Letter to the Editor

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Dexmedetomidine and propofol based total intravenous anesthesia in a case of Joubert syndrome

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Sir,

Joubert syndrome (JS) is a rare autosomal recessive/sporadic ciliopathy with an underreported prevalence of 1 in 80000 to 1 in 100000. These children present with developmental delays, hypotonia, effortless episodic tachypnea with or without apneic spells, ocular nystagmus, retinopathies, laryngomalacia, palate malformations, renal failure (sometimes requiring continuous renal replacement therapy), hepatic fibrosis/failure, and congenital heart disease [1,2]. The most significant neuropathological finding is dysgenesis of the cerebellar vermis [3]. The hallmark and obligatory criterion for diagnosing JS is the molar tooth sign (MTS) on neuroimaging. Few case reports describe the use of anesthetic agents for sedation in these patients, especially during magnetic resonance imaging (MRI). Here, we describe the case of an eight-year-old male who presented to us with severe dental caries and a known diagnosis of JS. The child experienced an uncomplicated vaginal delivery. On his second day of his life, he developed tachypnea and generalized tonic-clonic seizures. MRI images revealed the classic MTS, confirming the diagnosis of JS (Fig. 1A). On examination, the child had delayed motor skills, poor speech, and coordination. He also had a previous history of a period of 10-minute apnea. At his current age of eight years, he could stand and walk, and required only intermittent support. He continued to exhibit hypotonia, especially in his lower limbs. He also had polydactyly, a partially split tongue, and high arched palate (Fig. 1B & 1C). All these features further supported the diagnosis of JS with orofacial defects (JS-OFD) [1]. On further evaluation, he had episodic tachypnea with respiratory rates as high as 80 breaths per minute, but with no cardiac, renal, ocular, or hepatic involvement. An MRI brain scan did not reveal any hydrocephalus; however, the presence of MTS (Fig. 1A) and a split cerebellum confirmed the diagnosis. Airway examination revealed no micrognathia and macroglossia.

The child was scheduled for the extraction of carious teeth. Premedication was achieved with oral ketamine 3 mg/kg after confirming adequate preoperative fasting.

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Dr. Vishwendra Singh is now working as a Senior Resident in Government Medical College and Hospital, Chandigarh. The work should be attributed to PGIMER, Chandigarh.

