Research Paper

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A Child with Osteum Secundum Atrial Septal Defect and Down Syndrome Undergoing Adenotonsillectomy –An Anaesthetic Challenge

Medical Science

KEYWORDS : congenital heart disease, Down syndrome, left to right shunts, non-cardiac surgery.

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ABSTRACT

The presence of congenital heart disease (CHD) in a child undergoing non-cardiac surgery poses a great challenge for the anaesthesiologists. The challenges include the patient's age, complexity of the cardiac lesion present, coupled with patient's capacity to compensate for the physiological changes that occur under anaesthesia, urgency of surgery and the presence of other coexisting diseases(1).CHD occurs in about 40% of children with Down syndrome. Anaesthesia in Down syndrome carries a higher risk due to the prevalence of mental retardation, hypothyroidism, cardio-respiratory manifestations, airway abnormalities and postoperative pulmonary complications(2). Inability to secure an airway, congestive heart failure, arrhythmias, hypoxia,hypercarbia and paradoxical embolism are the factors that place childrenat increased risk during anaesthesia for non-cardiac surgery(3, 4). Adequatepost operative analgesia, prevention of hypoxia, hypercarbia, dehydration and hypothermia is vital (3).

Introduction :

Congenital heart disease(CHD) is one of the inborn defects occurring in approximately 0.8% of newborn infants.CHD, placed first among common birth defects, occurs approximately 1 in 125 live births[1]. The frequency of atrial septal defect is 9% among all acyanotic conditions[1]. Osteum secundum or fossa ovalis defect results from a deficiency in the region of the fossa ovalis. These defects may be associated with mitral valve prolapse or mitral regurgitation [4]. Children with CHD presenting for non-cardiac surgery can be grouped into three categories: a) Non-operated patient, b) Patients with previous corrective surgery and c) Patients with previous palliative surgery [3]. Apart from categorising, to fully optimise the patient, it is essential for the anaesthesiologist to obtain information about the cardiac lesion, altered physiology, implications under anaesthesia and also postoperative considerations[2].

Shunting through these defects depend upon the diameter of defect and factors responsible for the balance between systemic vascular resistance(SVR) and pulmonary vascular resistance(PVR) which is vital in the anaesthetic management of these patients [2]. L-R shunts are characterized by excessive pulmonary blood flow, shunting is increased if PVR is reduced for instance, with high fraction of inspired oxygen (FiO_2) and low partial pressure of carbon dioxide ($PaCo_2$). Patients are usually acyanotic but deterioration in gas exchange may result from pulmonary congestion [3, 4]. Trisomy 21, also known as Down syndrome is a genetic disorder that occurs in approximately 1 in every 800 live births [2].This condition may allow C1-C2 subluxation and subsequent spinal cord injury during airway manipulation. Factors affecting the upper airways include hypotonia, obesity, midface hypoplasia, relative glossoptosis, increased secretions, excessively large tonsils and adenoids [5]. Another concern to the anaesthetic management of these patients is tracheal stenosis associated with difficult intubation and possibility exists that all children with Down syndrome have a small subglottic area when compared to normal children. Obstructive airway disease has been recognized as a significant problem for children with Down syndrome [2].

CaseHistory :

A 10 year old female patient presented with history of recurrent attacks of nasal obstruction and inability to breathe through nose during sleep, snoring at night and difficulty in swallowing. There was no history of breathlessness, febrile seizures and cyanotic spells. She was diagnosed with Down syndrome at the age of 7years when she was referred to the hospital for similar complaints like nasal obstruction. On evaluation she was also diagnosed to have acyanotic congenital heart disease i.e. atrial septal defect with 7mm defect. History of delayed milestones were present i.e. neck holding at the age of 10 months, sitting with support-2years. On examination child was hyperactive, well-oriented, slurred speech, playful, weighed 23kgs, with typical features of Down syndrome-depressed nasal bridge, low set ears, high arched palate and macroglossia as shown in the picture1.There was no cyanosis or clubbing. On auscultation pan systolic murmur was heard best at the tricuspid area. The lung fields were clear. Oxygen saturation was 100% in room air, pulse rate 110beats/min, respiratory rate was 20cycles/min. Airway was graded as Mallampati class II with adequate mouth opening, tonsillar enlargement were noted, bilaterally extending to the midline i.e. kissing tonsils as shown in the picture 2. Normal neck movements were present.

