Intraoperative CT-guided Occipitocervical Decompression and Fusion for Mucopolysaccharidoses

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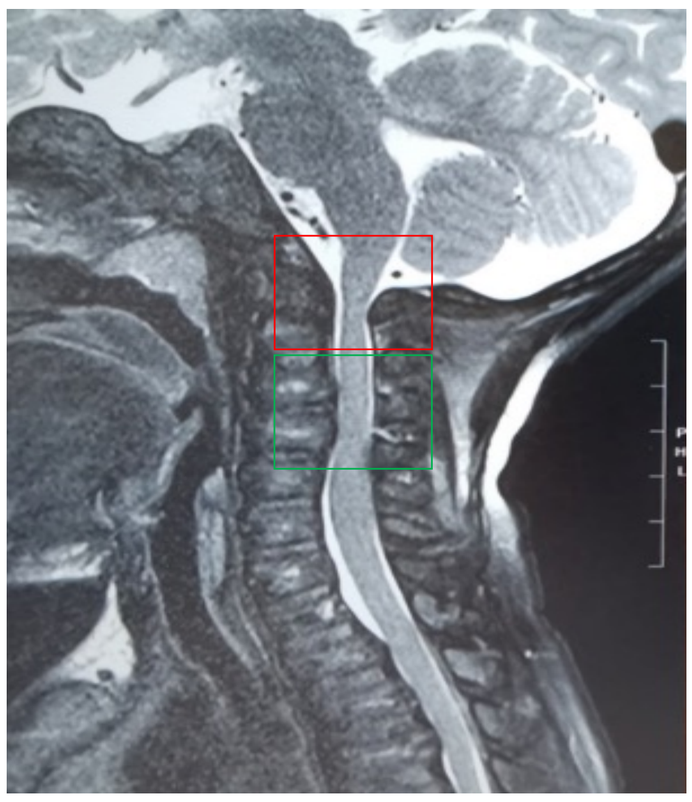
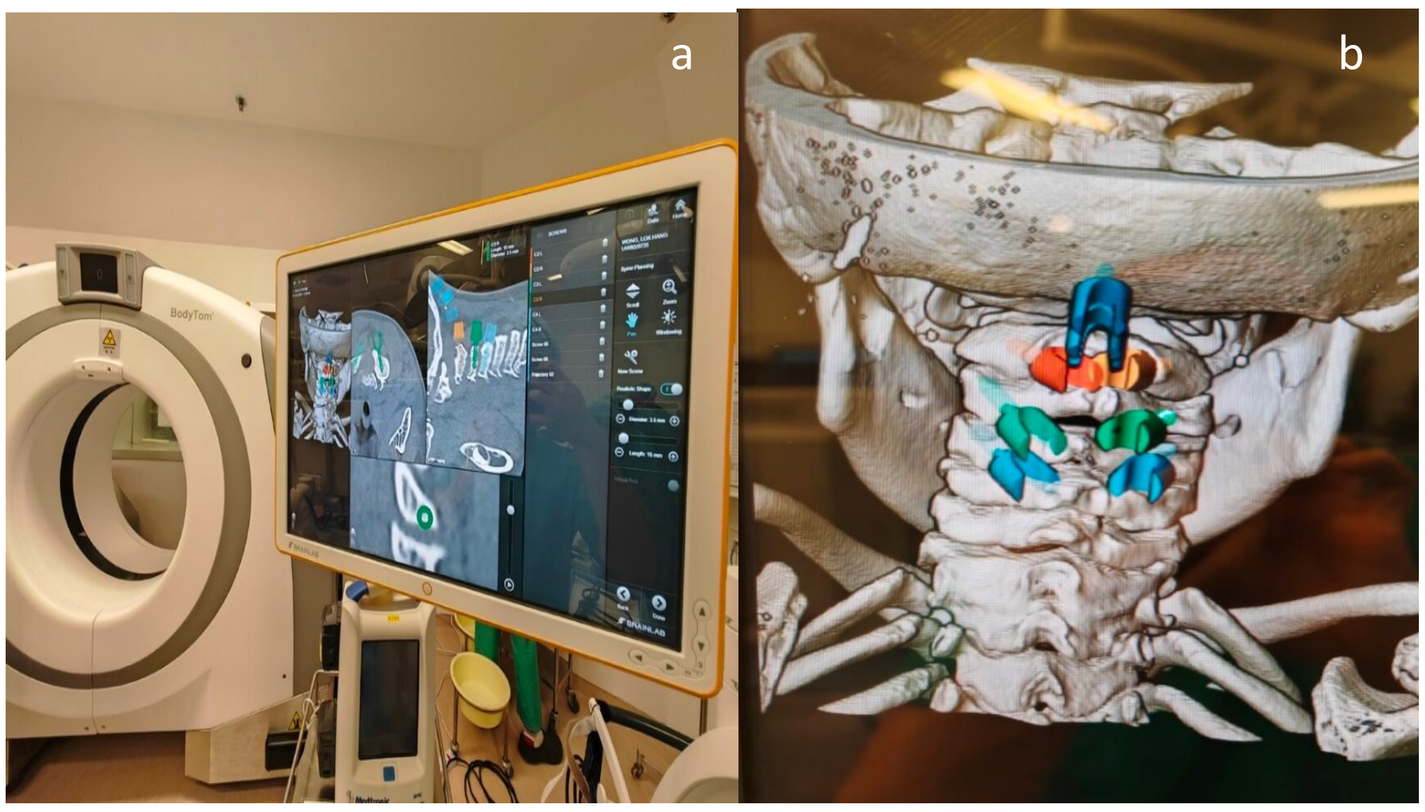
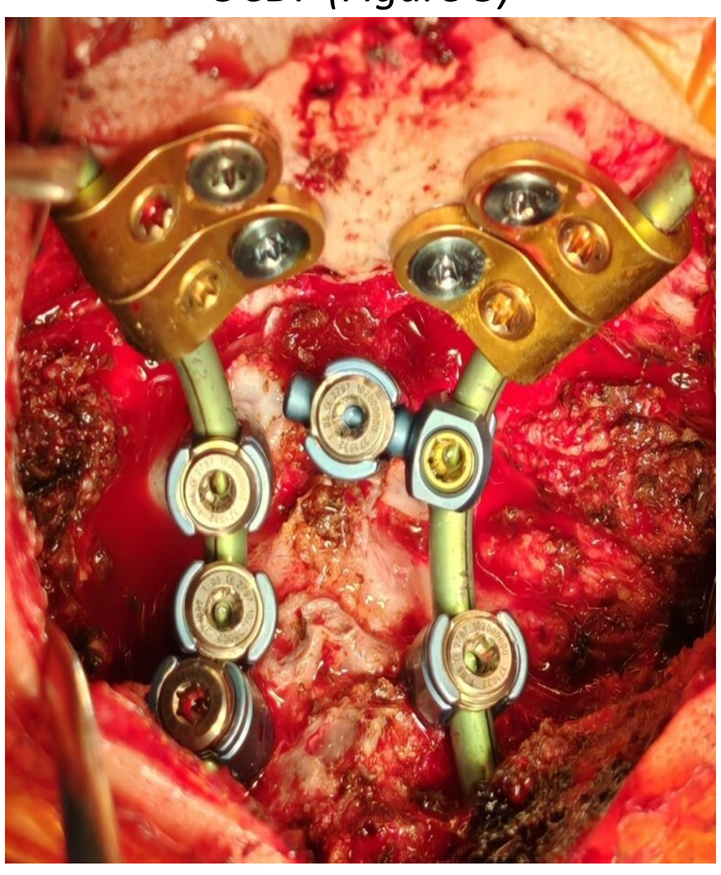
Pre-operative imaging (Figure 1)



