Chapter 2
Functional Aspects of an Organic Respiratory Disorder: Cystic Fibrosis

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Abstract The management of respiratory diseases with organic causes often can be complicated by the emergence of concurrent functional respiratory symptoms. Thus, clinicians must be alert to the possibility that functional symptoms may develop in this setting. For example, anxiety with associated dyspnea can affect the management of many patients with significant respiratory disease such as asthma, chronic obstructive pulmonary disease, and cystic fibrosis (CF). An understanding of functional aspects of respiratory disorders will aid the clinician in identifying the correct cause of symptoms. In turn, this will help guide the application of appropriate therapy, thus preventing unnecessary tests and treatments that may compound an existing medical problem or create a new one. As an example illustrating how clinicians can approach patients with a respiratory disease that can have both organic and functional components, this chapter focuses on patients with CF. Case studies are presented through which the clinician will gain a better appreciation for the interaction of functional and organic causes of symptoms in this setting.

Keywords Adherence • Anxiety • Cystic fibrosis • Depression • Dyspnea • Habit cough • Insomnia

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Background

The management of respiratory diseases with organic causes often can be complicated by the emergence of concurrent functional respiratory symptoms. Thus, clinicians must be alert to the possibility of the development of functional symptoms in this setting. For example, anxiety with associated dyspnea can affect the management of many patients with significant respiratory disease such as asthma, chronic obstructive pulmonary disease, and cystic fibrosis (CF). An understanding of functional aspects of respiratory disorders will aid the clinician in identifying the correct cause of symptoms. In turn, this will help guide application of appropriate therapy, thus preventing unnecessary tests and treatment that may compound an existing medical problem or create a new one.

As an example illustrating how clinicians can approach patients with a respiratory disease that can have both organic and functional components, this chapter focuses on patients with CF.

For readers who are not familiar with CF, the first part of this chapter provides a brief introduction to the clinical presentation of this chronic and eventually fatal illness, with a special focus on symptoms and signs that can overlap with the clinical presentation of functional symptoms.

The second part introduces functional conditions that have been described particularly affecting patients with CF. This part places emphasis regarding symptoms that can arise as a result of these functional conditions that potentially overlap with those caused by the physiological abnormalities encountered in CF.

The third part discusses how clinicians might differentiate functional from organic symptoms in patients with CF and how to effectively balance evaluation and treatment of organic and functional symptoms in this population. Case studies are presented through which the clinician will gain a better appreciation of the interaction of functional and organic symptoms in patients with CF.

Key Organic Features of Cystic Fibrosis [1–11]

General Considerations

CF is the most common, fatal, hereditary disorder of Caucasians. The disorder also is common in Latinos and Native Americans. It is less common in Blacks and Asian Americans. CF affects the exocrine or secretory glands that normally produce mucus, the thin, watery, and slippery substance that coats the lining of the mouth, nose, sinuses, throat, lungs, and gastrointestinal tract. In CF, this mucus is abnormally thick and sticky. Rather than functioning as a protective lubricant, CF mucus builds up and blocks the passage ways of the lungs, pancreas, and reproductive system. This blockage encourages the growth of bacteria associated with chronic infections and inflammation of the lungs, ultimately leading to irreversible lung

damage, respiratory failure, and an early death from pulmonary complications in 90% of the patients. Other manifestations include pathognomonically high sweat chloride levels, life-threatening respiratory complications, pancreatic dysfunction, a number of other serious digestive problems, and fertility issues.

Causes and Incidence

CF is an autosomal recessive disorder caused by a mutation of the CF transmembrane conductance regulator (CFTR) protein, which leads to abnormal ion transport. Autosomal recessive inheritance means that in this case, both parents carry a single copy of the CF mutation and each of their children has a 25% chance of receiving a copy from both parents. Having two copies of the CF mutation leads to development of the disease.

CFTR functions as a channel across the membrane of cells that produce mucus, sweat, saliva, tears, and digestive enzymes. The channel transports negatively charged chloride ions into and out of cells. The transport of chloride ions helps control the movement of water in tissues, which is necessary for the production of thin, freely flowing mucus. The CFTR protein also regulates the function of other channels, such as those that transport sodium ions across cell membranes and which are necessary for the normal function of organs such as the lungs and pancreas.

To date, approximately 160 gene mutations have been identified as causing clinical CF. (Most of the approximately 2,000 mutations in the CFTR protein are not thought to cause disease.) Nearly 90% of Americans who have CF carry at least one copy of the delta F508 gene. CF affects approximately 30,000 Americans and an additional 40,000 individuals worldwide. In United States Caucasians, about 1 out of 3,200 individuals has CF.

Clinical Findings [2]

Respiratory tract symptoms include coughing, dyspnea (shortness of breath), increased mucus in sinuses, sinus pain or pressure, sinus polyps (overgrowth of sinus tissue), sputum production from the lungs that can cause lung crackles, recurrent episodes of pneumonia, bronchiectasis (damaged airways), atelectasis (small areas of collapsed lung tissue), hemoptysis (coughing up blood), and pneumothorax (air leak from the lungs that can cause collapse of a large part of a lung).

Gastrointestinal manifestations may include pancreatic insufficiency (90% of patients) that leads to malabsorption (especially of fat) associated with steatorrhea (fat in the stools), pancreatitis, nausea, loss of appetite, weight loss, malnutrition, abdominal pain, bloating, gas, constipation often from distal intestinal obstruction syndrome, and meconium ileus (obstruction of the intestine due to overly thick fecal material) at birth.

Infertility is present in almost all men as result of congenital absence of the vas deferens with azoospermia. Women have thickened cervical mucus that results in a 20% decrease in fertility rate.

Other findings and complications include digital clubbing, delayed puberty, diabetes (which affects 50% of those over 30 years old), osteoporosis, arthritis, biliary cirrhosis, liver failure, gall stones, and rectal prolapse.

Patients with *advanced disease* may present with hypoxemia (low levels of oxygen), hypercapnia (high levels of carbon dioxide), and cor pulmonale (heart dysfunction that arises because of the severe lung disease).

Diagnosis

Newborn screening for CF accounts for most patients who currently are being diagnosed. The diagnosis is confirmed by genetic testing and/or a test of the salt content of the sweat.

Treatment

The goals of pulmonary treatment are to decrease airway mucus viscosity so that it can be coughed out more easily, improve clearance of sputum through provision of chest physiotherapy, decrease airway inflammation, and prevent and treat lung infections. The goals of gastrointestinal treatment are to replace pancreatic enzymes and vitamins and to provide aggressive nutritional support and therapy to prevent liver disease, if indicated.

