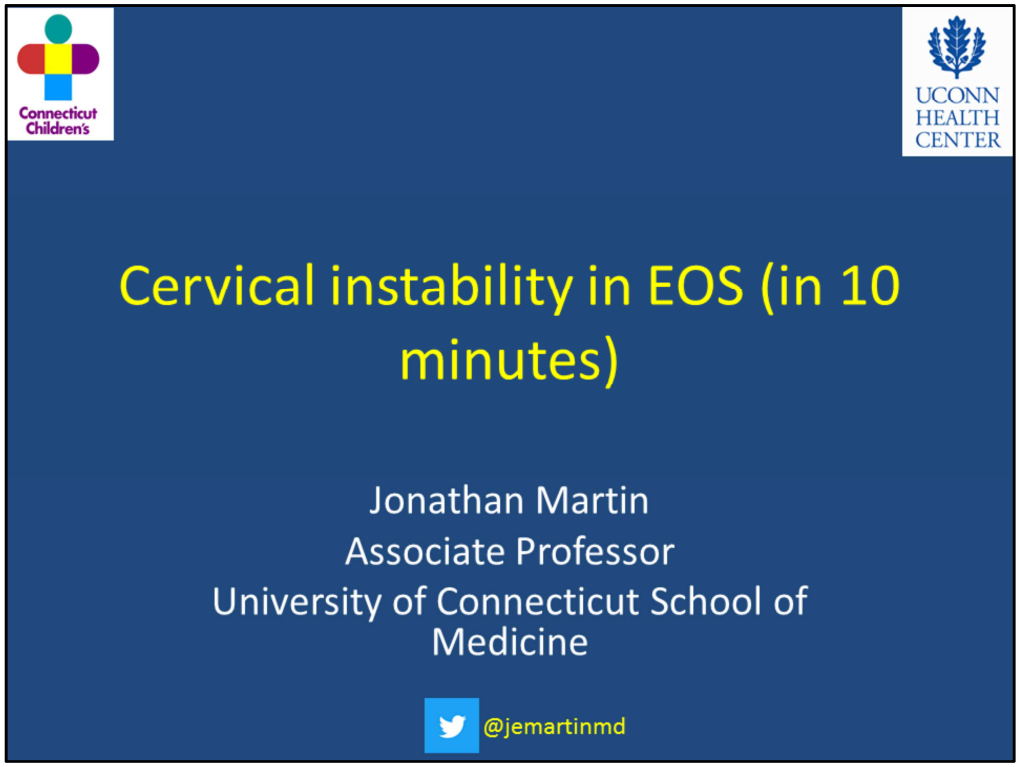
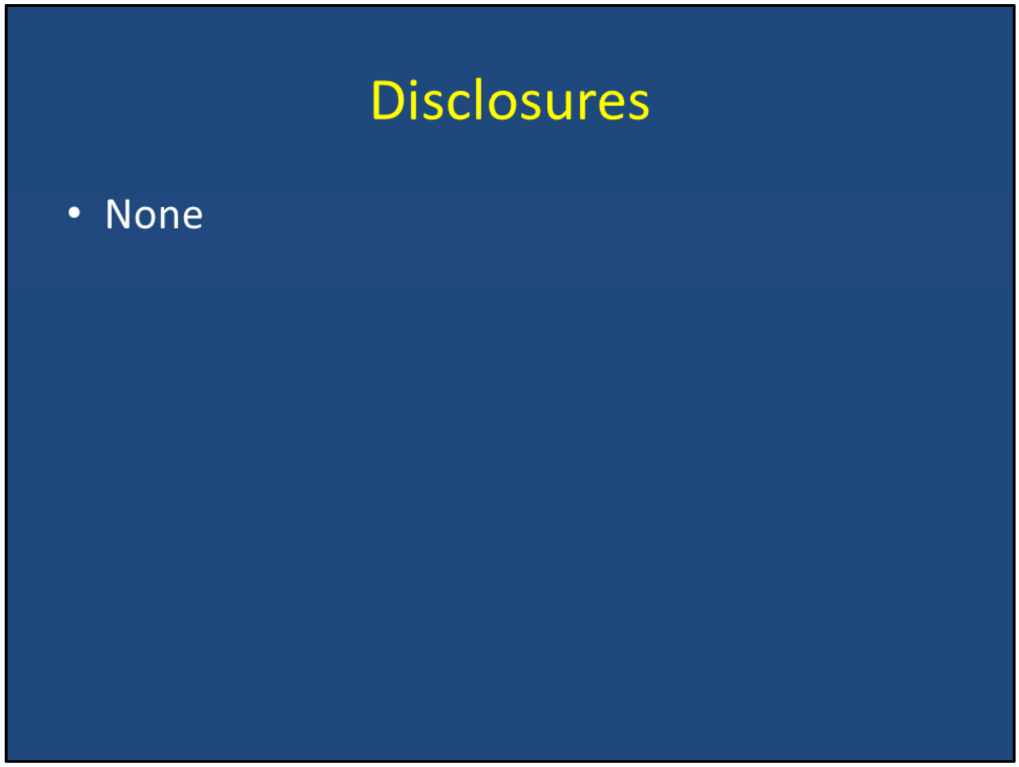
