General Health Status of Children With d-Transposition of the Great Arteries After the Arterial Switch Operation

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Background—To study the long-term impact on general health status of d-transposition of the great arteries (D-TGA) after the arterial switch operation (ASO) during infancy, we asked parents to complete the Child Health Questionnaire, Parent Form-50 when their children were 8 years old.

Methods and Results—Of 160 eligible patients, questionnaires were completed for 155 subjects (96%). Median age at surgery was 6 days (range 1 to 67 days), and median age at completion of the Child Health Questionnaire was 8.1 years (7.6 to 10.0 years). Subsequent to questionnaire completion, children underwent psychometric testing. Mean Physical Health Summary and Psychosocial Summary scores were 54.0 ±6.6 and 49.7 ±9.9, respectively, which were similar to those of normal subjects. Compared with the normative sample, parents of D-TGA patients reported more problems with attention, learning, and speech, as well as greater frequency of developmental delay (P<0.001 for each). Worse Psychosocial Summary scores were significantly associated with lower full-scale IQ (P=0.001) and lower achievement in reading (P=0.005) and math (P=0.007). Worse Physical Health Summary scores were associated with longer hospital stay after the ASO (P=0.02). General health status scores were not significantly related to presence of ventricular septal defect, age at surgery, perfusion variables during the ASO, sex, or history of cardiac reoperation.

Conclusions—At age 8 years, children with D-TGA after ASO have an overall physical and psychosocial health status similar to that of the general population. Lower IQ and academic achievement are associated with worse psychosocial health status, whereas longer hospital course after initial surgery is associated with worse physical health status. (Circulation. 2001;104[suppl I]:I-138-I-142.)

Key Words: heart defects, congenital ▪ transposition of great vessels ▪ surgery ▪ pediatrics ▪ quality of life

Studies on outcomes after infant heart surgery generally describe mortality and morbidity, but data are limited regarding the general health status or health-related quality of life of affected children. These children are encountering new challenges that affect their overall health as they reach school age. Although many instruments have been available to assess adult quality of life, tools for use in pediatric patients have been developed only recently. The Child Health Questionnaire, Parent Form-50 is a comprehensive, multidimensional health assessment tool designed to measure physical and psychosocial functioning in children and adolescents. The questionnaire is generic, allowing for comparison of health status among children in the general population and among children with specific medical conditions. The Child Health Questionnaire has been normed and validated in 391 children and benchmarked for children with asthma, juvenile rheumatoid arthritis (JRA), and attention deficit hyperactivity disorder (ADHD). We used the Child Health Questionnaire, Parent Form-50 to explore the general health status of 8-year-old children with d-transposition of the great arteries (D-TGA) who had undergone the arterial switch operation in early infancy. Our cohort had been enrolled preoperatively in a single-center, randomized clinical trial, the Boston Circulatory Arrest Study, with treatment assignment to a support method consisting of either predominantly deep hypothermic circulatory arrest or predominantly low-flow cardiopulmonary bypass. Treatment groups were compared with respect to neurological and cognitive outcomes in the perioperative period and then at ages 1, 4, and 8 years. Before evaluation at age 8, parents completed the Child Health Questionnaire Parent Form-50.

The present report is the first to examine the general health status of children with D-TGA after the arterial switch operation. Specifically, we compare the psychosocial and physical health status of our cohort with those of a normative
sample, as well as with those of children with other benchmarked diseases. Finally, we explore the relationship of psychosocial and physical health to risk factors, including perioperative course, interim medical history, and scores on tests of ability and achievement at age 8 years.

Methods

Subjects
Subjects were enrolled between April 1988 and February 1992 in a single-center, randomized clinical trial of children with D-TGA who underwent the arterial switch operation; the trial compared the incidence of brain injury after assignment to either predominantly circulatory arrest or predominantly low-flow bypass. Eligibility criteria included (1) a diagnosis of D-TGA with intact ventricular septum or ventricular septal defect, (2) scheduled repair by 3 months of age, and (3) coronary artery anatomy thought to be suitable for the arterial switch operation. Exclusion criteria included (1) birth weight of <2.5 kg, (2) recognizable syndrome of congenital anomalies, (3) associated extracardiac anomalies of more than minor severity, (4) previous cardiac surgery, or (5) associated cardiovascular anomalies requiring aortic arch reconstruction or additional open surgical procedures. Patients’ outcomes were assessed in the perioperative period and at ages 1, 4, and 8 years. Earlier results and methods have been described previously.4–6 Informed consent was obtained from the parents of all subjects according to the guidelines of the institutional human investigation committee.

