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The long-term outcome of patients treated operatively and non-operatively for scoliosis deformity secondary to spina bifida

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The purpose of this study was to evaluate the long-term outcome of adults with spina bifida cystica (SBC) who had been treated either operatively or non-operatively for scoliosis during childhood.

We reviewed 45 patients with a SBC scoliosis (Cobb angle ≥ 50º) who had been treated at one of two children's hospitals between 1991 and 2007. Of these, 34 (75.6%) had been treated operatively and 11 (24.4%) non-operatively. After a mean follow-up of 14.1 years (standard deviation (SD) 4.3) clinical, radiological and health-related quality of life (HRQOL) outcomes were evaluated using the Spina Bifida Spine Questionnaire (SBSQ) and the 36-Item Short Form Health Survey (SF-36).

Although patients in the two groups were demographically similar, those who had undergone surgery had a larger mean Cobb angle (88.0º (SD 20.5; 50.0 to 122.0); *versus* 65.7º (SD 22.0; 51.0 to 115.0); p < 0.01) and a larger mean clavicle–rib intersection difference (12.3 mm; (SD 8.5; 1 to 37); *versus* 4.1 mm, (SD 5.9; 0 to 16); p = 0.01) than those treated non-operatively. Both groups were statistically similar at follow-up with respect to walking capacity, neurological motor level, sitting balance and health-related quality of life (HRQOL) outcomes.

Spinal fusion in SBC scoliosis corrects coronal deformity and stops progression of the curve but has no clear effect on HRQOL.

Cite this article: *Bone Joint J* 2014; 96-B:1244–51

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doi:10.1302/0301-620X.96B9.33857 $2.00

Bone Joint J 2014;96-B:1244–51.
Received 18 February 2014; Accepted after revision 25 June 2014

Scoliosis affects up to 50% of patients with spina bifida cystica (SBC) and can progress to affect sitting balance.[1-5] Spinal fusion has been advocated to correct deformity, prevent curve progression, improve sitting balance and increase function.[1,6] Unfortunately, studies have reported a decreased ability to carry out the activities of daily living (ADL) after surgery.[2,7-11] Mazur et al[12] reported an improvement in sitting balance but a decreased ability to walk in patients with SBC after a spinal fusion. Other studies have reported no significant relationship between spinal deformity and physical function, self-perception or self-motivation.[3,13] Although spinal fusion is frequently undertaken, its functional benefit for patients with SBC remains uncertain.[3,14]

Despite complication rates approaching 80%, few studies have evaluated the post-operative health-related quality of life (HRQOL) of patients with SBC.[15-19] The purpose of this study was to evaluate the HRQOL outcomes of adults with SBC who had been treated either operatively or non-operatively for scoliosis during childhood.

Patients and Methods

This was a two-centre retrospective comparative review. All patients diagnosed with SBC and treated between 1 January 1991 and 31 December 2007 were identified from their respective hospital databases. Ethical approval was obtained before enrolling patients in the study.

We included patients with SBC (meningomyelocoele, meningocoele, lipomeningocoele or lipomeningomyelocoele) who were ≤ 18 years of age with a primary Cobb angle ≥ 50º and a fixed coronal curve on anteroposterior (AP) radiographs.[20] Patients with other causes of scoliosis (idiopathic or traumatic) and those undergoing a non-instrumented fusion or isolated kyphectomy (apical vertebrae resection, decancellation techniques or vertebral osteotomies) were excluded. SBC patients with congenital deformities (sacral agenesis, unsegmented bars or hemivertebrae) were included.

Of 170 eligible patients, 129 (75.8%) underwent fusion and 41 (24.1%) were treated non-operatively; 125 failed to respond to our request for participation (Table I), giving a

Table I. Baseline assessments of study responders *versus* non-responders

		Responders n = 45 (%)	Non-responders n = 125 (%)	p-value*
Gender	Male	21 (*46.7*)	49 (*39.2*)	0.39
	Female	24 (*53.3*)	76 (*60.8*)	
Institution	Hospital for Sick Children	35 (*77.8*)	87 (*69.6*)	0.34
	Children's Hospital of Eastern Ontario	10 (*22.2*)	38 (*30.4*)	
Treatment	Operative	34 (*75.6*)	95 (*76.0*)	0.99
	Non-operative	11 (*24.4*)	30 (*24.0*)	
Mean age at follow-up (yrs) (SD; range)		26.7 (SD 4.7; 18.8 to 34.7)	29.0 (SD 4.4; 19.4 to 38)	0.01
Mean Last Known Cobb Angle (°)		56.3 (SD 31.2; 12 to 162)	52.6 (SD 27.5; 8 to 154)	0.50