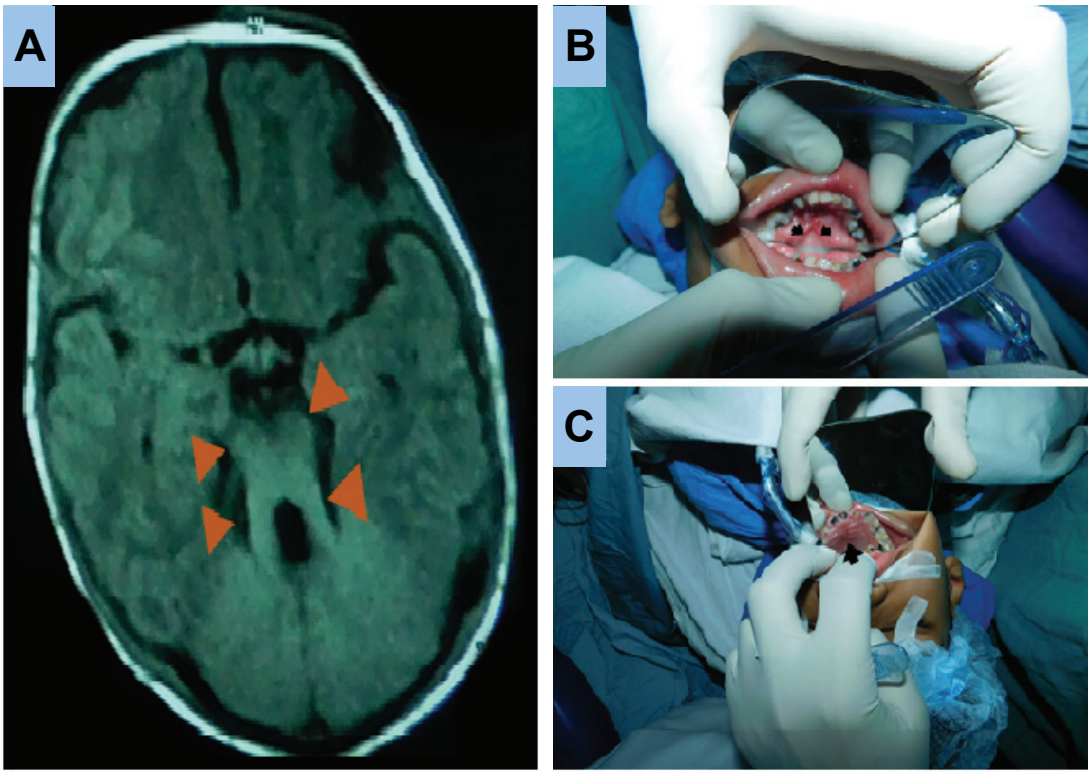

Fig. 1. (A) Molar tooth sign (orange arrowheads) – T1 weighted MRI image, (B) An incomplete cleft in the tongue (black arrows), (C) High arched palate and submucous cleft (black arrows).

General anesthesia was administered using 1 mg/kg propofol under bispectral index (BIS) monitoring. Once an adequate depth of anesthesia was achieved (BIS < 60), and bag-mask ventilation was confirmed, atracurium 0.5 mg/kg was administered. Plain lignocaine 1.5 mg/kg was administered to attenuate the laryngoscopy response. The airway was secured with a 6.5 cuffed endotracheal tube with no difficulty (Cormack-Lehane grade 2), once the train of four (TOF) count was zero. Anesthesia was maintained with dexmedetomidine 0.5 µg/kg/hour, and propofol 25–50 µg/kg/min with a 50:50 air/oxygen mixture on pressure-controlled ventilation (PCV) mode. No opioids were used. Analgesia was augmented with local infiltration of 0.125% bupivacaine. At the end of the surgery, the propofol was switched off, and the dexmedetomidine dose was reduced to 0.3 µg/kg/hour until spontaneous breathing resumed. The child was reversed with 50 µg/kg neostigmine and 10 mics/kg glycopyrrolate. There was no tachypnea, and the child was extubated smoothly at a TOF count of 4 and a TOF ratio of 0.9. Spontaneous regular breaths with a normal respiratory rate were achieved around five minutes post-extubation. Upon extubation, the dexmedetomidine was switched off, and one hour later, the child re-developed episodic tachypnea with respiratory rate of 75–80 breaths per minute, interspersed with apneic spells, which lasted for fewer than 10 seconds and were not associated with oxygen desaturation. These apneic spells resolved after 6 hours, and the episodic tachypnea reduced to 60 breaths per minute. The patient had an uneventful postoperative course, apart from the episodic tachypnea, and was discharged from the hospital after 48 hours.

This case highlights the importance of appropriate monitoring, such as the bispectral index and TOF, along with a multimodal opioid-sparing analgesic approach. Avoiding opioids is of paramount importance as these agents—except remifentanil—can exacerbate and prolong the apneic spells. Additionally, patients with JS are sensitive to opioid use [2]. Alpha-2 agonists such as

dexmedetomidine have been used safely to maintain regular spontaneous breathing without tachypnea in such patients, especially during MRI sedation [4]. The peri-operative use of dexmedetomidine facilitates smooth extubation, once reversal of the neuromuscular blockade has been confirmed.

Novelty:

The report highlights the importance of proper monitoring and opioid avoidance in patients with JS, which reports do not mention. Opioids can exacerbate and prolong apneic spells in these patients. Dexmedetomidine has been reportedly used for sedation for MRI; however, in the current case, it was used for a surgical procedure.

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AUTHOR CONTRIBUTIONS

Ankur Luthra: Conceptualization, Data curation, Formal analysis, Methodology, Supervision, Writing – original draft, Writing – review & editing

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