

Assessment of Quality of Life in Pediatric Patients with Pulmonary Hypertension

Brian J Hallis BA¹; Kathryn A Tillman APNP³; Cheryl L Brosig PhD³; Mary Krolikowski MSN RN³; Evelyn M Kuhn PhD²; Edward C Kirkpatrick DO³; Stephanie S Handler MD³

1: Medical College of Wisconsin
2: Children's Hospital of Wisconsin
3: Medical College of Wisconsin, Department of Pediatrics

Background: Mortality in pediatric pulmonary hypertension (PH) has improved with new therapies, now resulting in a greater focus on quality of life. The purpose of this study is to quantify functioning across four domains (physical, emotional, social and school) using a validated quality of life survey (PedsQL).

Methods: This was a prospective cohort study of pediatric patients 2-18 years of age with PH. Parents of all children and patients 8-18 years with appropriate developmental capacity completed PedsQL survey in clinic. Results were compared with published norms for pediatric patients, those with cancer and congenital heart disease (CHD).

Results: Twenty-five children were enrolled yielding 13 self and 25 parent-proxy surveys: 5 patients 2-4 years, 3 patients 5-7 years, 9 patients 8-12 years, and 8 patients 13-18 years of age. Four 8-12 year old patients did not complete self survey. Sixteen patients had WHO Group I PAH and 9 WHO Group III PH associated with lung disease. Eight patients received monotherapy, 8 dual therapy and 7 triple therapy for PH. Ten patients were NYHA Class 1 function, 4 Class 2, 10 Class 3, and 1 Class 4.

Overall, the PH cohort had significantly lower scores than healthy children in all PedsQL domains and CHD patients in physical, social and school functioning on both parent-proxy and self surveys [Figure 1]. During survey administration, deficiencies were noted in psychological and school functioning prompting immediate referral for 3 patients.

Conclusion: Quality of life in pediatric PH patients assessed by PedsQL survey revealed functional impairment in multiple domains. Early referral for psychology and/or school intervention may be beneficial to improve quality of life. Administration of the PedsQL during outpatient encounters may provide an easy, reproducible method to assess quality of life in physical, emotional, social and school functioning over time.

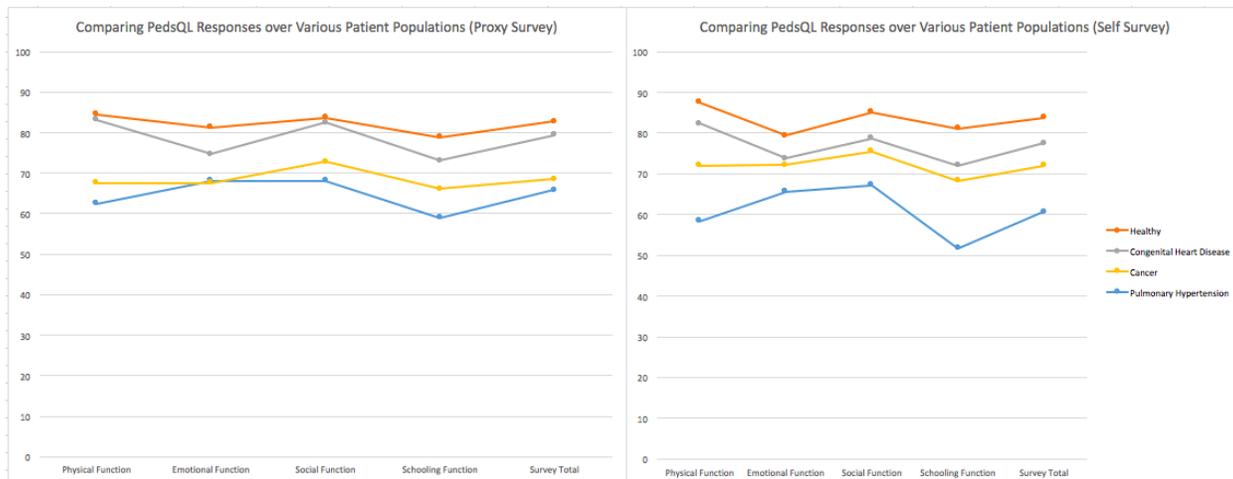


Figure 1