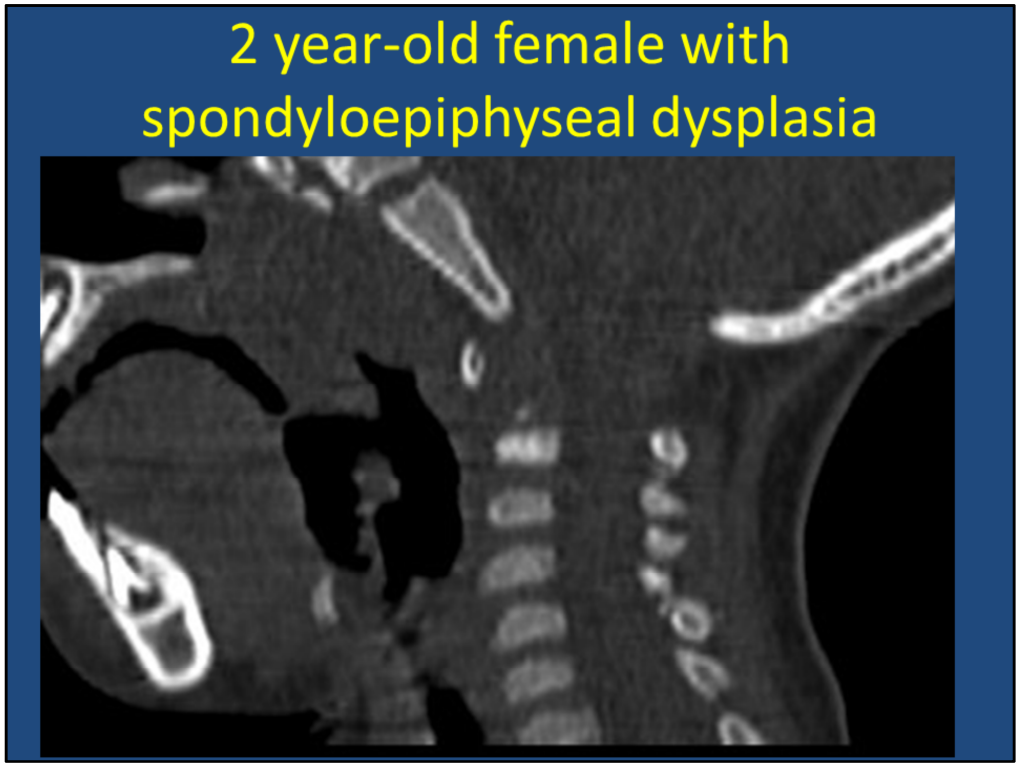