SD, standard deviation

* Continuous variables; independent *t*-test; categorical variables; chi-squared test

Table II. Baseline assessment of the study cohort (Mean values with standard deviations (SD) and range where appropriate)

		Operative n = 34 (%)	Non-operative n = 11 (%)	Total n = 45	p-value*
Gender (%)	Male	18 (*52.9*)	3 (*27.3*)	21 (*46.7*)	0.18
	Female	16 (*47.1*)	8 (*72.7*)	24 (*53.3*)	
Living status (%)	Home	32 (*94.1*)	11 (*100*)	43 (*95.6*)	0.41
	Long-term care facility	2 (*5.9*)	0 (*0*)	2 (*4.4*)	
Neurological motor level (%)	Thoracic-L3	25 (*73.5*)	7 (*63.6*)	32 (*71.1*)	0.70
	L4-Sacral	9 (*26.5*)	4 (*36.4*)	13 (*28.9*)	
Hoffer classification (%)	Ambulatory (I-II)	11 (*32.4*)	4 (*36.4*)	15 (*33.3*)	0.54
	Non-ambulatory (III-IV)	23 (*67.6*)	7 (*63.6*)	30 (*66.7*)	
Pressure ulcers (%)		9 (*26.5*)	1 (*9.1*)	10 (*22.2*)	0.41
Urinary incontinence (%)		31 (*91.2*)	10 (*90.9*)	41 (*91.1*)	0.69
Gastrostomy tube (%)		6 (*17.6*)	1 (*9.1*)	7 (*15.6*)	0.66
Ventriculoperitoneal shunt (%)		29 (*85.3*)	7 (*63.6*)	36 (*80.0*)	0.19
Baseline radiology	Cobb angle (o)	88.0 (SD 20.5; 50.0 to 122.0)	65.7 (SD 22.0; 51.0 to 115.0)	82.5 (SD 22.8; 50.0 to 122.0)	< 0.01
		n = 34	n = 11	n = 45	
	Kyphosis (o)	41.2 (SD 24.8; 2 to 90)	53.3 (SD 18.6; 22 to 75)	44.9 (SD 23.5; 2 to 90)	0.16
		n = 25	n = 11	n = 36	
	Lordosis (o)	61.8 (SD 45.2; 15 to 170)	54.9 (SD 38.5; 2 to 115)	59.6 (SD 42.7; 2 to 170)	0.67
		n = 23	n = 11	n = 34	
	Pelvic obliquity (o)	18.8 (SD 12.6; 5 to 45)	10.8 (SD 8.1; 0 to 25)	16.6 (SD 12.0; 0 to 45)	0.06
		n = 29	n = 11	n = 40	
	Coronal balance (mm)	40.1 (SD 28.0; 4 to 105)	22.9 (SD 17.8; 0 to 48)	35.0 (SD 26.4; 0 to 105)	0.07
		n = 26	n = 11	n = 37	
	Clavicle-rib intersection difference (mm)	12.3 (SD 8.5; 1 to 37)	4.1 (SD 5.9; 0 to 16)	9.9 (SD 8.6; 0 to 37)	0.01
		n = 27	n = 11	n = 38	

*Continuous variables: independent samples *t*-test; categorical variables: chi-squared test

participation rate of 26.5% (45 of 170 patients). This is similar to that achieved by another multicentre study of this population (~30%).[21] Reasons for not participating included lack of interest (n = 53) and inability to locate the patient (n = 63). In all, nine of the 125 (7.2%) patients had died. The mean age of non-responders at follow-up was 29.0 years (standard deviation (SD) 4.4; 19.4 to 38.0). For non-responders, the mean baseline Cobb angle of patients who had undergone fusion was 84.3° (SD 19.8; 50° to 130°) and the last known Cobb angle of patients treated non-operatively was 68.1° (SD 18.0; 50° to 110°). Overall, non-responders were pretty similar in their demographics and radiological measurements to the patients enrolled in the study

Of the 45 (26.5%) patients who participated in the study, 21 (46.7%) were men and 24 (53.3%) were women. Of these, 34 (75.6%) underwent fusion and 11 (24.4%) were treated non-operatively. With respect to location, 35 (77.8%) were treated at the Hospital for Sick Children, Toronto, Canada, and the other ten (22.2%) at The Children's Hospital of Eastern Ontario, Ottawa, Canada.

