

Safety of surgical treatment for patients with scoliosis and surgically corrected congenital cardiac malformations: a comparison with patients with scoliosis and normal hearts

Clinical article

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Object. The safety of spinal fusion has been poorly studied in children with surgically corrected congenital cardiac malformations (CCMs). The objective of this study was to evaluate the safety of spinal fusion in patients with CCMs following cardiac surgery.

Methods. A retrospective study was conducted on 32 patients with scoliosis who received surgical treatment for their CCMs (CCM group). Sixty-four age- and sex-matched patients with scoliosis and normal hearts who received spinal fusion served as the control group. These 2 groups were compared for demographic distribution, blood loss, transfusion requirements, and incidence of postoperative complications.

Results. The ages, curve pattern distributions, and number of levels fused were similar between the 2 groups before spinal fusion. Overall, a total of 7 patients in the CCM group (21.9%) and 5 (7.8%) in the control group had documented postoperative complications. The perioperative allogenic blood transfusion rate and mean red blood cell transfusion requirement in the CCM group were significantly higher than those found in patients in the control group (68.7% vs 28.1%, respectively, $p = 0.000$; and 2.68 ± 2.76 units/patient vs 0.76 ± 1.07 units/patient, respectively, $p = 0.011$). In the CCM group, a preoperative major curve magnitude $\geq 80^\circ$ was the most accurate indicator of an increased risk for a major complication ($p = 0.019$), whereas no statistically significant correlation was noted between postoperative complications and age, type of congenital heart disease, operative duration, and estimated blood loss during the operation and transfusion.

Conclusions. Spinal fusion subsequent to prior cardiac surgery is relatively safe and effective in correcting the spinal deformity for patients with scoliosis and surgically corrected CCMs. A preoperative major curve magnitude $\geq 80^\circ$ may be a risk factor in predicting postoperative complications in scoliotic patients with surgically corrected CCMs.

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KEY WORDS • scoliosis • postoperative complication • spinal deformity • congenital cardiac malformation • spine

IT has been reported that the prevalence of scoliosis in children is approximately 2%–3%.^{3,15} However, among children with congenital heart disease (CHD), scoliosis is much more common. It has been suggested that the association between the embryonic development of the cardiovascular and musculoskeletal systems could account for the high incidence of scoliosis in these children. Subsequent estimates of the rate of scoliosis in patients with CHD have ranged from 11% to 34%.^{4,13} Over the last 2 decades, advances in the fields of pediatric cardiology and pediatric cardiac surgery have led to an increase in the rates of survival of these patients,⁷

creating at the same time a subset of patients who will require spine surgery for the correction of their spinal deformity. However, there is limited published data about the safety of spinal fusion in children with surgically corrected congenital cardiac malformations (CCMs). This retrospective cohort study was undertaken to investigate the predictive factors of postoperative complications in unselected scoliotic patients with surgically corrected CCMs (CCM group) and to identify whether spinal fusion is safe in this population.

Methods

Congenital Cardiac Malformation Group

The medical records of 32 unselected patients with scoliosis (6 males and 26 females) and surgically corrected

Abbreviations used in this paper: BMI = body mass index; CCM = congenital cardiac malformation; CHD = congenital heart disease; EBL = estimated blood loss.

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CCMs operated on at Peking Union Medical College Hospital from January 2000 to January 2012 were identified from a single institutional database. Those patients with acquired cardiac disease were excluded. The diagnosis included congenital scoliosis in 12 cases, idiopathic scoliosis in 15 cases, and neuromuscular scoliosis in 5 cases. The most common heart defects were ventricular septal defect (n = 10), patent ductus arteriosus (n = 6), atrial septal defect (n = 7), and tetralogy of Fallot (n = 3; Table 1). These children had previously undergone cardiac surgical interventions at a mean age of 7.4 years (range 1.5–15.0 years), including median sternotomy, interventional treatment, and/or thoracotomy. The mean age of the patients who underwent spinal fusion in the CCM group was 13.8 years (range 9.2–18.3 years; Table 2). The average coronal Cobb angle was 68.2° (range 37°–145°). The surgical interval between prior cardiac surgical intervention and spinal fusion was 5.7 years (range 2.0–11.0 years).

