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TREATMENT OF KYPHOSCOLIOSIS AND TIS ASSOCIATED WITH MYELODYSPLASIA USING THE VEPTR IN AN EIFFEL TOWER CONSTRUCT

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Disclosures

A. Joshi, MD	Celling Technologies
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Introduction

- Thoracic Insufficiency Syndrome (TIS) is a source of morbidity and mortality in children with spinal and thoracic deformities
- VEPTR has been a successful treatment for TIS associated with spinal and thoracic deformities
- We analyzed the effect of VEPTR treatment on spinal, thoracic, and pelvic deformities and TIS in children with myelomeningocele

Background

- Orthopedic management of myelomeningocele
  - Correct spinal, thoracic, and pelvic deformities
  - Improve respiratory function
  - Maintain spinal growth
- Surgical management of spinal deformities in myelomeningocele is challenging
  - High infection rates
  - Poor soft tissue coverage
  - High hardware failure rates
- Myelomeningocele can result in both primary and secondary TIS

Secondary TIS

- Thoracic Insufficiency Syndrome
  - The inability of the thorax to support normal respiration or lung growth
- Secondary TIS
  - upward pressure of the abdominal contents against the diaphragm, interfering with normal respiration and lung growth
- It occurs in myelomeningocele due to:
  - Lack of active lumbar extension
  - Severe pelvic obliquity
- The marionette sign is a clinical manifestation of this phenomenon

Objectives

1. To describe an operative technique using VEPTR for children with myelomeningocele
2. To assess the effects of VEPTR on the spinal, thoracic, and pelvic deformities
3. To measure changes in respiratory status
4. To measure response of TIS to VEPTR treatment
5. To identify complications associated with the use of VEPTR in children with myelomeningocele

