



# Practical Evidence in Anaesthesia Knowledge

## CASE SERIES

# Anaesthetic management in syndromic paediatric patients undergoing dental procedures: A case series

Rochana Girish Bakhshi<sup>1</sup>, Mona Sanjeevkumar Jadhav<sup>1</sup> , Niharika Ranjan<sup>1</sup>, Sarvesh Mishra<sup>1</sup>

<sup>1</sup>Department of Anaesthesia, D.Y. Patil Hospital, Nerul, Navi Mumbai, India

### \*Corresponding author:

Dr. Rochana Girish Bakhshi,  
Department of Anaesthesia,  
D.Y. Patil Hospital, Nerul,  
Navi Mumbai, India.

[drrochana@yahoo.com](mailto:drrochana@yahoo.com)

Received: 26 November 2024

Accepted: 20 December 2024

Published: 15 February 2025

### DOI

10.25259/PEAK\_14\_2024

### Quick Response Code:



## ABSTRACT

Rare congenital disease patients are a challenge to the anaesthesiologist especially when conducted under general anaesthesia. A thorough clinical examination and accurate history regarding previous anaesthetic management needs to be sought. This is a case series of paediatric patients who presented with congenital rubella, West syndrome, Down's syndrome, mucopolysaccharidosis (MPS) and Pelizaeus-Merzbacher disease (PMD) for dental procedures. The key to managing West syndrome patients is to continue antiepileptic drugs perioperatively and avoiding epileptogenic drugs. Patients with PMD have poor pharyngeal muscle control and subsequent airway complications. Down's syndrome is associated with multi-system comorbidities and atlantoaxial instability. Accumulation of glycosaminoglycans results in anatomical abnormalities and organ dysfunction in MPS patients. Congenital rubella syndrome poses challenges like unanticipated difficult airway and uncorrected cardiac defects.

**Keywords:** Airway problems, challenges, congenital syndromes, general anaesthesia, paediatric patients

## INTRODUCTION

A special child is defined as having physical, mental, sensory, cognitive, emotional, or behavioural disabilities that require special medical intervention, specialised services, or programmes.<sup>[1]</sup>

Tooth decay is a common problem in this set of patients due to higher amounts of carbohydrates in the diet, sugar content in medications, and poor oral hygiene with lower salivary flow in the oral cavity.<sup>[2]</sup> Patients on long-term antiepileptics have gingival hyperplasia, bleeding, petechiae and decreased platelet aggregation. Single-session dental treatment is appropriate and feasible.<sup>[3]</sup> We present here a case series of paediatric patients who presented with congenital rubella, West syndrome, Down's syndrome, mucopolysaccharidosis (MPS) and Pelizaeus-Merzbacher disease (PMD) posted for full mouth rehabilitation under general anaesthesia (GA) [Figure 1].

## CASE SERIES

### General Anaesthesia Management

Informed consent from parents was taken, and standard fasting guidelines, basic investigations and standard monitoring were done. Echocardiography was done to rule out cardiac anomalies. Any upper respiratory tract infection (URTI) was treated, and patients were optimised for the procedure. Difficult airway equipment was kept ready: videolaryngoscope, bougie, stylet, endotracheal tubes

This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-Share Alike 4.0 License, which allows others to remix, transform, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

©2025 The Authors; Scientific Scholar on behalf of Practical Evidence in Anaesthesia Knowledge



**Figure 1:** Syndromic patients. (a) Congenital rubella syndrome, (b) Cerebral palsy with West syndrome, (c) Down's syndrome, (d) Mucopolysaccharidosis, (e) Pelizaeus-Merzbacher disease.

