Does the Law of Diminishing Returns Exist in EOS with Connective Tissue Disorders?

Disclosures

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Introduction

- Growth friendly rods (GFR) are effective in growing spinal lengths in deformity
- Law of diminishing returns (LoDR) is defined as a decrease in gain of coronal height over time
- Sankar et al. demonstrated the presence of LoDR in all-cause early onset scoliosis
Little is known about whether this same phenomenon occurs in patients with lax connective tissue disorders (CTD) – Marfan – Ehlers Danlos (EDS) – Loeys Dietz (LDS).
Hypothesis

• Connective tissue properties may modify this phenomenon

• Patients with CTD undergoing Growth Friendly (GF) surgery may not have evidence of diminishing returns.
Methods

• Retrospective review of prospective registry

• Inclusion criteria
  – Patients with CTD
  – <10 years old at initial surgery
  – Minimum 5 year follow up
  – Distal and at least 1 proximal spine anchor
  – No self growing techniques
Methods

- **21 patients** met inclusion criteria
  - 17 (81%) Marfan syndrome
  - 1 (5%) Ehlers Danlos Syndrome
  - 2 (10%) Loeys Dietz syndrome
  - 1 (5%) uncharacterized CTD
Patient Characteristics

- Mean follow-up was **8.4 years**
- Mean preoperative **Major Curve 77° ± 21°**
- 7 had **definitive fusion**, 7 had **implants retained** and 7 with continuing **growing rod distraction**
- Mean age at **final follow-up (definitive fusion) = 12.5 years**
Patient Characteristics

- Mean age at initial implantation was 4.9 ± 2.9 years
- Mean T1- T12 at first preop visits was 175 ± 33mm
- Total mean improvement in coronal height at final follow up, including implantation, was 102 ± 48mm
Methods

• Radiographic parameters were assessed at pre-index surgery, post index and prior to every lengthening
  – T1 - S1 Length
  – T1 - T12 Length
  – Major curve angle
  – Kyphosis
Methods

• Outcomes of interest:

  – Difference in mean height gain during early- set distractions vs late- set distractions
    • Early set distractions defined as: D1 - D6 and Late set distractions as D7 – D10

  – T1 – S1 height gain per year (mm/year)
    • Example gain per year between 5th and 4th lengthening:
      \[
      \frac{[\text{PreL5 (mm)} - \text{preL4 (mm)} \times 365]}{\text{Days between 5th and 4th lengthenings}}
      \]
Methods

• Paired sample t-test was performed
  – To determine difference in coronal height gained from early set distraction and late set distractions.
    • Only patients with radiographic data beyond 6 lengthenings were included in the analysis

• Analysis of variance (ANOVA) was performed
  – To determine difference in gain per year after each subsequent lengthening
Results

• Among patients with greater than 6 lengthenings:
  – The mean height change in late set distraction was 12mm greater than the mean height change in early set distraction (p=0.304)
Results

• When normalized for time, there was no significant difference in net gain per year at different lengthening time points (p=0.59)
Results

• 14 (67%) of patients had >22cm T1 – T12 lengths at final follow up
Limitations

- Retrospective nature
- Sample size
- Sagittal plane not considered
Conclusions

- Our results demonstrate that the LoDR is not see among patients with EOS and CTD.
- These results have implications for timing of GR surgery among patients with CTD.
Thank You !