pulmozyme) can all be useful, Harville says. “We may also use devices to rapidly inflate and deflate the lungs or vibrate the body, to attempt to break up mucous and help with its removal from the body with postural drainage, i.e., [putting the] head lower than [the] rest of [the] body.”

**Other causes.** The National Institutes of Health asserts that the above five conditions may not be the only causes of sinusitis. Other factors that will make PIDD patients susceptible to recurring sinus infections include air pollution and smoke; asthma; changes in altitude (such as flying or scuba diving); dental work; sticking a foreign body into the nose; frequent swimming or diving; hospitalization, especially if in the hospital because of a head injury or if a tube has been placed into the nose (a nasogastric tube); pregnancy; and overuse of nasal decongestants. Pay special attention to nasal decongestants. According to the National Institutes of Health, nasal decongestants may help at first, but using them beyond three to five days can actually worsen nasal congestion. Harville adds that injuries to the nose and face—whether they are from boxing, football, car accidents and so on—are also risk factors for chronic sinusitis.

## How to Avoid Sinusitis?

Patients can take some measures to keep their sinuses clear of infections, but they should be sure to get approval from their doctor before trying them. Getting proper and consistent sleep; maintaining a good diet; avoiding alcohol and triggers; drinking lots of fluids; and washing hands well are all common-sense steps to take. But, patients also may want to look into using a saline nasal rinse; making use of a steroidal nose spray or a nose spray with antibiotics; using a nasal antihistamine such as Astelin; getting nasal respiratory treatments; using humidifiers to prevent sinuses from getting too dry; taking prophylactic antibiotics; employing a daily proton pump inhibitor (if affected by GERD); utilizing H2 antagonists, such as Zantac; and taking Afrin a half hour before take-off when traveling by plane.

When patients are in the throes of an infection, Harville says the following might help provide relief (of course,

## Signs of a Sinus Infection

Sometimes it can be hard to tell whether sniffles are just a cold or a full-blown sinus infection. Most cases of sinusitis begin with a common cold. But those colds can inflame sinuses and cause sinusitis. Sinusitis can be acute, when it lasts two to eight weeks, or chronic, when the symptoms aggravate for much longer. According to the National Institutes of Health, these symptoms include:

- Bad breath or loss of smell. Many parents with kids who have PIDD recognize this as the first sign. The kids’ breath usually has a unique fetid odor.
- Cough. The cough is often more severe at night.
- Fatigue and general ill-feeling.
- Headache. Pressure-like pain behind the eyes or forehead, a toothache, or other facial tenderness point to sinusitis.
- Nasal congestion and discharge.
- Sore throat and postnasal drip. The postnasal drip usually consists of thick secretions that are yellow, green or spotted with blood.

patients should consult a doctor first to determine if these are appropriate for their condition): a nose spray with antibiotics; a steroidal nose spray; a nasal antihistamine nose spray; antibiotic ointment Bactroban Nasal; nebulized antibiotics; or oral antibiotics. In severe cases where bone is involved, IV antibiotics could become necessary. Kuntz says that she reserves nasal antibiotics for patients who have had sinus surgeries. As with nasal rinses, surgeries create easier passageways to the sinuses for the medicine. Without this, she says, the antibiotics will reach only the nasal passage, not sinus cavities, where the infection resides.

Ultimately, patients’ best defenses may be to have an ENT (not a primary care physician) take a culture to find out what, if any, kind of bug is growing to better diagnose the course of treatment needed. While a primary care physician can take a nose swab, that method doesn’t get at the bacteria causing the infection, explains Kuntz. ENTs, on the other hand, have special equipment to get past the turbinates and into the sinus area to get an effective culture. ■

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