(ETT), sizes: 3.5 to 5.5 mm microcuffed and uncuffed. Preoperative sedation: fentanyl 1 µg/kg and midazolam 0.03 mg/kg were given intravenously. Nasal xylometazoline drops were instilled. GA was administered with propofol 2 mg/kg followed by bolus dose of 0.5 mg/kg atracurium as muscle relaxant. After propofol, a well-lubricated nasal airway was inserted and kept in situ for three to four minutes till the patient was being ventilated post-atracurium. Throat packing was done post-intubation and removed at the end of procedure after thorough suctioning. Hypoxia, hypercapnia, hypothermia and medications that would decrease seizure threshold were avoided. Patients were positioned carefully, and joints and bony prominences were padded to prevent fractures. Every child received 0.1 mg/kg dexamethasone post-induction. Intravenous paracetamol 15 mg/kg body weight was used for analgesia. Anaesthesia was maintained with a mixture of air, oxygen and sevoflurane (1.5% to 2%). At the end of the procedure, after the return of airway reflexes, extubation was done. The paediatric intensive care unit (PICU) was reserved for every child but only the MPS child needed ICU admission for 24 hours.

#### **Case 1. Congenital rubella syndrome**

A 17-year-old boy, a known case of congenital rubella, had encephalopathy with attention deficit hyperactivity disorder, autism, bilateral hearing loss, absence of speech, microcephaly, and microcornea. He was mentally retarded, unable to walk, had spasticity of lower limbs, and was dependent on his mother for all activities. 2D echocardiography showed moderate pulmonary stenosis, Ebstein anomaly and severe pulmonary arterial hypertension.

Low tidal volume without positive end-expiratory pressure was used for controlled ventilation to avoid an increase in pulmonary vascular resistance (PVR). High-dose opioids and nitrous oxide were avoided for the same reason. The intraoperative course was uneventful.

#### **Case 2. West syndrome**

Fourteen-year-old known case of West syndrome had cerebral palsy, secondary hyperparathyroidism, delayed milestones and mental retardation. He was on oral pacitane, clonazepam, vitamin D supplements, and baclofen twice a day, which was continued preoperatively.

After the administration of GA as described above, a 6mm polyvinyl chloride nasal micro-cuffed ETT was placed. The intraoperative course was uneventful.

#### **Case 3. Down's syndrome (Trisomy 21)**

An eight-year-old child with Down's syndrome had a history of recurrent ear infections and deafness with snoring at night. He was cheerful, outgoing, a mouth-breather with typical features such as depressed bridge of the nose, frontal bossing, obstructive sleep apnoea (OSA) and impaired immune functions causing recurrent ear infections.

The patient was induced with sevoflurane and propofol 1 mg/kg. After ensuring ventilation, atracurium 0.5 mg/kg was given. Intubation was done using a 5 mm nasal ETT. At the time of induction and thereafter patient had junctional rhythm. After ruling out other causes, sevoflurane was replaced with isoflurane, and the junctional rhythm was resolved. The patient was extubated with a nasal airway in situ as the child had a depressed nasal bridge with a history of snoring.

#### **Case 4. Mucopolysaccharidosis**

A 13-year-old male, a diagnosed case of MPS type IIIc, was uncooperative, aggressive, unable to speak or follow commands, had macroglossia, a history of snoring, short neck, frontal bossing, depressed bridge of the nose, lower loose incisors and buck teeth.

On the day of surgery, the child was agitated and uncooperative and was hence sedated preoperatively. Induction was by inhalational technique using incremental doses of sevoflurane with oxygen and air (40:60) followed by propofol 1 mg/kg. Oral intubation was done with a 5.5 mm microcuffed ETT as nasal intubation was not possible due to the depressed nasal bridge. At the end of the procedure, a 4.5 mm nasal airway was inserted in the right nostril as there was snoring post-extubation. He was shifted to the PICU with nasal airway in situ for 24 hours considering

the difficulty in maintaining the airway and the need for sedation for postoperative aggressive behaviour.

#### **Case 5. Pelizaeus Merzbacher Disease**

A three year-old patient with PMD had delayed developmental milestones, nystagmus and abnormal muscle tone with hypotonia. The patient had a history of multiple episodes of URIs with a recent episode one week prior. There was a history of hypoglycaemia and a need for enteral feeding for which he was hospitalised, 2–3 times since birth.