I appreciate the opportunity to speak here today




I have no disclosures



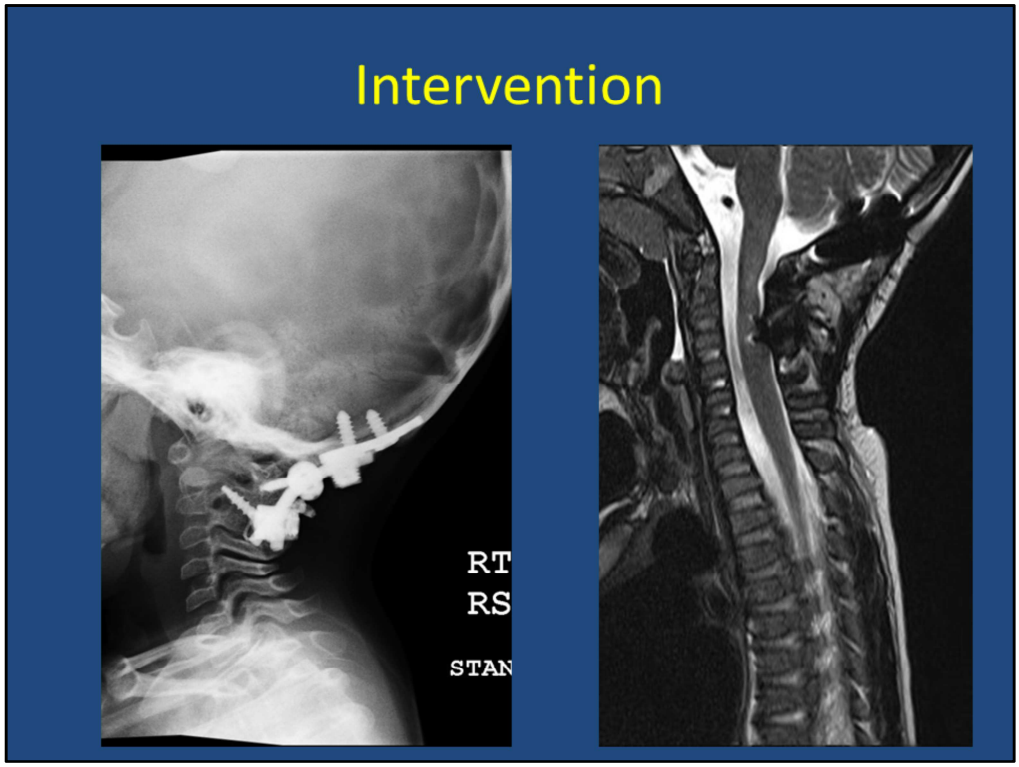

Let’s begin by considering this case of a child with spondyloepiphyseal dysplasia. She was referred to me as an infant for monitoring due to associated odontoid aplasia


And known atlantoaxial hypermobility, though I would challenge the group to quantify her laxity with a measurement. She had a baseline MRI, and was followed with serial clinical evaluation. We discussed use of a collar and the family declined. They were provided appropriate counseling regarding the need for a rear facing car seat for transportation.