Investigations: Blood group : 'A' positive, Hb : 13.9g/dl, INR: 1.46,platelet count : 3.8lakhs/cu mm, normal chest x-ray & echocardiogram revealed acyanotic congenital heart disease, left to right shunt, 5mm osteum secundum atrial septal defect with mild tricuspid regurgitation, ejection fraction: 74%, no pulmonary arterial hypertension and normal biventricular function. A preoperative diagnosis of chronic adenotonsillitis with atrial septal defect and Down syndrome was done and the child was posted for adeno-tonsillectomy under general anaesthesia. Case was accepted under ASA Grade III with moderate cardiac risk.

In the preoperative area the child was calm and co-operative in the presence of parents, fasting time was confirmed to be more than 6hours and intravenous access was obtained with 22G IV cannula in right upper limb. Antibiotics were administered as

per hospital protocol. Baby was shifted to operative room - pulse oximetry, ECG, non-invasive blood pressure monitors were connected. Child was premedicated with Inj. Ondansetron 0.1 mg/kg IV, Inj Midazolam 0.5mg IV, Inj Glycopyrrolate 0.01mg/kg IV, Inj pentazocine 0.5 mg/kg IV. Preoxygenated for 3min with 100% oxygen, child was induced with Inj Propofol 2mg/kg IV, adequacy of mask ventilation was confirmed by good chest rise and a normal capnogram. When the child was in deeper planes of anaesthesia with sevoflurane 2-3%, laryngoscopy was done with utmost care to visualize the vocal cords, and grading was done as Cormack Lehane grade II. Intubated with 5.0mm ID cuffed portex oral endotracheal tube in the first attempt, connected to Bain circuit with 1litre bag. Bilateral air entry was confirmed and tube was secured in the midline. Maintained on N_2O:O_2-2: 2, sevoflurane-0.8-1.5%.Non depolarizing muscle relaxant used was Inj Atracurium 0.5mg/kg for loading dose and 0.1 mg/kg for maintenance. Paracetamol suppositories 30mg/kg body weight was inserted. Intra op: PR: 110-130/min BP: 100-110/60-70mmhg, SpO_2:100% ECG: within normal limits (WNL), $EtCo_2$ = WNL. IV fluids were given according to Holiday Segar regime. At the end of the surgery, the child was fully awake, calm, with adequate muscle power and intact reflexes for suctioning, gentle laryngoscopy was done and haemostasis confirmed ,reversal agent Inj Neostigmine 0.05mg/kg IV, Inj Glycopyrrolate 0.01mg/kg was given IV, child was extubated and careful monitoring of respiratory and cardiac parameters were done. Child was shifted to post operative ward for further care and monitoring of pulse rate, BP, SpO_2 was done for 6hours. There was no bleeding from the tonsillar fossae. Entire post operative period was uneventful and adequate hydration was maintained. Post operative analgesia was given in the form of paracetamol suppositories 30mg/kg Q6th hourly.

Discussion:

Down syndrome is associated with multi-system problems that need thorough assessment pre-operatively. Assessment of airway, cervical spine for signs and symptoms of atlanto-axial instability, sub-glottic stenosis ,screening for associated congenital heart diseases and respiratory problems are important[2]. Sedative premedicants should be administered with caution keeping in mind the dangers of respiratory depression, airway obstruction leading on to hypoxia and hypercarbia. IV cannulation is difficult in these patients and care should be taken to prevent air embolism which increases the risk of paradoxical embolism. About 12% of patients with Down syndrome have atlantoaxial instability, cervical spine manipulations should be undertaken with the greatest care when positioning these patients for surgery because neck extension may cause spinal cord compression [5, 10].

The factors which can lead to difficulty in securing airway in Down syndrome includes obesity, small chin, inadequate mouth opening, macroglossia, short neck, laxity of airway muscles and presence of subglottic stenosis .Difficult intubation cart which includes emergency airway modalities, fiberoptic equipment, laryngeal mask airway, flexible bougie, emergency surgical airway should be kept ready[2]. Intubation should be done in a deeper plane of anaesthesia with or without muscle relaxant [2, 8, 10]. Cuffed endotracheal tube is preferred for tracheal intubation to reduce the chance of aspiration of blood, secretions and to reduce gas leaks around the tube [2, 8] .

