

Prevalence of and Predictive Factors for Scoliosis After Surgery for Congenital Heart Disease in the First Year of Life

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Background: The surgical treatment of congenital heart disease is reported to be associated with a high prevalence of scoliosis, although the detailed etiology is unknown. Surgical interventions involving the rib cage are considered to increase the risk of scoliosis. However, whether the cardiac condition or the procedure performed makes patients more susceptible to the development of spinal deformity is controversial.

Methods: The present study included 483 patients who underwent surgery for the treatment of congenital heart disease with use of procedures involving the immature rib cage (sternotomy and/or thoracotomy) during the first year of life, followed by the evaluation of standing chest radiographs at ≥ 10 years of age. Patients with congenital spinal deformity and potential neuromuscular disease were excluded. The prevalence of and predictive factors for scoliosis were evaluated. The presence of scoliosis (Cobb angle $\geq 10^\circ$ to $< 20^\circ$, $\geq 20^\circ$ to $< 30^\circ$, $\geq 30^\circ$ to $< 45^\circ$, $\geq 45^\circ$), the convex side of the curve, and the location of the curve were evaluated radiographically. Potential predictive factors that were analyzed included the age at the time of surgery, surgical approach, use of cardiopulmonary bypass, postoperative heart failure and/or cyanosis, New York Heart Association (NYHA) class, cardiomegaly, and age at the time of radiography.

Results: The mean age at the time of surgery was 112 days, and the mean age at the time of radiography was 14.4 years. The prevalence of scoliosis was 42.4%, and the prevalences of $\geq 10^\circ$ to $< 20^\circ$, $\geq 20^\circ$ to $< 30^\circ$, $\geq 30^\circ$ to $< 45^\circ$, and $\geq 45^\circ$ scoliosis were 31.7%, 5.8%, 2.5%, and 2.5%, respectively. Three patients underwent surgery for the treatment of progressive scoliosis. Multivariate analysis indicated that the predictive factors were female sex, left thoracotomy, bilateral thoracotomy, NYHA class, and age at the time of radiography for $\geq 10^\circ$ scoliosis; cardiomegaly, NYHA class, and age at the time of radiography for $\geq 20^\circ$ scoliosis; cardiomegaly, number of surgical procedures, and age at the time of radiography for $\geq 30^\circ$ scoliosis; and cardiomegaly for $\geq 45^\circ$ scoliosis. Age at the time of radiography was a predictor of $< 45^\circ$ scoliosis; however, the relative association was small.

Conclusions: Surgery for the treatment of congenital heart disease during the first year of life was associated with a high prevalence of scoliosis ($\geq 40\%$). While female sex was one of several predictors of $\geq 10^\circ$ scoliosis, cardiomegaly was the sole predictor of $\geq 45^\circ$ scoliosis.

Level of Evidence: Prognostic Level IV. See Instructions for Authors for a complete description of levels of evidence.

Congenital heart disease (CHD) is the most common type of birth defect, accounting for one-third of all major congenital anomalies. Worldwide, 1.35 million infants are born with CHD each year, with a worldwide occurrence of 7 per 1,000 live births¹. Before the advent of cardiac surgery, CHD, together with infectious disease, was the most prominent cause of death during the neonatal and

infantile periods. The advent of corrective cardiac surgery in the late 1950s and the increase in knowledge concerning the longitudinal care of patients with CHD have led to a spectacular increase in life expectancy. Today, $>90\%$ of children with CHD who survive the first year of life will live into adulthood². However, this long life expectancy has brought new challenges in clinical management as many patients

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develop late complications, including not only heart-related complications but also skeletal complications³. One of the skeletal complications is scoliosis. A high prevalence of scoliosis after cardiac surgery for the treatment of CHD has been reported, and surgical interventions involving the immature rib cage (sternotomy and/or thoracotomy) are considered to increase the risk of scoliosis⁴⁻⁹. However, controversy remains with regard to whether the cardiac condition or the procedure performed makes the patient more susceptible to the development of spinal deformity.