Commonly Used Medications

- Recombinant human DNase (dornase alpha), nebulized, to thin sputum (thereby acting as a mucolytic)
- Hypertonic saline, nebulized, to increase airway hydration and induce cough
- Inhaled bronchodilators to aid in opening the airways
- Antibiotics to treat airway infection either when needed or preventively
- Pancreatic enzyme replacement to aid the absorption of fats and proteins
- Vitamin supplements, especially of fat soluble vitamins A, D, E, and K

Therapeutic Procedures

- Mechanical interventions to clear lower airway secretions include manual chest physiotherapy consisting of chest percussion and postural drainage, high

frequency chest wall oscillation with a mechanical vest, and use of positive end expiratory pressure.
- High calorie, high fat, and high protein diet.
- Exercise—regular aerobic and strength training.
- Drink plenty of fluids in order to maintain hydration.
- Avoid—smoke, dust, dirt, fumes, household chemicals, mold, and mildew.

Surgery

Some patients require insertion of a gastrostomy feeding tube because of their malnutrition. Lung transplantation is an option for patients with CF who have end-stage lung disease. At this time, the 5-year survival following transplantation is approximately 50%.

Prognosis

Most children remain relatively healthy throughout school and finish college and/or find employment. The mean life span of a CF patient is 38.3 years [4], which is a remarkable improvement over the past few decades and is attributable to the introduction of many new therapies for the disease.

Functional Conditions Associated with Cystic Fibrosis

Many psychiatric and psychosocial factors can develop as patients with CF become older, which affect their clinical presentation and quality of life, as well as the appropriateness and efficacy of their treatment. For example, adolescents with chronic illnesses are at increased risk of developing adjustment problems and internalizing symptoms of depression and anxiety. Thus, patients presenting with CF may exhibit concurrent behavioral problems that cause physical symptoms such as abdominal discomfort, muscle tension, shortness of breath, and/or tremors (e.g., as a result of anxiety). Other behavioral issues can affect adherence to the prescribed medical therapies. While treatment for CF necessarily focuses on the medical condition, the underlying psychological factors should be addressed simultaneously in order to achieve optimal results.

Many primary care or pulmonary specialists fail to consider psychological evaluation of their patients because they are unfamiliar with how to diagnose psychiatric conditions or how to assess the psychosocial impact on their patients' clinical presentations. Further, children and adolescents, who may have difficulty clearly describing their symptoms, may be difficult to diagnose with a psychological disorder. Thus, this section reviews the diagnostic criteria for the psychiatric disorders and psychosocial considerations commonly associated with CF.

Psychiatric Considerations

Psychopathology within the CF community has been reported to be as high as 57% with a diagnosable psychiatric disorder [2]. The current basis for assessment of psychiatric disorders is the Diagnostic and Statistical Manual, fourth edition, Text Revision (DSM-IV-TR) that contains a chapter regarding Mental Disorders Due to General Medical Conditions. The criteria for such disorders include that the disturbance is the result of a physiological consequence of a general medical condition such as CF, is not better accounted for by another mental disorder, and does not occur exclusively during the course of delirium. Indications that a psychiatric disorder may be caused by a medical condition include acute onset without apparent cause; correlation between the onset, exacerbation, and remission of psychiatric symptoms with the course of the medical condition; and atypical features, such as aggression with anxiety.

Notably, DSM-IV-TR also recognizes that psychological factors can affect medical conditions (see Chap. 1). For example, generalized anxiety disorder (GAD) is a diagnosis commonly associated with a chronic respiratory disease such as CF that can affect its clinical presentation. GAD can cause worry, restlessness, fatigue, loss of concentration, irritability, muscle tension, and sleep difficulties. All of these behavioral signs and symptoms should be considered when evaluating a patient for GAD or assessing for other possible mental health disorders.

Although a thorough psychiatric assessment including mental status and neurological examination is ideal, it is rarely practical or immediately necessary, if the clinician has a basic knowledge of behavioral symptoms. For example, a patient might be suspected of having a psychiatric disturbance based on complaints of feelings of worthlessness and fatigue. A diagnosis can be made through consultation with a mental health care professional who is clinically experienced with patients who suffer from medical conditions.

Anxiety

A patient with CF reports experiencing nightly insomnia for 2 months after being discharged following a recent hospitalization for a respiratory exacerbation. He states that at night he develops nightmares about being readmitted to the hospital and during the day he worries about contracting respiratory infections that might trigger yet another exacerbation. Sometimes, when he focuses on his worries, he develops shortness of breath, chest tightness, dizziness, and paresthesia.

The most common mental health diagnosis for children and adolescents with CF is anxiety, which afflicts as many as 35% of school-aged children with CF. Forms of anxiety in CF include separation anxiety disorder (25%) GAD (15%), phobias (7.5%), obsessive compulsive disorder (5%), and panic disorder (4%) [12]. Notably, individual patients may suffer as a result of more than one type of anxiety at the same time.

Anxiety affects 30% of adolescents with CF, which makes it the most common psychiatric disorder in this age group, as well [2]. Anxiety in adolescence often is related to the patients' developmental emphasis on peer bonding, autonomy, conflict, and sexual exploration [13]. Paradoxically, anxiety has been associated with improved adherence to prescribed treatment because of adolescents' preoccupation with the idea that symptoms from their illness may adversely affect their peer relationships [2].

Parents of children with CF have been found to develop anxiety, worry, sadness, and hypervigilance when their children have been diagnosed with CF, even when the diagnosis was made following prenatal screening [14]. Caregivers of preschoolers with CF have been documented to have anxiety (16.4%) and depression (33%) associated with their children's sleep, eating, adherence, and mealtime issues [15, 16]. Parents of school-aged children demonstrated increased rates of anxiety (50%) often related to concerns over treatment adherence [16]. Similar issues are evident with parents of adolescents, who may find themselves in conflict with their children's desire for autonomy that can lead to risky behaviors and compromised health care [17].

Importantly, parental anxiety may adversely affect their children's mental health and treatment for CF. For example, sometimes children's anxiety can develop or intensify as a result of their parents' anxiety. Conversely, families who function well, as demonstrated through measures of cohesiveness, expressiveness, and organization, have been associated with psychological well-being in their CF adolescents [18].

Symptoms of anxiety that can overlap with organic issues in CF include dyspnea, chest pain, difficulty with swallowing, abdominal discomfort, tremor, and insomnia.

