As discussed by Kristin Riekert, PhD, a first hospitalization can sometimes be a turning point that may lead some patients to realize the potential seriousness of their illness and the need to take responsibility for their care. It is questionable, however, whether Ryan perceived his hospitalization as a serious health event, because his main recollection of the hospitalization was associated with him not being able to attend his high school’s basketball playoff game with his friends, something he still reminds his caregivers about.

Ryan is currently living with his parents and is in his first semester at the local university, where he is studying engineering. Ryan’s last clinic visit was nearly 5 months ago; he skipped a scheduled appointment because of a school conflict and has been too busy with his course work to reschedule. Recently, Ryan’s parents had noted a significant increase in his cough, particularly at night and during his morning shower, and scheduled a clinic appointment for him. During his visit, Ryan’s FEV1 was approximately 75% of predicted, which was somewhat lower than his baseline range. Ryan found the whole visit somewhat irritating and emphatically stated, “I’m fine—the cough bothers my parents more than it bothers me.” His focus during the visit was on the inconvenience of coming to clinic given his busy school schedule.

CHOOSING A TREATMENT PROTOCOL

Experienced CF caregivers will recognize the challenge of caring for a patient who is convinced that he has minimal CF lung disease and is dismissive of his current symptoms. These attitudes may often persist, even in the face of obvious symptoms (ie, increased cough and sputum production) and a decline in pulmonary function tests. If one were to follow published pulmonary treatment guidelines in Ryan’s situation, several initial treatment options might be considered, including treatment with an oral antibiotic along with his prescribed inhaled tobramycin and other medications; an oral and inhaled antibiotic along with hypertonic saline; or perhaps an oral and inhaled antibiotic, hypertonic saline, and an extra session of airway clear-
ance each day. Oral antibiotics, such as fluoroquinolones (eg, ciprofloxacin), are often used to treat pulmonary exacerbations in patients who are chronically infected with *P aeruginosa*.[1-3] As discussed in the review of the pulmonary treatment guidelines, many treatment options exist. Inhaled tobramycin and dornase alfa have been demonstrated to improve FEV1 when used chronically in appropriate patients with CF. Chronic use of inhaled tobramycin reduces exacerbations in patients aged 6 years and older who are asymptomatic or have mild lung disease. Chronic use of oral azithromycin has also been demonstrated to reduce the frequency of exacerbations in individuals with CF and *P aeruginosa* infection, with some evidence indicating a potential benefit even in *P aeruginosa*-negative patients. Hypertonic saline inhalation may be used to improve mucociliary clearance by increasing hydration of airway surface liquid, and has been demonstrated to reduce the frequency of exacerbations1 (see article by John Paul Clancy, MD, for more information on the CF pulmonary guidelines).

**PATIENT EDUCATION ON ADHERENCE AND COMPLIANCE**

In Ryan's case, a key factor to recognize is that regardless of the selected treatment regimen, the patient is at a very high risk for intelligent nonadherence, particularly if the regimen is complicated. As discussed by Dr Riekert, intelligent nonadherence is a typology where the patient deliberately avoids, alters, or discontinues therapy as a result of an underlying belief that therapy: (1) will not significantly impact health; (2) is not really required; or (3) may result in an unwanted side effect (see Dr Riekert's article for more information). Individuals with intelligent nonadherence often make a decision to be nonadherent while believing that they are making a sound decision about maintaining their health. With Ryan, prior to adding more medications to his current regimen in an effort to address his increasing symptoms, his caregivers decided to specifically address his adherence to his current medications. In assessing his adherence, Ryan was asked directly which, if any, of the prescribed medications he had been taking. Upon further discussion, it was discovered that Ryan's actual regimen consisted of enzymes, vitamins, and azithromycin, with only rare use of dornase alfa, and no use of inhaled tobramycin since his hospitalization last year. He reported that his main recent intervention to improve his health was attempting to go to the gym more often, which is consistent with his belief that exercise was the main way for him to stay healthy. Ryan's approach is classic for Dr Riekert's description of intelligent nonadherence: he considers many of his medications to be unnecessary given his "mild" CF lung disease and absence of major symptoms. Ryan also believes that the prescribed therapy is not the most effective way to maintain his health.

The strategies used to address Ryan's nonadherence were specific to those used for individuals falling into the intelligent nonadherence typology: (1) involving Ryan in the decision-making process for his medical regimen; (2) providing personalized feedback on the relationship between his adherence to medication and health outcomes; (3) providing CF education; and (4) linking therapy with personal goals. For Ryan, the most important aspect of CF education was to address his perception that he had such mild CF lung disease that he did not really need chronic medications. Ryan was shown a graph that included a plot of his FEV1 values for the last 5 years—which highlighted the fact that his lung function had declined during this time period, despite having relatively good lung function overall. In addition, he underwent a chest computed tomography (CT) scan during the clinic visit, and the details of the images were reviewed with him. These images demonstrated the presence of bronchiectasis, mucus plugging, and cystic changes in his lungs, all of which he was able to see for himself. This led to a somewhat eye-opening experience for Ryan, during which he realized for the first time that he had significant structural lung disease.

Ultimately, Ryan was prescribed a course of oral ciprofloxacin, and proper use of inhaled tobramycin (as previously prescribed) was reviewed in detail. Ryan was also convinced to take part in a “Ryan experiment,” where he would assess his symptoms and FEV1 before and after the use of his medication so that he could see the potential benefits of adhering with treatment. Ryan was scheduled to follow up in 3 weeks for a re-evaluation of his clinical status and for further support and education on the importance of treatment adherence.

**FOLLOW-UP**

Ryan returned to clinic in 3 weeks with obvious signs of improvement. His FEV1 had increased to 80% of predicted and he reported having much less cough and sputum production. He kept a copy of the CT scan image that was provided for him, and acknowledgments.
Acknowledging referring back to it occasionally when he needed to recall the importance of making his treatment a higher priority.

As caregivers, our initial tendency is often to add more medications to a patient's regimen when we see a decline. In Ryan's case, however, just adding several more medications to his regimen would have been a poor decision, because it would have only resulted in a longer list of medications that he was not taking. Instead, by addressing some of Ryan's underlying attitudes and providing him with an opportunity to participate in treatment decisions, his caregivers were able to help Ryan make a significant change, both in his current symptoms and in his chances for future adherence.

REFERENCES

