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Surgical and Health-related Quality of Life Outcomes in Children with Congenital Scoliosis during Five-Year Follow-up. Comparison to age and gender matched healthy controls

Abstract

Background: Congenital spinal anomalies represent a heterogeneous group of spinal deformities, of which only progressive or severe curves warrant surgical management. Only a limited number of studies have investigated the impact of surgery on the health-related quality of life (HRQoL) and very limited data exists comparing these outcomes to healthy controls.

Methods: A single surgeon series of 67 consecutive children with congenital scoliosis (mean age at surgery 8.0 years, range 1.0 – 18.3 years, 28 girls) undergoing hemivertebrectomy (HV, n=34), instrumented spinal fusion (ISF, n=20), or VEPTR procedure (n=13) with a mean follow-up of 5.8 years (range 2 years to 13 years). The comparison was made to age and gender matched healthy controls. Outcome measures included Scoliosis Research Society (SRS) questionnaire both pre- and postoperatively, radiographic outcomes, and complications.

Results: The average major curve correction was significantly better in the HV (60%) and ISF (51%) than in the VEPTR group (24%), respectively ($p<0.001$). Complications were noted in eight of 67 (12%) children, but all patients recovered fully during follow-up. Pain, self-image, and

function domains improved numerically from preoperative to final follow-up, but the pain score was the only one with a statistically significant change (p=0.033). The SRS pain, self-image, and function domain scores remained at a significantly lower level at final follow-up than in the healthy controls ($p \leq 0.05$), while activity score improved to the similar level.

Conclusion: Surgery for congenital scoliosis improved angular spinal deformities with reasonable risk of complications. Health-related quality of life outcomes improved from preoperative to final follow-up, but especially pain and function domains remained at a significantly lower level than in the age and gender matched healthy controls.

Level of Evidence: Therapeutic Level III.

Key Words: congenital scoliosis; hemivertebrectomy; VEPTR; health-related quality of life

Introduction

Congenital vertebral malformations are classified as defects of formation (hemivertebra), failure of segmentation (unilateral bar), and a mixed group.[1] Treatment of congenital spinal deformities vary from identification and observation to challenging spinal deformities including scoliosis or kyphosis with or without fused ribs resulting into pulmonary insufficiency[2] and spinal cord deficits.[3-5] Fully segmented hemivertebrae represents the most common indication for surgery in congenital scoliosis.[6,7] These are typically treated using hemivertebrectomy and a short pedicle screw instrumentation at an early age in order to prevent development of compensatory curves often requiring longer spinal fusion.[7] Failure(s) of segmentation have been traditionally treated with an early anteroposterior spinal fusion or growing rods.[3,4,8] Patients with fused ribs may develop thoracic insufficiency syndrome and can be treated using thoracoplasty and VEPTR instrumentation.[2]

Surgical outcome studies in children with congenital scoliosis are typically limited to reporting radiographic correction, risk of re-operation and complications. From the patient and family perspective, however, health-related quality of life (HRQoL) measurements including pain, function, self-image, and activity should also be documented using validated outcome questionnaires, such as EOSQ-24 and SRS.[9-11] In a previous study, the general HRQoL using Child Health Questionnaire (CHQ) remained at significantly lower level in children operated early for congenital scoliosis as compared with healthy children.[12]

We aimed to compare HRQoL in ambulatory children undergoing surgery for congenital scoliosis at a minimum of 2-years (mean 5.8 years) follow-up with healthy controls. We hypothesized that instrumented spinal fusion with or without hemivertebrectomy would improve HRQoL, but the quality of life would remain at a lower level than in healthy controls.

Methods

This was a retrospective study using prospectively collected data on 67 consecutive ambulatory children (28 girls) with congenital scoliosis (no neurological diagnosis) undergoing surgery by a single orthopedic spine surgeon. The mean age at surgery was 8.0 years, (range 1.0 – 18.3 years) and all had a minimum of two-year follow-up (mean follow-up 5.8 years, range 2 – 12.1 years). HRQoL outcomes were compared to healthy controls, matched by age and gender (Table 1). Age matching was performed based on the age at final follow-up. The study received approval from the Ethics Committee of the Hospital District (ETMK 96/1801/2020).

