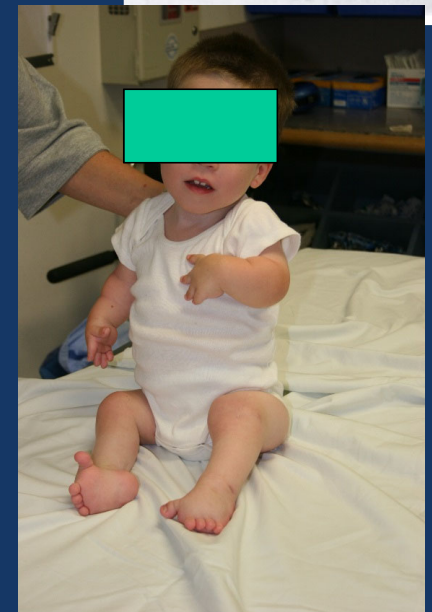
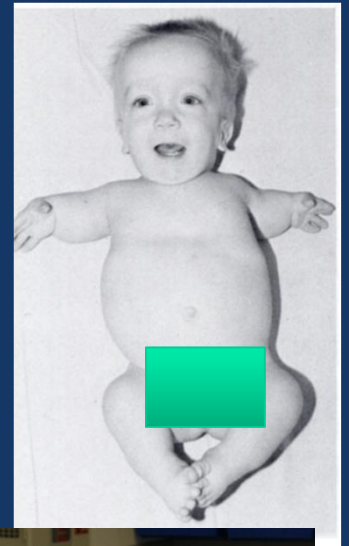


Anterior Vertebrectomy and Cervical Fusion: A Technique For Correction of Kyphotic Deformity in Diastrophic Dysplasia

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Background

- Diastrophic dysplasia first described in 1960
- Mutation in diastrophic dysplasia sulfate transporter gene (DTDST)
- Clinical abnormalities restricted to cartilage and bone due to decreased proteoglycan sulfation
- Short stature, laryngotracheomalacia, cleft palate, joint contractures, cauliflower ears, hitchhiker thumbs, symphalangism, equinovarus or skew feet
- Cervical kyphosis, scoliosis, hip dysplasia, lateral patellar dislocation, foot deformity, degenerative joint disease



Background

- Cervical spine kyphosis occurs in the mid cervical spine
- Usually present at birth - 30%
 - Factors include: vertebral body wedging, ligamentous laxity, incompetent posterior elements
- C3-C5 anterior hypoplasia, spina bifida occulta, and stenosis



Background

- Natural history of cervical spine kyphosis is spontaneous resolution, up to 60 degrees
- Severe progressive kyphosis + cord compression can occur but rare
- Many descriptions of posterior fusion techniques (Pakkasjarvi et al., Remes et al., Jalanko et al.)



Cervical cord compression in diastrophic dwarfism

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Fig. 1. Lateral view of the cervical spine showing marked dorsal kyphosis with small sized vertebral bodies of C₃, C₄, and C₅.

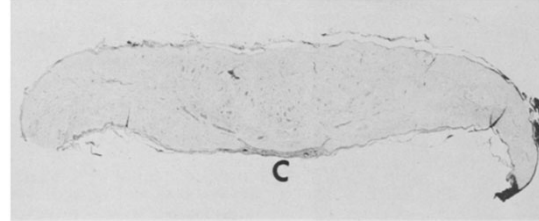


Fig. 3C. Section of spinal cord at the site of direct compression. There is a loss of virtually all the normal architecture of the spinal cord with replacement by astrocytosis. A few persistent myelinated tracts are barely visible in some areas. (Paraffin-embedded section; Luxol fast blue-PAS; $\times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

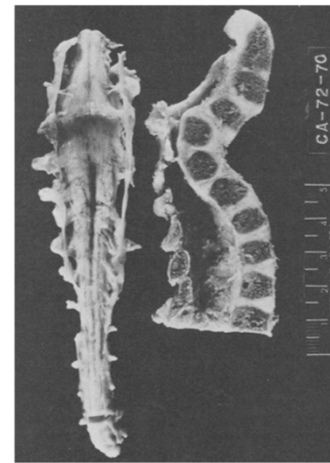


Fig. 2. Note the narrowing of the spinal canal at the region of spondylolisthetic slip and the corresponding compression of the spinal cord.

