IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 16, Issue 3 Ver. VIII (March. 2017), PP 126-127
www.iosrjournals.org

Dantrolene Sodium: A Need of the Day at Tertiary Centre.

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Abstract: *Malignant hyperthermia (MH) a rare genetic disorder is often associated with congenital disorders which are not diagnosed preoperatively. The incidence of MH being very low is still potentially a fatal disorder in anesthesia practice. Mortality still remains high despite its complex and demanding management. Reporting of MH and MH-like syndrome when treated timely with Dantrolene Sodium drastically decreses the mortality of the patients. This report describes a case of congenital cleft lip in a patient of 5 years with no any other congenital anomaly associated with it. Presented as MH like syndrome in post-operative period, which was diagnosed promptly and managed by Dantrolene sodium. We present this case report to emphasize the availability of DS at tertiary centers and at specialized centers where such surgeries are conducted routinely.*
Keywords: *Dantrolene Sodium (DS), Malignant Hyperthermia. (MH)*

I. Introduction

With advances in anesthesia, surgical technology and drugs, more and more complicated and congenitally diseased patient are being planned for surgery. The patients who are associated with one or more congenital defects or syndromes are not diagnosed clinically before surgery and may present as unwanted fatal emergencies during perioperative period. Malignant hyperthermia (MH) a rare genetic disorder with an incidence of 1:100000 has long association with many syndromes which are earlier been reported e.g Noonan syndrome, King Denborough Syndrome, Down's Syndrome, Muscular dystrophies, Arhthrogryposis etc [1,2] . The common features among all these syndrome remains to be congenital myopathy and cleft face abnormalities. Previous studies have also shown strong association with congenital cleft face abnormalities and MH (MH-like syndromes) [3]. Even after much research on MH and its association with cleft face abnormalities, its awareness at many tertiary centers remains questionable.

We present a case report of 5 year male child posted for cleft lip repair with no underlying disease at time of pre-anesthetic assessment. During intra-operative period there was rise in $EtCO_2$ from 35mmhg to 50mmhg associated with hyperpyrexia, tachycardia, flushing of whole body which continued in post-op period following extubation, mimicking like Malignant Hyperthermia (MH). Early clinical diagnosis of MH and prompt treatment with Dantrolene Sodium (DS) prevented fatal outcome in patient and was life saving. This case report emphasizes importance of prompt clinical diagnosis and treatment of Malignant Hyperthermia (MH) by life saving DS and also it emphasizes availability of DS at centers which are handling such category of patients.

II. Case Report

Five year old male child was scheduled to undergo cleft lip repair under general anesthesia, Preoperative anaesthesia evaluation revealed nothing significant. All relevant investigations were normal. There was no past history of anaesthetic exposure, malignant hyperthermia or neuromuscular disorders. There was no family history of any operation or such disease. After securing IV line patient was induced with Thiopentone 5 mg /kg, Succinyl Choline 2mg/kg, Fentanyl 2µg/kg. Anaesthesia was maintained with Halothane 0.6% + O_2 40% + N_2O 60% and Atracurium. After about $1^{1/2}$ hour of induction heart rate and $ETCO_2$ started rising from 35mmhg to 50mmhg with no further increase. The breathing circuit was changed to open and patient hyperventilated, also halothane and nitrous oxide was stopped. Anaesthesia depth was increased with midazolam and propofol 10 mg in incremental doses every 20 min. Temperature at this time was normal and chest was clear. Surgeon was asked to complete the surgery at relatively faster pace. Immediately after extubation child developed high grade fever (106^{o} F) and heart rate (HR) increased from 100/min to 232/min with no further increase in it. Blood pressure was 122/81mmhg, respiratory rate was 22/min initially and then increasing to 28/min, child was conscious but irritable.

After high clinical suspicion, high probability of malignant Hyperthermia was suspected and all

symptomatic measures were started immediately. Dantrolene sodium 1mg/kg bodyweight intravenously was given over ten minutes of the event and was repeated every 6 hour up to 24 hr. All vital monitoring was done every 10 min for first 12 hours and then every 30 min for next 12 hours. Gradually temperature came down to 99 F in 30 min, HR settled to 132min, BP was 110/72 mmhg.

Blood sample for Liver Function Test (LFT), serum Calcium, serum potassium, blood urea, serum creatinine, arterial blood gas analysis (ABG), Serum CPK and serum myoglobin level were sent to laboratory on urgent basis. Halothane caffeine test could not be sent. Patient was kept under observation for 24 hrs in Intensive care unit. There were no episodes of fever, tachycardia or any muscle rigidity and child was stable for 24 hrs. Urine output and all investigation were normal. No active intervention was needed thereafter. Patient was shifted to ward on second day and was discharged after five days. Follow-up of the patient was done in pre-anaesthetic clinic for next 6 months.

III. Discussion

Incidence of MH is 1:15000 in children and 1:150,000 in adult patients receiving general anesthesia. MH is a hyper metabolic state due to active of skeletal muscle sustained contraction in patients with inherited susceptibility and exposure to triggering agent. There is massive release of intracellular Ca++ and consumption of ATP reserve. In this case differential diagnosis of MH with Neurolept Malignant Syndrome (MNS), pheochromocytoma and thyrotoxicosis were ruled out on clinical basis. With Temperature increase >101with 10 points, inappropriately rapid increase in temperature (in anaesthesiologist's judgment) with 15 points and also inappropriate sinus tachycardia (HR =252/min) with 3 points cumulating RAW Score of 28 points was achieved on clinical assessment keeping patient in MH rank 4, indicating *somewhat greater possibility* of MH [5]. Hereby, prompt detection and timely management can reduce mortality associated with such a life threatening condition. Hereby to conclude, management of MH is incomplete without DS, but the availability seems to be a far sighted goal in tertiary center. We were lucky enough to have DS in hand to save a precious life. Even if the incidence of MH being 1:50000, makes us to think the cost effectiveness of DS and its availability on regular basis in hospitals. According to Malignant Hyperthermia Association of United States (MHAUS), stocking of 36 vials of DS costs approximately $3000 per 30 months, a tiny fraction of most facility budgets and a very small price to pay for patient safety. Although fulminate MH episodes are less common, they do happen and patients still die from MH. Dantrolene Sodium could be used similar as a defibrillator, when it comes to own a life saving therapy, drug or unit, it is kept ready for use at all times, even though the need is rare. Its use in MH-like syndromes, it can be a boon to life with its cost prorated among all patients [6].

To conclude, it's high time to emphasize strongly on diagnosis of MH and its association with different congenital facial anomalies and most importantly prompt availability of Dantrolene sodium for treatment at tertiary center.

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