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Developmental Screening in Children with Congenital Heart Disease: Ages and Stages Questionnaires

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Abstract

Objective: Standardized developmental screening tools are important for the evaluation and management of developmental disorders in children with congenital heart disease (CHD). However, psychometric properties and clinical utility of screening tools, such as the Ages and Stages Questionnaires, Third Edition (ASQ-3), have not been examined for the CHD population. We hypothesized the ASQ-3 would be clinically useful for this population.

Study Design: ASQ-3 developmental classifications for 163 children with CHD at 6, 12, 24 and/or 36 months of age were compared to those obtained from concurrent developmental testing with the Bayley Scales of Infant and Toddler Development, Third Edition.

Results: When ASQ-3 screening failure was defined as ≥ 1 standard deviations below the normative mean, specificity (81.9%) and negative predictive value (81.0%) were high across ASQ-3 areas. Sensitivity was high for gross motor (79.6%), increased with age for communication (35.7%–100%), and generally decreased with age for problem solving (73.1%–50.0%). When ASQ-3 screening failure was defined as ≥ 2 standard deviations below the normative mean, specificity (93.6%) and positive predictive value (74.5%) were generally high across ASQ-3 areas, but sensitivity was low (31.1%) to fair (62.8%). The ASQ-3 improved accuracy in predicting delays over clinical risk factors alone.

Conclusions: The ASQ-3 appears to be a clinically useful tool for screening development in children with CHD, although utility varied based on developmental area and time point. Clinicians are encouraged to refer children scoring ≥ 1 standard deviation below the normative mean on any ASQ-3 area for formal developmental evaluation.

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Conflicts of Interest: None.

Keywords

assessment; neurodevelopment; cardiology

Introduction

Children with congenital heart disease (CHD) who underwent surgical intervention during infancy are at high risk for developmental challenges, including language and motor delays, adaptive behavior deficits, and learning disabilities.¹ The American Heart Association published a statement in 2012 recommending periodic developmental surveillance, screening, evaluation, and reevaluation throughout childhood to identify deficits and provide appropriate therapies and education.² While several widely used developmental screening tools demonstrate good psychometric properties for the general pediatric population, including the Ages & Stages Questionnaires, Third Edition (ASQ-3)^{3, 4, 5, 6} their psychometric properties and clinical utility have not been examined for the CHD population.

The ASQ-3 is a set of parent questionnaires that measure development in communication, gross motor, fine motor, problem solving, and personal-social domains for children ages 1 to 66 months.⁶ The ASQ-3 has yielded mixed empirical support with select pediatric illness populations including premature birth,^{7, 8, 9} neurosensory disability,¹⁰ and hypoxic-ischaemic encephalopathy.¹¹ In the context of CHD, the ASQ-3 has been used for developmental outcomes research,^{12, 13, 14, 15} evaluated as a predictor of parent-reported developmental concerns three years later,¹⁶ and incorporated into national collaborative quality improvement efforts (e.g., National Pediatric Cardiology - Quality Improvement Collaborative), yet studies have not evaluated psychometric properties or clinical utility of the ASQ-3 for this patient population. This study aimed to evaluate the ASQ-3 as a developmental screening tool in infants and toddlers with CHD by comparing developmental classifications obtained via ASQ-3 to those obtained from the gold standard of developmental classification, the Bayley Scales of Infant and Toddler Development, Third Edition (Bayley-III).¹⁷ A secondary aim was to evaluate the added utility of the ASQ-3 after accounting for known clinical risk factors for developmental disorders in the CHD population. We hypothesized that the ASQ-3 would demonstrate clinical utility for developmental screening and improve accuracy in predicting developmental delays as compared to clinical risk factors alone.

