

## CASE SERIES

## PEDIATRIC ANESTHESIA

# Anesthetic management of pediatric patients with congenital heart disease posted for cochlear implant: a case series

Hina N. Gadani<sup>1</sup>, Nidhi D. Jariwala<sup>2</sup>, Mahir M. Kureshi<sup>3</sup>, Vishwa M. Shah<sup>4</sup>, Purvi D. Thakkar<sup>5</sup>

**Author affiliation:**

1. Hina N. Gadani, Associate Professor, Department of Anesthesia, GMERS Medical College, Gandhinagar, India; E-mail: [hinagadani@gmail.com](mailto:hinagadani@gmail.com)
2. Nidhi D. Jariwala, Resident doctor, Department of Anesthesia, GMERS Medical College, Gandhinagar, India; E-mail: [jariwalanidhi46@gmail.com](mailto:jariwalanidhi46@gmail.com)
3. Mahir M. Kureshi, Resident doctor, Department of Anesthesia, GMERS Medical College, Gandhinagar, India; E-mail: [mahirkureshik@gmail.com](mailto:mahirkureshik@gmail.com)
4. Vishwa M. Shah, Assistant Professor, Department of Anesthesia, GMERS Medical College, Gandhinagar, India; E-mail: [shahvishwa14@yahoo.com](mailto:shahvishwa14@yahoo.com)
5. Purvi D. Thakkar, Assistant Professor, Department of Anesthesia, GMERS Medical College, Gandhinagar, India; E-mail: [thakkarpurvid90@gmail.com](mailto:thakkarpurvid90@gmail.com)

**Correspondence:** Dr. Vishwa M. Shah; E-mail address: [shahvishwa14@yahoo.com](mailto:shahvishwa14@yahoo.com)

## ABSTRACT

Congenital heart disease (CHD) is the most common birth defect which might be a part of a broader syndrome. Such patients pose many challenges when subjected to anesthesia for non-cardiac surgery. Therefore, a patient-specific peri-operative management plan needs to be employed to reduce peri-operative morbidity.

We conducted 10 pediatric cases with CHD posted for cochlear implant surgery. The study was conducted in GMERS Medical College, Gandhinagar, India. Each case was managed peri-operatively with a standardized protocol. All patients had acyanotic CHD, and no patient had complex CHD. They were pre-operatively assessed in detail and optimized if required. As part of intra-operative management, care was taken to avoid sudden hemodynamic fluctuations by using etomidate as induction agent and atracurium as muscle relaxant. Inj. lignocaine was used to prevent hemodynamic response to intubation. Adequate analgesia was achieved. None of the patients had any adverse cardiac event peri-operatively. We were able to manage each patient successfully without any untoward consequences.

**Key words:** Congenital heart disease; Cochlear implant; General anesthesia; Pulmonary hypertension; Pulmonary vascular resistance; Systemic vascular resistance

**Citation:** Gadani HN, Jariwala ND, Kureshi MM, Shah VM, Thakkar PD. Anesthetic management of pediatric patients with congenital heart disease posted for cochlear implant: A case series. *Anaesth. pain intensive care* 2023;27(4):590–595; DOI: [10.35975/apic.v27i4.2272](https://doi.org/10.35975/apic.v27i4.2272)

**Received:** May 11, 2023; **Reviewed:** May 25, 2023; **Accepted:** Jun 07, 2023

## 1. INTRODUCTION

Congenital heart disease (CHD) is the most common birth defect, which occurs approximately in one in 125 live births.<sup>1</sup> CHDs may be a part of a syndrome with other anomalies. Therefore, when such patients are subjected to anesthesia for elective or emergency

surgeries, they warrant vigilance on the part of peri-operative physician. Detailed knowledge about anatomy and physiology of various CHDs and other anomalies, type of CHD and the type of circulation, i.e., normal, balanced (series) or single ventricle, is required in order to assess the peri-operative risk.<sup>1</sup> We managed 10 pediatric cases with CHD posted for cochlear implant surgery.

## 2. CASE HISTORY

### Case 1

An 11-y old male, weighing 25 kg, presented with deafness and mutism since birth was posted for right side cochlear implant. Patient was born at full term with normal vaginal delivery without any complication, weighing 3 kg. There was no history of jaundice, breathlessness, convulsions, and no NICU admission. Patient was fully vaccinated till date and achieved developmental milestones at normal age.

He was a known case of CHD and 2D-echo suggested moderate perimembranous VSD with left to right shunt and mild AR and aortic valve prolapse. Patient was operated for VSD before 7 y under general anesthesia, and 3 units packed cells were transfused intraoperatively. Postoperatively patient developed right sided pneumothorax. Patient was kept in ICU for 2 days for observation postoperatively.

At the time of presentation in our hospital, patient's vital signs were within normal limits. All investigations were also within normal limits with Hb 13.1 gm%. On chest X-ray sternal sutures were noted. Recent 2D echo was suggestive of ventricular septal defect (VSD) repair with normal biventricular function and no residual leak across VSD.

Fitness was given by the cardiologist.

### Case 2

A 5 months old male baby, weighing 11 kg being, mute and deaf since birth, as informed by his parents was posted for right cochlear implant. Patient was born at full term with normal vaginal delivery without any complications, with a birth weight of 2 kg. There was no history of jaundice, convulsions or respiratory distress or any NICU admission. Patient was fully vaccinated and achieved developmental milestones at normal age. There was no sibling or family history of similar complaints. Nor was any significant medical or surgical past history.

