Intravenous Sedation in Arnold-Chiari Malformation With Respiratory Failure

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Type II Arnold-Chiari malformation (ACM) is an abnormality in which the cerebellum, pons, and medulla oblongata are displaced downward into the spinal cord. Type II ACM is often complicated by respiratory depression, sleep-disordered breathing, and deglutition disorder as a result of medullary dysfunction and impairment of the lower cranial nerves. Bending and stretching of the neck is restricted, and anesthetic management is problematic in patients with the disorder. We performed dental treatment twice under intravenous sedation in a patient with intellectual disability with type II ACM complicated by hypercapnic respiratory failure. Propofol was used for the first sedation procedure. Repeated bouts of respiratory depression occurred on that occasion, so the airway was managed manually by lifting the jaw. However, aspiration pneumonia occurred postoperatively. A combination of dexmedetomidine and midazolam was used for sedation on the second occasion, and the intervention was completed uneventfully without any respiratory depression. Our experience with this patient highlights the need for selection of an agent for intravenous sedation that does not require neck extension and has minimal effect on respiration in patients with type II ACM, who are at high risk of respiratory depression and pulmonary aspiration.

Key Words: Arnold-Chiari malformation; Hypercapnic respiratory failure; Intravenous sedation; Dexmedetomidine; Midazolam; Propofol.

Arnold-Chiari malformations (ACMs) are congenital anomalies of the cerebellum, pons, and medulla oblongata and are classified as types I–IV.¹ In type I, the cerebellar tonsils and lower part of the medulla oblongata extend below the foramen magnum without displacement of the fourth ventricle. Type II ACM is usually associated with myelomeningocele and is characterized by caudal migration of the lower part of the cerebellum and downward displacement of the fourth ventricle, which appears lengthened, and the foramina open into the spinal subarachnoid space. In type III, the cerebellum and medulla oblongata are displaced into the cervical spinal canal, and an occipital meningocele is present. In type IV, the cerebellum is incomplete or underdeveloped.

Type II ACM is often complicated by respiratory depression as a result of upper airway obstruction caused by impairment of the lower cranial nerves,² sleep-disordered breathing caused by impairment of the medullary respiratory center,^{3,4} and deglutition disorder and aspiration caused by impairment of the medullary swallowing center.^{5,6} This type of ACM is often associated with myelomeningocele, other forms of spina bifida, and/or hydrocephalus. Respiratory and swallowing disorders caused by medullary dysfunction and impairment of the lower cranial nerves are common in infants with ACM. These complications have important prognostic implications.⁷ Unfortunately, prompt treatment of myelomeningocele and hydrocephalus in these patients does not always improve the clinical symptoms.⁸ Patients with type II ACM who develop severe upper airway obstruction or impaired swallowing may need to undergo tracheotomy or gastrostomy. Furthermore, several problems must be addressed when these patients require sedation/general anesthesia.

In patients with type II ACM, the cerebellum and fourth ventricle are displaced downward and press on the medulla oblongata. Therefore, rapid bending and stretching of the neck can worsen the impairment of the

Received March 29, 2018; accepted for publication August 15, 2018.

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Anesth Prog 66:37–41 2019 | DOI 10.2344/anpr-66-01-06
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respiratory and swallowing centers, and cooperation with a dental procedure under physical restraint may be poor. Anesthetic management that avoids movement of the neck as much as possible is important in these patients. Dexmedetomidine, which causes less respiratory depression than opioids, midazolam, or propofol,^{9,10} is often used when intravenous sedation is required for dental treatment^{11–13} and is recommended particularly for patients at risk of respiratory depression.¹⁰ We have performed intravenous sedation for dental treatment in an intellectually disabled patient with type II ACM and hypercapnic respiratory failure on 2 separate occasions: the first using propofol and the second using a combination of dexmedetomidine and midazolam. This report describes our experience with this patient. Consent to report this patient's case was obtained from the patient's legal guardian.

