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"AIRWAY ANOMALY IN SYNDROMIC CHILDREN" – ONE YEAR AUDIT OF THE PERIOPERATIVE ANAESTHETIC MANAGEMENT IN A TERTIARY CARE CENTRE

Dr. Sumalatha Radhakrishna Shetty B.Sc, MBBS, DA, DNB, Professor, Department of Anaesthesiology, K S Hegde Medical Acadamy,Nitte University,Mangaluru, Karnataka

Dr.Sindhu C* MBBS, Post Graduate, Department of Anaesthesiology, K S Hegde Medical Acadamy, Nitte University, Mangaluru, Karnataka *Corresponding Author

Dr.Ananda Bangera MBBS, MD, DNB, Professor and HOD, Department of Anaesthesiology, K S Hegde Medical Acadamy, Nitte University, Mangaluru, Karnataka

ABSTRACT

Background: Syndromic children can present with many congenital airway abnormalities which are relevant to anaesthesiologists. They may have difficult ventilation (eg. Pierre Robin syndrome), difficult intubation (eg.TMJ Ankylosis) or both(eg. Pierre Robin syndrome). This study evaluated the anaesthetic management of various syndromic children posted for craniofacial surgeries in a tertiary care centre.

Methods: This was a retrospective audit done on all syndromic children posted for craniofacial surgeries in one year from November 2015-16 in NitteMeenakshi Institute of Craniofacial surgery at K S Hegde Hospital. Their preoperative airway assessmentand perioperative management data were collected from case records and evaluated.

Results: In the study period, 475 maxillofacial cases were operated, out of which 15 were syndromic children with Pierre Robin syndrome (9),Congenital TMJ ankylosis(2), KlippelFeil syndrome(1), Goldenhar syndrome (1), Down's syndrome(1) and Treacher Collins syndrome(1). Majority of the cases were induced with propofol with spontaneous breathing and most of them were intubated with fibreoptic bronchoscope.

Conclusion: The management strategy of every syndromic child has to be planned based on history, anatomical and functional airway assessed during the pre anaesthetic evaluation. Proper preparation, planning and execution of the same with expertise is essential to manage all these patients.

Introduction

A syndromic child with associated airway abnormalities is a challenge for anaesthesiologist particularly so for an occasional paediatric anaesthesiologist. Reports regarding the real incidence of difficult airway management in children are sparse, but they are thought to be lower than in the adult population. Certain features predicting potential difficulties with airway management are often present in a number of syndromic children in paediatric anaesthesia practice. They can have dysmorphia, limited neck extension due to fusion of cervical vertebrae (Klippel– Feil syndrome), vertebral anomalies (Goldenhar's syndrome), restricted neck movements, atlanto-axial subluxation (Down's syndrome), limited submandibular space (Pierre Robin syndrome), maxillary and mandibular hypoplasia (Treachercollins syndrome).[1] Our study describes perioperative management of various syndromic children posted for craniofacial surgeries in our centre.

Methods

After approval by the Institutional Ethics Committee, we conducted a retrospective audit of all syndromic children who underwent elective maxillofacial surgery over a period of one year from November 2015-16. Data were collected from the case sheets obtained from Medical Record Section of the hospital and the photographs from computer system records of outpatient department of Nitte Meenakshi Institute for Craniofacial Surgeries. Information collected included demographic details, co-morbidities, associated syndromes, American Society of Anaesthesiologists(ASA) grade, preoperative airway assessment, premedication, intraoperative anaesthetic techniques and precautions taken for various syndromes.

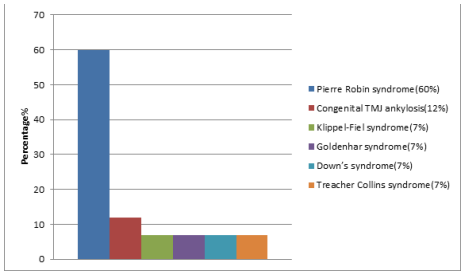
Results

In the study period, 475 maxillofacial cases were operated, out of which 15 were syndromicchildren. This included 9 males(60%) and 6 female(40%) children. The syndromes documented were Pierre Robin syndrome (9), Congenital TMJ ankylosis(2), KlippelFeil syndrome(1), Goldenhar syndrome (1), Down's syndrome(1) and Treacher Collins syndrome(1) (Figure 1). The clinical and demographic characteristics are shown in table 1.