Thirty-four children (mean age at surgery 6.4 years, range 1.1 – 18.3 years) underwent all-posterior hemivertebrectomy and a short pedicle screw instrumentation[7,8,13] (HV group) for a fully segmented posterolateral hemivertebra (n=19) or posterior hemivertebra producing congenital kyphosis or kyphoscoliosis (n=15) (Figure 1). Thirteen children (mean age at surgery 12 years, range 3 to 18 years) underwent an all-posterior pedicle screw instrumentation with posterior column osteotomies. A combined anteroposterior spinal fusion for a failure of segmentation was performed in five patients (instrumented spinal fusion, ISF group, 18 children). Twelve children (mean age at surgery 4.7 years, range 1 to 10 years) were treated using longitudinal rib osteotomies and a rib to rib or rib to spine VEPTR instrumentation. One 4-year-old child with extensive posterolateral thoracolumbar failure of segmentation was treated with multiple segmental posterior bony bar osteotomies and a unilateral VEPTR instrumentation from rib to pelvis. Two patients were treated using magnetically controlled growing rods for mixed spinal anomalies (Growth-friendly management group, 15 patients).

Perioperative data were collected, as well as radiographic outcomes (Table 1) and SRS-24 scores[11] preoperatively and at follow-up visits (6 months, 2 years, and final follow-up). HRQoL questionnaires were filled by the patient or caregivers depending on the age of the patient; patients over 10 years of age filled the forms independently and younger patients were assisted by their caregivers. Patients were examined before and after surgery for their lower limb neurological function, walking ability, sitting and standing balance. Preoperatively full spinal magnetic resonance imaging (MRI) and CT were obtained to plan spinal osteotomies and implant placement. To rule out occult anomalies all children underwent also preoperative cardiac and renal ultrasound.

Perioperative Management

Perioperative management was standardized and included cefuroxime and vancomycin as antibiotic prophylaxis, tranexamic acid bolus (30mg/kg) and infusion (10mg/kg/h until wound closure), and spinal cord monitoring (motor evoked potentials, somatosensory potentials, and lumbar nerve root electroneuromyography).[14] Postoperatively patients were monitored in the pediatric intensive care unit and the mean arterial pressure was maintained between 65 and 75 mmHg (24 hours). Young patients undergoing hemivertebrectomy (before the age of 5 years) were immobilized using a hip spica cast for 6 weeks and rigid thoracolumbosacral orthosis (TLSO) for three months. Older children with spinal column osteotomies were immobilized using a rigid thoracolumbosacral orthosis for 4 months.

Radiographic Parameters

The proximal thoracic (PT), main thoracic (MT), and thoracolumbar/lumbar (TL/L) curves were measured from standing anteroposterior radiographs and thoracic kyphosis (T5-T12), lumbar lordosis (T12-S1) and segmental kyphosis or lordosis were measured from the lateral radiographs using the Cobb technique.[15,16]

SRS-24 Questionnaire

SRS-24 -questionnaire[11] was filled out by the patients and their caregivers preoperatively and at follow-up visits. The questionnaire has 7 domains: pain, general self-image, function from back condition, general level of activity, postoperative self-image, postoperative function, and satisfaction. SRS-24 pain domain score <4.0 (1=severe pain; 5=pain free) was considered clinically relevant.

Healthy Controls

The healthy controls were obtained from our previous study.[18] In that study, two-hundred and seventy-two healthy controls were selected from a population register and were invited to complete and return the SRS-22r questionnaire[17,19,20] between January 2012 and December 2015.[18] One healthy control was matched for sex and age (±3.5 years) at final follow-up for each congenital scoliosis patient filling out the SRS questionnaire at final follow-up.

To compare congenital scoliosis patients with controls, we used the first 15 questions of the preoperative SRS-24 questionnaire. Questions 1 through 15 of the SRS-24 correspond with questions 1, 2, 4, 5, 6, 8, 9, 11, 12, 14, 15, and 17 through 20 of the SRS-22r. These questions were used as the basis for 4 domains (pain, self-image, general function, and activity) of comparison between preoperative SRS-24 scores for the congenital scoliosis treatment group and the healthy control groups (Table 2). The mean total score of the exactly same 8 questions in both questionnaires were also used when comparing the intervention group and the controls, these questions were 1 through 8 for the SRS-24 and 1, 2, 4, 5, 6, 8, 9, and 11 for the SRS-22r.

