Supplementary Online Content


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This supplementary material has been provided by the authors to give readers additional information about their work.
Prorating procedures were used in this study to harmonize constructs across multiple versions of the Wechsler Preschool and Primary Scale of Intelligence (WPPSI), Wechsler Intelligence Scale for Children (WISC) and Wechsler Adult Intelligence Scale (WAIS). For the WPPSI-R and WISC-R versions, the verbal comprehension (VC) and perceptual reasoning (PR) composites were calculated based on the four subtests from the respective Verbal IQ and Performance IQ scales that were found to have the highest correlations with the verbal and performance indexes according to the test manuals. This served to: a) eliminate the tests that were later found to have significant overlap with working memory (WM) or processing speed (PS) constructs, including Mazes, Arithmetic, and Coding; and b) retain a nearly identical set of subtests that were used to measure VC and PR in subsequent versions of the tests (i.e., WPPSI-III and WISC-III). Prorating followed the procedure outlined in the test manual. To calculate WM and PS scores from the WISC-R and WPPSI-R, index scores were derived from scaled scores from a single subtest. For the WISC-R, WM was estimated based on the Digit Span subtest and PS was estimated based on the Coding subtest. For the WPPSI-R, the Arithmetic and Animal Pegs subtests were used to estimate WM and PS, respectively. These individual measures were selected because WM and PS scales on subsequent versions of the tests were based on these tasks or similar variations; these subtests were also found to have high index score correlations within this factor structure. For patients who completed the WPPSI-III, a measure of WM was not available as no subtests measured this construct. For the WISC-V and the WPPSI-IV, the PR index was estimated using the Visual Spatial composite. The test manuals demonstrate that this index has a strong correlation with the perceptual reasoning index and PIQ scores on the previous versions of the Wechsler scales ($r > .60$). No prorating procedures were necessary for versions of the WAIS as a consistent 4-factor structure was retained in all versions used to follow patients in this study.
eFigure 1. Selection of Study Population

Patient Population

139 patients with childhood cALD completed allogeneic HSCT at UMN Medical Center between January 1, 1991 and October 20, 2014

74 patients excluded:
- 71 had pre-HSCT MRI severity score ≥ 10
- 3 had unavailable or insufficient baseline MRI data to determine pre-HSCT MRI severity

Study Selection

65 childhood cALD patients identified who completed allogeneic HSCT with pre-transplant MRI severity score <10

3 patients excluded:
- 2 patients had no available neuropsychological data
- 1 patient had pre-existing intellectual disability unrelated to cALD

Baseline

62 patients had valid pre-HSCT neurocognitive evaluation

29 patients lacking long-term follow up:
- 7 patients died within first two years
  - 4 died from transplant-related complications or infections
  - 1 died from cALD disease progression
  - 2 cause of death unknown
- 22 patients survived but did not return for 2-year post-HSCT evaluation or later

Long-term Follow Up

33 patients had long-term neurocognitive follow up data
eFigure 2. Correlation Between Pretransplant Measures of Neurocognitive Functioning and MRI Severity Scores in 62 Patients With cALD
**eFigure 3.** Individual Neurocognitive Trajectories of 62 Patients With cALD After HSCT on Three Standardized Neurocognitive Measures (Normative Mean = 100 ± 15)

Shaded regions depict age-typical performance.