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

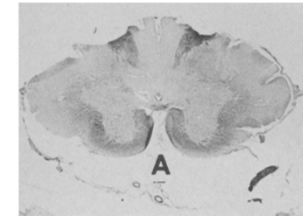


Fig. 3A. Section of spinal cord above the site of compression; ascending wallerian degeneration is present. (Paraffin-embedded section; Luxol fast blue-PAS; $\times 7.5$.)

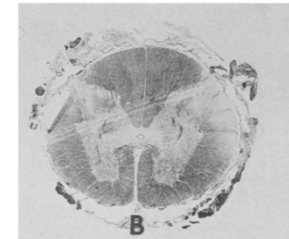
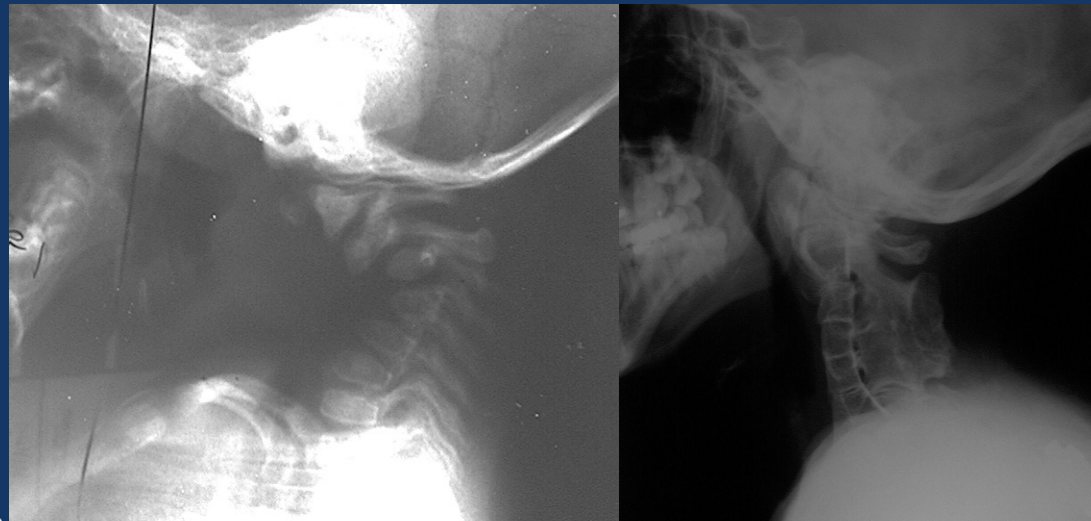
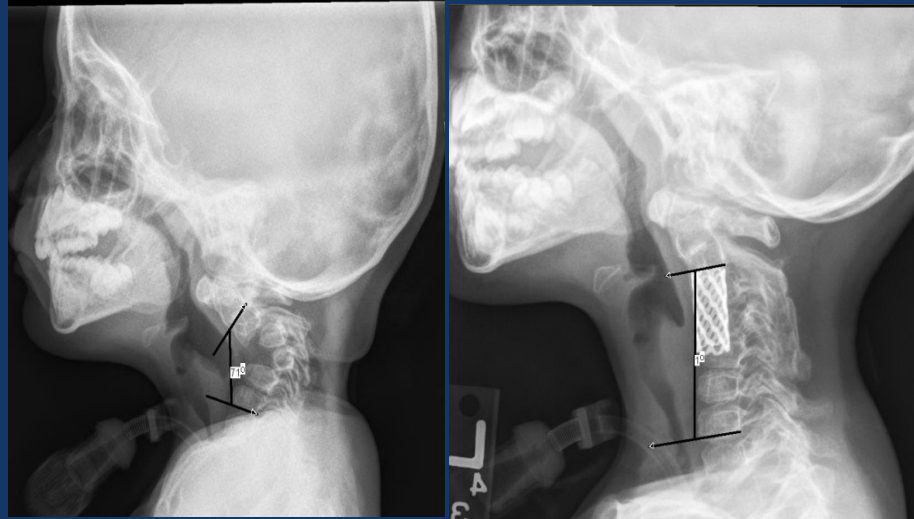


Fig. 3B. Section of spinal cord below the site of compression; descending wallerian degeneration is present. (Paraffin-embedded section; Luxol fast blue-PAS; $\times 7.5$.)

