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Title: Journal of alternative and complementary medicine (New York, N.Y.)  
 Title Abbrev: J Altern Complement Med  
 Citation: 2010 Sep;16(9):1007-10  
 Article: Reestablishment of hope as an intervention for a p  
 Author: Anbar R;Murthy V  
 NLM Unique ID: 9508124 Verify: PubMed  
 PubMed UI: 20799891  
 ISSN: 1075-5535 (Print) 1557-7708 (Electronic)  
 Fill from: **Any format**  
 Publisher: Mary Ann Liebert, Inc., New York, NY :  
 Copyright: Copyright Compliance Guidelines  
 Authorization: sayred  
 Need By: N/A  
 Maximum Cost: **\$16.00**  
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 Routing Reason: Routed to NYUMSK in Serial Routing - cell 1  
 Received: Sep 29, 2010 ( 11:02 AM ET )  
 Lender: Memorial Sloan-Kettering Cancer Center/ New York/ NY USA (NYUMSK)

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## Reestablishment of Hope as an Intervention for a Patient with Cystic Fibrosis Awaiting Lung Transplantation

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### Abstract

Patients with cystic fibrosis (CF) experience a progressive deterioration in health leading to a shortened life expectancy with concomitant psychologic challenges that also may require intervention. Approaches to address psychologic dysfunction include insight-oriented therapy, cognitive-behavioral therapy, biofeedback, hypnosis, massage, music therapy, relaxation, and family therapy. Patients who learn to use coping strategies are more likely to adhere to treatment, which helps improve both their physical and psychologic well-being. Unique stressors arise for patients awaiting lung transplant as a result of their advanced state of disease along with their fear of death before donor lungs become available. This case report demonstrates some of the psychologic struggles faced by a 10-year-old with CF as he awaited lung transplantation, which led to a loss of hope and his request for withdrawal of care. Encouraging of the patient to express how his life might be improved yielded changes in his management that restored his hopefulness, and allowed him to survive long enough to undergo successful lung transplantation. Thus, reestablishment of hope might be studied as an additional intervention to help improve the well-being of patients who are seriously ill.

### Introduction

**P**ATIENTS WITH CYSTIC FIBROSIS (CF) experience a progressive deterioration in health leading to a shortened median predicted survival age of under 38 years,<sup>1</sup> with concomitant psychologic challenges that may require intervention. Assessment of these challenges can be reflected through quality of life measures.<sup>2</sup> These have shown that patients with CF have an increased likelihood of psychiatric problems, such as depression,<sup>3</sup> and express lower levels of hope in comparison to well individuals.<sup>4</sup> Approaches to address psychologic dysfunction include insight-oriented therapy, cognitive-behavioral therapy, biofeedback, hypnosis, massage, music therapy, relaxation, and family therapy.<sup>5</sup> Patients with CF who learn to use coping strategies are more likely to adhere to treatment, helping to improve both their physical and psychologic well-being.<sup>5-7</sup>

The effect of life stressors on patients' psychologic status has been associated with impaired coping ability.<sup>8</sup> Furthermore, unique stressors arise for patients awaiting lung transplant as a result of their advanced state of disease along with their fear of death before donor lungs become available.<sup>9,10</sup> In such patients, optimism or active coping with psychosocial stressors have been associated with superior psychologic quality of life (QoL), as opposed to avoidance behaviour that prevents dealing with the stressors.<sup>6,11</sup> The

degree of coping has been shown to account for more variation in QoL than parameters such as the level of disease severity and time spent in therapy.<sup>12</sup> Improvement in QoL, including physical function, has been reported in patients awaiting lung transplantation who received a psychosocial intervention in the form of telephone-based supportive counseling that included education about stress and instruction in coping techniques such as relaxation, problem solving, and calming self-statements.<sup>13</sup> Thus, coping strategies may be an important target for intervention in patients with CF.<sup>12</sup>

