The Cervical Spine in Skeletal Dysplasia

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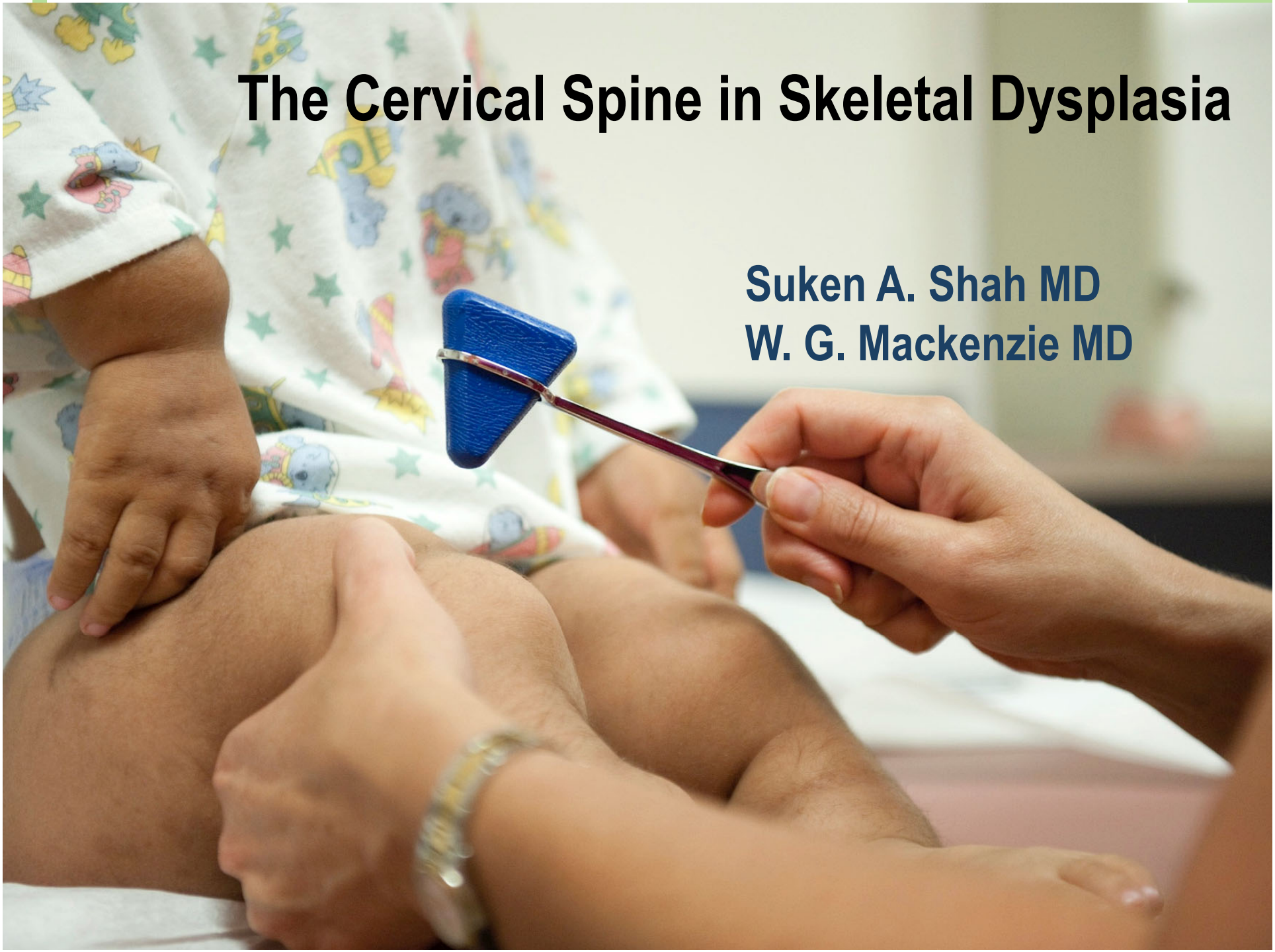


Cervical Spine Problems in Skeletal Dysplasia

Instability - O-C 1, C 1-2, Sub -axial

Stenosis – Cervical

Deformity – Kyphosis/Lordosis (rarely)

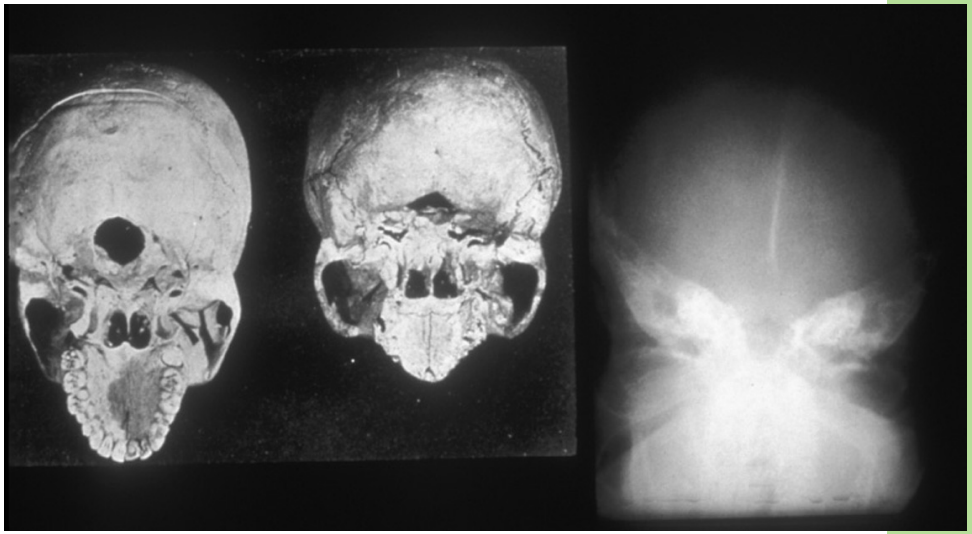
Clarify diagnosis prior to treatment as the natural history and associated problems vary with dysplasia type

Achondroplasia
Foramen Magnum Stenosis

Cervicomedullary Compression

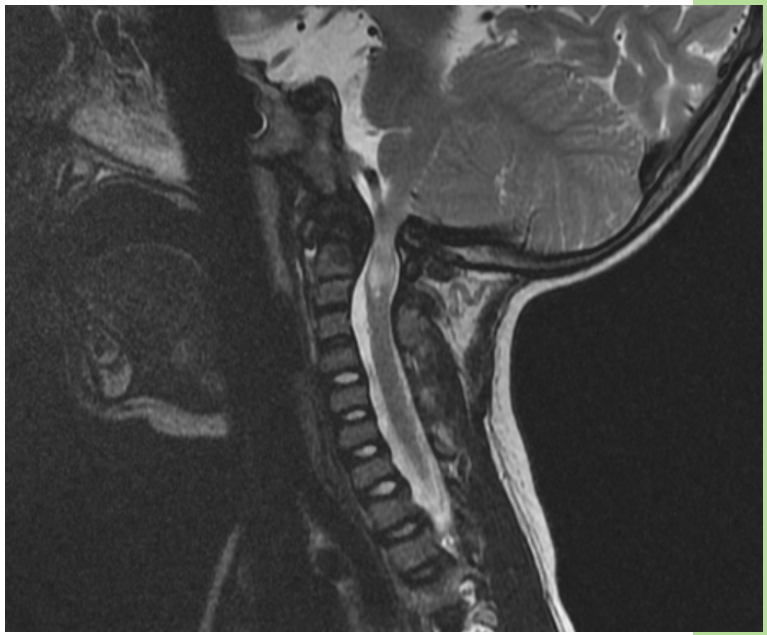
(60 % incidence)

- Common in achondroplasia but not in other types of skeletal dysplasia
- Can cause;
  - Developmental delay
  - Hypotonia
  - Sleep apnea
  - Feeding difficulties
  - Hemiplegia, quadriplegia
  - Sudden death



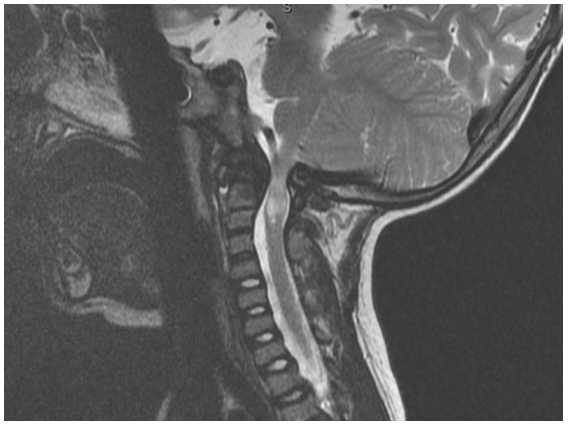
Foramen Magnum Stenosis

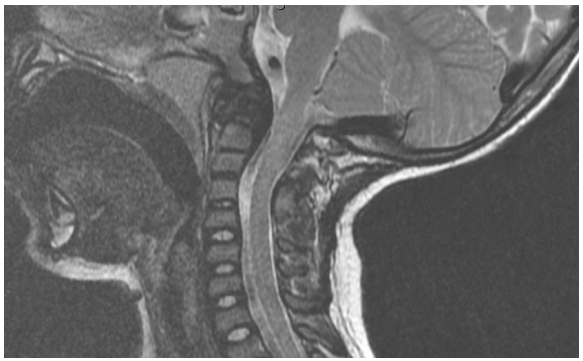
- Evaluation-
  - AAP recommendation for CT or MRI in perinatal period
  - Sleep study
- Natural History –
  - Can see spontaneous relief of compression over first 3 years of life



Foramen Magnum Stenosis

- Surgical Decompression
  - ~5% incidence
  - Fusion not required
  - significant complications
    - CSF dynamics
    - neurological injury
  - good results in symptomatic children but ? prophylactic surgery





Cervical Instability in Achondroplasia

- Instability defined as abnormal does not occur in Achondroplasia, BUT physiologic instability with FM and /or C1 stenosis can result in spinal cord compression

