

Dr Jayanthi Sripathi
Editorial Team
Chennai

Editorial

The Annual IAPA conference scheduled on 8-10 February 2019 at AIIMS, Delhi under the Chairmanship of Dr. Rajeswari Subramaniam, Prof and HOD of Anaesthesia, promises to be one of its kind. The theme of the meeting 'Art and Science of Paediatric Anaesthesia' is most appropriate in today's era. The Art and Science of Paediatric Anaesthesia can only be learnt 'hands-on' in the operating rooms contrary to the belief of some who think they can learn by 'reading online'. Topics at the conference include most of the basics and others to keep abreast of the latest developments in the field. There are special sessions on neonatal surgery, difficult airway, thoracic surgery and chronic pain with lectures delivered by renowned experts from India and abroad.

Dr Dean Kurth, the architect of the 'Wake up safe initiative' of the Society of Paediatric Anaesthesiology (SPA,USA) will deliver the oration on 'Game changing innovations in Paediatric Anaesthesiology'. Dr. David Polaner of the PRAN network will be elaborating on the outcomes of the path breaking PANDA and GAS studies. Hearing these stalwarts first hand will be a privilege for all budding Anaesthesiologists.

Hands on workshops on Difficult Airway, Perioperative Ultrasound Course (PoCUS), Ultrasound Guided Regional Anesthesia (USGRA) and Simulation in Paediatric Anaesthesia will give the delegates a feel of the subject. In addition, faculty from five institutions will be initiated into the 'Wake up safe' workshop in collaboration with the SPA, USA., to initiate the same in their institutions.

The Indian Association of Paediatric Anaesthesiologists (IAPA) has entered the 13th year of its formation in 2019. The specialty of Paediatric Anaesthesiology has certainly come of age! There is so much happening within our society and in our work. The office bearers headed by Dr Elsa Varghese (President) and Dr Subrahmanyam (Secretary) have infused so much enthusiasm to make IAPA vibrant and effective. The IAPA has been organizing educational initiatives over the length and breadth of the country during the last year with CMEs in Kolkata, Varanasi, Vellore and the Perioperative Paediatric Life Support (PPLS) and Training the Trainer TTT workshops in Bangalore. The demand for the IAPA Paediatric Anesthesiology fellowships has markedly increased. The curriculum of the IAPA fellowship is now streamlined and the Exit Examination is held half yearly at the venue of the IAPA annual conference in February and during the mid-term executive meeting in August. The examiners chosen by the IAPA at a national level give the course extra credibility.

"Why do you need the Fellowship in Paediatric Anaesthesiology?" is one of the questions we pose to postgraduates who are interested in joining the fellowship. Some of them reply – 'simply to take away the fear of handling children'. Others want to be able to handle neonates safely. Anaesthesia postgraduates have about 1 month per year of their MD course in Paediatric Anaesthesia. Not all are at centers with high volumes or major Paediatric work. This kind of limited exposure without hands-on independent management does not prepare them for the real world of providing anaesthesia to children (in private practice) which they embark on soon after completing the MD course. Starting an IV in a small infant, handling crisis situations or managing the difficult airway in a neonate or infant becomes a nightmare and disaster ensues.

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INSIDE THIS ISSUE**Editorial**

By Dr Jayanthi Sripathi
Chennai. Pg. 1 & 2

Anaesthesia for paediatric cardiac catheterization / intervention

By Dr. Muralidhar K
Bangalore. Pg. 3 & 4

Anaesthetic management of a case of Klippel–Feil syndrome posted for cleft palate corrective surgery

By Dr. Deepika Teckchandani
Mumbai. Pg. 5 & 6

Anaesthesia challenges in freeman sheldon syndrome – case reports

By Dr. Geeta Kamal, Delhi.
Pg. 7 & 8

Umbilical Arterial Cannulation - Access, Technique and Complications

By Dr Mahmood Mirza
Hyderabad. Pg. 9 & 10

IAPA Activities 2018 – 2019

Pg. 11

Pediatrics Anaesthesia MCQs for IAPA News Letter

By Dr Abhishek Kumar, Dr Rakesh Garg. Delhi Pg. 12

PG tutorial

Emergence delirium in children

Dr S Sanjay Prabhu, Chennai.
Pg. 13 & 14

Recent interesting reads in paediatric anaesthesia

Dr Ekta Rai, Vellore. Pg 15

Crossword

By Dr Esha Nilekani.
Chennai, Pg 16

The Paediatric Anaesthesiology Fellowships conducted under the aegis of the IAPA aim to make the fellows competent, confident and comfortable with handling children. From the premature neonate to the obese adolescent, they are equipped to take on all. In fact, the fellowship should be a must for every MD postgraduate even if they do not plan to be full time Paediatric Anaesthesia. It's a onetime worthwhile investment for their anaesthesia career which fetches handsome returns lifelong! For those interested in Paediatric Anaesthesia as a career the DM course in Paediatric Anaesthesia is a wonderful opportunity to take it to the next level. Hearty Congratulations to Dr Nandini Dave at KEM Hospital, Mumbai and Neerja Bhardwaj at PGIMER, Chandigarh who have been recognized by their respective universities to conduct the DM course in Paediatric Anaesthesia.

We at IAPA salute their efforts.

Our History

“The Indian Association of Paediatric Anaesthesiologists” was formed in March 2006 by a small group of Mumbai based paediatric anaesthetists.

Dr. Pradnya Sawant took initiative for formation of the association.

Answers to MCQs:

1. Ans-c

Infants respiratory muscles are composed of type II muscle fibers and they achieve adult configuration of type I muscle fibers by the age of 2 years. This explains the easy fatigability of infants in case of increased work of breathing with subsequent apnea and desaturation.

2. Ans-d

RSI and muscle paralysis is contraindicated in these cases as it may lead to immediate loss of airway with immediate desaturation and hypoxia. Slow anesthesia induction in presence of difficult airway cart should be done.

3. Ans- c

Congenital pyloric stenosis presents at the age of 2 to 6 weeks of life and the child should be resuscitated before taking the patients to surgery. USG is the most common modality to diagnose the condition and metabolic derangements are due to loss of gastric fluid and profound vomiting.

4. Ans-c

The ability to regulate temperature is much more limited in infants than adults. Nonshivering thermogenesis, from the metabolism of brown fat, is much more significant in infants than in adults. Evaporative heat losses are also greater as a result of decreased keratin.

5. Ans-a

The risk factors for postoperative vomiting are different than in adults. Gender differences have not been shown to be a major factor until after puberty. All other choices have been implicated as risk factors of PONV in children.

