



Quality of Life and Parental Adjustment in Pediatric Pulmonary Hypertension

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Background: This study examines the impact of pulmonary hypertension (PH) on the quality of life (QoL) of affected youth, as well as the relationships among PH disease severity, parental adjustment variables, and family relational functioning.

Methods: Subjects were 47 eligible parents of children with PH aged 2 to 18 years who were evaluated at Boston Children's Hospital. Measures of patient QoL and of parental stress, coping, and adjustment were administered to the caregivers. Clinicians rated illness severity and family relational functioning.

Results: Youth with PH scored significantly lower than healthy norms on a measure of parent-reported QoL (total, psychosocial, and physical QoL, each $P < .0001$). The parents reported encountering stressful events more frequently than published norms of parents of children with cancer ($P < .0001$). Thirty-four percent of parents of children with PH met criteria for presumed psychiatric diagnosis, and they reported using psychologic coping strategies significantly more often than a normative sample. A summary parental stress measure correlated inversely with child QoL, an effect that held true even after controlling for disease severity ($P = .03$).

Conclusions: PH takes a major toll on children and their families. Decreased QoL among youth with PH was significantly associated with high levels of parental stress, over and above the effect of illness severity. These findings suggest that interventions to better support the caretakers of affected children and adolescents should accompany medical treatment advances so as to improve QoL for patients facing pediatric PH. *CHEST 2014; 145(2):237–244*

Abbreviations: BSI = Brief Symptom Inventory; GARF = Global Assessment of Relational Function; GSI = Global Severity Index; PedsQL = Pediatric Quality of Life Inventory; PH = pulmonary hypertension; PHSI = pulmonary hypertension severity index; PIP = Pediatric Inventory for Parents; QoL = quality of life; RV = right ventricular; SES = socioeconomic status; WCQ = Ways of Coping Questionnaire; WHO = World Health Organization

Pediatric pulmonary hypertension (PH) is a heterogeneous disease marked by elevated pressure in the pulmonary vasculature, leading to progressive right-sided heart failure.^{1,2} Survival for patients diagnosed with pediatric PH has historically been measured in

months. However, new therapeutics and management strategies have significantly decreased mortality and, as a result, increased focus on clinical outcomes beyond survival. The emergence of pediatric PH as a chronic childhood disease requiring significant physical adaptation and complex medical management has increased attention to its impact on health-related quality of life (QoL) and parental adjustment.

Health-related QoL, a construct encompassing physical and psychosocial functioning, is an important indicator of patient well being, reflecting contributions from a variety of domains (eg, health status, family

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relationships, social life, and role function).³ On measures of QoL, youth facing chronic health conditions, including congenital heart disease, have shown decrements in physical and psychosocial functioning relative to healthy peers.³⁻⁶ While QoL has been shown to be severely impaired in adult studies of PH,⁷⁻⁹ few data exist regarding QoL in patients with pediatric PH. There are no data on how the disease affects parental adjustment or family functioning in this illness population.

This prospective pilot study was designed to characterize QoL of patients facing PH and to determine potential correlates of patient QoL, including demographics, PH disease severity, parental adjustment variables, and family relational functioning. We hypothesized that higher PH severity, lower socioeconomic status (SES), longer time since diagnosis, and poorer parent adjustment would be significantly associated with decrements in patient QoL and that the association between illness severity and patient QoL would be partly accounted for by parent adjustment. Finally, exploratory analyses on relationships between coping style and parent adjustment were planned to identify strategies potentially linked with positive adaptation.

MATERIALS AND METHODS

Participants

The study was conducted in the Pulmonary Hypertension Clinic at Boston Children's Hospital, Boston, Massachusetts, from November 2006 to August 2009 with approval of the Committee on Clinical Investigation (protocol number X06-07-0335). Patients had a primary cardiologist who provided longitudinal medical care supported as indicated by a cardiology social worker and/or a child psychiatry consultation. Eligible participants were the caregivers of patients aged 2 to 18 years with PH, identified by screening consecutive clinic schedules.