After giving informed consent, 28 (62.2%) patients attended outpatients for clinical and radiological assessment and were given a HRQOL questionnaire to complete. 17 patients (37.8%) who were interested in participating but unable to attend ('remote participants')

Table III. Peri- and post-operative assessments (Mean values with standard deviations (SD) and range where appropriate)

		Operative patients n = 34
Age at surgery (yrs)		12.2 (SD 2.8; 5.4 to 16)
Haematocrit		0.34 (SD 0.04; 0.3 to 0.4)
Surgical duration (min)		592.0 (SD D 102.4; 255 to 720)
Blood loss (ml/kg)		63.8 (SD 38.3; 5.1 to 175.4)
Peri-operative blood transfusion (%)		28 (*82.4*)
ASA* physical status classification > 3 (%)		17 (*50*)
Approach (%)	Anterior	4 (*11.8*)
	Anterior + posterior	25 (*73.5*)
	Posterior	5 (*14.7*)
Instrumentation (%)	Pedicle screws and rod	7 (*20.6*)
	Sublaminar wiring and rod	12 (*35.3*)
	Combination	15 (*44.1*)
Bone graft (%)	Autograft	8 (*24.2*)
	Allograft	5 (*15.2*)
	Combined	20 (*60.6*)
Distal extension (%)	Lumbar	10 (*29.4*)
	Sacrum/pelvis	24 (*70.6*)
Length of arthrodesis (number of movement segments)		12.9 (SD 3.5; 5 to 17)
Duration of admission (days)		12.0 (SD 6.2; 4 to 37)
Immediate post-operative	Cobb angle (o)	40.4 (SD 19.2)Min:10.4Max:72.9
	Kyphosis (o)	40.8 (SD 27.3; 3 to 119)
	Lordosis (o)	46.9 (SD 23.5; 5 to 86)
	Pelvic obliquity (o)	6.5 (SD 6.1; 0 to 25)
	Coronal balance (mm)	28.1 (SD 22.4; 0 to 80)
	Clavicle–rib intersection difference (mm)	9.0 (SD 8.0; 0 to 30)

* American Society of Anesthesiologists[25]

had their HRQOL questionnaire and ambulatory status determined by telephone.

Baseline assessment. The patients' demographic details and radiographs were retrieved from their records and their relevant medical comorbidities recorded (Table II).[14,16,22-24] Living status was categorised as living 'at home' or living in 'long-term care facilities' for four or more days per week. Peri- and post-operative complications were also recorded (Table III).[25] Surgical site infections (SSI) were categorised as early (less than three months) or late (three months or more),[26] and as superficial or deep.[27,28] Pseudarthrosis was defined as movement demonstrated radiologically or at surgical exploration.[29,30] Baseline pulmonary function tests (PFTs) were obtained and were undertaken six months before fusion.[31] Baseline PFTs were not available for patients who had been treated non-operatively.

The demographics of patients in the two groups were statistically similar, with the exception of the baseline Cobb angle ($p < 0.01$, independent *t*-test) (Table II).

The 34 fusions were performed by eight surgeons (five from The Hospital for Sick Children and three from the Children's Hospital of Eastern Ontario) (Table III) with a concurrent kyphectomy in five patients.

The implants used for fusion were classified as pedicle screws and rods, sublaminar wiring and rods, or a combination of these. The surgical approach in each case was categorised as anterior, combined anterior and posterior (staged or non-staged) or posterior.

All 11 patients treated non-operatively had treatment which consisted of routine clinical and radiological assessment, bracing for comfort, modification of their walking aids or chair, sacral ulcer prevention and other treatments as necessary, similar to those patients who had undergone fusion.