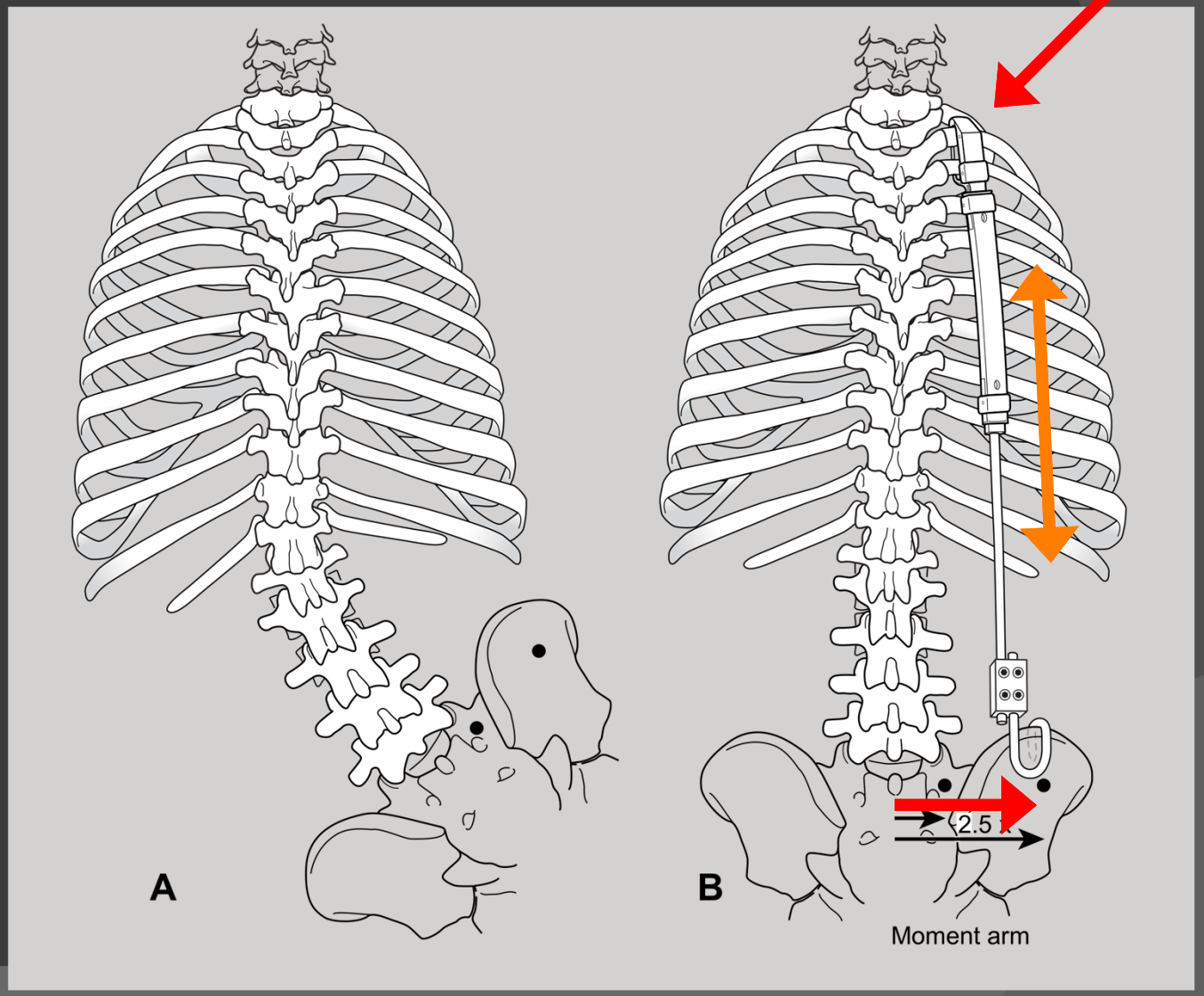
Materials & Methods

- Retrospective study
- Study Criteria
  - Inclusion:
    - Diagnosis of myelomeningocele with kyphoscoliosis
    - Selection for treatment using VEPTR instrumentation
    - Minimum 2 years follow-up
  - Exclusion: prior spinal or thoracic surgeries
- Patient selection
  - 10 patients met the inclusion criteria; none were excluded
  - 6 males; 4 females
  - Age at first surgery: 7.7 years (range, 1.2-14.1)
  - Length of follow-up: 6.2 years (range, 2.3-12.1)