ANAESTHESIA FOR PAEDIATRIC CARDIAC CATHETERIZATION / INTERVENTION

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For many years cardiac catheterization was the standard workup for obtaining both anatomical and physiological data to aid diagnosis in children with heart disease. Cardiac catheterization for diagnostic purposes in children is overtaken by other noninvasive imaging techniques. However, catheterization is done to resolve the anatomy in complex congenital heart disease patients and to perform haemodynamic measurements. The use of cardiac catheters for therapeutic treatment has increased steadily concomitant to the number decrease of diagnostic catheterizations^{1,2}. Interventional procedures are undertaken either as a non-surgical option or as an adjunct to operative treatment. The great advantage of interventional procedures is the avoidance of the use and complications of cardiac surgery and cardiopulmonary bypass. In addition, there is no postoperative pain and attendant ICU stay and hospital stay are dramatically reduced. Some of the specific concerns in providing care to the patients in catheterization suite are

- A. Haemodynamic disturbance: Some procedures are associated with marked haemodynamic disturbance; specific examples are aortic/pulmonary balloon valvuloplasty associated with loss of cardiac output during balloon inflation.
- B. High-risk patient: A significant number of children undergoing diagnostic/interventional procedures belong to high-risk category.
- C. Additional cardiac support: An occasional child may undergo catheterization whilst on extracorporeal membrane oxygenation. Familiarity with techniques and problems with institution and maintenance of mechanical support is mandatory for professionals caring for such patients.
- D. Arrhythmia and electrophysiology (EP) procedures: EP studies undertaken in catheter suite include demarcation and transcatheter ablation of abnormal conducting pathways and implantation of pacemakers.
- E. Environment: The cardiac catheterization laboratory is often located in a remote place, sometimes in a different floor altogether, from the operating room. The environment is frequently uncongenial for the anaesthesia, not brightly illuminated and crammed with equipment that makes patient access difficult. Hybrid suites are relatively new and larger in size; these complexes hold equipment for both catheter interventions and surgery, as well as circulatory support backup with extracorporeal membrane oxygenation or standard cardiopulmonary bypass.
- F. Risk of hypothermia: The catheterization suites are maintained at a cold ambient temperature and control of patient temperature is rather difficult paediatric population.
- G. Distant recovery room: Patients may be transferred some distance to recovery or intensive care facilities and the anaesthesiologist must be satisfied that the patient is in a stable condition prior to transfer. Monitoring, oxygen, resuscitation equipment and drugs, and sufficient personnel should accompany the patient, if this is the case.
- H. Design and space constraints: Competing "airspace" with suspended equipment (e.g., lights, injectors, echocardiography or ultrasound beams, and also gas columns and gas lines) can make the functional space very small regardless of the dimensions of the room. The movement of biplane fluoroscopy arms and their "parking place" frequently encroaches on the space at the head of the table.
- I. Recovery areas: post procedural recovery room should be immediately close by and have facility to monitor haemodynamics and institute mechanical ventilation.
- J. Exposure to radiation 3: A major concern during cardiac catheterization/intervention is exposure of both the patient and personnel to radiation. The risk of injury associated is increased with increasing dose (amount of energy absorbed); however, the magnitude of effect is not dose related.

Anaesthetic considerations

Optimal management goals of anaesthetic for paediatric cardiac catheterization/intervention are

- A) The patient free from pain and comfortable
- B) No respiratory depression,
- C) No cardiovascular depression
- D) Findings accurately reflect the pathophysiology
- E) Ensure no restlessness, delirium or agitation and
- F) Sedation light enough to allow a normal response at the end of the procedure.

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MONITORED ANAESTHETIC CARE FOR CARDIAC CATHETERIZATION / INTERVENTION IN CHILDREN-NH PROTOCOL4

1. Adequate fasting interval .
Water clear and fluids: up to 2 hours
Milk: up to 4 hours
Solid foods: up to 6 hours
2. Routine pre-anaesthesia check is carried out; chest must be clear both clinically and radiologically.
3. Echocardiographic evaluation/data/report is reviewed.
4. Consent for procedure is obtained.
5. Pre-procedure Hb and serum creatinine is estimated: HIV, HbSAg & HCV status of the patient is known.
6. Known allergies documented.
7. Blood grouping and cross matching necessary in infants < 5kg. All resuscitative equipment (especially endotracheal tubes and laryngoscopes)/drugs available and checked. Anaesthesia machine checked as per the OR protocol.
8. An overhead warming unit is needed for infants and small children.
9. Atropine, midazolam, ketamine IV in appropriate doses along with adequate local infiltration is used in stable children. Supplemental oxygen is given to all patients except those needing oximetry or those undergoing test of reactivity of pulmonary circulation for operability. In the latter, pulmonary pressure is documented prior to and after O₂ administration and subsequently O₂ is administration continued Sick babies and cyanotic infants and children have arterial blood gases done during and/after the procedure. Dose of atropine 20µg/Kg; glycopyrrolate 10µg/kg, midazolam 0.05mg/kg, ketamine 1-2 mg/kg and dexmedetomidine 0.5-1µg/ Kg/hr infusion.
10. ECG (for HR and rhythm), SaO₂, NIBP and respiration are monitored and recorded every 5-10 minutes throughout the procedure. Sick children and very tiny infants may need endotracheal intubation electively.
11. The following are included in the post-procedure care:
 - (a) O₂ inhalation (b) fluid administration@2ml/kg/hour (c) NPO Status for 2-4 hours (d) Monitoring of HR, SaO₂ continuously, RR & BP every 5-10minutes and record every 15 minutes
12. CPR ready @ all times; bradycardia give IV atropine 20µg/kg; VF = defibrillate @ 2 J/ Kg & adrenaline 10µg/ Kg; Chest compression @ 100/min, depth ≥1/3rd anteroposterior diameter of chest, allow chest recoil.

Typical cardiac catheterization data in normal children (pressure in mmHg)

	New Born	Older Children	Oxygen Saturation (%)
Right Atrium(mean)			
Mean	0-4	2-6	60-80
Right Ventricle			
Systolic	65-80	15-25	65-75
End Diastolic	2-7	3-8	
Pulmonary Artery			
Systolic	65-80	15-25	65-75
Diastolic	35-50	8-12	
Mean	40-70	10-16	
PA wedge			
Mean	5-8	7-13	95-100
Left atrium			
Mean	3-6	5-10	95-100
Left Ventricle			
Systolic	65-80	90-120	95-100
End Diastolic	3-7	2-5	
Aorta			
Systolic	65-80	90-120	95-100
Diastolic	45-60	60-75	
Mean	55-65	70-90	
Flows			
Pulmonary (Op)	3.5-5.0	3.5-5.0	95-100
Systemic (Qs)	3.5-5.0	3.5-5.0	
Resistance			
Pulmonary (Rp)	8-10	1-3	
Systemic (Rs)	10-15	15-30	

Anaesthetic management of a case of Klippel Feil syndrome posted for cleft palate corrective surgery

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Introduction :

Klippel Feil syndrome is a bone disease characterised by abnormal fusion of two or more cervical vertebrae.¹ Aetiology includes genetic mutation of GDF 6, GDF 3, MEOX1 genes and it has an autosomal dominant inheritance.² The clinical features include- short neck, low back hair line, restricted neck mobility and in severe cases, spinal stenosis all of which pose difficulty in managing the airway in these patient.³ This difficulty can get aggravated in airway surgeries like cleft palate repair surgery. We report this case to discuss the airway and systemic management in this patient.

Case report

A 2-year-old male child weighing 9 kg, clinically diagnosed as Klippel Feil Syndrome and with complete secondary cleft palate defect was posted for cleft palate repair. He had short webbed neck, restricted head and neck mobility, keel chest (Figures 1& 2) and Sprengel deformity, congenital high and under-developed scapula causing weakness of the shoulders (Figure 3). He also had history of seizures since 9 months of age with last episode 6 months back for which he was on Levetiracetam syrup. He underwent surgery for laryngo-tracheal cleft at the age of 4 months and required postoperative tracheostomy with paediatric intensive care stay for 10 days.