Radiological assessment. We reviewed all chest and full-length AP and lateral radiographs of the spine, with the exception of traction or bending films. At follow-up, new full-length sitting AP and lateral spine radiographs were obtained. Measurements were performed by a single observer to reduce variability.[20]

The Cobb angle and the clavicle-rib intersection difference (CRID) were measured as previously described.[32-34] Pelvic obliquity was measured by the method described by Osebold et al.[35] Coronal imbalance was measured using the method of Li et al.[33] Thoracic kyphosis and lumbar lordosis were measured using the techniques described by Jackson and McManus.[36] If radiographs were unavailable, the radiology reports were reviewed. If the radiological parameters were not included in the reports, they were not calculated.[37]

The imaging of patients who underwent fusion was reviewed at three intervals: zero to three months before fusion ('baseline'), less than three months after fusion ('post-operative'), and more than six years after fusion ('follow-up'). The imaging of patients treated non-operatively was reviewed at two time-points: a) first clinical assessment with Cobb angle ≥ 50° ('baseline'), and b) more than six years after the initial assessment ('follow-up').

Table IV. Follow-up assessments of the study cohort (Mean values with standard deviations (SD) and range where appropriate)

		Operative n = 34	Non-operative n = 11	Total n = 45	p-value*
Age at follow-up (years)		27.0 (SD 4.6; 18.8 to 34.7)	25.6 (SD 5.1; 20.2 to 33.6)	26.7 (SD 4.7; 18.8 to 34.7)	0.380
Neurological motor level (%)	Thoracic-L3	14 (*73.6*)	7 (*77.8*)	21 (*75.0*)	0.30
	L4-Sacral	5 (*26.3*)	2 (*22.2*)	7 (*25.0*)	
Hoffer classification[36] (%)	Ambulatory (I-II)	6 (*17.6*)	4 (*36.4*)	10 (*22.2*)	0.23
	Non-ambulatory (III-IV)	28 (*82.4*)	7 (*63.6*)	35 (*77.8*)	
Sitting Balance Scale (%)	Requiring arms	5 (*26.3*)	3 (*33.3*)	8 (*28.6*)	0.89
	Not requiring arms	0 (*0*)	0 (*0*)	0 (*0*)	
	Able to reach	6 (*31.6*)	3 (*33.3*)	9 (*32.1*)	
	Able to shift weight	8 (*42.1*)	3 (*33.3*)	11 (*39.3*)	
Follow-up radiology	Cobb angle (o)	47.3 (SD 22.4; 12 to 96) n = 32	85.4 (SD 38.4; 55 to 162) n = 10	56.3 (SD 31.2; 12 to 162) n = 42	< 0.01
	Kyphosis (o)	39.2 (SD 25.3; 5.6 to 140) n = 31	45.7 (SD 22.9; 10 to 76) n = 10	40.8 (SD 24.6; 5.6 to 140) n = 41	0.48
	Lordosis (o)	47.2 (SD 25.6; 5 to 102) n = 31	62.3 (SD 52.7; 5 to 169) n = 10	50.8 (SD 34.0; 5 to 169) n = 41	0.23
	Pelvic obliquity (o)	9.7 (SD 8.1; 0 to 28) n = 31	12.9 (SD 12.5; 1 to 40.5) n = 10	10.5 (SD 9.3, 0 to 40.5) n = 41	0.35
	Coronal balance (mm)	27.4 (SD 21.9; 0 to 78) n = 31	30.5 (SD 42.1; 0 to 143.5) n = 10	28.2 (SD 27.6; 0 to 143.5) n = 41	0.76
	Clavicle rib intersection difference (mm)	7.3 (SD 8.9; 0 to 35) n = 31	10.7 (SD 9.7; 0 to 24.8) n = 10	8.1 (SD 9.1; 0 to 35) n = 41	0.31

* Continuous variables: independent samples t-test; categorical variables: chi-squared test or Fisher's exact test

Follow-up clinical assessment. At follow-up, the most distal neurological motor level (NML) as described by the International Myelodysplasia Study Protocol, the Hoffer classification for ambulation, and the Sitting Balance Scale (SBS) were determined.[3,13,38] Follow-up NML or SBS scores were not available for remote participants.