Statistical analysis

Mean changes between baseline and two years were compared between the groups with linear mixed models for repeated measurements. Model included time as within factor and group as between factor, and group x time interaction was included in the model. Assumptions were checked with studentized residuals. Results for congenital scoliosis operated patients at final follow-up and control subjects were compared with Wilcoxon rank sum test. P-values less than 0.05 (two-sided) were considered as statistically significant. The data analysis was generated using SAS software, Version 9.4 of the SAS System for Windows (SAS Institute Inc., Cary, NC, USA).

Results

Patients undergoing hemivertebrectomy (mean age 6.4 years) and growth-friendly instrumentation (4.0 years) were significantly younger at the time of surgery when compared to the children undergoing instrumented spinal fusion (13.9 years, $p<0.001$) (Table 1). The mean major curves were significantly smaller in the hemivertebrectomy group as compared with the instrumented spinal fusion or the growth-friendly management groups ($p<0.001$). The mean major coronal curve correction was significantly higher in the HV (60%) (Figure 1) and ISF (51%) groups than in the growth-friendly management group (28%), respectively ($p<0.001$). The mean intraoperative blood loss was 458 mL (range 10 mL to 1400 mL).

Complications

Complication and re-operation data was available for all 67 patients during the entire follow-up. Complications were noted in eight of the 67 (12%) patients, but all patients recovered fully during follow-up (Table 3). One patient with a severe mixed congenital scoliosis and fused ribs developed cardiac arrest while being in prone position for a growing rod procedure. She was turned supine and cardiopulmonary resuscitation restored sinus rhythm. The procedure was finished in a lateral

decubitus position without further sequalae. Postoperative neurological deficit was encountered in one of 67 patients (1.5%). This was an iatrogenic medullar contusion and resulted into right-sided paraplegia postoperatively resolving fully in six months. One patient developed adding on and needed revision and extension of instrumentation. The other complications included three deep surgical site infections necessitating irrigation and debridement, one cerebrospinal fluid leakage requiring suture of the leak and one pneumothorax requiring a chest drain.

Health-Related Quality-of-Life

SRS-24 questionnaires were available for analysis in 38 patients preoperatively, 36 patients at six months follow-up, 42 patients at 2-year follow-up, and in 49 patients at final follow-up. Pain, self-image, and function domains improved during the follow-up, but the pain score was the only one with a statistically significant change ($p=0.033$) (Table 4). Total score and activity domain remained at a relatively high level from preoperative to final follow-up. Interestingly, total score and activity domain reduced significantly from preoperative to 6 months follow-up, possibly reflecting the 4 months immobilization postoperatively ($p<0.05$). The number of patients scoring under 4 in the SRS pain domain indicating moderate pain was 37% (14/38) preoperatively and 14% (7/49) at final follow-up ($p=0.015$).

Comparison to healthy controls

The SRS total score, pain, self-image, and function domain scores remained at a significantly lower level in the congenital scoliosis group at final follow-up than in the healthy controls ($p<0.05$), with no difference in the activity score between groups (Table **5**). The differences in the pain (mean 4.42 ± 0.79 vs. 4.73 ± 0.51, $p=0.0091$) and function domains (4.00 ± 0.75 vs. 4.53 ± 0.29, $p<0.001$) were most apparent between the patients and the healthy controls, while the difference in the self-image domain reached borderline significance (4.26 ± 0.72 vs. 4.53 ± 0.49, $p=0.055$). Activity score

improved to similar levels in the surgically treated patients (4.51 ± 0.80) at final follow-up as compared to controls (4.69 ± 0.41, p=0.67).

Discussion

Surgery for congenital scoliosis improved angular spinal deformities with a reasonable risk of complications and as such prevented neurological deficits typical of kyphotic congenital deformities.[1,3,4] Health-related quality of life improved or were maintained from preoperative to final follow-up, but especially pain and function domains remained at a significantly lower level at final follow-up than in age and gender matched healthy controls.