CASE PRESENTATION

The patient was a 27-year-old man (height 153 cm, weight 33 kg) who had been diagnosed with type II ACM at birth. Shortly after birth, a surgical repair was performed for a myelomeningocele that was associated with spina bifida, and a ventriculoperitoneal shunt was placed for hydrocephalus. The medulla oblongata and cerebellar tonsils were compressing the spinal cord at the C1 level, and the patient's physician had counseled on the hazards of excessive bending and stretching of the neck. He had deglutition disorder and had developed acute respiratory distress syndrome caused by aspiration pneumonitis a year earlier at the age of 26 years. At that time, he was admitted in a critical state but was treated successfully. However, his course at that time was complicated by hypoxemia and hypercapnia. He was diagnosed with type 2 respiratory failure characterized by hypoxemia and hypercapnia with a $\text{PaO}_2 < 60$ mm Hg and a $\text{PaCO}_2 > 50$ mm Hg. Home oxygen therapy was initiated, but tracheotomy and gastrostomy were not performed.

The patient's mother had noticed that the patient was experiencing pain in the oral cavity, so she brought him to our hospital. He was resistant to examination of his mouth because of his disability, so general anesthesia was considered. However, his respiratory physician predicted that there would be a high risk of difficulty weaning the patient off mechanical ventilation and a high likelihood of perioperative complications, including lengthy artificial ventilation, pneumonitis, and worsening of his type 2 respiratory failure and so advised against general anesthesia. Therefore, day-stay hospitalization for dental treatment under intravenous sedation was scheduled.

Preoperative evaluation revealed the patient's blood pressure and heart rate to be 118/78 mm Hg and 75 bpm, respectively. His SpO_2 was 90%–92%, and his PaCO_2 was 60–65 mm Hg on room air. Preoperative blood tests and an electrocardiogram did not reveal any abnormalities. He weighed 33 kg with a height of 153 cm. Communication was impossible because of the patient's intellectual disability. He was wheelchair bound because of complete paraplegia. Scoliosis and thoracic deformity were observed. The patient had a history of latex allergy with 3 previous episodes of systemic inflammatory response to latex. Sleep-disordered breathing was reportedly observed at night.

First Intravenous Sedation Procedure

Intravenous sedation using a continuous infusion of propofol was planned. We anticipated problems with the patient's airway, so preparations for tracheal intubation were made. All the devices used were latex free. On arrival in the operating room, oxygen was administered at a flow rate of 3 L/min via a nasal cannula equipped with a capnometer. The patient was monitored by pulse oximetry, 3-lead electrocardiography, and noninvasive blood pressure measurement. The intravenous line was secured under restraint. Propofol was administered as a bolus dose of 15 mg followed by an initial continuous infusion of 3 mg/kg/h. When the patient's Observers Assessment of Alertness/Sedation Scale (OAA/S) score was 3, the oral cavity was examined and radiographic images were obtained. Next, infiltration anesthesia was achieved using 3.6 mL of 2% lidocaine containing 1:80,000 epinephrine. The mandibular right first molar tooth was then extracted, and a restorative procedure was performed on the mandibular left second molar tooth, followed by dental scaling. There was no detectable body movement during the procedure, but frequent episodes of upper airway obstruction caused by displacement of the tongue were observed. When these episodes occurred, the propofol infusion rate was decreased, and the airway was managed manually by lifting the jaw to avoid bending and stretching of the neck. Propofol was administered intraoperatively at a dose of 2–3 mg/kg/h and was discontinued on completion of surgery. The intraoperative OAA/S score was 2–3, with a mean arterial pressure of 65–80 mm Hg, a heart rate of 60–65 beats/min, an SpO_2 of 93%–96%, and an end-tidal CO_2 of 55–65 mm Hg. The total dose of propofol used was 140 mg over 55 minutes, and the duration of anesthesia was 70 minutes. The patient's vital signs and respiratory status stabilized immediately after the surgery. However, on

the following day, he developed a fever and aspiration pneumonitis and so was kept in hospital for treatment. His symptoms steadily improved, and he was discharged on postoperative day 7.