Health-related quality of life outcomes. Two HRQOL questionnaires were administered: the Spina Bifida Spine Questionnaire (SBSQ)[39] and the United States English-language Medical Outcome Study 36-item Short Form Health Survey (SF-36v2, QualityMetric, Inc., Lincoln, Rhode Island).[40] Remote patients completed their questionnaires by telephone, a previously used practice for spine-specific conditions.[41-44] The SBSQ is a spina bifida and scoliosis specific HRQOL instrument previously developed by our group and has high test-retest reliability.[2,3,45] The SF-36 survey has high validity in adults with SBC.[20,23,40,46-48] The composite physical component summary (PCS) and mental component summary (MCS) scores were calculated as described by Ware and Sherbourne.[40]

Statistical analysis. Data were analysed by a bio-statistician who used independent samples *t*-tests for continuous variables, and the chi-squared test or Fisher's exact test for categorical variables.[37] Mean differences were calculated for continuous variables with 95% confidence intervals (CI).[37] Linear regression models (Pearson) with R^2 and p-values were used to evaluate relationships between HRQOL and Cobb angles.[37] Statistical significance was set a p-value < 0.05.

Results

There were no peri-operative deaths. Peri-operative complications included two patients with hypotension and one each of respiratory depression, lumbar vertebral body fracture and latex anaphylaxis. Post-operative complications included four patients with leakage of cerebrospinal fluid or shunt externalisations which required repair; seven respiratory or urinary tract infections; two cases each of pneumothorax or pulmonary oedema and delirium; and one case of sepsis.

Peri-operative records for one patient were not available. PFTs followed a restrictive pattern, with reduced mean percent-predicted force vital capacity (%FVC) (53.8, SD 18.6; 28 to 92) and mean percent-predicted forced expiratory volume in one second (%FEV1) (57.7, SD 19.2; 32 to 100) values.[31]

There were 11 (32.4%) surgical site infections. A total of five of the six early infections and two of the five late infections were superficial. Six patients (17.6%) developed a pseudarthrosis and seven (20.6%) needed to have their instrumentation removed, four for infection and three due to its distal prominence.

The mean age of the patients at follow-up was 27.0 years (SD 4.6; 18.8 to 34.7) for patients who had undergone fusion and 25.6 years (SD 5.1; 20.2 to 33.6) for those treated non-operatively. Overall, the mean time to follow-up was 14.1 years (SD 4.3; 6.7 to 21.4). The mean time to follow-up (from fusion to the administration of HRQOL outcomes) was 14.9 years (SD 3.9; 6.7 to 21.4) and 11.6 years (SD 4.7; 6.7 to 20.9) (from the date of being a candidate for surgery to the administration of HRQOL outcomes) for patients treated non-operatively (Table IV).

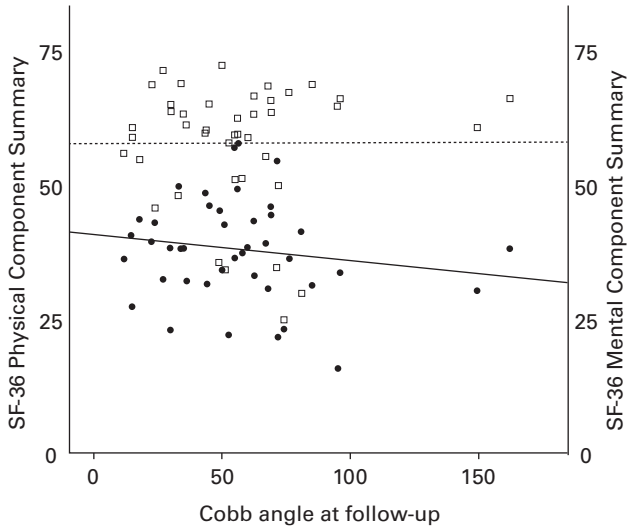

Fig. 1

Follow-up Cobb angles plotted against 36-Item Short Form Health Survey physical component summary (PCS) (filled circles) and mental component summary (MCS) (unfilled squares) scores for all patients (n = 45). Best-fit lines for PCS (solid line) and MCS (dashed line) also depicted.