The purpose of the present study was to elucidate the prevalence of and predictive factors for scoliosis among patients with CHD who underwent cardiac surgery with use of procedures involving the immature rib cage (sternotomy and/or thoracotomy) in the first year of life. We hypothesized that the prevalence of scoliosis is high in such patients and that both cardiac and operative factors can affect the severity of scoliosis.

Materials and Methods

Patients

Institutional review board approval was obtained prior to the initiation of this study, and the requirement for written informed consent of patients and their parents was waived because of the retrospective nature of the study. The 10-year period from 1995 to 2004 was chosen in order to obtain a sizable group of patients and to ensure a minimum duration of follow-up of 10 years from the time of cardiac surgery. Among a total of 975 patients who underwent cardiac surgery for the treatment of CHD in the first year of life, 477 were excluded because of a lack of standing chest radiographs at ≥ 10 years of age (reported death, $n = 80$; lost to follow-up including unreported death, $n = 397$). Among the remaining 498 patients, 5 who demonstrated congenital spinal deformity and 10 who were also affected by neuromuscular diseases also were excluded. The upper age limit at the time of surgery (< 1 year) was selected to achieve a uniform skeletal age, which is reported to affect the development of scoliosis. Therefore, 483 patients (257 males and 226 females) were included in the final analysis. The mean age at the time of surgery was 112 days (range, 0 to 364 days), and the mean age at the time of the last standing chest radiograph was 14.4 years (range, 10 to 21 years) (Table I).

TABLE I Patient Demographics

No. of patients	483
Female:male ratio (<i>no. of patients</i>)	226:257
Interval between birth and initial surgery* (<i>d</i>)	112 \pm 107 (0-364)
Age at radiography* (<i>yr</i>)	14.4 \pm 2.9 (10-21)
*The data are given as the mean and the standard deviation, with the range in parentheses.	

TABLE II Analyzed Factors*

Cardiac and other factors	
Sex (female or male)	
No. of days between birth and surgery	
Age at radiography (in years)	
Use of cardiopulmonary bypass	
Sternotomy	
Right thoracotomy	
Left thoracotomy	
Both right and left thoracotomy	
Number of surgical procedures	
Cardiomegaly (CTR $\geq 55\%$)	
Postop. cyanosis	
Postop. heart failure	
NYHA class (1, 2, 3, or 4)†	
Radiographic parameters	
Scoliosis ($\geq 10^\circ$)	
Curve grade: mild ($\geq 10^\circ$ to $< 20^\circ$), moderate ($\geq 20^\circ$ to $< 30^\circ$), pronounced ($\geq 30^\circ$ to $< 45^\circ$), surgical candidate ($\geq 45^\circ$)	
Convex side (right or left)	
Curve type: HT (high thoracic curve only), LT (low thoracic curve only), DT (double thoracic curve), T and L (thoracic and lumbar curves)	
*CTR = cardiothoracic ratio, and NYHA = New York Heart Association. †N = 475 (data not available for 8 patients).	

Radiographic Evaluation

All posteroanterior radiographs included the L2 level or lower. If no tilt of L2 was observed, the lumbar spine was considered to be straight (no lumbar scoliosis). Spinal curvature was measured according to the Cobb method¹⁰ on a flat monitor with use of built-in imaging software (Centricity CDS; M & H). Scoliosis was defined as curvature of $> 10^\circ$ and was graded as mild ($\geq 10^\circ$ to $< 20^\circ$), moderate ($\geq 20^\circ$ to $< 30^\circ$), pronounced ($\geq 30^\circ$ to $< 45^\circ$), or a surgical candidate ($\geq 45^\circ$) (Table II). Curve locations were classified as high thoracic, low thoracic, or lumbar. For patients with > 1 curve, the existence of a vertebral apex that was prominent to the contralateral side of the proximal or distal curve with a Cobb angle of $\geq 10^\circ$ was classified as a double thoracic curve or a thoracic and lumbar curve, depending on the location of the curvature. The curvature with the largest Cobb angle was used in the subsequent analysis.

The convexity of each curve to the left or right also was recorded. One of the authors, who is a board-certified spine surgeon, measured all of the radiographic parameters.