3 years old died of respiratory infection, quadriparetic

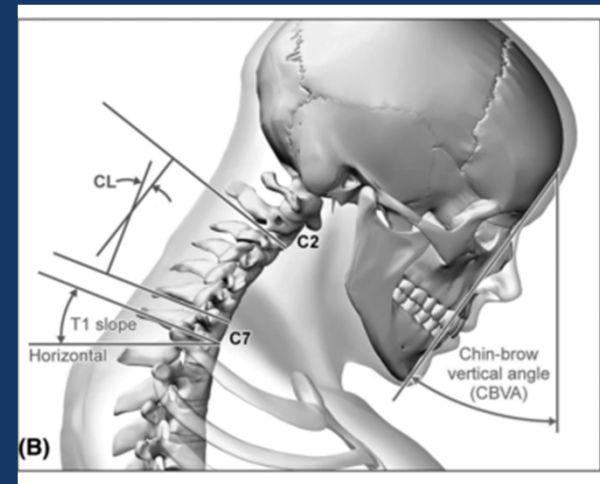
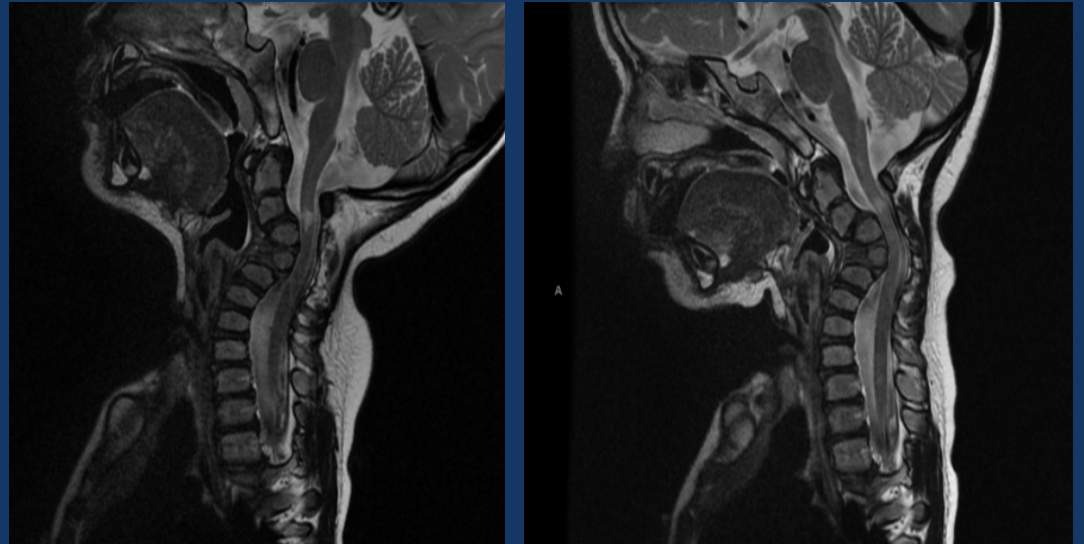
Methods

- 8 children with Diastrophic Dysplasia who underwent cervical spinal surgery
 - 4 with anterior vertebrectomy and 360° fusion
 - 4 with posterior only fusion



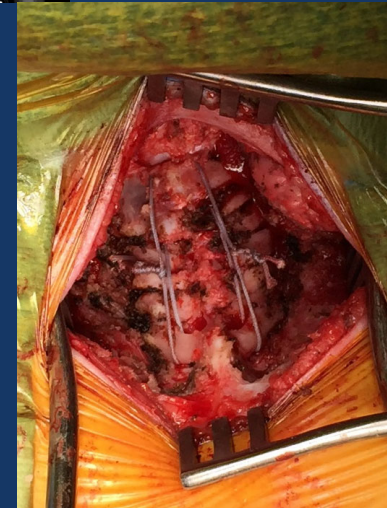
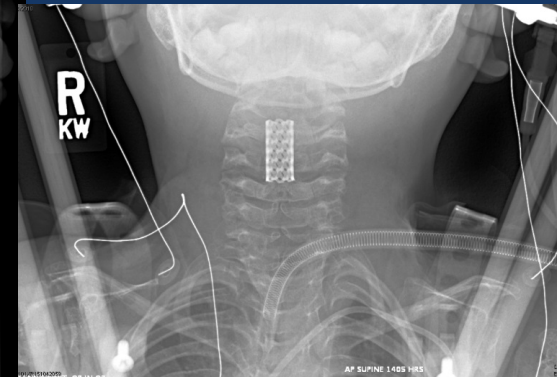
Methods