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Atlantoaxial Instability

- Spondyloepiphyseal Dysplasia and variants
- Morquio's Syndrome
- Pseudoachondroplasia
- Metatropic Dysplasia
- Kniest Dysplasia
- Metaphyseal Chondrodysplasia
- MPS

Every diagnosis except for Achondroplasia

Evaluation

- Careful history and physical examination
  - Hypotonic baby
    - Delayed motor milestones
  - Functional loss
  - Change in endurance
  - Change in bowel or bladder function
  - Neurological findings
    - Initial evaluation and changes at subsequent visits

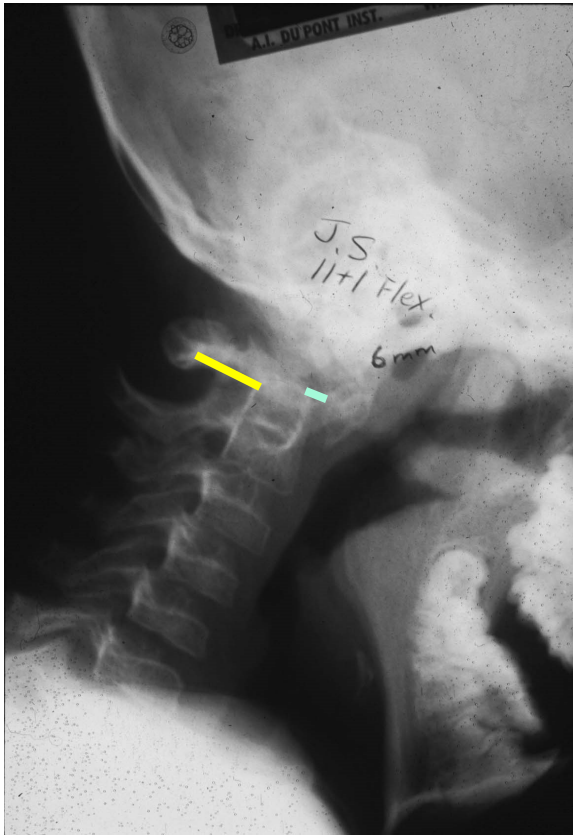
Evaluation

- Flexion/extension lateral cervical X-rays
- Flexion/extension MRI
  - Safe under sedation- Mackenzie et al J Pediatr Orthop 2013
- CT
  - to assess C1 arch, anatomy for C1-2 screw , lateral mass morphology, vascular abnormality
- Urodynamics

Atlantoaxial Instability

Etiology

- Ligamentous laxity
- Odontoid hypoplasia/aplasia
- Os odontoideum (common)
- Associated stenosis common



Atlanto-dens interval

Space available for cord

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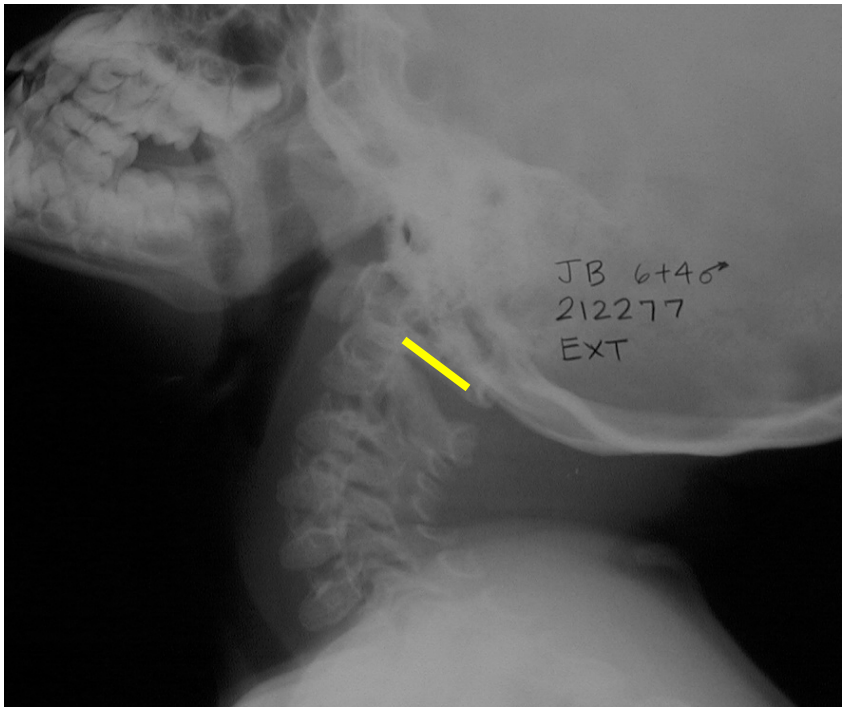
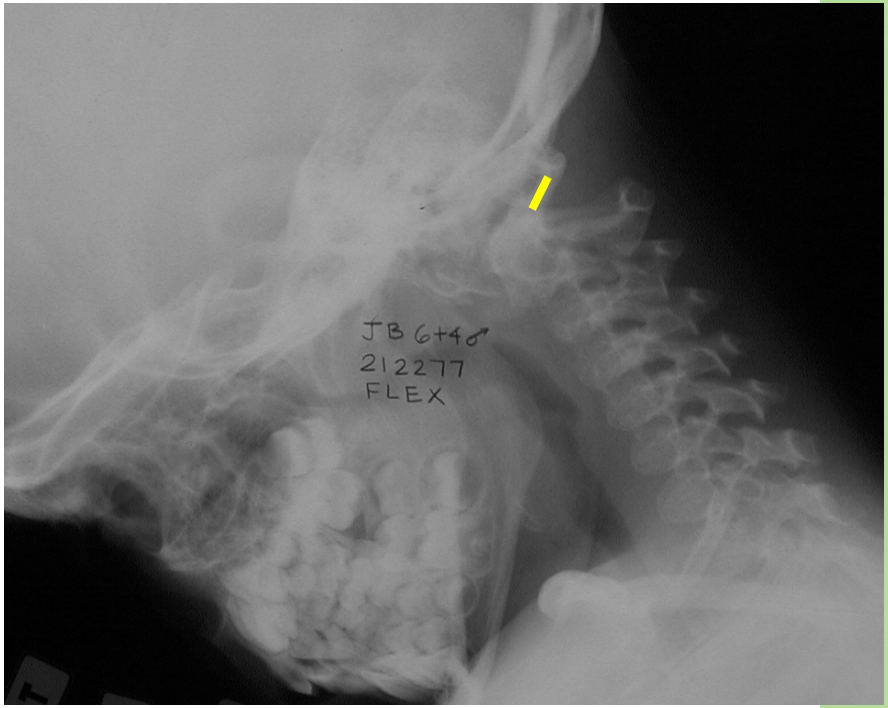
Pseudoachondroplasia





Space available for the cord

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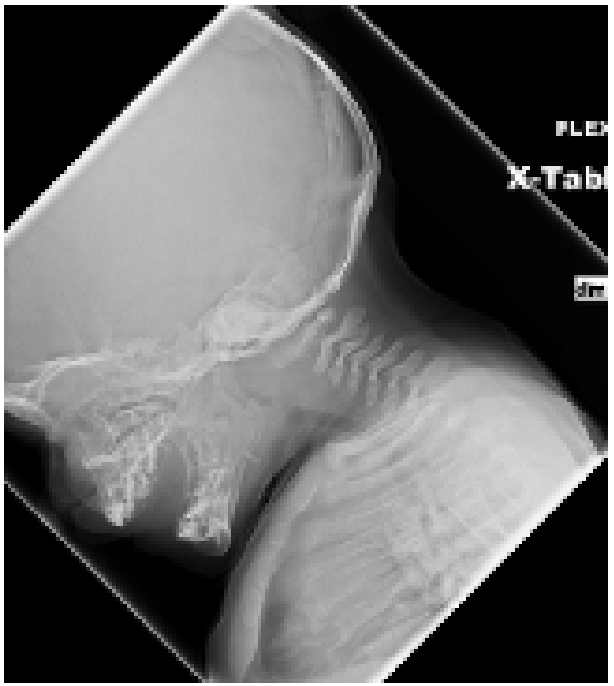
Flexion/Extension Cervical Spine X-Rays
SED Tarda 18 months



Interpretation is challenging

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Flex/Ext MRI
SED Tarda 18 months

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Metatropic Dysplasia- 10 yo, asymptomatic but moderate C1-2 instability (Flexion SAC -12mm)

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Metatropic Dysplasia- 10 yo Flex/ext Cervical MRI
- no cord compression

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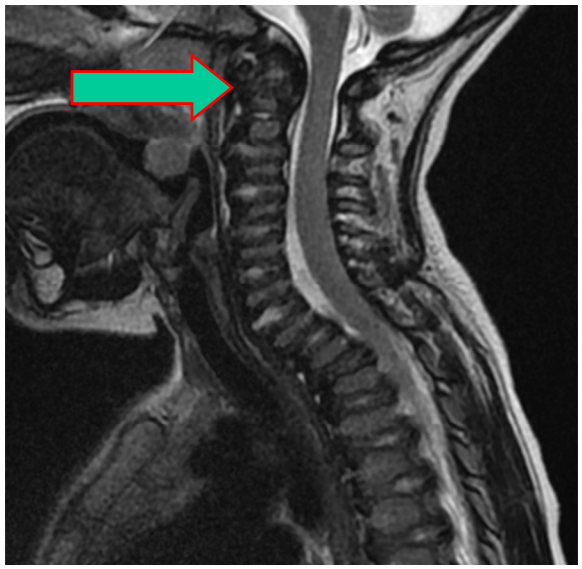
Cervical Stenosis

- Congenital
- Instability
- Secondary to extradural thickening
  - MPS

RCDP



Cervical Fusion +/- Decompression

- **Indications**
  - **Fusion**- Significant instability without cord compression
    - ADI > 8 mm or SAC < 13 mm
      - Commonly quoted as indications in the literature ?
  - Instability with cord compression
    - **Fusion**- Vertebral translation can be reduced, relieving cord compression
    - **Decompression and fusion-** when vertebral translation cannot be reduced
  - Cervical myelopathy