Table 1: The clinical and demographic characteristics of various syndromic children undergoing craniofacial surgeries at K S Hegde Hospital, Mangaluru (n = 15)

Characteristic	n (%)
Gender	
Male	9(60)
Female	6(40)
Age group(years)	
<1	1(6.6)
1-3	4(26.6)
4-6	7(46.6)
7-12	2(13.3)
13-18	1(6.6)
Syndrome	
Pierre Robin syndrome	9(60)
Congenital TMJ Akylosis	2(12)
Down's syndrome	1(7)
Klippel–Feil syndrome	1(7)
Goldenhar syndrome	1(7)
Treacher Collins syndrome	1(7)

Figure 1: Incidence of various syndromes included in our study



The perioperative management is outlined in table 2. Airway assessment was done preoperatively for all the patients. Retrognathia was present in most of the cases(8),cleft palate in 4 cases, short neck in 2 cases, mal-aligned teeth in 6 cases, limited neck extension with atlantoaxial dislocation in one case, temporo-

mandibular joint immobility and limited mouth opening in one case. However Mallampati class was I in most of the cases(14). With regard to premedication, 6 children received syrup pedichloryl(trichlorfos), 7 received oral midazolam and 2 received tablet diazepam. Majority of the children were classified ASA I.

Table 2: The perioperative anaesthetic management of various syndromic children undergoing craniofacial surgeries at K S Hegde Hospital, Mangaluru (n = 15)

Characteristic	n(%)
Airway	
Retrognathia	8(53.3)
Cleft palate	4(26.6)
Short neck	2(13.3)
Mal-aligned teeth	6(40)
Atlantoaxial dislocation	1(6.6)
Limited mouth opening	1(6.6)
Premedication	
Syrup pedichloryl (trichlorfos)	6(40)
Oral midazolam	7(46.6)
Tablet diazepam	2(13.3)
Intubation	
Sedation with spontaneous breathing	13(86.6)
Airway block	1(6.6)
Paralysis	1(6.6)
Equipment	
Fibreoptic scope	12(80)
Ambuscope	3(20)

Airway management

Intraoperative events like induction and intubation were noted. All children were planned for fibreoptic intubation, 12 cases withKarl Storzfibreoptic-scope and 3 cases with Ambuscope ® . Of the total 15 children, 13 were intubated after sedation with propofol under spontaneous breathing and one under airway block. Only one patient aged 17years was intubated after paralysis with intravenous succinylcholine. Tracheostomy consent was taken preoperatively for all the patients. No direct laryngoscopy was tried in any of these children. Down's syndrome child with atlanto-axial instability was intubated with soft cervical collar in place. All cases were uneventful and were extubated on table once fully awake and conscious.

Discussion

Clinical evaluation of the syndromic child with a potentially difficult airway should focus on signs and symptoms of airway obstruction, including a history of any apnoeic episodes, daytime somnolence and also evidence of noisy breathing, stridor, snoring and increased work of breathing. The airway assessment must include dentition, extent of mouth opening, head–neck mobility, facial anomalies, anomalies of the palate and mandibular floor, and assessment of the thyromental distance. Congenital heart diseases are very commonly assosiated and must be assessed thoroughly. None of the children in study group had congenital heart disease.

