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Anaesthetic Management of A Child With Aicardi Syndrome Undergoing Total Mouth Rehabilitation

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INTRODUCTION

This case report describes anesthetic considerations in a 7 year old boy with Aicardi syndrome posted for total mouth rehabilitation. The patient had history of generalized convulsions for which he was receiving antiepileptics. Child underwent circumcision under general anesthesia at 4 years of age which was uneventful. This had a positive impact on our anesthetic plan.

Aicardi syndrome (AS) is a rare congenital syndrome and is characterized by the triad of infantile spasm, agenesis of the corpus callosum and anomaly of chorioretinal lacunae. We here report a case of a patient with AS done under general anaesthesia. Although there is no report in which specific muscle relaxants were used in AS patients, vecuronium bromide was used as muscle relaxant in this case. Careful management is important for AS patients during an operation that significantly affects respiratory function. In addition, it is possible that muscle relaxants be administered safely in AS patients. Careful monitoring such as epileptiform electroencephalogram and bispectral index monitors may be needed for the early detection of epileptic activities.

SPECIAL FEATURES OF AS

It is a genetic mutational dominant disease.The following are features of Aicardi syndrome

Cortical malformations,periventricular anom alies.It occurs extensively in females than in males,found in klinefelter syndrome. Etiology:It is x-linked and Sub cortical heterotopia,-cysts around third ventricle and choroid plexus,Papilloma of Choroid Plexus,Verte bral and costal abnormalities,Microphthalmia.

Symptoms**:**
Coloboma refering to eye gaping,Microcephaly,Hand deformities,Developmental delay,Spasticity,Gastro

oesophageal reflux,Scoliosis.

Diagnosis**:**
Aicardi syndrome is diagnosed by Ocular examination,Electoencephalogram,MRI brain imaging.

MRI pictures of the patient revealed the presenceof genu associated with agenesis of the rest of corpus callosum.

A funduscopic examination revealed bilateral small, solitary, pale areas with sharp borders, some of which had minimal surrounding pigmentation (chorioretinal lacunae), especially clustered around the disc, and they were more prominent on the left side. We report here on the unusual findings of a complex partial seizure, myoclonic seizure and the atypical EEG finding in addition to the well-known characteristic clinical and imaging findings of a patient with Aicardi syndrome.

Anaesthetic Management**:**
A 7year old boy, weighing 22 kg, was posted for total mouth rehabilitation. The diagnosis of Aicardi syndrome was made when he was 4 years of age. Since early childhood he had frequent respiratory tract infection, cough and cold. The child had also history of delayed developmental milestones, palpitation and shortness of breath on exertion. He has undergone circumcision under general anaesthesia at the age of 4. Thereafter, the child had normal growth with no significant medical history. He was on oral Frisium 5mg per day.There was no significant family history for anesthetic complications or congenital abnormalities.

On examination, the child had, pulse rate 92/min, blood pressure 110/64 mmHg, Mallampati score – grade 2, normal neck movement, adequate mouth opening. On auscultation, a systolic murmur was heard on left second intercostal space. The hemoglobin was 12.8 gm/dl. Serum electrolytes, blood glucose and creatinine were normal. Hematological investigations showed no coagulation or platelet defects. Echocardiography showed mild pulmonary regurgitation.

On arrival in the operating room, pulse oximeter, cardiac monitor, blood pressure cuff were attached. The child was cooperative. Peripheral venous access was secured using 22 G cannula. Antibiotic prophylaxis was given with IV 500 mg Taxim and 250 mg amikacin.. Premedication was given with 1 mg midazolam, 0.1 mg glycopyrrolate, 50 µg fentanyl IV. Induction was done with propofol 50 mg IV. After establishing adequate bag mask ventilation, inj vecuronium 2.2 mg was given IV. After 3 min bag mask ventilation, intubation was done with 5 mm cuffed nasotracheal tube. Neuromuscular blockade was monitored using a peripheral nerve stimulator. Temperature was monitored. Maintenance of anesthesia was done with oxygen, nitrous oxide, sevoflurane, vecuronium bromide in the 1.5 hour procedure. Paracetamol infusion 450 mg was given as analgesic. Ventilation was controlled to achieve an $EtCO_2$ of 35-40 mmHg, and the SpO_2 to be maintained between 95% and 100%. To fulfil the fluid deficit 550 ml lactated ringers solution was infused. At the end of the procedure, neuromuscular blockade was reversed with the intravenous administration of glycopyrrolate and neostigmine. Extubation was done after the return of rhythmic breathing and return of protective airway reflexes. The child was given postoperative oxygenation for 5 min and kept in post anesthesia care unit and careful monitoring was done for 24 hours. Diclofenac sodium was given as postoperative analgesic in infusion. Postoperative period was uneventful. The child was extubated and when fully awake shifed to high dependency unit..

General Anesthetic considerations

Infantile spasm causing seizure is drug resistant.Swallowing functionis underdeveloped due to corpus callosum agenesis.Difficult airway due to facial anomaly is another challenge during intubation Respiratory dysfunction occurs dur ing general anesthesia.Aspiration pneumonia is common cause of death.

Delayed recovery after surgery during extubation can oc cur which can be reduced by optimal doses of muscle relaxants.

RESULTS:

No significant cardiovascular changes occurred during general anaesthe-

sia. SPO2 maintained between 97-100% . End tidal CO2, meanly higher in this child, but never exceeded 45 mmHg. Similarly, peak inspiratory pressure was increased, but was always maintained within acceptable values. Finally, gentle and slower surgery was required, whose length seems to be negatively influenced by age. No intraoperative complication occurred. Recovery was comparatively smooth and uneventful.

CONCLUSION

The spectrum of motor and mental disabilities is wide and, their severity is to some extent determined by the severity of underlying brain abnormalities. Some patient with milder brain malformations may have some understanding of the language, an independent motility, and they may be responsive to their enviornment.The treatment is symptomatic for Aicardi syndrome, and this generally involves management of the epileptic seizures and an intervention program for the motor and mental retardation. The prognosis of the patients varies with the severity of underlying brain abnormalities and symptoms, and the life expectancy can be severely limited from several months to only a few years. In our case, a favorable prognosis could be expected because the infantile spasms had not developed since birth. Also, a favorable prognosis is expected because the agenesis of corpus callosum is partial and the chorioretinal lacunae are relatively small in size, isolated and they spare the macula. In conclusion, we were presented with a 7 year old male patient with Aicardi syndrome who manifested the unusual findings of a complex partial seizure, myoclonic seizure and atypical EEG finding in addtion to the well-known characteristic clinical and imaging findings of the syndrome. We have reported on this case along with a review of the literature.

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