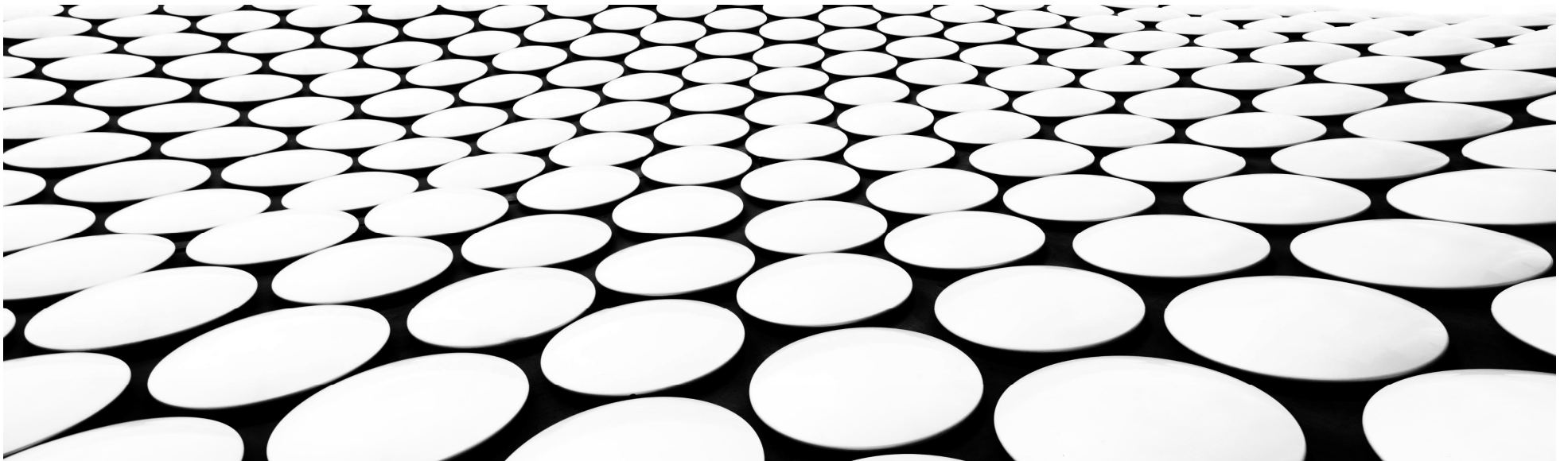




Response Evaluation In Neurofibromatosis Schwannomatosis INTERNATIONAL COLLABORATION

- If sharing any data or information from these slides generated by the REINS International Collaboration, please acknowledge the authors, group chairs, and specific working group.
- If using any information presented with a citation, please reference the primary source.

DYSTROPHIC SCOLIOSIS AND NF1: PARENT PERSPECTIVE.



LESSON 1 - SCOLIOSIS IN NF1 CAN DEVELOP FAST

LESSON 2 - CAN'T FUSE THE ENTIRE SPINE UNTIL KIDS ARE DONE GROWING

LESSON 3 - SECOND OPINIONS DON'T ALWAYS MAKE THINGS EASIER



May 2009 (Age 7)



Oct 2009



Dec 7, 2009 (Age 8)