He was a full term, was delivered normally and cried immediately after birth but required neonatal intensive care for respiratory distress immediately after birth. Developmentally he showed delayed speech and spoke in mono and bi syllables only.

On examination, cardiovascular system was normal. In respiratory system he showed presence of keel chest, raised scapulae with equal air entry on both sides. Airway examination revealed Mallampati class 2 with thyromental distance less than 2 fingers and reduced neck movement pointing towards difficult intubation (Figure 4).

Blood investigations were within normal limits. 2-D echocardiography was not suggestive of any structural heart disease. MRI of the cervical spine revealed segmental anomalies, spina bifida from C3-C6. Computed tomography of cervical spine showed partial atlanto-occipital assimilation with fusion of the dens with left C4 anterior arch. Bilateral omovertebral bones extending to C5 with bilateral Sprengel defect. C1 vertebra was not visualised in neck X-ray suggestive of occipitalised C1 vertebra. Chest radiograph showed bilateral high clavicles and scapulae with a slight scoliotic spine (figure 5,6). EEG was suggestive of abnormal discharges from the left parieto-central area. Ultrasonography of the pelvis and abdomen ruled out renal abnormalities .

Direct laryngoscopy procedure few days before the cleft repair surgery in view of noisy breathing revealed a post epiglottectomy stub, no evidence of laryngotracheal cleft, bilateral normal vocal cords, short length of trachea and an 'A' shaped trachea.

On the day of surgery after an informed consent from parent, the patient was premedicated using nasal midazolam 0.3mg/kg. The operating room was kept ready with a difficult airway trolley and the ambient temperature was adjusted. Appropriate monitors were attached i.e. SpO₂, 5 lead ECG and NIBP. Intravenous induction with 1 µg/kg fentanyl and titrated doses of propofol with the aim of preserving spontaneous ventilation was carried out. We found that mask ventilation was easy in this patient.

After pre-oxygenation with 100% oxygen, direct laryngoscopy was attempted with size 2 Magill blade but the vocal cords were not visualised and only the epiglottic stub was seen. Second attempt of direct laryngoscopy done with McCoy blade size 2 revealed similar findings. The decision to move on to plan B using a 0 degree endoscope (diameter 4 mm, length 30 cm) by the ENT surgeon was taken. A gum elastic bougie was passed through the vocal cords under vision as seen on the 0 degree endoscope view and a size 4.5 south pole tube was passed over the bougie. Due to inability to negotiate the tube beyond the subglottic area, this tube was removed. During this period patient desaturated to 65% but could be ventilated back to 100%. A repeat attempt of 0 degree endoscope was done and size 3.5 uncuffed regular endotracheal tube was successfully placed. Air entry was confirmed and the tube was fixed. Wheeze was found on auscultation which was managed with injection dexamethasone 0.1 mg/kg intravenous and salbutamol puffs given through endotracheal tube. Patient was then paralyzed. Throat was packed, patient positioned and handed over to surgeon for repair. Intra-operatively patient was maintained on oxygen, air and isoflurane. Haemodynamics were stable throughout surgery which lasted for 50 minutes. Blood loss was around 20 mL which was replaced with Ringer's lactate solution.

At the end of surgery, throat pack was removed. A leak was detected along with rising trend of EtCO₂, hence decision to change tube to size 4.0 mm ID over bougie was taken. After successful change of tube the patient was shifted to PICU for elective ventilation in view of difficult intubation, airway manipulation and risk of bleeding post operatively. Patient was safely extubated the next day.

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Discussion

Difficult airway should be anticipated for Klippel Feil syndrome due to poor neck extension.^{4,5} In this patient, prior direct laryngoscopy finding of narrow airway, epiglottic stub and a narrow 'A' shaped trachea was suggestive of difficult intubation. Since the patient had adequate mouth opening and prior direct laryngoscopy was possible, our plan A was to perform direct laryngoscopy under anaesthesia preserving spontaneous ventilation. In anticipation of a narrow airway, small size tubes along with other aids to intubation were kept ready on difficult airway cart. Our plan B was intubation over 0 degree endoscope by ENT surgeon which was performed after the failure of Plan A. Plan C for our case was tracheostomy if plan B failed. Other possibilities are the use of video laryngoscope or flexible fiberoptic bronchoscope, both of which were not available in our institute at the time of the surgery.

All patients with difficult intubation should be anticipated for difficulty in extubation and need careful planning. In this case bleeding from raw area of palate after palatoplasty could have added to the difficulty. Hence we decided to electively ventilate the patient for 24 hours in PICU.

Conclusion

This case emphasizes careful planning for difficult airway in a patient with abnormal airway scheduled for airway surgery. Active involvement of surgical colleagues and using their expertise in Plan B and C in the absence of fiberoptic bronchoscope should be considered.

Fig 1 Front view of child showing webbed neck



Fig 2 Side view of child showing restricted neck extension



Fig 3 Back view of child showing Sprengel deformity



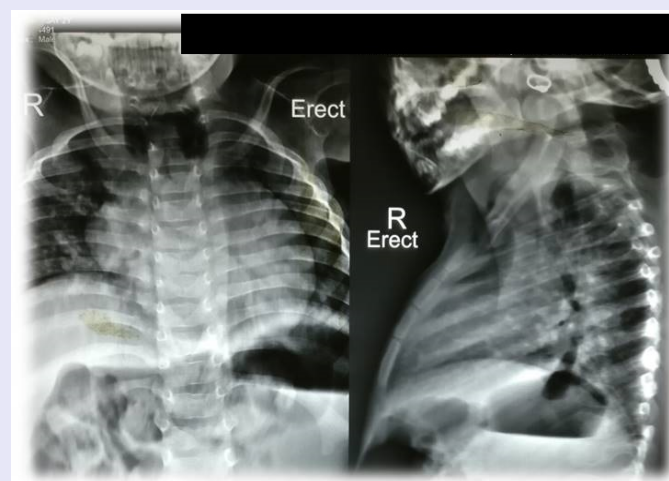
Fig 4 Child demonstrating adequate mouth opening



Fig 5 X- Ray cervical spine lateral view showing occipitalised C1 vertebra



Fig 6 CXR showing high clavicles and scapulae



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ANAESTHESIA CHALLENGES IN FREEMAN SHELDON SYNDROME – CASE REPORTS

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Introduction

Freeman-Sheldon syndrome (FSS) is a non progressive or slowly progressive congenital dysplasia characterised by contractures of facial, limb and respiratory muscles. The condition was first described in 1938 by Ernest Freeman, a British orthopaedic surgeon and Joseph Sheldon, a British physician.¹ The underlying myopathy produces weakness, increased tone or fibrosis. The three common dysmorphic features are: microstomia with pouting lips, camptodactyly with ulnar deviation of the fingers and talipes equino varus.^{2,3} FSS is also known as the craniocarpotarsal syndrome describing the parts affected; or “whistling face syndrome” due to the pursed mouth. “Windmill-Vane-Hand” syndrome and distal arthrogyriposis type 2A are used alternatively.³ The concerns related to anaesthesia are airway management, intravenous access, regional anaesthesia, a possible risk of malignant hyperthermia (MH) and body positioning.^{3,4,5} We describe the anaesthetic challenges faced during the management of patients with this rare syndrome.