Second Intravenous Sedation Procedure

Eight months after the first intravenous sedation procedure, the patient was brought back to the hospital when his mother again noticed that he was experiencing pain in the oral cavity. On this occasion, a plan was made for intravenous sedation using dexmedetomidine and midazolam. No interval change in medical history was noted, and baseline vital signs were similar to the first procedure. Oxygen was administered at a flow rate of 3 L/min via a nasal cannula equipped with a capnometry sampling cannula. After securing an intravenous line, 1.0 mg of midazolam was administered simultaneously with an initial bolus of dexmedetomidine 1 µg/kg administered over 10 minutes. The patient's OAA/S score reached 3 after the initial bolus of dexmedetomidine, and a continuous infusion at a dose of 0.5 µg/kg/h was started. The oral cavity was then examined, and radiographic images were obtained. Local infiltration anesthesia consisting of 2.6 mL of 2% lidocaine containing 1:80,000 epinephrine was then administered, after which restorative procedures were performed for the mandibular first and second premolar teeth on the right. Slight body movement was observed at 5 and 30 minutes intraoperatively; intravenous midazolam 0.25 mg was administered on both occasions. There were no intraoperative episodes of upper airway obstruction requiring manual repositioning of the airway. Dexmedetomidine was administered intraoperatively at a dose of 0.5–0.7 µg/kg/h and discontinued upon completion of surgery. His intraoperative OAA/S score was 3, with a mean arterial pressure of 62–80 mm Hg, a heart rate of 55–65 beats/min, an SpO₂ of 94%–97%, and an end-tidal CO₂ of 55–60 mm Hg. The total doses of midazolam and dexmedetomidine administered were 1.5 mg and 44 µg, respectively. The surgical intervention time was 45 minutes, and the duration of anesthesia was 68 minutes. The patient's vital signs and respiratory status stabilized soon after his surgery, and he was discharged on the following day.

DISCUSSION

The issues to be addressed during dental treatment and sedation in this patient included worsening of respiratory depression and deglutition disorder in response to

bending and stretching of the neck, aspiration because of a weak cough reflex and deglutition disorder, CO₂ narcosis as a result of hypoventilation, and potential anaphylactic shock because of latex allergy.

Propofol was selected for the first intravenous sedation procedure as it could be easily titrated as needed. Although deep sedation was achieved without movement of the patient's head during treatment, aspiration pneumonitis occurred postoperatively. Propofol is a general anesthetic agent that is used for intravenous sedation during dental procedures, but it causes respiratory depression that can trigger apnea during surgery.¹⁴ In this patient, there were several episodes of airway obstruction during sedation with propofol. These episodes were managed by decreasing the propofol infusion rate and repositioning the airway manually by lifting the jaw. The association between propofol and postoperative aspiration pneumonia at the time of the first intravenous sedation procedure is not clear. However, when propofol is administered to patients with type II ACM to suppress body movements, it is possible that obstruction of the airway by the tongue adds to suppression of the swallowing center, creating conditions that allow aspiration. Furthermore, because bending and stretching of the neck may worsen impairment of the respiratory and swallowing centers in patients with type II ACM, deep sedation/nonintubated general anesthesia should be avoided, if possible, to limit the need for airway positioning. If airway opening/repositioning is necessary, it should be performed by a jaw lift maneuver without neck extension. Intubation should be performed with in-line cervical stabilization.