Prior to the spine surgery, M-mode and 2D echocardiography, and Doppler ultrasonography of all patients were performed by the sonographer using a GE Vivid7 scanner (GE Medical Systems) equipped with a 3.4-MHz phased array transducer. Recordings and measurements were made following international guidelines.^{2,14} Electrocardiography and perioperative medication histories were also noted by checking the ward physicians' notes. Although some minor residual structural malformations were found (mild pulmonary valve stenosis in 1 case, residual leakage at membranous ventricular septal defect in 1 case, mild mitral valve regurgitation in 2 cases), all the patients had normal cardiac function with a mean ejection fraction of 63.7% (range 55%–75%) preoperatively.

Control Group

We also reviewed our database of patients with scoliosis who received surgical treatment during the same period (from January 2000 to January 2012). These patients with scoliosis but without cardiac malformations (control group) were chosen and best-matched for sex, age, and curve pattern and magnitude for the CCM group, and then matched at a 2:1 ratio, respectively. In total, 64 patients with scoliosis, including 24 patients with congenital scoliosis, 30 with idiopathic scoliosis, and 10 with neuromuscular scoliosis, were selected to serve as the control group.

TABLE 2: Clinical and radiographic patient data*

Variable	Controls	CCM
no. of patients	64	32
age (yrs)	14.0 ± 3.8	13.8 ± 5.3
sex		
male	7	6
female	57	26
BMI (kg/m ²)	18.4 ± 3.6	17.5 ± 4.0
preop curve magnitude (°)	62.5 ± 24.8	68.2 ± 31.3
curve pattern		
single thoracic	20	10
double major	44	22
no. of levels fused	8.19 ± 5.42	9.22 ± 4.32
op duration (mins)	266.1 ± 93.8	285.4 ± 79.2
EBL (ml)	664.5 ± 413.8	720.4 ± 468.1
transfusion (units/patient)	0.76 ± 1.07	2.68 ± 2.76

* Comparisons of all variables between groups were nonsignificant except for transfusion (p = 0.011). Mean values are presented ± SD.

Surgical Procedures

In the CCM group, the surgical procedures included anterior spinal fusion in 2 cases and posterior spinal fusion in 30 cases. In the control group, the surgical procedures included anterior spinal fusion in 5 cases, posterior spinal fusion in 58 cases, and a combined single-staged anterior/posterior spinal fusion in 1 case. Surgical approach was determined by curve type and curve rigidity, anterior fusion and instrumentation was indicated for thoracolumbar and lumbar curves, and anterior release and posterior fusion was required for rigid curves (> 60° on convex bending radiographs).

In the control group, special anesthetic regimens such as central venous line access and arterial blood pressure monitoring were used. A Cell Saver system (Haemonetics Cell Saver 5, Haemonetics Corporation) was used intraoperatively to reinfuse shed blood in all patients. The volume of blood and hematocrit collected by the cell saver system were recorded, and estimated blood loss (EBL) was calculated by measuring loss through the cell saver

TABLE 1: Distribution of congenital heart defects in the patients and repair status at the time of spinal fusion*

CHD Type	No. of Patients	CHD Postop Status
tetralogy of Fallot	3	complete repair
PDA	6	PDA ligation
pulmonary stenosis, isolated	2	percutaneous pulmonary valvuloplasty
ASD	7	ASD repair
mitral valve prolapse	1	mitral valve annuloplasty
aortic dilation & mitral valve prolapse	1	aortic valve replacement, mitral valve annuloplasty
aortic dilation & PDA	1	aortic valve replacement, PDA ligation
ASD & PDA	1	ASD repair, PDA ligation
VSD	10	VSD closure

* ASD = atrial septal defect; PDA = patent ductus arteriosus; VSD = ventricular septal defect.

Postoperative complications in patients with cardiac surgery

suction system and by weighing the surgical sponges. Patients who did not use anticoagulant medication received tranexamic acid at a loading dose of 30 mg/kg shortly after the induction of anesthesia, followed immediately by continuous infusion of 1 mg/kg/hr during surgery and for 5 hours after the operation. The same blood transfusion and prophylactic antibiotic guidelines were used for all patients. Allogenic blood transfusion was performed if hemoglobin decreased to less than 7.0 mg/dl or if anemic symptoms developed, such as a decrease in blood pressure to less than 100 mm Hg systolic, tachycardia greater than 100 beats per minute, or a low urine output of less than 30 ml/hour, even after initial fluid challenge with 500 ml normal saline in patients with a hemoglobin level between 7.0 and 8.0 mg/dl.^{9,10} All patients were given 1 gram of first-generation cephalosporin 30 minutes before skin incision, and received antibiotic coverage for 24 hours with 3 additional doses.