Case 1

A 2-year male child weighing 10 kg presented with bilateral inguinal hernia. His face was noticeably expressionless with a puckered mouth (Figure 1). The interincisor distance was 1.5 cm and Mallampatti grade IV (Figure 2). The thyromental distance and neck movements were normal.

He had deep set eyes with hypertelorism and low set ears. Wrists showed ulnar deviation and contractures of fingers with no visible veins (Figure 3). A modest degree of developmental delay was noticed. Examination of the spine revealed scoliosis involving the thoracic spine. He had no previous anaesthesia exposures. Past medical history and family history was unremarkable. He was scheduled for a bilateral herniotomy.

He was referred to the child developmental clinic and was diagnosed as a case of Freeman Sheldon Syndrome. Preoperative haematological and biochemical investigations were normal. He was ASA physical status II on preoperative assessment. General anaesthesia supplemented with regional analgesia was planned. Written informed consent was obtained from the parents of the child after explaining the risks involved. Consent was also taken for the publication of the child’s photographs in scientific journals.

Size appropriate airway, fiberoptic bronchoscope and an emergency tracheostomy tray were kept ready. A MH free anaesthesia machine was prepared. Eutectic mixture of local anaesthetic (EMLA) was applied on his feet 1 hour before intravenous cannulation. Baseline vitals were recorded with non invasive monitors like ECG, pulse oximeter, NIBP and an axillary thermistor. By distracting the child’s attention, 24 G IV cannula was secured over the EMLA applied limb. Anaesthesia was induced with intravenous injections of 2 µg/kg fentanyl and 2 mg/kg propofol. The mouth opening did not improve even after jaw relaxation. A proseal laryngeal mask airway (PLMA) was chosen to preserve spontaneous ventilation. A lubricated completely deflated PLMA size 2 was easily inserted. The PLMA cuff was inflated at the oropharyngeal seal pressure of 50 cm H₂O.

USG guided bilateral ilioinguinal nerve block was given with 4mL of 0.25% bupivacaine. 150 mg paracetamol was given intravenously. Anaesthesia was maintained with 50% N₂O in O₂ and propofol infusion @ 3 mg/kg/hr on pressure control ventilation. Muscle relaxants were avoided. Intraoperative vitals including ETCO₂ and temperature remained stable. There were no signs and symptoms of MH. The 1.5 hr surgery was uneventful. LMA was removed when the patient was awake with return of reflexes. The postoperative period was uneventful.

Figure 1: Expressionless “whistling” facies with hypertelorism and low set ears. (Photo produced with parental permission)



Figure 2: Airway assessment revealing mouth opening of 1.5 cm and mallampatti grade



Figure 3: Bilateral ulnar deviation and contractures of fingers with adduction of thumb (windmill vaale position)



Case 2

A 10 month old, 6.4 kg girl presented with bilateral flat rocker bottom feet (Figure 4). She showed similar facial features (as case 1). Her lips were puckered as if whistling (Figure 5). She also had a limited mouth opening and a chin dimple. Her wrists showed ulnar deviation with contractures of fingers. Her ankle joints were stiff. Her muscle tone was generally increased. Perinatal, medical and family history was not significant. She underwent multiple corrective castings for foot deformity without prior anaesthesia exposure. Now she was scheduled for bilateral percutaneous tendo-achilles tenotomy.

On genetic workup, she was diagnosed as a case of Freeman Sheldon syndrome. Preoperative investigations were within normal limits. A written informed consent explaining the risks involved was obtained from the parents of the child. Consent was also taken for publication of the child's photographs in scientific journals.

As in previous case, difficult airway equipment was made readily available. The anaesthesia machine was prepared to avoid the possibility of MH. One hour prior to surgery, EMLA was applied on the forearm. O₂ was insufflated by a face mask held above the face of child with 50% O₂ N₂O mixture. 24 G intravenous cannula was secured over the EMLA applied area. Anaesthesia was induced with intravenous injection of 2 µg/kg fentanyl and 3 mcg/kg propofol. A well lubricated completely deflated size 1.5 PLMA was inserted through the restricted oral aperture. Perioperative analgesia was supplemented via caudal block with 5 cc of 0.25 % bupivacaine and 1 mcg/kg clonidine. Anaesthesia was maintained with 50% N₂O in O₂ and propofol infusion @ 4 mg/kg/hr on pressure control ventilation. Muscle relaxants and inhalational anaesthetics were avoided. The half hour surgery was uneventful without signs and symptoms of MH. LMA was removed after the patient demonstrated spontaneous eye opening and purposeful movements. The patient was discharged later the same day in good health.

Discussion

The fibrotic nature of FSS manifests with varying dysmorphic features. These patients pose several anaesthetic challenges. The most obvious concern for the anaesthesiologist is the difficult airway. Fibrosis of the facial muscles produce a mask like facies.³ Perioral fibrosis (microstomia) which is barely affected by neuromuscular blockade and restricted neck movements make laryngoscopy and intubation difficult.⁶ The mandibular hypoplasia, small nasal passages and kyphoscoliosis also contribute to the challenging airway. However microglossia and a high arched palate aid the proper placement of LMA.⁶ A characteristic dimple on the chin, hypertelorism with deep set eyes and short downslanting palpebral fissures are common.² The ears may be low set with a hearing deficit. The nasal cartilage is underdeveloped and alae nasi are notched.

Microglossia and the limited movement of soft palate cause a nasal speech. Feeding problems may result from microstomia and difficulties with swallowing. The pharyngeal muscles may be affected causing upper airway obstruction, gastroesophageal reflux and aspiration. They are at increased risk of POPC and recurrent respiratory infections. The associated scoliosis and pectus excavatum may result in abnormal respiratory mechanics. Sleep apnea and cor pulmonale due to chronic airway obstruction have been reported.²

The underlying myopathy may predispose these patients to malignant hyperthermia.² Though this association between FSS and MH is not supported by review of literature, abnormal responses to succinylcholine and halogenated volatile agents are well described. Unpredictable responses to neuromuscular blocking agents also increase the risk of POPC.⁵

Thus our technique of securing airway with LMA and maintaining spontaneous ventilation with a non-triggering anaesthetic technique is advocated in these patients. But at the same time emergency airway equipment should be kept ready. Peripheral nerve locator and USG guided blocks can minimise the use of sedatives and opioids and prevent POPC. So an ilioinguinal nerve block was considered for perioperative analgesia. However contracture and limited joint movement can make access to peripheral nerves difficult. Central neuraxial block can be used wherever possible in older children.^{4,7} But associated vertebral anomalies may be considered a contradiction to central blockade.

Deformed limbs with thickened subcutaneous tissues and frequent extremity surgery can result in difficult venous access, although this was not a problem in our patient. Central venous access may also be difficult due to the limited movement of the short neck.

