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Approaches to neurodevelopmental assessment in congenital diaphragmatic hernia survivors

Catherine Chen[a,*], Sandra Friedman[b], Samantha Butler[b], Stefanie Jeruss[c], Norma Terrin[c], Hocine Tighiouart[c], Janice Ware[b], Jay M. Wilson[a], Susan K. Parsons[c]

[a]*Department of Surgery, Children's Hospital, Boston, MA 02115, USA*
[b]*Department of Medicine, Children's Hospital, Boston, MA 02115, USA*
[c]*Institute for Clinical Research and Health Policy Studies, Tufts-New England Medical Center, Boston, MA 02111, USA*

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Abstract

Background: Infants with congenital diaphragmatic hernia require complex surgical care and may have neurodevelopmental morbidity. We examined the performance of reports of motor functioning in 25 congenital diaphragmatic hernia survivors using the parent-completed Developmental Profile-II and a clinical evaluation by a neurodevelopmental pediatrician (MD) measured against the Bayley motor scale.

Methods: Bayley motor scores were dichotomized as normal or abnormal. Sensitivity and specificity were calculated for each test.

Results: The median age at assessment was 25 months. Bayley motor scores were abnormal in 77% of infants tested (10/13). The MD examinations detected motor problems in 92% (12/13). Sensitivity and specificity of the MD examination were 1.0 and 0.33, respectively. Developmental Profile-II physical scores were abnormal in 15% (2/13); sensitivity and specificity were 0.2 and 1.0, respectively.

Conclusions: The high rate of abnormal motor findings in this study supports the need for ongoing screening and evaluation. The sensitivity of MD examinations was excellent, but hypotonia findings were not universally corroborated by the Bayley. Although specificity of parent-reported motor findings was high, parents underreported abnormal motor findings. Parental reports of neurodevelopmental problems should be heeded, and physicians should perform screening motor examinations. Bayley assessments may be warranted to determine the functional implications of observed abnormalities.

Congenital diaphragmatic hernia (CDH) is a congenital malformation of an incompletely formed diaphragm, requiring surgical repair as a neonate. In addition to pulmonary hypoplasia, neonates with CDH may also have associated cardiac anomalies, pulmonary hypertension, or other genetic syndromes, which complicate their postnatal management. The complexity of care often required in the peri- and postoperative period for CDH patients renders CDH the most expensive of any pediatric surgical condition based on the number of diagnosis related group units [1]. Congenital diaphragmatic hernia survivors have been reported to experience ongoing medical morbidity, including pulmonary and nutritional morbidity, and gastroesophageal reflux [2-4]. In case series of CDH survivors treated with neo-

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* Corresponding author. Tel.: +1 617 355 0535; fax: +1 617 730 0298.
E-mail address: catherine.chen@childrens.harvard.edu (C. Chen).

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natal extracorporeal membrane oxygenation (ECMO), difficulties with developmental delay, hearing loss, and musculoskeletal development have been described [5,6]. We have found that a subset of long-term CDH survivors with ongoing clinical problems a median of 8 years after surgery has lower functional status [7]. In addition, family impact is profound and long-standing for this subset of CDH survivors with more severe conditions and ongoing morbidity (Chen et al, unpublished data).

The CDH population provides a unique model of a complex pediatric surgical condition, which has become a paradigm of chronic disease. Few studies have reported the neurodevelopmental outcomes of CDH survivors in the contemporary era since 2000, when in-hospital survival has been reported to be greater than 90% at several institutions [8,9]. In this pilot study, we examined 3 independent approaches to neurodevelopmental assessment of young CDH survivors, focusing on motor functioning. We evaluated the performance of reports of motor functioning using 2 screening measures, the parent-completed Developmental Profile-II (DP-II) and a clinical evaluation by a neurodevelopmental pediatrician (MD), measured against the diagnostic Bayley motor scale administered by a psychologist.

1. Methods

1.1. Patient selection and data collection

We identified 68 patients with CDH who survived surgical repair to hospital discharge at Children's Hospital Boston from January 1, 2000 through December 31, 2003. The in-hospital survival rate during this period was 87%. Patients were excluded from this Institutional Review Board approved study (#X03-02-009) if they received preoperative or postoperative care at Children's Hospital Boston but underwent surgical repair of CDH at another institution. Of the 68 survivors, 19 patients could not be reached, either by mail or by telephone, or via an identifiable primary care physician. The evaluable cohort of 49 patients was recruited by mail to participate in this study and asked to return an opt-out postcard to provide consent. Twenty-five patients participated in this pilot study. Reasons cited for nonparticipation included "not interested" for 8 patients, "too much to participate" for 6 patients, and "would if asked at a different time" for 1 patient.

Perinatal and perioperative data were obtained for the cohort of 68 established CDH survivors by review of hospital medical records. Medical issues at the time of hospital discharge and at the time of the study were defined by the presence of any of the following: cardiac issues (need for diuretics, inotropes, or antihypertensive medications); pulmonary issues (need for bronchodilators, steroids, or home oxygen therapy); or gastrointestinal issues (need for parenteral nutrition, gastrostomy tube feeds, or medications for gastroesophageal reflux).

1.2. Study instruments and assessments

Three independent approaches to neurodevelopmental assessment of motor functioning were used in this study.

1.2.1. Parent report

All 25 study participants completed the DP-II, a norm-referenced developmental screening measure administered as a structured interview (by author S.J.) that asks parents to estimate their child's current level of functioning [10]. The DP-II contains 217 developmental tasks for the age range from birth to 12 years and addresses 5 functional domains: physical, adaptive, social, academic, and communication. Domain scores are obtained using established scoring algorithms and are interpreted in terms of the degree of developmental lag between chronological age and current levels of functioning. Norm tables are used to determine the significance of lags. This study focused on the DP-II physical domain score as a measure of parent-reported motor functioning. Delayed or borderline delayed DP-II physical scores were classified as 'abnormal.'

1.2.2. Physician's clinical evaluation

Twenty-three study participants received a clinical evaluation by a neurodevelopmental pediatrician (author S.F.; referred herein as MD) as part of their routine follow-up in the CDH Outpatient Multidisciplinary Clinic (referred herein as CDH Clinic). Two patients did not see the MD at a time compatible with the DP-II interview or the Bayley assessment. Clinic notes were extracted, and motor findings on physical examination, if any, were tabulated for each patient.

1.2.3. Bayley Scales of Infant Development-II

Thirteen study participants provided consent and completed the psychologist-administered (author S.B.)-administered diagnostic Bayley Scales of Infant Development-II, which was used as our gold standard. The Bayley Scales of Infant Development-II measures mental and motor development and tests the behavior of infants from 1 to 42 months of age [11]. Twelve patients did not consent to the Bayley, which was obtained for research purposes in this study and not referred based on clinical examinations. This study focused on the motor scale, which assesses the degree of body control, large muscle coordination, fine motor skills of the hands and fingers, dynamic movement, postural imitation, and stereognosis. Bayley motor scores were obtained using established scoring algorithms and compared with age appropriate norms. Scores were further dichotomized as normal or abnormal by the psychologist. Abnormal scores reflect those classified as "mildly delayed performance" (score range, 70-84) or "significantly delayed" (score below 69).

1.3. Statistical analysis

Summary statistics were calculated for each outcome variable, and descriptive results were summarized for each cohort tested. Sensitivity and specificity were calculated for