Pierre Robin syndrome

Pierre Robin syndrome is characterized by the presence of the clinical triad of micrognathia, glossoptosis, and a U- or V-shaped cleft palate. It occurs in 1:8500 live births with an equal male-to-female ratio(60% PR sequence in our study). It is considered to be a sequence, since multiple secondary abnormalities could be caused by a single anomaly. Hypoplasia of the mandible prevents the palatal shelf from fusion between the 8th and 10th weeks of gestation resulting in retrognathiaand glossoptosis, thereby causing airway obstruction of variable severity.[2]

The degree of upper airway obstruction in the presence of mandibular hypoplasia has been divided into four types. In type 1, the obstruction describes true glossoptosis where the tongue touches the posterior pharynx at the level just below the soft palate. In type 2, the tongue touches at or just above the level of the soft palate, resulting in the soft palate being compressed between the tongue and the posterior pharyngeal wall. In type 3, obstruction is the result of medial compression by the lateral pharyngeal walls and in type 4, the pharynx constricts the airway in a sphincteric manner.

Surgical interventions depend on the type of obstruction. Most type 1 and 2 obstructions are managed with a nasopharyngeal airway or mandibular distraction procedures, although some type 2 obstructions require relief with tracheostomy. Most type 3 and 4 patients require tracheostomy for a more definitive relief of airway obstruction.[3] In the elective setting as in our case, fibreoptic intubation is the gold standard, although numerous other approaches have been used successfully.

Downs syndrome

Trisomy 21 is the most common chromosomal abnormality occurring in 1:600–800 live births(7% in our study). The main airway issues in these patients are related to their short neck, relative macroglossia, microdontia, mid-facial and mandibular hypoplasia, atlanto-axial instability with vertebral ligamentous abnormalities, and a higher incidence of congenital subglottic and/or tracheal stenosis. Tonsillar and adenoidal hypertrophy often contributeto upper airway obstruction, including obstructive sleep apnoea syndrome.

Patients with trisomy 21 often have tracheal narrowing and it is recommended that a tracheal tube 0.5–1 mm smaller than the calculated size for the age of the child be used to avoid subglottic trauma. Atlanto-axial instability in trisomy 21 is caused by the laxity of the transverseligament and/or bony abnormalities such as a malformation ofthe odontoid process.[4]The occurrence of symptoms and signs of myelopathiccompromise such as motor abnormalities (change in gait and weakness of arms or legs, spasticity, hyperreflexia), a change in bowel or bladder function, significant neck or radicular pain, head tilt, or torticollis should be investigated before anaesthesia.

Neck flexion-extension and rotation movements should be kept to a minimum in all these patients and proper precautions need to be taken for positioning in order to maintain the neck in a neutral position. In our case, position was maintained with the use of a soft cervical collar after induction to preserve the position of the neck.

Treachercollins syndrome

This disorder of neural crest formation involves the first and second branchial arches and is caused by a genetic mutation on chromosome 5. It occurs in 1:10,000 live births(7% in our study). Sixty per cent of cases arise from new mutations. The clinical features involve the head and neck and tend to be bilateral and symmetrical.[5]In contrast to Pierre Robin sequence, airway management in general and intubation in particular becomes more difficult with increasing age (mainly due to decreased mandibular growth).Successful intubation was done with ambuscope ® in our centre.

Goldenhar syndrome

This form of hemifacialmicrosomia, also known as oculo-auriculovertebralsyndrome or facio-auriculo-vertebral sequence, affects the first and second branchial arch resulting in ipsi- and unilateral (90% of cases) underdevelopment of the eye, ear, nose, soft palate, lip, and mandible. The aetiology is thought to be multifactorial, but very rarely familial causes have been described.Maintaining a seal for facemask ventilation can be difficult due to facial asymmetry. Difficult intubation arises from a combination of asymmetrical mandibular hypoplasia, hemifacialmicrosomia, tracheal deviation to one side, and craniovertebral abnormalities such as the possibility of C1–2 subluxation and potentially limited neck mobility.[6] The radiological evaluation of the cranio-facial-vertebral abnormalities using three-dimensional CT scanning has been recommended for selected patients. In our case, patient had mandibular hypoplasia, difficult

airway was suspected and fibreoptic intubation was planned.

Klippel–Feil syndrome

This syndrome consists of the classic triad of short neck, low posterior hairline, and limited neck mobility caused by the absence or fusion of cervical vertebrae. Abnormalities at the atlanto-occipital joint and spinal canal stenosis can co-exist. Sprengel's deformity, where one shoulder sits higher on the back than the other is seen in 25–35% of patients, contributing to decreased ventilatory capacity and difficulties with surgical positioning.[7] The syndrome occurs in 1:40 000 live births(7% in our study).