There is an association with undescended testis and inguinal hernia.² Cardiac involvement is rare. Intelligence, general health and life expectancy are usually normal.

Conclusion

Every syndrome has a unique constellation of abnormalities which could be challenging from anaesthesiologists perspective. The difficult airway and a possibility of malignant hyperthermia are the chief anaesthesia concerns in a patient of FSS. A LMA and a non triggering anaesthesia technique should be considered in these patients

Figure 4: Bilateral congenital talipes equino varus



Figure 5: Puckered mouth – typical facies of Freeman Sheldon syndrome



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Umbilical Arterial Cannulation -Access, Technique and Complications

Mirza M.
Princess Esra Hospital
Hyderabad

Introduction:

The ethos of modern-day management of the sick neonate has increasingly become "proactive" rather than "reactive". Decisions in the neonatal intensive care are taken based on the hard evidence of quantitative measurements. Inherent in this transformation is the trend for greater intensive monitoring and invasive management, which is achieved by reliable access to the vascular space. The availability of high quality, sophisticated lines has made it possible to access the vascular space in the smallest of neonates in a manner which, till recently, was possible only in larger children and adults.

Umbilical Arterial Access:

Indications:

Umbilical arterial access is used in sick neonates for

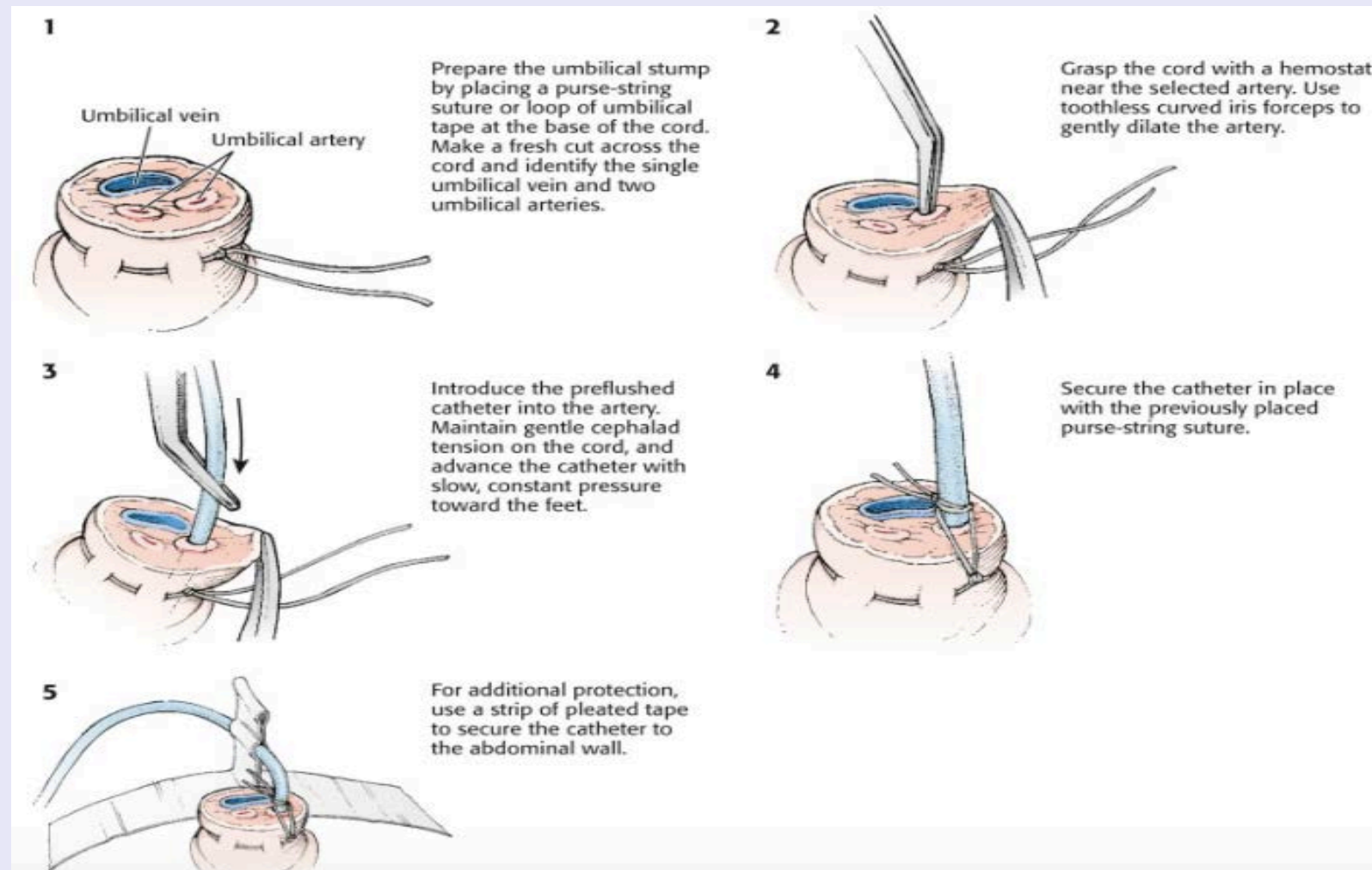
1. continuous intra-arterial blood pressure monitoring,
2. frequent blood gas determinations, and
3. isovolumetric exchange transfusions.

Contraindications

1. Omphalitis
2. Necrotizing enterocolitis
3. Peritonitis/acute abdomen
4. Omphalocele
5. Evidence of vascular compromise of lower limbs or buttock areas.

Technique:

Umbilical arterial catheterization is performed in neonates up to 1 week of age, and are most easily cannulated in the first few hours of life. After preparation of the skin and applying a tie to the base of the umbilical cord, the artery is identified. The umbilical cord contains two narrow lumens, thick-walled arteries, which have a tendency to project beyond the cut surface of the cord when it is fresh. The mouth of the artery is dilated using an iris forceps (plain) by inserting first one, then both limbs and applying gentle outward traction. Next, choose the position of the catheter, There is two safe positions 'high position' at the level of T6-9 thoracic vertebrae (above the origin of the celiac artery) and 'low position' below L2 vertebra (below the origin of the renal arteries). However, there are reports suggesting less risk of vascular compromise with the high position. Measure shoulder umbilical length (Shoulder umbilical length is the perpendicular distance between parallel horizontal lines at the level of the umbilicus and through the distal end of the clavicles).



To be continued

A size 3.5 to 5 Fr Umbilical catheter flushed with saline, is then threaded into the artery, which traverses posteriorly and caudad in its initial course and then upwards, a fact well delineated on a lateral radiograph of the abdomen when the catheter is in place. Correct placement is confirmed radiologically; as the 'high' position at T6-9 or 'low' position below L2 avoiding the origin of the renal arteries. The umbilical arterial line is then secured by a purse string suture, followed by a clear transparent dressing. A three-way stopcock attached to the hub of the line permits easy sampling as well as the provision of continuous infusion and intra-arterial monitoring of blood pressure.

Monitoring and care of the catheter

The patency of the arterial lines is by flushing it continuously with 1-2 mL/h of saline containing 1 unit/mL of heparin. Flushing the line after sampling is a must.