Management

- Careful preparation
  - Pre-op measurement for Halo-vest (custom) or other bracing if required
  - Neuromonitoring
    - Sensory AND Motor evoked potentials
    - ALL spine cases and some extremity cases (kyphosis)
  - Consider instrumentation and other intraop needs
    - Mayfield apparatus, halo adaptor
  - Personnel
    - Anaesthesia (video intubation), Neurosurgery, General Surgery
- PICU and Medically Complex Team for post-op support
  - Respiratory complications

Hurler/MPS 1- 12y- Flex/Ext Cervical Spine X-Rays

SAC in flexion- 12mm, extension- 21mm

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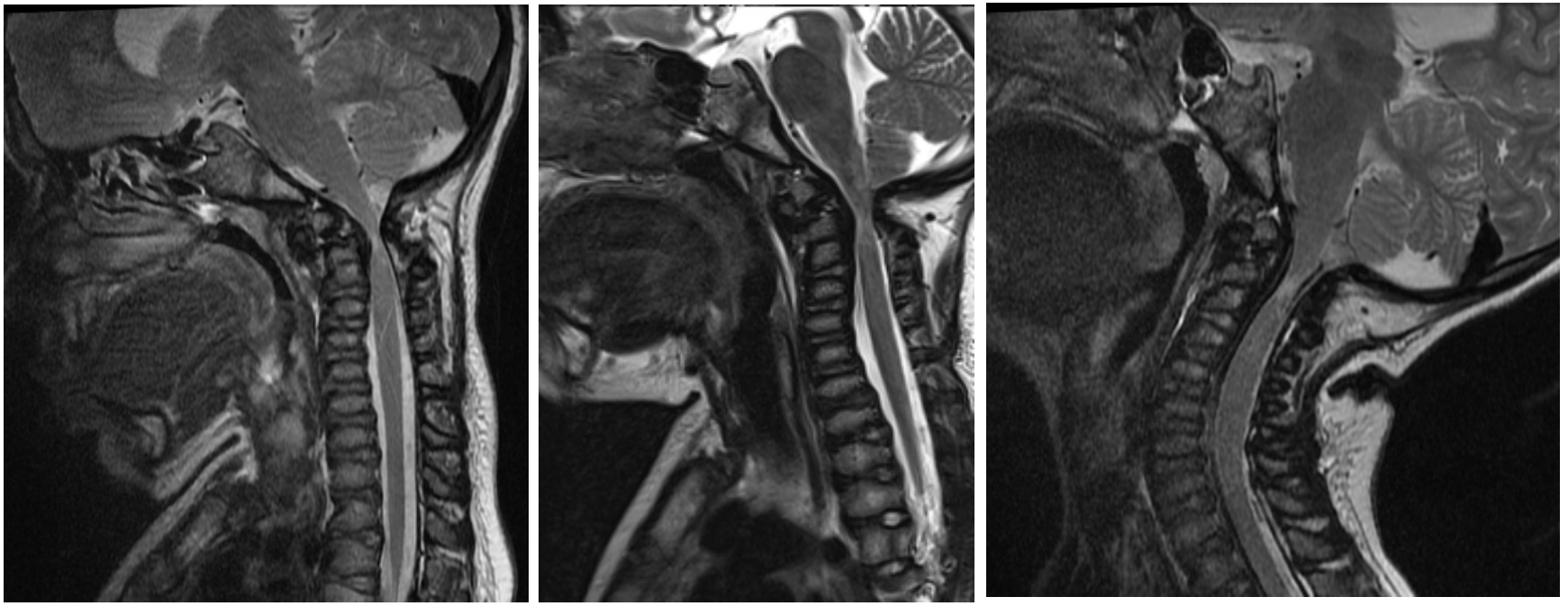
Awake Flex/Ext MRI



C 1-2 instability, stenosis, extradural MPS deposition, myelomalacia

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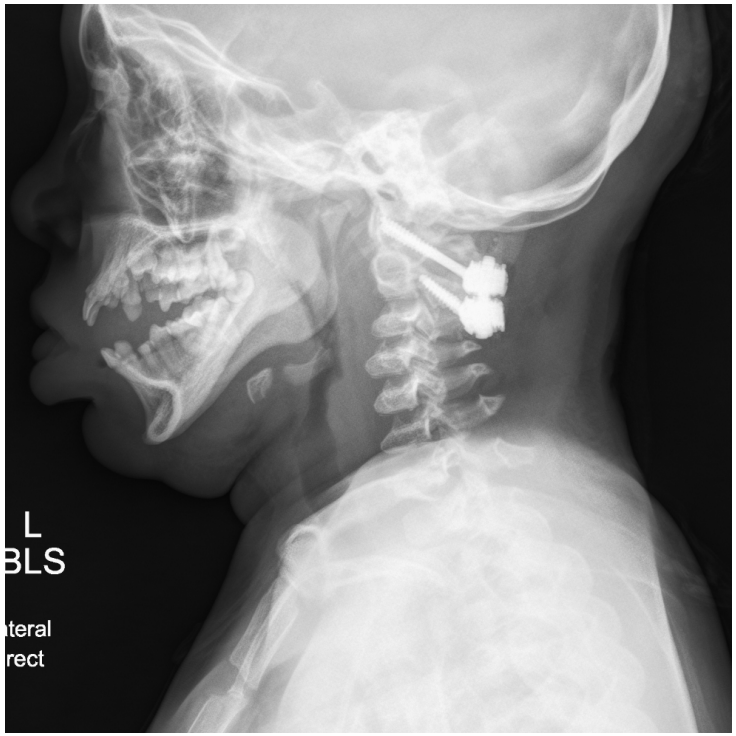
C1 decompression with C1 lateral mass and C2 pedicle fixation



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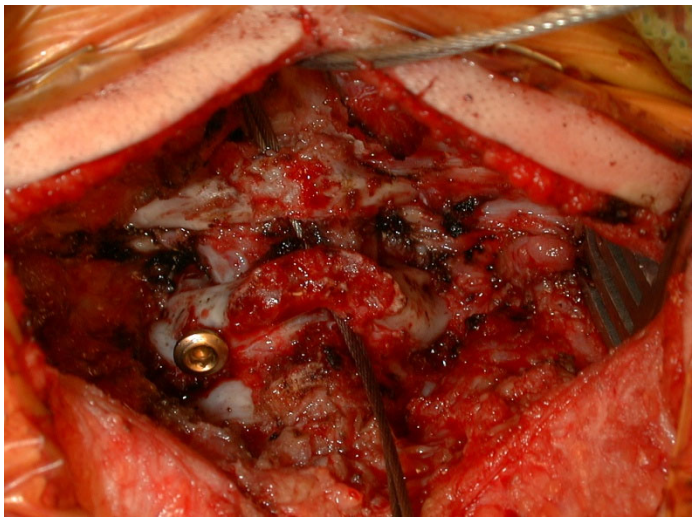
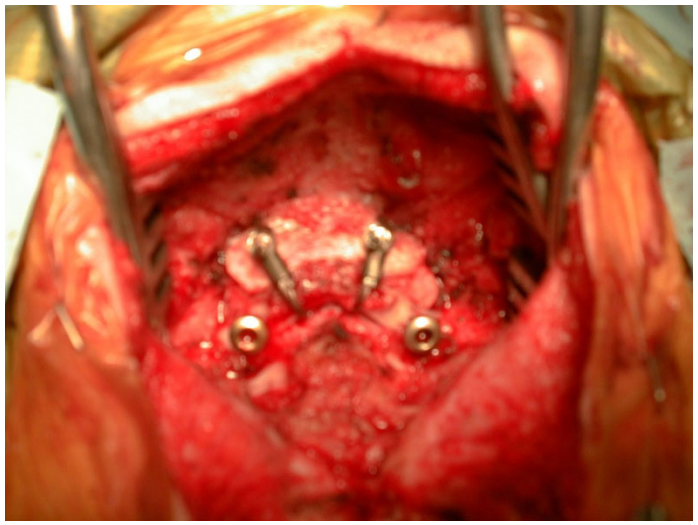
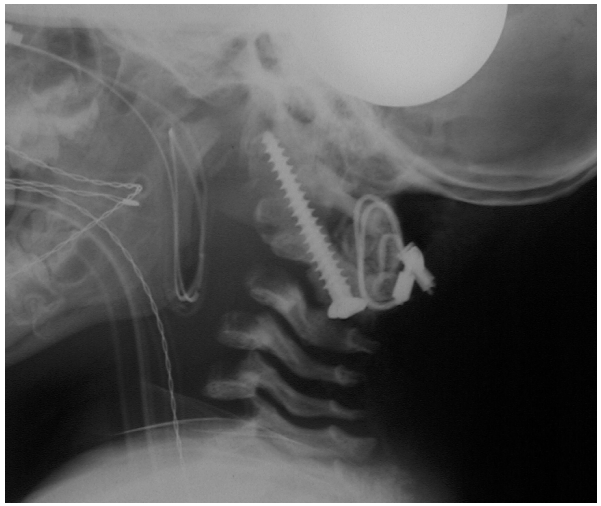
C 1-2 Fixation

Transarticular screws, Brooks wiring







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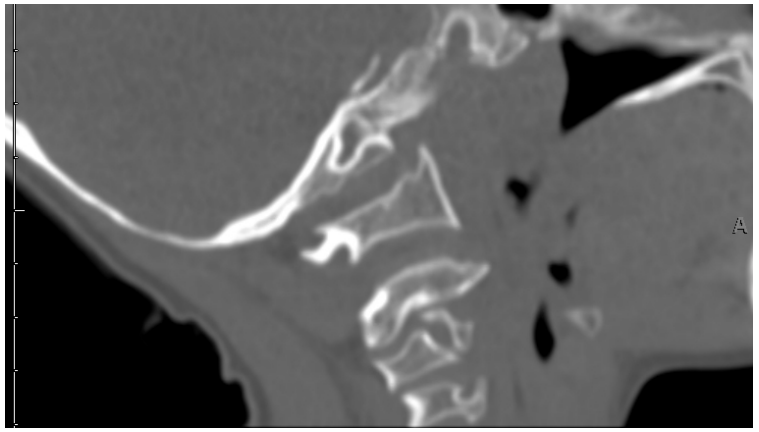