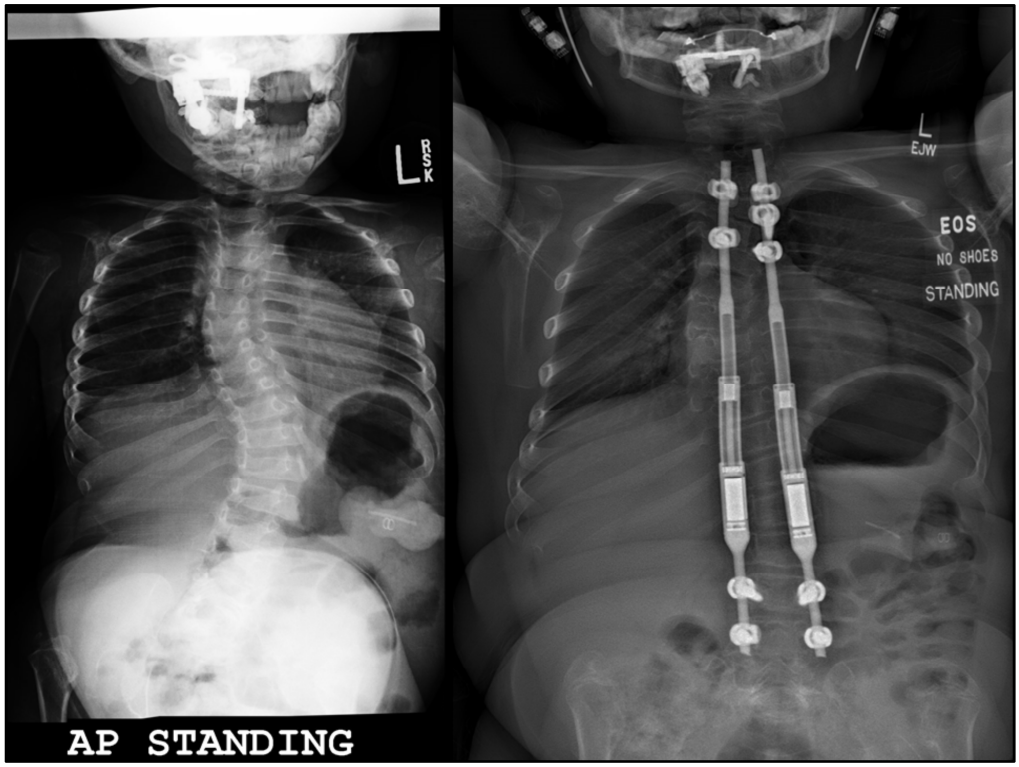
## Longitudinal follow-up

- 2 years of age- hyperreflexia
- MRI- gliotic change CCJ

At 2 years of age, she developed hyperreflexia; MRI demonstrated craniocervical junction narrowing with cord signal change




Prompting instrumented OC fusion. She has done well neurologically with no subsequent change in her exam




Yes, an EOS story with a ‘happy ending’, at least by orthopedic standards

# Cervical instability in EOS

- Abnormalities of formation (aplastic odontoid, congenital kyphosis)
  - SED, MPS patients (Morquio MPS-4) with aplastic odontoid, Larsen
- Ligamentous laxity
  - Marfans/EDS
- Secondary deformity
  - NF-1

Exacerbated by stenosis

The case illustrates some of the challenges with definitions of cervical instability, and frames a multidimensional approach to these patients

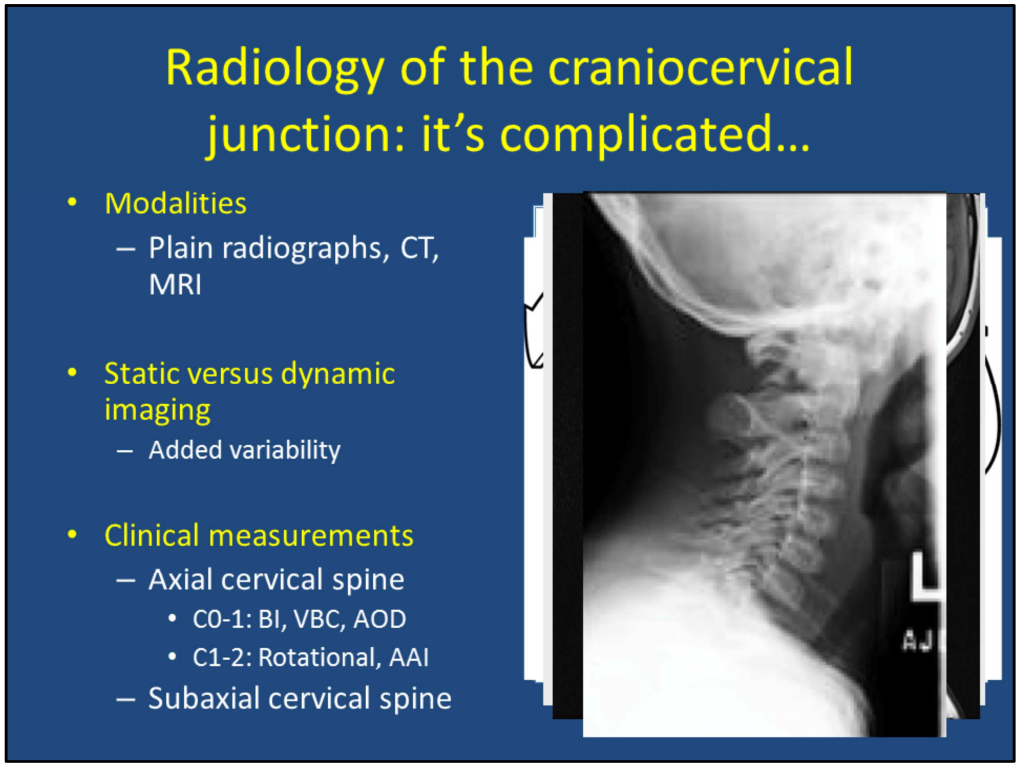
They are challenging- that's why we are sitting here. They are often structurally different due to structural deficiencies of bone or ligament. In some cases, this can be accelerated by what we as a healthcare team do to the. For example, laminectomy for resection of an intradural mass in an NF-1 patient