Owing to the cervical (and thoracic) spine anomalies, neck motion can be severely limited, which together with micrognathia and mandibular anomalies may render airway management difficult. There is a potential risk of spinal cord injury during laryngoscopy, intubation, and positioning of the patient. Sudden rotatory head movements inducing syncope due to compromised blood supply secondary to vascular anomalies (e.g. unilateral carotid artery agenesis) have been described. Careful neck movement during airway management and positioning is therefore mandatory.

Congenital TMJ ankylosis

Temporomandibular joint (TMJ) ankylosis results in inability to open mouth either partial or complete. In India incidence of TMJ ankylosis is still high. It is observed from 2 years of age to 60 years of age. Congenital TMJ ankylosis is very rare. Trauma or infection is the cause of TMJ ankylosis.[8] Facial asymmetry, malocclusion, anaemia, malnutrition may are the consequences. It also leads to increased airway obstruction, obstructive sleep apnoea and cor-pulmonale. Airway obstruction is secondary to structural encroachment on or pharyngeal and hypo pharyngeal lumen, subatmosphericintrapharyngeal pressure and hypotonocity of oropharyngeal muscles. All these structural deformities lead to difficulty in ventilation, intubation and extubation.[9]

Awake nasal intubation is the safest approach of intubation. Nasal intubation either blind or fibre-optic guided & awake or under anaesthesia, retrograde intubation & tracheostomy are the different techniques of securing airway in these patients. If anaesthetic agents are used there is risk of perioperative apnea, desaturation &dysarhythmia . Due to extreme sensitivity to central depressant drugs benzodiazepines and opioids should be used in titrated dose only. In our case awake nasal fibreoptic intubation was done under airway blocks. Child tolerated the procedure well and was uneventful.

Conclusion

The management strategy of every syndromic child includes proper history and the airway assessment of both anatomical and functional airway before induction of anaesthesia. Optimal airway management of patients with craniofacial abnormalities comprises-

- Good assistance
- Putting together an airway plan
- Knowing the airway equipment's use
- Modifying techniques
- Adequate training for difficult airway management

References

1. Raj D, Luginbuehl I. Managing the difficult airway in the syndromic child. Continuing Education in Anaesthesia, Critical Care & Pain. 2014 Apr 3;15(1):7-13
2. Meyer AC, Lidsky ME, Sampson DE, Lander TA, Liu M, Sidman JD. Airwayinterventions in children with Pierre Robin Sequence. Otolaryngol HeadNeck Surg 2008; 138: 782–7
3. Sher AE, Shprintzen RJ, Thorpy MJ. Endoscopic observations of obstructivesleep apnea in children with anomalous upper airways: predictive andtherapeutic value. Int J PediatrOtorhinolaryngol 1986; 11: 135–46
4. Brockmeyer D. Down syndrome and craniovertebral instability. PediatrNeurosurg 1999; 31: 71–7
5. Hosking J, Zoanetti D, Carlyle A, Anderson P, Costi D. Anesthesia forTreacher Collins syndrome: a review of airway management in 240 pediatriccases. PaediatrAnaesth 2012; 22: 752–8
6. Al-Abri AS, Khan RM, Haris A, Kaul N. Successful airway management inpatient with Goldenhar's syndrome using TruviewPCDw laryngoscope.PaediatrAnaesth 2012; 22: 1229–31
7. Bevan, J C. Congenital Syndromes in Paediatric Anaesthesia: Whatis Important to Know. Canadian Journal of Anaesthesia 1998;45:5, R3-R9
8. Neelima Anil Malik, Ankylosis of temporomandibular joint & its management. Ch. No. 22, Text Book of oral & maxillofacial surgeryFirst Edition, 207-219, 2002
9. Shah F R,Sharma R K, Hillowall R N ,Karandikar A D, J Indian Soc. PedoPrev Dentistry 2002 ;20;1,16-20