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From the Editor's Desk

Dear members,

We are happy to bring out the 3rd edition of OFISA NEWSLETTER, and we thank all our members for their continued support and contributions towards the issue.

In this issue, we deal with preterm, cardiac patients with pacemakers, topical anaesthesia for cataract and management of patient posted for eye surgery with ectodermal dysplasia.

Dr. Renu Sinha and others from AIIMS, present the anaesthetic concerns of preterm babies for ophthalmic procedures with an interesting video clipping demonstrating the abnormal respiratory pattern in such babies.

Dr. Multan Mal Sharma of Aditya Jyot Eye Hospital , Mumbai presents the ophthalmic management in patients presenting with permanent pacemakers and with Automated Implantable Cardioverter Defibrillators(AICD).

Dr. Shubhangi Shah and others from Raghudeep Eye Clinic, Ahmedabad present topical anaesthetic regime for cataract surgery at their Clinic.

Dr Jaichandran V V, Consultant Anaesthesiologist at Sankara Nethralaya, Chennai presents Anaesthetic and ophthalmic implications in a case of Ectodermal dysplasia posted for eye surgery.

Wishing you all a happy and interesting reading

With regards

Dr. Kannan R

Editor, OFISA Newsletter.



Anaesthetic concern in preterm infants for ophthalmic procedures

Dr. Renu Sinha

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In recent years, number of preterm infants for ophthalmic examination and procedures has increased due to improved survival rates and better screening methodologies. Most common procedures in preterm infants are congenital cataract, congenital glaucoma and retinopathy of prematurity (ROP). Multiple anaesthesia exposures are required for screening, diagnosis, surgery and further progress of the disease as treatment is primarily surgical.

Topical ophthalmic drugs:

Pupillary dilatation is essential for ocular examination and surgery. Paediatric concentrations of ophthalmic medications are not readily available, using the present drops may lead to overdosing and various undesirable systemic effects.

Apart from using micro droppers, wiping off any excess drug, application of digital pressure over the lacrimal duct, increasing the viscosity of the ocular drug can reduce the systemic effects. Pupillary dilatation should be evaluated in preoperative room and topical tropicamide can be instilled and if still needed one microdrop of 1.0 % phenylephrine with digital pressure over the lacrimal duct may be administered.

Application of digital pressure over the lacrimal duct for 5 minutes after instillation or keeping the eye gently closed for 5 minutes may reduce systemic absorption by 67%. Alternative drugs to augment pupillary dilatation like proparacaine, diclofenac and flurbiprofen can be used in infants with difficult mydriasis.

Drugs used for treatment of glaucoma are mainly acetazolamide, pilocarpine and betaxolol. Acetazolamide can cause metabolic acidosis, hypokalemia and dehydration; pilocarpine can cause bronchospasm,

bradycardia; betaxolol can lead to bradycardia and even sinus arrest.

Concerns in the preterm infants:

Preterm infants may have associated problems like low birth weight, cardiac defect, bronchopulmonary dysplasia, recurrent apnea, episodic bradydysrhythmias, anaemia, intraventricular haemorrhage and necrotizing enterocolitis. Bronchopulmonary dysplasia is associated with increased airway resistance and reactivity, diminished lung compliance and hypoxemia. Preterm babies are more prone for bradycardia, difficult intubation, hypothermia, hypoglycemia, hypocalcemia, air embolism and postoperative apnea.

Incidence of postoperative apnea is as high as 26% in infants less than 44 weeks post conceptual age (PCA) where as incidence of apnea reduces to 3% in infants more than 44 weeks PCA. Postoperative apnea is strongly correlated with gestational age and PCA, with previous history of apnea, anemia (hematocrit<30).

Preterm infants less than 44 to 46 weeks PCA should be admitted and monitored for postoperative apnea following general anaesthesia for atleast 18-24 hours. Preterm infants upto 55 weeks should be monitored continuously for atleast 12 hours after general anaesthesia.