## Cervical stability defined

- **Spinal stability**: the ability of the spine to limit displacement under physiologic loads in order to prevent *pain*, ***deformity***, and *neurologic injury*
- Focus on defining radiographic instability
  - Challenges

I'm sure the group is familiar with this general definition of spinal stability

When it comes to the determination of cervical spine stability, there is typically a large focus on radiographic definitions of instability. These can be challenging in any patient population.




Why? Well for starters, we have multiple modalities to look at, each with its own strength and weakness in their ability to demonstrate regional anatomy *CLICK* *CLICK*

We can add in additional variables by introducing dynamic movement of the spine *CLICK*

And when we begin to set parameters on the static and dynamic 'stability' of the spine in an area with such potential for rotational and translational movement, things get messy fast. *CLICK*

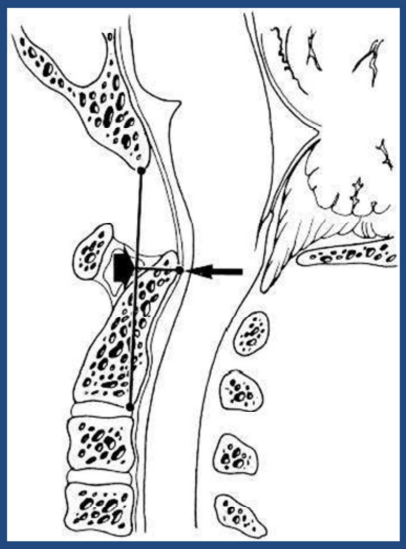
Occipital-atlantal instability
- Basilar invagination/impression
- Ventral brainstem compression
- Atlanto-occipital dislocation *CLICK*

Atlanto-axial instability
- Rotational C1-2 instability
- Atlanto-axial translational instability *CLICK*

Subaxial angular and translational instability

## The problem with measurements

- Small magnitude, high consequence measurements
- Most measurements have not been validated for inter-rater reliability
- Should they be rigidly to guide clinical management?



Underlying the inherent complexity of imaging assessment is the elephant in the room regarding measurement fidelity. Most of these measurements are small in magnitude and large in consequence, yet few have be validated for the reliability of their performance. This raises the question: should they be used to rigidly define patient management?

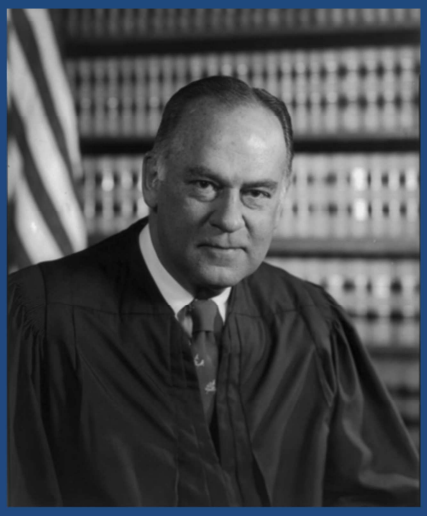
## Further challenges with cervical spine measurements in EOS

- Particularly problematic in EOS when required landmarks for measurement can be challenging to see!