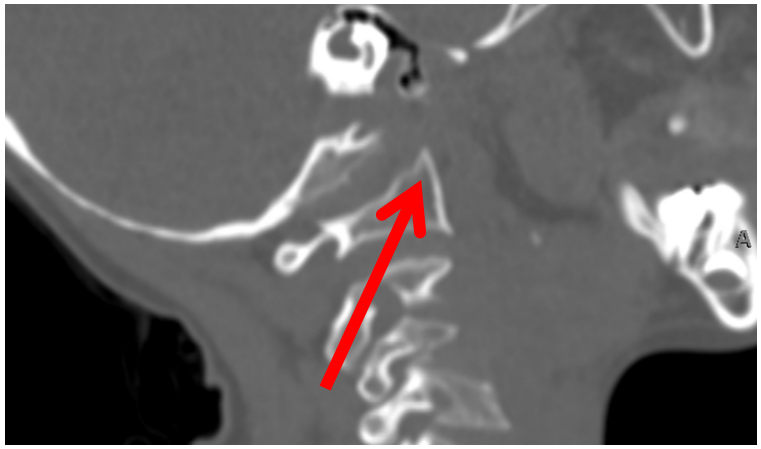

Metatropic Dysplasia with C1-2 instability that reduces
Pre-op CT

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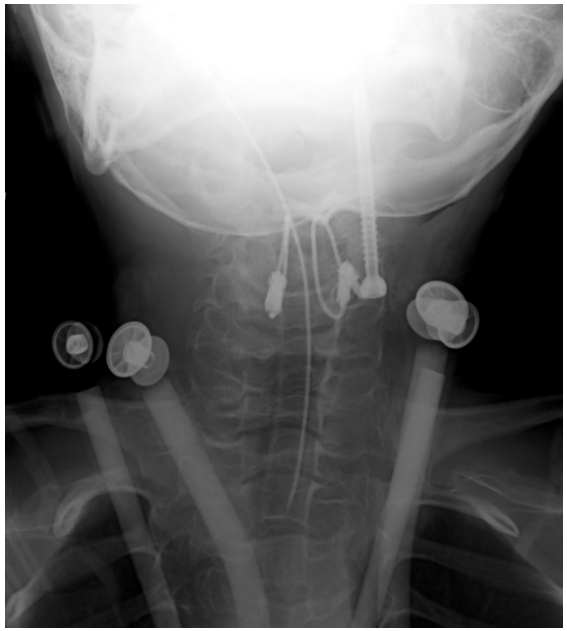
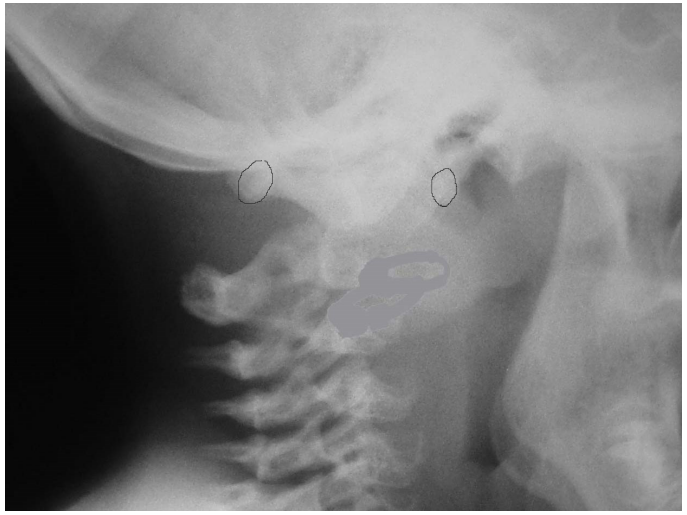
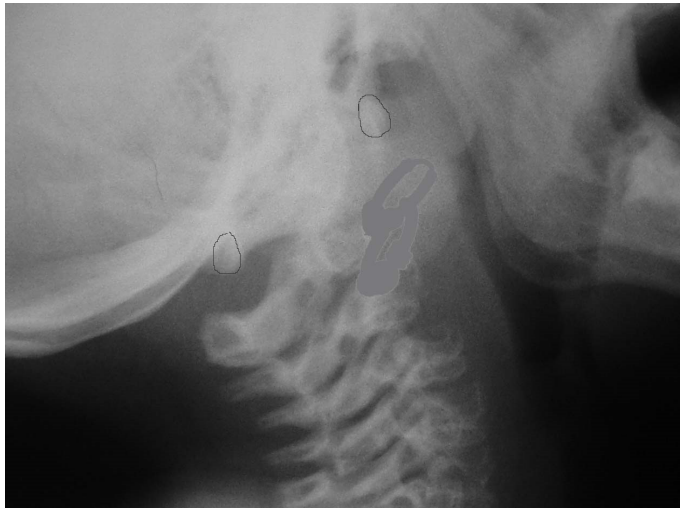
- One Solution
  - Single transarticular screw (Magerl) with posterior interlaminar wiring

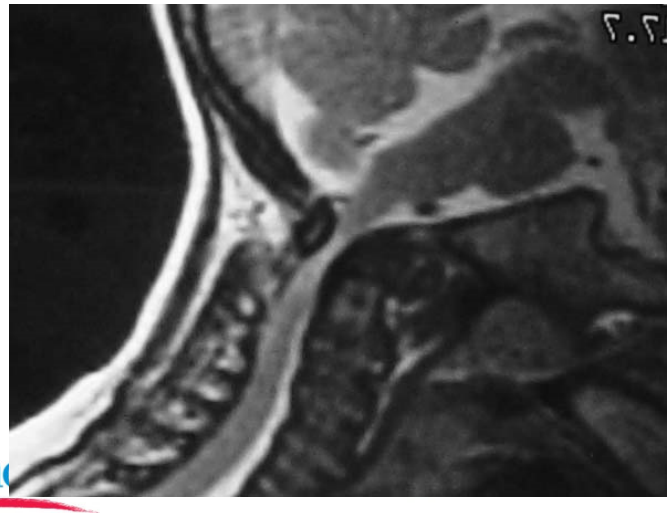


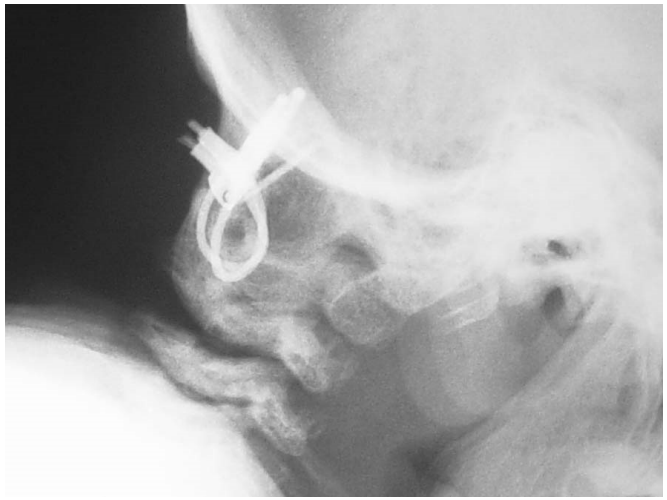
Morquio Syndrome

Irreducible subluxation, Decompression and O-C2 fusion

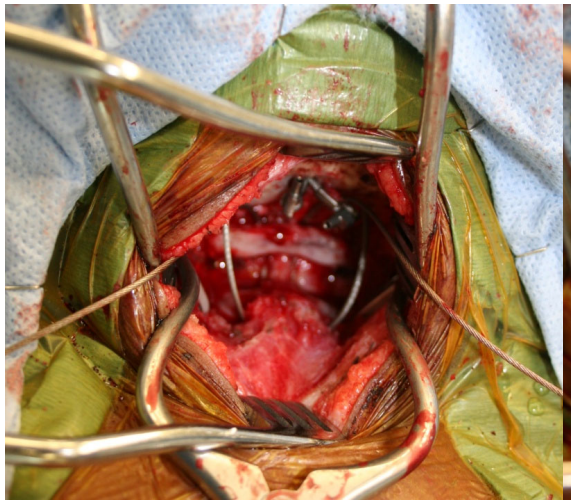
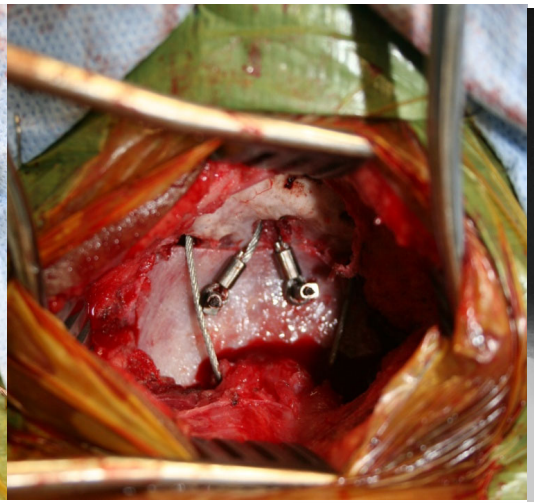
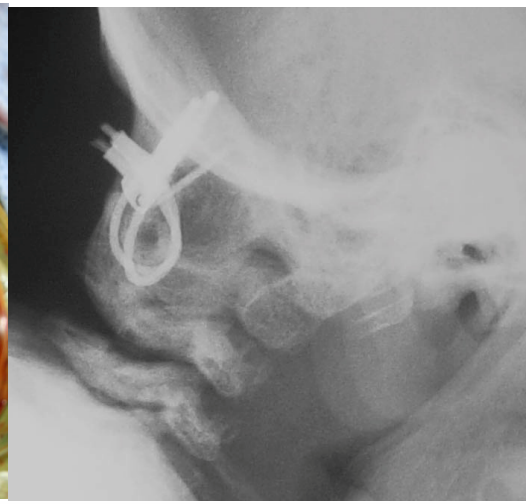