Pre Anaesthetic evaluation:

History of NICU stay, prematurity, birth asphyxia, oxygen requirement, mechanical ventilation, apnea, bradyrhythmias, cyanosis, convulsion and drug history should be obtain. Airway abnormalities should be ruled out as many syndromes has associated ophthalmic anomalies. They may have cleft lip and palate, high arch palate, micrognathia, macroglossia, short neck, craniosynostosis etc. as a part of syndrome.



Anaesthetic concern in preterm infants for ophthalmic procedures



Appropriate investigation should be done to rule out metabolic disorders (e.g. serum calcium levels), and if present these should be corrected prior

[Click here to open video](#) to any procedure. Apart from routine haemogram, chest X ray and echocardiography should be done in premature babies to rule out congenital heart disease or if there is history of cyanosis, feeding difficulty, repeated chest infections, failure to thrive and delayed mile stones.

Counseling of the parents:

Apart from visual prognosis, parents must be counselled about the risk associated with anaesthesia and surgery in the premature infants as they may have other congenital anomalies including cardiac defects or hypoxia induced brain damage. The need for postoperative nursery care and ventilatory support must be emphasized to the parents. They should be explained about need of repeated examination.

Perioperative management:

The objectives of anaesthesia include safety, akinesia, satisfactory analgesia, minimal bleeding, avoidance or obtundation of the oculocardiac reflex and prevention of intraocular hypertension.

Anaesthesiologist should be aware of the problems of preterm infants and also about potential interactions between ophthalmic drugs and anaesthetic agents. Phenylephrine is an alpha agonist mydriatic and can cause transient hypertension, pulmonary oedema and even cardiac arrest. There are case reports of renal failure with tropicamide 0.5% and phenylephrine 0.5%, transient paralytic ileus with cyclopentolate 0.2% and phenylephrine 1%, bradycardia with tropicamide 1% and

heart failure with phenylephrine 1% and cyclopentolate 1%.

Laser for ROP can be done in NICU to avoid problems associated with shifting the infant to the operating room (OR). A wide range of techniques have been described for laser surgery such as topical anesthesia alone, topical anesthesia with ketamine or morphine or remifentanyl infusion, with or without endotracheal intubation leading to varying reports of apnea, bradycardia and prolonged need of mechanical ventilation. However for surgical procedures (trabeculectomy, lense aspiration, vitreo retinal surgery) infant is shifted to OR. Glaucoma surgery and vitreo retinal surgery are moderately painful.

General anaesthesia for ophthalmic procedures in children is similar to that provided for other surgical procedures. Clear communication with the ophthalmologist can delineate which diagnostic or therapeutic procedure is to be performed. Consideration should be given to the proposed duration of the procedure as the access to the airway may be difficult. A wide assortment of anaesthetic agents can be used safely and effectively in ophthalmic procedures. There is little evidence to support any one single method of sedation, analgesia or anaesthesia. Regardless of which agent is selected, it should have short duration of action and should be administered in titrated doses.

Ocular akinesia is required for accurate placement of laser burns in ROP surgery and to prevent the risk of light threatening foveal burns, thus necessitating general anaesthesia with neuromuscular blockade. Neuromuscular paralysis is essential not only for complete akinesia of the eyeball but also to provide controlled ventilation which ensures normocarbida and normal IOP.



Anaesthetic concern in preterm infants for ophthalmic procedures

Endotracheal intubation with controlled ventilation provided continuous end tidal CO₂ monitoring to avoid hypercarbia and its ocular hypertensive effect as well as to detect inadvertent disconnection of the endotracheal tube from the anaesthesia circuit, a dangerous event that can be obscured by the surgical drapes. Non depolarising muscle relaxants are used. Atracurium or cisatracurium is a better choice in preterm and low birth weight babies as enzyme systems to metabolize other non-depolarizing drugs are not well developed.

Simple, short, screening or diagnostic procedures can be performed with inhalation general anaesthesia using a face mask or using supraglottic devices like the laryngeal mask airway (LMA). For short procedures, placement of LMA provides a suitable airway, even for patients with history of mild to moderate bronchopulmonary dysplasia, and causes less cardiovascular stimulation than laryngoscopy and endotracheal intubation. It does not impede the ophthalmologist's access to the eyes, and is associated with reduced coughing.