PH was defined as resting mean pulmonary artery pressure > 25 mm Hg or right ventricular (RV) pressure greater than one-half the systemic arterial systolic pressure.^{2,10} Inclusion criteria consisted of PH diagnosis made at least 3 months prior to study participation and caregiver's ability to complete study questionnaires in English. Children younger than age 2 years were not included due to limitations of validated study instruments. Subjects received a hospital parking voucher and a \$20 gift certificate for participation.

Parents gave consent for the study during scheduled clinic visits. Study questionnaires were distributed by the research staff and completed in writing at the visit. The parent self-identifying as the primary caregiver completed the study questionnaires. Caregivers reported their child's date of birth, ethnicity, sex, and age at PH diagnosis. In addition, parents reported their own date of birth, ethnicity, sex, marital status, level of education, and employment status. Private/public insurance status was recorded as a proxy for SES.

PH Severity Measure

The following variables of medical severity were abstracted from medical records: echocardiographic assessments of RV pressure and function, hemodynamic measurements from most recent cardiac catheterization (cardiac index and response to acute vasodilator testing with response defined as decrease in mean pulmo-

nary artery pressure > 10% to achieve a pulmonary artery pressure \leq 40 mm Hg with no decrease in cardiac output),¹¹ presence of *BMP2* mutation,¹² and current pulmonary vasodilator therapy. To assess symptom severity and physical limitations, cardiology practitioners assigned a four-level World Health Organization (WHO) functional class for each patient based upon age-appropriate activity at the time of questionnaire completion.¹³

A PH severity index (PHSI) was developed to integrate the seven variables into a single index of overall medical severity (Table 1). In selecting variables for the PHSI, we chose measures that have been shown to predict prognosis in patients with PH, including disease etiology, hemodynamics, response to acute vasodilator testing, functional capacity, and current PH therapy. The PHSI score was determined by the total of component scores, with a possible range of 0 to 13. For subjects who had not undergone recent cardiac catheterization (n = 5), PHSI scores were prorated using the five available indexes (maximum sum: 11) such that prorated PHSI = (sum of available scores)/11 \times 13. Two cardiologists independently rated each patient's disease severity for the study; interrater agreement was 100%.

Patient QoL and Parental Adjustment Measures

Caregivers completed a series of survey instruments. These included a patient QoL measure; parental adjustment measures of parental stress, psychopathology, and coping strategies; and a measure of family relational functioning.

Patient QoL Measure: The Pediatric Quality of Life Inventory (PedsQL), Parent Report version, was selected to assess child QoL. With norms for a sample of 8,836 physically healthy children, the PedsQL is a well-established measure designed to reflect the physical, psychological, and social health dimensions of QoL, as delineated by the WHO.¹⁴ The PedsQL has been used with a variety of pediatric populations and has demonstrated sensitivity to group differences in medical status, as well as changes in disease severity.³⁻⁵

Table 1—Pulmonary Hypertension Severity Index^a: Items and Scoring Weights

Items	Scoring Weights
1. RV pressure	0 \leq 0.5 SASP 1 \geq 0.5 SASP, less than SASP 2 \geq SASP
2. RV function	0 = normal 1 = mild dysfunction 2 = moderate dysfunction 3 = severe dysfunction
3. Cardiac index	0 \geq 2.5 L/min/m ² 1 < 2.5 L/min/m ²
4. Response to acute vasodilator testing	0 = response 1 = no response
5. <i>BMP2</i> mutation	0 = not present or unknown 1 = present
6. Medical therapy	0 = none 1 = oral/inhaled 2 = IV prostanoid
7. WHO functional classification	0 = class I (no limitation) 1 = class II (mild limitation) 2 = class III (marked limitation) 3 = class IV (severe limitation)

RV = right ventricular; SASP = systemic arterial systolic pressure; WHO = World Health Organization.

^aCumulative pulmonary hypertension severity index: total of items 1 through 7.

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