Original Article

Increased cautiousness in adolescent idiopathic scoliosis patients concordant with syringomyelia fails to improve overall patient outcomes

ABSTRACT

Background: Adolescent idiopathic scoliosis (AIS) is a common cause of spinal deformity in adolescents. AIS can be associated with certain intraspinal anomalies such as syringomyelia (SM). This study assessed the rate of SM in AIS patients and compared trends in surgical approach and postoperative outcomes in AIS patients with and without SM.

Methods: The database was queried using ICD-9 codes for AIS patients from 2003–2012 (737.1–3, 737.39, 737.8, 737.85, and 756.1) and SM (336.0). The patients were separated into two groups: AIS-SM and AIS-N. Groups were compared using *t*-tests and Chi-squared tests for categorical and discrete variables, respectively.

Results: Totally 77,183 AIS patients were included in the study (15.2 years, 64% F): 821 (1.2%) – AIS-SM (13.7 years, 58% F) and 76,362 – AIS-N (15.2 years, 64% F). The incidence of SM increased from 2003–2012 (0.9 to 1.2%, $P = 0.036$). AIS-SM had higher comorbidity rates (79 vs. 56%, $P < 0.001$). Comorbidities were assessed between AIS-SM and AIS-N, demonstrating significantly more neurological and pulmonary in AIS-SM patients. 41.2% of the patients were operative, 48% of AIS-SM, compared to 41.6% AIS-N. AIS-SM had fewer surgeries with fusion (anterior or posterior) and interbody device placement. AIS-SM patients had lower invasiveness scores (2.72 vs. 3.02, $P = 0.049$) and less LOS (5.0 vs. 6.1 days, $P = 0.001$). AIS-SM patients underwent more routine discharges (92.7 vs. 90.9%). AIS-SM had more nervous system complications, including hemiplegia and paraplegia, brain compression, hydrocephalous and cerebrovascular complications, all $P < 0.001$. After controlling for respiratory, renal, cardiovascular, and musculoskeletal comorbidities, invasiveness score remained lower for AIS-SM patients ($P < 0.001$).

Conclusions: These results indicate that patients concordant with AIS and SM may be treated more cautiously (lower invasiveness score and less fusions) than those without SM.

Keywords: Adolescent, deformity, idiopathic, scoliosis, syringomyelia

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INTRODUCTION

Adolescent idiopathic scoliosis (AIS) refers to a three-dimensional alteration of the normal curvature of the spine arising around puberty in otherwise typical children.[1,2] Approximately 3% of children under the age of 16 years are diagnosed with AIS, some of which may be based on genetic contribution, otherwise the etiology has yet to be clearly elucidated.[3] AIS is a difficult condition to treat, as the patients and their spinal curvatures are continuing to grow and change following the time of diagnosis. Therefore,

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bracing is the primary avenue for skeletally immature patients with Cobb angles <20°, along with periodic observation through serial radiographs.[4] However, bracing is not suggested if the patient's Cobb angle is severely pronounced, or if they present with rapid curve progression in follow-up. In these cases, surgical intervention often becomes the primary recommended treatment.

In an effort to preserve the segmental range of motion for AIS surgical patients, metrics such as the Lenke classification have been proposed to facilitate the operative selection of the proper upper and lower instrumented vertebra.[5,6] Nevertheless, planning of AIS procedures is inherently complicated by the nature of the disease, but idiopathic scoliosis is often not the patient's sole morbidity. Surgical planning for AIS patients becomes more nuanced with concurrent intraspinal or extraspinal conditions to additionally take into account. For example, Chiari malformations, or the extension of brain tissue into the spinal canal, and syringomyelia (SM), the development of a fluid-filled cyst in the spinal cord, are serious intraspinal abnormalities that may occur in conjunction with AIS. Do *et al*. and Swarup *et al*. demonstrated concomitant intraspinal anomaly and AIS rates for Chiari malformations as 1.2% to 4.2%, respectively, and SM from 0.6% to 5%, respectively.[7,8] Regardless, there exists limited research concerning postoperative outcomes of AIS corrective procedures for patients concurrently affected by SM. Godzik *et al*. analyzed a small cohort of AIS patients concomitantly affected by Chiari Type 1 malformation (CIM) and SM from a single institution.[9] They discovered that while alignment correction and health-related quality of life scores from fusion procedures were similar between AIS patients with and without CIM and SM, those with all three conditions experienced higher rates of neuromonitoring challenges and postoperative complications.