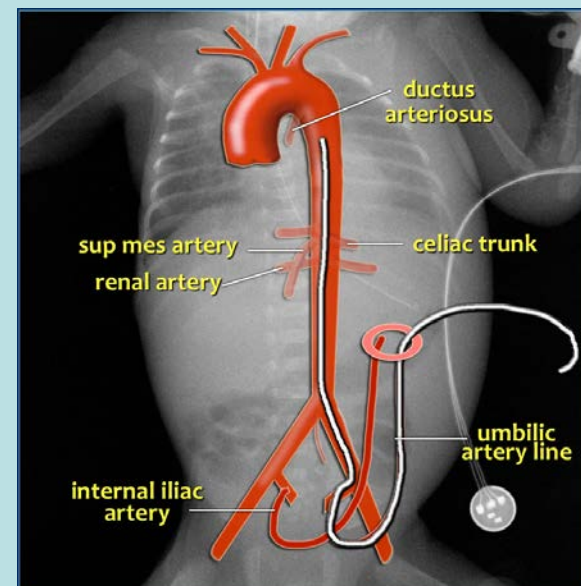
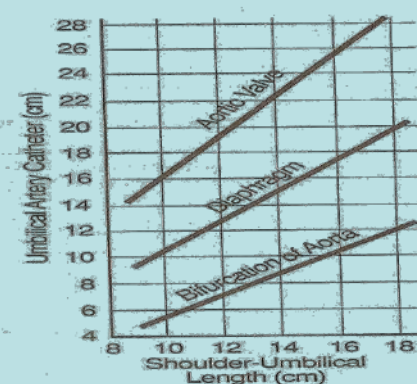
The line should be monitored for evidence of inflammation, distal ischemia, and local infection. Capillary circulation, the temperature of the distal limb, and pulses should be meticulously examined and recorded.

Hypertonic solutions, calcium and pressor agents should not be infused into them.

Complications

These include:

- Creation of false track
- Arterial spasm (which may be diagnosed by finding blanching of the opposite limb) leading to ischemic necrosis of the limb or gluteal region
- Embolism from blood clot or air
- Thrombosis of
 - mesenteric artery – leading to gut ischemia, necrotizing enterocolitis
 - renal artery [commoner in low umbilical arterial line] – renal failure
 - Femoral artery – limb ischemia or gangrene. Sudden cyanosis or pallor of a part below umbilicus especially a limb or toes may be related to embolism or spasm which warrants warming opposite leg with a warm towel. If the limb does not return to normal color within 5 min or if the gluteal region involved its advisable to remove the line.



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“IAPA Newsletter allows you to advertise your paediatric fellowships , vacancies related to paediatric anaesthesia.

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Activities 2018 - 2019

1. PPLS and TTT (ASPA)Jan 2019

IAPA and ASPA together have organised the Pediatric Perioperative Life Support (PPLS) Workshop and Training the Trainer (TTT) Programme in Bangalore on January 5th and 6th 2019. This was conducted at the Indira Gandhi Institute of Child Health, Bangalore under the able leadership of Dr Chandrika along with her enthusiastic team.

The ASPA Faculty included Dr Agnes Ng, Dr Josephine Tan and Dr Shu Ying from KK Hospital Singapore, Dr Lucy Chan and Dr Sushila Sivasubramanyam from the University of Malaysia, Kuala Lumpur, Malaysia, Dr Rebecca Jacob, Dr Vrushali Ponde and Dr Elsa Varghese from India The PPLS programme on Feb 5th 2019 was an intensive one-day programme. Thirty anesthesiologists participated.

The TTT (half day) programme was conducted on Jan 6th 2019 with fifteen participants (all had participated the previous day) and three faculty (Dr Agnes Ng, Dr Rebecca Jacob and Dr Elsa Varghese). The participants wholeheartedly participated and excellent role playing in the various scenarios. We look forward to taking the PPLS programme forward in a big way across India.

2. The FAST and Pediatrics association Anesthesia postgraduates one day conference- Nov 2011

The FAST and Pediatrics association Anesthesia postgraduates one day conference, attended by more than 100 participants, was held on 04-11-18 at L V Prasad eye hospital, KAR campus which was organized by the department of Anesthesia ,LVPEI . It was attended by Practicing anesthesiologists and post graduates from Telangana.

The first half of the day various current topics on pediatric and Ophthalmic Anesthesia were discussed. A panel discussion on operative sedation and day care surgery protocols were also talked about in detail . Effect of repeated exposure of anesthetic agents in children was spoken about as literature review .

The second half of the day four interesting hands on Anesthesia sessions were held.

1 Difficult venous cannulation (Indo-American Cancer Institute, Hyderabad) ;

2.Peribular block-Hands on Practice(Shankar Nethralaya, Chennai);

3.Airway management technique(LVPEI) ;

4.Pediatric CPR(LVPEI). It was well appreciated by all the faculty and delegates .

There were more than 100 participants who have attended this work-shop from different hospitals and medical colleges.



PPLS Course Bangalore

3. CME 2018 August

An update on Paediatric Anaesthesia was conducted by the Department of Anaesthesia along with IAPA, on 25th August, 2018 with focus on safe anaesthesia for neonates and infants. Dr. Sajjan Philip George was the organizing chairman and Dr. Ekta Rai was the organizing secretary.

The CME was attended by one hundred and thirty delegates and five external faculties. The renowned resource persons were Dr. Rebecca Jacob, President of the Asian Society of Paediatric Anaesthesia, Dr. Elsa Varghese, President of Indian Association Paediatric Anaesthesia, Dr. Krishnan N. Head of the Department ICH, Dr. H.M. Krishna, Head of Unit, KMC Manipal and Dr. Elizabeth Joseph, Trissur Medical College.

The sessions consisted of lectures highlighting neonatal physiology, pharmacology, fluid management, ventilation, anesthesia management for emergencies, difficult airway in neonates and infants. The post-lunch session had five skill stations on simulation, POCUS, pediatric resuscitation, vascular access and difficult airway. All the participants received a CME Workshop Manual which included write ups on all the above topics. As part of the CME, a quiz competition was organised in which more than fifty enthusiastic post graduates participated.



FAST and Pediatrics association Anesthesia postgraduates one day conference



CME Vellore

Pediatrics Anaesthesia MCQs for IAPA News Letter

Dr Abhishek Kumar, Dr Rakesh Garg

New Delhi

1. Compared to adults, which of the following changes in pulmonary system in pediatrics patients is incorrect?

- Dead space ventilation is similar to adults
- Oxygen consumption and work of breathing is more in infants
- Infant's diaphragmatic and intercostals muscles are composed of type I muscle fibers
- Chest wall and airways of infants are more compliant than adults.

2. Which of the following statement regarding airway management in a child with acute airway obstruction with stridor is incorrect?

- Inhalational based induction will be slowed in view of airway obstruction
- Halothane is preferred over sevoflurane for induction
- Mild laryngospasm can be relieved by applying PEEP of 10 to 15 cm H₂O
- RSI with succinylcholine administration should be done.