Continuous electrocardiogram, non-invasive blood pressure, pulse-oximetry, capnography and temperature should be monitored. Preterm babies are prone to hypothermia, therefore due precautions must be taken during the transport to and from the operation room.

Intraoperatively, oxygen saturation should be maintained between 87% and 92%. In infants with chronic lung disease, higher FiO₂ during surgery and anaesthesia reduces the likelihood of severe hypoxemia, lowers pulmonary arterial pressure, and decreases airway resistance. As fluctuation between hypoxia and hyperoxia produces greater neovascularization, oxygen saturation should be maintained within a restricted, tight range. The anaesthesiologist may consider keeping peri-operative oxygen saturations within the NICU prescribed limits.

Since concentration, duration, timing, and fluctuations of oxygen levels may all have a role in ROP, the optimal intraoperative oxygen saturation for these patients has yet to be clearly elucidated.

Intraoperative analgesia can be maintained with short acting opioids (fentanyl) or remifentanyl. Remifentanyl has the advantage of rapid recovery so preferred in preterm infants. However use of opioid may result in respiratory depression and apnoea, they should be used with proper intra and postoperative monitoring. Acetaminophen can be administered intravenously or rectally. Topical anaesthetic and regional block can reduce the total dose of opioid, thus decreasing the associated side effects.

During surgery normothermia and normoglycemia should be maintained. Blood sugar should be monitored at regular interval and dextrose containing fluid should be transfused accordingly. Warmed air and warmed fluid should be administered apart from maintaining higher ambient OT temperature. Use of warming mattress, blanket and radiant heater, especially in the pre-operative room and during intubation and extubation, is recommended, as shown in figure 1. Preterm infants are also prone to hypocalcaemia. If needed, calcium levels should be measured.



Fig - 1 Baby wrapped and with over head radiant warmer

Anaesthetic concern in preterm infants for ophthalmic procedures

Except for preterm and low birth weight babies, any residual neuromuscular blockade can be reversed after completion of surgery. If the child is breathing adequately and no bradypnoea or apnoea is present, extubation can be attempted in OR.

Postoperative complications

Postoperative breath holding and apnea are potentially serious complications. There may be associated episodic bradycardia. Risk factors for postoperative apnea include PCA, gestational age, history of apnea, chronic lung disease (CLD), anemia, use of opioid and/or muscle relaxants. Apnea at emergence from anaesthesia and periodic breathing in the recovery room confer moderate additional risk for delayed breath holding. Preterm infants should be observed after surgery with pulse-oximetry and apnea monitoring in NICU.

To conclude, a timely and detailed history and physical examination, complimented with indicated diagnostic tests, generally ensure a safe anaesthetic course. Preterm or ex-premature infants undergoing any ophthalmic procedure require proper institutional support, and need postoperative transportation from the ophthalmology specialty centre to a paediatric intensive care facility for further monitoring. Skillful anaesthetic management is an absolute requirement for optimal outcomes after ophthalmic surgery. Because the complications of ophthalmic anaesthesia can be vision threatening or life threatening, it is imperative that the ophthalmologist and the anaesthesiologist understand the complex and dynamic interaction among a multitude of factors, including the disease, anaesthetic agents, ophthalmic drugs, and surgical manipulation.



Ophthalmic Anesthesia Management in patients with Permanent Pace-maker & Automated Implantable Cardioverter Defibrillator(AICD)

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The challenges for Ophthalmic Anesthesiologists are increasing as many patients come with complex cardiac diseases for ophthalmic surgery, Anesthesiologist will have to give attention to initial pathology that led to pacemaker insertion as well as to co-existing diseases & its treatment, as well as specific information regarding kind of pacemaker/AICD used.

Primarily there are two types of permanent pacemakers commonly used.

1. Standard pacemaker that triggers the chambers of heart, see Figure 1
2. Automated Implantable cardiac internal defibrillator or AICD, see Figure 2 and pacemaker combination, this works as standard pacemaker as well delivers a shock to stop "lethal cardiac rhythm".

