Determinants of Spinal Bracing Success in Mitigating Spine Deformity Progression in Spinal Muscular Atrophy (SMA)

Hiroko Matsumoto[1,2], Yuri Ota[1], Joyce Huang[1], Timothy Hresko[1,2], Ron El-Hawary[1,2], Grant Hogue[1,2], Craig Birch[1,2], John Emans[1,2], Dennis Anderson[2,3], Ryan Murdock[4], Brian Snyder[1,2]

[1]Boston Children's Hospital, Boston, MA, [2]Harvard University, Cambridge, MA, [3]Beth Israel Deaconess Medical Center, Boston, MA, [4]Thinks Works, PBC, Chicago, IL

Email: Hiroko.Matsumoto@childrens.harvard.edu

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INTRODUCTION: Neuromuscular spinal deformity (NMSD), presenting as scoliosis projected in the coronal plane and kyphosis projected in the sagittal plane, is prevalent in ~90% of children afflicted by SMA. The spine deformity is often accompanied by pelvic obliquity and rib cage distortion that contribute to physical disability, cardiopulmonary compromise, reduced health-related quality of life, and early mortality[1]. While spinal instrumentation effectively improves spine deformity, it requires repeated surgeries in growing children that can be associated with complications prolonging hospitalization and recovery[2]. Spinal bracing provides a non-operative treatment for spinal deformity that may delay or reduce the need for surgery by superimposing asymmetric corrective forces and bending moments on the dynamic forces and moments that the spine and axial skeleton are typically exposed during daily activities to induce correction of the spine deformity through viscoelastic CREEP and modulation of spinal growth. While bracing has been proven to mitigate adolescent idiopathic scoliosis[3], its role in managing NMSD remains unclear. In growing children with NMSD, bracing may defer surgical interventions until the child is bigger, potentially improving safety and reducing the cumulative burden of repeated operations for these fragile children and their families. Previous NMSD brace studies raised concerns that bracing could exacerbate rib deformities contributing to thoracic insufficiency, restrict thoracic and diaphragmatic inspiratory motion leading to dyspnea, compress the abdomen provoking gastro-esophageal reflux, irritate skin, aggravate overheating, and cause discomfort[4,5]. However, these prior investigations were limited by poor methodology and the use of outdated brace constructs that modern 3D thoraco-lumbar-sacral-orthosis (TLSO) may overcome. This retrospective clinical study evaluates the efficacy a 3D-TLSO to moderate NMSD progression in SMA children depending on the effectiveness of the brace to reduce deformity magnitude and the ability of the patient to wear the brace for sufficient time to modulate deformity progression.

METHODS: This single-center, IRB approved, retrospective study included SMA patients with NMSD, ≤18 years old, treated with a patient specific 3D-TLSO (Fig. 1). Major deformity magnitude, in-brace correction and rib deformity (Parasol Score ≤0.56[6]) were assessed on serial sitting spine X-rays in & out of TLSO at baseline, 1-year and 2-year follow-up. Brace use compliance (daily hours of wear) was self- or proxy reported. Treatment was considered successful if a child <10 years old, remained surgery-free and/or maintained NMSD <50° until adolescence; for an adolescent (≥10 years old), NMSD progression was limited to ≤10°/year over 2-years. Predictors of success were analyzed using multiple regression models.

RESULTS: 30 SMA patients (16 males, 14 females) were evaluated; 57% had 2 *SMN*2 gene copies, 37% had 3 *SMN*2 gene copies, and 6% had 4 *SMN*2 gene copies. All patients received disease-modifying therapies at a mean age of 2.4±3.4 years (range 0–13): 77% received Nusinersen, 23% received Onasemnogene Abeparvovec. At brace initiation, 11% of patients could walk independently, 63% could sit independently, and 26% were non-ambulatory. To treat an average scoliosis of 54°±16°, 3D-TLSO was initiated at a mean age of 5.9±3.9 years, achieving a mean in-brace curve magnitude reduction of 24% ± 22% (mean in-brace residual scoliosis 44°±18°). Significant parasol rib deformity was observed in 42% of patients prior to bracing; one patient improved, while the rib deformity did not change with bracing. NMSD progressed in 17 patients (57%) over 2-years. After adjusting for initial curve magnitude, treatment success required ≥25% in-brace deformity correction (Fig 2.). 75% of patients who wore the brace ≥7 hours/day achieved success, compared to 38% who wore the brace <7 hours/day (Fig 3). Treatment success was not associated with age at brace initiation, sex, *SMN2* copy number, or functional status.

DISCUSSION: This preliminary study of SMA patients with NMSD indicates that a 3D TLSO can delay NMSD progression, without exacerbating rib deformity, provided that the TLSO reduces the deformity by ≥25% and the patient wears the TLSO ≥7 hours/day consistently during waking hours, which is important since many SMA patients cannot tolerate nighttime brace wear due to nocturnal non-invasive ventilatory support and/or G-tube feeding.

SIGNIFICANCE/CLINICAL RELEVANCE: This evidence-based retrospective study demonstrates that a 3D TLSO can delay NMSD progression in SMA children and adolescents without exacerbating rib deformity, provided that the TLSO effectively reduces the spine deformity by ≥25% and the patient wears the TLSO consistently ≥7 hours/day during waking hours.

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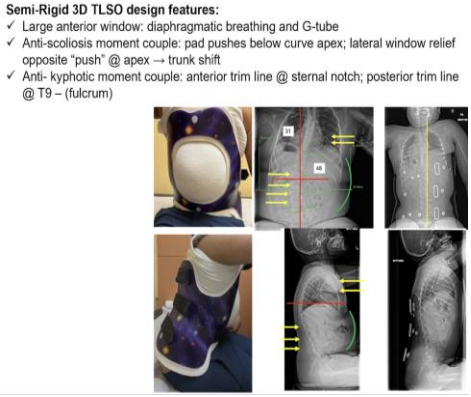

Fig. 1. Salient design features posterior opening TLSO

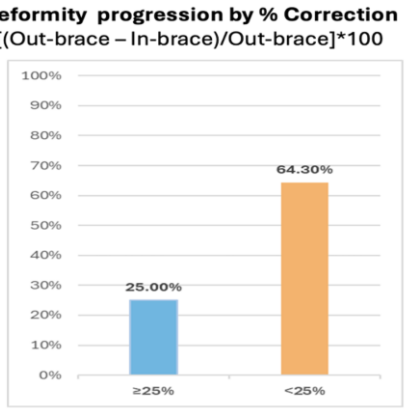

Fig. 2. Incidence of deformity progression based on in brace correction ≥25% vs. >25%

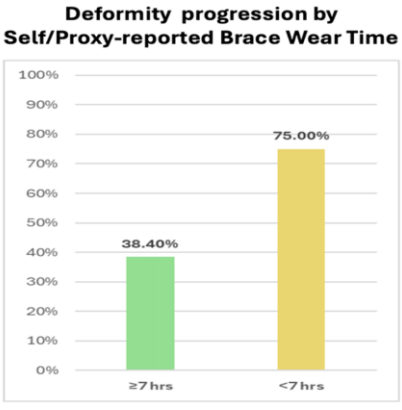

Fig. 3. Incidence of deformity progression based on brace wear compliance ≥7 hours vs <7 hours