Methods

Participants and Procedures

Participants were 163 children ages four to 40 months who underwent cardiac surgery during infancy and completed at least one neurodevelopmental evaluation through the Nemours Cardiac Center's developmental follow-up program between 2012–2014. Neurodevelopmental evaluations were administered at the approximate ages of 6, 12, 24 and/or 36 months with the Bayley-III and ASQ-3. Parents also completed a brief sociodemographic questionnaire. Select clinical variables were extracted from the medical record, including gestational age at birth, confirmed or suspected genetic syndromes, single

versus two-ventricle cardiac anatomy, age at first cardiac surgery, length of hospital stay, and cumulative lifetime duration of cardiopulmonary bypass and deep hypothermic circulatory arrest. This study was reviewed and approved by the Nemours Institutional Review Board with waiver of the requirement for obtaining informed consent/parental permission.

Measures

Bayley-III.—The Bayley-III assesses developmental functioning of children one to 42 months of age through structured play activities. Scaled scores (mean of 10, standard deviation of 3) for cognitive, receptive communication, expressive communication, fine motor and gross motor development are calculated from raw scores, and composite scores (mean of 100, standard deviation of 15) for cognitive, language, and motor development are calculated from scaled scores. Psychometric properties are well defined and acceptable.¹⁷ The Bayley-III was administered by a clinical psychologist or psychology postdoctoral fellow. An interpreter was utilized for Spanish speaking families. Bayley-III adaptive behavior and social-emotional parent questionnaires were not utilized as part of the neurodevelopmental evaluation and were therefore not included in this study.

ASQ-3.—The ASQ-3 is a set of parent-report age-specific developmental screening questionnaires assessing communication, gross motor, fine motor, problem solving, and personal-social skills.⁶ Each age-specific questionnaire has 40 items: 6 items per area and 10 open-ended questions. Response options for closed-ended questions are: “yes” when the behavior is present (10 points), “sometimes” when the behavior is emerging (5 points), or “not yet” when the behavior is absent (0 points). Points within each developmental area are summed and resulting scores are compared against screening cutoff scores corresponding to one (“Monitor”) and two (“Refer”) standard deviations below the normative mean. The ASQ-3 requires 5–10 minutes to complete and a sixth grade reading level. The Spanish version of the ASQ-3 was utilized when appropriate. Parents completed the ASQ-3 without examiner input during each neurodevelopmental evaluation visit, following administration of the Bayley-III.

Data Analyses

Receiver operating characteristic analyses using a 2×2 pass/fail design were conducted to determine sensitivity (rate of true positives), specificity (rate of true negatives), positive predictive value (probability that screening failure reflects true delay), and negative predictive value (probability that a score in the normal range reflects true normative development) of the ASQ-3 in identifying cognitive, language, fine motor, and gross motor developmental delays as measured by the Bayley-III. ASQ-3 areas were compared to Bayley-III domains measuring similar developmental skills: problem solving area of the ASQ-3 to the cognitive scaled score of the Bayley-III, communication area to the language composite score, fine motor area to the fine motor scaled score, gross motor area to the gross motor scaled score. The Bayley-III language composite was used rather than receptive and expressive communication scaled scores as the ASQ-3 does not screen for receptive and expressive communication delays separately. The personal-social area of the ASQ-3 was not included in analyses, as Bayley-III parent questionnaires measuring similar constructs (i.e., adaptive behavior, social-emotional) were not administered.

The first set of receiver operating characteristic analyses defined screening failure on the ASQ-3 as 1.0 standard deviation below the normative mean (i.e., score within the “Monitor” or “Refer” ranges).¹⁸ A second set of receiver operating characteristic analyses utilized a more stringent definition of screening failure: 2.0 standard deviations below the normative mean (i.e., score within the “Refer” range). Developmental delay as measured by the Bayley-III was defined as 1.0 standard deviation below the normative mean (scaled score 7, composite score 85) for all analyses. The decision to utilize a broad definition of developmental delay, capturing both those children with a mild or emerging delay as well as those exhibiting severe delay, was made by considering the purpose of developmental screening for identifying those children who would benefit from formal evaluation but may not be identified through surveillance alone. In this patient population, mild to moderate developmental delays are quite common and warrant parent anticipatory guidance, recommendation for home activities, and/or early intervention.¹⁹ Receiver operating characteristic analyses were performed separately for each time point (6, 12, 24, and 36 months) and using aggregated data across all time points.

