Abstract:
13 years male born of non-consanguineous parents presented with progressive spastic weakness of upper and lower limbs with atrophy and contracture of muscles 18 months duration. No specific family history. Sibling was normal. Ophthalmological examination showed bilateral corneal haziness. Skeletal survey showed skeletal dysplasia around joints. These features are consistent with Morquios disease. Atlantoaxial Dislocation due to odontoid hypoplasia was considered to be the cause of quadriparesis. C1- C2 pars screw fixation with bone grafting of the AA joints was done.

Keyword:
Atlantoaxial dislocation, C1-C2 fixation, Morquios disease, Mucopolysaccharidoses

Introduction:
MORQUIO-BRAILSFORD SYNDROME (MBS) is Type IV mucopolysaccharidoses (MPS). MPS are lysosomal storage disorders due to a deficiency of lysosomal enzyme resulting in accumulation of substrate in the cell. Accumulation of substrate in the cells near joints results in metaphyseal and epiphyseal bone dysplasia with degeneration of connective tissues near the joints.

Clinical Presentation
13 year old male born to non-consanguineous parents admitted with c/o difficulty in using and weakness of upper and lower limbs –18 months duration, progressive deformity-initial walking on toes with associated flexion deformity of both knees, deformity of the spine and occasional difficulty in breathing. No specific family history. Sibling -Normal General examination showed Short Neck / Low Hair Line, High Arched Palate, Pectus Carinatum, Genu Valgum, Flat Foot Ophthalmological examination showed bilateral corneal haziness

Higher functions-Appropriate for age Cranial nerves –Normal Spinomotor system Patient had atrophy of Lt. Trapezius and bilateral Gastrosoleus, Contracture of both tendoachillies, Spasticity of both upper limbs and lower limbs, Power - Upper limbs-4+/5 & Lower limbs-4/-5, all deep tendon reflexes exaggerated, Ankle clonus ,Plantar –bilateral withdrawal and Spastic gait

**Investigation:**
X-ray Cervical Spine revealed atlantoaxial instability.

MRI showed odontoid hypoplasia, thinned out cord, platyspondyly, beaking of vertebrae
Skeletal survey showed skeletal dysplasia around joints
The clinical and radiological examinations showed no evidence of dislocation of hip joints The urinary examination for glycosaminoglycan showed increased urinary excretion.

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**Diagnosis:**
Atlantoaxial Dislocation due to Morquio’s Disease

Post-operative X-Rays;

Patient recovered from quadriparesis and subsequently underwent soft tissue release of the knee joints to gain better mobility.

**Conclusion:**
Odontoid hypoplasia and AAD are known to be associated with MBS and is often associated with significant morbidity and even mortality. Some authors have even recommended prophylactic stabilization of C1-C2 in this condition to prevent sudden death. Therefore, C1-C2 arthrodesis forms an important and viable modality of treatment. Till date, C1-C2 polyaxial screw rod fixation for Morquio’s induced AAD has not been reported in the literature.

**Reference**