

# Quality of Life of Children After Repair of Transposition of the Great Arteries

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**Background**—We sought to assess quality of life of children with transposition of the great arteries (TGA) enrolled during transition in management strategy from atrial to arterial switch operation.

**Methods and Results**—Neonates enrolled by the Congenital Heart Surgeons Society in a prospective study of TGA between 1985 and 1989 were eligible. A Child Health Questionnaire was sent for completion by the child between February and June 2000. Data were compared with published normative values. Child Health Questionnaires were completed by 306 of 708 survivors at a mean age of  $13\pm 1$  years. Diagnosis included TGA (n=202, 66%), TGA/ventricular septal defect (VSD) (n=84, 27%), and TGA/VSD/pulmonary stenosis (n=20, 7%). Repair type was arterial switch (n=189, 62%), atrial switch (n=105, 34%; Senning=58, Mustard=47), or Rastelli (n=12, 4%). Children with TGA scored significantly higher than published norms in all categories except self-esteem. TGA/VSD/pulmonary stenosis was associated with lower scores than TGA and TGA/VSD in physical functioning ( $P=0.002$ ), general health perceptions ( $P=0.012$ ), and mental health ( $P=0.048$ ). Arterial repair was associated with higher scores than atrial or Rastelli repair in physical functioning ( $P<0.001$ ), pain ( $P=0.004$ ), mental health ( $P=0.019$ ), self-esteem ( $P=0.004$ ), and general health perceptions ( $P<0.001$ ). By multivariable analyses, the most common independent factors impacting scores were repair type, perfusion parameters, and gender.

**Conclusions**—Quality of life and health status as perceived by children 11 to 15 years after TGA repair is excellent when compared with published normative data and is better after arterial switch operation than after atrial repair. (*Circulation*. 2003;108:857-862.)

**Key Words:** heart defects, congenital ■ transposition of great vessels ■ pediatrics ■ quality of life

Traditionally, outcomes in children with transposition of the great arteries (TGA) have been measured in terms of morbidity and mortality. Because children with TGA now have excellent long-term survival after surgical repair, quality of life of these children is increasingly important. The World Health Organization defines health as “a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity.”<sup>1</sup> Children’s health is conceptualized to have physical and psychosocial (emotional, behavioral, and social) dimensions, and deficits in either may affect the ability to perform important social roles.<sup>2</sup>

Quality of life assessment instruments measure these dimensions of health and assess impact of disease on the child’s daily life. Global quality of life instruments have recently been developed for use in children.<sup>2-5</sup> One previous study used a parent-completed instrument to assess quality of life in children surviving repair of TGA.<sup>6</sup> Using an instrument designed for completion by the child, we sought to assess

quality of life of children undergoing repair of TGA during transition in management strategy from atrial repair to arterial switch operation (ASO).

## Methods

### Patients

Neonates with TGA (n=829) admitted to 1 of 24 Congenital Heart Surgeons Society (CHSS) institutions at age <15 days were prospectively enrolled in a multi-institutional study between January 1985 and March 1989.<sup>7-11</sup> During this period, a major transition in surgical management of TGA occurred from atrial inversion technique (Mustard or Senning operation) to ASO. Treatment was nonrandomized and selected by the enrolling institution based on surgeon and institutional knowledge, experience, and preference. There were 708 surviving children at the time of ongoing cross-sectional follow-up between February and June 2000 eligible for participation.

### Measurements

Contact of surviving children was attempted by the CHSS Data Center (n=478) or enrolling institution (n=230). Only children

Received March 5, 2003; revision received May 29, 2003; accepted May 29, 2003.

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*Circulation* is available at <http://www.circulationaha.org>

DOI: 10.1161/01.CIR.0000084547.93252.9A

**TABLE 1. Diagnosis and Repair Type**

Morphology	Repair Type				Total
	ASO	Senning	Mustard	Rastelli	
Simple TGA	123	44	35	0	202
TGA+VSD	61	11	10	2	84
TGA+VSD+PS	5	3	2	10	20
Total	189	58	47	12	306

whose enrolling institution had obtained institutional review board approval were contacted, thereby excluding 113 children from follow-up contact. A copy of Child Health Questionnaire (CHQ) Version CF-87 was sent to 595 children. If no response was received within 6 weeks of initial mailing, a reminder was sent, followed by 2 attempts to complete follow-up by telephone.

### Child Health Questionnaire

Version CF-87 was chosen for our study because children in our population were age 11 years or older at follow-up. It is an 87-item validated questionnaire that assesses self-perceived physical and psychosocial well-being of children aged 11 years and older.<sup>2</sup> Most pediatric quality of life instruments focus on parents' perception of children's disease.<sup>3-5</sup> In contrast, the CHQ focuses on an individual's subjective perception of his or her health. Version CF-87 is designed for completion by the child.