An optimistic attitude has been reported to be associated with improved health in several populations. Among healthy individuals, a positive outlook has been found to permit better coping with stressful situations, thereby reducing the harmful effects of stress on the body.<sup>14</sup> Furthermore, happiness may protect patients against becoming ill, as there is a strong association of happiness with longevity in healthy populations.<sup>15</sup> In patients with human immunodeficiency virus (HIV), optimism, active coping, and spirituality were associated with slower disease progression, as demonstrated by monitoring of CD4 cell counts.<sup>16</sup> Optimism has been associated with lower ambulatory blood pressure<sup>17</sup> and lower risk of rehospitalization following coronary artery bypass graft (CABG) surgery.<sup>18</sup> In patients with cancer, finding meaning in life and having hope appears to enhance

psychospiritual well-being, which allows for improved coping with the illness,<sup>19</sup> while hopelessness has been associated with an increased rate of metastasis and pain.<sup>20</sup>

The following case demonstrates some of the psychologic struggles faced by a 10-year-old boy with CF as he awaited lung transplantation, which led to a loss of hope and his request for withdrawal of care. In order to help improve this patient's coping ability, he was encouraged to express his ideas on how his life might be improved. This yielded changes in his management that helped restore his hopefulness and allowed him to survive long enough to undergo successful lung transplantation.

### Case Report

This patient was diagnosed with CF at 2 weeks of life, when he had meconium ileus. He was homozygous for the  $\Delta F508$  CF mutation. Complications of his disease included recurrent pneumonia (necessitating nine hospitalizations), recurrent sinusitis, renal stones, and severe gastroesophageal reflux for which he underwent a Nissen fundoplication at 8 years of age. Despite aggressive therapy, by the time he was 9 years old, his lung function had deteriorated such that his forced expiratory volume in 1 second ( $FEV_1$ ) was 0.37 L (29% of predicted for height and age), and he required full-time supplemental oxygen via nasal cannula at 2 L/minute. Therefore, he was placed on a waiting list for lung transplantation in July 2003. At that time, the patient lived with his mother, stepfather, and 1 and 3 year-old half-sisters. He was in third grade.

As the patient remained on the transplantation list for 13 months, he became increasingly fatigued, with increased coughing, chest tightness, and shortness of breath. He required home-schooling for the second half of fourth grade because of his worsening medical condition. Generally, the patient was cooperative with, and adherent to, his extensive therapeutic regimen including use of daytime bilevel positive airway pressure (BiPAP) administered through nasal prongs for 2 hours, along with 5 L/minute of oxygen, as well as overnight BiPAP. He used a Vest<sup>®</sup> Airway Clearance System (American Biosystems, St. Paul, MN) for 30 minutes, 3–4 times a day, and a TheraPEP<sup>®</sup> Positive Expiratory Pressure System (DHD Health Care Corporation, Wampsville, NY) during breaks from the Vest<sup>®</sup> therapy. His medications included: three oral antibiotics; nebulized antibiotic twice daily; nebulized hypertonic saline twice daily; nebulized mucolytic twice daily; nebulized asthma medications six times daily; an oral asthma medication; three oral pancreatic enzymes with each meal and snack; two oral vitamin and mineral supplements; an oral antihistamine; an oral hydrogen-proton blocker; two oral diuretics; and an oral appetite stimulant. In addition, he was receiving 1.5 L of normal saline intravenously (I.V.) overnight to maintain his hydration.

During the week prior to a visit at his CF center, the patient stated to his mother that he did not want any more physiotherapy. He was removed from BiPAP and had great difficulty breathing, which frightened him. Subsequently, he said he no longer wanted to receive a lung transplant and wanted to stop fighting. He explained that he was tired of spending entire days maintaining his health. He said he was not afraid of undergoing lung transplant surgery, but did not believe lungs would become available for him in time to save

him. This patient was told that, if he no longer wanted to fight CF, his parents and attending physician would respect his decision. The patient discussed his belief in heaven and said that, no matter what happened, he believed he would be "okay." His parents confirmed they shared these beliefs, but also expressed that they wished very much that the patient would survive long enough to be able to undergo lung transplantation. This patient asked for videotapes to be made of him so that his sisters could view them when they were older.

On physical examination, this patient appeared to be in moderate respiratory distress and sad. His responses to queries were thoughtful and well-reasoned. The patient was not assessed formally for depression, but his mother said she did not feel he was depressed. His height was 46.5" (at approximately the 50th percentile for a 6-year-old) and his weight was 58.7 lbs. His body-mass index was 19.1 (approximately the 80th percentile for his age.) His respiratory rate was 30. A lung examination revealed diffuse crackles. He had 3+ digital clubbing. Pulmonary function testing revealed an  $FEV_1$  of 0.31 L (23 % of predicted). His hemoglobin saturation, as measured by pulse oximetry, was 92%, while receiving 5 L/minute of oxygen via nasal cannula.