Data Collection

Clinical data were reviewed for surgical history and cardiac condition. Preoperative, perioperative, and postoperative variables are listed in Table II. Diagnoses of CHD were classified into 20 groups based on primary diagnosis (Table III). Cyanosis

TABLE III Primary Diagnosis of Enrolled and Excluded Patients

Primary Diagnosis	Enrolled (no. of patients)	Excluded (no. of patients)
Tetralogy of Fallot	95	31
Ventricular septal defect	77	162
Coarctation/interruption of the aorta	47	39
Single ventricle, left ventricle	38	14
Dextro-transposition of the great arteries	43	29
Atrioventricular septal defect	30	23
Single ventricle, right ventricle	27	45
Single ventricle, heterotaxy	24	20
Total anomalous pulmonary venous connection	22	17
Double outlet right ventricle	16	17
Congenital atrioventricular block	9	6
Patent ductus arteriosus	9	39
Atrial septal defect	7	9
Corrected congenital transposition of the great arteries	7	3
Pulmonary atresia, intact ventricular septum	7	9
Congenital mitral stenosis/regurgitation	6	3
Single ventricle, other	5	1
Truncus arteriosus communis	4	5
Congenital aortic stenosis/regurgitation	3	6
Others	7	14
Total	483	492

was defined if patients were diagnosed as having a single functional ventricle or relatively balanced ventricles but surgically untreatable intracardiac defect(s) with right-to-left shunting. Heart failure was defined if the patient was currently being managed with medications such as digoxin, diuretics, angiotensin-converting enzyme inhibitors, or beta blockers. On the basis of posteroanterior chest radiographs, which were used for evaluation of scoliosis, the cardiothoracic ratio (CTR) was measured, and cardiomegaly was defined if the CTR was $>0.55^{11}$.

Statistical Analysis

Statistical analyses were performed with use of PASW Statistics (version 24; SPSS). For univariate analysis, the Mann-Whitney U test was used to compare age at initial surgery, age at radiography, and number of procedures, whereas the chi-square test was used to compare sex, use of cardiopulmonary bypass, sternotomy, thoracotomy, postoperative cyanosis/heart failure, and New York Heart Association (NYHA) class. To assess

predictive factors for each degree of scoliosis, multivariate logistic regression analysis was performed with use of variables with a p value of <0.20 in univariate analysis. P values of ≤ 0.05 and ≤ 0.0083 were considered significant for 2-group and 4-group comparisons, respectively.

Results

Prevalence and Characteristics of Scoliosis

The mean age at the time of the initial cardiac procedure was 112 days (range, 0 to 364 days), and the mean age at the time of radiography was 14.4 years (range, 10 to 21 years). The prevalence of scoliosis was 42.4% (205 of 483), and those of $\geq 10^\circ$ to $<20^\circ$, $\geq 20^\circ$ to $<30^\circ$, $\geq 30^\circ$ to $<45^\circ$, and $\geq 45^\circ$ scoliosis were 31.7% (153 of 483), 5.8% (28 of 483), 2.5% (12 of 483), and 2.5% (12 of 483), respectively. Curve locations were high thoracic in 33.2% of the patients, low thoracic in 30.2%, double thoracic in 31.2%, and thoracic and lumbar in 5.4%. Convexity was to the left in $>70\%$ of high thoracic curves, to the right in $>70\%$ of low thoracic curves, and to the left in $>70\%$ of lumbar curves (Table IV). Data regarding nonoperative treatment with serial casting or a brace could not be collected. Only 3 patients underwent surgery for the treatment of progressive scoliosis during the follow-up period. The degrees of curvature at the time of scoliosis surgery were 83° (T4 to T11) at the age of 13 years for 1 of those patients and 48° (T1 to T6) and 56° (T6 to T12) at the age of 17 years for another; the preoperative radiograph for the third patient was unavailable, but she underwent scoliosis surgery at the age of 12 years and demonstrated 37° (T5 to T12) of curvature even after the surgical correction. However, 12 patients (2.5%) demonstrated curvature of $>45^\circ$, which is the threshold for surgical