The CHQ measures 11 child health concepts, with each being scored on a scale from 0 to 100 (Appendix [Table 6]). Higher scores indicate better self-perceived function. Published scores obtained from 278 healthy children aged 10 to 15 years from a middle school in northeast United States were used as normative reference.<sup>12</sup> Although ethnic and gender distribution of the reference population is not similar to our study subjects, sample size and age distribution are similar.

### Data Analysis

Data are described as frequencies, medians with ranges, or means with standard deviations. Response bias was sought by comparing categorical data for responders versus nonresponders using  $\chi^2$  or Fisher's exact tests. General linear regression was used to assess possible associations between each CHQ concept and gender, TGA morphology, and type of surgical repair. Multiple linear regression was performed for scores on each of 11 CHQ scales using variables listed in the Appendix (Table 7). Variables meeting significance criterion of  $P < 0.15$  were entered into multivariable analysis, and those with  $P < 0.10$  were retained in final multivariable models.

**TABLE 2. Mean CHQ Scores for TGA Group and Published Norms**

CHQ Concept	TGA Group (n=306)	Published Norms	P
Physical functioning	93.2±11.3	88.8±14.0	<0.001
Bodily pain	83.7±19.7	74.4±23.1	<0.001
General health perceptions	74.6±16.4	66.4±14.6	<0.001
Self-esteem	82.6±14.8	81.8±15.8	0.33
Mental health	78.7±13.8	72.7±16.0	<0.001
Behavior	78.7±14.2	76.6±14.6	0.009
Social limits, physical	93.2±11.3	88.3±21.0	<0.001
Social limits, emotional	92.3±17.4	85.9±21.0	<0.001
Social limits, behavioral	94.2±15.3	86.5±21.5	<0.001
Family activities*	85.4±18.2	...	...
Family cohesion*	74.5±22.8	...	...

\*Control values not available.<sup>2</sup>

Analyses were performed using SAS statistical software Version 8 (SAS Institute).

### Results

Of 595 children sent a CHQ, 317 (53%) returned a CHQ to the Data Center. Of these, 306 CHQs were analyzed. Eleven CHQs were excluded from analysis for the following reasons: parent completion (n=1), insufficiently completed CHQ (n=3), or unable to complete CHQ because of severe learning or intellectual disabilities (n=7). The latter had either ASO (simple TGA, n=2; TGA/VSD, n=1) or Mustard (simple TGA, n=3; TGA/VSD/pulmonary stenosis [PS], n=1) repair. The following potential explanatory factors for neurological deficits were identified in 5 subjects: prolonged circulatory arrest of 86 minutes (n=1), persistent severe cyanosis before Mustard (n=1), severe ventricular arrhythmia after ASO (n=1), and persistent low cardiac output syndrome (Mustard, n=1; ASO, n=1).

Mean age at follow-up was 13±1 years. Morphology and type of repair for responders are given in Table 1. There were no significant differences between CHQ responders and

**TABLE 3. Mean CHQ Scores by Morphology**

CHQ Concept	Morphology			P
	Simple TGA (n=202)	TGA/VSD (n=84)	TGA/VSD/PS (n=2)	
Physical functioning	93.9±8.6	93.9±8.2	83.8±28.1	0.002
Bodily pain	82.4±20.2	88.0±17.6	79.5±20.9	0.052
General health perception	75.2±16.7	75.7±15.3	64.5±14.6	0.012
Self-esteem	81.9±15.6	85.1±13.1	79.4±12.2	0.15
Mental health	78.4±14.4	80.9±12.1	72.8±13.5	0.048
Behavior	78.4±14.6	79.7±13.8	77.4±12.2	0.71
Social limits, physical	96.0±11.4	94.4±14.5	96.6±10.1	0.56
Social limits, emotional	91.1±19.4	95.4±12.2	91.5±12.6	0.15
Social limits, behavioral	94.4±15.1	95.8±10.2	94.0±15.8	0.87
Family activities	85.3±18.7	85.8±17.5	83.7±17.7	0.90
Family cohesion	73.9±22.4	76.5±23.2	73.8±24.7	0.65

