

Neuromonitoring decision algorithm for improved safety in children with Hurler Syndrome undergoing non-spine surgery: Our experience

Careskey M, Kandil, A, Pettit C, Berry L, Chestnut E, Habeych M, Buck D, Leslie N, McAuliffe J, Chidambaran V



Background

- Patients with Hurler syndrome pose unique challenges to the anesthesiologist.
- Treatment with bone marrow transplants improve airway challenges;
- Spinal deformities place Hurler pts at risk for spinal cord ischemia (SCI) during non-spinal surgeries.¹
- Presence of kyphosis and stiff spines require increased attention to maintaining spinal cord perfusion in these patients
- Intraoperative neuromonitoring (IONM) is a useful modality for identifying and preventing irreversible SCI.

Methods

- We held multidisciplinary meetings (genetics, orthopedics, neurology, anesthesia, radiology),
- Reviewed studies noting significantly increased intramedullary pressure with critical kyphotic angles (4),
- Discussed the standard of care at other institutions
- Developed our own IONM decision algorithm (figure 2) for children with Hurler Syndrome undergoing non-spine surgery.

Our Care Algorithm for Anesthetic Care of MPS Patients

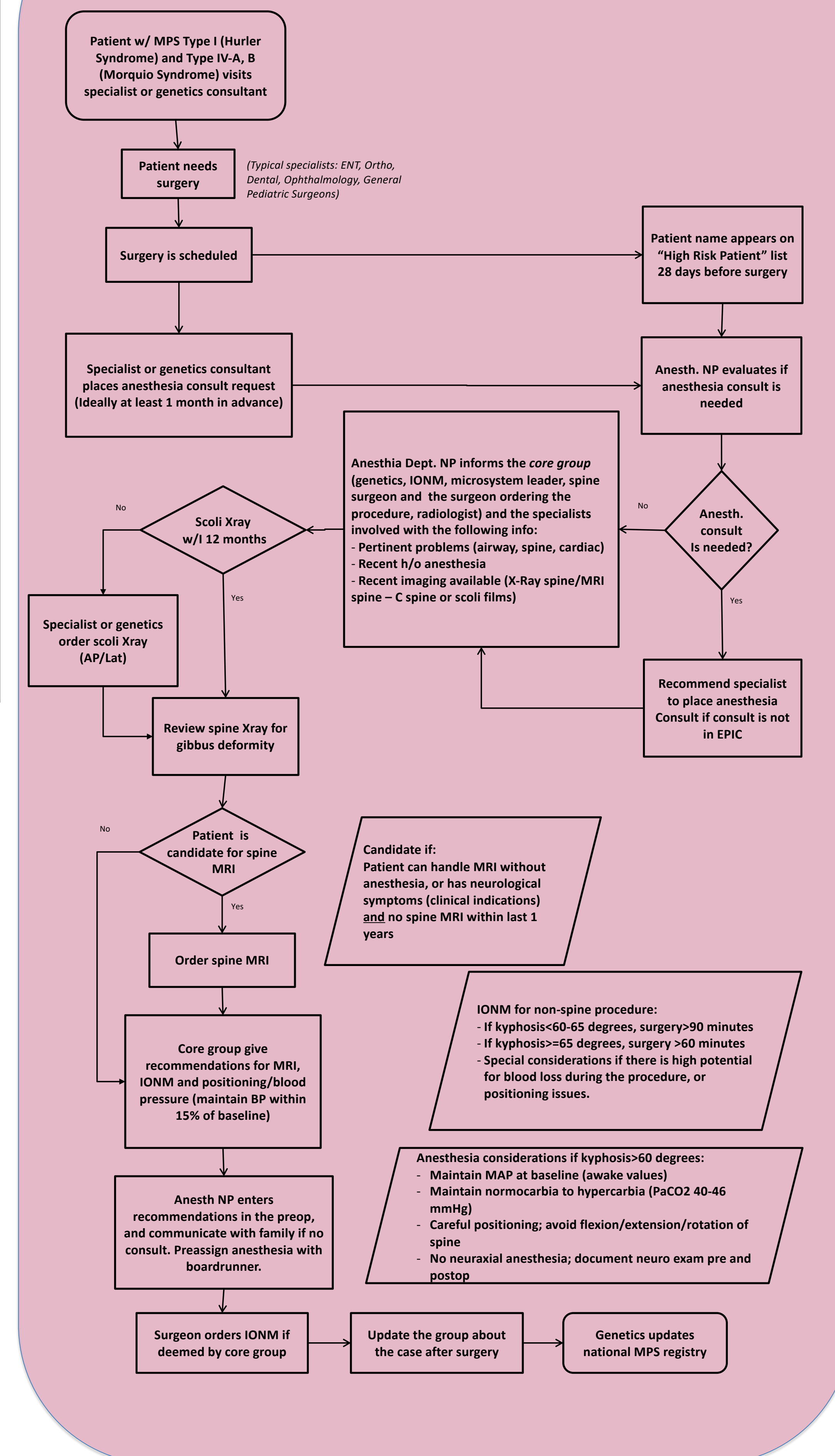


Figure 2) CCHMC's institutional algorithm for children with Hurler's undergoing non-spine surgery

