Indonesian Journal of Anesthesiology and Critical Care Medicine

Journal homepage : https://ijaccm.org

Procedural Sedation Management in Patients Suspected of West Syndrome

Lyanda Watung[1], Eka Yudha Lantang[1],Antonio B. Lengkong[1]

[1]Department Anestesiologi dan Terapi Intensif, Fakultas Kedokteran, universitas Sam Ratulangi, Manado, Indonesia

ARTICLE INFO

Keywords :
West syndrome,
Peripheral venous access difficulty,
Infantile spasms

ABSTRACT

West syndrome is a rare epileptic disorder characterized by a triad of infantile spasms, mental retardation, and an interictal electroencephalogram (EEG) pattern known as hypsarrhythmia. Comprehensive examination is necessary in anesthetic preparation for these patients due to potential difficulties in airway management resulting from anatomical malformations, challenges in peripheral venous access, mobility issues caused by contractures, seizure activity, and adverse effects of medications therapy. This report details the anesthetic management of a patient suspected of West syndrome undergoing procedural sedation for magnetic resonance imaging (MRI), utilizing total intravenous anesthesia (TIVA).

Citation: Watung L, Lantang EY, Lengkong AB. Procedural Sedation Management in Patients Suspected of West Syndrome. *Indonesian Journal of Anesthesiology and Critical Care Medicine*.2026;1(2):80-85

1. Introduction

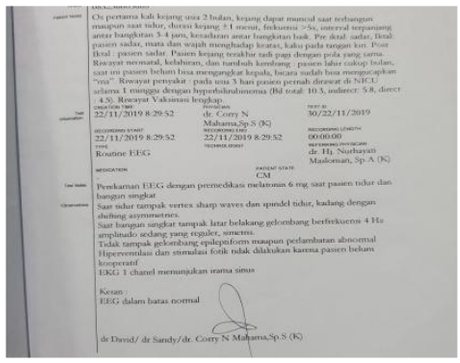
West syndrome is an uncommon disorder predominantly affecting children, characterized by a triad comprising infantile spasms, mental retardation, and an interictal EEG pattern called hypsarrhythmia. First described by British physician W.J. West in 1841, diagnosis can be made even if one component of the triad is absent. Its prevalence ranges from 1 in 2,000 to 6,000 live births, typically manifesting within the first 3 to 7 months of life, often before age 2. The syndrome can result from various causes including hypoxic-ischemic brain injury, hemorrhage, trauma, infections, genetic syndromes, or malformations occurring before, during, or after birth. The underlying pathophysiology is believed to involve abnormal connections between the cerebral cortex and brainstem. Patients with West syndrome frequently require anesthesia, whether for diagnostic procedures or surgical interventions. Preoperative evaluation must be comprehensive to anticipate challenges such as vascular access, airway management, seizure control, and interactions of anticonvulsant medications with anesthetics. This case report discusses perioperative management of a suspected West syndrome patient undergoing MRI under sedation.

2. Case Report

Figure 1. Patient in sedation preparation

A male infant aged 8 months weighing 7.6 kg underwent a series of diagnostic procedures to investigate West syndrome. The patient experienced seizures starting at 2 months of age characterized by unpredictable episodes occurring during activity or sleep, lasting approximately one minute, with a frequency exceeding five episodes per day. The longest interval between episodes was 3–4 hours, with preserved consciousness between seizures. During seizures, the eyes and face turned upward, with limbs extended; post-ictally, consciousness was regained. The patient's birth history was unremarkable, with normal term delivery and no perinatal complications. He was hospitalized briefly at 3 days of age for hyperbilirubinemia (total bilirubin 10.3 mg/dL). No history of falls was reported. Developmental delays included inability to lift the head or prone position, and limited speech (only uttering "Ma"). The immunization status was complete. The infant was exclusively breastfed for six months, with no known allergies. No prior anesthesia or surgeries were documented. Vital signs were within normal limits. Physical examination revealed no craniofacial malformations, interincisor distance, hyoid-mental distance, thyroid to hyoid distance all within normal range. Airway assessment show complete visualization of the soft palate. Cardiorespiratory, abdominal, and extremity examinations were unremarkable. Laboratory tests, including hematology, biochemistry, and coagulation profiles, were normal. EEG during sleep showed vertex sharp waves and sleep spindles, sometimes with asymmetrical shifting; awake EEG demonstrated rhythmic 4 Hz waves of moderate amplitude, symmetrical, without epileptiform discharges or slowing. Hyperventilation and photic stimulation were not performed due to