Strengths and limitations

This study was a retrospective analysis of consecutively treated children with congenital scoliosis. Another limitation of our study is the somewhat different questionnaires used in the study groups. However, we opted to keep the same original SRS-24 questionnaire in the surgical cohort to provide comparable data from preoperative to 5-year follow-up. In order to do a reliable comparison, we chose the questions from both questionnaires, which were the same to build up the pain, activity, self-image, and function domains. We used the fifteen preoperative questions of SRS-24 and the preoperative domains in SRS-24. Our results were supported using the eight of the fifteen preoperative questions, which were exactly alike with similar scoring. In the remaining seven questions the scoring was not totally identical, but close enough to make a valid comparison. Not all patients filled out the questionnaires at each follow-up visit. The relatively small number of patients operated using different techniques (hemivertebrectomy, instrumented spinal fusion or growth-friendly management) limited us from performing subgroup analyses. Even after a mean follow-up of 5 years most of our patients were skeletally immature and growing. Therefore, their

results need to be re-assessed at skeletal maturity to evaluate the final outcomes. The EOSQ-24 questionnaire[10] would have been more suitable HRQoL outcome tool to assess the impact of surgical management in the current study on congenital scoliosis. Additionally, SRS questionnaires were planned to be filled out by the patient, but the young age of our hemivertebrectomy and growth-friendly management patients prevented themselves to fill out this questionnaire.

The study has several strengths. Despite the retrospective analysis, the data collection was prospective on consecutively enrolled patients. They were all operated by the same senior surgeon using standardized surgical and anesthesiological techniques, early hemivertebrectomy with short fusion, instrumented spinal fusion with segmental pedicle screws or VEPTR instrumentation. The mean follow-up was over 5 years with a relatively high follow-up rate (100% for reoperation and complications; 73% for HRQoL). We used validated standardized questionnaires, and the data comprises of patients' own report and results are compared to age and gender matched healthy controls. All children in the current study were ambulatory and without developmental delay or disorders.

Comparison with previous data

EOSQ-24 was developed as a disease specific instrument that measures HRQoL outcomes in early onset scoliosis (EOS) and was validated for this patient group including congenital scoliosis in 2018.[10] EOSQ-24 is completed by the caregivers. The Scoliosis Research Society outcome questionnaire 24 (SRS-24) was developed and published in 1999 to evaluate the outcomes of adolescent idiopathic scoliosis and to be completed by the patient.[11] As SRS-24 was published and validated over 20 years ago it has been used as an estimate of the HRQoL of patients in pediatric spine surgery including EOS and congenital scoliosis.[6,21,22] Although SRS-24 does not measure caregiver burden, it includes domains for pain, self-image, function, activity, and satisfaction. Recently, Li et al.[21] reported a strong correlation between EOSQ-24 and SRS-22 in pain, function,

and mental health, and reasonable correlation with satisfaction especially in group of patients aged 10 or above. Thus, to present a long-term outcome study including preoperative assessment of HRQoL in children with congenital scoliosis the SRS outcome questionnaire appear valid. The minimum clinically important difference (MCID) values for the change in the SRS-22r have been reported for the pain domain (0.20), self-image 0.98, and activity 0.08.[23] In the current study only pain domain showed an increase of 0.27, which exceeds the MCID value, while self-image (0.24) and activity domains (-0.05) did not reach these reported values.

Ramo et al.[24] reported baseline EOSQ-outcomes in 610 children with early onset scoliosis (EOS). The etiological classification of EOS (C-EOS)[25] had the most significant effects on the HRQoL. Children with congenital scoliosis had similar baseline EOSQ domains than children with idiopathic early onset scoliosis. Ambulatory status, developmental delay, and medical comorbidities had a relatively limited effect on the HRQoL outcomes. The current study shows that pain, self-image, and function remain at a significantly lower level in the congenital scoliosis patients compared to healthy controls. While the activity domain improves to a similar level as that of the age and gender matched healthy children. These findings support the previous studies, that surgery for congenital scoliosis improves or maintains HRQoL,[9] but general assessment of physical function remains at a lower level than in healthy children.[12] We hypothesize that postoperative immobilization reduced the HRQoL domains in the current study at the six months follow-up, which then improved to baseline by the two-year follow-up.