These challenges are magnified in EOS patients, who have landmarks that can be challenging to see. I'll return to our SED friend for illustrative purposes

## ‘Potter Stewart’ Criteria

- Jacobellis vs Ohio 1964
  - Ohio pursued ban on public viewing of ‘The Lovers’
- Stewart provided majority opinion, with telling quote regarding the definition of pornography: ‘I know it when I see it’
  - Utility in illustrating CI in EOS?



Allow me to regress for a moment and provide a historical anecdote for perspective.

United States Supreme Court decision handed down in 1964 involving whether the state of Ohio could, consistent with the First Amendment, ban the showing of the Louis Malle film *The Lovers* (*Les Amants*), which the state had deemed obscene.

The most famous opinion from *Jacobellis*, however, was Justice Potter Stewart's concurrence, stating that the Constitution protected all obscenity except "hard-core pornography".

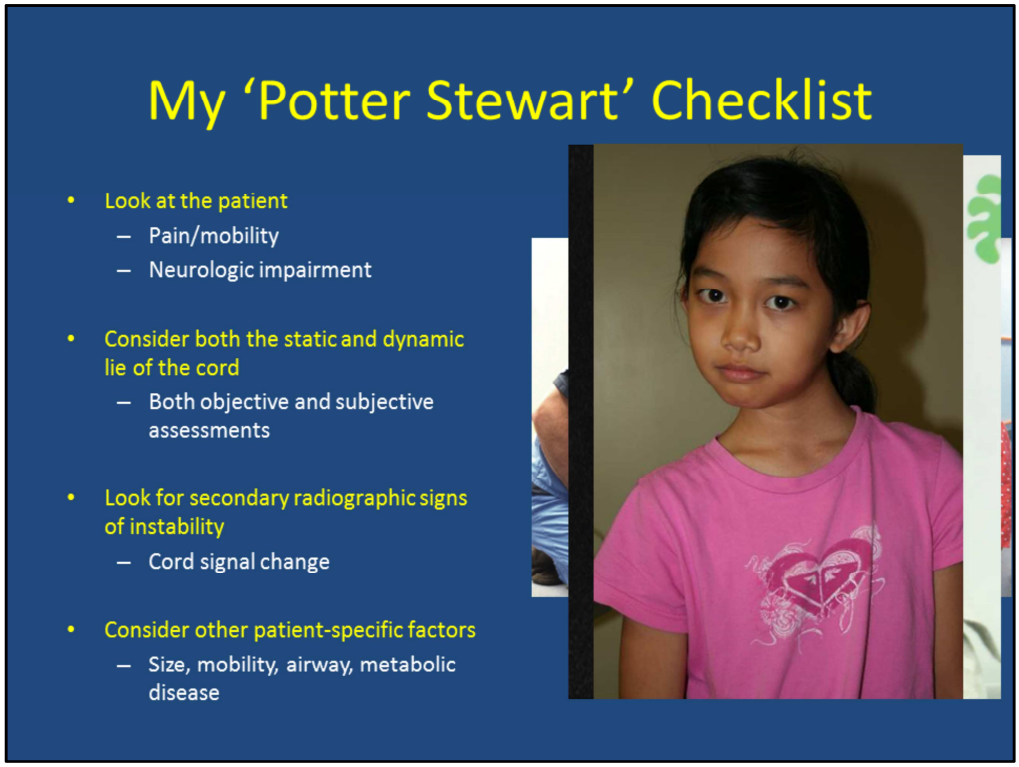
# White/Panjabi Checklist for Instability as a model

- General
  - Spinal elements destroyed
  - Positive stretch test
- Radiographic mobility
  - Translation > 3.5mm / > 20%
  - Greater 11 degrees angulation
- Spinal canal diameter
  - Pavlov's ratio <0.8
- Neurologic impairment