TABLE IV Radiographic Evaluation of Scoliosis*

Cobb angle	
Overall ($\geq 10^\circ$)	42.4% (205/483)
Mild ($\geq 10^\circ$ to $<20^\circ$)	31.7% (153/483)
Moderate ($\geq 20^\circ$ to $<30^\circ$)	5.8% (28/483)
Pronounced ($\geq 30^\circ$ to $<45^\circ$)	2.5% (12/483)
Surgical candidate ($\geq 45^\circ$)	2.5% (12/483)
Curve type	
HT	33.2% (68/205)
LT	30.2% (62/205)
DT	31.2% (64/205)
T and L	5.4% (11/205)
Convex side	
HT (left)	79.8%
LT (right)	70.6%
L (left)	75.0%

*HT = high thoracic curve, LT = low thoracic curve, DT = double thoracic curve, T and L = thoracic and lumbar curve, and L = lumbar curve.

TABLE V Comparison of Curve Severity Distribution Between Patients ≥ 18 Years Old (Skeletally Mature) and Patients < 18 Years Old

	≥ 18 Years Old (N = 67)	< 18 Years Old (N = 416)	P Value
Mean age and std. dev. (yr)	19.3 \pm 0.9	13.6 \pm 2.2	< 0.001
Cobb angle (no. of patients)			
Overall ($\geq 10^\circ$)	35 (51%)	170 (41%)	0.080
Mild ($\geq 10^\circ$ to $< 20^\circ$)	25 (37%)	128 (31%)	0.285
Moderate ($\geq 20^\circ$ to $< 30^\circ$)	6 (9%)	22 (5%)	0.233
Pronounced ($\geq 30^\circ$ to $< 45^\circ$)	2 (3%)	10 (2%)	0.777
Surgical candidate ($\geq 45^\circ$)	2 (3%)	10 (2%)	0.777

treatment, and 2 of them had progression to $> 90^\circ$. However, because of their cardiac conditions (with 1 patient showing failure of the Fontan circulation and the other patient having postoperative heart failure with severe pulmonary artery stenosis), neither of those patients underwent scoliosis surgery. Both patients died from heart failure at the age of 13 years.

Comparison of Curve Severity Distribution Between Patients ≥ 18 Years of Age (Skeletally Mature) and Patients < 18 Years of Age

To clarify the effect of skeletal maturity on the distribution of curve severities, the prevalence was compared between patients ≥ 18 years of age (i.e., skeletally mature patients) (mean age, 19.3 years) and patients < 18 years of age (mean age; 13.6 years). The distribution of curve severities was not significantly different between the groups (Table V).

Effects of Thoracotomy on the Development and Convex Side of the High Thoracic Curve

The prevalence of a high thoracic curve type was significantly higher for patients who underwent right or left thoracotomy compared with patients who did not undergo thoracotomy.

However, the sides of the apex were not different between the groups (Table VI).

Univariate Analysis of Predictive Factors for Scoliosis

In patients with $\geq 10^\circ$ scoliosis, the female:male ratio, rate of left thoracotomy, average number of surgical procedures, rate of postoperative heart failure, average NYHA class, and average age at the time of radiography were significantly higher than those in patients without scoliosis (see Appendix). In patients with $\geq 20^\circ$ scoliosis, the average number of surgical procedures, rate of cardiomegaly, rate of postoperative heart failure, average NYHA class, and average age at radiography were significantly higher than those in patients with less-than-moderate scoliosis (see Appendix). In patients with $\geq 30^\circ$ scoliosis, the average number of surgical procedures, rate of bilateral thoracotomy, rate of cardiomegaly, average NYHA class, and average age at the time of radiography were significantly higher than those in patients with less-than-pronounced scoliosis (see Appendix). In patients with $\geq 45^\circ$ scoliosis, the female:male ratio, rate of bilateral thoracotomy, average number of surgical procedures, and rate of cardiomegaly were significantly higher than those in patients with curves of $< 45^\circ$ (Table VII).

