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Anaesthetic Management in a Congenital Catharact Patient with Down Syndrome

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ABSTRACT
We have reported a case of down's syndrome with congenital catharact on both eyes in 5 month old male. He was admitted to Comprehensive Ophthalmology Sumatera Eye Center reporting a progressive decrease in vision and worsening glare in both eyes. Examination of oral cavity revealed a high arched palate with grade II Mallampati. No abnormal findings in X-ray examination. Chest x-ray showed bronchopneumonia with pleural thickening in lateral basal left hemithorax. Echocardiogram revealed minimal atrium septal defect and moderate tricuspid regurgitation with an ejection fraction of 81%. Other examinations were within normal limits.
Keyword: *Down's syndrome, congenital catharact, mallampati, bronchopneumonia, atrium septal defect.*

INTRODUCTION

Down's syndrome is also referred to as Trisomy 21 due to the presence ofan extra copy of chromosome number 21and the commonest of congenital anomalies occurring 1 in 800 live births[1,2]. The children are disabled by mental retardation (100% have IQ's below 65), seizure disorder (2-9%) . In addition to the stigmata of the syndrome, other congenital defects are frequently found in these patients. Cardiac lesions are particularly prominent [3]. Congenital heart disease such as endocardial cushion defects, VSD, ASD, isolated secondum atrial defect, PDA, Tetralogy of Fallot.Children with Down syndrome are more prone to sleep apnea and airway obstruction due to a large tongue, short neck and laryngomalacia [4].There is also increased incidence of atlanto axial instability and risk of spinal cord injury. These children are susceptible to infection and they are also considered to be hypersensitive to the effect or atropine. These all factors modify the anesthetic implication and also anesthetic management in these cases[2].

OBJECTIVE

- To report anaesthetic management incongenital catharact patient withDown syndrome.

CASE

An 5-month-old male, 7 kg with a history of Down syndrome presented to the comprehensive ophthalmology Sumatera Eye Center reporting a progressive decrease in vision and worsening

glare in both eyes during the last few months. He has been examined by ophthalmologists before and was told that his lenses were clouded in both of his eyes.

Family History

Ocular examination: externally normal, visual acuity is difficult to assess,pupils were isochoric with no defect, ocular motility full OU, no abnormality in anterior segment, corneal lens is foggy in either eye, funduscopic exam: hard to assess. Examination of the oral cavity revealed a high arched palate, grade II Mallampati indicating possible intubation problem. Patient lengthis 55 cm. X ray neck was ordered with special emphasis on the atlanto axial joint to rule out atlantoaxial ligament laxity which is commonly seen in patients with downs syndrome. X-ray revealed no abnormal findings. Chest x-ray showed bronchopneumonia with pleural thickening in lateral basal left hemithorax. Echocardiogram revealed minimal atrium septal defect and moderate tricuspid regurgitation with an ejection fraction of 81%. Other examinations were within normal limits. Anaesthetic management: after taking patient on operation table, proper positioning and limb support was given to reduce joint instability. Hemodynamic baseline is within 140-160 bpm for pulse and SpO_2 (99%) were noted. Monitoring of ECG, NIBP, and pulse oximetry was then started. Anesthesia is given starting with preoxygenation for 5 minutes. premedication Premedication given include injection of0,25 mg Dexamethasone IV, 0,1 mg SA IV, 30 mcg Fentanyl IV,0.7 mg Midazolam IV, and 10 mg Lidocain IV. Patient was induced with 15 mg Propofol IV to induce deep sleep. Muscle relaxants were not given since difficult intubation was anticipated. Visualization of larynx by Laryngoscopy was difficult due to high arched palate. External pressure was given on larynx by an assistant, different blade sizes (long sizes) were prepared. After epiglottis could be visualized,10 mg Atracurium IV was injected, No.3 cuffed endotracheal tube was passed through right mouth tip and the tip was manipulated below the epiglottis to pass in the larynx smoothly to prevent the increase of intraocular pressure. Anaesthesia was maintained by Oxygen: N2O (50%:50%), Sevoflurane (1% - 2%).Regular doses of Fentanyl and Atracurium were given via boluses and hemodynamicwas closely monitored. Intraoperati-vely, patient hemodynamic remain stable with minimal blood loss was 5-10 ml.Surgery lasted for 1.5 hours and in the end patient was extubated in operating room. Coughing or gagging is minimized by extubating the patient at a moderately deep level of anesthesia. As the end of the surgical procedure approaches, muscle relaxant reversal is used and spontaneous respiration is allowed to return. Extubation proceeds thereafter during spontaneous respiration with 100% oxygen. Patient transferred to recovery room and hemodynamic was closely monitored. Postoperative pain management is contained with Novalgin 40 mg IV and infection control is done by cefotaxime 150 mg IV.

DISCUSSION

Children with Down's syndromes have usually polycythemic (>70% hematocrit) and phlebotomy may be required if hematocrit>80%, otherwise there is chances of circulatory failure[5]. There is a well-recognised association between DS and endocrine disease, particularly thyroid. Congenital hypothyroidism occurs 30 times more frequently[6]. Such children have antithyroid antibodies and usually there are chances of hypothyroidism in adulthood which may precipitate the hypothermia and ultimately may be causative factor of delay recovery from anesthesia. Down's syndrome is associated with anomalies in various systems. Cardiovascular, respiratory and central nervous system may be involved in Down's syndrome as an isolated system involvement or multiple system involvement, thus posing several anaesthetic challenges. The most frequent cardiac malforma-tions are interauricular septal defect (IASD), atrioventricular septal defect (AVSD) interventr-icular septal defect (IVSD) and patent ductus

arteriousus (PDA) (90%)[7]. Clinical features of Down's syndrome include microcephaly, macroglossia, andligamentous laxity at atlanto occipital joint and subglottic stenosis, which can pose problems for securing airway[8].

Patient presenting with sign of difficult airway such as microcephaly and macroglossia, then the patient must be prepared for multiple attempt of intubation, so it is necessary to premedicate the patient with dexamethasone. Anesthesia induction is preferred with awake or sleep non apnea technique in terms to preserved spontaneous ventilation. The risk is greater in term of hipersensitivity in airway which will results in laryngeal oedema.

Patient also suffers bronchopneumonia, which is an acute symptom of pulmonary infection. Patient presenting with productive cough and sneezing, hardness to breathe, and nostril work of breathing will increase perioperative risk. Those conditions will also affect difficulty in airway management of this patient. Patient is also presenting with ASD as cardiac problem. Thus, induction agent and maintenance must be done to preserve maximum cardiac function and prevent systemic failure. So, in this patient, we need to ensure adequate ventilation and proper induction to achieve tracheal intubation by using selective drugs which will less impair hemodynamic and respiratory function.

CONCLUSION

A detailed preoperative assessment and optimization was done. There was no atlanto-axial instability but there is. However, child was evaluated thoroughly and anaesthetic management was planned with anticipation for difficult airway, sensitivity to drugs and ventilation challenges during surgery

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