Case Report- IONM Changes Affecting Intra-Op Management

Case

- 7yo, 24kg boy
- Hurler's Syndrome
- Severe Scoliosis s/p Correction
- Unilateral Pelvic Osteotomy

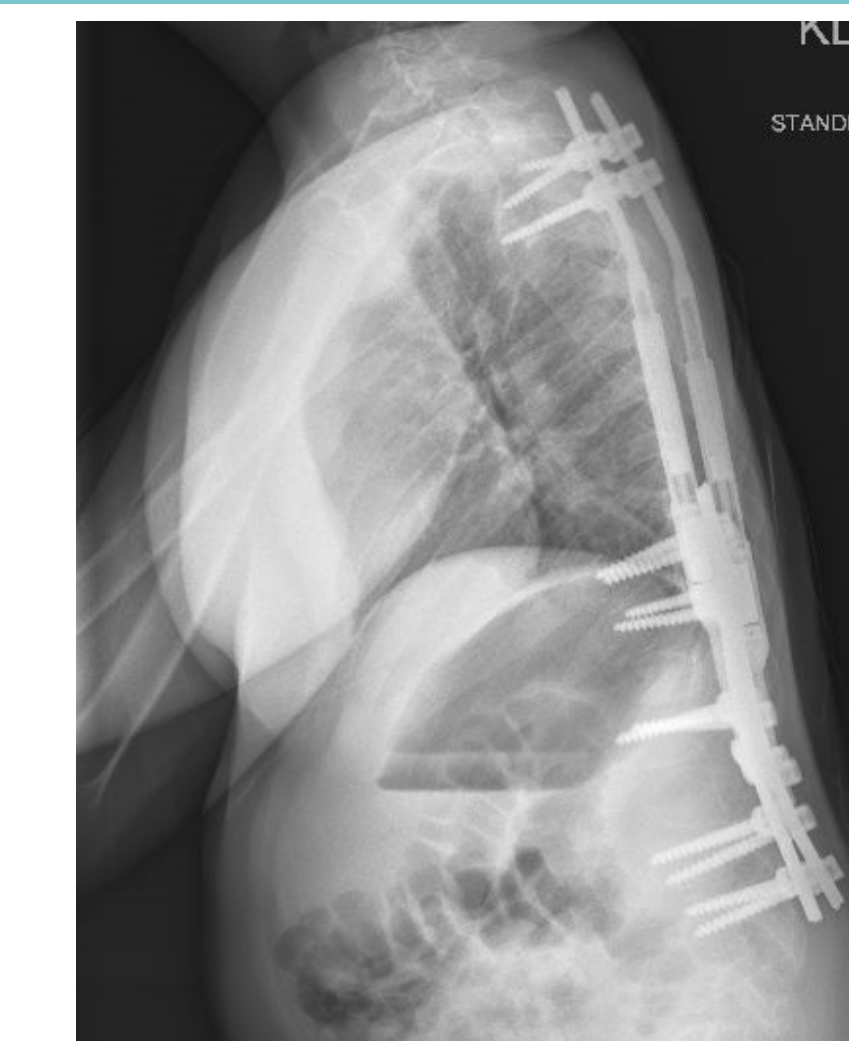
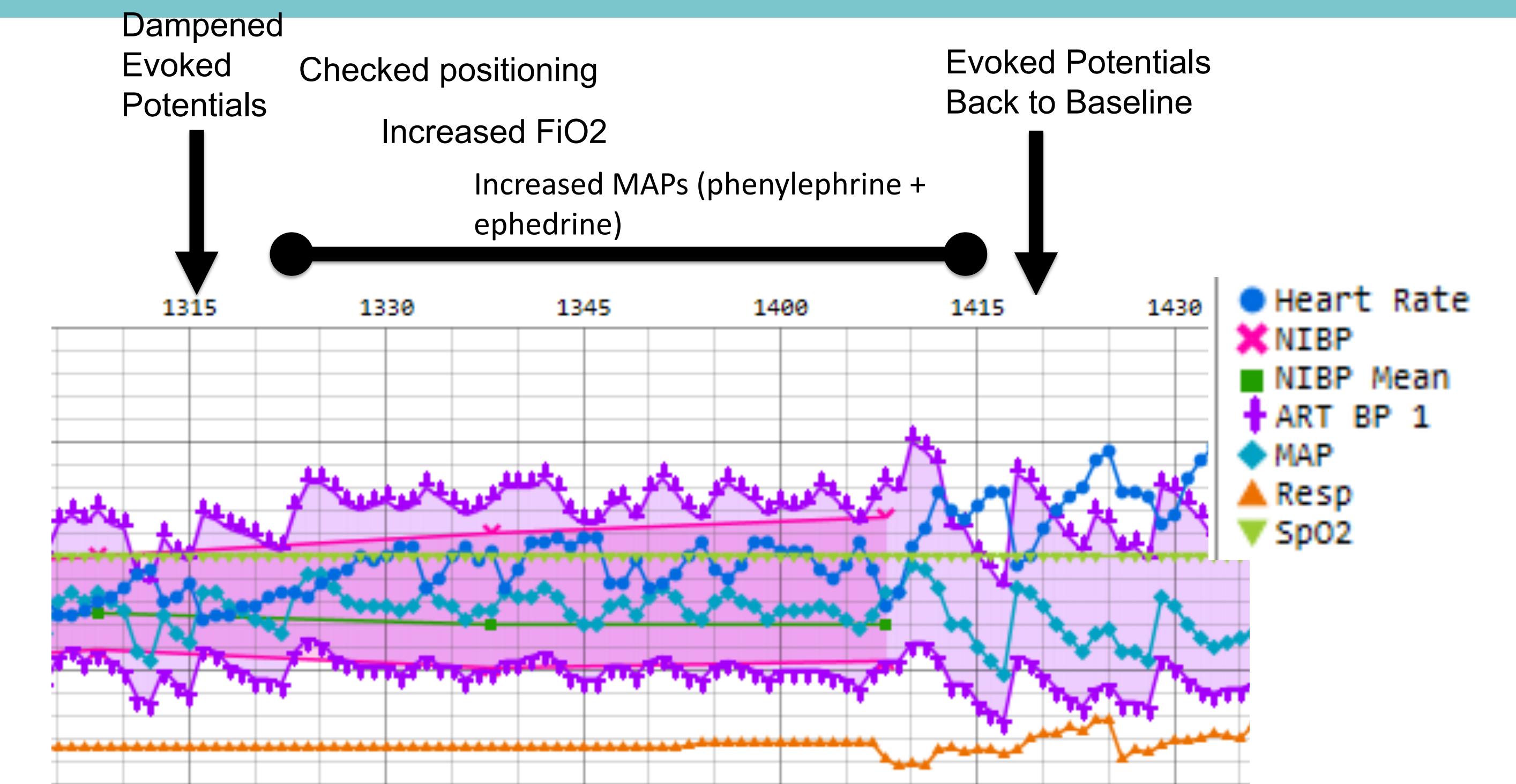