Depression

An adolescent with CF comes to your office complaining of feeling constantly sad and guilty about the death of her brother, which occurred over a year ago as a result of his CF. She explains that she feels as if she were the one who should have died since he took all of his therapies, while she frequently skipped therapies. As a result of her sad feelings, she reports that she has been unable to focus on her schoolwork and her grades have suffered. She also is concerned because some of her friends have left her because they told her it is no longer fun to be around her. She has experimented on one occasion with snorting cocaine to help her feel better.

The second most common mental health disorder in children and adolescents with CF is depression, which affects as many as 29% of patients in both age groups, as assessed by the children's depression inventory (CDI) [19]. Parents and caregivers of recently diagnosed children with CF have been found to have at least mild depressive symptoms at a frequency as high as 44% in mothers and 33% in fathers using the Beck Depression Inventory (BDI) [20].

Major depression is known to adversely affect cognition, attention, energy, and motivation, which leads to a decrease in task completion, adherence to medical regimens, missed clinic appointments, increased at risk behaviors (smoking and drinking), eating disturbances, poor body image, and decreased quality of life [21, 22]. Depression has been shown to have an impact on the daily functioning of CF patients and their caregivers [23]. Parent and child depression has been shown to correlate with worse adherence to airway clearance in school-aged children and adolescents [24]. Depression in the parent/caregiver has been associated with a decreased adherence to use of pancreatic enzymes in their children, with resultant poor weight gain [25].

Health-related quality of life (HRQOL) measurements on standardized screening tools like the cystic fibrosis questionnaire-revised (CFQ-R) were worse in CF patients with depressive symptoms. Those with moderate to severe lung disease and depression had poorer scores than those with severe lung disease without depression, which supports the hypothesis that depression heightens the effects of chronic illness such as CF [26]. Depressed CF patients also had decreased HRQOL scores on body image scales, eating problems, and emotional functioning [22].

Symptoms of depression that can overlap with organic issues in CF include fatigue, malaise, loss of appetite, weight loss, insomnia, night awakenings, and an increase of any CF-associated symptom as a result of poor adherence to prescribed therapy.

Comorbidity of Anxiety and Depression

A 17-year-old with CF tells you that over the past year, he has felt more breathless, keyed up with muscle tension, and in the past 2 months has had recurrent thoughts of death.

In the general US population, anxiety and depression have been reported as occurring in 25–29% of all children and adolescents [27]. Patients in medical settings have been found to have comorbidities at frequencies as high as 75% [28]. In CF adolescents, strong positive associations have been found between anxiety and depression [29], although there has been scant research on this relationship in CF [29].

No systematic studies have been undertaken to determine what specific effects mental illness has on CF health care outcomes [30]. However, patients with chronic lung diseases and anxiety or depression have been noted to have increased symptoms and functional impairment, including adverse effects on cognition, attention, energy, and motivation that lead to decreased task completion [21, 31].

Evaluation of anxiety and depression may initially be improved by the implementation of annual, routine, standardized screening procedures, such as the Hospital Anxiety and Depression Scale [32], or the Revised Child Anxiety and Depression Scales [33], which are useful with children and adolescents, and the Beck Anxiety and Depression Inventories [34, 35] or the Multidimensional Anxiety Questionnaire [36], which are applicable to adult patients and caregivers [30]. These

tests are simple and quick to take and score. Such screening followed by treatment improves outcomes especially when offered at specialized CF centers [23].

Treatment for preschool children with mild to moderate depression is recommended to be family therapy [37]. First-line treatments for school-aged children with anxiety and depression include behavioral interventions such as interpersonal or cognitive behavioral therapy (CBT, Chap. 9) [37–43]. Interpersonal therapy (IPT) is a diagnosis-targeted, time-limited, present-focused treatment that makes a practical link between the patient's mood and disturbing life events (such as bereavement, loss of social support, or life transitions) that either trigger or follow from the onset of a mood disorder [40]. IPT application may be of particular value during the transition from adolescence to adulthood [41]. CBT and family therapy have been shown to help improve short-term knowledge and, combined with behavioral interventions, aid nutrition and growth [42].

Psychopharmacological medication also has been used in patients with anxiety and depression (Chap. 14) [44, 45]. Use of psychotherapy in conjunction with medications may be a more effective treatment than either alone [38].

Mood Disorders

A young adult with CF confides that she has not been adherent to her CF treatment regimen. She reports that she has been feeling much better by staying up all night. She has not been tired during the day for over 2 weeks. She is going to the race track daily in order to bet on the horses with the expectation of winning a large race and then quitting her job.

Mood disorders encompass not only depression but also bipolar, dysthymic, and cyclothymic disorders. The differential diagnosis includes borderline personality disorders and mental disorders due to a general medical condition [46]. Although the initial functional presentation may appear to be related to depression, bipolar disorder should be suspected if the constellation of symptoms includes inflated or irritable mood, decreased need for sleep, pressured speech, inflated self-esteem, and increased psychomotor agitation, lasting a week or more. Such a modification in diagnosis will dramatically change the treatment regimen.

Symptoms of a mood disorder that can overlap with organic issues in CF include irritability, insomnia, and increase of any CF-associated symptom as the result of poor adherence to prescribed therapy.

Somatoform Disorders

Your 32-year-old patient with CF who been stable clinically for a number of years reports that he is concerned he has had increased mucus plugging in his lungs with

excruciating associated chest pain, although objective findings including his physical examination, pulmonary function testing, and chest x-ray have not changed in over a year.

Somatoform disorders can be defined as a history of physical complaints without identified medical abnormalities. An overview of somatoform disorders has been presented in Chap. 1 and summarized in Table 1.1.

Somatization disorder should be considered when a patient experiences pain in four or more body areas or functions. Patients with CF often report pain in association with their disease, including 59% of children and 89% of adults [47, 48]. For example, patients frequently report chest pain associated with mucus plugging or recurrent lung infections. In patients with CF, somatization disorder may be associated with repeated hospitalizations due to personal or family issues, poor living conditions, or parental psychiatric conditions [49].

Pain disorder is another somatoform classification in which the patient presents with pain in one or more sites, e.g., chest pain [50]. This pain may or may not be related to a CF-related organic abnormality. For example, pain disorder may occur in the lungs simply due to the phobic anxiety of developing recurrent pneumonia when a minor unrelated chest pain occurs.

