

## Editorials

## Quality of life in cystic fibrosis

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In recent years the role of behavioural and psychological variables in the management of patients with cystic fibrosis has been noted.<sup>1–3</sup> This interest has grown from the observation that improved diagnosis and treatment mean that many patients can now anticipate burdensome daily management routines continuing for some decades. Recent studies in this and other publications have included demonstrations that the health related quality of life (HRQOL) of adults with cystic fibrosis is in some respects impaired,<sup>1–3</sup> and that HRQOL is a valuable outcome measure in clinical trials.<sup>4–5</sup> Such findings are in contrast with earlier literature which tended to show little evidence of psychosocial impact in children<sup>6</sup> and adolescents<sup>7</sup> with cystic fibrosis. There is, however, an important distinction between the psychosocial outcomes identified in the 1980s, often using instruments intended to detect psychopathology, and more recent conceptualisations of HRQOL.

HRQOL is a multidimensional construct encompassing a range of impacts including physical, functional, social, and emotional components. Definitions frequently emphasise not only this breadth of impact but the subjectivity of reports<sup>8</sup>—that is, HRQOL differs from reports of psychological adjustment or functional status in targeting the importance of the impairment to patients, assessed through their own reports where possible. This reliance upon self reporting is one reason for the dearth of paediatric studies since clinicians and researchers have doubted the capacity of children to self report.<sup>9</sup> Even supposing the reliability of children as reporters can be established, there are currently few if any instruments suitable for the assessment of children's HRQOL.<sup>10</sup>

The paper by Staab and colleagues<sup>11</sup> in this issue of *Thorax* therefore makes an important contribution to this poorly researched field. These authors have explored determinants of HRQOL in patients aged 12 years and over using the patients' own reports of physical, functional, social, and emotional impacts. Importantly, they have also addressed HRQOL impacts for the parents of those patients in their sample who live at home and for the parents of children too young to self report. This is a significant advance in a literature which has previously taken a descriptive approach to the impact of cystic fibrosis on children's families<sup>12–13</sup> or has again focused on psychopathology.<sup>14</sup> Since the study assesses only patients' self reports and parents' reports of their own HRQOL, and does not use proxy reports of the HRQOL of younger children, it avoids the problem that parents' own adjustment is a major predictor of parents' reports of child adjustment,<sup>15</sup> and thus a potential confounder in proxy reports of HRQOL.

Staab *et al*<sup>11</sup> have shown that, once clinical severity has been accounted for, there are other significant predictors of HRQOL for patients and their parents. Hours spent each day on therapy add significant unique variance to the pre-

dictive equation for both patients' and parents' HRQOL. Subjective perception of health is also a predictor for patients. This in itself is unremarkable—patients' perceptions of their health are not so far removed from their perceptions of their health related quality of life that this finding should be notable alone. Much more interesting and potentially valuable for patient management are the findings regarding the role of coping. Coping has been conceptualised in a variety of ways, the most salient distinction being between those theories and resultant measures that identify coping *style* as a personal trait<sup>16</sup> and those that see coping as a set of *strategies* or ways of coping which may differ in response to different situations.<sup>17</sup> Both approaches have been used in past studies of cystic fibrosis where investigations have been made of the role of trait coping in patient adjustment,<sup>18</sup> coping strategies in parent adjustment,<sup>19</sup> and parent coping strategies in child adjustment.<sup>20</sup> Notable by its absence is any study of coping as a predictor of HRQOL. Since psychosocial adjustment, especially in children, has proved to be an insensitive measure of the impact of living with cystic fibrosis, such a study is overdue.

Staab *et al*<sup>11</sup> take the coping *strategies* approach, obtaining self reports of illness related ways of coping from both patients and parents. The German language measure used is not well known in the English speaking literature, but its reported subscales are not dissimilar to better known measures. For example, problem solving, cognitive avoidance, and self-encouragement have strong parallels in the better known Ways of Coping questionnaire.<sup>17</sup> The inclusion of coping subscales in the study by Staab *et al*<sup>11</sup> explains unique variance in HRQOL for both patients and parents. For both groups reports of depressive coping are predictive of poor HRQOL. Such a process is likely to involve self blame and a preoccupation with emotional thoughts—a process associated with poor psychosocial outcomes in many patient groups including the mothers of children with cystic fibrosis.<sup>19</sup> The next most predictive variable for patients is cognitive avoidance or denial, which is also a negative indicator, while for parents the most important predictor is a response involving retreat from social relations and control of emotions. For both groups strategies which indicate failure to confront the illness and its impact predict the poorest HRQOL. This is consistent with past research in which the outcome measure has been the psychosocial adjustment of parents,<sup>14–19</sup> but in studies of patients a denying or avoiding style has been associated with more positive psychological outcomes.<sup>18–21</sup> This may of course be the result of an artefact—those who use a denying response to their illness may also deny psychosocial problems. It is therefore of great value to learn that, when the outcome measure is appropriately characterised as HRQOL, patients with cystic fibrosis show a pattern

more consistent with that of other patient groups dealing with long term illness.<sup>22</sup>

Smaller degrees of predictive power are also associated with some positive strategies in the study by Staab *et al.*<sup>11</sup> Together these offer valuable insights into the kinds of approaches that might be offered to patients as ways of coping with the daily demands of managing cystic fibrosis. The study also points to the value of further investigations of HRQOL in this patient group. The measure used here is a generic tool designed to assess a range of impacts in a range of illnesses. Whilst there are no publications to date that offer measures of HRQOL specifically for cystic fibrosis,<sup>23</sup> at least two are under development.<sup>11 24</sup> The current study highlights the importance of such endeavours, as do suggestions that generic measures which sum across HRQOL domains may obscure important illness related differences in patterns of impact.<sup>25</sup> The significant need at present is for a measure which is both specific for cystic fibrosis and multidimensional. Since studies in other illness groups have shown the value of self reports for children from primary age upwards,<sup>26 27</sup> there is also a need for measures which can address the paediatric age range. Outcome measurement is now a sophisticated science and the advent of such instruments has a great deal to offer in the future investigation of management options for patients with cystic fibrosis.