Worldwide nearly 4 million patients have pacemakers and 4 lac have AICDs Standard pacemaker Pacemaker with AICD



Figure :1 Standard pacemaker
Courtesy: dancewithshadows.com

See Figure :2 Pacemaker with AICD

In 1995 Drs. Badrinath/ Bhaskaran/ Sundararaj retrospectively reviewed ophthalmic surgery cases and reported that presence of pacemakers significantly increased the probability of mortal events within 6 weeks of post op. regardless of type of anesthesia

AICD can be distinguished from conventional pacemaker by presence of shock-coils on Right ventricular.

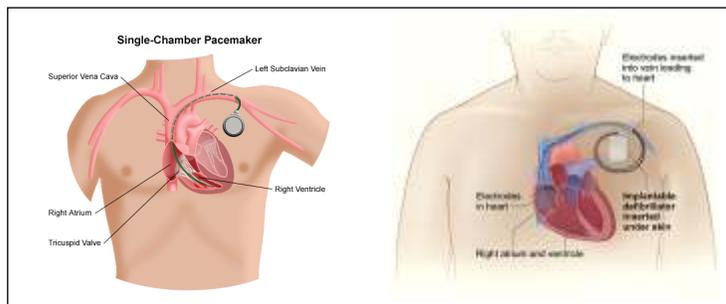


Figure3: Courtesy: www.lpch.org/media/images/conditions/ei



Figure 4: Courtesy: sciencephoto.com

American College of Cardiology(ACC) guidelines and an American Society of Anesthesiologists(ASA) advisory suggested that bipolar electrocautery reduced the risk of malfunction of implanted devices, compared with monopolar electrocautery. These documents do not specifically address modifying perioperative management including temporarily inactivating an AICD, if only bipolar electrocautery is used.

An informal survey by (American) Ophthalmic Anesthesia Society (OAS) revealed that 96% of respondents did not use a magnet to reprogram or convert the pacemaker to asynchronous mode. 86% did not use a magnet to reprogram or inactivate an AICD before exposure to bipolar electrocautery. No adverse effects in any of these pts reported.



Ophthalmic Anesthesia Management in patients with Permanent Pace-maker & Automated Implantable Cardioverter Defibrillator(AICD)

Placement of magnet over generator usually inactivates some pacemakers but in others might not produce change in pacing as not all the pacemakers have continuous asynchronous mode and it will not prevent aberrant AICD behavior.

Pre-anesthetic evaluation and pacemaker reprogramming.

1. Evaluation and optimization of co-existing diseases.
2. No special Lab. Tests needed.
3. Chest film (x-ray) to document the position of lead especially coronary sinus lead and confirming the type of device present.
4. There is no history of any arrhythmia that will predispose to shock (VT, VF, SVT or paroxysmal atrial fibrillation).
5. Direct interrogation with programmer remains the only method for evaluating battery status lead performance & adequacy of reliable current settings.

Intra and post operative management of pacemakers.

Monopolar electrocautery use remains the principle intra operative issue, coagulation electrocautery is more likely to cause problem than cutting electro-surgery.

Cautery pad must be placed in such a way that electro-surgery current does not cross the chest.

However if General Anesthesia is planned inactivating the AICD should be strongly considered,

this is due to the additional risks of use of peripheral nerve stimulators or succinylcholine(via microdepolarization of muscle) causing AICD to discharge inappropriately.

ECG monitoring must include the ability to detect the pacemaker discharges('artifact filter' disabled)

Perfused (peripheral)pulse must be monitored in waveform display.

HOcm (hypertrophic obstructive cardiomyopathy), DCM(dilated cardiomyopathy), BiV patients might need cardiac output monitoring.

SpO₂, NIBP / IBP must be measured. Pacemaker rate might need to be increase due to an increased O₂ demand.

Appropriate equipment must be readily available to provide back up pacing or defibrillation.

Avoid use of drugs that suppress AV or SA node, as potent opiates or dexmedetomidine.