**Checklist for the Diagnosis of Clinical Instability in the Middle and Lower Cervical Spine**

| Element | Point Value |
|---|---|
| Anterior elements destroyed or unable to function | 2 |
| Posterior elements destroyed or unable to function | 2 |
| Positive stretch test | 2 |
| Radiographic criteria* | 4 |
| A. Flexion/extension x-rays | |
| 1. Sagittal plane translation > 3.5 mm or 20% (2 pts) | |
| 2. Sagittal plane rotation > 20° (2 pts) | |
| OR | |
| B. Resting x-rays | |
| 1. Sagittal plane displacement > 3.5 mm or 20% (2 pts) | |
| 2. Relative sagittal plane angulation > 11° (2 pts) | |
| Abnormal disc narrowing | 1 |
| Developmentally narrow spinal canal | 1 |
| 1. Sagittal diameter < 13 mm | |
| OR | |
| 2. Pavlov's ratio < 0.8† | |
| Spinal cord damage | 2 |
| Nerve root damage | 1 |
| Dangerous loading anticipated | 1 |
| **Total of 5 or more = unstable** | |

Back to the problem at hand, I've borrowed a multi-model model for the determination of spinal instability from White and Panjabi, one that incorporates clinical and radiographic variables to consider the question of instability



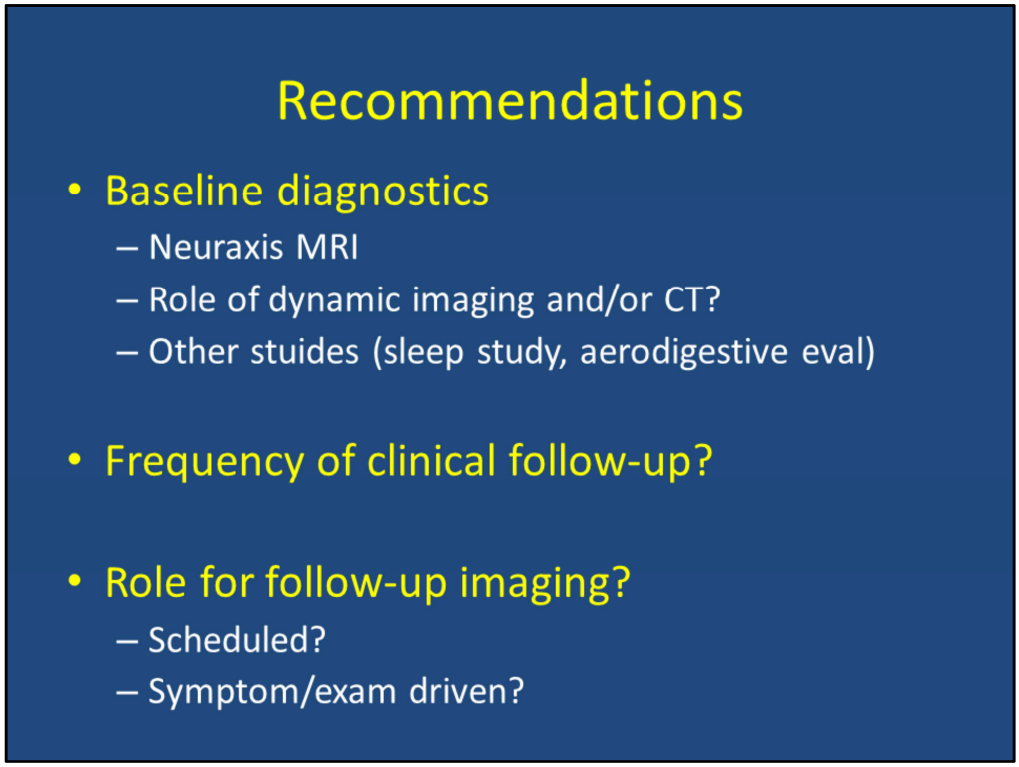

And applied it through a 'Potter Stewart' lens

Starting with the patient, with a focus on functional manifestations of spinal instability. *CLICK* Is the spine doing it's job for the patient? *CLICK*

Radiographic findings absolutely matter, even if the objective measurements don't always from a quantitative perspective *CLICK*

Qualitiative radiographic findings are equally important, perhaps more so in my algorithm *CLICK*