Child Health Questionnaire, Parent Form-50

The Child Health Questionnaire measures the overall health status in children aged 5 to 18 years. The questionnaire contains 50 questions and generates 2 summary scores: the Psychosocial and Physical Health Summary scores. Higher summary scores indicate better psychosocial and physical functioning. The summary scores are derived from 12 subscales, which capture 14 health concepts. The health concepts that contribute to the Psychosocial Summary score are mental health, role/social limitations (emotional), role/social limitations (behavioral), behavior, and self-esteem. The health concepts that contribute to the Physical Health Summary score are physical functioning, role/social limitations (physical), bodily pain/discomfort, and general health perceptions. The health concepts of parental impact/emotional and parental impact/time contribute to both summary scores. In addition, parents are asked to indicate on a checklist whether their child has had specified medical conditions or diagnoses.3

Parents completed the questionnaire before their child’s 8-year neurological and developmental assessment. The questionnaire was mailed to parents along with other questionnaires regarding their child’s health and behavior. Questionnaires were returned to study staff on the day of each child’s neurological and developmental in-person assessment. Members of the data management staff obtained information on incomplete questionnaires via telephone.

Developmental Testing

At age 8 years, 1 investigator (D.C.B.) administered a 5-hour comprehensive battery of standardized tests to assess academic and intellectual performance. From among the standardized tests administered, we chose for analysis only the primary developmental outcome variables. We assessed general intelligence using the Wechsler Intelligence Scale for Children, Third Edition,7 generating scores for full-scale IQ, verbal IQ, and performance IQ. Academic achievement was measured using the Wechsler Individual Achievement Test,8 from which we chose to analyze summary scores for reading (Composite Reading) and mathematics (Composite Math).

Statistical Methods

The primary outcomes in this study were the Physical Health Summary and Psychosocial Summary scores of the Child Health Questionnaire, Parent Form 50. Patient groups were compared with respect to variables measured on a continuous scale using the t test for independent samples or the nonparametric Wilcoxon rank sum test for skewed responses. Categorical variables were compared by Fisher’s exact test. Exact tests for trend were used to analyze ordered categorical variables (eg, quartile of hospital stay). We examined the association between variables using the Spearman correlation coefficient and multiple regression techniques.

Results

Of 171 children enrolled in infancy in the Boston Circulatory Arrest Study, 6 died, 5 lived outside the United States, and 160 were eligible for study participation at age 8 years. Of these, the families of 155 (96%) completed the Child Health Questionnaire; 154 also underwent neuropsychological testing. The family of 1 additional child who returned for this testing did not complete the Child Health Questionnaire. Five parents declined to complete the questionnaire. No patients were lost to follow-up.

Characteristics of Children With D-TGA

<table>
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<th>Characteristics of Children With D-TGA</th>
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<td>Composite Reading achievement‡</td>
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<td>Composite Math achievement‡</td>
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Values are mean±SD.  
*Score on Hollingshead Four-Factor Index of Social Class, with higher scores indicating high social status.  
†Wechsler Intelligence Scale for Children, Third Edition.  
‡Wechsler Individual Achievement Test.

Of 171 children enrolled in infancy in the Boston Circulatory Arrest Study, 6 died, 5 lived outside the United States, and 160 were eligible for study participation at age 8 years. Of these, the families of 155 (96%) completed the Child Health Questionnaire; 154 also underwent neuropsychological testing. The family of 1 additional child who returned for this testing did not complete the Child Health Questionnaire. Five parents declined to complete the questionnaire. No patients were lost to follow-up.

Characteristics of the 155 study subjects are presented in the Table. Of the 155 subjects, 118 (76%) had D-TGA with intact ventricular septum and 37 (24%) had D-TGA with ventricular septal defect. The cohort included 76% males and 90% whites. Approximately half of each had been assigned to predominantly circulatory arrest (78, or 50.3%) or predominantly low-flow bypass (77, or 49.7%) strategy of vital organ support during their arterial switch operation. Median ages at surgery, questionnaire completion, and neuropsychological testing were 6 days (range 1 to 67 days), 8.1 years (range 7.6 to 10.0 years), and 8.3 years (range 7.6 to 10.1 years), respectively. By the 8-year evaluation, 7 patients (4.5%) had
undergone cardiac reoperation and only 1 patient was on cardiac medications (digoxin and furosemide [Lasix]). Somatic growth was excellent, with height and weight percentiles (mean±SD) of 54±30 and 59±32, respectively.