Also sevoflurane, isoflurane and desflurane may exacerbate the long Q-T interval.

Post-operative shivering may trigger AICD falsely. Any pacemaker/ AICD that was reprogrammed for operation should be reset appropriately.

Summary

Most ophthalmic surgery patients with AICD may not have their devices inactivated when only bipolar electro surgery is used, however if General Anesthesia is planned than inactivating AICD should be considered.



Ophthalmic Anesthesia Management in patients with Permanent Pace-maker & Automated Implantable Cardioverter Defibrillator(AICD)

Appropriate equipment must be on hand to provide backup pacing and / or defibrillation. Monitor cardiac rhythm / peripheral pulse mechanically with pulse oximeter or arterial waveform.

Avoid use of succinylcholine, potent opiates, high concentration of inhalational anesthetic gases during GA.

To reduce the risk of eye injury from patient movement in the event of an AICD discharging (shock), inappropriately during surgery, we must monitor the ECG carefully if a ventricular arrhythmia occurs in a patient with functioning AICD ,we have about 15 seconds to warn the surgeon to adjust or remove the instruments and prepare for possible movement.

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Topical Anaesthetic Regime for Cataract Surgery at Raghudeep Eye Clinic

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There is a wide range of options for ophthalmic anaesthesia, including general, peribulbar, retrobulbar, subconjunctival, sub-Tenon, topical, and intracameral.² Topical and intracameral anaesthesia are the most popular anaesthetic methods, and they have been shown to be both safe and effective ^{1–3} because they tend to avoid the complications associated with traditional peribulbar and retrobulbar injection methods, such as globe perforation, retrobulbar hemorrhage, optic nerve trauma, brainstem anaesthesia, and extraocular muscle injury.^{4–5}

The advantage of Topical Anaesthesia is well established and understood.

- Faster with greater patient comfort
- No patient anxiety over receiving a deep injection next to the eyeball
- Immediate visual recovery
- Eliminates need for an eye patch

Our Regime at Raghudeep Eye Clinic:

Topical anesthesia for Cataract surgery is the anesthesia of choice at Raghudeep Eye Clinic, for over 10 years. This has been possible because of improvised surgical technique and technology.

Pre-operative counseling: Every patient is assessed by an anaesthesiologist few days prior to surgery. During this preoperative evaluation, all adult cataract patients are assessed for suitability of topical anaesthesia, their medical condition, and are counseled for the steps they will undergo during surgery.

Preparation on Day of Surgery:

Patients are informed about the technique of Topical anesthesia and an informed consent is obtained. An Intravenous access is established. Aseptic preparation is carried out over the operative eye.

Topical anaesthesia is given with 2 % Lignocaine Gel instilled in conjunctival sac 10 minutes before surgery. Patients are instructed about their fixating on microscope light and keeping both eyes open during the entire surgery. Before beginning of surgery patients receive Inj. Midazolam 0.01 mg/Kg. and Inj. Fentanyl 0.5 to 0.7 mcg/Kg. slowly intravenously. NIBP, SaO₂ and ECG are standard monitors used for all patients. Intracameral Injection of 0.2 ml of 1 % preservative free Lignocaine Average duration of surgery is 25 to 40 minutes. During surgery patients receive a combination of 0.5 mg. Inj Midazolam and 10 to 20 µgm of Inj. Fentanyl diluted in normal saline intravenously by infusion pump per hour. The infusion rate is adjusted according to requirement. During surgery 2 % preservative free Lignocaine is made available for supplementation if the duration of surgery extends more than 30 minutes. Post operatively, oral paracetamol is the choice of analgesic for a day.

We have done a prospective, observational study with our regime. The objective of this study was to assess the efficacy of our regime in controlling Anxiety, Pain management during surgery, Surgeon's and Patients' comfort and sedation level at the end of surgery.