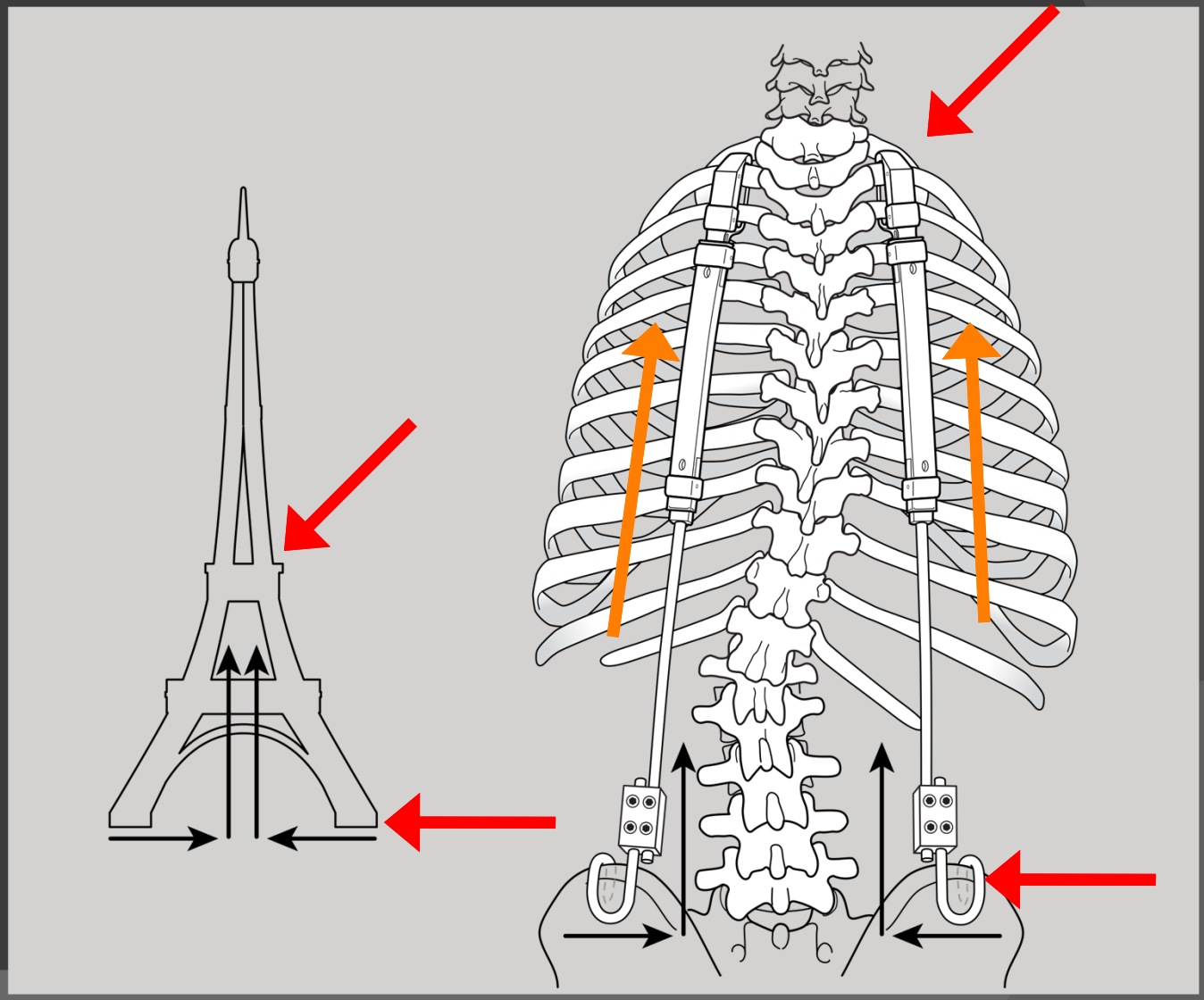
Materials & Methods

- Chart review
  - Demographic data
  - Operative technique and frequency
  - Respiratory status: Assisted Ventilatory Rating (AVR), respiratory rate, capillary blood gases, marionette sign
  - Complications
- Radiographic Assessment
  - Cobb angle
  - Lumbar kyphosis
  - Pelvic obliquity
  - Space Available for Lungs (SAL)
- Data analyzed using paired student *t*-test and Mann-Whitney U test

Operative Technique: Eiffel Tower Construct



Operative Technique: Eiffel Tower Construct



Results

- Operative Technique
  - Implantation
    - 8 unilateral rib-to-pelvis device
    - 2 staged bilateral rib-to-pelvis devices
    - All received lateral rib-to-rib devices (6 bilateral; 4 unilateral)
  - Replacements
    - Mean of 2.5 surgeries (25 total)
    - 2 for migration; 3 for DM hook fracture; 2 for infection
  - Expansions
    - Mean of 7.8 surgeries (range, 3-15)

Results

- Spinal, Thoracic, & Pelvic Alignment

	n	**Pre-Implant**	**Final F/u**	***p*-value***
Cobb Angle	10	70.8° ±18.4°	46.9° ±17.2°	0.004
Lumbar Kyphosis	6	42.5° ±13.7°	22.3° ±18.9°	0.07
Pelvic Obliquity	10	24.7° ±11.6°	8.6° ±6.6°	0.009
SAL	10	0.659 ±0.100	0.880 ±0.057	0.002
* paired student *t*-test				

Results

- Respiratory Status

	n	Pre-Implant	Final F/u	*p*-value
AVR	10	8 unchanged (AVR 0) 1 improved (AVR 1 to AVR 0) 1 declined (AVR 0 to AVR 1)		>0.05*
Respiratory Rate	10	29.3 ±5.4	24.0 ±1.7	n/a
CBG: pCO_2	10	34.5 ±2.9	38.8 ±2.1	0.01**
Marionette Sign	10	6 positive	0 positive	n/a
AVR: (0=no assistance; 4=full-time ventilator dependence) * Mann-Whitney U test ** paired student *t*-test				

Results

- Complications
  - Device migrations: 6 (in 4 patients)
    - 2 superior cradles
    - 2 D-M hooks
    - 2 inferior cradles
  - Hardware fatigue fractures: 3 (in 3 patients)
    - all D-M hook fractures
  - Wound Infections: 4 (in 4 patients)
    - 3 superficial
    - 1 deep
    - all successfully treated

Conclusions

- VEPTR treatment is a useful technique for addressing primary and secondary TIS in myelomeningocele
- Spinal, thoracic and pelvic deformity correction was observed
- The ‘Eiffel Tower’ construct using bilateral or unilateral rib-to-pelvis hybrid devices with wide base and narrow apex might be most effective for correcting lumbar kyphosis and pelvic obliquity
- Infections were surgically treatable with implant salvage and acceptable morbidity

Thank you