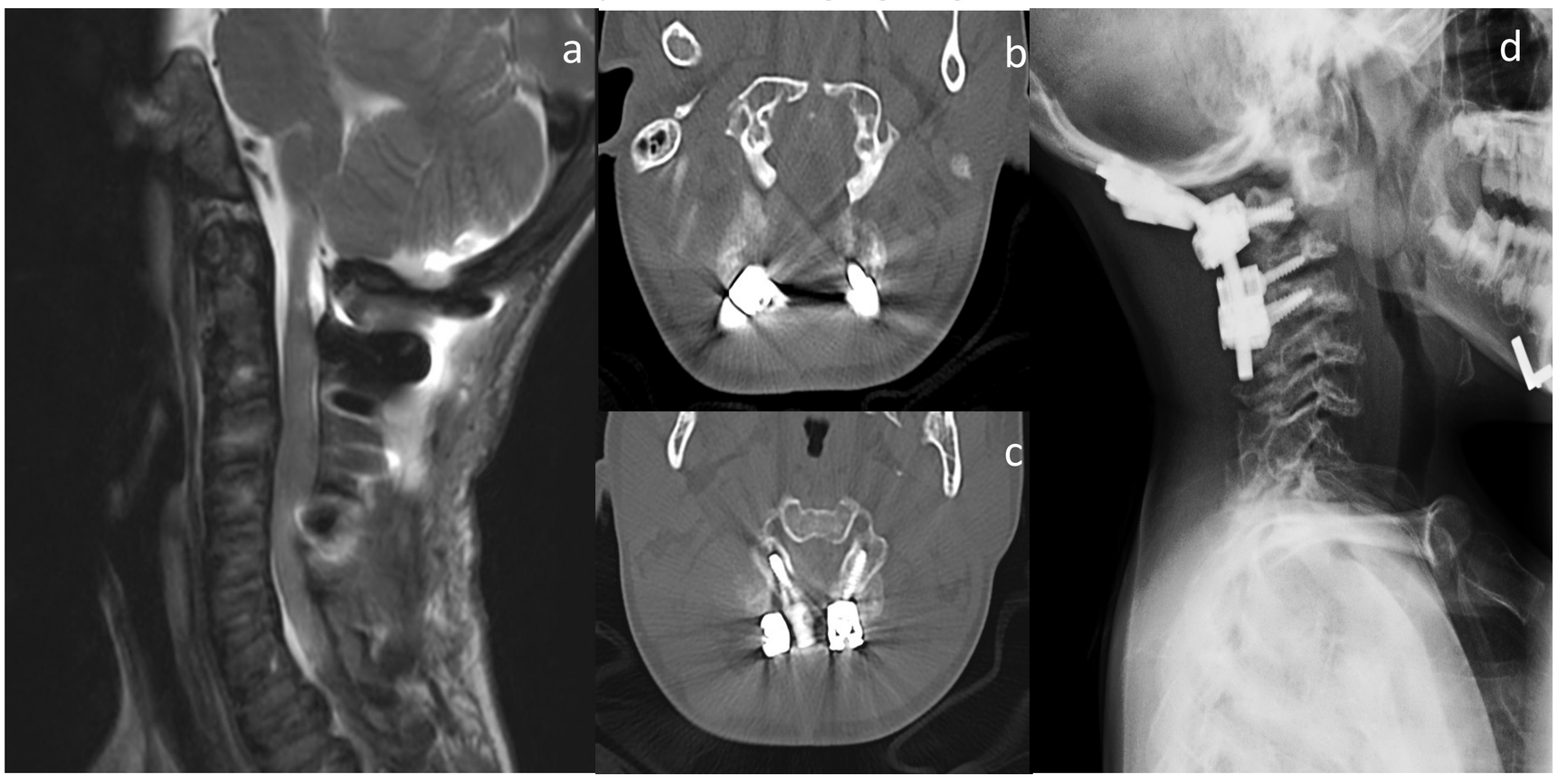
iCT-guided OCDF workflow (Figure 2)