Conversion disorder occurs when a patient has a psychologically based motor or sensory symptom or deficit that is unintentionally produced and may be preceded by conflicts or stressors [46].

Hypochondriasis or the preoccupation with fears of having a serious disease despite nonsupportive medical evaluation has been reported in adults with CF [51].

Symptoms of somatoform disorders that can overlap organic issues in CF include cough, stridor, or dyspnea related to conversion symptoms; reports of pain in the lungs, heart, pancreas, and joints; gastrointestinal symptoms such as nausea and vomiting; and loss of libido.

Sleep Disorders

A 24-year-old with CF is transferring to you from another CF center, where he had just been started on albuterol prior to administration of hypertonic saline. He has developed anxiety, tremor, and insomnia and wonders about the origin of these symptoms.

It is well recognized that patients with CF have disrupted sleep histories [52–56]. Contributors to CF sleep abnormalities may include the use of certain medications such as β_2-agonists before sleep [53], vitamin deficiencies [54], chronic anxiety, and depression [55]. Other underlying etiologies of sleep disturbances in CF include nocturnal coughing, obstructive apnea as a result of sinus disease or enlarged tonsils and adenoids, increased work of breathing due to progressive lung disease, and periodic leg movements. By the time they develop end-stage lung disease, patients' sleep also can be disrupted by hypoxemia and hypercapnia [52].

A sleep disturbance in a patient with CF can cause exacerbation of anxiety, depression, irritability, and attention deficit hyperactivity disorder (ADHD), all of which can lead to increased nonadherence to prescribed therapy and poorer medical outcomes.

As a sleep disturbance in CF can be the result of organic and/or psychological factors, both possible etiologies must be evaluated and addressed.

Eating Disorders

An adolescent girl with CF comes to your office complaining of an intolerable stomach ache. You ask her about the regularity of her treatment regimen and she says that she strictly adheres to the treatment guidelines. You notice she is tugging at her slacks and commenting that she is concerned that she is gaining weight. She explains that she wants to remain slim in order to make the cheerleading squad.

Nutritional issues are of concern for patients with CF as they usually require a high calorie, high fat diet. Adherence to this dietary regimen is limited in preschool CF children, 32% of whom have been reported to have eating problems [56]. Nutritional difficulties are of particular concern in the treatment of adolescent girls who tend to have poorer adherence to nutritional recommendations [57] because of their heightened awareness regarding how others perceive their appearance. Notably, there are no reports that CF predisposes to the development of anorexia nervosa or bulimia nervosa [58].

Symptoms of eating disorders that can overlap with organic issues in CF include loss of appetite, nausea, emesis, abdominal discomfort, and weight loss.

Oppositional Defiant Disorder

A 12-year-old with CF has begun smoking cigarettes and refuses to cooperate with airway clearance and nebulized medication treatments. He complains that his treatments take too much time. He continues to smoke and remain noncooperative with his therapies even though he acknowledges the adverse health consequences of his behaviors. He says he is not interested in being rewarded with small financial prizes for improved adherence to his therapies.

Of children 7–12 years old with CF with a parent-reported behavior problem, 62% met the criteria for a DSM diagnosis based on clinical interview. Diagnoses of anxiety and oppositional disorder were most frequent [59] with a prevalence of 22.5% for oppositional defiant disorder [12]. Oppositional problems involve children and adolescents and can progress to conduct disorder, which in adulthood can manifest as antisocial personality disorder.

Patients with CF often oppose the pervasive intrusion into their lives of their demanding treatment regimen. Their extensive prescribed therapies require time out of their daily schedule, remind them that they are different than their peers, and can reveal their condition to others. Opposition to therapy often results in conflicted relationships with parents, especially in combination with the usual child and adolescent drive to separate from parental influence and become increasing reliant on peers [49]. Further, symptoms resulting from CF usually worsen with age, which adds to the burden of therapy and associated conflict regarding its use.

Attention Deficit Hyperactivity Disorder

A teenager with CF becomes distracted easily while doing his therapies and therefore curtails their use regularly. He often forgets to take his pancreatic enzymes, cannot remember where he put his medication containers, and has been unable to follow through on recommendations to organize his medications.

His older young adult sibling, who also has CF, complains about his employer's blatant lack of regard for his medical condition by not allowing him to take breaks in order to follow his treatment regimen. He jumps up and begins pacing and yelling about his mistreatment and the life-altering effects of his condition.

ADHD has been reported as being a common comorbidity in patients with CF [60–62]. This condition can be confused with oppositional defiant disorder as a cause of persistent, angry, resentful behaviors in which patients are easily annoyed by others, lose their temper, and blame others. It has been suggested that stimulant, non-stimulant, and combination ADHD treatments for patients with CF can help improve adherence to their prescribed therapies [61, 62].

This disorder, which was originally considered a child and adolescent condition, is known to carry over into adulthood.

Post-traumatic Stress Disorder

A 30-year-old woman who just was discharged from the ICU following a life-threatening exacerbation of her CF appears distressed in your office. When questioned, she admits to recently avoiding her treatment regimen, even though she has a long history of treatment adherence. She reports recurrent nightmares about her experiences in the ICU, from which she awakens profusely sweating, and is unable to fall back asleep for "hours."

Post-traumatic stress disorder (PTSD) has been shown to occur after extensive or traumatic hospital stays [47, 63, 64]. Risk factors include length of stay in the hospital or ICU and a history of requiring mechanical ventilation or high level sedative medication. Other factors may involve female gender, younger age, past psychiatric

history, and the number of traumatic and delusional memories [63]. As many as 50% of patients who have been in the ICU have been diagnosed with PTSD [47]. Having a patient with severe symptoms that require serious treatment, such as delivered in an ICU, should alert the treating clinician that the patient is at risk of developing this condition.

Symptoms of PTSD that can overlap with organic issues in CF include insomnia and an increase of any CF-associated symptom as result of poor adherence to prescribed therapy.

Psychosocial Considerations

Gender

A 15-year-old girl refuses to follow a high calorie diet because she says she wants to remain thin and pretty.

Morbidity and mortality rates are higher in females with CF [65–67]. Girls, 10–21 years old, reported significantly more illness-related concerns, including emotional issues, discouragement about treatment, and lower self-esteem [68]. These issues have been associated with behaviors that have been detrimental to their health including poor adherence to prescribed treatment regimens, engagement in cough suppression, and an inadequate fat and caloric intake. The situation is complicated further because poor adherence can lead to further discouragement regarding the effectiveness of therapy and even lower self-esteem [57, 69]. A sense of isolation due to repeated respiratory exacerbations can lead to a fear of early death, depression [65], PTSD, and possible suicidal ideation [70].