Perioperative complications include bradycardia, bronchospasm, aspiration and rarely, neurological problems due to atlanto-axial subluxation [2, 8].All efforts should be made for early detection of hypoxia in perioperative period by intense monitoring using apnea alarm, capnograph, pulse oximeter, precordial stethoscope, and electrocardiogram. Monitoring in the post operative period is essential to prevent complications like post-extubation stridor, bronchospasm, hypoxia, bradycardia and seizures [11].

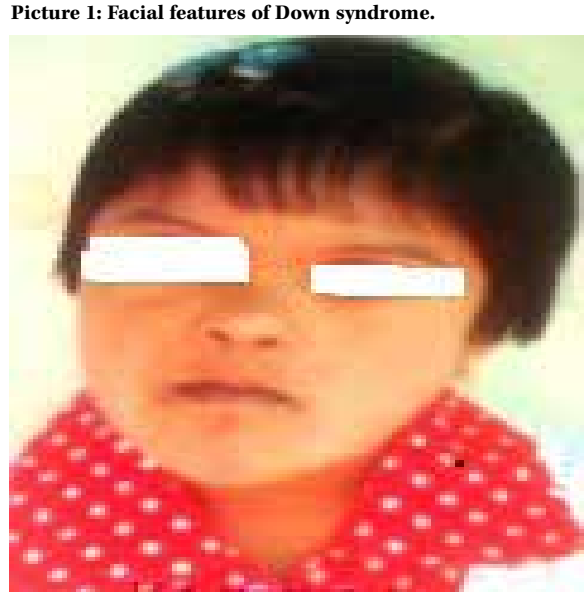
A left-to-right shunt seen in an atrial septal defect does not cause cyanosis but the high volume pumped to the right side may result in pulmonary hypertension and, if this builds up and exceeds systemic pressure, the shunt may reverse from right to left[1,3,6,7,9]. In unrestricted left to right shunts there is excess pulmonary blood flow which can result in congestive heart failure (CHF) and pulmonary hypertension. Hypothermia, stress, pain, acidosis, hypercarbia, hypoxia and elevated intrathoracic pressure are the factors under anaesthesia which can worsen pulmonary hypertension. Entrapment of air in the IV tubing's is strictly avoided.

The anaesthetic goals in managing such a patient is to prevent increase in PVR, decrease in SVR and depression of myocardial function.1-1.5 minimum alveolar concentration of isoflurane, halothane and sevoflurane has no effect in patients with isolated ASD during mechanical ventilation [3] and the use of isoflurane and sevoflurane for maintenance of anaesthesia is widely reported. Both agents have minimal effect on myocardial contractility or shunt fraction [6]. 100% oxygen and hyperventilation in patients with L-R shunt will result in exaggerated pulmonary vasodilatation, which in turn will increase pulmonary congestion, and thus should be avoided[1,3]. The effects of shunting on the speed of inhalation inductions are more pronounced for N_2O than for the more soluble volatile anaesthetic agents. The choice of specific neuromuscular blocking agents in children with congenital heart disease is usually made on the basis of the drug's predicted cardiovascular effects as well as their duration of action [4,9]. Maintenance of euvolaemia, pain management and monitoring of vitals to prevent any complications are critical components of successfully managing a child with acyanotic congenital heart disease in the postoperative period.

Considering the challenging nature of such cases it is prudent that the anesthesiologist should be well aware of the physiological changes and the complications that may arise at any time of anaesthesia or surgery. The timely intervention in managing these complications is of utmost importance in reducing the morbidity and mortality

Acknowledgement: ---

Picture 1: Facial features of Down syndrome.



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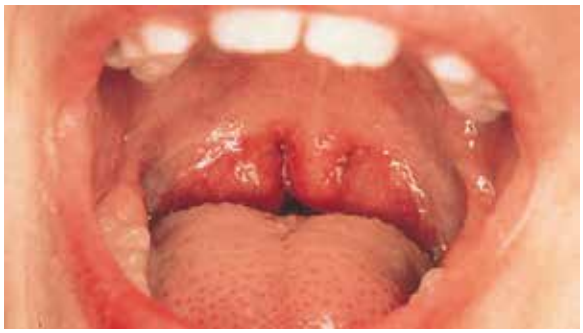
Picture 2: Mouth opening, enlarged tonsils extending to midline i.e; kissing tonsils



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