Children's Hospital of Philadelphia
PolicyLab

Neurodevelopmental outcome of infants with congenital diaphragmatic hernia prospectively enrolled in an interdisciplinary follow-up program

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PURPOSE: The purpose of the study was to evaluate the neurodevelopmental outcome in infants with congenital diaphragmatic hernia (CDH).

METHODS: Between June 2004 and September 2007, 41 CDH survivors were prospectively enrolled in an interdisciplinary follow-up program. Neurodevelopmental status was evaluated using the Bayley Scales of Infant Development II (prior 2006, n = 9), the Bayley Scales of Infant Development III (after 2006, n = 27), or the Wechsler Preschool and Primary Scale of Intelligence III (children older than 4 years, n = 5). Scores were grouped as average, mildly delayed, and severely delayed by standard deviation intervals (115-85, 71-84, <70), and mixed if average and mildly delayed in either cognitive or language.

RESULTS: Median age at last assessment was 24 months (range, 6-62). Average, mixed, mildly delayed, and severely delayed scores for neurocognitive and language skills were found in 49%, 19%, 17%, and 15%, respectively. Psychomotor scores were normal, mildly delayed, and severely delayed in 46%, 23%, and 31%, respectively. Autism was present in 7%. Abnormal muscle tonicity was found in 51% (49% hypotonic, 2% hypertonic). Multivariate risk factors for borderline or delayed neurodevelopmental, neurocognitive, and/or psychomotor outcome were intrathoracic liver position (P = .02), presence of a right-sided CDH (P = .02), extracorporeal membrane oxygenation need (P < .001), Gore-Tex patch repair (P = .02), O(2) requirement at 30 days of life (P < .01), and hypotonicity (P < .01).

CONCLUSIONS: The prospective evaluation in an interdisciplinary follow-up program uncovered striking morbidities in neurodevelopmental status in approximately half of the CDH infants. The most common neurologic sequelae are neuromuscular hypotonicity and psychomotor dysfunction. Patient-specific factors are important determinants of adverse neurologic outcome.

Authors:

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