Topical Anaesthetic Regime for Cataract Surgery at Raghudeep Eye Clinic

Method: 627 consecutive adult cataract patients from October 2010 to January 2011, were recruited for study. The patients to be operated under general anaesthesia and complicated eye surgeries like subluxated cataract and scleral fixation of lens and combined cataract and glaucoma surgeries were excluded from the study. The heart rate, systolic, diastolic and mean blood pressure was documented on pre-operative day, in the pre-operative holding area on operative day and during surgery for every 10 minutes. The type of cataract was noted and the difficulty in surgery was graded in the form Surgical Challenge Grade 1-4, according the challenge assessed in pre-operative evaluation of Eye. Following difficulties were observed in the pre-operative evaluation.

1. Nuclear sclerosis Gr 4,
2. Anterior Chamber Depth < 2.2 mm.
3. Posterior Polar Cataract
4. Small Pupil < 5 mm.
5. White Mature cataract
6. Uveitic Eye with synechia.
7. High Myopia AL > 27mm
8. Corneal disorders limiting Visibility

Table 1	
Number of Difficulty	Challenge Grade
No Difficulty	Gr. 1
1 Difficulty	Gr. 2
2 Difficulties	Gr. 3
More then 2	Gr. 4

Grading was done according to the number of type of challenges observed in the eye from the list above. The challenge was graded as shown in Table 1.

Table 5 Surgical Challenge Gr.No. of Patients(n-627) %Gr. 127143.22Gr. 225941.3Gr. 38012.76Gr. 4172.71 Considering there is no restriction of eye movements during surgery under topical anesthesia, the discomfort to the surgeon during surgery was documented as per the Table 2 below.

This assessment was done separately for the consultants and the trainee surgeons operating independently.

Table 2		
Score	Consultant	Trainees
1	No eye Movements	No eye movements
2	Movements does not affect	Movements does not affect
3	Movements inflected surgery	Movements inflected surgery
4	Not Applicable	Hand over to consultant

At the end of surgery patient's sedation score was documented with the help of Ramsay's Sedation Scale. Sedation score was assessed according to the Ramsay's scale as shown in Table 3

Table 3 Ramsay sedation Scale	
Anxious or Agitated	1
Oriented and cooperative	2
Responding to commands	3
Brisk response to glabellar tap or Loud auditory stimulus	4
Sluggish response to glabellar tap or Loud auditory stimulus	5
No response	6



Topical Anaesthetic Regime for Cataract Surgery at Raghudeep Eye Clinic

Over all surgeons' and patients' comfort was asked and graded according to the Table 4

Table 4	
	Score
Not Comfortable	0
Moderately Comfortable	1
Very Comfortable	2

Post operatively patients were asked about pain during surgery and to measure the pain Verbal Analogue Scale was used.

Observations: 627 consecutive patients as per the inclusion and exclusion criteria were recruited for observation.

Table 5 shows the number of patients with percent in each difficulty grade according to the classification mentioned in Table 1.

Out of these 11 patients received peribulbar anaesthesia because of either dementia, hard of hearing, language barrier and non –communication. These eleven patients were in Cataract Challenge Gr. 3 or 4

Table 5		
Surgical Challenges Gr.	No. of patients (n=627)	%
Gr. 1	271	43.22
Gr. 2	259	41.3
Gr. 3	80	12.76
Gr. 4	17	2.71

Table 6 shows the mean vital parameters data documented at various stages of Peri-operative period. There was a decrease in average heart rate and Blood Pressure after the pre-operative counseling and initiation of sedation.

Table 6	Sys.	Dias.	Mean	HR
Pre-Op	144.48	86.57	106.67	76.75
Holding Area	155.23	84.76	111.23	77.45
On Table	146.56	83.45	105.26	74.55
10 min	132.24	78.43	97.71	68.45
20 min	127.37	76.34	93.45	66.45
30 min	125.67	76.76	94.65	66.6

Table 7 shows the other parameters of observation made during and after the surgery.

Table 7 **Parameters Scores** Eye Movements - Consultant 0.63, Eye Movements – Trainee surgeon 0.72, Surgeon's comfort 2.21, Ramsay Sedation Scale 2.19 and VAS for pain 0.12. Trainee doctors were allotted only Cataract Challenge category Gr.1 patients according to Table 1..

