Williams Syndrome and difficult intubation

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Citation

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Abstract

Williams Syndrome is a genetic condition characterised by a distinct facial appearance, cardiovascular anomalies, neonatal hypercalcemia and craniofacial abnormalities. An 18-year-old girl with Williams's syndrome was scheduled for appendicectomy. This patient had a difficult intubation. Since we had identified her as a difficult airway patient in the pre-operative assessment, our preoperative preparation helped us in successfully managing the airway.

We read with interest " Anaesthesia management of a patient with Williams Syndrome" by S Sahin, A Colak and I Gunday in Volume 14 of The Internet Journal of Anesthesiology. Williams syndrome, as first described by Williams, Barratt-Boyes and Lowe in 1961, is a rare genetic condition caused by a sub microscopic deletion of a number of genes on the long arm of chromosome 7. Its incidence is about 1:20000 in the general population and the clinical manifestations are usually multisystem and include a distinct facial appearance, cardiovascular anomalies, neonatal hypercalcemia, and a characteristic neurodevelopment profile. The deletion of the elastin gene and the characteristic craniofacial abnormalities has always prompted proper pre anaesthetic assessment of these patients with a view of identifying difficult mask ventilation or difficult intubation or both. However there are no reports as yet in the anaesthetic literature of a difficult intubation.

We report the perioperative management of 18-year-old girl weighing 56kg, with Williams's syndrome admitted with clinical and biochemical signs of acute appendicitis, who required anesthesia for laparoscopic appendicectomy. She had undergone a herniotomy in 1991, which was performed under mask anaesthesia uneventfully. Her most recent cardiac echo was in 1998, which showed no Left ventricular outflow Tract obstruction. Preoperative evaluation was unremarkable and she was fit and healthy. Lab investigations showed a mild medullary nephrocalcinosis. Preoperative airway assessment showed a Mallampati grade 3 with mandibular subluxation of -1. She had good neck extension with a mentohyoid distance of only 1.5cm.After adequate fasting period, she was taken to the anaesthetic room and IV access secured and a rapid sequence induction was done after preoxygenation with thiopentone 5mg/kg and suxamethonium 1.5 mg/kg. In view of her Mallampati Grade and poor mandibular subluxation, we had the difficult intubation equipment ready. There was increased masseter muscle tone, which resolved 45sec after Suxamethonium. Laryngoscopy was done with a McCoy Blade size 3 and the laryngoscopy view was Cormack and Lehane grade 3b. A bougie was inserted just underneath the epiglottis and a 6.5mm ID ETT was railroaded over the bougie. Placement was confirmed with a capnographic trace and auscultation of breath sounds. The anaesthetic was maintained with oxygen and nitrous oxide and propofol infusion and rocuronium was used for muscle relaxation .The rest of the anaesthetic was uneventful and the patient neuromuscular blockade was reversed end operatively and extubated awake at the conclusion of the surgery.

As far as we are aware this is the first reported case of difficult intubation in Williams syndrome in the anaesthetic literature. Airway problems were anticipated in this patient and we had all the necessary equipment. A combination of mild mental retardation and anxiety precluded the use of awake fibreoptic intubation in this patient. We decided to make one good attempt with the right head positioning and the right laryngoscopy blade before falling back on asleep fibreoptic intubation. Despite good positioning and the use of the McCoy Blade, we still had a poor laryngoscopy view and had to use a bougie to place the Endotracheal tube.

References

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