Quality of Life and Parental Adjustment in Pediatric Pulmonary Hypertension

Some Methodologic Considerations

To the Editor:

We commend Mullen et al1 for their recent article in CHEST (February 2014) on quality of life and parental adjustment in pediatric pulmonary hypertension. It is indeed a very valuable research question that the authors have made an attempt to answer. However, we would like to point out a number of methodologic concerns that we fear might limit the validity of the study results.

The small sample size (N = 47) is likely to lower the statistical power of the study design. We are of the opinion that a generic scale like the Pediatric Quality of Life Inventory (PedsQL), Parent Report version, is not ideal for measurement of quality of life of the study population. A more disease-specific instrument such as the Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR) scale, which was designed specifically for patients with pulmonary arterial hypertension and is primarily a patient report version, would have better suited the study purpose.2

Lack of adjustment for multiple comparisons and lack of generalizability of the single-site study pose further questions as to the validity of the study results. We would also like to point out that the assessment of psychiatric morbidity in children with pediatric pulmonary hypertension and inclusion of psychologic morbidity detected as a factor influencing both patient quality of life and parental stress would have been appropriate, considering the high psychiatric morbidity demonstrated in earlier studies of patients with pulmonary arterial hypertension.3 Not assessing and including the same (psychiatric morbidity in parents) as a confounding variable results in a lacuna that cannot be filled. We would recommend similar studies on a larger scale (preferably multisite) that would take into account the various methodologic challenges to shed more light on this valuable research question.

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Response

To the Editor:

We appreciate the comments of Drs Gnanavel and Robert regarding our recent article in CHEST.1 This study represents the largest sample to date on the psychosocial aspects of pediatric pulmonary hypertension (PH). In our article, we acknowledged limitations of the study, including small sample size and single-center design. We agree that a disease-specific instrument to assess quality of life in patients with PH could be useful; no such instrument has been validated for the pediatric age group. Optimally, this should use both self report and parental report. We agree that assessment of psychiatric morbidity in children with PH may be informative and could be addressed in additional studies. Finally, as we stated in our article, future multisite studies will be essential to further delineate the important psychosocial consequences of PH in pediatric patients.

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6-Minute Walk Distance

Effect of Instructions

To the Editor:

We read with great interest the article by Weir et al1 published in CHEST (December 2013). The authors compared the effect of the instruction “walk as fast as you can” (fast walk) with the standard instruction “walk as far as you can in 6 min” (standard walk) for the 6-min walk test (6MWT) in patients with pulmonary arterial hypertension, idiopathic pulmonary fibrosis, and other forms of pulmonary hypertension. We would like to point out a few additional considerations.

The small sample size (N = 47) is likely to lower the statistical power of the study design. We are of the opinion that a generic scale like the Pediatric Quality of Life Inventory (PedsQL), Parent Report version, is not ideal for measurement of quality of life of the study population. A more disease-specific instrument such as the Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR) scale, which was designed specifically for patients with pulmonary arterial hypertension and is primarily a patient report version, would have better suited the study purpose.

Lack of adjustment for multiple comparisons and lack of generalizability of the single-site study pose further questions as to the validity of the study results. We would also like to point out that the assessment of psychiatric morbidity in children with pediatric pulmonary hypertension and inclusion of psychologic morbidity detected as a factor influencing both patient quality of life and parental stress would have been appropriate, considering the high psychiatric morbidity demonstrated in earlier studies of patients with pulmonary arterial hypertension.

Not assessing and including the same (psychiatric morbidity in parents) as a confounding variable results in a lacuna that cannot be filled. We would recommend similar studies on a larger scale (preferably multisite) that would take into account the various methodologic challenges to shed more light on this valuable research question.

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of interstitial lung diseases. They found that patients walked longer distances in 6 min with the fast walk compared with the standard walk. We would like to emphasize that an important component of the applicability of the 6MWT is its ability to reflect activities of daily living over other walk tests. In day-to-day life, patients are used to walking at their own pace and not fast. As a result, the reduction in distance of fast walk may not truly reflect an equivalent reduction in patients’ activities of daily living. In addition, for measuring functional capacity, we believe that repeatability of the test, which the authors did not study, is more important.