The studies related to intraspinal anomalies in the setting of AIS corrective surgery are generally from single centers with small patient cohorts, making generalizable conclusions ultimately challenging. The benefit of large dataset analyses lies in the volume of patients and ability to examine epidemiological trends. However, there are also limitations to the use of national databases, as they rely upon ICD-9 codes, which may result in clerical errors and lack of follow-up. Nevertheless, careful and diligent selection of relevant ICD-9 codes has proved insightful for past studies to explore diagnosis and surgical trends in a large population.[10-12] Therefore, this study was designed to examine the largest national pediatric inpatient database to assess the incidence of concomitant AIS and SM and compare surgical characteristics and postoperative outcomes between AIS-SM and AIS patients without SM (AIS-N).

METHODS

Study design and data source

The Kids' Inpatient Database (KID) is a part of the Healthcare Cost and Utilization Project, and stands as the leading source for all-payer pediatric (<21 years at admission) inpatient hospital visits in the United States. It contains data for approximately 3 million discharges a year. The study is exempt from the International Review Board approval, with preservation of patient confidentiality due to the lack of state and hospital identifiers. Further information regarding the KID initiative can be found at: https://www.hcup-us.ahrq.gov/kidoverview.jsp. Each institution obtained approval from their local IRB, and patient consent was not required. Patients from 2003–2013 were retrospectively analyzed in the KID database for the present study. Data source and statistics are similar to a previous study (Segreto *et al*., 2019).

Inclusion criteria

The database was queried for ICD-9 codes of operative and nonoperative patient discharges pertaining to the diagnosis of AIS (737.1–3, 737.39, 737.8, 737.85, and 756.1). The database was further queried for patients AIS who also had SM (336.0). Patients with an age of < 21 years were included in the analysis. AIS patients were stratified into two groups: those with concurrent SM (AIS-SM) and those without SM (AIS-N).

Data collection and outcome measures

Demographics consisted of age, sex, and race. Comorbidity severity was quantified utilizing the Deyo adaption of the Charlson Comorbidity Index for administrative databases relying on ICD-9-CM diagnosis and procedural codes. Invasiveness scores (developed by Mirza *et al*.) were assessed between the comorbidity types based on body system (neurologic, musculoskeletal, pulmonary, cardiovascular, and renal), individually specified comorbidities, surgical details (approach, construct length, and techniques), perioperative inpatient complications, inpatient length of stay, mortality, and discharge destination, which were isolated utilizing ICD-9-CM diagnosis codes, procedure codes, and preestablished available data elements within the KID database.

Statistical analysis

National estimates for annual AIS hospitalization incidence were quantified using KID-weighted discharges. Descriptive, Chi-square, and independent sample *t*-test analyses assessed frequencies and means of demographic variables among AIS-SM and AIS-N patients. One-way ANOVA ascertained significant variation for continuous variables. All tests were two-sided, and significance was set to a *P* value of less than 0.05. All statistical analyses were performed utilizing

IBM Statistical Package for the Social Sciences (SPSS) version 23.0 132 (Armonk, NY, USA: IBM).