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- 1 de Jong W, Kaptein AA, van der Schans CP, *et al.* Quality of life in patients with cystic fibrosis. *Pediatr Pulmonol* 1997;23:95-100.
- 2 Blair C, Cull A, Freeman CP. Psychosocial functioning of young adults with cystic fibrosis and their families. *Thorax* 1994;49:798-802.
- 3 Congleton J, Hodson ME, Duncan-Skingle F. Quality of life in adults with cystic fibrosis. *Thorax* 1996;51:936-40.
- 4 Orenstein DM, Pattishall EN, Nixon PA, *et al.* Quality of well-being before and after antibiotic treatment of pulmonary exacerbation in patients with cystic fibrosis. *Chest* 1990;98:1081-4.
- 5 Busschbach JVV, Horikx PE, van den Bosch JMM, *et al.* Measuring the quality of life before and after bilateral lung transplantation in patients with cystic fibrosis. *Chest* 1994;105:911-17.
- 6 Drotar D, Doershuk CF, Stern RC, *et al.* Psychosocial functioning of children with cystic fibrosis. *Pediatrics* 1981;67:338-43.
- 7 Mador JA, Smith DH. The psychosocial adaptation of adolescents with cystic fibrosis. *J Adolesc Health Care* 1989;10:136-42.
- 8 Schipper H, Clinch J, Powell V. Definitions and conceptual issues. In: Spilker B, ed. *Quality of life assessments in clinical trials*. New York: Raven Press, 1990: 11-24.
- 9 Hyland M. Panel discussant. *Respir Med* 1991;85(Suppl B):34.
- 10 Czyzewski DI, Mariotto MJ, Bartholomew K, *et al.* Measurement of quality of well-being in a child and adolescent cystic fibrosis population. *Med Care* 1994;32:965-72.
- 11 Staab D, Wenninger K, Gebert N, *et al.* Quality of life in patients with cystic fibrosis and their parents: what is important besides disease severity? *Thorax* 1998;53:727-31.
- 12 Eiser C, Zoritch B, Hiller J, *et al.* Routine stresses in caring for a child with cystic fibrosis. *J Psychosom Res* 1995;39:641-6.
- 13 Quittner AL, Opiari LC, Regoli MJ, *et al.* The impact of caregiving and role strain on family life: comparisons between the mothers of children with cystic fibrosis and matched controls. *Rehab Psychol* 1992;37:275-90.
- 14 Mullins LL, Olson RA, Reyes S, *et al.* Risk and resistance factors in the adaptation of mothers of children with cystic fibrosis. *J Pediatr Psychol* 1991;16:701-15.
- 15 Thompson RJ, Gustafson KE, Hamlett KW, *et al.* Psychological adjustment of children with cystic fibrosis: the role of child cognitive processes and maternal adjustment. *J Pediatr Psychol* 1992;17:741-55.
- 16 Byrne C. The repression-sensitisation scale: rationale, reliability and validity. *J Pers* 1961;29:334-49.
- 17 Lazarus RS, Folkman S. *Stress, appraisal and coping*. New York: Springer, 1984.
- 18 Moise JR, Drotar D, Doershuk CF, *et al.* Correlates of psychosocial adjustment among young adults with cystic fibrosis. *J Dev Behav Pediatr* 1987;8: 141-8.
- 19 Thompson RJ, Gustafson KE, Hamlett KW, *et al.* Stress, coping and family functioning in the psychological adjustment of mothers of children with cystic fibrosis. *J Pediatr Psychol* 1992;17:573-85.
- 20 Cappelli M, McGrath PJ, MacDonald NE, *et al.* Parent, family and disease factors as predictors of psychosocial functioning in children with cystic fibrosis. *Can J Behav Sci* 1988;20:413-23.
- 21 Aspin AJ. Psychological consequences of cystic fibrosis in adults. *Br J Hosp Med* 1991;45:368-71.
- 22 Suls J, Fletcher B. The relative efficacy of avoidant and non-avoidant coping strategies: a meta-analysis. *Health Psychol* 1985;4:249-88.
- 23 Tullis DE, Guyatt GH. Quality of life in cystic fibrosis. *Pharmacoeconomics* 1995;8:23-33.
- 24 Moriarty C. Quality of life and the effects of dornase alfa in patients with cystic fibrosis. Poster presented at the 2nd Australian Cystic Fibrosis Conference, Perth, Australia, November 1997.
- 25 Keilen M, Treasure T, Schmidt U, *et al.* Quality of life in eating disorders, angina and transplant candidates: are they comparable? *J R Soc Med* 1994; 87:441-4.
- 26 French DJ, Christie MJ, Sowden AJ. The reproducibility of the childhood asthma questionnaires: measures of quality of life for children with asthma aged 4-16 years. *Qual Life Res* 1994;3:215-24.
- 27 Juniper EF, Guyatt GH, Feeny DH, *et al.* Measuring quality of life in children with asthma. *Qual Life Res* 1996;5:35-46.