The main concern in a patient with PMD is prolonged neuromuscular blockade for which neuromuscular monitoring is advised. Preoperative blood sugars were checked and glucose-containing intravenous fluid was given preoperatively. The patient was given nebulisation preoperatively due to the recent URI. Lateral radiograph of the skull revealed adenoidal hypertrophy.

Nasal intubation was avoided due to adenoid hypertrophy and subsequent chance of bleeding, traumatic adenoid avulsion and aspiration. Oral endotracheal intubation was performed using 4.5 mm micro cuffed ETT.

## **DISCUSSION**

Multisystem involvement affecting syndromic children poses challenges to the anaesthesiologist. Apart from routine investigations, thyroid function, serum calcium and serum glucose may be required. Echocardiography is done to rule out congenital heart disease. Parents need to be counselled for the high risk involved in general anaesthesia. Placement of an intravenous cannula can be challenging due to poor nutrition, muscle wasting and spasticity of limbs. Premedication with opioids carries the risk of respiratory depression but is often required for separation from parents. These patients need anticholinergic premedication as they have excessive secretions due to overactive salivary glands. Dysfunction of cranial nerves reduces clearance of pharyngeal secretions which needs thorough suctioning at the time of induction and extubation. Difficult intubation cart should be ready in craniofacial malformed patients.<sup>[4]</sup> Short trachea, tracheomalacia and bronchomalacia are some associated airway anomalies. Macroglossia, abnormal dentition and mandibular abnormalities cause difficult mask holding. Most dental procedures need nasal intubation, so adenoidal hypertrophy must be ruled out preoperatively. Pre-existing OSA and velopharyngeal dysfunction predispose to postoperative airway obstruction. Long-term anticonvulsants can alter liver function, hence anaesthetic drugs need careful titration. Fluid restriction may be needed in patients with cardiac abnormality. Padding of pressure points and careful

positioning are required in osteoporotic and spastic children. Body temperature is maintained by warming blankets as malnourishment and hypothalamic dysfunction predispose to hypothermia and delayed emergence.

The syndromic child is not a good candidate for dental chair anaesthesia as the margin of safety for any type of sedation or inhalational technique is narrow due to limited resources, inadequate monitoring and lack of expertise in resuscitation.

West syndrome patients have spasticity of limbs, which is reduced after induction. Careful padding and positioning is done to prevent iatrogenic fractures.<sup>[5]</sup>

PMD is a type of leukodystrophy affecting the nervous system. Due to pharyngeal muscle hypotonia, these patients need enteral feeding to prevent poor nutrition and aspiration.<sup>[6]</sup> Due to prolonged neuromuscular blockade, a low dose of muscle relaxants and neuromuscular monitoring is advised under GA.

The significant features of Down's syndrome are atlantoaxial instability (15%), subglottic stenosis, obstructive sleep apnoea, enlarged tongue and tonsils. Associated cardiac defects are atrial septal defect, tetralogy of Fallot, ventricular septal defect and pulmonary hypertension (PHT). These patients are prone to bradycardia, hypotension and arrhythmias if induced with a large dose of sevoflurane, so gradual dosing is advised.<sup>[7]</sup>

The MPS patient in this case series was type IIIC (Sanfilippo), with a deficiency of acetyl coenzyme A. Progressive cellular and multisystemic damage occurs due to partially degraded glycosaminoglycans (GAG). This defect occurs due to a deficiency of specific lysosomal enzymes. As years pass, new GAG deposition may change the previous airway anatomy, affect cardiac and pulmonary functions, causing problems like OSA. Airway abnormalities predispose to obstruction and difficult intubation, with possible "cannot intubate/cannot ventilate" scenarios. It is safer to have the patient spontaneously breathing until successful intubation is done or (at least) confirmation of mask ventilation. He was extubated and shifted to the PICU with a nasal airway inserted for 24 hours, similar to the case managed by Walker R et al.<sup>[8]</sup>

