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Case Report

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# Anaesthetic Management of Fracture Neck of Femur in a Case of Hurler's Syndrome – A Case Report

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## Abstract

Hurler's syndrome is a type of mucopolysaccharidoses, a rare lysosomal storage disorder with a prevalence of 1 in 100,000 individuals, results from deficient α-L-iduronidase enzyme activity, leading to the accumulation of glycosaminoglycans within lysosomes. This condition presents unique challenges to the attending anaesthesiologist as it manifests with profound anomalies, including severe airway and cervical spine manifestation. Difficult intubation is reported in 54% of cases, with a 23% incidence of failed intubation, highlighting the critical need for specialized preoperative care.

We describe a case of a 20-year-old male diagnosed with Hurler's syndrome since birth, presenting with a fracture of the neck of femur. Given the risks associated with manipulation of the airway, the patient underwent CC-screw fixation under regional anaesthesia. The anaesthetic plan focused on avoiding airway manipulation and optimizing perioperative conditions under the guidance of a multidisciplinary team, including experienced anaesthesiologists.

**Keywords:- Hurler's syndrome, anaesthetic management, central neuraxial blockade, perioperative care**



## INTRODUCTION

Mucopolysaccharidoses (MPS) is a rare inherited lysosomal storage disorder associated with the progressive accumulation of glycosaminoglycans (GAGs) in tissues and organs. Anaesthesia and surgery in these patients carry a high mortality risk due to serious complications, particularly airway obstruction, which can cause ventilation and oxygenation difficulties and significant cardiovascular compromise.[1]

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Hurler's syndrome, also known as MPS I, is characterized by a deficiency in the lysosomal enzyme α-L-iduronidase. This deficiency results in the accumulation of GAGs, leading to progressive multisystem involvement. Patients with Hurler's syndrome typically present with distinctive facial features, skeletal abnormalities, organomegaly, and severe neurological impairment. Anaesthetic management in these patients is particularly challenging due to potential difficulties with airway management, cervical spine instability, and the presence of multiple comorbidities.[2] Central neuraxial blockade is another challenge in such patients as they present with deformities of the spine like kyphosis and a smaller volume of spinal cerebrospinal fluid, potentially affecting the spread of local anaesthetics. This report describes the anaesthetic considerations and management of a patient with Hurler's syndrome undergoing surgery for a fracture of the neck of the femur which was managed with regional anaesthesia, avoiding the need for intubation.[3,4]

# CASE REPORT

A 20-year-old male presented with a fracture of the neck of the femur on the right side, posted for CC screw fixation. The patient was diagnosed with Hurler's syndrome at birth and was known to have a seizure disorder since childhood, on regular medication with tablet Phenobarbitone and tablet Phenytoin. His intellectual functions were normal for his age. The patient weighed 30 kilograms and was 138 centimetres tall. Airway examination revealed adequate mouth opening, macroglossia, modified Mallampati class III, and short neck with an adequate range of neck movements [Figure 1].

**Figure 1: Airway examination showing macroglossia and a short neck.**

On palpation of the spine, the patient had significant kyphosis at the thoracolumbar region resulting in gibbus deformity with reduced intervertebral spaces [Figure 2].