TABLE VI Effects of Thoracotomy on the Prevalence and Convex Side of a High Thoracic Curve*

	Left Thoracotomy (N = 79)	Right Thoracotomy (N = 55)	Bilateral Thoracotomy (N = 18)	No Thoracotomy (N = 331)
≥ 2 thoracotomies on same side†	19 (24%)	7 (13%)	0 (0%)	NA
HT curve†	32 (40.5%)	21 (38%)	8 (44%)	71 (21%)
P value (vs. no thoracotomy)	< 0.001 ‡	0.007‡	0.023	NA
Convex side of HT curve				
Left†	27 (84%) of 32	13 (62%) of 21	5 (63%) of 8	59 (83%) of 71
Right†	5 (16%) of 32	8 (38%) of 21	3 (38%) of 8	12 (17%) of 71
P value (for left, right, and bilateral thoracotomy vs. no thoracotomy)	0.872	0.039	0.159	NA

*HT = high thoracic, and NA = not applicable. †The values are given as the number of patients, with the percentage in parentheses. ‡P < 0.0083 according to the Bonferonni method for 4-group comparison.

TABLE VII Univariate Analysis for $\geq 45^\circ$ (Surgical-Candidate) Scoliosis*

	With $\geq 45^\circ$ Scoliosis (N = 12)	Without $\geq 45^\circ$ Scoliosis (N = 471)	P Value
Female:male ratio (no. of patients)	9:3	217:254	0.047†
Interval between birth and surgery‡ (d)	58.6 ± 55.46	113.5 ± 107.4	0.350§
Cardiopulmonary bypass (no. of patients)	11 (92%)	448 (95%)	0.587†
Sternotomy (no. of patients)	12 (100%)	458 (97%)	0.560†
Left thoracotomy (no. of patients)	2 (17%)	77 (16%)	0.977†
Right thoracotomy (no. of patients)	1 (8%)	54 (11%)	0.736†
Bilateral thoracotomy (no. of patients)	2 (17%)	16 (3%)	0.017†
Number of surgical procedures‡	2.7 ± 1.4	1.9 ± 1.2	0.025§
Cardiomegaly (CTR $\geq 55\%$) (no. of patients)	7 (58%)	64 (14%)	<0.001†
Postop. cyanosis (no. of patients)	1 (8%)	57 (12%)	0.692†
Postop. heart failure (no. of patients)	4 (33%)	135 (29%)	0.724†
NYHA class (1/2/3/4) (n = 475) (no. of patients)	0/6/4/0	1/394/65/5	0.145†
Age at radiography‡ (yr)	15.2 ± 3.1	14.4 ± 2.9	0.312§

*CTR = cardiothoracic ratio, and NYHA = New York Heart Association. †Chi-square test. ‡The values are given as the mean and the standard deviation. §Mann-Whitney U test.

Multivariate Logistic Regression Analysis

Multivariate logistic regression analysis demonstrated that predictive factors were female sex (odds ratio [OR] = 2.15, $p < 0.001$), left thoracotomy (OR = 1.74, $p = 0.040$), bilateral

thoracotomy (OR = 4.20, $p = 0.015$), NYHA class (OR = 2.80, $p < 0.001$), and age at radiography (OR = 1.13, $p = 0.001$) for $\geq 10^\circ$ scoliosis; cardiomegaly (OR = 4.15, $p = 0.001$), NYHA class (OR = 2.80, $p < 0.001$), and age at radiography (OR = 1.22, $p < 0.001$) for $\geq 20^\circ$ scoliosis; cardiomegaly (OR = 3.29, $p = 0.016$), number of surgical procedures (OR = 1.41, $p = 0.033$), and age at radiography (OR = 1.19, $p = 0.026$) for $\geq 30^\circ$ scoliosis; and cardiomegaly (OR = 6.13, $p = 0.005$) for $\geq 45^\circ$ scoliosis (Table VIII).