Anterior Fusion

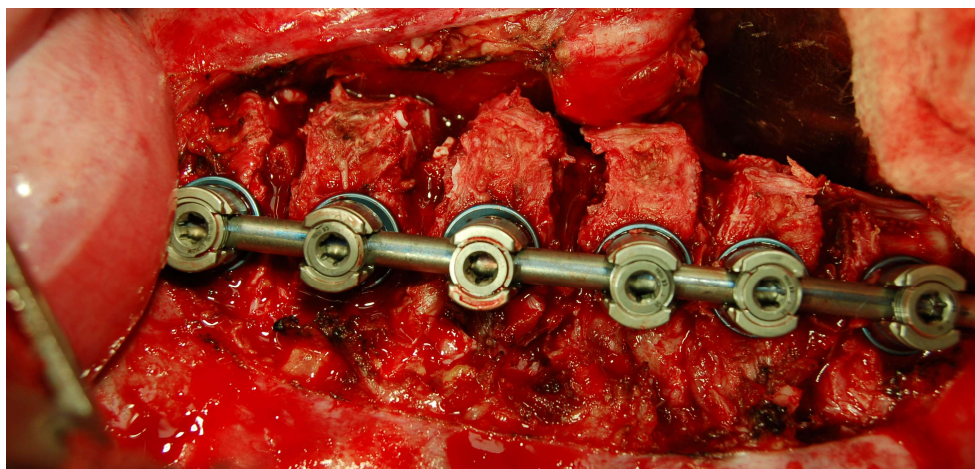
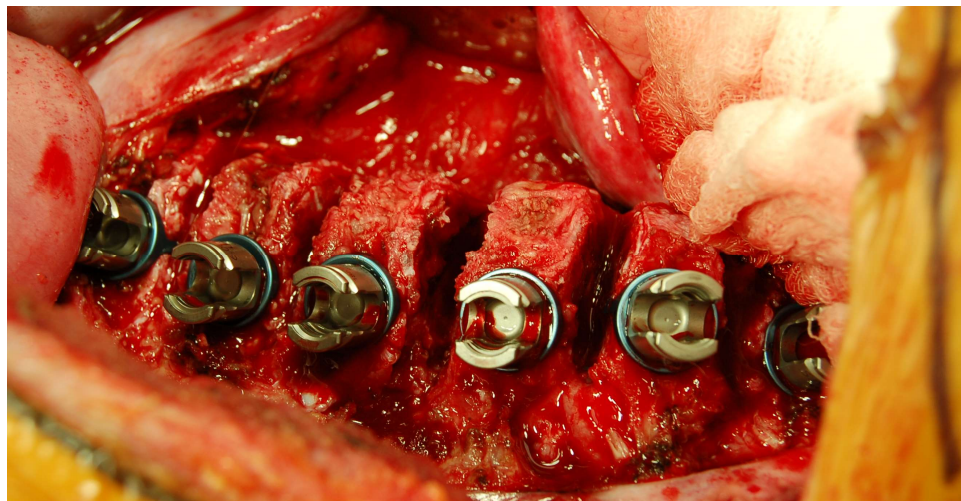
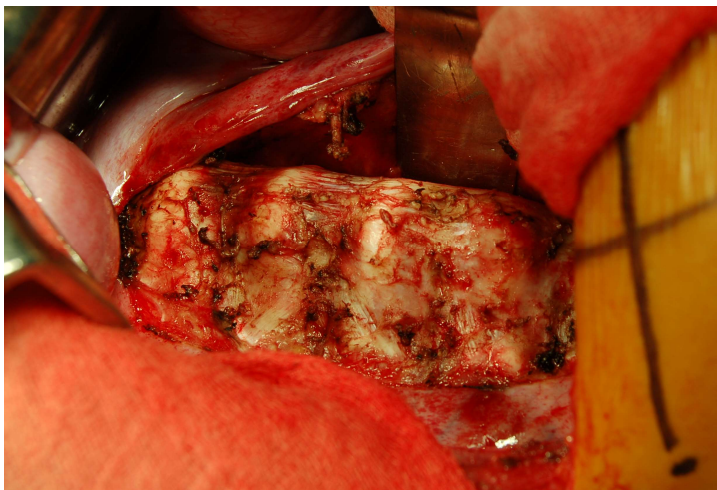
Our doctor

VS.

Growing Rods

2nd opinion

SPINE SURGERY IS INTENSE: ANTERIOR FUSION (DECEMBER 2009)



Surgical Access:
Chest wall
Remove a rib
Collapse the lung

DECEMBER 14, 2009: ANTERIOR FUSION T7-T12

POSITIVE RESULT



May 2009



Oct 2009



Dec 2009



Jan 2010

CHAPTER 2: NF1 SPINES DO WHAT THEY WANT

(Durotomy at T8 – Bone Chip Cut the Dural Sac)