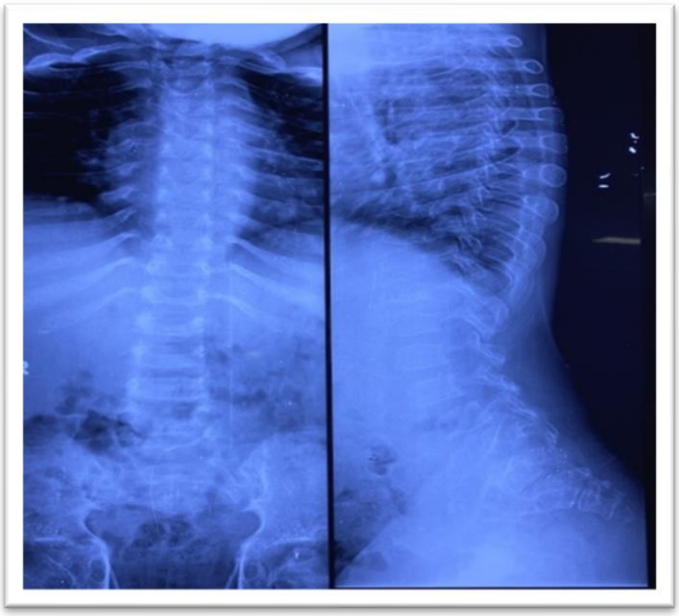

**Figure 2: X-Ray showing significant kyphosis at the thoracolumbar level with reduced intervertebral spaces.**

The rest of the systemic examination was normal. No abnormal values were found on routine blood investigations and 2D echocardiography. A written and informed consent was obtained from the patient. The operating room was prepared with a difficult airway cart and the patient was approached with multidisciplinary care involving experienced anaesthesiologists. After attaching standard monitoring and taking baseline recordings, an 18-gauge intravenous cannula was secured. The patient was placed in a sitting position and the skin was prepared with 0.5% chlorhexidine and draped in a sterile manner. A small amount of local anaesthetic (2% Lignocaine) was injected into the L2-L3 and L3-L4 interspaces. Using the midline approach, the epidural space was accessed using the loss of resistance technique at the level of L3-L4 with an 18-gauge Tuohy's needle. The epidural catheter was inserted and fixed to the skin at a depth of 9 cm. At the level of L2-L3 interspace, the dura was pierced using a 25-gauge Quincke's spinal needle. Subsequently, 1.8 mL of 0.5% hyperbaric Bupivacaine was injected along with an adjuvant of Clonidine 15mcg. Intraoperatively, the patient was hemodynamically stable. The surgical procedure lasted for two hours and was uneventful. The patient was transferred to the post anaesthesia care unit with stable vital signs and analgesia was maintained through the epidural

catheter with an infusion of 0.2% Ropivacaine at 4ml/hr for a period of two days.

# DISCUSSION

Mucopolysaccharidoses type I - Hurler's syndrome has a prevalence of 1 in 100,000 people and is caused by a deficiency of the lysosomal enzyme α-L-iduronidase, leading to the accumulation of glycosaminoglycans in multiple systems in the body. Patients with mucopolysaccharidoses have profound anomalies of the airway and the spine like macroglossia, cervical spine instability, restricted movement of the temporomandibular joints and anteriorly placed larynx, resulting in challenges in managing the airway.[5] They often have multisystemic involvement including abnormalities of the cardio-respiratory and skeletal system, which manifests as restrictive lung disease. Progressive neurodegenerative disease such as brain atrophy are more prominent in Hurler's disease. Evidence of obstructive sleep apnoea should be sought; preoperative sleep studies may be required for full evaluation and risk stratification. The high anaesthetic risk for these patients primarily is due to predicted difficult airway and the presence of comorbidity like seizure disorder and anomalies of the spine like kyphoscoliosis. They pose certain challenges to the attending anaesthesiologist in terms of providing neuraxial anaesthesia. An experienced anaesthetic team should always manage these patients. Adequate preparation and resuscitation equipment should be readily available for management of the airway. Always anticipate a difficult airway in patients with MPS and ensure the full range of difficult airway adjuncts is available. Securing an intravenous access is another challenge in such patients due to the presence of tissue deposits, contractures, and bony defects. Difficult airway should always be anticipated as they have joint and bony involvement and a difficult airway cart should be kept standby before the administration of anaesthesia. Preferably, sedative premedication should be avoided to prevent airway involvement.[6] Instability of the cervical spine should be presumed in the absence of flexion–extension cervical spine films and the cervical spine must be immobilized during tracheal intubation. Short stature leads to a smaller volume of spinal cerebrospinal fluid, potentially affecting the spread of local anaesthetics. The deposition of excessive intra-lysosomal GAGs thickens the soft tissues, resulting in the narrowing of the nasal airways, oral cavity, pharyngeal, and laryngeal tissues. Perioperative mortality rates averaging 20-30% have been reported for patients with this disease, with failure to control the airway as the largest single cause of mortality.[7] The major benefit of regional anaesthesia (RA) is related to the non-manipulation of the airway, thus avoiding the risk of difficult or failed intubation. The deposition of mucopolysaccharides in the epidural space or around nerve sheaths can prevent direct access of local anaesthetics to the nerve.[8] Nonetheless, apart from deposition of GAGs in the epidural space, MPS is known to have other vertebral anomalies and neurological involvement which can influence RA techniques. The common problems of spines include high lumbar kyphosis, thoracic scoliosis, antero-inferior beaking of hypoplastic thoracolumbar vertebral bodies, thickening of the ligamentum flavum, and dural thickening. This can cause technical difficulty in performing neuraxial techniques and also influence intrathecal drug spread.[9,10]

# CONCLUSION

The MPS patient poses a major challenge to the anaesthetist. The anaesthetic risk can be reduced considerably if the anaesthetist anticipates potential problems that may arise in these patients during and after the procedure, including difficult intubation and ventilation, and cardiac and cervical spine issues. This requires a thorough preoperative evaluation and knowledge of the pathophysiology underlying the respiratory and cardiac manifestations, as well as cervical and tracheo-laryngeal anatomy in these patients. Therefore, these difficult decisions should ideally be made by a multidisciplinary team in a tertiary referral centre experienced in the perioperative management of MPS patients. Anaesthesia in patients with an unstable spine or for spine surgery is particularly difficult and requires additional care and thought in the choice of anaesthetic, monitoring and postoperative care.

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