Adherence to Treatment

Recently, one of your 17-year-old CF patients has noticeably lost weight and has had frequent pulmonary exacerbations followed by brief hospitalizations. You are concerned and question why. He says that it is winter and he has been unlucky in catching a series of colds. He appears fatigued and seems indecisive.

Current treatments for CF require extensive, consistent, often tedious, and time-consuming interventions that involve complex pulmonary, nutritional, medication, and exercise regimens [24, 71]. Daily treatments may necessitate 2–3 h or more. The consequences of inadequate adherence to prescribed therapies are considerable and associated with more exacerbations, increased hospital admissions, and increased mortality [72]. Overall, the mean rate of nonadherence in children with CF is reported to be between 30% and 50% or more with a trend of adolescents being less adherent than younger children and adults even less adherent [72, 73].

Factors associated with poor adherence include psychiatric, psychological, and emotional considerations [24]. Depression may follow serious chronic illness and lead to compromised adherence. One study indicated that depressed patients were three times more likely than those not depressed to be nonadherent with medical treatment recommendations [74]. As mentioned earlier, adults with CF and depression are known to have impaired lung function and poorer HRQOL [75]. Anxiety, oppositional disorders, and eating disorders can also impact the level of adherence. Therefore, we recommend that nonadherent CF patients be screened routinely for coexisting psychiatric and psychosocial issues. (For more information about screening tools, see Chap. 8.)

Parental and Family Issues

During an appointment with a family with twin adolescent boys who have CF, you notice that the one boy who gets along well with his parents seems well adjusted and content, whereas the boy with oppositional behavior toward his parents is much less open and conversant. He is wearing a teen club jacket, appears bored and irritated, and complaints bitterly about all the wasted time spent on CF treatments while watching his parents carefully for their reaction. He appears short of breath as he speaks. The parents appear worried and anxious.

For children and adolescents, family life is central to their self-esteem and adherence [76] and thus influences their pulmonary health [24]. Positive family relationships were reported to be associated with greater adherence to airway clearance and aerosolized medications [76].

Parental reaction to a diagnosis of CF in their infant often involves development of stress and guilt that can alter the family structure. Affected parents have demonstrated more controlling, serious, and less encouraging behaviors, and their children were less responsive to parental guidance [77].

Among parents of preschool children, as many as 40% blame themselves for their child's CF. Such self-blame has been associated with poorer emotional adjustment and possible anxiety and depression in children and their parents [78]. Families whose preschool children have CF can develop significant difficulties with interpersonal interactions, setting appropriate behavioral expectations and enforcement, and marital role strain [79].

Adolescents with CF whose parents supervise their treatment time have better adherence [80]. For adolescents, peers provide more emotional and companionship support than do their families, and such support may serve as a cushion for negative family interactions [81].

Peer Relationships and Socialization

The troubled parents of the twin teenage boys with CF indicate that the oppositional twin hangs out with his peers and is rarely home, so they are unaware of his level of

adherence. He attempts to hide or deemphasize his CF. He is participating in risky behaviors including smoking and drinking that are adversely affecting his health and quality of life, which ironically makes him even more upset and dependent on his parents.

The importance of peer interactions and socialization increases with age. With children's entrance into school, they begin to compare themselves to peers as a gauge for their identity and sense of social competence. Such comparisons serve to underscore how having CF has made them different from others. Their desire often is to deemphasize the importance of their treatment regimen in order to appear as normal as possible, which often entails keeping their diagnosis of CF as a secret [49]. This may lead to an inability to develop and maintain intimate friendships, which is an essential part of socialization that is necessary for completion of mature development [82].

Adolescence is the time when peers become even more influential as children distance themselves from their parents as part of the process of their identity formation [13].

Teenagers with CF have been found to have a higher rate of risky behaviors as compared to healthy peers [17]. Even given the health hazards, 20% of CF teenagers admitted to having smoked cigarettes [57], as well as 19% of females and 7% of males who revealed self-imposed weight-loss regimens [70].

Differentiation of Functional from Organic Symptoms in CF

As demonstrated in the first two sections of this chapter, many symptoms of patients with CF can be triggered by both organic and functional causes, and often, these occur in the same patient. The question for clinicians is how to tease out organic from functional causes so that the appropriate therapeutic course can be followed.

We believe that as long as organic causes of symptoms are considered and treated, concurrent assessment of their possible functional components can only be of benefit. It should be kept in mind that because of the high frequency of psychological and psychosocial issues in patients with CF, development of functional symptoms should be expected, and therefore, evaluation for such symptoms should be part of routine care for these patients. It should be emphasized that functional symptoms can arise as a result of a primary psychological disorder or as a consequence of the organic illness.

Symptoms that often reflect underlying functional components are listed in Table 1.2. Further, a functional component should be suspected if an organic symptom causes excessive distress or impairment or if the symptom appears to be intentionally produced or feigned.

The following brief case studies demonstrate how functional and organic respiratory symptoms frequently overlap in patients with CF. The reader is invited to reflect on how the cases might have unfolded if the functional contribution to the patients' symptoms had been addressed earlier, as opposed to the possible outcomes if the functional components were not recognized or addressed.

Case 1: 4-Year-Old with Cough [83]

The patient was an almost 5-year-old who was diagnosed with CF following birth with meconium ileus. He developed pneumonia at 3 months of age, and by a year of age, was found to harbor *Pseudomonas aeruginosa* in his airway. Subsequently, he was treated with every-other-month tobramycin by inhalation (TOBI®) and daily dornase alpha. When he was well, he did not cough.

At 4½ years of age, the patient manifested severe coughing after contracting influenza. At that time, he was diagnosed with a second pneumonia and treated with intravenous antibiotics. Two months later, computerized tomography revealed diffuse sinusitis, which was thought to be aggravating his pulmonary disease. Therefore, he underwent sinus surgery. A week after removal of the postsurgical nasal packing, he developed a loud, harsh cough that occurred every other second for hours at a time, while awake. The cough ceased as soon as he fell asleep.

The patient was treated with oral ciprofloxacin and prednisolone in addition to TOBI®, but there was no improvement in his cough. No new abnormalities were documented on his physical examination. His lung function was normal.

Because of the cough, the patient was unable to attend summer camp.

The patient lived with his parents and 6-year-old sister who also had CF. The sister's respiratory status was good, but she had a gastrostomy tube for supplemental feedings.