TABLE VIII Multivariate Analysis*

	Odds Ratio (95% CI)	P Value
$\geq 10^\circ$ (mild, moderate, pronounced, and surgical-candidate) scoliosis		
Female sex	2.15 (1.45-3.18)	<0.001
Left thoracotomy	1.74 (1.24-3.40)	0.040
Bilateral thoracotomy	4.20 (1.32-13.3)	0.015
NYHA class	2.80 (1.66-4.70)	<0.001
Age at radiography	1.13 (1.05-1.21)	0.001
$\geq 20^\circ$ (moderate, pronounced, and surgical-candidate) scoliosis		
Cardiomegaly	4.15 (2.06-8.38)	0.001
NYHA class	2.80 (1.53-5.10)	<0.001
Age at radiography	1.22 (1.10-1.36)	<0.001
$\geq 30^\circ$ (pronounced and surgical-candidate) scoliosis		
Cardiomegaly	3.29 (1.25-8.67)	0.016
No. of surgical procedures	1.41 (1.03-1.94)	0.033
Age at radiography	1.19 (1.01-1.35)	0.026
$\geq 45^\circ$ (surgical-candidate) scoliosis		
Cardiomegaly	6.13 (1.71-22.0)	0.005

*CI = confidence interval, and NYHA = New York Heart Association.

Discussion

The present study demonstrated that approximately half of patients with CHD who underwent cardiac surgery during the first year of life developed scoliosis. The prevalence of pronounced or surgical-candidate scoliosis ($\geq 30^\circ$) also was high (5.0%). While female sex and surgical approach were predictors of $\geq 10^\circ$ scoliosis, cardiomegaly and heart failure were associated with more-pronounced scoliosis. Particularly, cardiomegaly was the most significant predictive factor for $\geq 20^\circ$ and $\geq 30^\circ$ scoliosis. Age at the time of radiography was a predictor of $< 45^\circ$ scoliosis; however, the relative association was small.

Prevalence of Scoliosis After Surgical Treatment of CHD and Related Factors

The prevalence of idiopathic scoliosis has been reported to be 2% to 3% for mild scoliosis ($\geq 10^\circ$) and 0.1% to 0.3% for pronounced scoliosis ($\geq 30^\circ$)^{12,13}. However, the prevalence of $\geq 10^\circ$ scoliosis in the present study was 10 to 20 times higher than that of idiopathic scoliosis, similar to the findings in a previous report by Ruiz-Iban et al., in which the prevalence of scoliosis was 46% in patients who underwent cardiac surgery before the age of 18 months⁷. Coronal spinal deformity caused

by mechanical injury to the costovertebral joints after sternotomy or thoracotomy¹⁴ is thought to result in the high prevalence of scoliosis after congenital heart surgery during the early period of life, when the skeletal structure is immature and grows rapidly.

All of the identified predictive factors for scoliosis in the present study, including female sex, heart-related factors (NYHA class), surgical approach, number of surgical procedures, and age at radiography, have already been reported⁴⁻⁹. The slightly higher prevalence of scoliosis in female patients is similar to that in previous reports⁴⁻⁹; however, multivariate analysis revealed that female sex was not a predictor of $\geq 20^\circ$, $\geq 30^\circ$ or $\geq 45^\circ$ scoliosis (female-to-male ratios: 1.4 for $\geq 20^\circ$ scoliosis, 1.7 for $\geq 30^\circ$ scoliosis, and 3.0 for $\geq 45^\circ$ scoliosis). This trend toward increased female-to-male ratios with curve progression is not clear compared with that observed for idiopathic scoliosis, for which the female-to-male ratio increases with curve progression (2.0 for mild, 5.4 for moderate, and 10.0 for pronounced scoliosis)^{12,13}.

Multivariate analysis demonstrated that left and bilateral thoracotomy were predictive factors for $\geq 10^\circ$ scoliosis in the present study. However, the direct influence of thoracotomy on the occurrence of scoliosis cannot be proved from this study because 68 (86.1%) of 79 patients who underwent surgery via left thoracotomy and all 18 patients (100%) who underwent surgery via bilateral thoracotomy had undergone subsequent or previous surgery via median sternotomy. Hence, combined thoracotomy and sternotomy, rather than single left thoracotomy, which was typically applied at the closure of patent ductus arteriosus or repair of isolated coarctation of the aorta, would be a predictor of scoliosis.