HRQoL outcomes including pain, self-image, and function are important for patients, families, and the treating health care system. However, it should be noted that patients with severe untreated congenital and especially kyphotic spinal deformities may develop neurological deficits and even paraplegia.[1,3,4] Thus, it is possible that HRQoL outcomes do not fully reflect the effectiveness of timely and technically appropriate surgical management of congenital spinal deformities preventing severe neurological complications.

Conclusion

Surgery for congenital scoliosis improved angular spinal deformities with a reasonable risk of complications none of which were permanent. Health-related quality of life outcomes and especially pain improved from preoperative to final follow-up. However, SRS total score as well as pain and function domains remained at a significantly lower level at a mean 5-year follow-up compared to the age and gender matched healthy controls, while activity score was improved to a similar level as in the healthy controls.

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Table and Figure Legends

TABLE 1. Clinical Characteristics of the Patients with Congenital Scoliosis and Healthy Controls.

	Hemivertebrectomy N=34	Growth-friendly N=15	Instrumented Spinal fusion, N=18	All N=67	Healthy controls, N=51
Age at the time of surgery	6.4 ± 5.4	4.0 ± 3.0	13.9 ± 4.0 (p<0.001)‡	8.0 ± 5.6	NA
Age at Final FU	11.6 ± 5.3	11.4 ± 3.2	18.7 ± 4.0 (p<0.001)‡	13.2 ± 5.6	16.0 ± 4.6
Gender, females	15 (44 %)	6 (40 %)	6 (33 %)	28 (42 %)	21 (41 %)
Major coronal curve (◦)					
Preoperative	38 ± 17	50 ± 19	45 ± 12 (p<0.001)‡	40 ± 15	NA
Postoperative	15 ± 14	43 ± 20	23 ± 11 (p<0.001)‡	22 ± 18	
Final FU	15 ± 13	36 ± 16	20 ± 13 (p<0.001)‡	23 ± 13	
Major sagittal curve (◦)					
Preoperative	34 ± 19	48 ± 15	64 ± 14 (p<0.001)‡	45 ± 25	NA
Postoperative	30 ± 16	51 ± 14	27 ± 12 (p<0.001)‡	32 ± 18	
Final FU	29 ± 13	42 ± 13	34 ± 16 (p<0.001)‡	33 ± 15	
Revision surgery†, number (%)	4 (12 %)	10 (67 %)	2 (11 %) (p<0.001)‡	16 (24 %)	NA
Number of procedures per patient	1.1	5.2	1.1 (p<0.001)‡	2.0	NA
Associated anomaly**					
Cardiac	2 (6 %)	2 (15 %)	3 (15 %)	7 (10 %)	NA
GI	2 (6 %)	0	0	2 (3 %)	
Other orthopaedic	1 (3 %)	4 (31 %)	3 (15 %)	8 (12 %)	

*Data presented as mean and SD unless otherwise stated, FU=follow-up, NA=data not available,

†Includes surgical lengthening procedures. ‡Statistical comparison between the study groups.

**Associated cardiac anomalies/comorbidities included surgically treated tetralogy of Fallot (2 patients),

atrial septal defect, ventral septal defect, univentricular heart (child with three operations), Wolf-Parkinson-White, dextrocardia. Associated gastrointestinal anomalies included surgically treated anorectal malformation, esophageal atresia. Other associated anomalies included Jeune syndrome, Goldenhar syndrome, absent kidney, Down syndrome, Talus verticalis (reverse Ponseti treated), arthrogryposis (2 patients), hydrocephalus (ventriculoperitoneal shunt).

TABLE 2. Questions used to compare SRS-24 (patients) and SRS-22r (controls) for the domains.

SRS Domain	SRS-24	SRS-22r
Pain	1, 2, 3, 6, 8	1, 2, 4, 8, 11
General self-image	5, 14, 15	6, 19, 20
General function	7, 12, 13	9, 15, 18
General activity	4, 9, 10	5, 12, 17

TABLE 3. Complications of The Surgical Group.