Fig 3) Pre-op Scoliosis X-ray- curvature previously corrected surgically

7 > 12 < 185
33



Post-op Course

- No neuro deficits
- D/C home on POD 3

Results & Discussion

- The decision tree in the developed algorithm is dictated by degree of kyphosis from scoliosis films and surgical risk (duration, positioning and blood loss) (Fig 1).
- Based on this algorithm, IONM was instituted for 17 children with Hurler's requiring non-spinal surgeries (Table 1).
- We detected changes in transcranial motor evoked potentials (TcMEP) and somatosensory evoked potentials (SSEP) in 2 patients, for whom we intervened intraoperatively, mostly with blood pressure management and re-positioning, and prevented sustained injury to the spinal cord and upper extremity.
- **None of these patients sustained neurological deficits.**

Conclusion

- IONM may add to anesthesia safety in children with Hurler's syndrome, based on risk-benefit profiles
- Neuraxial blocks should be avoided
- Epic anesthesia alerts may be beneficial in pointing to safety considerations

Anesthetic Considerations for Patients with Hurler Syndrome

Figure 1) Patient with Morquio Syndrome (MPS IV, similar to Hurler Syndrome)



Difficult Airway

Macrosomia
Large tonsils/adenoids
redundant tissue
Thoracic cage abnormalities
Limited TMJ movement

Cardiac Disease

Valvular disorders
Coronary disease
Arrhythmias
Systemic vasculopathy

Spine Abnormalities

Atlanto-axial instability
Severe kyphoscoliosis
Risk of Spinal Cord Ischemia
Restrictive lung disease

Variable	Mean±SD
Age (years)	7.8 ± 5.6
Duration of surgery (hours)	5.1 ± 2.7
Degree of Kyphosis	35.2± 26.3
Surgery Type (n=17)	%(n)
Hip Osteotomy	24% (4)
Dental rehabilitation	24% (4)
ENT Cases	13% (3)
Other Orthopedic Cases	35% (6)
IONM Changes	12% (2)

Table 1) Demographics and Perioperative Information of children with Hurler's syndrome who were monitored with IONM; IONM= Intraoperative Neuromonitoring (either somatosensory evoked potentials or transcranial motor evoked potentials)

References

1. Pruszczynski B, et al.. Clin Orthop Relat Res. 2015 Oct;473(10):3315-3320.
2. Tong CK, et al.. J Neurosurg Pediatr. 2012;9:608-612.
3. Othman Z, et al. Spine (Phila Pa 1976). 2004;29:E258-E265.