The family was reassured that the cough would resolve in time. In order to help the cough resolve, it was suggested the patient be told, "Are you sure you need to be coughing?" However, the mother felt ambivalent about this advice because she was aware that coughing is beneficial for patients with CF.

The cough resolved a month later after application of hypnotic imagery, and the mother's reassuring of herself that the patient's cough was not related to his CF and thus its resolution would be safe for her child.

Questions:

1. The differential diagnosis for this patient's persistent cough includes all of the following EXCEPT:

   (a) Chronic sinusitis
   (b) Foreign body in the airway
   (c) Gastroesophageal reflux
   (d) Habit
   (e) Pneumonia

2. All of the following therapies have been reported to be beneficial in the treatment of habit cough EXCEPT:

   (a) Bedsheet wrap technique
   (b) Clonazepam

(c) Hypnosis
(d) Reassurance
(e) Suggestion therapy with inhaled lidocaine

3. How might the mother's attitude regarding the cough affect its persistence?

(a) The child might have sensed the mother's ambivalence about its resolution and acted accordingly.
(b) The mother's concern about the cough increased her own anxiety, which affected the child.
(c) The child might have received additional attention for the cough that helped reinforce it.
(d) All of the above.
(e) None of the above.

Answers:

1. (e): Of the listed diagnosis, the most likely is habit cough, given the disruptive nature of the cough and as it resolved when he fell asleep. Cough associated with (a), (b), and (c) typically would occur during sleep as well. Persistent pneumonia is unlikely, as other symptoms likely would have manifested.
2. (b): There is no indication for clonazepam in this setting. Reassurance usually is ineffective. The bedsheet wrap technique involves the patient tightening a bedsheet around his midsection in order to suppress the urge to cough [84]. Suggestion therapy (Chap. 5) and hypnosis (Chap. 12) have been reported as very effective.
3. (d): Habit cough can persist for all of the listed reasons, as well as in association with various psychosocial stressors such as divorce, loss of a significant other, or difficulties in school. Sometimes, the psychosocial stressors must be addressed in order to permit resolution of the cough.

Case 2: 10-Year-Old with Shortness of Breath [85]

The patient was diagnosed with CF at 2 weeks of life, after he was born with meconium ileus. Complications of his disease included recurrent pneumonia necessitating nine hospitalizations, severe gastroesophageal reflux for which he underwent a Nissen fundoplication at 8 years of age, and renal stones. Despite aggressive therapy, by the time he was 9 years old, he was placed on the waiting list for lung transplantation.

As the patient remained on the transplantation list for 13 months, he became increasingly fatigued, with increased coughing, chest tightness, and shortness of breath. His blood concentration of carbon dioxide was mildly elevated. The burden of his treatments included continual administration of oxygen and assisted ventilation with the aid of tight-fitting nasal prongs (BiPAP) for 2 h in the daytime and throughout the night. He received chest physiotherapy for 30 min, 3–4 times a day. He received three nebulized medications each twice daily, in addition to many pills.

The patient complained that his shortness of breath frightened him. He was asked what changes could be made to make his life more tolerable. The patient stated that he had nothing to look forward to, since his lungs did not appear to be forthcoming. The physician asked him what he wished he could do, and the patient replied he would like to go crab fishing on the Atlantic seaboard. The physician thought this was an excellent goal, and the patient became excited by this prospect. Plans were made for the patient and his family to travel 250 miles to the New Jersey shore 3 weeks later. The patient was prescribed an anxiolytic, twice daily, as needed, for difficulty sleeping or feeling scared.

Subsequently, the patient experienced severe respiratory distress following chest physiotherapy, for which he was hospitalized for 4 days. Upon discharge, the patient resumed planning for his trip and reported that his shortness of breath, while present, did not bother him much. He used his anxiolytic medication on only a few occasions.

The patient's lungs became available during his trip, and he underwent successful lung transplantation.

Questions:

1. The differential diagnosis for this patient's dyspnea is likely to include all of the following EXCEPT:

   (a) Anxiety
   (b) Bronchospasm
   (c) End-stage lung disease
   (d) High blood concentration of carbon dioxide
   (e) Side effect of the multiple nebulized medications

2. Which of the following therapies would be most likely to be helpful for control of this patient's dyspnea in a short time frame?

   (a) Breathing into a paper bag
   (b) Cognitive behavioral therapy
   (c) Hypnotherapy
   (d) Speech therapy
   (e) All of the above

3. How might the patient's dyspnea have improved as a result of his planning for his trip and with hypnotherapy?

   (a) It did not improve; the patient just said it did.
   (b) Thinking about the trip distracted him.
   (c) Shifting his mind's focus to pleasant thoughts with use of hypnosis changed the way the patient perceived the sensations that caused him to feel dyspneic.
   (d) The improvement reflected the natural course of his dyspnea that waxed and waned and was unrelated to the interventions.
   (e) None of the above.

Answers:

1. (e): The patient's lung disease had progressed to the point that the patient had developed significant dyspnea, which could have been related to (b), (c), and (d). In such a setting, it is common for anxiety to occur, which can lead to worsening of dyspnea. This symptom occurred despite aggressive medical therapy, which typically serves to relieve (rather than cause) dyspnea in patients who are not as ill as this boy.
2. (c): A year before his transplant, this patient was taught self-hypnosis techniques in one session, which have been reported to decrease anxiety in patients with cystic fibrosis [86]. Just before the patient's trip, use of hypnotic imagery of going crab fishing to calm himself was associated with improvement in his dyspnea. This patient's dyspnea was not the result of hyperventilation given that his blood concentration of carbon dioxide was mildly elevated, and therefore, breathing into a paper bag would have caused harm. Cognitive behavioral and speech therapies typically take a few sessions and would be unlikely to have a rapid effect in this setting. Chronic dyspnea has been shown to improve with use of biofeedback and breathing techniques (see Chaps. 10 and 11).
3. (c): Hypnosis has been shown to change the way the brain processes pain perceptions and thus may have a similar effect in the processing of the sensation of dyspnea. Distraction has not been shown to have the same effects as hypnosis in the control of pain. A controlled study will need to be conducted in patients with end-stage CF in order to demonstrate whether the improvement in dyspnea can be attributed to hypnosis or planning for an exciting event.