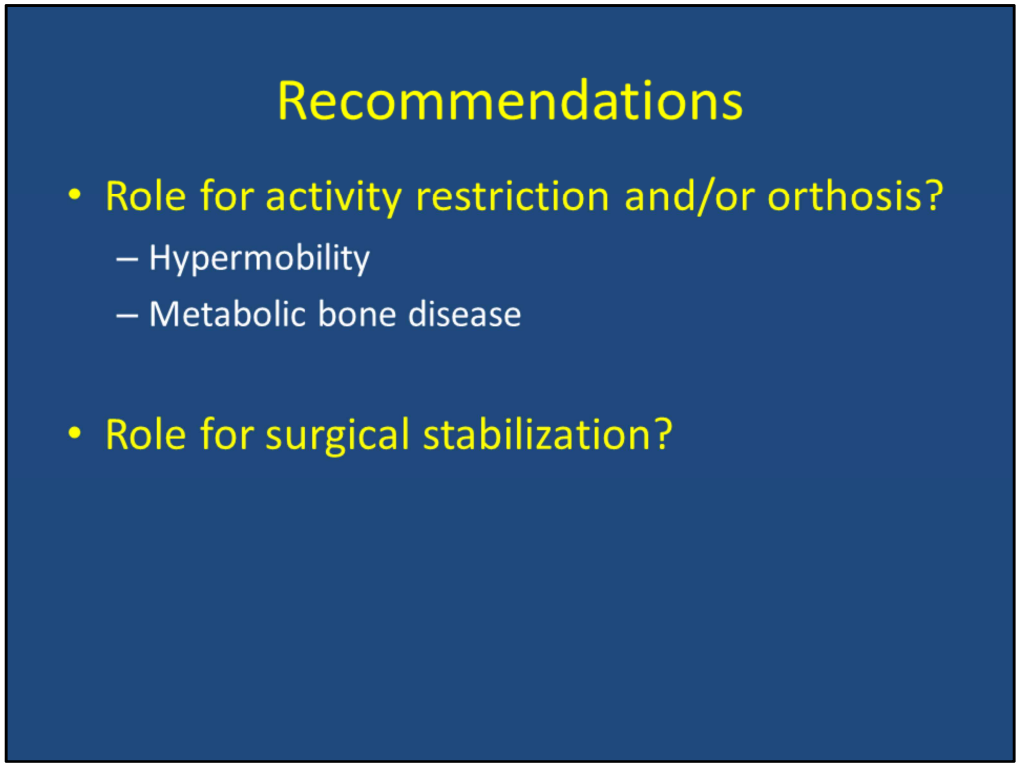
Finally, patient-specific factors profoundly influence how to proceed: observe? Treat? When? Why?




1) Baseline diagnostics: essentially all of these children require a baseline neuraxis MRI. It eliminates surprises, and allows for comparison if/when circumstances change. I'm less adamant about the need for dynamic imaging or CT outside of pre-operative planning. If it works for you, great. Based on the lack of established standards, most dynamic studies are interpreted subjectively. Sleep studies and aerodigestive evaluations are also key studies in patients with craniocervical junction pathology

2) Frequency of clinical follow-up: this is my work horse in these patients. 3-6 month interval follow-up visits are essential to me in this population, with a focus on review of systems and exam to target cranial neuropathy and myelopathy

3) I typically do not obtain scheduled follow-up cross sectional imaging based on the need for anesthesia in most of these patients




4) Activity restriction and orthotic use: I am relatively militant regarding common sense activity restrictions (climbing, tumbling) and liberal with prescribing orthotics to these patients if I have concerns for laxity. I'm sure this group is under no illusion that a collar is truly protective; it serves as a reminder to the patient and those around him/her and generally results in increase precaution. In the specific case of metabolic bone disease with basilar impression, orthosis is in my opinion the only effective solution we have to progressive occipito-cervical settling

5) And finally surgical stabilization: when all else fails, do it.

# Why does this talk matter?

Because the consequences when we fail to recognize an issue can be grave

# Questions?