International Journal of Biomedical Research
ISSN: 0976-9633 (Online); 2455-0566 (Print)
Journal DOI: https://doi.org/10.7439/ijbr
CODEN: IJBRFA

Letter to Editor

Anaesthetic management of a child with West Syndrome: A case report

Sachinkumar Wagh[*1], P S Garcha[1], Sachin Hiradeve[2]

[1]*Departments of Anaesthesiology, D. Y. Patil Dental College, Pune, Maharashtra, India*
[2]*School of Pharmacy, G.H. Raisoni University, Chhindwara, Madhya Pradesh, India*

***Correspondence Info:**
Dr. Sachinkumar Wagh,
Department of Anaesthesiology,
D. Y. Patil Dental College, Pimpri,
Pune, Maharashtra- 411018, India

***Article History:**
Received: 17/06/2019
Revised: 26/06/2019
Accepted: 27/06/2019
DOI: https://doi.org/10.7439/ijbr.v10i6.5206

QR Code

How to cite: Wagh S, Garcha P and Hiradeve S. Anaesthetic management of a child with West Syndrome: A case report. *International Journal of Biomedical Research* 2019; 10(06): e5206. DOI: 10.7439/ijbr.v10i6.5206
Available from: https://ssjournals.com/index.php/ijbr/article/view/5206

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Dear Editor,

West syndrome (WS) is one of the malignant epilepsies rarely seen in the infantile period, which consists of the hypsarrhythmia triad, characterised by infantile spasm, mental disorder and electroencephalography (EEG) abnormality.[1,2] This syndrome can result from brain dysfunction in the prenatal, perinatal (during delivery) or postnatal period. Its aetiology includes infections, trauma, hypoxic-ischemic or haemorrhagic causes, malformation syndromes and genetic syndromes. Its pathophysiology contains an abnormal interaction between the brain stem and cortex.[3] Prednisone, adrenocorticotropic hormone (ACTH), antiepileptic drugs and benzodiazepine are used for the treatment of West syndrome.

A preoperative evaluation should be carefully performed for these patients, considering aetiology of disease, possible difficulties in intubation, anatomic malformations, side effects of the drugs and epileptic seizures. This case report aims to present anaesthesia management in a patient with WS who was planned to undergo Dental Rehabilitation under General Anaesthesia.

The patient was 6 years old and 18 kg male with WS who required general anaesthesia for dental rehabilitation. The written and informed consent was obtained from guardian. The patient was diagnosed as case of WS when he presented with infantile spasm and developmental delay at the age of one and half year old. He was treated with polytherapy which included steroids, zonisamide, valproic acid and leviracetam. The physical examination of his appearance revealed a prominent ear, development delay and micrognathia. The preoperative investigations Haemogram, Renal function test and Liver function test, 2-D echo, Electroencephalogram was found to be within normal limits.

At present, the child showed no seizures since two years on leviracetam 500 mg/day. Aspiration prophylaxis with injectable ranitidine (1 mg/kg) and ondansetron (0.15 mg/kg) was given. Protection such as a cushion was used at bony prominences to prevent pressure sores. The patient was tilted 30° head-up, pre-oxygenated and rapid sequence induction was carried out using Thiopentone 5mg/kg preceded with Midazolam 0.05 mg/kg. Muscle relaxation was provided with rocuronium 1 mg/kg. The trachea was nasally intubated with a size 6 North Pole tube.

Maintenance of anaesthesia was achieved by sevoflurane. Following the procedure that lasted 90 min, residual neuromuscular blockade was reversed with neostigmine 1 mg and glycopyrrolate 0.08 mg. After the patient demonstrated airway protective reflexes, spontaneous eye opening, and a regular respiratory pattern, the trachea was extubated. Postoperative analgesia was obtained with paracetamol (15 mg/kg). Recovery was uneventful and the patient was subsequently discharged.

West syndrome consisted of the triad of “infantile spasm, physical and mental disorder and characteristic EEG findings”. The characteristic EEG finding was named “hypsarrhythmia. The aetiology for the occurrence of West syndrome includes, especially genetic and a malformation syndrome, hypoxic-ischemic or haemorrhagic causes central nervous system infections and trauma. Tuberous sclerosis accounts for 10%–30% of the causes in the prenatal period. Since tuberous sclerosis is a syndrome

involving cardiac and kidney tumours, it is important for anaesthesia.[3] the underlying causes should be carefully investigated in the preoperative evaluation of cases.

Infantile spasms are generally resistant to antiepileptic drugs. The mechanism of steroid therapy is not known (ACTH or prednisone), it can yield better results. Recently, positive results have been reported, particularly with vigabatrin treatment, among antiepileptic drugs. However, long-term use of high-dose vigabatrin leads to visual field constriction in nearly half of the patients.[4] Despite the fact that ACTH is effective in seizure control for most WS patients, it has serious side effects, including osteoporosis, cardiac hypertrophy, hypertension, tendency to infection, electrolyte imbalance, behavioural changes and weight gain.[5] While evaluating patients in the preoperative period, the drugs that they use should be investigated in detail, and laboratory findings should be assessed. Therefore, the underlying causes should be carefully investigated in the preoperative evaluation of cases, and the appropriate anaesthetic method and agents should be preferred.

In these patients developmental delay and micrognathia reminding difficult intubation. No problem was encountered during intubation carried out in the supine position. Nonetheless, this physiological posture of the patient requires attention to the intraoperative position. Positioning was performed carefully in order to avoid re-occurrence of the fracture. The joints of the patients were supported with pads. Convulsions should be taken into consideration while administering anaesthetic agents in patients with West syndrome.[6-8] In our case, midazolam was used for premedication due to its anxiolytic, sedative and anticonvulsive effects and on day of surgery continued Leviracetam 500 mg/day. It was revealed that the agitation occurring during sevoflurane anaesthesia was not associated with seizures and that sevoflurane was not contraindicated in patients with epilepsy.[9-12]

Thiopentone was employed in induction in order to avoid the side effects of high-dose inhalation agents. Sevoflurane, which is an anaesthetic commonly used in child and has become clear for convulsion, even a little, was preferred for maintenance.

We have described the successful management of a child with West Syndrome. We have mentioned all the investigations, and as a rare case, documentation is always helpful in further evolving management in medicine.

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