RESULTS

Study sample

Totally 77,183 AIS patients met inclusion criteria. The Incidence of AIS was 613.97 patients per 100,000 annual discharges. The mean patient age for AIS patients overall was 15.2 years, with 64% females. Eight hundred and twenty-one AIS patients (1.1%) had concomitant SM, with a slightly younger mean patient age 13.7 years and 58% females. The incidence of AIS-SM per 100,000 patients was 6.53. AIS-SM patients were significantly younger ($P < 0.001$) and had less females ($P < 0.001$). White patients comprised a significantly higher proportion of patients with concomitant AIS and SM, while patients without SM had a greater incidence in the Hispanic and Black populations ($P < 0.001$). The incidence of SM in the AIS population increased from 2003 to 2012, 0.9% in 2003 to 1.2% in 2012 ($P = 0.036$) [Table 1].

Comorbidity severity, quantification, and clustering

The mean overall patient Deyo-Charlson score was 0.842, where AIS-N patients presented with significantly greater scores 0.846 compared to AIS-SM patients, 0.478 ($P < 0.001$). By body system, AIS-N presented with 36% neurological, 45.3% pulmonary, 4.1% renal, 8%, cardiovascular, and 5.2% musculoskeletal comorbidities. In comparison, AIS-SM presented with 71.2% neurological, 74% pulmonary, 2% renal, 5.4% cardiovascular, and 5% musculoskeletal comorbidities. AIS-SM had higher overall comorbidity rates (79% vs. 56%, $P < 0.001$). The top body system comorbidity associations included concurrent pulmonary/neurologic comorbidities for both AIS N (31%) and AIS SM (68%) patients ($P < 0.001$) [Table 2].

Surgical details for operative patients

41.2% of the patients were operative, 48% of AIS-SM, compared to 41.6% AIS-N. AIS-SM patients had fewer surgeries with posterior fusion (AIS-SM: 24.5%, AIS-N: 32.7%, $P < 0.001$), anterior fusion (AIS-SM: 1.7%, AIS-N: 2.7%, $P = 0.077$), any fusion (AIS-SM: 29%, AIS-N: 39.1%, $P < 0.001$), and interbody device placement (AIS-SM: 2.8%, AIS-N: 5.7%, $P < 0.001$) than AIS-N patients. AIS-SM patients had lower mean surgical invasiveness score (AIS-SM: 2.72, AIS-N: 3.02, $P = 0.049$) and less LOS (AIS-SM: 5.0, AIS-N: 6.1 days, $P = 0.001$). After controlling for respiratory, renal, cardiovascular, and musculoskeletal comorbidities, invasiveness score remained significantly lower for AIS-SM patients ($P < 0.001$).

Perioperative outcomes

When assessing postoperative outcomes, AIS-SM patients underwent more routine discharges (AIS-SM: 92.7%, AIS-N: 90.9%). AIS-SM had more nervous system complications (AIS-SM: 2.3%, AIS-N: 0.7%), including hemiplegia (AIS-SM: 1.5%, AIS-N: 0.4%), paraplegia (AIS-SM: 2.4%, AIS-N: 0.9%), brain compression (AIS-SM: 32.1%, AIS-N: 0.6%), hydrocephalous (AIS-SM: 7.7%, AIS-N: 1.3%), and cerebrovascular complications (AIS-SM: 0.87%, AIS-N: 0.26%), all $P < 0.001$.

DISCUSSION

The findings of this study indicate that patients with concordant AIS and SM may ultimately be treated earlier (younger at time of diagnosis) and more cautiously (lower invasiveness scores and fewer fusion procedures) compared to AIS patients without SM. However, despite the guarded approach, AIS-SM patients experienced increased documented adverse events, especially neurological complications. Consequently, we proposed that the added precaution taken in cases of AIS-SM seems to fail to improve patient outcomes.

