Case Report: Dexmedetomidine Sedation for tracheostomy in a child with McCune-Albright Syndrome

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Introduction: A 13 y/o girl, with a massive cranio-facial tumor, progressively compromising the child’s airway presented for tracheostomy. Her mother detailed a history of limited physical activity, obstructed sleep apnea, visual impairment and progressive dysphagia. The child had been experiencing increased shortness of breath while supine and maintained a sitting position during sleep. We present the intraoperative use of Dexmedetomine during a tracheostomy performed under local anesthesia, in an extremely advanced case of McCune-Albright Syndrome.

Perioperative Course: On physical exam she exhibited a massive facial bone tumor totally obstructing her nares and most of her oropharynx. She had a very limited range of mouth opening with abnormal scant dentition and the neck had a limited extension. She had finger clubbing, kyphoscoliosis and anterior bowing of the tibias. She was somewhat agitated preoperatively, however she allowed us to gain intravenous access following EMLA cream on the dorsum of her hand.

A Dexmedetomidine (Dex) infusion (0.04mg/ml) was started at a rate of 1mcg/kg for 10 minutes. At this point she was sedated with closed eyes which she would open to verbal command. The rate of infusion was then continued at 0.05mcg/kg/hr. Lidocaine 1% with epinephrine was injected subcutaneously without complaint. 1 mg of midazolam IV was administered at the time of incision. Her heart rate had dropped to 86/min from 120 after the Dex infusion was started. SpO2 remained between 97 and 100% throughout the procedure and other vital signs were stable. Upon completion of the tracheostomy general anesthesia was induced with sevoflurane. The infusion of Dexmedetomidine was decreased to 0.03mcg/kg/hr for the remainder of the case which involved biopsy of the tumor from several sites. Furthermore it was noted that a fiber optic endoscope failed to pass the hypopharynx due to the tumor mass.

The patient, who breathed spontaneously for the entire procedure, made an uneventful recovery.

Discussion: Here we were presented with the challenging situation of a child who needed a surgical airway because of progressively worsening upper airway obstruction; making intubation via the nose or mouth impossible.

Dexmedetomidine is an alpha adrenergic agonist with a high, (1620: 1) alpha 2: alpha 1 receptor affinity. It produces sedative hypnotic effects by acting on the Alpha 2 receptors in the locus ceruleus. Given as an infusion intravenously, it has a rapid onset of action with a time to peak effect at approximately 15 – 20 minutes. Dex sedation is not associated with an altered CO2 response.

Dexmedetomidine has been used successfully in children to improve sedation during mechanical ventilation in the intensive care setting. (1) It has also been described to be effective as a sole sedation agent for certain non invasive procedures without producing respiratory depression. (2).

Dexmedetomidine has been used for “awake” fiber optic intubations in adults, as an adjunct to topical anesthesia (3, 4). In this case, Dex was successfully used in a child to sedate safely and adequately until a tracheostomy was performed.
Reference:


Figure 1
Figure 2
Figure 3