**TABLE 4. Mean CHQ Scores by Repair Type**

CHQ Concept	Repair Type			P
	ASO (n=189)	Atrial (n=105)	Rastelli (n=12)	
Physical functioning	95.7±7.6	91.2±8.9	90.1±9.1	<0.001
Bodily pain	86.5±18.3	79.2±21.4	76.7±18.3	0.004
General health perceptions	78.6±14.9	69.9±16.8	62.1±14.4	<0.001
Self-esteem	85.3±13.1	78.8±16.6	76.8±12.1	0.004
Mental health	80.6±12.8	76.0±15.3	76.8±8.5	0.019
Behavior	80.1±14.0	76.4±15.1	79.3±9.9	0.10
Social limits, physical	95.9±13.0	95.3±10.9	96.3±9.9	0.90
Social limits, emotional	93.2±17.2	90.4±18.5	96.3±7.2	0.29
Social limits, behavioral	94.5±15.2	93.3±16.3	99.1±3.2	0.45
Family activities	86.9±18.3	82.5±18.1	84.4±18.9	0.15
Family cohesion	75.3±22.9	73.5±21.9	79.6±24.1	0.62

nonresponders in gender (70% versus 71% male; *P*=0.88), morphology (67% versus 64% simple, 27% versus 30% TGA/VSD, 6% versus 6% TGA/VSD/PS; *P*=0.27), and repair type (62% versus 63% ASO, 15% versus 12% Mustard, 19% versus 22% Senning, 4% versus 3% Rastelli; *P*=0.73).

CHQ scores of children with TGA were significantly higher than those from the published normative population (indicating better self-perceived health status) in all categories except self-esteem (Table 2). By univariable comparison, girls had higher mean scores in behavioral social limits (97 versus 87, *P*=0.003) and boys had higher general health perception scores (68 versus 60, *P*=0.02). Children with simple TGA or TGA/VSD had significantly higher CHQ scores than those with TGA/VSD/PS in the categories of physical functioning, general health perceptions, and mental health (Table 3). Children having ASO scored significantly higher in the categories of physical functioning, bodily pain, mental health, self-esteem, and general health perceptions than those having atrial repair or Rastelli procedure (Table 4). Children who had ASO scored higher than either atrial repair subgroup (Mustard or Senning). There were no significant differences between children who had Mustard or Senning.

Multivariable regression analysis was performed for scores on each CHQ concept (Table 5). Although statistically significant, the percent variation in CHQ concept scores explained by the factors is small, with *R*<sup>2</sup> values ranging from 0.024 to 0.26. Repair type was associated with scores on all but 2 CHQ concepts (emotional and behavior social limits). For each, atrial repair was associated with lower scores than ASO. Perfusion parameters such as circulatory arrest, ischemic time, duration of cardiopulmonary bypass, and cooling temperature were associated with lower scores on social limits and behavior concepts. Male gender was associated with lower scores in emotional and behavioral concepts, and female gender was associated with lower general health perceptions.

### Discussion

Compared with the normal population, children after TGA repair have been reported to have more frequent neurologic

**TABLE 5. Summary of Multiple Regression Analysis for Independent Factors Associated With Lower Scores on Individual CHQ Concepts**

Variable	Parameter Estimate (SEM)	P
Physical functioning		
Repair other than ASO	4.6 (1.0)	<0.001
Associated noncardiac anomalies	7.1 (3.6)	0.051
Bodily pain		
Repair other than ASO	6.9 (2.3)	0.003
Diagnosis other than TGA/VSD	5.1 (2.5)	0.043
General health		
Repair other than ASO	9.2 (1.8)	<0.001
Female	3.4 (1.9)	0.081
Self-esteem		
Repair other than ASO	9.1 (2.1)	<0.001
Repair other than Senning	4.6 (2.6)	0.081
Mental health		
Repair other than ASO	4.5 (1.6)	0.005
Behavior		
Mustard operation	6.1 (2.2)	0.006
Lower perfusion temperature, per 1°C	0.7 (0.3)	0.053
Social limits, physical		
Use of circulatory arrest	3.0 (1.7)	0.080
Longer cardiopulmonary bypass time, per 20 minutes	0.8 (0.4)	0.080
Associated noncardiac anomalies	8.9 (5.5)	0.10
Mustard operation	3.2 (1.9)	0.10
Social limits, emotional		
Male	4.4 (2.1)	0.041
Use of circulatory arrest	4.9 (2.4)	0.046
Longer ischemic time, per 10 minutes	1.1 (0.6)	0.068
Social limits, behavioral		
Male	4.2 (2.2)	0.053
Use of circulatory arrest	4.7 (2.4)	0.057
Family activities		
Mustard operation	7.3 (2.9)	0.012
Family cohesion		
Mustard operation	6.9 (3.5)	0.050
Global general health		
Repair other than ASO	5.2 (2.3)	0.025
Mustard operation	5.5 (3.1)	0.076
Global behavior		
Lower perfusion temperature, per 1° C	0.9 (0.4)	0.059
Male	4.3 (2.4)	0.073
Postoperative seizure	11.9 (6.7)	0.078
Mustard operation	5.3 (3.1)	0.084

impairment, learning disabilities, behavior disorders, and poorer motor, vocabulary, and acquired abilities.<sup>6,13</sup> Our study suggests that children and adolescents surviving repair of TGA seem to be functioning well, both physically and

psychosocially. Our study population scored significantly higher than published norms in all categories except self-esteem. Although statistically significant, the magnitude of differences between TGA and control children is small. At present, this difference is of unknown clinical relevance.