uncooperative patient. MRI imaging was scheduled after a six-hour fast, as prior Brainstem Evoked Response Audiometry (BERA) was not feasible.



Cb pertama kali kejang usia 2 bulan, kejang dapat muncul saat terbangun maupun saat tidur, durasi kejang ± 1 menit, frekuensi >5x, interval terpanjang antar bangkitan 3-4 jam, kesadaran antar bangkitan baik. Pre ictal: sadar, Ictal: pasien sadar, mata dan wajah menghadap kanan, kaku pada tangan kiri. Post Ictal : pasien sadar. Pasien kejang terakhir tadi pagi dengan pola yang sama. Riwayat neonatal, kelahiran, dan tumbuh kembang : pasien lahir cukup bulan, saat ini pasien belum bisa mengangkat kepala, bicara sudah bisa mengucapkan "ma". Riwayat penyakit : pada usia 3 hari pasien pernah dirawat di NICU selama 1 minggu dengan hiperbilirubinemia (Bil total: 10.3, indirect: 5.8, direct 4.5). Riwayat Vaksinasi lengkap.

CREATION TIME 22/11/2019 8:29:52 | PHYSICIAN dr. Corry N Mahama,Sp.S (K) | TEST ID 30/22/11/2019

RECORDING START 22/11/2019 8:29:52 | RECORDING END 22/11/2019 8:29:52 | RECORDING LENGTH 00:06:00

TYPE Routine EEG | TECHNICIAN dr. Hj. Nurhayati Maliomsan, Sp.A (K)

MEDICATION | PATIENT STATE CM

Pemeriksaan EEG dengan premedikasi melatonin 6 mg saat pasien tidur dan bangun singkat

Saat tidur tampak vertex sharp waves dan spindel tidur, kadang dengan shifting asymmetries.

Saat bangun singkat tampak latar belakang gelombang berfrekuensi 4 Hz amplitudo sedang yang reguler, simetris.

Tidak tampak gelombang epileptiform maupun perlambatan abnormal

Hiperventilasi dan stimulasi fotik tidak dilakukan karena pasien belum kooperatif

EKG 1 chanel menunjukan irama sinus

Kesan :

EEG dalam batas normal

dr David/ dr Sandy/dr. Corry N. Mahama,Sp.S (K)

Figure 2. EEG Output of patient

Informed consent was obtained from the parents after explaining potential risks. Fasting instructions, given to the patient before the procedure. During preparation, peripheral venous access was challenging, requiring approximately ten attempts in different limbs. Prior to induction, vital signs included a heart rate of 135 breaths per minute, respiratory rate of 26 breaths per minute, sinus rhythm on ECG, and oxygen saturation of 98% with room air. Monitors were set up in accordance with American Society of Anesthesiologists (ASA) standards. Premedication was administered with midazolam 0.03 mg/kg. Induction with intravenous propofol 1 mg/kg, titrated according to patient response, with spontaneous breathing through a nasal cannula at 2 L/min. Anesthesia was maintained with intermittent boluses of propofol. Body temperature is monitored and the air in the room is controlled to prevent hypothermia including using blankets during the procedure. Hemodynamic parameters remained stable throughout the procedure. Post-procedure, the patient was observed in the Post-Anesthesia Care Unit and discharged after reaching a Pediatric Anesthesia Discharge Score (PADS) of 10, with parental education provided regarding outpatient anesthesia care.