Table V. Health-related quality of life assessments of the study cohort (Mean values with standard deviations (SD) and range where appropriate)

	Operative n = 34	Non-operative n = 11	Mean difference (MD)	95% CI* of MD - lower	95% CI of MD - upper	p-value†
SF-36						
Physical functioning	26.1 (SD 24.6; 0 to 100)	37.3 (SD 32.7; 0 to 100)	–11.2	–34.2	11.9	0.24
Role-physical	73.5 (SD 34.8; 0 to 100)	90.9 (SD 23.1; 25 to 100)	–17.4	–36.2	1.5	0.13
Bodily pain	79.7 (SD 27.4; 22 to 100)	78.6 (SD 26.1; 20 to 100)	1.1	–18.2	20.4	0.91
General health perception	74.8 (SD 19.0; 25 to 100)	69.0 (SD 22.1; 30 to 100)	5.8	–10.0	21.6	0.40
Energy/vitality	67.7 (SD 19.8; 10 to 95)	73.2 (SD 20.8; 40 to 95)	–5.5	–20.6	9.6	0.43
Social functioning	83.5 (SD 20.3; 25 to 100)	86.4 (SD 19.7; 50 to 100)	–2.9	–17.4	11.6	0.68
Role-emotional	89.2 (SD 24.2; 0 to 100)	85.8 (SD 32.1, 10 to 100)	3.5	–19.2	26.1	0.71
Mental health	78.8 (SD 16.7; 40 to 100)	79.9 (SD 14.8; 44 to 96)	–1.1	–12.2	10.0	0.84
Physical component summary	36.7 (SD 9.1; 15.6 to 57.8)	40.6 (SD 10.3; 25.2 to 56.9)	–3.9	–11.2	3.5	0.24
Mental Component summary	57.8 (SD 10.7; 24.7 to 72.2)	57.3 (SD 13.3; 29.7 to 68.7)	0.5	–9.0	10.0	0.90
Spina bifida scoliosis questionnaire	65.2 (SD 20.7; 20.7 to 97.9)	65.2 (SD 28.5; 14.6 to 98.4)	0.0	–20.0	20.0	0.99

* CI, confidence interval
† Independent samples *t*-test

At follow-up, patients who had undergone fusion had a smaller mean Cobb angle ($p < 0.01$, independent *t*-test) but similar pelvic obliquity, coronal imbalance and CRID to patients who had been treated non-operatively. The Cobb angles at follow-up were not related to SBSQ ($R^2 = 0.04$, $p = 0.21$), PCS ($R^2 = 0.24$, $p = 0.33$) or MCS (R-< 0.01, $p = 0.99$) (Fig. 1). Furthermore, patients treated operatively did not differ in SBSQ or SF-36 PCS and MCS scores from those treated non-operatively (Table V). Similarly, Cobb angles were not related to SF-36 component scores in those treated operatively (PCS: $R^2=0.08$, $p=0.13$, MCS: $R^2 < 0.01$, $p = 0.73$) or non-operatively (PCS: $R^2 = 0.30$, $p = 0.10$, MCS: $R^2 = 0.03$, $p = 0.61$) (Fig. 2).

Subgroup analysis revealed that, irrespective of treatment, patients who could walk had a significantly higher mean PCS score (43.9, SD 9.4; 25.2 to 57.8) than those who could not (35.9, SD 8.7; 15.6 to 54.4), $p = 0.03$ (independent *t*-test); mean difference: 8.0 (95% CI 0.9 to 15.1) (Fig. 3a). Although not statistically significant, non-walkers had a slightly higher mean MCS score (59.6, SD 10.4; 24.7 to 72.2) than those who could walk (50.9, SD 11.7; 29.7 to 63.1), $p = 0.052$ (independent *t*-test); mean difference: 8.8 (95% CI 0.1