The physician asked if the patient would be willing to receive lungs for transplantation if they became available on that day. The patient said that he would. The physician responded that perhaps the patient was not quite ready to give up fighting given that he would still accept lungs. The physician asked what changes could be made to make the patient's life more tolerable. The patient replied that he did not want to do as many therapies and that he disliked use of the Vest.<sup>®</sup> Therefore, the physician suggested that the patient experiment instead with a cough-assist device. Also, the patient stated that he had nothing to look forward to, because new 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 with sleeping or feeling scared.

The patient returned to the CF center 2 weeks later. In the interim, he had experienced severe respiratory distress following chest physiotherapy with hemoglobin saturations to the 60s, for which he was hospitalized for 4 days, during which time he was maintained on BiPAP for 20 hours/day, and started on therapy with two I.V. antibiotics, two oral antibiotics, and an oral steroid. Upon discharge, he felt much better, and by his follow-up his  $FEV_1$  was 0.36 L (27% of predicted). In response to a query by his physician, the patient stated that, if he had to be intubated, he could accept it for 2 weeks in case lungs became available. The physician suggested that, because the patient felt better as a result of looking forward to his vacation a week later, he should think about other enjoyable events in which he might engage after his return. For example, the physician suggested that the patient could host a party for his friends.

Six (6) days later, while the patient was at the New Jersey shore, lungs became available at his transplant center that was 300 miles away. The patient's initial reaction, when told

that lungs were available, was that he did not want to go through with the transplant because he was having too much fun at the shore. After a bit of cajoling by the physician and by the patient's family, he agreed to be transported to the transplant center through a hastily arranged flight and arrived 6 hours later. Upon arrival, the patient expressed optimism that he would do well with the transplant, and urged his surgeons to "get the show on the road." The patient tolerated the transplant well. He returned to school 4 months later as an active and happy member of the fifth grade. Thus far, he has lived an additional 5 years.

### Discussion

This report illustrates how reestablishment of a hopeful attitude can strengthen a patient even with end-stage pulmonary disease and passive suicidal ideation. The physician's approach to dealing with the patient's expressed wish for withdrawal of care was based on familiarity and a relationship with the patient and his family over a number of years. For example, based on knowledge that the patient tended to be cooperative with, and adherent to, his therapies, the physician believed that the patient had demonstrated long-term motivation to remain as healthy as possible. Furthermore, the physician determined that the patient's cognitive and emotional development was sufficient to allow him to be an active participant in the ensuing discussion regarding why his motivation had changed in a way that led to the request for withdrawal of care.

The physician's question about whether or not the patient would be willing to undergo lung transplantation if it were possible on the day of the clinic visit helped this patient recognize his own ambivalence about his request for withdrawal of care and realize that he still had capacity for hope. Furthermore, the physician's offer to discuss how the patient's life might be changed to address his concerns implied that options remained for the patient, and thus demonstrated further hope and modeled it for this patient.<sup>21</sup> The negotiations regarding modification of the patient's medical care showed the physician's respect for the patient's thoughts and feelings and allowed the patient to become engaged in directing his own care. These negotiations helped empower this patient by giving him more control and providing significant ego-strengthening, which may have helped him cope with his situation better.<sup>22</sup> Allowing the patient to choose a short-term goal raised his spirits, in part, because this helped ensure that the intervention was based appropriately on the patient's developmental age, including accounting for any regression that may have occurred as a result of his severe illness.<sup>23</sup> Thus, the intervention helped rebalance the patient's ambivalence and shift his focus from how he lacked control as a result of his disease, and how he might orchestrate his death, to how he could live while having increased control of his therapeutic regimen and pleasurable activities.<sup>24</sup> When the patient became enthusiastic about a goal that involved a social activity with his family, the physician reinforced these positive feelings by suggesting that the patient might consider other social activities that included his friends after the family vacation.

QoL was not assessed formally in this case, but such a measurement may have alerted his health care team earlier that a psychological intervention could have been helpful.<sup>25</sup>

### Conclusions

In conclusion, the experience with the patient in this report suggests that reestablishment of hope might be studied as an additional intervention to help improve the well-being of patients who are seriously ill.

### Disclosure Statement

The authors have no relevant financial interests to disclose.

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