Maintaining the systemic vascular resistance (SVR) to PVR ratio is of utmost importance in congenital rubella patients posted for non-cardiac surgery. It is prudent to avoid hypercarbia, hypoxia, hypothermia and acidosis to maintain PVR. An increase in right to left shunt, reduced pulmonary perfusion and hypoxaemia may result due to a fall in SVR. Nitrous oxide is avoided due to evident PHT. It is beneficial to maintain a slightly lower heart rate and to augment preload in patients with pulmonary stenosis.<sup>[9]</sup>

## CONCLUSION

When preparing a child with a syndrome for GA, it is important to consider all systems that may be affected. Factors that may impact overall care and outcomes, like poor nutrition, repeated URTI, difficult intubation and chronic anticonvulsant drugs should be well managed for the safe conduction of syndromic patients. Multidisciplinary team involvement is mandatory for better care.

Careful planning includes anticipation and preparedness to deal with the airway and altered physiology with choice and dosing of anaesthetic agent and proper patient positioning. Postoperative care must be individualised and depending on the severity of clinical condition, high-level care may be required.

**Declaration of Patient Consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship:** Nil.

**Conflicts of interest:** There are no conflicts of interest.

**Use of artificial intelligence (AI)-assisted technology for manuscript preparation:** The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

## REFERENCES

- Alfarraj J, Louise Gray J, Alargan T, Alkathiri M, Alshehri R, Almarwan M. Dental treatment provided to special needs children under general anesthesia in a tertiary care hospital – A cross sectional retrospective study. *Saudi Dent J* 2024;36:579–83.
- Ahuja R, Jyoti B, Shewale V, Shetty S, Subudhi SK, Kaur M. Comparative evaluation of pediatric patients with mental retardation undergoing dental treatment under general anesthesia: A retrospective analysis. *J Contemp Dent Pract* 2016;17:675–8.
- Solanki N, Kumar A, Awasthi N, Kundu A, Mathur S, Bidhumadhav S. Assessment of oral status in pediatric patients with special health care needs receiving dental rehabilitation procedures under general anesthesia: A retrospective analysis. *J Contemp Dent Pract* 2016;17:476–9.
- Ciccozzi A, Pizzi B, Vittori A, Piroli A, Marrocco G, Della Vecchia F, et al. The perioperative anesthetic management of the pediatric patient with special needs: An overview of literature. *Children (Basel)* 2022;9:1438.
- Şahin SH, Çopuroğlu E, Uğur H, Sağroğlu G, Çolak A. Anaesthesia management of a child with West syndrome. *Turk J Anaesthesiol Reanim* 2014;42:362–4.
- Kapoor R, Zavala AM, Van Meter A, Williams UU, Porche VH, Owusu-Agyemang P. Anesthetic challenges and successful management of a child with Pelizaeus-Merzbacher disease using general and caudal anesthesia. *J Anaesthesiol Clin Pharmacol* 2018;34:250–1.
- Borland LM, Colligan J, Brandom BW. Frequency of anesthesia-related complications in children with Down syndrome under general anesthesia for noncardiac procedures. *Paediatr Anaesth* 2004;14:733–8.
- Walker R, Belani KG, Braunlin EA, Bruce IA, Hack H, Harmatz PR, et al. Anaesthesia and airway management in mucopolysaccharidosis. *J Inher Metab Dis* 2013;36:211–9.
- Manjula VR, Koshy PG, Prasad MS, Saradadevi V. Congenital rubella syndrome and anesthetic considerations. *MRIMS J Health Sciences* 2018;6:51–3.

**How to cite this article:** Bakhshi RG, Jadhav MS, Ranjan N, Mishra S. Anaesthetic management in syndromic paediatric patients undergoing dental procedures: A case series. *Pract Evid Anaesth Knowl*. 2025;1:40–43. doi:10.25259/PEAK\_14\_2024