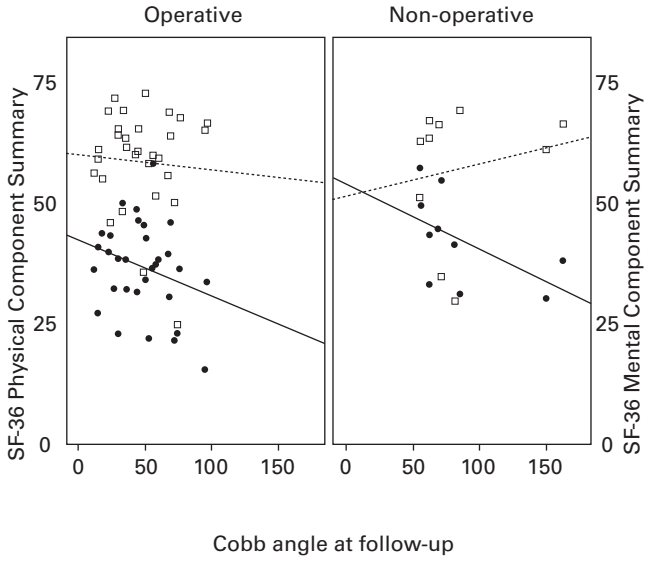

Fig. 2

Follow-up Cobb angles plotted against 36-Item Short Form Health Survey physical component summary (PCS) (filled circles) and mental component summary (MCS) (unfilled squares) scores for operative (n = 34) and non-operative (n = 11) patients. Best-fit lines for PCS (solid line) and MCS (dashed line) also depicted.

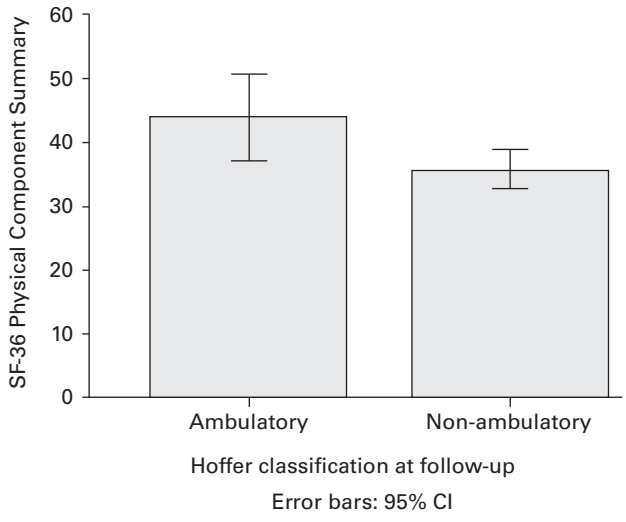

Fig. 3a

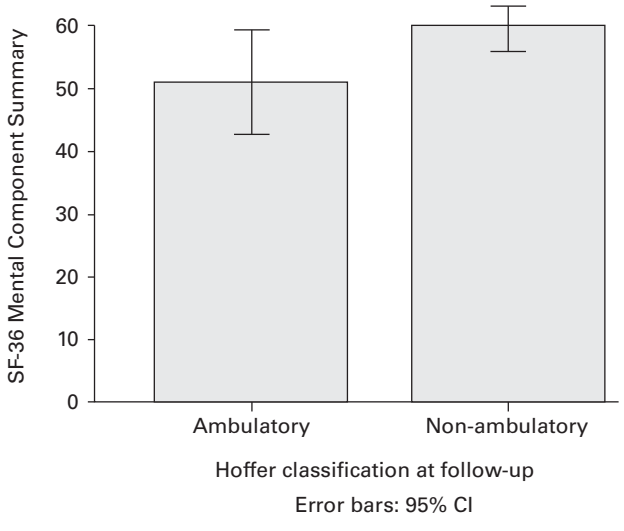

Fig. 3b

Follow-up 36-Item Short Form Health Survey (SF-36) a) physical and b) component summary scores of ambulatory (n = 10) and non-ambulatory patients (n = 35).

to 17.6) (Fig. 3b). There was no difference in mean Cobb angle between the two groups at follow-up (p = 0.39; ambulatory mean 49.4; SD 21.4; 15 to 81 *versus* non-ambulatory mean 58.2; SD 33.4; 12 to 162). A total of five patients who underwent fusion could walk at 'baseline' but were unable to at follow-up. There were no patients who were unable to walk at 'baseline' who could walk at follow-up. Although not significant, the five patients whose ability to walk declined had lower mean PCS scores (31.6, SD 12.0; 15.6 to 48.4) than the other 40 patients who did not decline (38.5, SD 8.9; 21.6 to 57.8) (p = 0.12, independent *t*-test). These five patients had slightly higher mean MCS scores (62.2, SD 4.2; 58.8 to 68.4 *vs* 57.1, SD 11.7; 24.7 to 72.2) (p = 0.35, independent *t*-test).

Discussion

This study adds to the small body of literature about scoliosis in SBC and has the advantage of being the first to evaluate patients with SBC and include patients treated operatively as well as some treated non-operatively, and also include long-term follow-up and quality of life assessment.