Logistic regression models assessed the utility of the ASQ-3 for predicting developmental delays after accounting for a set of known clinical risk factors in the CHD population.² Length of hospital stay for first surgical admission and presence versus absence of premature birth, single ventricle anatomy, and confirmed or suspected genetic syndrome were entered in block 1 of each model and ASQ-3 screening failure defined as 1.0 standard deviation below the normative mean was entered in block 2 (one model for each of the four developmental domains). Participants who had not undergone cardiopulmonary bypass were excluded. To reduce the number of analyses and risk for type II error, logistic regression analyses were conducted for the 12 month time point only as it provided the largest sample size.

Results

Sample Characteristics

A total of 250 neurodevelopmental evaluations (ASQ-3 and Bayley-III) from 163 patients were included (6-months: n = 59; 12-months: n = 90; 24-months: n = 69; 36 months: n = 32). Mean ages at each evaluation were as follows: 6.4±1.0 months; 12.9±1.5 months; 24.3±2.2 months; 36.5±1.5 months. Ninety participants completed one evaluation, 59 completed two, and 14 completed three. Sociodemographic, clinical, and neurodevelopmental characteristics of participants are displayed in Table 1. Bayley-III motor composite scores tended to be lower than cognitive and language composite scores across time points.

ASQ-3 Screening Failure Defined as 1.0 Standard Deviation below the Normative Mean

Table 2 displays area under the curve, sensitivity, specificity, positive predictive value, and negative predictive value for each ASQ-3 area with ASQ-3 screening failure defined as 1.0 standard deviation below the normative mean. Across all developmental areas, specificity was excellent (81.9%) and the probability that a score in the normal range reflected true normative development was high (negative predictive value 81.0%). Sensitivity and

positive predictive value varied by developmental area. The gross motor area demonstrated excellent sensitivity (79.6%) in identifying children with delay and screening failure was associated with a high probability of true delay (positive predictive value = 84.9%). Screening failure in the communication area was also associated with a high probability of true delay (positive predictive value = 81.4%), but its sensitivity was lower (67.8%). The problem solving and fine motor areas demonstrated only fair sensitivity and relatively low positive predictive value (<70%).

Examination of the ASQ-3 at individual time points (Supplemental Table 1) revealed that sensitivity increased with age for the communication area (6 months = 35.7%; 12 months = 74.3%; 24 months = 90.0%, 36 months = 100.0%) and decreased between 12- and 36-months of age for the problem solving area (12 months = 73.1%; 24 months = 66.7%, 36 months = 50.0%) (Figure 1).

ASQ-3 Screening Failure Defined as 2.0 Standard Deviation below the Normative Mean

Table 2 displays area under the curve, sensitivity, specificity, positive predictive value, and negative predictive value using a more stringent definition of screening failure: 2.0 standard deviations below the normative mean. Specificity was excellent across all areas (93.6%). However, sensitivity values were low to fair, ranging from 31.1% (communication) to 62.8% (gross motor). Screening failure was associated with a high probability of true delay for the communication, fine motor and gross motor areas (positive predictive value 85.1%), but with a somewhat lower probability of true delay for the problem solving area (positive predictive value = 74.5%). The probability that a score in the normal range reflects true normative development was high for the problem solving area (negative predictive value = 81.6%), but lower for the communication, fine motor and gross motor areas (negative predictive value ranging from 68.7 – 75.0%).

The probability that a score in the normal range reflects true normative development increased with age for the communication area (negative predictive value: 6 months = 53.9%; 12 months = 64.5%; 24 months = 84.0%, 36 months = 85.0%) (Supplemental Table 2).