Our study is unique because data were obtained directly from children. Most studies of this type survey parents, and few investigators have relied on direct responses from children with congenital heart defects.<sup>14</sup> Guyatt et al<sup>15</sup> noted that parents of children aged 11 years or older provided little information regarding their child's quality of life beyond that obtained directly from the child.

Clearly, self-perception of disease is multifactorial. The finding of low  $R^2$  values in our study indicates that a large portion of variability in CHQ scores is explained by variables not included in our data set. Such variables may include home and school environment, economic status, and genetic or geographic factors. In a study of cognitive development after Fontan operation by Wernovsky et al,<sup>16</sup> surgical and diagnostic factors explained 6% of variation in achievement and IQ scores. The addition of socioeconomic status to their analysis increased explained variation nearly 4-fold. The inference from our study is that diagnostic and surgical factors are small but important determinants in children's psychosocial development.

Previous reports show that 96% of children are without symptoms or limitation of physical activity up to 5 years after ASO.<sup>17,18</sup> Masterson et al<sup>6</sup> used a parent-completed version of the CHQ to evaluate general health status in 160 children at 8 years of age. They found that the overall physical and psychosocial health status after ASO was similar to a normative population. Furthermore, children who underwent ASO have preserved systemic ventricular function with sustained sinus rhythm and near-total avoidance of cardiac medication.<sup>18–20</sup> Functional outcomes published to date are reassuring to the findings of our study. There is concordance between children's self-perception of physical and psychosocial health and that objectively documented in clinical studies. Although our study extends follow-up of physical and psychosocial outcomes of ASO patients to 13 years, later evaluation is necessary to monitor for subsequent deterioration in subjective and objective measures of health status.

In contrast to ASO patients, there is often discordance between subjective history and objective findings in patients having atrial repair of TGA. Paul and Wessel<sup>21</sup> found that most patients with atrial repair report a self-perception of normal physical function and ability. However, when assessed systematically, others have found that 40% to 77% of patients report some degree of physical restriction.<sup>22,23</sup> Objective exercise testing after atrial repair reveals diminished aerobic capacity with limited cardiac output because of reduced stroke volume and attenuated heart rate response.<sup>21,24</sup>

In our study, multivariable regression analyses reveal that repair type was significant for all but 2 CHQ concepts, with atrial switch being associated with lower scores than ASO. Although no significant differences were found between scores for children with Mustard and Senning by univariable comparison, Mustard repair was independently associated with lower scores on several CHQ concepts. This finding

cannot be readily explained. Previous data suggest a survival advantage for patients with Mustard and a rhythm advantage for patients with Senning, but no important differences in functional class have been found to date.<sup>22,25</sup>

Long-term psychological outcomes after atrial repair remain unclear. Alden et al<sup>26</sup> reported that 26% of children had behavioral problems, although psychosocial performance and self-esteem are often normal. Their patients were repaired 20 years earlier than our patients during a time when definitive correction was delayed for months. Longer duration of cyanosis in these children may be a factor explaining differences in behavior and cognitive outcomes.<sup>27</sup>

Other factors may explain higher scores in ASO children. Because they were operated on at younger age than those after atrial or Rastelli repair, they had a shorter cumulative duration of cyanosis. They may also have fewer long-term limitations because of ventricular failure and arrhythmia. Ellerbeck et al<sup>28</sup> compared the cognitive and motor development of children after atrial and ASO repair. They noted no difference in neurologic impairment, learning disabilities, behavior disorders, and motor, vocabulary, and acquired abilities. Our finding that the ASO group had better CHQ scores may be attributable to a difference in the age of our patients, their duration of follow-up, or our method of data collection that excluded a few patients with important learning disabilities.

