



# DYSTONIA AND MYELOPATHY

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# DYSTONIA AND MYELOPATHY

- 23 yo Dx at age 4 with CP
- Prior to dx mother states developing normal motor function
- Symptoms started in legs and progressed to arms then neck over a few years
- Dx later with dystonia by neurologist
- Baclofen pump placed 10 years ago with improvement to legs and some arms but not neck
- 2 yrs ago started developing weakness in legs and hands
- 2 mo prior to presentation had to use wheelchair and was using walker several months prior to this for weakness and balance
- Referred for DBS evaluation, no genetic studies but felt possible DYT1

# DYSTONIA AND MYELOPATHY



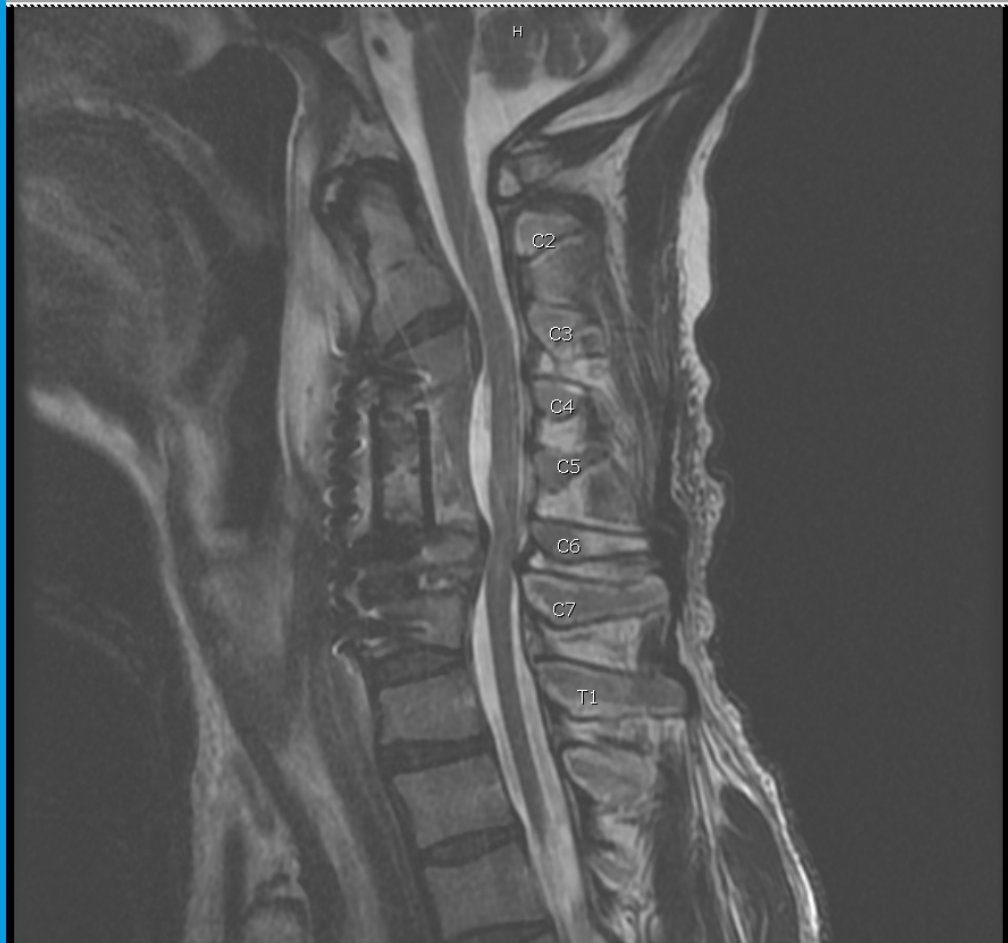
- Examination
  - Dysarthria but stable per mom
  - Dynamic left lateral collis
  - Contracture of arms and hands with dynamic components but able to move passively and some active
- Strength
  - UE 4/5 delt, bicep, WE, WF, HI
  - LE HF 4/5, difficult to assess PF and DF ow 5/5
- Sensory
  - Decreased proprioception ow normal
- Reflex
  - 2+ with no UMN signs