3. Congenital pyloric stenosis is characterized by

- Is a surgical emergency
- Normally presents at 6 months of life.
- Hypokalemia, hypochloremia and metabolic acidosis.
- Diagnosis is made by barium swallow study

4. Which of the following mechanisms best explains why infants, relative to adults, are more predisposed to hypothermia?

- Lower skin surface area to body mass ratio
- Increased keratin content in skin
- Higher thermoneural temperature
- None of the above

5. Which of the following does not increases the risk of postoperative vomiting in a prepubescent child?

- Female gender
- Strabismus surgery
- Age 3 or more
- Duration of surgery 30 minutes or longer

(Find answers on Page 2)

Answers to crossword

19. Polio (5)
 18. Archie(6)
 15. Murphy (6)
 13. POCA(4)
 10. Lemon (5)
 9. RDS (4)
 8. Grunt(5)
 7. Fastack (8)
 5. Jackson(7)
 3. Cockpit (7)
 1. Guedel(6)
- Down**
20. Whoosh(6)
 17. Chang(5)
 16. Magill (6)
 14. Saygo(5)
 12. Sprocket (8)
 11. Water bottle (11)
 6. Malt(4)
 4. Gregory(7)
 2. Cronzon(7)
- Across**

Emergence delirium in children

Dr S Sanjay Prabhu
Apollo Children's Hospital
Chennai

Children commonly display early postoperative negative behavior (e-PONB) after general anesthesia, which includes emergence delirium (ED), discomfort, temperament, and pain

Emergence delirium is a type of PONB characterized by a *self-limiting disturbance* in the child's awareness and attention

Definition

Emergence delirium (ED) can be defined as a "dissociated state of consciousness in which the child is irritable, uncompromising, uncooperative, incoherent and inconsolably crying, moaning, kicking or thrashing" ¹

Sikich and Lerman defined ED as "a disturbance in a child's awareness of and attention to his/her environment with disorientation and perceptual alterations including hypersensitivity to stimuli and hyperactive motor behavior in the immediate postanesthesia period." ²

Emergence agitation is a state of restlessness and mental distress that, unlike delirium, does not always suggest a significant change in behavior. It can arise from a number of sources including pain, anxiety or physiological disturbances. Not all children with agitation have delirium.

Causes

- Pain** - although analgesia has been found to prevent emergence delirium (see below), the involvement of pain in the genesis of emergence delirium remains controversial.
- Psychological** -psychological immaturity of children and their lack of adaptation to the perioperative anxiety during the perioperative period
- Pharmacokinetic factors** – clearance of anaesthetic agents leads to differential recovery rate from anesthesia of brain functions [cognitive function recovers later than audition and locomotion]

Risk factors

- Age: Occurs in 2- 5 yrs. age group
- Type of surgery: ENT, Eye surgeries > Urological or general surgery ³
- Parental anxiety : Parental anxiety can be passed on to their children.
- Patient anxiety :A child's degree of preoperative anxiety is correlated with their risk of exhibiting ED. A higher score on the modified Yale Preoperative Anxiety Scale (mYPAS) posed an increased risk of ED; specifically, for every additional 10-point increment, the risk of ED increased by 10% ⁴
- Preoperative psychological state :The patient's pre-existing temperament, anxiety, and coping behaviour are important factors in predicting emergence delirium.
- Interactions with health care providers : Negative behaviors displayed during interaction has a higher risk of ED
- Anesthesia Induction : negative behavior at induction is associated with a higher risk of ED ⁵
- Type of anesthesia:
 - More likely with inhalational agents [Higher with shorter acting than longer acting. More likely with desflurane and sevoflurane].It has been postulated that rapid awakening after the use of the insoluble anesthetics may initiate EA/ED by worsening a child's underlying sense of apprehension when finding himself in an unfamiliar environment
 - Absence of RA or inadequate analgesia seems to be a contributing factor

Clinical features

- Manifest most commonly in the first 15-30 min after awakening.
- Disorientation, perceptual disturbances [hypersensitive to stimuli], hyperactive motor behavior
- Typically, these children do not recognize or identify familiar objects or people

Impact of emergence delirium

Children experiencing e-PONB may suffer injuries and remove bandages, dressings, intravenous catheters, or drains. They require extra nursing care or additional sedative drugs in recovery room resulting in increased stress and anxiety in parents as well as caregivers and reduce overall satisfaction with anesthesia.

Children with ED are at a greater risk of having maladaptive behavioural change⁴ .

Scoring systems

3 scales are most commonly used to assess ED

1. Watcha – simple scale to detect presence of delirium
2. PAED scale – to quantify delirium, most validated scale
3. Cravero scale

Watcha Scale

Behaviour	Score
Asleep	0
Calm	1
Crying, but can be consoled	2
Crying, but cannot be consoled	3
Agitated and thrashing around	4

PAED Scale

Total score – 20. For a PAED score > 10, sensitivity of 0.64 and a specificity of 0.86 for ED

Behaviour	Not at all	Just a little	Quite a bit	Very much	Extremely
Makes eye contact with caregiver	4	3	2	1	0
Actions are purposeful	4	3	2	1	0
Aware of surroundings	4	3	2	1	0
Restless	0	1	2	3	4
Inconsolable	0	1	2	3	4

Inconsolability and restlessness have both been associated with delirium and may also reflect pain and both these features are part of two scoring systems – PAED scale [for delirium] and FLACC scale [for pain]. On the other hand, ‘no eye contact’, ‘no purposeful movements’, and ‘unawareness of surroundings’ were identified as core behaviors of ED in young children and hence are more useful for clinicians to assess ED in the recovery.

Cravero Scale

Behaviour	Score
Obtunded with no response to stimulation	1
Asleep but responsive to movement or stimulation	2
Awake and responsive	3
Crying (for >3 min)	4
Thrashing behaviour that requires restraint	5

The definition for ED in this scale is reached if level 4 or 5 was evident and present for at least 3 minutes despite active calming efforts. The Cravero scale has the advantage of simplicity.

Strategies to decrease ED**Behavioural**

A family-centred approach, **ADVANCE**, aims at Anxiety reduction, Distraction on the day of surgery, Video modelling and education before the day of the operation, Adding parents to the child’s surgical experience, No excessive reassurance, Coaching of parents by staff, and Exposure/shaping of the child via mask practice.

Anesthesia

Stressful induction possibly influences the development of ED. A smooth induction [by whatever methods possible] without restraints could be beneficial

TIVA seems to offer benefits over volatile agents in decreasing the incidence of ED ⁶ Propofol maintenance after sevoflurane induction decreases the possibility of ED. Subhypnotic doses of Propofol at the end of anesthesia have been shown to effective in preventing ED.

The time to emergence has been proposed as a contributor to EA/ED. A slower awakening may possibly decrease the risk of ED

Analgesia

Pain behaviour can be misinterpreted as EA or ED. It is also a good idea to rule out pain before treating ED

Pharmacological agents

Benzodiazepines - Though they do not directly decrease the risk of ED [7], it is still useful in alleviating preoperative anxiety which might indirectly affect ED.

Alpha agonists – Dexmedetomidine is superior to clonidine

Premedication with melatonin [0.2-0.4 mg/kg] , gabapentin [15 mg/kg], ketamine [1 mg/kg] has also shown to decrease ED ^[8,9,10.] Magnesium also seem to decrease the risk of ED

Narcotics have no effect on ED

Management of ED

Best method to deal with ED is by prevention rather than treatment. If a patient exhibits ED it is best to rule out pain first, as differentiating pain and ED can be challenging.