Added Utility of ASQ-3 beyond Clinical Risk Factors

After accounting for known clinical risk factors for developmental disorders, the ASQ-3 significantly predicted Bayley-III developmental classifications at 12 months of age. Compared to the model with clinical risk factors only, the addition of the ASQ-3 increased the percentage of correctly classified participants from 72.3 to 80.7% for the Bayley-III cognitive domain, 68.8 to 87.5% for language, 71.4 to 78.6% for fine motor, and 76.2 to 86.9% for gross motor (Table 3). ASQ-3 screening failure in problem solving, communication, fine motor, and gross motor areas was associated with odds ratios of 7.65 (95% CI = 2.33–25.15), 209.20 (14.49–3020.20), 3.82 (1.29–11.33), and 49.96 (8.05–309.94), respectively, for developmental delay in corresponding Bayley-III domains.

Discussion

While standardized developmental screening tools are important for the evaluation and management of developmental disorders in children with CHD, this is the first study to our knowledge examining the psychometric properties and clinical utility of a specific developmental screening tool for this population. We have shown that the ASQ-3 has clinical utility for screening development in infants and toddlers with CHD and improves the accuracy in predicting delays over clinical risk factors alone.

The ASQ-3 appears particularly useful for screening gross motor development, with 80% of mild to severe gross motor delays and 88% of typical development identified using the 1 standard deviation cutoff. This is in contrast to a prior investigation of preterm infants in which the ASQ-3 failed to accurately discriminate participants with motor delays.⁷ While these varied results may be partially attributable to methodological differences between the studies (second versus third edition of the Bayley Scales; mode of ASQ-3 administration), it is possible that the ASQ-3 has increased utility in the gross motor domain for infants and toddlers with CHD as compared with preterm infants. A developmental screening tool with high sensitivity and specificity in the area of gross motor is relevant for this population as gross motor delays are among the most common developmental challenges following infant heart surgery.^{20, 21} Furthermore, the ASQ-3 demonstrated clear benefit over a review of risk factors alone for prediction of gross motor delay. It is not known how the ASQ-3 gross motor area would fare in comparison to routine physician-based surveillance and examination, and this should be examined in future studies.

Clinical utility of the ASQ-3 for screening cognitive and language development varied by time point, similar to ASQ-3 findings reported with hypoxic-ischemic encephalopathy¹¹ and preterm birth.⁷ The ASQ-3 appears useful for screening cognitive development in infants but not toddlers, with only 67% and 50% of cognitive delays identified in the 24- and 36-month samples, respectively. Some items used to assess cognitive/problem solving development on the ASQ-3 at 24- and 36-months of age may not be commonly observed in the home environment (e.g., “Show your child how to make a bridge with blocks [...] Does your child copy you by making one like it?”), when compared to 6-months of age (e.g., “Does your baby play by banging a toy up and down on the floor or table?”) and therefore may not be as accurately reported. Conversely, the ASQ-3 appears useful for screening language development in toddlers but not infants. Prelinguistic communication in infancy, such as responsiveness to sounds and non-word vocalizations/gestures, may be particularly susceptible to subjective interpretation. Clinicians should use caution when interpreting problem solving and communication results for these age groups and consider supplementing the ASQ-3 with parent interview and behavioral observations.

The ASQ-3 has two screening cutoff scores corresponding to one standard deviation (“Monitor”) and two standard deviations (“Refer”) below the normative mean. While continuing to monitor children from the general population who score between one and two standard deviations below the normative mean may be appropriate, children with CHD exhibit a pattern of low-severity or combined developmental disabilities that can impact functioning if not addressed.^{1,20} Recognition of this pattern has led clinicians and

researchers to recommend formal developmental evaluation and intervention even in the case of mild delays.^{2,20} Results of the present study suggest that referring only children whose scores fall 2 or more standard deviations below the normative mean for evaluation would result in missing as many as 69% of children with a mild to severe delay, although the rate of false positives would decrease from 10–18% with the 1 standard deviation cutoff to as low as 1–6% with the 2 standard deviations cutoff. As with any screening tool, the optimal cutoff score depends on a balance of sensitivity and specificity dependent on risk associated with over- or under-identifying the condition. Formal developmental evaluation has no physical risks and is often helpful even in the absence of significant delay, as it is an opportunity to provide parent education/consultation and make recommendations for home activities to promote development¹⁹ and could prevent more significant problems from developing. Therefore, clinicians are encouraged to refer children scoring one or more standard deviations below the normative mean on any ASQ-3 area for developmental evaluation.