Posterior Fusion: L3 to T3



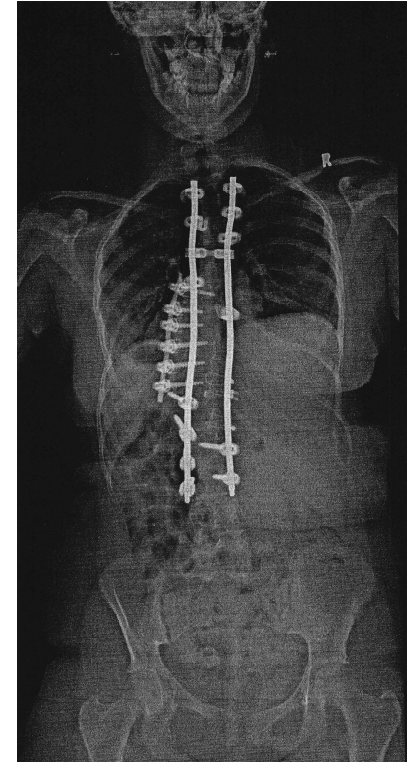
Jan 2010



Oct 2013



Dec 2013



2014→

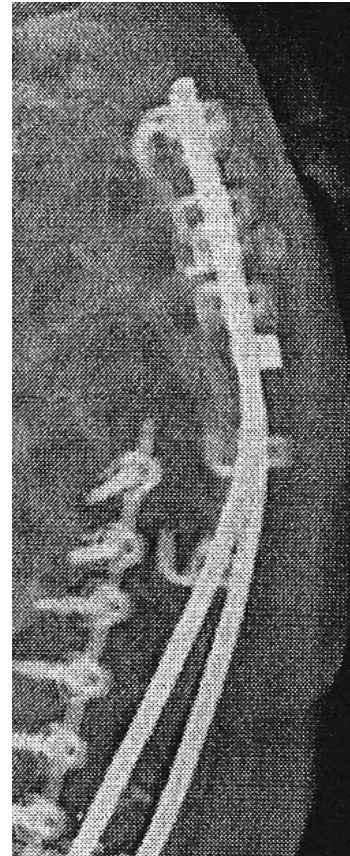
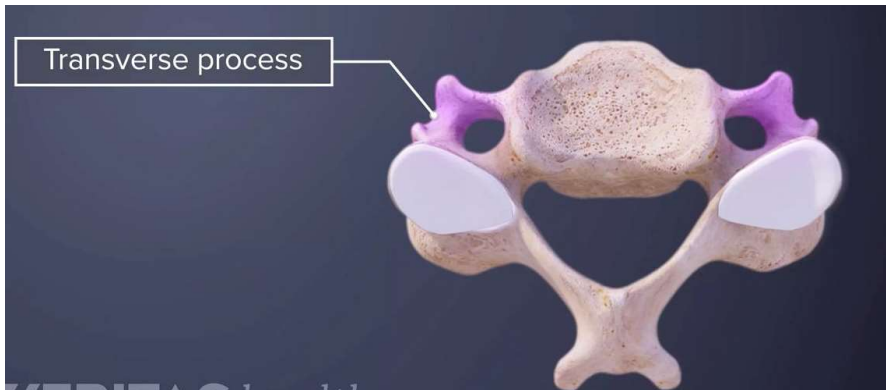
**PARASPINAL TUMORS:
COMMON, COMPLICATING, AND CONTRIBUTING FACTOR(?)**



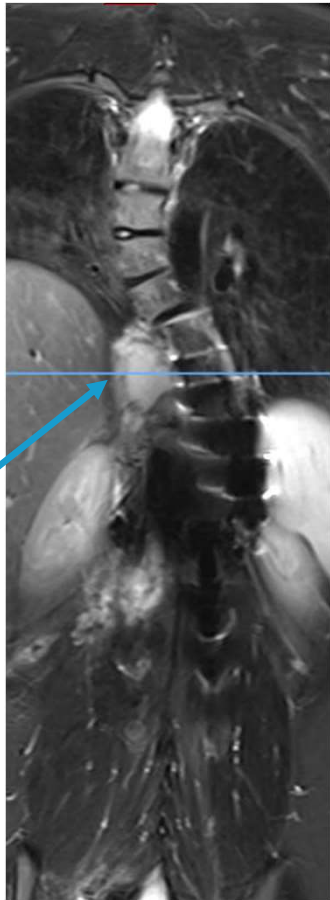
'Plexiform Tumor'

NF1 WEAK BONES – ANOTHER COMPLICATING FACTOR

- **Non-standard hardware due to bone strength**
 - Hooks instead of screws in upper spine
- **Impact: Upper back pain (4 mo. post fusion)**
 - Follow-on surgery (#3) to fix hardware issue
 - Doc: “Hook had pulled through the transverse process”