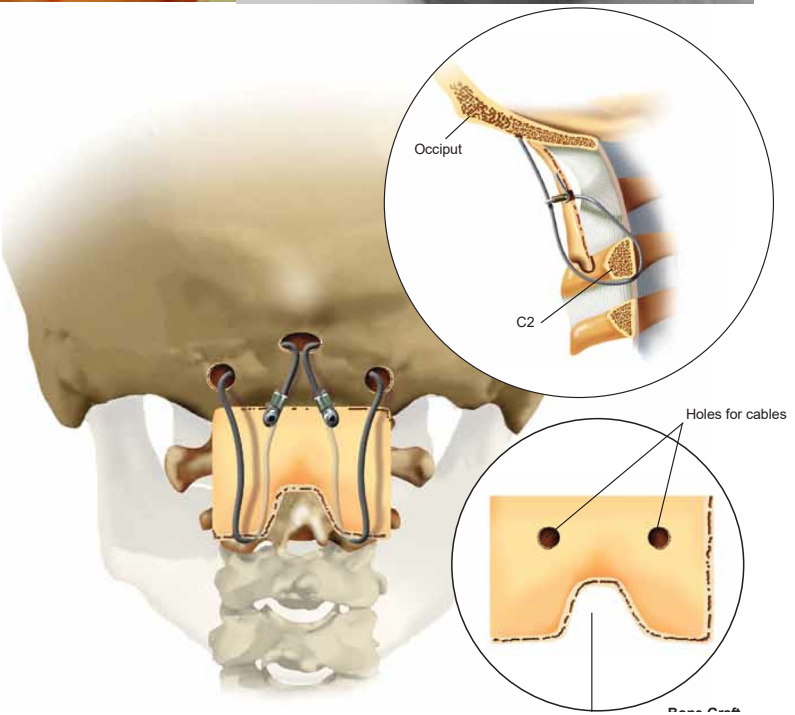


Occipitocervical Fusion

Dual Cable fixation from Occiptal burr holes to C2 lamina with illiac crest or rib graft provides good alignment, stable fixation and reliable fusion

Sitoula et al *Spine* 2014



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Anaesthetic complications

- All patients with MPS present major anaesthetic risks, and death can result if appropriate precautions are not taken. (Baines 1983)
- The most important complications relate to the following:
  - Dysostosis multiplex leading to rigidity of the neck and spine.
  - Cervical instability
  - Induction may be difficult because of an inability to maintain an adequate airway. (Kempthorne)
  - **Intubation may require smaller-than-anticipated endotracheal tubes and a narrow,deformed trachea and thickened vocal cords will impede intubation.(Theroux et al 2015)**
  - **Recovery from anesthesia may be slow, and post-operative airway obstruction is always a risk.**

MPS IVA- Morquio Syndrome

- Mid tracheal obstruction 2° to Innominate artery compression and distal tracheal obstruction

Head and Neck in Extension Absolutely Essential for airway in MPS *(Walker et al, Pritzker et al, Theroux et al)*

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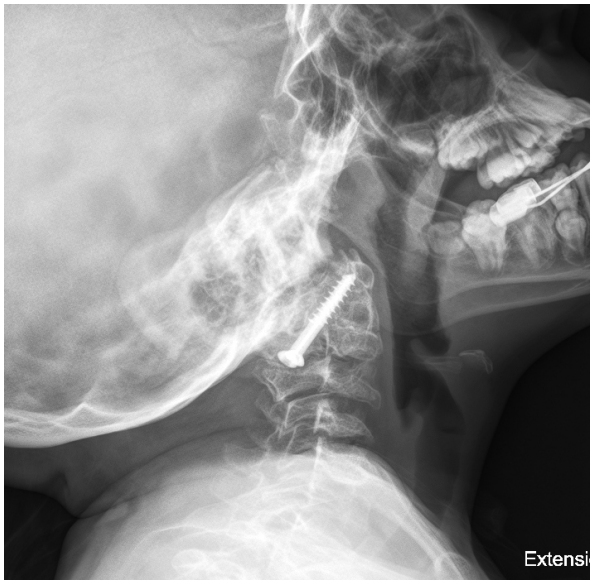
Late instability below successful fusion



- Morquio Syndrome ~ 30 % incidence of instability under O-C3 fusion (high incidence in SEDC)
  - Dede et al JBJS 2013

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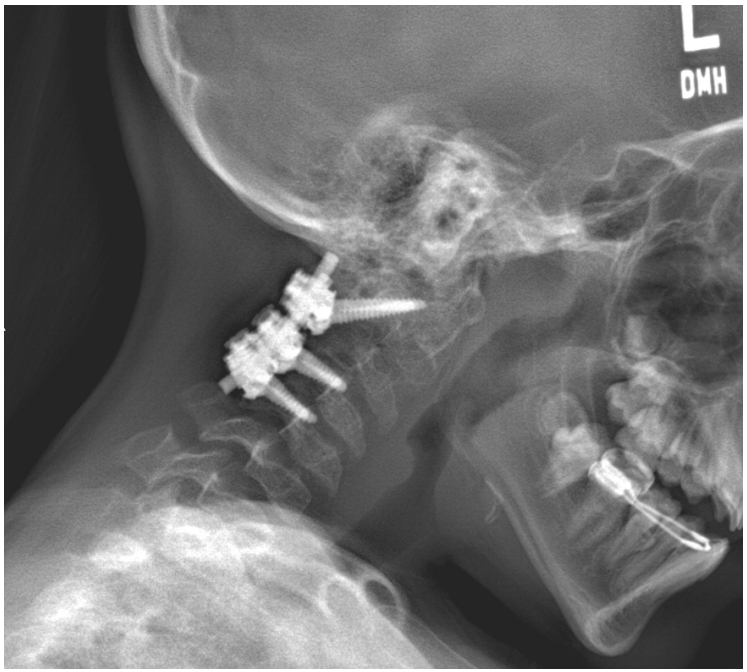
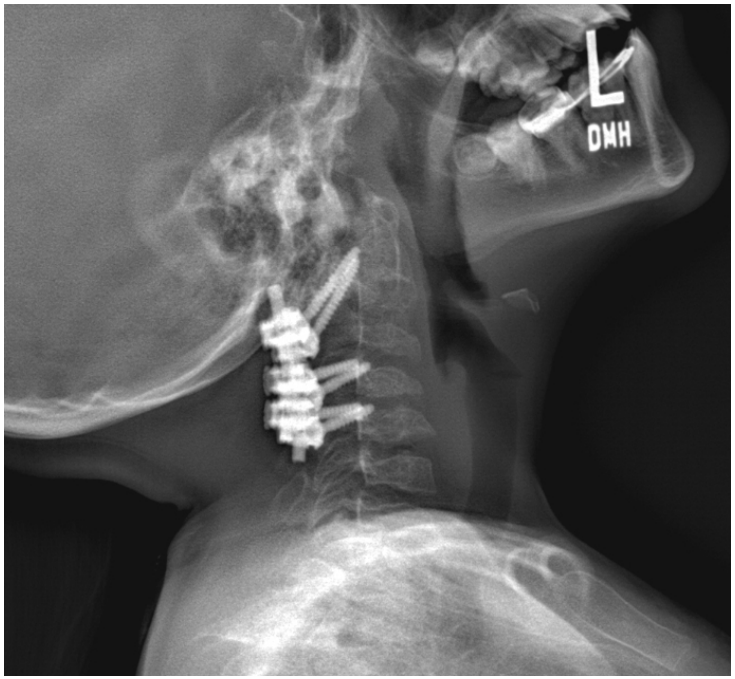




- Extension of fusion

- Pediatric Upper Cervical fusion Techniques
  - Koop et al 1984
  - Magerl, Seeman 1986
  - Dormans et al 1995
  - Rogers et al 1999
  - Harms, Melcher 2001
  - Gluf, Brockmeyer 2005
  - Ahmed, Traynelis, Menezes 2008
  - Hedequist 2009
  - Sitoula, Mackenzie 2014

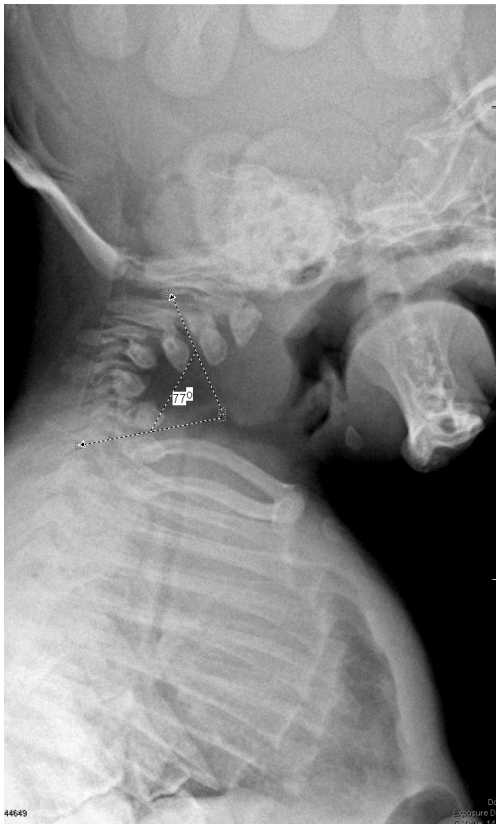
- **Upper Cervical Fusion in Skeletal Dysplasia**
  - Svensson, Aaro 1988
  - Miyoshi et al 2004
  - Ain et al 2006
  - Sitoula, Mackenzie et al 2006
  - Dede, Mackenzie et al 2013
  - Helenius et al 2015
- Overview of results
  - Most have multiple dx's, few cases
  - Fusion rates ~ 80% overall, **instrumented** ~ 100%
    - Use autograft
  - Neurologic recovery worse with chronic myelopathy
  - Complication rate low
    - Do not pass sublaminar wires if stenosis present
    - Beware vertebral artery