Mean general health status scores (ie, Psychosocial Summary and Physical Health Summary scores) in children with D-TGA were similar to national norms (Figure 1). The Psychosocial Health Summary score (mean±SD) in the D-TGA patients was 49.7±9.9 compared with 51.2±9.1 in the normative sample. The Physical Health Summary score for the study cohort (mean±SD) was 54.0±6.1 compared with 53.0±8.8 in the normative sample.

We also compared the general health status of children with D-TGA with that of children who had other chronic illnesses benchmarked by the Child Health Questionnaire: asthma, JRA, and ADHD (Figures 2 and 3). Children with D-TGA had significantly worse Psychosocial Summary scores (49.7±9.9) than did patients with asthma (51.6±8.1, \( P=0.04 \)) and JRA (53.4±9.2, \( P=0.006 \)), but significantly better scores than those with ADHD (36.9±10.9, \( P<0.001 \)). Conversely, Physical Health Summary scores were better in children with D-TGA (54.0±6.1) than in those with asthma (46.6±7.8, \( P<0.001 \)) and JRA (42.1±13.9, \( P<0.001 \)) but worse than those in children with ADHD (57.6±6.2, \( P<0.001 \)).

The Child Health Questionnaire, Parent Form-50 asks parents whether their children have certain “medical conditions.” Compared with the normative sample, patients with D-TGA were reported by their parents to have significantly more frequent attention problems (34% versus 19.4%, \( P<0.001 \)), developmental delay/mental retardation (13% versus 2.8%, \( P<0.001 \)), learning problems (27% versus 11.7%, \( P<0.001 \)), and speech problems (32% versus 12.2%, \( P<0.001 \)). In contrast, the D-TGA cohort was similar to the normative population in their prevalence of each of the other queried conditions, including anxiety, asthma, behavior, chronic allergies, orthopedic joint and bone problems, chronic rheumatic disease, depression, diabetes, epilepsy, hearing impairment or deafness, vision problems, sleep disturbances, and respiratory, lung, or breathing problems.

Within the D-TGA cohort, we explored the relation of summary scores to sociodemographic variables, perioperative factors, subsequent medical events, and neuropsychological test scores at age 8 years. Psychosocial Summary scores were most highly associated with academic ability and achievement at age 8 years. Specifically, worse Psychosocial Summary scores were associated with lower full-scale, verbal, and performance IQ scores (\( r_s=0.26, P=0.001 \); \( r_s=0.21, P=0.009 \); and \( r_s=0.27, P=0.001 \), respectively), as well as with worse academic achievement as measured by scores on the Wechsler Individual Achievement Test Composite Reading (\( r_s=0.23, P=0.005 \)) and Composite Math (\( r_s=0.22, P=0.007 \)). Lower Psychosocial Summary scores were also significantly associated with an increased number of “other medical conditions” (\( r_s=-0.47, P<0.001 \)), which in this cohort largely represented measures of neuropsychological performance. The only variable significantly associated with worse Physical Health Summary scores was longer hospital stay after the arterial switch operation (\( r_s=-0.19, P=0.02 \)). However, there was a trend in the same direction for days in the intensive care unit (\( r_s=-0.14, P=0.07 \)). Neither summary score was significantly associated with diagnosis (D-TGA with intact ventricular septum versus with ventricular septal defect), treatment group assignment, age at surgery, duration of circulatory arrest, sex, or history of cardiac reoperation. The highest parental education achieved was significantly greater for families of children with D-TGA than for those in the normative sample (\( P<0.001 \)), but neither summary score was significantly related to the parent’s socioeconomic status within the D-TGA sample.

Discussion

D-TGA is one of the most common forms of congenital heart disease, constituting 5% to 7% of cardiac malformations.
Since the original description of the arterial switch operation in 1975 by Jatene et al., the arterial switch operation has become the procedure of choice for repair of D-TGA with intact ventricular septum or ventricular septal defect. Because of the rapid decline in the work capacity of the left ventricle after birth in children with D-TGA with an intact ventricular septum or a small ventricular septal defect, anatomic correction with the arterial switch operation must be performed during the first weeks of life. This practice provides a uniformity of age at repair that is rare among congenital heart defects. Furthermore, the arterial switch operation is performed with extremely low early mortality rates, negligible late mortality rates, and infrequent need for reoperation. We previously reported that neurocognitive performance in children with D-TGA has been below expected in several domains, including IQ, expressive language, visual-motor integration, motor planning and organization, and oromotor control. The present report is the first description of the general health status and quality of life in survivors.