Case 3: 13-Year-Old with Recurrent Exacerbations of Her Lung Disease

By the time this patient was 13 years old, her CF-associated lung disease had become very advanced. The patient required frequent hospitalizations and courses of intravenous antibiotics in order to treat her lung disease effectively. The patient's symptoms included a productive cough, dyspnea, occasional episodes of coughing up small amounts of blood (hemoptysis), and recurrent abdominal discomfort. While receiving intravenous antibiotics, the patient required supplemental oxygen via nasal cannula. During hospitalizations, the patient stated that supplemental oxygen was helpful to her even when her hemoglobin saturations were normal in room air (i.e., >97%).

The patient was an only child, whose parents had separated when she was a toddler. The patient lived with her mother and had virtually no contact with her father. Because of difficulties with the economy, and in being a single parent of a child with a severe chronic illness, the mother had lost jobs on four occasions. The mother had established and broken off relationships with a number of boyfriends during the patient's life. While not formally assessed, the mother often reported being distressed and anxious about the patient's poor health condition and prognosis.

The patient's CF care team noted that the patient tended to report that she was more symptomatic beginning in September (when school started) and when her mother had broken off a relationship or lost her job.

Questions:

1. All of the following of this patient's symptoms could have a functional component EXCEPT:

   (a) Cough
   (b) Dyspnea
   (c) Hemoptysis
   (d) Abdominal discomfort
   (e) Need for supplemental oxygen

2. Possible reasons that the patient's lung disease exacerbations tended to coincide with crises in her mother's life likely include all of the following EXCEPT:

   (a) The mother paid less attention to the patient when she was preoccupied with her own issues, and therefore, the patient received insufficient therapy at those times.
   (b) The patient drew attention to herself when she became more symptomatic, and she wanted increased attention when her mother was preoccupied.
   (c) The patient wanted to feel sicker so that she could share in her mother's misery.
   (d) The mother felt better when the patient was receiving aggressive care, which was important especially when the mother felt poorly as a result of her own issues. Therefore, the mother was more likely to complain of the patient's symptoms in such circumstances.
   (e) By drawing the mother's attention to the patient's symptoms, the patient was distracting her mother from thinking about the mother's issues.

3. The mother's anxiety about the patient's health could have contributed to the patient's reported development of exacerbations through all of the following means EXCEPT:

   (a) The mother became too nervous to assess the patient properly, which led her to ignore some of the patient's symptoms until they became severe enough to require hospitalization.
   (b) The anxiety led the mother to be hypervigilant and therefore more likely to interpret the patient's symptoms as necessitating medical evaluation.
   (c) The patient became anxious as a result of the mother's anxiety, which led to intensification of her symptoms.
   (d) The anxiety caused the mother to doubt her own capability to assess and deal with the patient properly, and therefore, the mother was more likely to seek medical intervention for the patient.
   (e) The anxiety led the mother to think of the worst possible outcomes when the patient reported symptoms and therefore caused her to believe that the patient was in danger.

4. Given that the mother's anxiety may have contributed to the difficulties in managing this patient, appropriate interventions for the mother included all of the following EXCEPT:

   (a) Counseling for the mother
   (b) Family therapy
   (c) Anxiolytic therapy for both mother and patient
   (d) Hypnotherapy for both mother and patient
   (e) Reassurance

Answers:

1. (c): There is no known functional cause of hemoptysis, which typically occurs in patients with CF as a result of erosion of a bronchial artery by airway inflammation. Cough, dyspnea, and abdominal discomfort can all occur as a result of physiologic issues in CF but also can present as functional symptoms. The patient's need for supplemental oxygen despite her normal hemoglobin saturations probably was related to anxiety.
2. (c): All of the other options can help perpetuate functional symptoms. Options (b) and (e) represent examples of secondary gain that tend to perpetuate functional symptoms. Options (a) and (d) demonstrate how parents' reactions to their children's illness may affect its actual or reported severity. Patients do not usually want to feel more ill, as suggested in option (c).
3. (a): Anxious individuals tend to be hypervigilant, and thus, it is unlikely that the patient's mother would tend to ignore symptoms as a result of anxiety.
4. (c): While reassurance may not have been sufficient to address this mother's anxiety, it is an appropriate first course of action. Counseling, family therapy, and hypnotherapy could all have helped with anxiety reduction. Anxiolytic therapy by itself for the mother is unlikely to have worked as it would not have addressed her concerns regarding the patient's health. Furthermore, anxiolytic therapy is not indicated for the patient if her anxiety arose as a reaction to her mother's anxiety.

Case 4: 19-Year-Old with Chest Pain

The patient was a 19-year-old with CF who emigrated with his mother and older brother from an Eastern European country to the United States at the age of 8 years in order to join his father who already had been in America for 2 years. The patient had received only oral antibiotics sporadically for his CF-related lung disease and pancreatic enzyme supplementation. Upon arrival, he was found to be malnourished and to have moderate lung disease. The father left the family shortly after their arrival.

The patient was started on standard therapy for CF including dornase alpha, chest physiotherapy with a high frequency chest wall oscillating vest, pancreatic enzymes, and vitamins. Within a year, his lung function had improved markedly, but he continued to be malnourished despite oral nutritional supplements. Insertion of a gastrostomy feeding tube was recommended when the patient was 10 years old, but because they hoped he could eat enough on his own, his family did not consent to its placement until he was 13 years old. He gained weight rapidly thereafter with the use of nighttime feedings.

The patient did well from a pulmonary and nutritional perspective for the subsequent 5 years. However, he complained that he had no appetite during the days, which was attributed to his being fed overnight. In his later adolescence, the patient reported spending many hours a day fantasizing that he was powerfully built. He reported that he awaked frequently at night and was tired in the daytime. He became very focused on building muscles; however, he was unable to maintain motivation to lift weights on a regular basis. He lost interest in other activities outside of his home.

The patient asked that his CF center physician arrange for him to have a personal trainer, but this was not possible. The patient became increasingly noncommunicative with his physician over the subsequent few months. He began complaining of significant chest pain that made it hard for him to cough. He denied shortness of breath. He denied episodes of regurgitation or emesis. On examination of his chest, his pain was elicited when his mid-sternum was pressed. He also complained of pain when his costochondral joints were pressed bilaterally. His lungs were clear. His pulmonary function was normal. His chest x-ray revealed mild upper lobe bronchiectasis that had not changed in appearance over the past 2 years. The patient reported that use of ibuprofen and acetaminophen did not help with his pain.

The chest pain persisted for the subsequent 6 months. During this time, the patient removed his gastrostomy tube against medical advice and stopped taking his therapies. His appetite remained poor. He began losing weight rapidly and his pulmonary function declined. He refused admission to the hospital or referral to a psychiatrist.