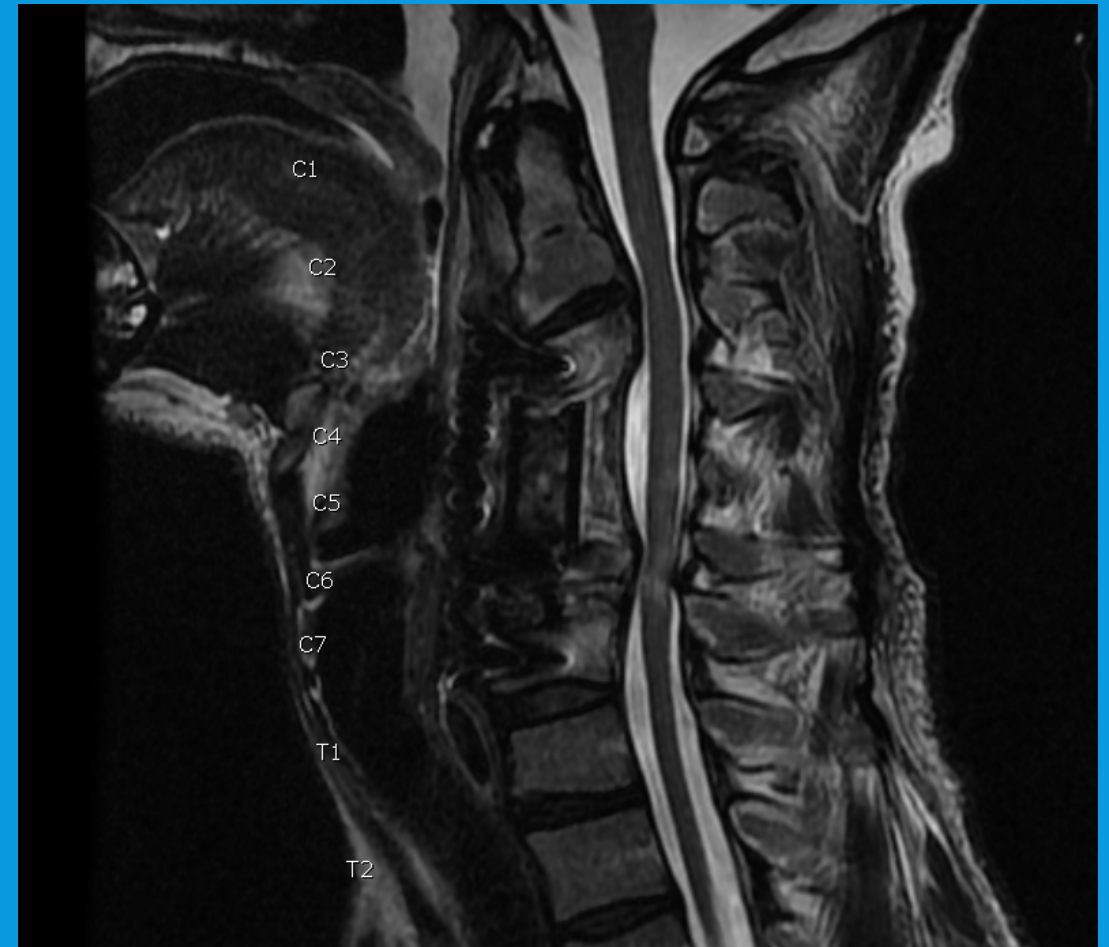
# DYSTONIA AND MYELOPATHY

- Operative
  - C<sub>4</sub>, C<sub>5</sub> corpectomy, C<sub>6-7</sub> ACDF
  - Plan for staged posterior stabilization
- Post operative
  - Very weak diffusely and somnolent
  - Baclofen decreased to 250 mcg/day from 300 mcg/day
  - Improvement in motor and cognition / alertness
  - Improved over 3 days from decrease
- Plan
  - At this time it was planned to optimize Baclofen levels and PT to improve motor function and gait
  - Plan to perform posterior cervical in 1 month
- Clinic follow up from rehab facility
  - Had been non ambulatory
  - Urinary retention and catheter placed
  - Complained of feeling too loose and weak

# DYSTONIA AND MYELOPATHY



Post Op day 1



Follow up



# DYSTONIA AND MYELOPATHY



X Table Portable

Post op Day 1



Follow up



# DYSTONIA AND MYELOPATHY

- Readmitted with repeat MRI demonstrating no new compression or cord signal
- Baclofen decreased to 100 mcg/day
- Urology consulted and felt atonic bladder, possible chronic and from Baclofen
- Over 3 days started to feel stronger
- Day 5 able to stand and ambulate with balance assistance and catheter removed and PVD 30 cc
- Plan for posterior stabilization
  - C2-T2 posterior
  - Case went uneventfully
- Post operative was ambulating but complaining of increased pulling in arms and legs and head
- Baclofen increased over 3 days to 200 mcg/day



# DYSTONIA AND MYELOPATHY



2 week post op





# DYSTONIA AND MYELOPATHY

- Follow up 2 weeks
  - Unsteady gait and feeling loose again
  - Strength in hands and legs at 5/5
  - Increased dystonic head torsion
  - Cervical xray demonstrate rod displaced from screw
- Plan
  - Revise posteriorly
  - DBS for dystonia
- Thoughts