We found that at age 8 years, children with D-TGA who underwent a single-stage arterial switch procedure in early infancy had general health status similar to that in the normal population. Children with D-TGA had significantly worse psychosocial health status than did patients with asthma and JRA but significantly better status than did those with ADHD. Conversely, physical health status in children with D-TGA was better than that in those with asthma and JRA but worse than that in children with ADHD. Compared with parents of the normative sample, parents of children with D-TGA reported a significantly greater frequency of problems with attention, learning, and speech, as well as a greater frequency of developmental delay. Within the D-TGA cohort, worse psychosocial health status was associated with lower general intelligence and academic achievement. Worse physical health status at age 8 years was significantly associated with longer duration of hospitalization after the arterial switch operation. Neither physical nor psychosocial health status was related to associated presence of a ventricular septal defect, age at surgery, intraoperative perfusion variables during the arterial switch operation, sex, or cardiac reoperation.

Measures of academic ability and achievement emerged as the most significant correlates of psychosocial functioning in our population at age 8 years. This observation is consonant with the greater frequency of neurocognitive problems in children with D-TGA and the association of learning disabilities with psychosocial dysfunction in the general population. Learning disabilities and ADHD affect ≈5% to 10% of school-age children, but their manifestations often persist throughout life. Effects during the school years include underachievement; poor social relations; lowered self-esteem; lowered expectations by the child, family, and school; and behavioral problems. Educators suspect that the etiology of illiteracy in adults is often unrecognized and untreated learning disabilities. School dropout and truancy are more common in children with learning problems, and there are reports of increased substance abuse and delinquency as well. The long-term effects of ADHD have been studied more extensively. Adolescents with a history of ADHD have a greater incidence of dropout from school, problems with the law, psychiatric disorders, automobile accidents, and lowered school achievement. These findings continue into adulthood and include lower-level jobs and more frequent job changes. Lower academic ability and achievement in children with D-TGA and their association with worse psychosocial functioning suggest that early neurodevelopmental assessment and intervention may be warranted in children who have undergone infant heart surgery.

The present study should be viewed in light of certain limitations. General health status questionnaires in the present study were completed by parents rather than by the patients themselves, raising the study limitation that health perceptions of the children may have differed from those of their parents. Indeed, the assessment of quality of life in pediatrics often necessitates the use of proxy responders, usually the mother of the patient. Parents can be expected to evaluate their children accurately or reliably when the items are concrete and objective. Parents who live with the patient and guide his or her medical care are the most appropriate responders for many health questions. Reported potential parental biases in assessment of their children’s health include idealized views or expectations of their child, congruence of the parent’s own subjective experiences with the perceived experiences of their child, and transference of the parent’s own sense of debilitated function onto the child. The age at which children are able to give valid and reliable responses varies with the type of information sought, as well as with the complexity of the questionnaire. The ideal general health status or quality-of-life instrument would include both a child report and a parent-proxy report.

The families in our sample had achieved higher educational status than those in the Child Health Questionnaire, Parent Form-30 normative sample. Because lower socioeconomic status has been associated with lower maternal rating of child health, it is possible that the similar general health status scores in our population and the normative sample underestimate the negative impact of infant heart surgery. In analyses within our sample, however, we did not find a significant relationship between summary scores and social class. Medical conditions, including attention problems, developmental delay, learning problems, and speech problems, were reported more frequently in children with D-TGA than in the normative sample. However, at age 8 years, these problems did not have sufficient influence on parents’ perceptions of their children’s general health status to cause their scores to be significantly lower than those of the normative sample. An additional study limitation is that we did not include sibling controls in our study. Children who have undergone open heart surgery have been reported to have worse cognitive function than their siblings. Interestingly, Ellerbeck et al. found that the siblings of children with D-TGA had more learning problems than expected, suggesting that a familial tendency for learning problems might be present. An additional study limitation is that the Child Health Questionnaire is a generic instrument applicable to a broad spectrum of children and adolescents and, like other pediatric measures of quality of life in children, does not
have a disease-specific module for children with congenital heart disease.

In summary, using the Child Health Questionnaire, Parent Form-50 to assess health-related quality of life, we found that physical and psychosocial health status in children with D-TGA repaired by the arterial switch operation in infancy is similar to that of a normative sample. Eight-year academic ability and achievement emerged as the strongest correlates of psychosocial function. From among the many variables explored, only hospital length of stay was significantly associated with physical health status at age 8 years. In the future, disease-specific quality-of-life instruments should be refined for use in children with congenital heart disease and should include measurement of the child's self-perception.

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