Perfusion techniques are often implicated in neuropsychological outcomes. Others have shown that use of circulatory arrest during TGA repair is associated with fine and gross motor deficits but is generally well tolerated, with no cognitive or other neurologic deficits identified by formal testing.<sup>13,27</sup> By multivariable regression, our study revealed that perfusion parameters may be important in behavior and social limits concepts. Use of circulatory arrest, but not duration, was associated with lower CHQ scores, suggesting that any period of cerebral ischemia may be detrimental. That lower nadir core temperature may be associated with lower scores in behavior concepts is perhaps unexpected. We speculate that attaining lower nadir temperature could be associated with longer duration of cardiopulmonary bypass, higher flow rates, and more extreme rates of cooling and rewarming in the 1985 to 1989 era. Variations in such parameters were previously linked to lower scores on formal neurologic testing.<sup>29</sup> Inferences regarding perfusion parameters would require additional information, such as pH and blood-gas management.

### Study Limitations

The response rate was suboptimal, possibly contributing to undetected response bias. Although similar to responders in age, diagnosis, and repair type, nonresponders may differ in other important ways that potentially impact health-related quality of life. Albeit few, children with severe disabilities are not reflected in CHQ scores because they could not complete the questionnaire. Lastly, there may be important differences in social, demographic, and economic characteristics of our study children and published norms.

## Summary

Self-perceived quality of life is excellent in most children who underwent repair of TGA 10 to 15 years earlier. Diagnostic and surgical factors are small but important determinants of chil-

dren's perception of health-related quality of life. ASO is associated with improved physical and psychosocial functioning compared with other methods of repair. These differences may be additionally exacerbated with longer follow-up.

## Appendix

**TABLE 6. Description and Interpretation of Child Health Concepts Measured in Child Health Questionnaire Version CF-87**

Concepts	Description	Interpretation
<b>Physical health</b>		
Physical functioning scale	Presence and extent of physical limitations in self-care, mobility, and strenuous activities attributable to health-related problems.	Perfect score: able to perform all types of physical activities. Low score: very limited in performing physical activities, including self-care, because of health.
Bodily pain scale	Intensity and frequency of general pain.	Perfect score: no pain. Low score: extremely severe, frequent, and limiting pain.
General health perceptions scale	Evaluation of current and future physical health status.	Perfect score: perceives current and future health to be excellent. Low score: perceives health to be poor and likely to get worse.
<b>Emotional health</b>		
Self-esteem scale	Satisfaction with school and athletic ability, appearance, ability to get along with others and family, and life overall along a continuum.	Perfect score: very satisfied with all of the above. Low score: very dissatisfied with all of the above.
Mental health scale	Frequency of positive and negative states (anxiety and depression).	Perfect score: peaceful, calm, and happy all of the time. Low score: anxious and depressed all of the time.
<b>Behavioral health</b>		
Behavior scale	Overt behavior, such as frequency of behavior problems and ability to get along with others.	Perfect score: never exhibits aggressive, immature, or delinquent behavior. Low score: often exhibits aggressive, immature, or delinquent behavior.
<b>Social health</b>		
Role/social limitations scales	Limitations in kind, amount, and performance of schoolwork and activities with friends because of physical, emotional, or behavioral problems.	Perfect score: no limitations. Low score: severely limited in schoolwork or activities with friends as a result of physical, emotional, or behavioral problems.
Family activities and cohesion scales	Limitation experienced by family because of child's health and well-being as well as cohesiveness of the family unit.	Perfect score: family relationships are "excellent" and child's health never limits family activities nor is source of family tension. Low Score: family relationships are poor and child's health often interrupts family activities or is source of family tension.

**TABLE 7. Variables Considered in Multivariable Regression Analysis of CHQ Concept Scores**

Variable	Value (%)	Missing
Patient and morphological		
Male	216 (70)	0
Mean birth weight, kg	3.4±0.5	76
Major noncardiac anomalies	5 (2)	0
Morphology		
Simple TGA	202 (66)	0
TGA/VSD	84 (27)	0
TGA/VSD/PS	20 (7)	0
Major associated cardiac anomalies	14 (5)	0
Multiple VSD	14 (5)	0
Coarctation	8 (3)	0
History of right ventricular outflow tract obstruction	9 (3)	0
No. of palliative operations		
0	91 (30)	0
1	180 (58)	0
2	35 (12)	0
Surgical		
Mean cardiopulmonary bypass time, min	130±43	163
Mean coldest temperature, °C	18±3	92
Mean ischemic time, min	70±22	143
Total circulatory arrest	210 (76)	33
Mean circulatory arrest time, min	47±22	87
Repair		
ASO	189 (62)	0
Atrial	105 (34)	0
Mustard	47 (15)	0
Senning	58 (19)	0
Rastelli	12 (4)	0
Left ventricular outflow tract procedure	12 (4)	0
Right ventricular outflow tract procedure	12 (4)	0
Pacemaker	13 (4)	0
Postoperative seizures	9 (8)	199

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