Cervical Kyphosis

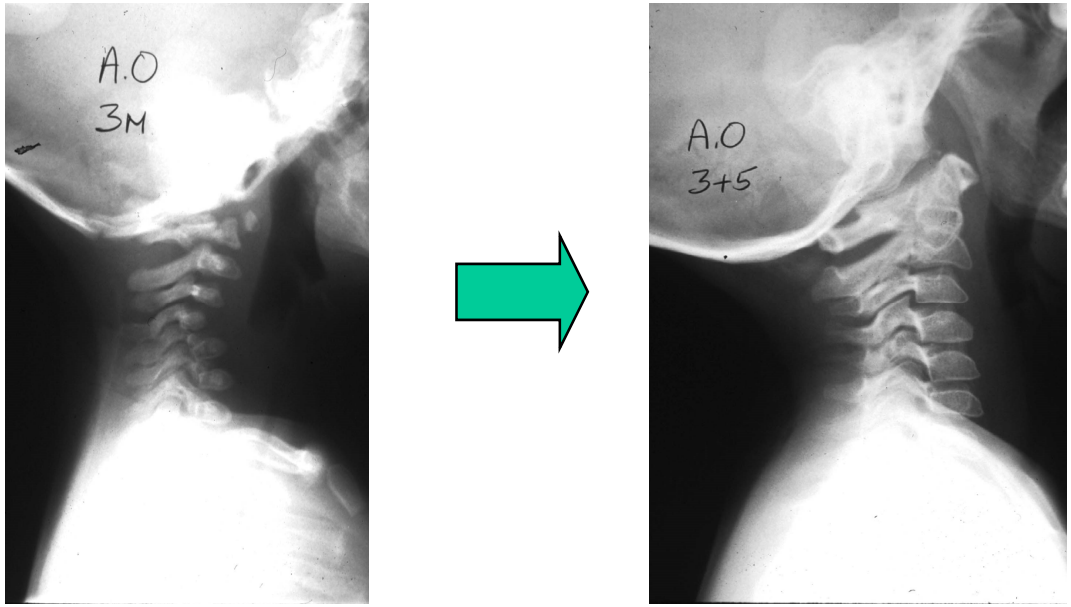
- Diastrophic Dysplasia
- Camptomelic Dysplasia
- Larsen Syndrome

Cervical Kyphosis in Diastrophic Dysplasia

- Incidence- 20-44%
  - Probably very common in newborn
    - Bethem et al 1980, Poussa et al 1991, Remes et al 1999
- Likely develops in utero
  - Factors thought to include; vertebral body wedging, ligamentous laxity and incompetent posterior restraints
- C-3,4,5 anterior hypoplasia
- Dysraphic posterior elements common

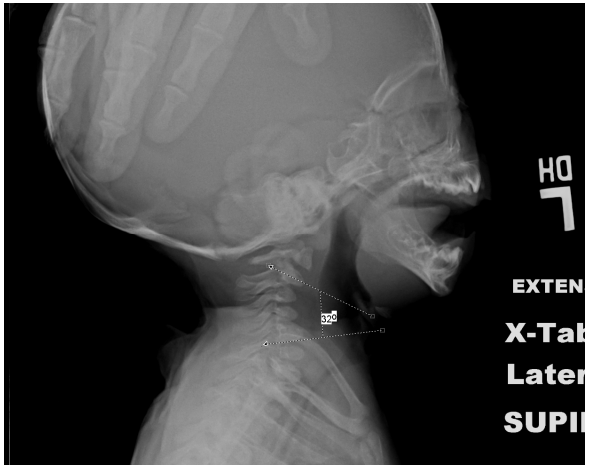


Cervical Kyphosis- Natural History

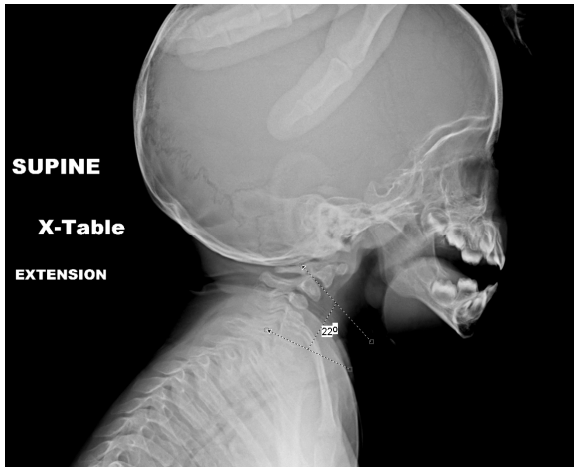


- Spontaneous improvement in > 75% occurs by age 5 years
  - Factors
    - < 60 °, no ovoid or severely wedged vertebrae at apex
      - Poussa et al 1991, Remes et al 1999
- Rarely see progression

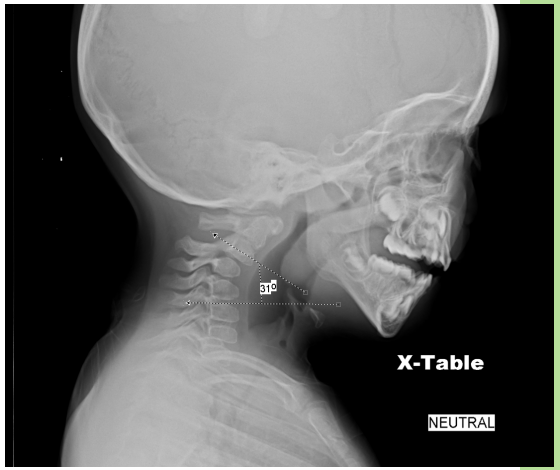
Typical Spontaneous Correction of Cervical Kyphosis (OV)



4 m



15 m



2+6 y

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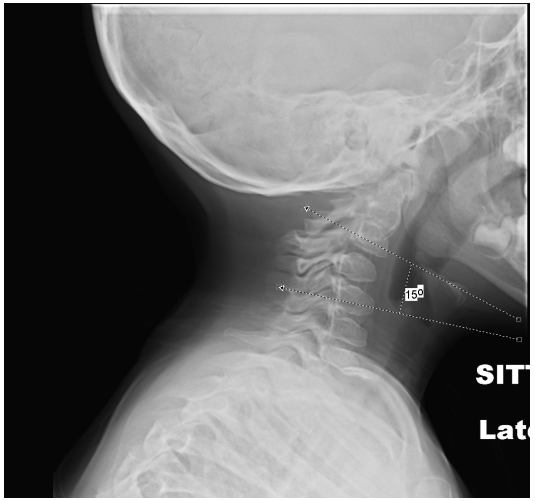
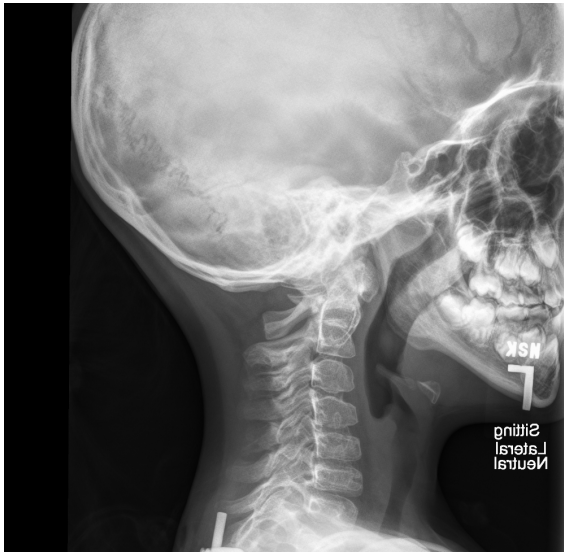


4 y

6 y



8 y

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Progressive Cervical Kyphosis

- Uncommon
  - 5/122 patients in Finland had a persistent kyphosis (average 90°)
    - Only 2 pts were alive at publication
      - 32 y with 165°
    - 3 pts. with severe kyphosis died
      - 2 died (severe tracheomalacia) at 1 y with 65 and 76°, 1 with 130° became quadriplegic after anaesthesia for a foot procedure
        - » Remes et al 1999
- Factors associated with progression
  - > 60°, ovoid or very wedged apical vertebra, posterior translation of apical vertebrae

Cervical cord compression in diastrophic dwarfism

Irwin J. Kash, M.D.,* *Pittsburgh, Pa.,* **Shashikant M. Sane, M.D.,** *Minneapolis, Minn.,* **Frederick J. Samaha, M.D.,** *Pittsburgh, Pa.,* **and Jakob Briner, M.D.,** *Switzerland*

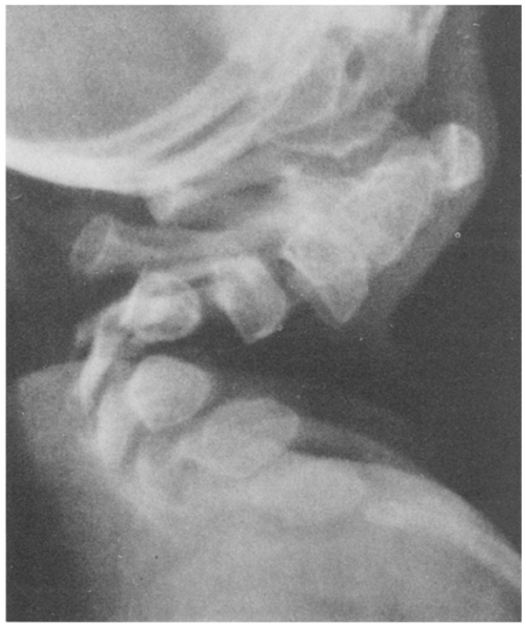

Fig. 1. Lateral view of the cervical spine showing marked dorsal kyphosis with small sized vertebral bodies of C_3, C_4, and C_5.

Journal of Pediatrics 1974

35 months old died of respiratory infection, quadriparetic

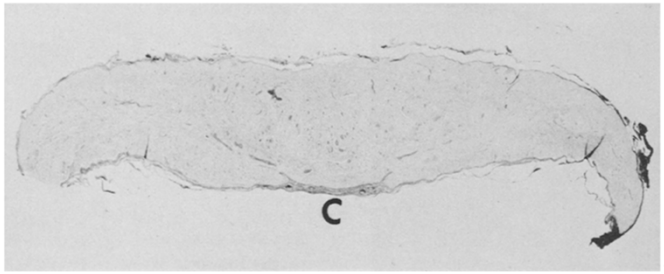

Fig. 3C. Section of spinal cord at the site of direct compression. There is a loss of virtually all the normal architecture of the spinal cord with replacement by astrocytosis. A few persistent myelinated tracts are barely visible in some areas. (Paraffin-embedded section; Luxol fast blue-PAS; × 7.5.)

