International Journal of Anesthesiology Sciences
Online ISSN: 2664-9276, Print ISSN: 2664-9268
Received: 06-05-2019; Accepted: 07-06-2019; Published: 14-06-2019
www.anesthesiologyjournals.com
Volume 1; Issue 1; 2019; Page No. 16-17

Aicardi syndrome: What the anaesthesiologist should know about management of a child with Aicardi syndrome

Dr. Sonal T Shah[1], Dr. Abhilasha D Motghare[2]

[1] Consultant Anesthesiologist, Department of Anesthesiology, Saifee Hospital, Maharshi Karve Marg, Charni Road, Mumbai, Maharashtra, India

[2] Anaesthesiology Resident(ex), Department of Anesthesiology, Saifee Hospital, Maharshi Karve Marg, Charni Road, Mumbai, Maharashtra, India

Abstract
Aicardi syndrome is a rare congenital disorder characterised by a triad of callosal agenesis, infantile spasms and chorioretinal lacunae along with other congenital defects of the eyes, ribs, vertebrae and frequently associated with mental retardation and intractable seizures. The rare syndromes are challenge to mange by the physicians and when such patients are posted for any surgery same challenge is faced by the anesthesiologist. We report a case of Aicardi syndrome posted for oral rehabilitation procedure.

Keywords: Aicardi syndrome, anesthesia management, intractable seizures

Introduction
Aicardi syndrome [1]. is a rare genetic malformation characterized by a classical triad of infantile spasms, corpus callosum agenesis and chorioretinal lacunae. The additional characteristics include abnormal facies, cleft lip and palate, vertebral body anamolies and abnormalities of neuronal migration. It is X linked dominant condition [2] found especially in females. It's a rare disorder [3] with very few cases worldwide (although the exact prevalence and incidence is unknown). The estimated incidence [4] in the United States is 1 in 105000 live births. These patients have profound psychomotor impairment with no meaningful speech. Their seizures start in early childhood, require multiple anti-epileptic medications and are usually intractable. We present the anaesthesia management of a child with multiple caries teeth posted for oral rehabilitation procedure under general anaesthesia. A short discussion on the perioperative anaesthetic concerns is presented.

Case Report
A 5 years old, 16 kg female child presented with multiple caries teeth posted for full mouth rehabilitation procedure which was to last for two to three hours. She had history of generalized convulsions 3-4 times a day especially on awakening. She was on syrup sodium valproate 200mg and tab nitrazepam 2 mg in divided doses. The parents used nasal midazolam spray as and when required for control of seizures. She had delayed milestones with no speech, inability to walk and no bladder bowel control. The patient was evaluated preoperatively and investigations like complete blood count, chest X ray, liver function test, renal function test, blood glucose levels were done along with a clearance fitness from a pediatric neurophysician.

The child was premedicated with oral nitrazepam 2 mg previous night and the anticonvulsants were continued on the day of surgery. Prilox gel was applied 90 mins prior to IV canulation. After IV canulation patient was sedated with inj Midazolam 1.0 mg and inj Fentanyl 15 mcg and shifted to operating room. She was induced with Inj Thiopentone sodium 80 mg and Inj Atracurium 8 mg and a nasotracheal ETT 5.0 cuffed was inserted and throat packing done. She was maintained on O_2 and N_2O (50:50) with isoflurane to maintain minimal alveolar concentration (MAC) 1. After 120 minutes of uneventful intra-operative course we repeated inj Midazolam 0.5 mg before neuromuscular blockade reversal with inj Neostigmine 0.8 mg and inj Glycopyrolate 128 mcg and extubated her in a deep plane of anesthesia keeping in mind that she had h/o convulsions on awakening. Further she was observed in recovery room for an hour and then shifted to pediatric intensive care unit.

Discussion
Aicardi syndrome is a severe neurodegenerative encephalopathy. It pose a significant challenge to anesthesiologist in view of resistant convulsions. They have poor swallowing reflexes, generalized hypertrophied subcutaneous tissue causing difficult venous canulation, severe vertebral malformation making caudal anesthesia difficult [5], tongue hypotonia and macroglossia making intubation difficult. We must also consider the impact of anticonvulsants on organ function, coagulation and interaction with anesthesia drugs. Co-existing sedation produced by anticonvulsants may have an additive effect with anesthetics, whereas drug induced enzyme induction could alter response to other drugs or contribute to systemic toxicity associated with administration of halothane and enflurane (though not used).

In view of availability of safe drugs which do not lower seizure threshold, it would seem reasonable to avoid potentially epileptogenic drugs. Thiobarbiturates, opioids, benzodiazepines, isoflurane, desflurane would all seem good choices. Regardless of the drugs used for anesthesia, it is important to maintain treatment with pre-established anticonvulsants throughout the perioperative period. Also

advanced monitoring like electroencephalogram and bispectral index for early detection of eplipetiform activity is suggested wherever feasible.

We wanted to report that above approach helped us in uneventful management of this rare syndromic disorder.

Conflicts of interest: None

References

1. O'Brien, Lowe D. Anesthetic implications of Aicardi's syndrome; European Journal of Anesthesiology (EJA): November. 2002; 19(11):845-846.
2. Guadagni MG, Faggella A. Aicardi syndrome: a case report; European Journal of Pediatric dentistry Vol 11/3, 2010.
3. https://emedicine.medscape.com Aicardi syndrome: Background, Pathophysiology, Epidemiology accessed on 13/9/18
4. Kroner BL, Preiss LR, Ardini MA, Gaillard WD. New incidence, prevalence, and survival of Aicardi syndrome from 408 cases. J Child Neurol. 2008; 23(5):531-5.
5. Lacobucci T. Anesthesia management in a patient with Aicardi's syndrome. Anesthesia. 2003; 58(1):95-95.
6. Aicardi J. Aicardi syndrome. Brain Dev. 2005; 27(3):164-71.
7. Aicardi J, Lefebvre J, Lerique-Koechlin A. A new syndrome: spasm in flexion, callosal agenesis, ocular abnormalities. Electroencephalogra Clin Neurophysiol, 1965; 19:609-10.
8. Dennis J, Bower BD. The Aicardi syndrome. Dev Med Child Neurol.1972 Jun.14 (3):382-90.
9. Iacobucci T, Galeone M, De Francisci G. Anaesthesia management in a patient with Aicardi's Syndrome. Anesthesia, 2003; 58:95-95.
10. Terakawa Y, Miwa T, Mizuno Y, Ichinohe T, Kaneko Y, Ka K. *et al*. Anesthetic management of a child with Aicardi syndrome undergoing laparoscopic Nissen's fundoplication: a case report. J Anesth. 2011; 25(1):123-6.