Session 1. Safe Anesthesia for Children with Co-Morbidity

Anaesthesia for Patient with Mucopolysaccharidosis

Learning Objectives:

Mucopolysaccharidosis (MPS) is a form of lysosomal storage disorders and it accounts for about one-third of all lysosomal storage disorders. Eleven enzyme deficiencies and 7 distinct forms of MPS have been identified. Presentation can be heterogeneous and common clinical manifestations include coarse facial features, macroglossia, macrocephaly, corneal clouding, hearing impairment, hernias, short stature, developmental delays, joint and skeletal deformities, organomegaly, angiokeratoma, muscle weakness or lack of control, neurological abnormalities including regression.

Patients with MPS often require anaesthesia for surgical interventions and sedation for imaging studies. Common surgical procedures include tonsillectomy and adenoidectomy, hernia repair, shunt operation for obstructive hydrocephalus, release of carpel tunnel and corneal transplants for corneal clouding. More commonly they need magnetic resonance and computer tomographic studies for skeletal deformities including cervical spine abnormalities. Apart from supportive treatment, enzyme replacement therapy (ERT) and hematopoietic stem cell transplant (HSCT) have emerged as newer and more effective treatment for MPS enabling a number of these patients to survive into adulthood resulting in even greater needs for more surgical interventions and surveillance imaging studies.

In this lecture we will go through the considerations for management of children with MPS requiring anaesthesia and sedation.