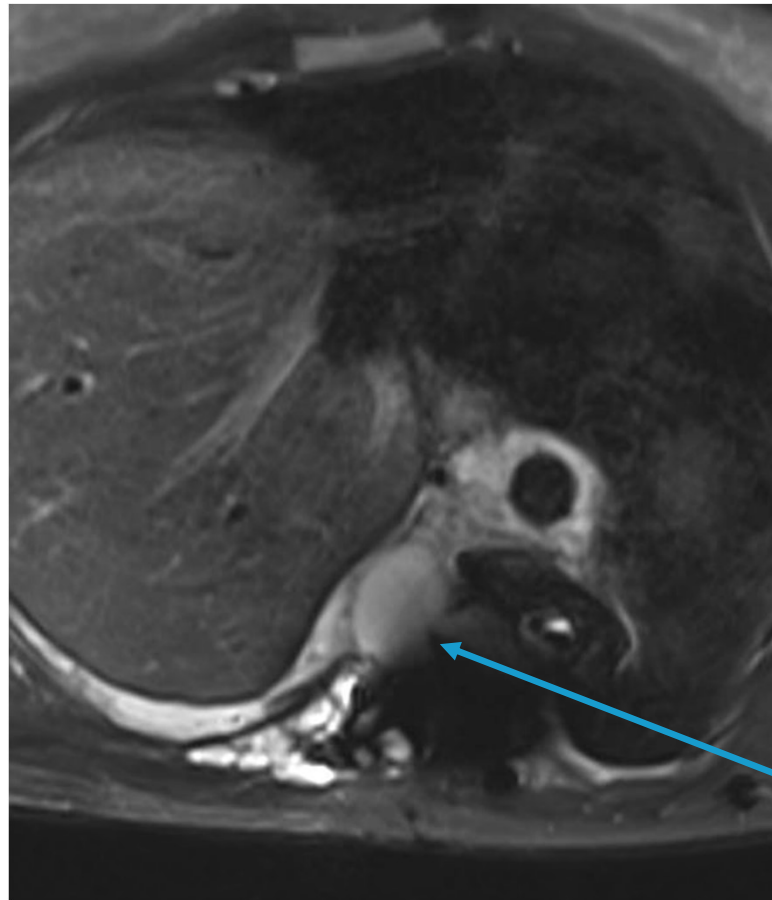


CHAPTER 3: NON-NF1 SURGICAL COMPLICATIONS CAN OCCUR



June,
2015

CT
Pet/MRI



Cause:

Durotomy 2013?
NF1 related?

Three years later (2018):

Upper back pain
Spine Surgery #4
Lost sensation waist down
Dx@NIH: Tethered spinal cord
Two more CT Scans
Spine (Cord) Surgery #5

'Pseudomeningocele'
Damaged Dural Sac @ T8

APPENDIX: OTHER BONE ISSUES → NON-OSSIFYING FIBROMA



Surgery #6

CUMULATIVE RADIATION EXPOSURE: 75+ X-RAYS; 3 CT SCANS; PET

Parent's Fear: Excess radiation of Plexiform Tumor (& healthy tissue) Creating 2nd hits --- Risk of cancer down the road

- Battle with X-Ray Technicians - "Cover her up more!"
- Battle with Spine Doc – "Do we absolutely need the x-ray this time? "Can we skip the side view?" "Can we do x-rays every other visit?" "Can we do 2nd one only if 1st shows issues?"
- EOS Machine – Newer technology, much less radiation

Unfounded fears? – 25mSV total; 8 yrs of background radiation

"Less radiation than a plane flight" "We might miss something...."

- Pathology of large tumor resected in 2016 showed signs of atypia (abnormal cells)
- Diagnosed with Hashimoto's – Thyroid auto-immune issue
- Future issues? Cause/Effect can be delayed by many years.



**EOS MACHINE
Micro-dose Radiation**

LESSON FROM JULIA: BRACES ARE “EVIL”



LESSON 99:

TRUST YOUR KIDS AS MUCH AS THE DOC

LESSON 100:

NF KIDS ARE REALLY TOUGH!