OCDF (Figure 3)



Post-operative imaging (Figure 4)



Background:

Mucopolysaccharidoses (MPS) constitute a group of rare inherited lysosomal enzyme deficiencies in which the accumulation of glycosaminoglycans can result in spinal deformities like cervical stenosis and atlantoaxial instability. It has been well demonstrated in the literature that surgical intervention is the only effective option for MPS-associated spinal deformities, yet there is no consensus on the timing of surgical intervention and surgical approach.

Aim:

Given the variety of surgical management options, this study illustrates the pre-operative planning and the surgical technique of intraoperative CT (iCT)-guided occipitocervical decompression and fusion (OCDF) in mucopolysaccharidosis (MPS) at a tertiary referral neurosurgical center.

Case Illustration:

A 6-year old boy with MPS type IVA-associated cranio-cervical stenosis and already on long-term enzyme replacement, was referred for neurosurgical evaluation. Preoperative sagittal T2W MRI of the cranio-cervical junction and cervical spine revealed foramen magnum stenosis (Figure 1; red box) and at the C3-4 spinal canal stenosis with cord indentation and T2-hyperintensities in the cervical spinal cord that was suggestive of early cervical myelopathy (Figure 1; green box).

Surgical Technique and Outcome:

Operative setup (Figure 2a) includes neuro-navigation (Figure 2b) and intraoperative neuromonitoring. Intraoperatively, atlantoaxial subluxation was observed with the C1 laminae compressing into the foramen magnum with cord compression. C1 laminectomy and C3-4 laminotomy were first performed to achieve foramen magnum and cord decompression, followed by iCT-guided occipital screws, C2 laminar screws, C3-4 lateral mass screws (all 3.0x12mm) placement, then posterior fusion (Figure 3). Postoperatively, the patient had an uneventful neurological recovery. Post-operative sagittal T2W MRI cervical spine showed increased central canal diameter and MRI-safe implants are seen (Figure 4a). Axial CT spine showed screws in-situ in C2 (Figure 4b-c). Spinal alignment is also restored as seen on lateral cervical X-ray (Figure 4d).

Conclusion:

To the best of the authors' knowledge, this is the first reported iCT-guided OCDF in a pediatric MPS patient. iCT-guided OCDF can safely achieve the surgical goals of early decompression, stabilization and restoration of alignment to prevent further irreversible spinal cord injury secondary to cranio-cervical stenosis in MPS.

References:

[1]Zairi, Mohamed et al. "Cranio-cervical decompression associated with non-instrumented occipito-C2 fusion in children with mucopolysaccharidoses: Report of twenty-one cases." *North American Spine Society journal* vol. 12 100183. 19 Nov. 2022, doi:10.1016/j.xnsj.2022.100183