Data Collection

Information was compiled about perioperative complications occurring within 30 days after the operation. This information included both surgical (postoperative bleeding, wound breakdown, infection, implant failure, neurological compromise, and others) and nonsurgical complications (pulmonary disease, arrhythmia, hemodynamic instability requiring inotropic support, and others). All patients had a minimum of 1 year of follow-up. Follow-up data collected included postoperative Cobb angle, unanticipated late reoperation, and other complications.

Statistical Analysis

Factors associated with the occurrence of postoperative complications were identified using univariate analysis. The data analysis was performed using SPSS (version 19.0, IBM). Continuous data were compared between the CCM and control groups using the Student t-test, whereas discontinuous data were analyzed using the chi-square test. The Fisher exact test was used for small data subsets ($n < 5$). All significance tests were 2-tailed, with $p < 0.05$ representing statistical significance.

Results

Demographic Data

A summary of the clinical and radiographic data before spinal fusion for the CCM and control groups is presented in Table 2. At the time of spinal fusion, the average age was 13.8 ± 5.3 years (range 9.2–18.3 years) for the CCM group and 14.0 ± 3.8 years (range 9.8–18.5 years) for the control group. The average primary curve magnitude for the CCM and control groups was $68.2^\circ \pm 31.3^\circ$ (range 37° – 135°) and $62.5^\circ \pm 24.8^\circ$ (range 42° – 115°), respectively. Curve patterns were classified as follows: in the CCM group, there were 10 single thoracic curves (4 left and 6 right) and 22 double major curves; in the control group, there were 20 single thoracic and 44 double major curves. No significant differences were observed between the 2 groups in age, sex ratio, body mass index (BMI), curve pattern distributions, or the number of levels fused.

Operative Data

Average operative time was 285.4 ± 79.2 minutes (range 150–425 minutes) in the CCM group and 266.1 ± 93.8 minutes (range 140–430 minutes) in the control group. Operation time was unrelated to the type of spinal deformity (congenital vs noncongenital), type of CHD (isolated CHD vs complicated CHD), or history of cardiac surgery.

Average EBL was 720.4 ± 468.1 ml (range 300–2160 ml) in the CCM group and 664.5 ± 413.8 ml (range 420–1500 ml) in the control group. In the CCM group, using univariate analysis, EBL was directly related to patient age ($R^2 = 0.15$, $p = 0.023$) and magnitude of the Cobb angle ($R^2 = 0.19$, $p = 0.031$). Twenty-two patients (68.7%) in the CCM group received allogenic blood perioperatively, either as red blood cells or whole blood. On average, 2.68 ± 2.76 units/patient were given perioperatively. However, in the control group, 18 patients (28.1%) received allogenic blood transfusion perioperatively, either as red blood cells or whole blood. On average 0.76 ± 1.07 units/patient were given perioperatively in the control group. The perioperative allogenic blood transfusion rate and mean red blood cell transfusion requirement in the CCM group were both significantly higher than those found in patients in the control group ($p = 0.000$ and $p = 0.011$, respectively).

Postoperative Complications

Of the 32 patients with CCMs, 7 (21.9%) had postoperative complications (Table 3), including increased requirement of postoperative ventilatory support in 2 cases, pneumonia in 2 cases, urinary tract infection in 1 case, and transfusion-related complications in 2 cases. Among the 64 patients with noncongenital cardiac deformities, 5 (7.8%) had postoperative complications, including increased requirement of postoperative ventilatory support in 1 case, atelectasis in 1 case, wound infection in 1 case, pancreatitis in 1 case, and transfusion-related complications in 1 case. No patient had documented cardiac arrhythmia or death postoperatively in either group.

Predictive Factors of Postoperative Complications

Postoperative complications were also evaluated with respect to age, sex, curve magnitude, curve pattern, surgical procedure, type of CHD, operative duration, and EBL during operation and transfusion (Table 4). In both

TABLE 3: Number of postoperative complications in each group

Complication	Controls	CCM
increased requirement of postop ventilatory support	1	2
pneumonia	0	2
atelectasis	1	0
wound infection	1	0
urinary tract infection	0	1
pancreatitis	1	0
transfusion-related complications	1	2