Further Progression of Scoliosis After Surgery for CHD and Related Factors

NYHA class and cardiomegaly were identified as predictive factors for further progression of scoliosis. Although a theoretical explanation for the direct relationship between heart failure and scoliosis is difficult, cardiomegaly was the sole predictor of $\geq 45^\circ$ (surgical-candidate) scoliosis. Thus, our results provide new evidence on the controversial issue of the effect of cardiomegaly on scoliosis.

Anterior chest deformity due to cardiomegaly is a well-known clinical phenomenon associated with skeletal immaturity in younger patients. The chest wall on the same side as the cardiac ventricular apex usually deforms anteriorly as a result of repetitive mechanical pulsation of dilated ventricles of the heart and has been shown to be an isolated predictive factor for scoliosis in animal experiments¹⁵. Thus, cardiomegaly can be a compounding factor of scoliosis caused by the interventions on the immature rib cage during the surgical treatment of congenital heart disease.

Associated congenital anomalies or genetic disorders may be related to the high prevalence of scoliosis in patients with CHD after cardiac surgery¹⁶⁻¹⁹. However, Reckles et al. found only 1 case (<1%) of congenital scoliosis in a study of

377 patients⁸. In the present study, only 5 (1%) of 498 patients who underwent standing radiography demonstrated congenital spinal anomaly and were excluded from analysis. These results suggest that congenital anomalies or genetic disorders do not have a profound influence on the development of scoliosis in patients with CHD^{18,19}, whereas genetic disorders recently have been shown to have an influence on the development of idiopathic scoliosis^{20,21}.


Study Limitations

The present study had several limitations. First, the follow-up rate at ≥ 10 years after surgery was only 50%, largely because of the severity of the heart disorders and the fact that the patients were located all over the country; nevertheless, the follow-up rate was sufficient for statistical analysis. Second, the orthosis prescription rate for patients with more-than-moderate scoliosis is unclear. The effectiveness of prevention of curve progression with use of bracing for skeletally immature patients has been demonstrated²², and brace treatment with sufficient wearing time can significantly reduce the risk of surgery²³. Adequate intervention with bracing might have altered the final prevalence of surgical-magnitude curves in the study population. Third, although sternal and chest wall deformities caused by congenital heart surgery may result in scoliosis, the relationship between the existence of sternal and chest wall deformities and the development of scoliosis was not revealed objectively. To estimate their relationship more accurately, lateral chest radiographs are now being made for the patients in the present study and data are being accumulated. Fourth, the mean age at the latest follow-up in the present study was 14.4 years, which was slightly younger than the age at which skeletal maturity is reached. A longer follow-up period may increase the prevalence of scoliosis and affect the results of the statistical analysis. However, it is generally difficult to achieve both a long follow-up period and a high follow-up rate. Finally, Cobb angle measurement including the L2 vertebra as the lowest level in a majority of patients might lead to under-measurement or mismeasurement of the curvature, and this could result in an underreporting of patients with a lumbar curve.

Overview

In conclusion, cardiac surgery for CHD performed in the first year of life was associated with a high prevalence of scoliosis ($\geq 40\%$). Female sex and surgical approach were among the predictors of $\geq 10^\circ$ scoliosis, whereas cardiomegaly, cardiac function, and the number of surgical procedures were associated with $\geq 20^\circ$ and $\geq 30^\circ$ scoliosis. Cardiomegaly was the sole predictor of $\geq 45^\circ$ scoliosis. Thus, our results provide new evidence on the controversial issue of the effect of cardiomegaly on scoliosis. Further improvement of surgical strategies, periodic checkups involving the evaluation of standing chest radiographs for evidence of scoliosis, and cooperation between cardiac and orthopaedic surgeons should be expected to stave off the deterioration in quality of life due to spinal deformity.

Appendix

 Tables showing the results of univariate analysis are available with the online version of this article as a data supplement at [jbjs.org \(http://links.lww.com/JBJSOA/A35\)](http://links.lww.com/JBJSOA/A35). ■

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