Positive predictive value and negative predictive value provide important information that clinicians can reference when counseling families about a child's risk for delay and need for formal evaluation. Children scoring at or below the 1 standard deviation cutoff for the gross motor and communication areas had an 85% and 81% probability of true delay, respectively, and those scoring at or below the 2 standard deviations cutoff for these same areas had a 94% and 93% probability of true delay, respectively. Therefore, clinicians can be confident in their referral for evaluation in the context of low ASQ-3 gross motor and communication scores. The probability of true delay among children scoring 1 or 2 standard deviations below the normative mean on the problem solving or fine motor areas of the ASQ-3 was somewhat lower, likely because these delays are less obvious to parents and have a lower base rate in this patient population during infancy/toddlerhood.^{15, 20}

The probability that a score in the normal range reflects true normative development ranged from 81% to 87% with the 1 standard deviation cutoff and from 69% to 82% with the 2 standard deviations cutoff, underscoring the importance of periodic developmental evaluation and reevaluation for children considered high-risk for developmental delay based on medical history, regardless of screening results.² Basing referral for evaluation on a screening tool alone would likely result in missing a number of children who may be experiencing subtle challenges that warrant consultation and intervention. Similarly, a screening tool cannot replace formal evaluation, even when a clinician suspects delay based on screening results. Comprehensive evaluation is necessary to identify strengths and weaknesses, provide recommendations, and develop treatment plans.^{2, 19}

The added utility of the ASQ-3 to risk factor analysis is supportive of a model of combined developmental surveillance (including risk factor review) and screening. While many pediatric cardiac centers now offer developmental evaluation as part of routine care for children with CHD,¹⁹ this is not yet universal. The availability of a brief, validated screening tool that can be completed during a medical visit may be particularly helpful in the absence of an on-site developmental follow-up program. Individualized information about a child's development (e.g. positive predictive value and negative predictive value data indicating a child's risk of delay) can be used to counsel families on the importance of formal

developmental evaluation, which may be more compelling to parents than clinical risk factors alone.

This study utilized data from a single institution's developmental follow-up program, which serves predominantly patients who are at high-risk for developmental disorders, precluding separate examination of ASQ-3 utility in low-risk patients. The ASQ-3 was not administered at the specific time points recommended by American Heart Association guidelines as the structure of this developmental follow-up program predated publication of the guidelines. However, the time points do align with other national collaborative efforts to implement ASQ-3 screening within this patient population (e.g., National Pediatric Cardiology - Quality Improvement Collaborative). Future research should evaluate the clinical utility of the ASQ-3 at time points recommended by American Heart Association guidelines, with separate analyses for subgroups known to be at varying risk for developmental delays, such as children with CHD within the context of a genetic syndrome, those with single ventricle anatomy, or those classified as low-risk for developmental disorders based on medical history. Most children completed one or two evaluations during the period of study, resulting in a different subset of children for each time point. Future studies are needed to evaluate performance trends as well as changes in ASQ-3 psychometric properties and clinical utility longitudinally with the same cohort. Lastly, the personal-social area of the ASQ-3 was not examined in this study, but should be evaluated in future studies given research suggesting impairments in adaptive behavior and social cognition in this patient population².

Conclusion

The ASQ-3 demonstrates utility for the developmental screening of infants and toddlers with CHD and provides additional predictive accuracy above and beyond clinical risk factors. Additional research is needed to evaluate ASQ-3 psychometric properties and clinical utility when administered longitudinally at multiple congenital heart centers to parents of children with varying CHD severity and comorbidities. A developmental screening tool validated for this patient population provides the opportunity to identify developmental concerns early and refer for formal evaluation and intervention to minimize impact on quality of life.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Figure 1.

