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Disclosures:

Consultant:

- Medtronics
- Depuy/Synthes Spine
- Royalties
 - Depuy/Synthes Spine (VEPTR II)





- Consequences:
 - High visibility troubling cosmetic deformity
 - Major cosmetic effect for minor curve

Surgical challenges:

- Vertebral arteries
- Nerve roots of functional significance
- Associated anomalies compromise anchors, correction
- 'Distraction' anchors difficult
- Circumferential approach, osteotomy difficult
- No easy/safe surgical solutions









- Klippel- Feil
 - Common association/overlap
 - Implications:
 - Assess/follow for instability
 - Limited cervical mobility exaggerates any cervicothoracic deformity – <u>compensatory cervical curve</u> <u>above C-T deformity not</u> <u>possible</u>









Occipito-Cervical anomalies

- Possible instability
- Associated head tilt affects C-T cosmetic deformity, limits compensatory curve
- Halo traction may be compromise
- Sprengel's
 - If ipsilateral to convexity of C-T curve, makes visual deformity much worse
 - If contralateral to convexity of C-T curve, may hide/compensate deformity







- Natural History
 - Progression dependent upon:
 - Balance of growth potential:
 - R vs L
 - Tethering bars or ribs
 - <u>Infant xray</u> useful in defining anatomy, predicting progression.







- Natural History
 - <u>Progression with rapid</u>
 <u>growth</u>. Pattern same as other congenital spine deformity:
 - Growth rate:
 - Highest age 0-2 and preadolescent growth spurt
 - <u>Early adolescent</u>
 <u>progression often missed</u>
 <u>after years of childhood</u>
 <u>stability</u>







- Consequences of C-T deformity
 - Thoracic Insufficiency Syndrome
- Pulmonary compromise
 - Upper thoracic vertebral growth arrest/retardation:
 - <u>Disproportionately profound effect</u> <u>of upper thoracic spine disturbance</u> <u>on pulmonary function</u>:
 - Canavese rabbit model
 - Synder rabbit model
 - Emans early fusion series
 - Karol early fusion series
 - Pulmonary compromise correlated with:
 - More levels fused
 - Earlier fusion
 - More proximal fusion
 - < 18-22 cm thoracic height at maturity (normal 28 cm)





Dorsal Arthrodesis of Thoracic Spine and Effects on Thorax Growth in Prepubertal New Zealand White Rabbits

Federico Canavese, MD,* Alain Dimeglio, MD,* Donatella Volpatti, APD,†

Pulmonary Function Following Early Thoracic Fusion in Non-Neuromuscular Scoliosis



- Compensatory curves:
 (MRI to r/o tethering)
- Cervical compensatory cu useful cosmetically
 - Longer term pain issues
- Thoracic compensatory curve
 - Usually quickly structural
 - Potentially very progressive, with rotation
 - Progressive thoracic deformity, <u>TIS often a more medically</u> <u>urgent issue than the C-T</u> <u>curve</u>







Age 30 months

AP



- Progressive
- Treatment Options?
 - Observation
 - Brace
 - In situ fusion
 - Convex epiphyseodesis
 - Instrumented
 - With concave distraction
 - Distraction Rx
 - GR, VEPTR
 - -VCR
 - Combinations?
- What to do?







Treatment Options:

- Observation:
 - Some C-T deformities nonprogressive
 - Deformity in thorax below may be more significant
- <u>Non-operative treatment</u>?
 - TOT-collar, C-T orthosis, Minerva, Pin-less Halo options
 - <u>Questionable effect</u> possibly helps create compensatory curve and improves cosmetic appearance







- In-situ fusion?
 - Safe option for progressive deformity BUT
 - Need anterior fusion also if lordotic deformity
 - All the *disadvantages* of fusion
 - Lost spine height, thoracic volume
 - No advantages of instrumentation, fusion:
 - Correction of deformity
 - Improvement in compensatory curve





Convex epiphyseodesis?

- Open A/P or Heinig posterior transpedicular A/P convex epiphyseodesis
 - Skill-sensitive technique with some risk
 - In-Situ fusion effect or correction with growth?
- Uzumcugil, A., A. Cil, et al. (2004).
 - "Convex growth arrest in the treatment of congenital spinal deformities, revisited." JPO 24(6): 658-666.
 - Mean age 29 months
 - 41% progressive improvement
 - 47% fusion effect
 - 12% progression
- Effect on Thoracic Height?





- Instrumented convex epiphyseodesis
 - Demirkiran, G., G. Yilmaz, Yazici et al. (2014). "Safety and efficacy of instrumented convex growth arrest in treatment of congenital scoliosis." JPO 34(3): 275-281.

– >/=5 degrees improvement in 9/12 pts

- Low risk, low tech
 - Does not compromise other operations or future techniques
 - Later VCR not altered
 - Simultaneous GR or VEPTR possible
 - ?Progressive correction?
 - Inadvertent lordosis from posterior tether?





Instrumented convex epiphyseodesis –







Instrumented convex epiphyseodesis –



- Convex epiphyseodesis and concave distraction
 - Case D Skaggs, CHLA









- Growing rods (spine based distraction)
 - Rarely feasible for C-T curve alone:
 - Anchor points in cervical spine problematic

<u>GR Combined with C-T</u> fusion or convex epiphyseodesis

 Common technique to control C-T deformity with worsening thoracic deformity below











• GR Combined with C-T fusion

- Klippel Feil and Occ-Cerv anomalies
 - <u>Unsolved head tilt</u>



- VEPTR
 - Concave expansion thoracostomy and VEPTR (rib based distraction)
 - Campbell JBJS -Improvement of cervical tilt, shoulder asymmetry, deformity more thoracic than cervical
 - Late effects of anchor drift, scarring/fusion beneath VEPTR?



- VEPTR
 - Concave expansion thoracostomy and VEPTR (rib based distraction)



Boston Children's Hospital Orthopedic Center







3- column osteotomies

- VCR /hemivertebra excision
- Best available correction
- Biggest risks
- VCR Caveats:



- Assess location of vertebral arteries - anomalous paths common
- Where are vertebrals <u>relative to osteotomy?</u>
- What roots are at risk?

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Example from T Zhang MD, PUMC

- VCR Caveats:
 - Anchor points in C-spine problematic
 - Undesirable extension of fusion into c-spine
 - Correction difficult, helped by:
 - Convex rib resection
 - Concave rib osteotomy
 - Halo vest
 - Useful to achieve/maintain correction

VCR Risks:

- Vertebral arteries
- C8, T1 roots, anomalous C-T roots
- Excessive bleeding anteriorly
- Anterior sympathetic chain
- Brachial plexus, recurrent laryngeal, phrenic indirect injuries
- Pneumo/hemo-thorax





VCR for C-T dysgenesis/dislocation







VCR for progressive head tilt













- VCR for failed instrumented in-situ fusion
 - Instrumented convex epiphyseodesis age 26m









- VCR for failed instrumented in-situ fusion
 - Anterior column deficiency not recognized, progression after convex epiphyseodesis



• Conclusion?

- Variable natural history
- Variable significance
 - Thoracic function
 - Appearance
- Multiple surgical options
 - Instrumented convex epiphyseodesis
 - -+/- concave distraction
 - VCR
- Beware:
 - Upper thoracic length, upper thorax volume important to long term pulmonary function.



