

Morquio Syndrome

40 year old male booked for right TKR in September. His operation was cancelled on day of surgery in a regional hospital last week.

Background

- Morquio syndrome:- type IV mucopolysaccharidosis. Usually presents with skeletal deformities, short stature and possible upper cervical instability and cardiac deformities. Potential for narrowed tracheal lumen, sleep apnoea, hypermobility and prominent sternum.
- Short stature (135cm)
- OSA diagnosed 5 years ago. Has not taken up CPAP therapy as recommended due to cost
- Smoker
- Glucose intolerance
- Previous bilateral hip replacements – reportedly under general anaesthetics

He works at the local Leagues club and his exercise is limited by his osteoarthritis. His only regular medication is paracetamol PRN. Unfortunately he continues to smoke - had been advised to cease. He reports no concerns with previous anaesthetics. His surgery was cancelled in Tamworth due to concerns about his high-risk status for anaesthesia.

His vital signs were normal. His cardio-respiratory exam was essentially normal, and he had a reasonably normal airway exam. He has previously had a Grade II laryngoscopy. Recent pathology was normal.

He has had a spirometry, echocardiogram, flexion/extension cervical spine x-rays, and MRI of his spine in work-up for this surgery. His spirometry was essentially normal with an FEV1 of 2.3L. His echocardiogram and flexion/extension cervical spine x-rays were normal. His MRI of his spine showed no spinal cord abnormalities, with the conus medullaris at the level of L1. His vertebral bodies were abnormal as expected due to his Morquio syndrome.

Discussion

- GA vs Spinal – there were debates about what is the best approach. There was agreement that there are no absolute contra-indications to either approach.
- Patient reasonably optimized for surgery given the results of investigations
- Many different reasons why possible cancellation on day of surgery. Patient reported he had been told that he was sent to us because “we have a spinal needle with a camera on the end of it!”
- Note this is a Mucopolysaccharidosis type IV – less severe subtype of these metabolic disorders

Outcome

Patient to proceed to planned surgery
Assess for HDU postoperatively due to history of possible OSA.