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Assessing Person-Centered Health Outcomes in Sickle Cell Disease Patients

These recommendations are offered as a starting point for consideration. They are not necessarily the best choices for every application and do not substitute for a comprehensive literature review.

Key Domains to Consider in Sickle Cell Disease

We recommend a "life-course" approach as particularly important to measuring health for people with SCD due to the historic focus in SCD care on pediatric health, the importance of transitioning from adolescent to adult SCD care, and the need for advances in our understanding of adults with SCD. Longitudinal assessments depend on patients' symptoms and functional impairment as patients move from childhood to adolescence and throughout adulthood.

Summary

- Recommended primary domains include physical function, cognitive function and social function.
- HealthMeasures offers brief, psychometrically sound measures for these domains.
- We recommend the use of Neuro-QOL. PROMIS contains measures of the same or similar domains as Neuro-QoL and may be useful when comparing functioning across multiple conditions.

Suggested Pediatric HealthMeasures for Primary Domains in Sickle Cell Disease

In childhood, the primary domains of health and functional status that are impacted are **pain, fatigue, emotions and cognition**. PROMIS pediatric measures have been validated in children with sickle cell disease for the domains listed in the left column of the above table entitled Primary HealthMeasures for Sickle Cell Disease. Additional domains that focus on pain (e.g., pain behavior and quality) and physical health (e.g., physical activity) are new and not yet tested in children with sickle cell disease.

Suggested Adult HealthMeasures for Primary Domains in Sickle Cell Disease

Primary Domains: Pain, Stiffness & Fatigue: The Adult Sickle Cell Quality of Life Measurement Information System (ASCQ-Me®) includes assessments of the frequency and severity of pain episodes as well as the impact of **pain**. These measure acute and chronic SCD pain and will indicate how your scores compare to a large sample of patients with SCD seen in various clinics throughout the US. The ASCQ-Me pain episode questionnaire is appropriate if you want to describe the frequency or severity of pain episodes, in particular. The Patient-Reported Outcomes Measurement Information System® (PROMIS®) Pain Interference V1.0 6a (6 questions) short form or CAT are a good alternative assessment of pain, especially for those seeking to compare the severity of SCD pain to pain in other chronic diseases, or to the average amount of pain in the general population. Many adults living with SCD suffer from profound **stiffness** which impacts their ability to move. We recommend use of the ASCQ-Me Stiffness Impact CAT or 5-item short form. These assess the intensity of stiffness and its impact on physical activities. ASCQ-Me was designed to complement PROMIS thus ASCQ-Me does not include a **fatigue** assessment. We recommend use of the PROMIS Fatigue

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CAT or PROMIS Fatigue 13a short form for adults with SCD. These assess frequency, duration, and intensity of fatigue, and its impact on physical, mental and social activities.

Primary HealthMesures for Sickle Cell Disease Patients

Pediatrics	Adults
PROMIS Pediatric Physical Functioning Mobility and Upper Extremity CAT of 8-item SF	ASCQ-Me Pain Impact and Stiffness Impact CAT or 5-item SF
PROMIS Pediatric Pain Interference CAT or 8-item SF	ASCQ-Me Pain Episodes 5-item SF
PROMIS Pediatric Fatigue CAT or 8-item SF	PROMIS Fatigue CAT or PROMIS Fatigue 13a SF
PROMIS Pediatric Depressive Symptoms CAT or 8-item SF	PROMIS Cognitive Functioning CAT and PROMIS Cognitive Function 8a SF
PROMIS Pediatric Anxiety CAT or 8-item SF	ASCQ-Me Emotional, Social Functioning, and Sleep impact CATs and 5-item SF
PROMIS Pediatric Anger CAT or 8-item SF	PROMIS 10-item Global SF

Suggested HealthMeasures for Secondary Domains in Sickle Cell Disease

Other domains of health also can be profoundly affected by SCD and, if resources allow, should be assessed. These include cognitive function, physical function, sleep, social functioning, emotional impact, and global health.

Assessment Times

Assessment times depend on which domains are relevant to your research or clinical practice. Establishing baseline PROs for pain, fatigue, cognition, and other relevant domains is important for determining if and when functional status has returned to baseline after exacerbations subside with or without treatment, or if status is worsening.

Secondary HealthMeasures for Sickle Cell Disease Patients

- PROMIS Cognitive Function CATs, short forms or NIH Toolbox Cognition Battery
- PROMIS Physical Function CAT or PROMIS Physical Function Short form 10a
- ASCQ-Me Sleep Impact CAT or 5-item short form
- ASCQ-Me Social Functioning Impact CAT or 5-item, short form
- ASCQ-Me Emotional Impact CAT or 5-item, short form
- PROMIS Global Health Scale

Learn More!

You can read about CATs and watch a video tutorial at the HealthMeasures.net website here!

Additional Information

The www.ASCQ-Me.org website provides additional information related to the recommended measures including copies of the measures, evidence to support these recommendations, detailed information about scoring and interpretation, an 800 number [866-744-5746] or email address [ascqmeinfo@air.org] for questions. The www.HealthMeasures.net website includes more information about measurement selection, scoring, and interpretation in general and copies of the measures. A Forum on the www.HealthMeasures.net website allows for questions and responses from the HealthMeasures community. The HealthMeasures team is also available for collaboration or consultation via help@healthmeasures.net.

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