Questions:

1. The differential diagnosis for this patient's chest pain includes all of the following EXCEPT:

   (a) Asthma
   (b) Costochondritis (inflammation and associated tenderness of the cartilage that attaches the front of the ribs to the breastbone)
   (c) Gastroesophageal reflux
   (d) Pneumothorax
   (e) Somatoform disorder

2. All of the following complaints were suggestive of depression EXCEPT:

   (a) Lack of appetite
   (b) Loss of interest in activities
   (c) Sleep disturbance
   (d) Preoccupation with body image
   (e) Lack of adherence to his therapies

3. Referral to all of the following services would be appropriate for treatment of this patient's chest pain EXCEPT:

   (a) Cognitive behavioral therapy
   (b) Hypnotherapy
   (c) Pain clinic for appropriate management of narcotic therapy
   (d) Psychiatry
   (e) Psychology

Answers:

1. (d): The patient is unlikely to have a pneumothorax given the absence of shortness of breath, decreased breath sounds, or x-ray evidence of this condition. Bronchospasm associated with asthma can cause chest pain. Costochondritis was suggested given that the pain was elicited with pressure applied during the physical examination. While the patient did not have symptoms suggestive of gastroesophageal reflux, his nighttime gastrostomy tube feedings placed him at increased risk for this condition. The likelihood of somatoform disorder as a diagnosis was increased given the patient's multiple psychological and family issues.
2. (d): Preoccupation with body image is common in an adolescent. While poor sleep hygiene and lack of adherence to therapies also are typical for adolescents, the extent of these behaviors in this patient was extreme. Loss of interest in favorite activities is common for patients with depression. This patient might have developed his lack of appetite as a result of his gastrostomy tube feedings, but this was unlikely as his appetite did not improve when he did not use the tube. Patients with advanced CF lung disease often lose their appetite, but this patient had only mild lung disease.
3. (c): Given this patient's complex psychosocial difficulties, it is unlikely that the chest pain would resolve with an intervention that mostly targets the symptom, as opposed to the underlying depression.

Case 5: 20-Year-Old with Insomnia [87]

The patient was a 20-year-old young man with advanced cystic fibrosis who had suffered a nearly fatal episode of hemoptysis, for which he necessitated being placed

on a ventilator for 6 days, followed by prolonged rehabilitation. Two months later, he reported recurrent nightmares about hemoptysis, which awakened him approximately five times a night. The patient explained that he was concerned about again suffering from massive hemoptysis and that this thought prevented him from falling asleep easily. Also, he was worried about being too physically active in case this might trigger further bleeding. Occasionally, when the patient tasted a small amount of blood, such as a result of a nosebleed, he has had flashbacks to his near fatal episode. He said that even thinking about the possibility of hemoptysis caused his chest to tighten and his heart to beat more strongly.

The patient had been taught how to use self-hypnosis for relaxation 2 years prior to his development of anxiety and nightmares about bleeding. It was suggested that the patient could view his beating heart as a metronome that was sending a wave of relaxation with each beat. When the patient applied this imagery, he reported that he no longer felt his heart was beating too strongly. Also, it was suggested that whenever the patient tasted blood that he might reaffirm his life by telling himself, "I am bleeding; therefore, I am alive."

To help with his nightmares, the patient was instructed to go into hypnosis and to allow his subconscious to show him a dream representative of those that woke him up. After a 3-min hypnosis session, the patient reported he had dreamed he was having another massive hemoptysis episode, which caused him to be very fearful. When he continued to dream, he perceived two possible endings to the dream: In one, he survived, and in the other, he died. The patient concluded that his most important realization was that no matter what happened, he would be "all right" and that the discomfort of the hemoptysis episode would be short-lived.

During the subsequent month, the patient reported having only two further nightmares, which he was able to "work through" while remaining asleep.

Questions:

1. The adverse effect of the patient's concerns about recurrent hemoptysis could have included all of the following EXCEPT:

   (a) He could have interpreted the associated chest tightness as indicating that he was physically ill, which could have led to the need for medical attention and increased anxiety.
   (b) His concerns about his level of activity could have translated into a more sedentary lifestyle that would have decreased his pulmonary toilet.
   (c) His sleep disruption as a result of his nightmares affected his quality of life.
   (d) When he had flashbacks to the hemoptysis, he imagined tasting blood, which is consistent with post-traumatic stress disorder leading to the development of gustatory hallucinations.
   (e) His concerns might have led him to be less adherent to his prescribed inhaled therapies, for fear that they might trigger recurrent hemoptysis.

2. All of the following referrals or therapies would be appropriate for this patient EXCEPT:

   (a) Cognitive behavioral therapy
   (b) Diphenhydramine, oral
   (c) Hypnosis
   (d) Mindful meditation
   (e) Sleep study

3. In addition to use of hypnosis, what other form of psychological therapy was this patient given?

   (a) Cognitive behavioral therapy
   (b) Meditation
   (c) Play therapy
   (d) Sleep hygiene education
   (e) None of the above

Answers:

1. (d): There is no reason to suspect that this patient imagined blood, as he pointed out that he suffers from epistaxis. The potential adverse events that could have arisen as a result of the patient's anxiety about recurrent hemoptysis illustrate the importance of addressing this issue effectively.
2. (b): Use of diphenhydramine could help the patient fall asleep but would not address his anxiety. A sleep study could help define whether there was an organic cause for the patient's sleep disturbance, such as obstructive sleep apnea. The other options could have helped improve his anxiety.
3. (a): By helping the patient identify the fear that was underlying his nightmares, he was able to resolve it. Sleep hygiene education includes establishing a consistent sleep schedule and bedtime routine and modification of the bedroom environment and food intake.

Conclusions

The detailed discussion provided in this chapter of the potential impact of functional symptoms on patients with CF serves as a model for a comprehensive approach to patients with chronic pulmonary diseases. For example, psychiatric and psychosocial factors are well-recognized as affecting patients with asthma and chronic obstructive pulmonary disease [88–91].

Thus, clinicians providing care for patients with chronic pulmonary disease should consider the possible impact of a functional contribution to the clinical presentation of every patient and offer treatment for this relatively early in the therapeutic algorithm. Treatment strategies for functional respiratory symptoms

are discussed in the third section of this book, "Treatment of Functional Respiratory Disorders." Such an approach will lead to more efficient and effective management of patients with chronic pulmonary disease.

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