Figure 3. Sedated patient undergoing mri examination

3. Discussion

Dr. William James West first described infantile spasms in his son, James Edwin West, at four months of age in 1841, with clinical features including head flexion and limb extension (2). The syndrome was later named West syndrome by Milichap in the early 1960s. Historically, treatment options were limited, but reports by Sorel and Dausaucy described the use of adrenocorticotropic hormone (ACTH) for infantile spasms. The prognosis for West syndrome is generally poor, often resistant to conventional antiepileptic drugs. Etiologically, it is heterogeneous, with hypoxic-ischemic encephalopathy and infections being common causes; tuberous sclerosis accounts for 10–30% of cases, often associated with cardiac and renal tumors (1). Patients with West syndrome typically exhibit global developmental delays and mental retardation. Anesthetic management

presents challenges during preoperative, intraoperative, and postoperative phases, including potential airway malformations, excessive salivation, and difficulties in airway management. If excessive salivation is present, suctioning of mucus is necessary during intubation and extubation. Post-anesthetic respiratory support and prolonged observation in intensive care are recommended for optimal recovery. Hemodynamic instability and hypothermia are potential intraoperative concerns, especially considering nutritional deficits and hypothalamic dysfunction. Proper patient positioning with padding is crucial to prevent fractures due to skeletal fragility (3).

Anesthesia procedures in pediatric patients differ significantly from those in adults. Children exhibit changes in hemodynamics, immune responses, hormonal regulation, and stress-related metabolic responses, which can impair recovery and increase morbidity, as discussed by Mahajan et al (4). The most significant challenge our patient faced was the establishment of peripheral vascular access. As noted by Naik et al. (5), establishing intravenous access in children can be difficult and stressful, especially for the child and family, due to small vein caliber, anatomical variations, and anxiety. Based on the Difficult Intravenous Access (DIVA) score, our patient scored 7 out of 10, indicating a high degree of difficulty in peripheral access placement. Obstruction during cannulation can also occur due to infantile vasospasm, which is common in West syndrome. However, this was not the case in our case. MRI procedures in children require sedation to relieve agitation, anxiety, pain, and reduce movement, as restraint is not recommended due to potential risks. Pre-sedation protocols should adhere to American Society of Anesthesiologists (ASA) guidelines, which include appropriate equipment, monitoring, and medications.

When planning anesthesia for a patient, it is important to evaluate the side effects and interactions of medications the patient is routinely taking. Patients with West syndrome take adrenocorticotropic hormone (ACTH), vigabatrin, and anticonvulsant drugs such as diazepam and phenytoin (6). ACTH or other corticosteroid therapy can effectively control seizures but has side effects such as osteoporosis, cardiac hypertrophy, hypertension, risk of infection, electrolyte imbalance, behavioral disturbances, and weight gain. Meanwhile, long-term use of vigabatrin can cause visual disturbances. Administration of antiepileptic drugs also often causes periodontal problems, the most common of which are dental caries, gingival hyperplasia with bleeding and petechiae, and decreased platelet aggregation, such as with long-term use of valproate. Chronic anticonvulsant use can also impair hepatic enzyme activity, affecting anesthetic drug metabolism. Various side effects and long-term effects of drugs, especially those given to West syndrome, increase the risks of surgery and anesthesia.

Anesthetic agents should ideally provide sedation, analgesia, hemodynamic stability, rapid recovery, minimal adverse reactions, and availability of an antidote. The drugs recommended for short-term procedures come from the opioid group, such as fentanyl. The required dose starts at 1-4 mcg/kg, then titrates to 0.5-2 mcg/kg. For airway management challenges, nasotracheal intubation with fiber-optic techniques under sedation with remifentanil has shown safety. Because low doses of fentanyl may not achieve sufficient sedation, its combination with midazolam is recommended. Both drugs are considered safe and commonly used in pediatric sedation procedures. In this case, premedication with midazolam was performed at a dose of 0.03 mg/kg. Induction was performed with intravenous injection of propofol 1 mg/kg, titrated according to the patient's response. Perks et al. (7) suggested that benzodiazepines, such as midazolam, have anticonvulsant properties, making them beneficial for patients experiencing seizures such as West Syndrome. The conclusion of this study is that procedural sedation in pediatric patients should avoid hypoxia and hypercarbia, which can lower the patient's seizure threshold. The use of midazolam in a study by Akbulut et al. (8) was shown to provide comfort for pediatric patients