Another important limitation of the study is the lack of a comparative arm of healthy subjects. The 6-min walk distance is compared with the predicted values obtained from the healthy population for making an interpretation. It is logical that healthy subjects will also walk more distance with the fast walk compared with the standard walk. Hence, it would have been interesting to see whether the increase in walk distance with the fast walk is also seen in terms of percent predicted. We hypothesize that percent predicted values might in fact be lower in patient populations with the fast walk compared with the standard walk.

Guidelines by the American Thoracic Society recommend giving a rest of 1 h between successive 6MWTs. Subjects were given a rest of about 15 min between tests in this study, which may potentially have an effect on subsequent tests. We believe that this effect will be larger on tests done after the fast walk compared with the standard walk. If that is true, this effect cannot be nullified by randomization of test instructions. Hence, we believe that it is important to statistically analyze and adjust the effect of order or sequence on the primary outcome.

We appreciate the efforts of the authors in bringing attention to the importance of standardized instructions for 6MWT and the potential effect different instructions can have on the distance walked. However, we believe that the increase in distance walked with the instruction “walk as fast as you can” does not truly reflect the activities of daily living of the patients. We also believe that the effects of previous tests on subsequent tests were not adequately adjusted in analysis to draw reliable conclusions from the study.

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**Table 1**—Variation and Reproducibility in Walk Distance With Differing Instructions (n = 12)

<table>
<thead>
<tr>
<th></th>
<th>Distance, m</th>
</tr>
</thead>
<tbody>
<tr>
<td>6MWT</td>
<td>Visit 1</td>
</tr>
<tr>
<td>Standard</td>
<td>491 (91)</td>
</tr>
<tr>
<td>Fast</td>
<td>495 (71)</td>
</tr>
<tr>
<td>Furious</td>
<td>511 (55)</td>
</tr>
</tbody>
</table>

Data are presented as median (interquartile range). 6MWT = 6-min walk test.

**Response**

To the Editor:

In our recent article in CHEST, we showed that replacing “far” with “fast” in the instruction provided to patients resulted in an average improvement of about 50 m in the distance attained. It is intriguing that a one-letter substitution (“f” for “y”) can stimulate such an improvement, as well as an ongoing debate. Dr Vanjare and colleagues question the value of optimizing the 6-min walk test (6MWT) distance as a reflection of a patient’s ability to do daily activities. However, the 6MWT is a submaximal exercise test designed to determine the patient’s capacity to perform activities of daily living. The American Thoracic Society guidelines on the 6MWT state that “the self-paced 6MWT assesses the submaximal level of functional capacity.” Although some patients achieve their submaximal functional capacity in activities of daily living, many do not. It is the capacity that needs to be assessed and not the true activity level at home. Several studies that are investigating sedentary lifestyles and means to improve upon them address this very topic.

Our findings are pertinent to the very salient issue of the use of the 6MWT in clinical trials. Although the baseline 6MWT predicts outcomes, the change in the 6MWT distance does not. Therefore, the 6MWT has fallen into disfavor as a study end point. We hypothesized in our article that improving the 6MWT precision and reproducibility may be facilitated by maximizing the distance attained. Indeed, this concept is the foundation for our follow-up study (termed “the fast and the furious”), which is currently in the recruitment phase. Patients perform three 6MWTs on 2 separate days: (1) a standard 6MWT with the “far” instruction, (2) a 6MWT with the “fast” instruction, and (3) a 6MWT test with the fast instruction but a stronger (“furious”) menu of scripted instructions each minute to encourage more effort. The hypothesis being tested is that the greater the effort and distance, the less variability in subsequent 6MWTs. We can report that there is a trend supporting this hypothesis among the 12 patients studied thus far, as depicted in Table 1.

Dr Vanjare and colleagues also raise the issue of whether sufficient time (15 min) was allowed between walks. To our knowledge, the American Thoracic Society recommendation for 1 h of rest between walks has never been tested or validated, and it is likely based on intuition and expert opinion. Unfortunately, adhering to this recommendation would increase the duration of four random walk tests in a day to 5 h or longer, thereby reducing the feasibility of our study. If patients were increasingly tired from