Table 7	
Parameters	Score
Eye Movement s - Consultant	0.63
Eye Movement s – Trainee surgeon	0.72
Surgeon's comfort	2.21
Ramsay sedation Scale	2.19
VAS for pain	0.12



Topical Anaesthetic Regime for Cataract Surgery at Raghudeep Eye Clinic

Discussion:

Cataract surgery by Phacoemulsification technique is minimally invasive to the patient and it is well established that Topical anaesthesia technique is adequate for surgery. However some factors related to eye like complicated cataracts or challenging environment mentioned early in the chapter, make the choice difficult. Some factors related to patients like, dementia, language barrier, hard of hearing also make the application of Topical anaesthesia difficult.

Alleviating fear and anxiety by counseling and intravenous Midazolam are very useful when cataract surgery is performed under local anaesthesia. Inj. Fentanyl 0.7 µgm./Kg. body weight has supplemental effects on analgesia during cataract surgery under topical anesthesia and increases patient comfort and surgeon satisfaction.⁶

We also use Inj. Fentanyl in a calculated dose during surgery. With the combination of these two predictably short acting drugs like Midazolam and Fentanyl, we are able to offer topical anaesthesia to a large number of our cataract patients.

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A case of Ectodermal dysplasia posted for eye surgery: Anaesthetic and ophthalmic implications

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Introduction:

Ectodermal dysplasia is a group of rare inherited disorders that results from the abnormal function of the ectodermal layer in the developing embryo. The ectoderm of the embryo forms the hair, teeth, nails, sweat glands and parts of the eyes.

Here we report a case of seventeen year old boy who developed bilateral juvenile open angle glaucoma posted for Trabeculectomy +Mitomycin in right eye under general anaesthesia.

Case report:

Full term normally delivered male child with history of delayed milestones and a unique history of teeth falling spontaneously came to our hospital for decreased vision in both eyes. Ophthalmic examination was done and he was diagnosed as bilateral juvenile open angle glaucoma for which Trabeculectomy was planned under general anaesthesia in the right eye.

General examination:

During pre-operative evaluation for fitness for surgery under general anaesthesia we found the boy to be short stature with stunted growth and facial dysmorphism. His body weight was 23 Kg. He had a triangular facies, maxillary and mandibular hypoplasia with high arched



Figure :1



Figure :2 & 3

The clinical features seen in him are genu valgum, spindle shaped fingers and arachnodactyly. He is known to develop allergic reaction to sulpha group of drugs. The cardiovascular and respiratory system were found to be within normal limits.

Investigations:

The routine laboratory investigations were within normal limits.

Random blood sugar: 97 mg/dl

Haemoglobin: 14.3 gm %

Blood urea: 11 mg/dl

Serum creatinine: 0.7 mg/dl

Na⁺: 142 mEq/L

K⁺: 4.1 mEq/L

Cl⁻: 106 mEq/L

HCO₃⁻: 18 mEq/L



Figure :4



A case of Ectodermal dysplasia posted for eye surgery: Anaesthetic and ophthalmic implications

His chest X-ray PA view (Figure 4) and 12-lead ECG was found to be normal. 2-D Echocardiogram showed normal bi-ventricular function with mild tricuspid regurgitation.



Figure : 5

X-ray findings of digits and hands showed osteopenia, widening of diaphysis, notching of metaphysis, distal phalanges acrolysis and tapering of diaphysis of proximal phalanges, see figure 5 .



Figure : 6 & 7

CT scan brain showed fusion of cranial sutures, narrowing of the optic canals and thinned out optic nerves, see figure 6 and 7.