Patient age, Sex	Complication	Time point	Outcome
2.5 years, female, MCGR revision	Cardiac arrest	Intraoperative	Sinus rhythm obtained after cardiopulmonary resuscitation
15.4 years, male, VCR	Spinal cord contusion	Intraoperative	Wide decompression, resolved during FU
1.8 years, female, Hemivertebrectomy	Deep surgical site infection	1 month postoperative	Two post-operative irrigation and debridement, 6-month antibiotic treatment. Implants retained
1,0 years, female, Hemivertebrectomy	Screw penetrated skin, deep surgical site infection	24 months postoperative	Irrigation and debridement, local muscle coverage. Implants retained
9.1 years, male, VCR	Deep surgical site infection	26 months postoperative	Irrigation and debridement, 6-month antibiotic treatment. Implants retained
16.6 years, male, Instrumented spinal fusion	Adding on	11 month postoperative	Reoperation, instrumentation was extended from T2-T11 to T2-L3
5.8 years, male, Hemivertebrectomy	Cerebrospinal fluid leakage	Intraoperative	Suture repair of the dural leak
3.7 years, female, Hemivertebrectomy	Pneumothorax	Intraoperative	Pleural drainage

TABLE 4. SRS Domains in Surgically Treated Patients*

SRS-24 Domain	Preoperative (n=38)	6-month FU (n=36)	2-year FU (n=42)	Final FU (n=49)	P Value†
Pain	4.04 ± 0.70	4.22 ± 0.58	4.16 ± 1.05	4.31 ± 0.75	0.033
Self-image	4.06 ± 0.92	4.22 ± 0.81	4.15 ± 1.09	4.30 ± 0.67	0.097
Function	4.05 ± 0.65	4.02 ± 0.48	4.00 ± 0.83	4.05 ± 0.70	0.38
Activity	4.60 ± 0.74	4.18 ± 0.94	4.17 ± 0.46	4.55 ± 0.74	0.95
Postop self-image	NA	3.28 ± 0.53	2.97 ± 1.04	3.16 ± 0.69	NA
Postop function	NA	2.24 ± 1.18	2.63 ± 1.13	3.05 ± 0.89	NA
Satisfaction	NA	4.18 ± 0.64	3.62 ± 1.62	4.24 ± 0.74	NA
Total Score	4.12 ± 0.51	3.88 ± 0.45	4.01 ± 0.61	4.04 ± 0.58	0.57

*Values given as mean and SD. †Comparison between preoperative and final follow-up, NA = not applicable, FU = follow-up. The preoperative SRS-24 questionnaires were available for 17 in the hemivertebrectomy cohort, six for the growth-friendly cohort, and 15 for the instrumented spinal fusion cohort. Similarly, this questionnaire was available at final follow-up for 24 in the hemivertebrectomy cohort, 8 for the growth-friendly cohort, and 17 for the instrumented spinal fusion cohort. Thirty-two patients had both preoperative and final follow-up SRS-24 questionnaires available.

TABLE 5. Comparison of SRS Domain Scores Between Congenital Scoliosis and Healthy Controls*

SRS domain	Congenital scoliosis at final FU	Healthy controls	p value†
Pain§	4.42 ± 0.79	4.73 ± 0.51	0.0091
Self-image	4.26 ± 0.72	4.53 ± 0.49	0.055
Function	4.00 ± 0.75	4.89 ± 0.29	<0.001
Activity	4.51 ± 0.80	4.69 ± 0.41	0.67
Total of 8 same questions	4.40 ± 0.70	4.65 ± 0.47	0.035

*Values are given as mean and SD, †Comparison of whole congenital scoliosis group with healthy controls.

§ Pain domain does not contain postoperative questions.

FIGURE 1 (A-D). Three-year-old girl with cervicothoracic congenital scoliosis due to multiple hemivertebrae demonstrated in preoperative radiograph (A) and 3D CT (B). A single stage, all posterior, two level hemivertebrectomy (C7, T1) and instrumentation between C2 and T6 was performed after three weeks halogravity traction. Two-year follow-up images of cervical spine (C, D).