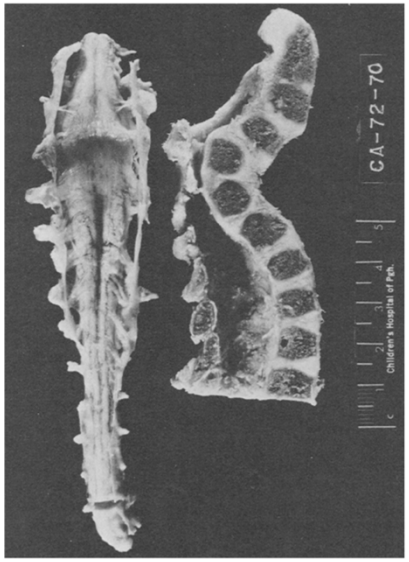

Fig. 2. Note the narrowing of the spinal canal at the region of spondylolisthetic slip and the corresponding compression of the spinal cord.

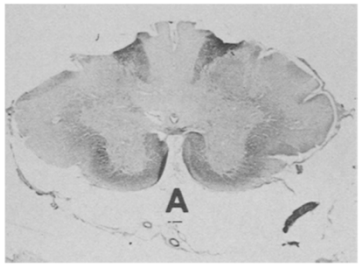

Fig. 3A. Section of spinal cord above the site of compression; ascending wallerian degeneration is present. (Paraffin-embedded section; Luxol fast blue–PAS; × 7.5.)

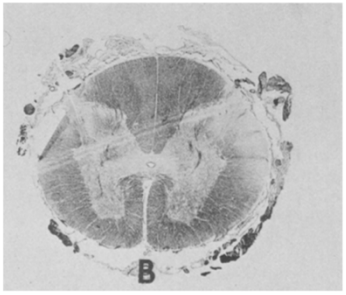

Fig. 3B. Section of spinal cord below the site of compression; descending wallerian degeneration is present. (Paraffin-embedded section; Luxol fast blue–PAS; × 7.5.)

tebrae, spinal cord compression has not been previously emphasized. This report describes a child with kyphosis

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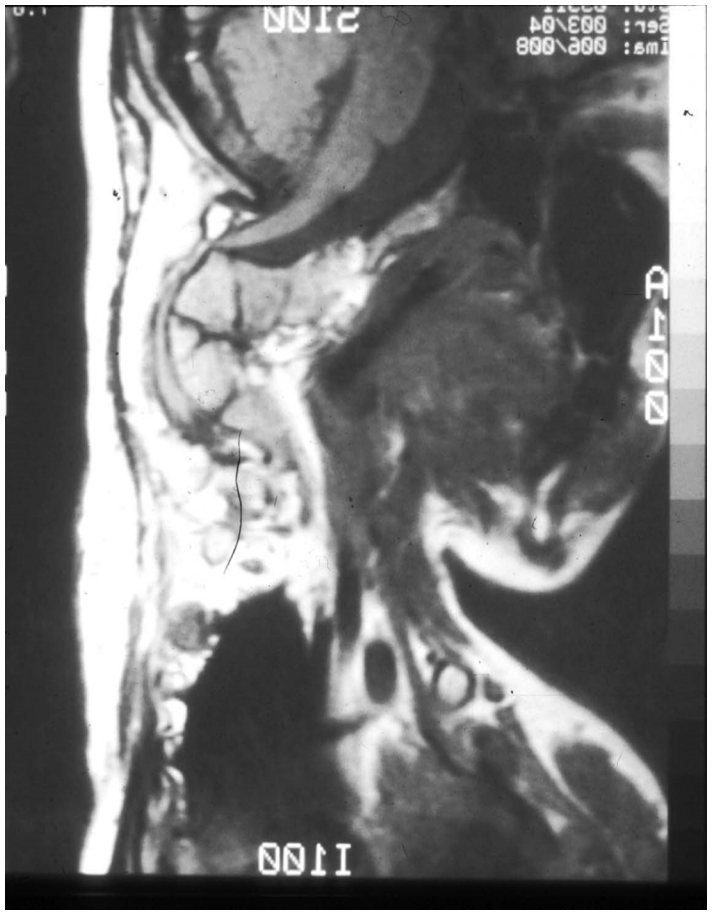
Progressive Cervical Kyphosis

- Treatment
  - Observation
  - Brace
    - No good evidence that it is effective
      - Bethem et al- ? Successful use of a Milwaukee brace vs. natural history
    - Indications for bracing in Literature
      - Progressive
      - No instability, < 60 degrees, < 4 yo and with normal neurological function

Severe Cervical Kyphosis

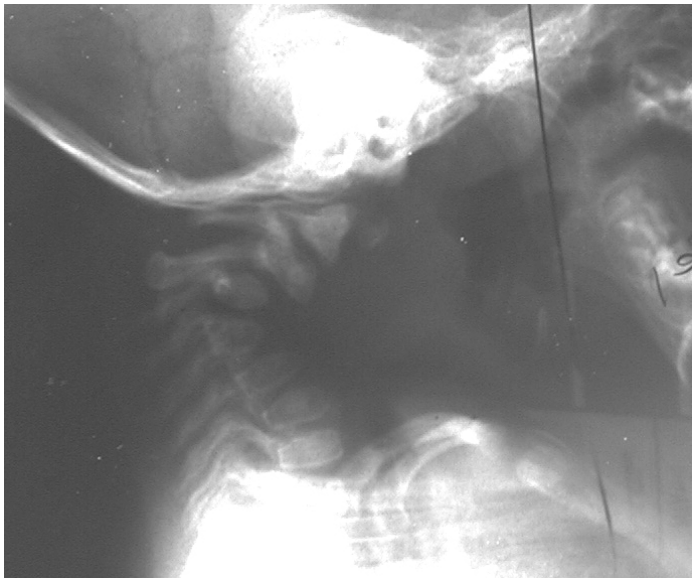
Treatment

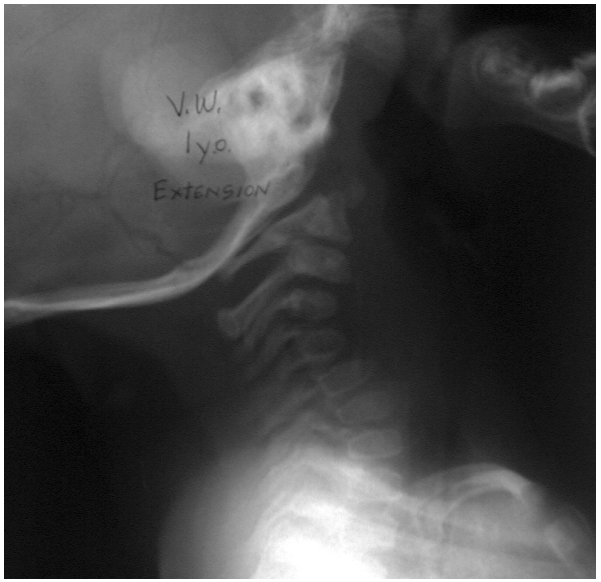
- Surgical
  - Indications
    - cord compression
    - severe or progressive kyphosis



Nemours. Children's Health System

Progressive kyphosis without cord compression

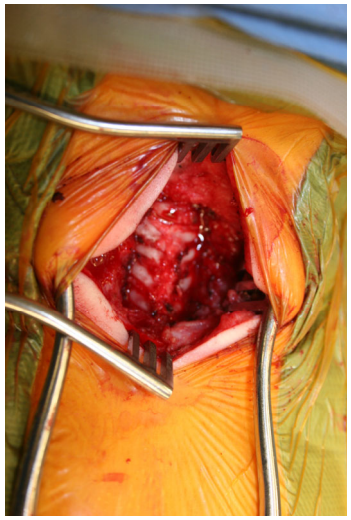




1 yo with 90° kyphosis, progressive, moderate flexibility and no neurologic symptoms or cord compression

- In situ PSF with halo-vest immobilization

Cervical spina bifida common !



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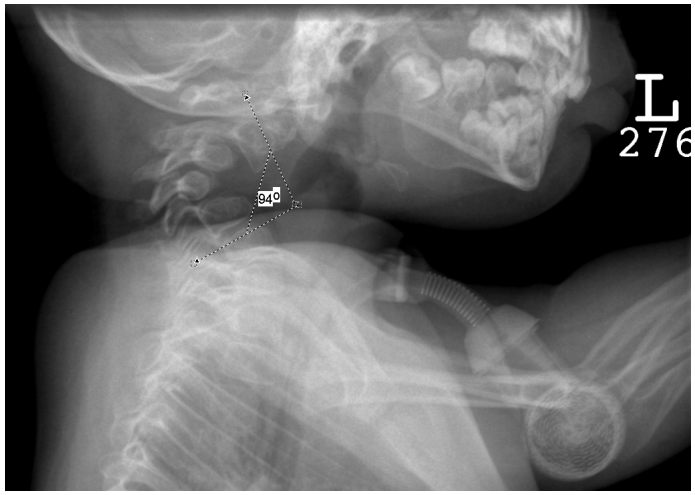
20 y

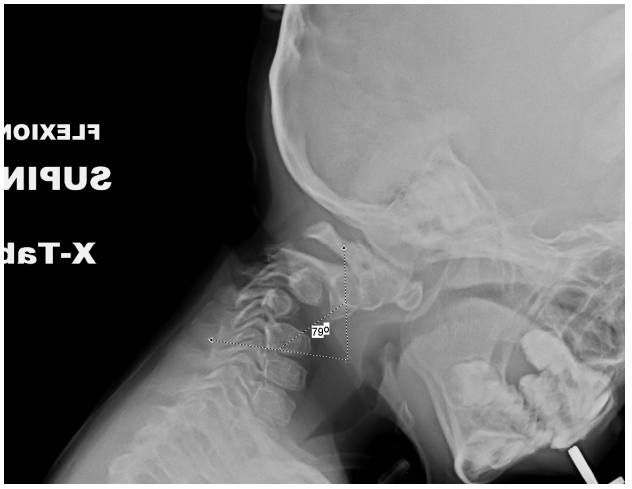
Progressive correction of kyphosis after posterior fusion