- Flexion/Extension MRI studies displayed:
 - Cord compression
 - Cord compression + myelomalacia
 - Cervical spine instability
- Plain radiographic measurements included cervical kyphosis Cobb angle from C2-C7
- T1 slope



Results

- The average age at the time of surgery:
 - Anterior 40 months
 - Posterior 55 months
- Surgical indication: symptomatic cord compression, spinal instability, severe or progressive kyphosis
 - All of the anterior pts had symptomatic cord compression, 2 with myelomalacia seen on MRI
 - 3 of the posterior pts had instability, one with progressive kyphosis



Results

| Subject ID | Procedure | Gender | Age at surgery (mo) | Indication for Surgery | EBL (mL) | Transfusion (mL) | Halo Post-operatively | Complications | Outcome |
|------------|-----------|--------|---------------------|------------------------------------|----------|------------------|-----------------------|--|---------------|
| 1 | A | F | 56 | Cord compression with myelomalacia | 250 | 130 | Y | None | Healed fusion |
| 2 | A | M | 24 | Cord compression | 150 | 189 | Y | None | Healed fusion |
| 3 | A | M | 39 | Cord compression with myelomalacia | 275 | 160 | Y | Dural tear during posterior portion. Erythematous halo pin-sites | Healed fusion |
| 4 | A | M | 41 | Cord compression | 500 | 297 | Y | None | Healed fusion |
| 5 | P | F | 13 | Cervical instability C3-C4 | 50 | 0 | Y | None | Healed fusion |
| 6 | P | M | 115 | Cervical instability C2-C3 | 300 | 0 | Y | None | Healed fusion |
| 7 | P | F | 62 | Cervical instability C2-C3 | 60 | 0 | Y | None | Healed fusion |
| 8 | P | M | 28 | Progressive kyphosis C2-C7 | 100 | 0 | Y | None | Healed fusion |

A=Anterior, P=Posterior, M=Male, F=Female, Y=Yes, N=No, mL=milliliters, mo=months

Table 1: Patient Demographics

| Subject ID | Pre-op C2-C7 Cobb (degrees) | Post-op C2-C7 Cobb (degrees) | Pre-op T1 Slope (degrees) | Post-op T1 Slope (degrees) |
|------------|-----------------------------|------------------------------|---------------------------|----------------------------|
| 1 | 71 | 1 | 12 | 8 |
| 2 | 111 | 11 | -43 | 18 |
| 3 | 88 | 26 | 14 | 24 |
| 4 | 91 | 43 | -21 | 18 |
| 5 | 66 | -30 | -14 | 31 |
| 6 | 8 | -1 | 26 | 23 |
| 7 | 5 | -12 | 22 | 25 |
| 8 | 74 | -16 | 30 | 24 |

A negative value indicates lordosis

Table 2: Cobb angles and T1 slope pre and post op

Results

- **All patients in the anterior group had more blood loss than posterior group**
 - Cell saver used in anterior group to help decrease volume loss and blood was returned
- **All patients were placed in a halo post operatively**
- **All patients healed fusion mass**
- **Complications:**
 - One pt with dural tear (repaired)
 - Same patient developed erythema of halo pinsites (treated successfully)

Conclusions

- There are many ways to address cervical kyphosis in patients with Diastrophic Dysplasia
- Spontaneous resolution is common
- Posterior fusion results in progressive correction due to posterior tethering and continued growth if pre-op spine is flexible
- Rigid kyphosis with anterior cord compression \pm myelomalacia can be safely addressed with this technique of anterior corpectomy and 360° fusion

An aerial photograph of the Alfred I. duPont Hospital for Children. The hospital is a large, multi-story building with a modern design, featuring a prominent section with a blue and white geometric pattern. It is surrounded by lush green trees and a large parking lot filled with cars. In the background, a city skyline and a body of water are visible under a blue sky with scattered clouds.

Thank You