TABLE 4: Predictive factors for postoperative complications*

Characteristic	Controls			CCM		
	Complications (%)	No Complications (%)	p Value	Complications (%)	No Complications (%)	p Value
age (yrs)						
9–13	2 (20.0)	8 (80.0)	0.112†	2 (40.0)	3 (60.0)	0.296†
≥13	2 (3.7)	52 (96.3)		5 (18.5)	22 (81.5)	
sex						
male	1 (14.3)	6 (85.7)	0.451†	2 (40.0)	3 (60.0)	0.296†
female	4 (7.0)	53 (93.0)		5 (18.5)	22 (81.5)	
curve magnitude (°)						
<80	2 (4.5)	42 (95.5)	0.171†	2 (9.1)	20 (90.9)	0.019†
≥80	3 (15.0)	17 (85.0)		5 (50.0)	5 (50.0)	
curve pattern						
single thoracic	2 (13.3)	13 (86.7)	0.583†	1 (12.5)	7 (87.5)	0.646†
double major	3 (6.1)	46 (93.9)		6 (25.0)	18 (75.0)	
forced vital capacity (%)	68.2 ± 19.8	72.1 ± 20.6	0.712	63.4 ± 26.2	68.5 ± 20.1	0.614
no. of levels fused	8.44 ± 5.11	7.81 ± 4.51	0.751	9.53 ± 4.26	8.61 ± 4.21	0.543
surgical procedure			0.553			1.000
ant spinal fusion	1 (20.0)	4 (80.0)		0 (0)	2 (100)	
pst spinal fusion	4 (6.9)	54 (93.1)		7 (23.3)	23 (76.7)	
1-stage ant/pst spinal fusion	0 (0)	1 (100)		NA	NA	
type of CHD						0.590
isolated	NA	NA		5 (19.2)	21 (80.8)	
complicated	NA	NA		2 (33.3)	4 (66.7)	
op duration (mins)	262.2 ± 108.8	271.3 ± 88.9	0.813	294.6 ± 84.7	261.8 ± 61.8	0.651
EBL (ml)	624.29 ± 328.17	707.60 ± 302.56	0.209	680.2 ± 479.4	768.5 ± 264.5	0.728
transfusion (units/patient)	0.82 ± 1.66	0.52 ± 1.40	0.558	2.14 ± 2.38	2.82 ± 3.06	0.503

* Mean values are presented ± SD. ant = anterior; NA = not applicable; pst = posterior.

† Fisher exact test.

groups, the rates of postoperative complications were similar between female and male patients. However, a higher frequency of postoperative complications was observed in younger patients (9–13 years) than in older patients (≥ 13 years). A higher frequency of postoperative complications was also observed in patients with larger curve magnitudes (≥ 80°) than in those with smaller curve magnitudes (< 80°). Based on univariate analyses, the curve magnitude was determined to be significantly associated with postoperative complications in the CCM group ($p = 0.019$), whereas no statistically significant correlation was noted between postoperative complications and age, type of CHD, operative duration, or EBL during operation and transfusion ($p > 0.05$).

Discussion

Surgical intervention has been widely used for the treatment of children with congenitally malformed hearts and is currently considered as an effective modality for preventing the secondary complications caused by cardiac malformations. With the development of techniques in surgical and medical management, more than 85% of children with congenitally malformed hearts undergoing

surgical treatment now reach adulthood.¹² Progressive spinal deformity can induce problems related to a significant reduction of pulmonary function in patients with previous cardiovascular lesions. Therefore, the number of patients with scoliosis and surgically corrected CCMs needing spinal deformity corrective surgery for scoliosis has increased in recent years. However, in patients with scoliosis who have already received cardiac surgery, the safety of spinal fusion still remains virtually unknown. To our knowledge, this study involves the largest sample size so far in the literature evaluating the safety of spinal fusion in patients with scoliosis and surgically corrected CCMs with a follow-up of at least 3 years.

As a reference and for comparison, the outcomes of spinal fusion in a cohort of age- and sex-matched patients with scoliosis and normal hearts were also evaluated. The indications for spine surgery were similar between the CCM and control groups, which consisted of a Risser sign > 1 and primary curve angles of more than 40°, or 30°–40° accompanied with established curve progression (at least 5° in 6 months). Moreover, for the sake of delaying the spinal fusion surgery, bracing is also recommended for young patients (< 9 years) who have curve angles of 40°–50°.