In the Adolescents with Spina Bifida in the Netherlands study (ASPINE), 138 adolescents with spina bifida were compared with a control population using SF-36.[23] However, the size of the curve and the surgical history of the participants were not included.[23] Our findings confirm previous studies showing that spinal fusion improves coronal deformity but has an uncertain effect on HRQOL.[12-14,18] Cobb angles did not correlate with HRQOL for all SF-36 and SBSQ scores for either group of patients. These findings support the work of Wai et al[13] who reported no relationship between spinal deformity and self-perception or physical function, and Sibinski et al[3] who reported that the size of the curve was not related to functional outcome.

Spinal fusion has been advocated to prevent the loss of physical function and the ability to walk.[14,18] However, Kahanovitz and Duncan[7] reported that the ability of patients with SBC to walk declined over the 20 years after spinal fusion. Muller et al,[8] in a series of 14 patients, showed that there was no difference in a patient's ability to manage the ADL post-operatively but half lost their ability to walk independently. In our study, 11 patients who underwent spinal fusion could walk before their operation but only six could do so at follow-up. By contrast, the same number of patients treated non-operatively (n = 4) could walk both at the time of initial assessment (baseline) and at follow-up. Schoenmakers et al[15] reported that independent mobility was the most important determinant of HRQOL in patients with SBC. Similarly, Buffart et al[21] found that the ability to walk independently was the main determinant of SF-36 PCS scores. Although surgery may achieve and maintain correction of the scoliosis, the reduction in ability to walk and the increased stiffness after fusion may have a detrimental effect on HRQOL.

Spinal fusion can help sitting balance and correct pelvic obliquity.[12,31] Curves of lesser degree have been associated with lower sitting pressures, although this is not consistently associated with a decrease in the occurrence of pressure induced ulceration.[11,31] Ouellet et al[11] reported that, despite significant curve correction, fusion did not reduce the risk of skin ulceration. In our study, clinical sitting balance did not differ at follow-up between patients treated operatively and non-operatively.

The life expectancy of patients with SBC has increased; consequently, iatrogenic morbidity has become more important.[23] Registry studies have reported complication rates of approximately 17.9% for children undergoing surgery for a neuromuscular scoliosis.[49,50] In our study 32.4% of the patients had a post-operative infection, with many requiring subsequent procedures and revisions.

The unique aspect of our study was the inclusion of patients who had been treated non-operatively. However, the groups were not comparable at the initial assessment. In addition to the concern that the two groups of patients were not matched, our study had other potential limitations. First, we lacked HRQOL measurements at initial assessment and immediately after operation, and were therefore unable to comment on any interval changes. The HRQOL for fused patients may initially have been lower than that of patients treated non-operatively, resulting in selection bias. Second, our sample size was small, especially for the cohort treated non-operatively, resulting in low statistical power.

We incorporated two study centres in an attempt to minimise bias in terms of patient characteristics, surgical technique, surgeon preference and post-operative care. Third, no late outcomes such as PFTs were collected. Therefore, we were unable to assess whether surgery had any long-term influence on pulmonary function. Fourth, our recruitment rate was relatively low but our response rate and reasons for non-participation were consistent with those of other large studies.[21-23]

While the mean difference between the HRQOL scores of patients treated operatively and non-operatively was strikingly small, the 95% CIs were large. In these cases, HRQOL outcome measures across or within groups can be evaluated against the 'minimal important difference' (MID), described as the 'smallest change that is important to patients'.[51,52] Based on the way in which it is constructed, the MID for the SF-36 PCS measure, for various spinal procedures in adults, has ranged from 1.26 to 5.95.[51] In our study, the mean difference in PCS scores was 3.9, which is lower than the upper threshold of the MID. However, given that the large CIs encompass the MID threshold, larger studies are still needed to conclude that HRQOL is not different between the two groups. Thus, although this study cannot be used to conclude that scoliosis surgery is not beneficial for patients with SBC, it is the only study to have long-term results, to use validated HRQOL outcomes, and to evaluate patients treated operatively and non-operatively.

In conclusion, spinal fusion for an SBC scoliosis corrects coronal deformity and halts its progression but has a high complication rate and no clear effect on HRQOL.

We would like to thank Dr K. Doughty and Ms L. Caspi for their assistance throughout this project.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

This article was primary edited by A. C. Ross and first proof edited by G. Scott.

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