Sensitivity of ASQ-3 Areas by Time Point

*ASQ-3 screening failure defined as 1.0 standard deviation below the normative mean

Table 1.**Participant Characteristics and Neurodevelopmental Outcomes**

Sociodemographic Characteristics		Frequency (%)			
Child Gender (Male)		87 (53%)			
Child Race/Ethnicity					
Non-Hispanic White		103 (63%)			
Black or African-American		27 (17%)			
Hispanic/Latino		21 (13%)			
Other		12 (8%)			
Education Level of Parent Respondent ^a					
High School		50 (31%)			
Partial College		30 (18%)			
College Degree or Higher		64 (39%)			
Clinical Characteristics		Median (1Q^b – 3Q^c)			
Age at First Cardiac Surgery (Days)		24 (6 – 64)			
Days in CICU ^d (First Surgical Admission)		7 (3 – 14)			
Days in Hospital (First Surgical Admission)		16 (9 – 30)			
		Frequency (%)			
Prematurity (<37 weeks)		32 (20%)			
Single Ventricle Anatomy		29 (18%)			
Confirmed Genetic Syndrome		35 (22%)			
Suspected Genetic Syndrome		20 (12%)			
Surgery with CPB ^e (Lifetime)		144 (88%)			
Surgery with DHCA ^f (Lifetime)		99 (61%)			
Neurodevelopmental Outcomes		Mean (SD)			
Bayley-III Composite		6 Months	12 Months	24 Months	36 Months
Cognitive		96.3 (17.7)	92.7 (14.9)	91.5 (15.5)	95.2 (9.2)
Language		84.1 (15.0)	88.9 (17.0)	89.3 (18.9)	94.7 (17.4)
Motor		86.4 (20.5)	84.8 (18.4)	87.8 (14.6)	91.6 (14.1)

^aMissing data for 19 participants (11%);^b1Q = first quartile;^c3Q = third quartile;^dCICU = cardiac intensive care unit;^eCPB = cardiopulmonary bypass;^fDHCA = deep hypothermic circulatory arrest;^eBayley-III = Bayley Scales of Infant and Toddler Development, Third Edition.

Table 2.

Area Under the Curve

ASQ-3 Area	n ^a	Prevalence of Delay	AUC ^b	Sensitivity	Specificity	PPV ^c	NPV ^d
ASQ-3 Screening Failure Defined as 1.0 SD^e Below the Normative Mean							
Problem Solving	239	28%	0.76	0.69	0.82	0.60	0.87
Communication	228	39%	0.79	0.68	0.90	0.81	0.81
Fine Motor	242	35%	0.74	0.66	0.82	0.67	0.82
Gross Motor	244	46%	0.84	0.80	0.88	0.85	0.83
ASQ-3 Screening Failure Defined as 2.0 SD^e Below the Normative Mean							
Problem Solving	239	28%	0.70	0.47	0.94	0.75	0.82
Communication	228	39%	0.65	0.31	0.99	0.94	0.69
Fine Motor	242	35%	0.68	0.40	0.96	0.85	0.75
Gross Motor	244	46%	0.80	0.63	0.96	0.93	0.75

^a n = number of assessments;

^b AUC = area under the curve;

^c PPV = positive predictive value;

^d NPV = negative predictive value;

^e SD = standard deviation;

Table 3.

Percentage of Correctly Classified Participants at 12 Months

Bayley-III Domain ^a	n	Clinical Variables Alone ^b	Clinical Variables + ASQ-3 ^c
Cognitive			
No Delay	53	93.0%	91.2%
Delay	30	26.9%	57.7%
Language			
No Delay	54	87.0%	97.8%
Delay	26	44.1%	73.5%
Fine Motor			
No Delay	53	88.7%	92.5%
Delay	31	41.9%	54.8%
Gross Motor			
No Delay	47	85.4%	85.4%
Delay	37	67.4%	88.4%

^aDelay defined as 1.0 SD below the normative mean;^bIncludes premature birth, single ventricle anatomy, confirmed/suspected genetic syndrome, and length of hospital stay (first surgical admission);^cIncludes ASQ-3 delay defined as 1.0 SD below the normative mean and clinical variables.