These findings are, in part, a contradiction to current research into the surgical outcomes experienced by AIS-SM patients. Godzik *et al*. noted that patients with concurrent SM and

Table 1: Demographics between adolescent idiopathic scoliosis patients with and without syringomyelia as well as the overall mean of within the adolescent idiopathic scoliosis cohort

	AIS patients without SM (%)	AIS patients with SM (%)	*P*	Overall mean (%)
Sample size (*n*)	76,362	821	-	77,183
Age (years)	15.2	13.7	<0.001	15.2
Sex (female)	64	58	<0.001	64
Race				
White	50.7	60.9	<0.001	50.8
Hispanic	12.5	7.94	<0.001	12.45
Black	12.8	8.4	<0.001	12.8
Other	0.42	0.65	0.311	0.42
Asian	1.74	2.57	0.073	1.75

AIS - Adolescent idiopathic scoliosis, SM - Syringomyelia

Table 2: Comorbidity information between adolescent idiopathic scoliosis patients with and without syringomyelia

	AIS patients without SM (%)	AIS patients with SM (%)	*P*
Comorbidities			
Overall	79	56	<0.001
Neurological	36	71.2	<0.001
Pulmonary	45.3	74	<0.001
Renal	4.1	2	0.005
Cardiovascular	8	5.4	0.007
Musculoskeletal	5.2	5	0.731
Comorbidity associations			
Pulmonary/neurologic	31	68	<0.001
Renal/cardiovascular	0.52	0.18	0.171
Muscular/cardiovascular	0.69	0.20	0.087
Deyo-Charlson score	0.846	0.478	<0.001

AIS - Adolescent idiopathic scoliosis, SM - Syringomyelia

Chiari malformations did experience more neurological complications compared to those without comorbid AIS, but comparable alignment corrections, suggesting similar patterns of invasiveness.[9] Further, Li *et al*. noted that when comparing surgical outcomes for patients with idiopathic scoliosis versus SM-associated scoliosis, both groups experienced similar patient-reported scores as well as complication rates but demonstrated global alignment differences.[13]

While prior studies have varying postoperative conclusions, confounding variables need to be taken into consideration. The limited pools of outcome studies on AIS-SM patients are from single institutions with limited patient cohorts of less than 100 patients. Other studies have explored intraspinal complications including SM, but in the context of additional spinal conditions such as tethered cord syndrome or Chiari malformations, as well as without control cases.[9,14,15] It is possible that the conclusions of a small group of patients from single institutions are not generalizable, despite the fact that prospective randomized trials are considered the gold standard in orthopedic research.[16] In addition, there has been a dramatic increase in the amount of orthopedic and neurosurgery journals publishing studies based in national health-care datasets.[17] However, large datasets come with their own pro et contra as well, which have been well documented and discussed at length.[12]

The postoperative outcomes for an AIS-SM patient are not consistently comparable to AIS controls, nor are their outcomes consistently inferior. In the present study, AIS-SM patients were at a younger age at diagnosis with decreased procedural invasiveness but ultimately presented with increased neurologic complications. Although, additional factors need to be accounted for in future research that has not been incorporated or directly addressed in this study nor previous studies of concordant AIS and SM. For instance, Samdani *et al*. noted that patients with syrinxes greater than 4 mm frequently had more levels fused, a greater EBL, and less curve correction.[18] Incorporation of the SM syrinx size had not been noted in the studies mentioned previously, nor did the KID database allow for quantifying patients in this study. Further, the plans made for surgical interventions seem to be influenced by the perspective of a scoliosis patient with concordant SM as opposed to a patient with SM who is affected by scoliosis.[19] Therefore, there are most likely minor details about cases that are not well captured by looking at billing codes. There are limitations to the bold conclusion from the present study that extra precaution taken on AIS patients with concomitant SM fails to improve patient outcomes, as we do not know the curve magnitude or progression of the patients. Still, surgeons could benefit from keeping in mind the trends noted in the paper, as well as the results from prior research in order to best optimize their surgical planning for patients.

CONCLUSIONS

These results indicate that patients concordant with AIS and SM may be treated more cautiously (lower invasiveness score and less fusions) than those without SM. Despite the guarded approach, AIS patients with SM documented more adverse events, especially neurological complications. This, overall, signifies that the extra precaution taken in these adolescents fails to improve patient outcomes.

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Conflicts of interest

There are no conflicts of interest.

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