In case of intense agitation with a risk of self-injury, pharmacological treatment with Propofol, ketamine or dexmedetomidine should be considered. In situations with less intense agitation, it is important to reassure the parents and caregivers that it is self-limiting.

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Have you read this?

Recent interesting reads in paediatric anaesthesia

Dr Ekta Rai, Vellore

1. Feinstein MM, Pannunzio AE, Castro P. Frequency of medication error in pediatric anesthesia: A systematic review and meta-analytic estimate. *Pediatr Anesth* 2018;28(12): 1071-77.
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1. Rainbow Hospitals, Hyderabad.

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To my dear little one

By Paediatric Anaesthetist

Don't cry little one

I don't want to take you away (from mum)

I will give you some juice

If only 2 hours before

To keep you from being hungry

And upset with me

I will give you some stuff

That will put you to sleep

While we take you in as we listen to the Beep!

EMLA has been on – its more than an hour

The IV goes in

No howl in the corridor

Hush – the baby's asleep

Is not what they say

For its only now that the music's turned on (by surgeon)

As loud as can be

Be it Carnatic or Bollywood or the latest movie hits

There's always a new theme baby

that you have no clue about !

Its not the ET tube that I next ask for

She's been more than replaced by her sister

The LMA

A caudal, a block or a local infiltrate

She's all yours to start

The surgeon we beckon

Position the child, Tie the tourniquet, adjust the light

There are many little things we paediatric anaesthetists do.

Though the surgeon chides us for going at 11 for tea,

There's not a thing he can do in a child without us indeed!

Our greatest reward is when the baby awakes

Looking around and cooing happily

The caudal ,the midaz or the wonder drug

Dexmed

She is happy to go back to mum

Not knowing at all –(what went on all along inside)

Bye bye little one, see you sometime

Maybe you will come back to see your

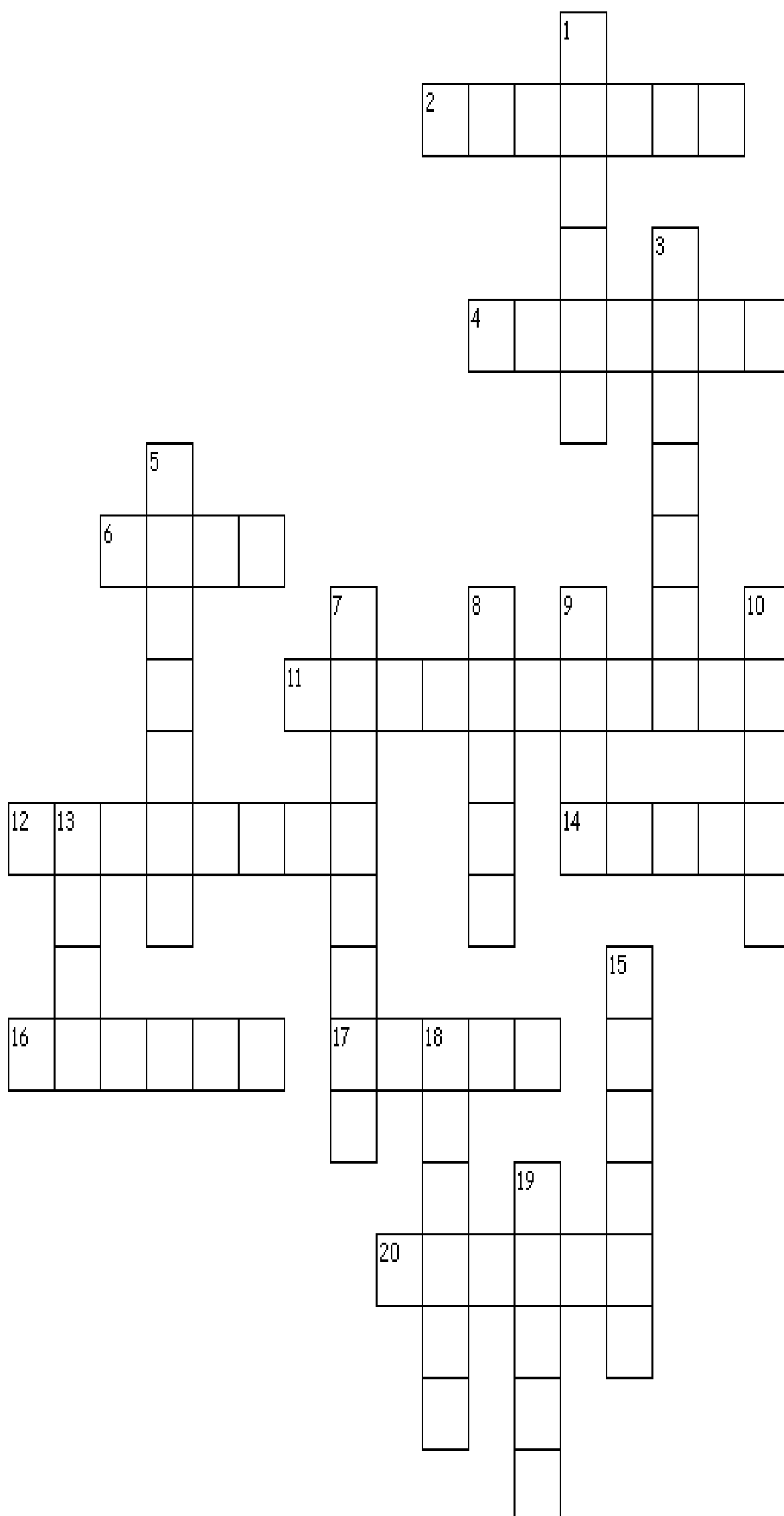
anaesthetist too

Who cared for you just like your mother did.

By Jay.

Crossword

Dr. Esha Nilekani, Kanchi Kamakoti Childs trust Hospital, Chennai.



Across

2. Syndrome which Rhymes with toasted bread pieces (7)
4. Name of Oscar winning Actor in "To kill a Mocking Bird"/
Discovered CPAP (7)
6. Fermented drink from barley/
Tonsil is a component (4)
11. It can be hot for pain relief/Xray
sign in pericardial effusion (11)
12. Pouch containing kings
tooth/Needle (8)
14. LA technique in FOB (5)
16. Irish born Anaesthetist (6)
17. Harry potter's one-time crush/
Textbook in mechanical
ventilation, Author(5)
20. Rapid gushing sound /Epidural
test (6)

Down

1. Stages of Ether anaesthesia/
Airway (6)
3. Pilot drill used in anaesthesia (7)
5. Singer whose death popularised
an inducing agent (7)
7. Watches/Quick recovery and
discharge of patients/ (8)
8. Snort like a Pig/Sign of distress(5)
9. mnemonic for difficult SAD
insertion (4)
10. Tropical Fruit/Quick Airway
assessment (5)
13. Registry for anaesthesia related
cardiac arrests in children (4)
15. What's meant to happen will
happen; law,IV set chamber)
18. First name of LMA pioneer (6)/
Comic book set in Riverdale
19. Preventable vaccine/blade (5)

(Find answers on Page 12)