Based on the above-mentioned inclusion criteria, the

Postoperative complications in patients with cardiac surgery

current study demonstrated that the overall incidence rate of postoperative complications in the CCM patients during hospitalization was 21.9% as opposed to a rate of complications of 7.8% in patients without heart disease ($p = 0.097$). This finding appears to suggest that postoperative complications are a significant problem for scoliotic patients with surgically corrected CCMs in clinical practice. Our complication rate was noted to be similar to earlier published data,¹¹ but the complications were relatively mild and reversible. No deaths occurred in the perioperative period within our cohort as a result of postoperative complications. These findings strengthen the evidence that spinal fusion is a safe procedure in the majority of children with surgically corrected CCMs. Our results also indicate that in contrast to age, type of CHD, operative duration, EBL during operation, and other clinical characteristics, curve magnitude is the only predictor of postoperative complications.

The mean red blood cell transfusion requirement in the CCM group was significantly higher than that found in patients with normal hearts undergoing spine surgery. Our results also showed significant differences between the groups in perioperative allogenic blood transfusion rate, as 68.7% of patients in the CCM group in the current study received allogenic blood perioperatively, compared with 28.1% of the patients in the control group ($p = 0.000$). In our study, we had a uniform perioperative blood management strategy. No patient predonated blood, no patient had hemodilution, and all patients received recombinant human erythropoietin administration postoperatively. We hypothesize that the differences in these 2 groups of patients with their underlying etiologies can be suggested to explain the higher allogenic blood transfusion rate in the CCM group. The association of cardiomyopathy and coagulopathy due to platelet and coagulation dysfunction is well established.⁶ Moreover, some of these patients had been treated chronically with coumarin anticoagulants because of their cardiac disease, having been switched to heparin prior to the spine surgery. The need to maintain a higher central venous pressure during the surgery also limits the use of controlled hypotension to minimize the bleeding.

Patients with congenital heart malformations have been documented to be associated with higher morbidity and mortality rates in spine surgery.^{1,11} Pérez-Caballero et al. reported that 1 patient (6%) died in the immediate postoperative period due to hypovolemic shock caused by massive bleeding.¹¹ Bitan et al. came to a similar conclusion in another series of 26 patients in which 2 patients died (7.7%), with another 2 developing severe complications (7.7%).¹ However, in the present study, no deaths occurred in the perioperative period and no serious complications were recorded. In the cohort, 2 patients suffered pneumonia attributable to mechanical ventilation and 1 patient suffered urinary tract infection caused by an indwelling catheter; this was resolved with a course of antibiotics. None of our patients developed deep wound infections. The difference noted in our study as compared with other previously reported studies might be due to the fact that our center performs very thorough and preoperative planning. We also have very good cooperation among the various specialized disciplines caring for our

patients. We believe that we should not deny patients who have undergone corrective congenital cardiac surgery the chance of undergoing corrective surgery for scoliosis, as our serious complication rates are low.

It has been suggested previously that preoperative major curve magnitude was associated with an increased prevalence of major complications.^{5,8} Master et al. reported 46 major complications in 37 patients with neuromuscular scoliosis (28% prevalence), including 2 deaths, and concluded that a preoperative curve magnitude is directly associated with an increased risk for major complications.⁸ Liang et al. also found that preoperative Cobb angle correlated with postoperative pulmonary complications in patients with scoliosis with moderate or severe pulmonary dysfunction.⁵ Consistent with the studies of Master et al. and Liang et al., our study demonstrated that a preoperative major curve magnitude $\geq 80^\circ$ was the most accurate predictor of an increased risk for a major complication, suggesting spinal fusion should be carefully considered in patients with scoliosis and surgically corrected CCMs whose preoperative major curve magnitude is greater than 80° .

Several potential limitations of this study should be mentioned. First, this study was retrospective in design and did not contain a control group of patients with scoliosis and CCMs whose cardiac malformations were treated by observation alone, followed by spinal fusion. Moreover, the results are limited by a relatively small sample size and the broad time period covered. In addition, changes in anesthetic and surgical practices over time may have affected patient outcomes. We also acknowledge the limitations introduced by our patients' clinical heterogeneity.

Conclusions

Patients with scoliosis and surgically corrected CCMs who undergo spinal fusion may present to the surgeon with many possible complications due to their underlying pathology. Our findings suggest that the risk of surgically related complications in these patients correlates with their preoperative major curve magnitude. We believe that the experience of a multidisciplinary team, consisting of anesthesiologists, cardiologists, and intensive care pediatricians with experience in congenital cardiac disease, is essential for the appropriate management of these patients.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Shen. Acquisition of data: Liang, Ding, Li. Analysis and interpretation of data: Liang, Ding. Drafting the article: Liang, Ding. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Shen. Statistical analysis: Chua, Li. Administrative/technical/material support: Chua.

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