Anaesthetic Management:

The boy was pre-medicated with Inj Glycopyrrolate 0.005 mg/kg IM 45 minutes before surgery. Pre-operative heart rate was 80-90/min and blood pressure was 90/70 mmHg. Since we anticipated difficult in airway a “trial of laryngoscopy”

was done with inhalational technique using sevoflurane 6%/O₂ : N₂O 3:3 flow. The vocal cords were visualized. Hence, we reduced the sevoflurane concentration to 2% /Induced with Inj Propofol 0.5mg/Kg IV and Inj Atracurim besylate 0.5mg/kg IV was given for intubation. Inj Fentanyl 1mcg/kg IV was given. Intubation was done using portex 6.0 ID mm cuffed Endotracheal tube. The tube position was confirmed by auscultation and end tidal carbondioxide. ECG, NIBP, pulse oximetry and end tidal carbondioxide were monitored. The surgery was done and surgery lasted for one hour. The child was reversed with neostigmine 0.08 mg/kg IV and atropine 0.02 mg/kg IV and extubated fully awake.

Case discussion:

Ectodermal dysplasia (anhidrotic ectodermal dysplasia, Christ-Siemens-Touraine syndrome) are described as “heritable conditions in which there are abnormalities of two or more ectodermal structures such as the hair, teeth, nails, sweat glands, cranial-facial structure, digits and other parts of the body.” It is not a single disorder, but a group of inherited disorders. The mode of inheritance varies among the different disorders. As an X-linked anhidrotic (non-sweating) trait, it mainly affects males. There is also an autosomal dominant form affecting males and females alike. However, transmission is from the female carrier, who usually appears normal and unaffected.

The main signs and symptoms found are:

Hair: Scalp and body hair may be thin, sparse and very light in colour.



A case of Ectodermal dysplasia posted for eye surgery: Anaesthetic and ophthalmic implications

Nails: Finger and toenails may be thick, abnormally shaped, discolored, ridged, slow growing or brittle.

Skin: Skin may be dry, thin and hypopigmented. It can be prone to rashes or infections, hence care must be taken to prevent cracking, bleeding and infection.

Teeth: Abnormal or missing teeth; lower-than-normal number of teeth; peg-shaped or pointed.

Sweat glands: It may function abnormally or may not have developed at all leading to inability to sweat. The body cannot regulate temperature properly and history of heat intolerance should be looked for. Children with the disease may have difficulty in controlling fevers.

Facial dysmorphism: Frontal bossing, longer or more pronounced chin, broader and low nasal bridge with or without maxillary and/ or mandibular abnormalities.

Eye involvement and the likely ophthalmic surgeries:

Tears: The function of lacrimal gland can be affected in Ectodermal dysplasia. This can predispose to infection, corneal ulceration and delay healing.

Tear drainage: The tear drainage system drains tears from the eye into the nose. Obstruction in this system can lead to infection of the tear drainage sac (dacryocystitis). Surgery required are syringing and probing the system (usually done in childhood) and dacryocystorhinostomy (DCR).

Cornea: The outer surface of the cornea epithelium, forms from the ectoderm. Thus in these patients corneal defects, ulcers and scarring can be present. Corneal problems are commonly seen in Ectrodactyly-ectodermal Dysplasia and Ectrodactyly, Ectodermal Dysplasia and Cleft Lip/Palate syndrome (EEC syndrome). Corneal ulcers are treated by corneal transplantation and ocular surface reconstruction.

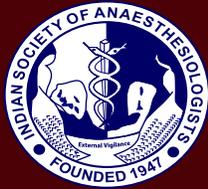
Lens: The lens forms from the ectoderm (surface skin) and hence cataract (congenital) can occur, which requires removal of cataractous lens surgery.

Anaesthetic implications:

The main anaesthetic implications are difficult in securing the airway owing to the presence maxillary and/or mandibular abnormalities and absence of teeth. A thorough examination of airway is needed during pre-operative evaluation of such cases. Hyperthermia may occur due to the defect of sweat glands and body temperature must be monitored continuously. Therefore, it is recommended to use humidified inspired gases both during surgery and in the post-operative unit too. Respiratory tract infections occurs frequently due to the absence of seromucosal glands.

The main aim of reporting this article is to emphasize that both ophthalmologists and anaesthesiologists should have an high index of suspicion of this rare disease in mind in patients with the above mentioned signs and symptoms and thus necessary measures to be taken must be planned well a head.





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