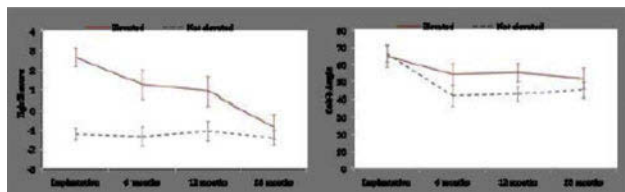


of elevated HgB in patients with EOS who require surgery and 2) quantify the response of HgB levels to treatment.

Methods: HgB laboratory values were prospectively collected in a multicenter database prior to initial implantation and following surgery at 6, 12, and 18 months. Because normal HgB values vary with age, HgB values were converted to Z scores, calculated by dividing age-adjusted mean HgB levels by the age-adjusted standard deviation. Elevated HgB was defined by a Z-score > 1. Change in HgB Z-score and Cobb angle over time were assessed using piecewise linear mixed modeling.

Results: 160 EOS patients were enrolled, comprising GR (12%), MCGR (33%), and VEPTR (54%) treatment. The average age at implantation was 6.7 years. The prevalence of elevated HgB (Z > 1) was 16%. HgB Z-scores in elevated HgB patients decreased preoperative to 6 months ($p < 0.001$), showed no change 6 to 12 months ($p = 0.73$), and decreased 12 to 18 months ($p = 0.01$) for an overall decrease of -3.7 (95% CI = [-6.9 to -0.6]). Patients with non-elevated HgB preoperative showed no postoperative HgB change over time ($p = 0.40$). Patients with elevated HgB showed less average Cobb angle correction with initial implantation than non-elevated, but showed similar average Cobb angle at 18 months.

Conclusions: Patients with elevated HgB before surgery respond differently to VEPTR and GR implantation than those with non-elevated HgB, but appear to eventually have similar results.



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Paper #27

Improvement of Pulmonary Function Measured by Patient-Reported Outcomes in Patients with Spinal Muscular Atrophy After VEPTR Surgery

Michael G. Vitale, Hiroko Matsumoto, John D. Mueller, Patrick J. Cahill, Peter F. Sturm, David P. Roye, John T. Smith

Summary: Quality of Life and PFT values from pediatric Spinal Muscular Atrophy (SMA) patients post-VEPTR implantation were evaluated. There was improvement in pulmonary function assessed by patient-reported outcomes measured at 1 year and 2 years postoperatively.

Hypothesis: Surgical intervention using VEPTR will improve pulmonary function in SMA patients.

Design: Retrospective cohort study.

Introduction: Surgical intervention with constructs such as the Vertical Expandable Prosthetic Titanium Rib (VEPTR) can halve the rate of decline of pulmonary function and allow lung growth. While pulmonary function has traditionally been assessed via PFTs, disease severity and young age in a number of patients precludes them from taking PFTs. This study aimed to demonstrate that the Early Onset Scoliosis Questionnaire (EOSQ-24) PF domain could be used to evaluate pulmonary function changes in SMA patients after VEPTR implantation.

Methods: We conducted a multi-center retrospective cohort study and queried a national Early Onset Scoliosis registry for patients with SMA

operated on between 2005 and 2015. Patients diagnosed with SMA and treated with VEPTR implantation were eligible for our study. Pre-operative and post-operative EOSQ24 pulmonary function domain scores at 1-year and 2-year were collected and PFT results measured by forced vital capacity (FVC) were also assessed.

Results: We identified 49 patients meeting inclusion criteria (averages: Cobb=58.2[deg]; 55%F; 6.9 years at implant). Average initial Cobb correction was 19.5[deg]. EOSQ24 pulmonary function scores increased significantly from 60.71 pre-operatively to 90.0 ($p = 0.039$) at one year post-operatively and to 86.31 ($p = 0.057$) at two years. There was no significant difference observed between pre-operative and post-operative FVC values.

Conclusions: Pulmonary function in SMA patients measured by patient-reported outcomes demonstrated significant improvement after VEPTR surgery. Although it is difficult to objectively measure pulmonary function, quality of life may be a good indicator for well-being and satisfaction toward pulmonary function.

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Paper #28

Quality of Life and Burden of Care in Patients with EOS Undergoing Casting



Emily Auran, Hiroko Matsumoto, David P. Roye, Michael G. Vitale, Peter F. Sturm, James O. Sanders, Matthew Oetgen, Sumeet Garg, Children's Spine Study Group/Growing Spine Study Group

Summary: Serial body casting plays an important role in the treatment of early onset scoliosis (EOS), serving as an effective, safer method compared to surgical intervention. The health-related quality of life (HRQoL) of patients with EOS of idiopathic etiology begins within a normal range, but declines while in cast and remains lower after the casting treatment is complete. While patients with non-idiopathic EOS begin with a lower HRQoL, their in-cast deterioration is smaller and recovers after casting is completed.

Hypothesis: HRQoL declines and burden of care increases during casting treatment and is restored post-casting for patients with both idiopathic and non-idiopathic EOS.

Design: Retrospective cohort.

Introduction: The treatment of children with EOS is controversial and evolving. Serial body casting plays an important role as a safer method compared to surgical intervention in correcting spinal deformity. It is thus critical to discern the impact of casting on patients and their caregivers. The purpose of this study is to compare HRQoL of patients with EOS and their caregivers' burden of care before, during, and after casting.

Methods: 59 EOS patients were identified from 2 multi-center databases. Mean index casting age was 2.2[plusmn]1.2y; mean Cobb pre-cast was 52[plusmn]21[deg]. 25 had non-idiopathic and 34 idiopathic EOS. HRQoL and burden of care were assessed by the EOSQ24. Child and Parent Satisfaction were also assessed. EOSQ scores were compared pre-, during, and post-casting (in or post brace). Patient scores were compared to those of age-matched healthy norms.

Results: In all EOSQ domains, non-idiopathic patients had lower scores than idiopathic for all time periods. Pre-cast, idiopathic scores were equivalent to age-matched norms except in burden domains, while non-idiopathic's were consistently lower. In cast, non-idiopathic patients declined in Transfer, Emotion and Child Satisfaction ($p < 0.01$), while idiopathic patients declined in those 3 plus Physical Function, Daily Living, Overall HRQoL and Burden ($p < 0.01$). In-cast scores for both groups were significantly lower than norms. In brace, non-idiopathic scores slightly increased while idiopathic scores did not change. Post-brace,