Dexmedetomidine is an α_2 -receptor agonist with sedative, antianxiety, and analgesic actions and reduces sympathetic tone, thus having minimal inhibitory effects on respiration.¹⁵ It has been reported that intravenous dexmedetomidine is useful for sedation during dental treatment,^{11–13} particularly in patients at high risk of respiratory depression.¹⁰ Postoperative aspiration pneumonia did not occur after the second intravenous sedation procedure in our patient, possibly because administration of dexmedetomidine to suppress body movements caused less displacement of the tongue and suppression of the swallowing center than propofol. However, the sympathetic reaction in response to stimulation of central nervous system α_2 receptors is inhibited, resulting in decreased blood pressure and heart rate, so careful monitoring of circulatory dynamics is required. The level of consciousness during intravenous sedation with dexmedetomidine at clinical concentrations is similar to that during natural sleep, and patients are easily arousable in response to stimulation.¹⁶ Therefore, we considered that sedation of a patient with an intellectual disability using dexmedetomidine alone would be insufficient, and the

addition of another sedative would be necessary. We considered low-dose propofol as the concomitant agent but felt that its inhibitory effect on the sympathetic nervous system would worsen the decrease in blood pressure and heart rate already caused by dexmedetomidine and could trigger hypotension and bradycardia. Therefore, we selected midazolam as the concomitant drug. Agitation in this patient prior to dexmedetomidine exerting its effect would likely have caused body movement, so intravenous midazolam was administered at the start of the dexmedetomidine infusion. Given that midazolam was required on 2 occasions to suppress body movement during surgery in this patient, dental treatment with dexmedetomidine alone would likely have been difficult in this patient with intellectual disability. Moreover, adequate adjuvant local anesthesia was deemed necessary. Sedatives such as dexmedetomidine and midazolam act synergistically,¹⁷ allowing a reduction in the dose of each agent when they are used concomitantly.¹⁸ In this patient, the combination of dexmedetomidine and midazolam achieved an adequate sedative effect with no hypotension or bradycardia. Moreover, dexmedetomidine has the advantage of suppressing secretion of saliva,¹⁹ which may also have contributed to creating a more favorable environment that reduced the risk of aspiration.

Type 2 respiratory failure is characterized by hypoxemia and hypercapnia ($\text{PaO}_2 < 60$ mm Hg, $\text{PaCO}_2 > 50$ mm Hg), and caution is needed in patients with type 2 respiratory failure because of the risk of CO_2 narcosis in response to hypoventilation. Moreover, there is a marked increase in PaCO_2 in patients with type 2 respiratory failure that creates a predominantly hypoxic drive in the respiratory center, so there is the risk of cessation of respiration when a high concentration of oxygen is administered. In this patient, oxygen was administered with careful monitoring of the concentration inhaled via the nasal cannula. A capnometer and pulse oximeter were used to monitor the patient's respiration during surgery, and care was taken to avoid an excessive increase in SpO_2 . The combination of dexmedetomidine and midazolam causes little respiratory depression,²⁰ so it is a suitable method for sedation in patients with type 2 respiratory failure.

Latex allergy is common in patients with a history of repeated surgery, and the rate in patients with spina bifida has been reported to be as high as 38%.²¹ Our patient had a history of 3 episodes of a systemic inflammatory response to latex and so was at high risk of significant allergic reaction or anaphylactic shock. Therefore, latex-free products were used. The patient's skin and vital signs were monitored closely during surgery, and there were no signs of perioperative latex allergy. Nevertheless, patients with type II ACM are

likely to be at high risk for latex allergy, so caution is always required.

In conclusion, type II ACM is often complicated by respiratory depression and deglutition disorder, which can be worsened by tilting of the head and stretching of the neck. Therefore, an anesthetic agent that causes minimal respiratory depression but adequately inhibits body movement should be selected for intravenous sedation when undertaking dental treatment in these patients. We performed a successful intravenous sedation procedure using a combination of midazolam and dexmedetomidine in a patient with type II ACM and hypercapnic respiratory failure. Safe and effective management of sedation with minimal variation in respiratory depression and circulatory dynamics was achieved.

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