Nemours. Children's Health System

Progressive kyphosis with cord compression



3 y



4 y

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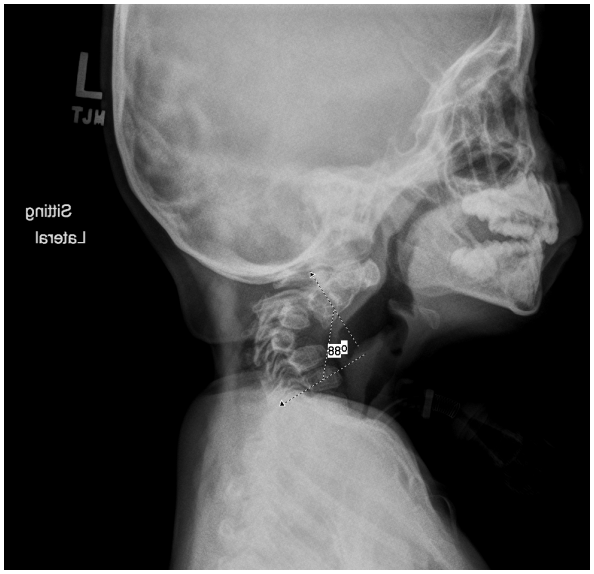
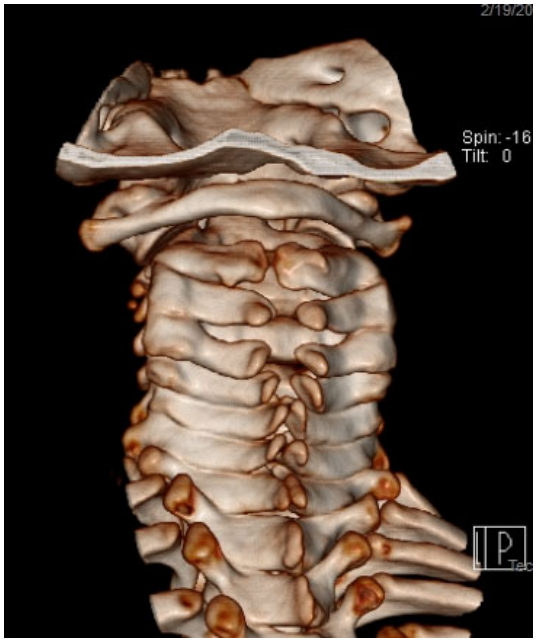
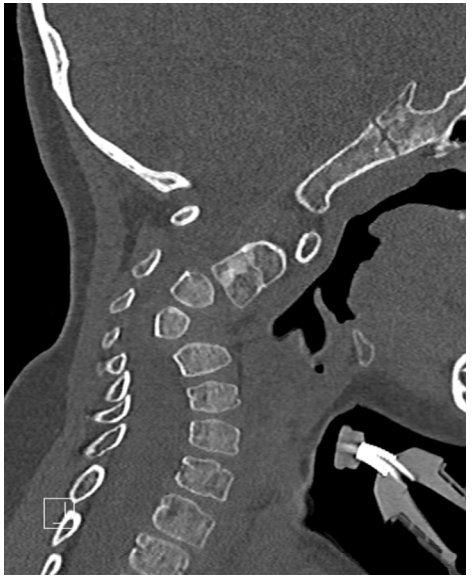
Neutral and Flexion/Extension X-Rays







5 y, weak upper and lower exremities

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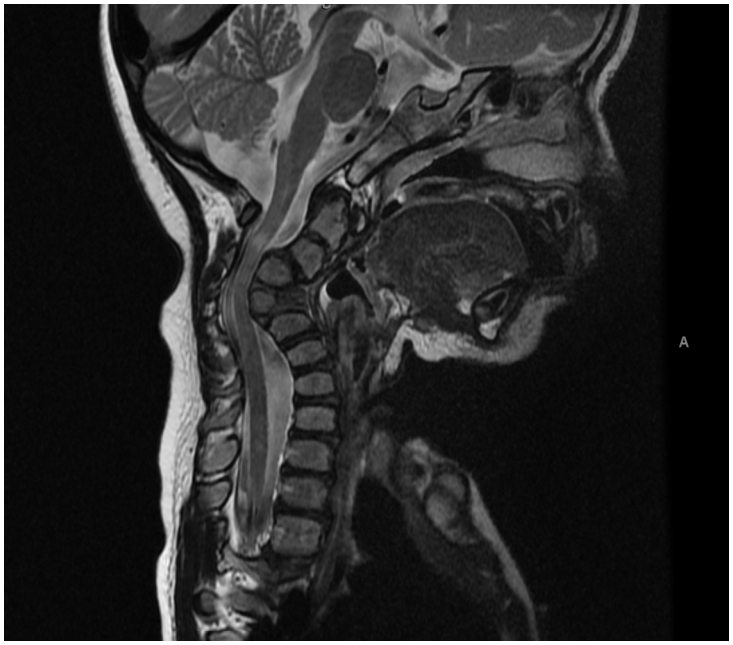

Cord compression and myelomalacia

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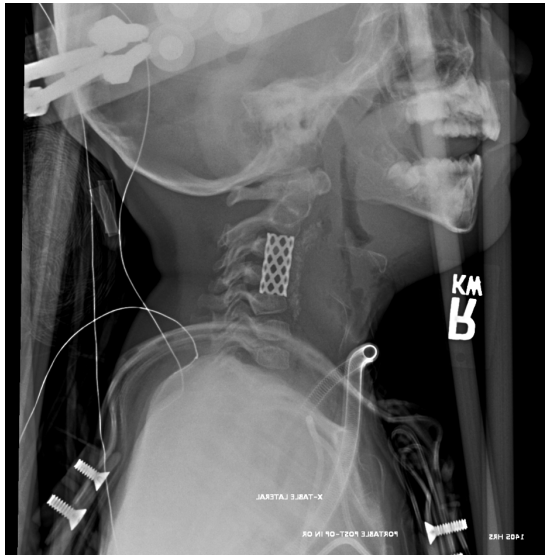
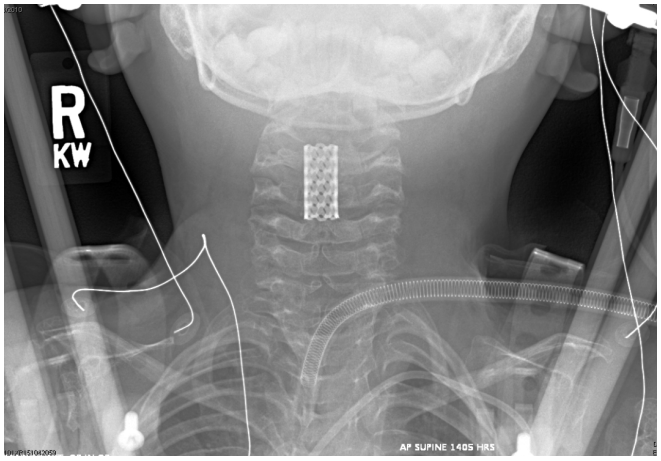
C-3,4 Anterior Vertebrectomy, Anterior and Posterior fusion



5 y



5+6 y

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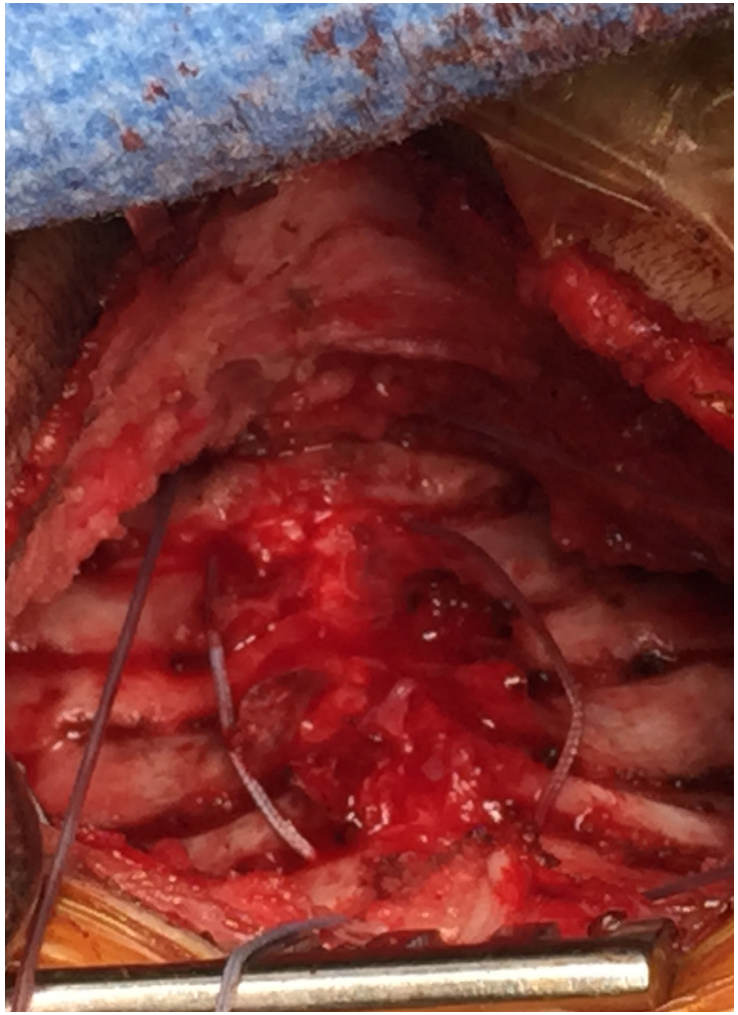
Posterior Suture Technique



Nemours. Children's Health System

Conclusions

- Diagnosis is important
  - Natural history
- Check for upper cervical spine instability and stenosis in all children with skeletal dysplasia
- Flex/ext MRI if poor ossification or any concerns about neurological symptoms or signs
- Have appropriately sized instrumentation and immobilization available
- Neurological recovery much better in young and those with minimal spinal cord changes
- Long term follow-up important
  - Adjacent instability

Nemours. Children's Health System

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