undergoing endoscopy. This study compared the combination of midazolam-ketamine and fentanyl-propofol, with results concluding that the use of midazolam-ketamine provided a better sedative effect for patients, but the use of both drugs required a longer recovery time compared to fentanyl-propofol. The use of propofol is starting to expand in the sedation of pediatric patients, although some studies have shown contradictory results. Etchu et al. in 2000 (9) reported the occurrence of intraoperative seizures due to propofol in a 75-year-old patient undergoing microlaryngeal surgery under total intravenous anesthesia using propofol. Yamaguchi (10) presented a different view in his study using Propofol for induction and maintenance of anesthesia in patients with West Syndrome. This study suggested that the use of propofol is an adequate anesthetic for this case, when used cautiously will not lower the seizure threshold. Another clinical investigation presented by Havel et al. (11) showed that the use of propofol can induce sedation more quickly than midazolam and has a faster recovery time after sedation. In this case, at the end of the MRI procedure, no significant problems or difficulties were found related to the use of midazolam and propofol. During anesthesia, there were no seizures or other medical problems. This indicates that midazolam and propofol do not lower the seizure threshold in our patient. Furthermore, recovery from sedation with both drugs occurred in a relatively short time in our patient, thus also indicating that midazolam and propofol are safe both intra- and post-sedation.

4. Conclusion

Comprehensive preoperative assessment, meticulous anesthetic planning and measures to prevent seizure precipitations are essential in managing patients with West syndrome. The use of midazolam and propofol for procedural sedation appears safe and effective based on this case. Continuous evaluation and tailored anesthesia protocols are recommended to ensure safety and optimal outcomes in this vulnerable patient population, with particular attention to avoiding hypoxia and hypercarbia, which can lower seizure thresholds.

Funding

None

Acknowledgments

None

REFERENCES

1. Phulkar P, Waghalkar P. Anesthetic Management of a Patient with West Syndrome. Journal of Anesthesia and Critical Care Case Reports. 2018;4(2):11-13.
2. Eling P, Renier WO, Baram TZ. The Mystery of the Doctor's Son, or The Riddle of West Syndrome.Neurology. 2002;58(6):953-955.
3. Dálonzo R, Rigante D, Mencaroni E, Esposito S. West Syndrome: A Review and Guide for Paediatricians. Clin Drug Investig. 2019;38(2):113-124.
4. Mahajan C, Dash HH. Procedural Sedation and Analgesia in Pediatric Patients. J Pediatr Neurosci. 2014;9(1):1-6.

5. Naik VM, Mantha SS, Rayani BK. Vascular Access in Children. Indian J Anaesth. 2019;63(9):737-745.
6. Sahin SH, Copuru E, Colak A. Anaesthesia Management of a Child with West Syndrome. Turk J Anaesthesiol Reanim. 2014;42(6):362-364.
7. Perks A, Cheema S, Mohanraj R. Anaesthesia and Epilepsy. British Journal of Anaesthesia. 2012;108(4):562-571.
8. Akbulut et al. A Comparison of Sedation with Midazolam-Ketamine versus Propofol-Fentanyl during endoscopy in Children: a randomized trial. European Journal of Gastroenterology and Hepatology. 2017;29(1):112–118.
9. Etchu H, Kurahashi K, Kitamura S. A Case of Convulsion Induced by Propofol. Masui. 1998;47(1):77-8
10. Yamaguchi S, Mishio M, Okuda Y, Kitajima T. Anesthetic management of a patient with West Syndrome. Masui. 2000;94:69–71.
11. Havel CJ, et al. A Clinical Trial of Propofol vs. Midazolam for Procedural Sedation in a Pediatric Emergency Department. Academic Emergency Medicine. 1999;6:10.