Patient was a known case of CHD with 2D-echo suggestive of a small size ostium secundum atrial septal defect (ASD), L to R shunt, a tiny patent ductus arteriosus (PDA) with left to right shunt, mild tricuspid regurgitation (TR), no pulmonary artery hypertension (PAH) and mild left atrial (LA) and left ventricle (LV) enlargement.

At the time of presentation in our hospital, patient's vital signs were within normal limits. On cardiovascular system (CVS) examination a continuous murmur was heard. MRI brain was suggestive of periventricular leukomalacia. Parents also gave history of upper

respiratory tract infection (URTI) twice in a month which was optimized and controlled preoperatively. All investigations were within normal limits with Hb 10.0 gm%.

Fitness was given by the cardiologist.

### Case 3

A 3-y old male patient, weighing 14 kg, presented with complaints of being mute and deaf since birth as informed by his parents. He was posted for right cochlear implant. Patient was born at 8 months of gestational age with normal vaginal delivery without any complications, weighing 2.3 kg, twin child, and had a history of NICU admission for 2 days. Patient was vaccinated till date, with developmental milestones achieved at normal age. There was no sibling or family history of similar complaints.

Patient was a known case of acyanotic CHD with a 2D-echo suggestive of trivial TR. At the time of presentation in our hospital, patient's vital signs were within normal limits, on CVS examination grade-II systolic murmur was heard.

MRI brain suggested T2W hyperintense / mild hypointense T1W signal white matter abnormality, involving periventricular white matter adjacent to lateral ventricle.

All investigations were within normal limits with Hb 12.6 gm%

### Case 4

A 5-y old female patient weighing 13 kg, presented with history of being deaf and mute since birth as informed by parents, was posted for right cochlear implant. Patient was born at full term with normal vaginal delivery weighing 2.5 kg without any complications. There was no history of NICU admission, convulsions or respiratory distress or cyanosis. Patient was fully vaccinated till date and had achieved developmental milestones at normal age. There was no surgical or medical past history. There was no sibling or family history of similar complaints.

Patient was a known case of CHD with 2D-echo suggestive of a tiny PDA with left to right shunt. There was no PAH. His LV and LA were normal. At the time of presentation in our hospital, patient's vital signs were within normal limits. On CVS examination Grade-1 systolic murmur was heard. high-resolution computed tomography (HRCT) brain was normal.

There was a history of active URTI which was controlled with antibiotics preoperatively.

All investigations were within normal limits with Hb 10.9 gm%.

As per cardiologist opinion no other cardiac interventions were needed and patient was asked to follow up after 6 months for PDA.

### Case 5

A 2-y old male patient weighing 10 kg, presented with complaints of being mute and deaf since birth as informed by parents. Patient was born at full term with normal vaginal delivery weighing 3 kg without any complications. There was no history of cyanosis, jaundice, convulsions, respiratory distress, or an NICU admission. Patient was vaccinated till date and had achieved developmental milestones at normal age. There was no previous medical or surgical history. There was no sibling or family history of similar complaints.

Patient was a known case of CHD, with 2D-echo suggestive of a large ostium secundum ASD with left to right shunt. There was no PAH.

At the time of presentation in our hospital, patient's vital signs were within normal limits.

HRCT brain suggested asymmetrical 'fluid attenuated inversion recovery' (FLAIR). Hyperintense areas seen in B/L peritrigonal, periventricular white matter and a few FLAIR hyperintense lesions in subcortical white matter of both parietal lobes. All investigations were within normal limits with Hb 9.1 gm%

As per cardiologist opinion patient could undergo ENT surgery with mild cardiac risk.

### Case 6

A 4-y old female child weighing 12 kg, presented with complaints of being mute and deaf since birth as informed by parents. Patient was born at full term with normal vaginal delivery weighing 3.3 kg without any complications. There was no history of cyanosis, jaundice, convulsions, respiratory distress, or NICU admission. Patient was vaccinated till date and had achieved developmental milestones at normal age. There was no previous medical or surgical history. There was no sibling or family history of similar complaints.

Patient was a known case of CHD with 2D-echo suggestive of a small PDA with left to right shunt. Vital signs were within normal limits, and CVS examination revealed a continuous systolic murmur. HRCT and MRI brain were normal. All investigations were within normal limits with Hb 11 gm%.

Cardiologist fitness was obtained for surgical intervention.

### Case 7

A 4-y old female patient, weighing 12 kg, presented with complaint of being mute and deaf since birth as

informed by her parents. The child was born at full term with normal vaginal delivery weighing 3.3 kg without any complications. There was no history of cyanosis, jaundice, convulsions, respiratory distress, or NICU admission. Patient was vaccinated till date and had achieved developmental milestones at normal age. There was no previous medical or surgical history. There was no sibling or family history of similar complaints.

Patient was a known case of CHD with 2D-echo suggestive of a small PDA with left to right shunt. At the time of presentation in our hospital, patient's vital signs were within normal limits. On CVS examination a continuous systolic murmur was heard. HRCT and MRI brain were normal. All investigations were within normal limits with Hb 11 gm%.

Cardiologist fitness was given for surgical intervention.

### Case 8

An 8-y old female, weighing 20 kg, presented with deafness and mutism since birth, and was posted for right side cochlear implant. Patient was born preterm with cesarean section without any complication, weighing 2.0 kg. There was no history of jaundice, breathlessness, convulsions, or NICU admission. Patient was fully vaccinated till date and achieved developmental milestones at normal age.

Patient was a known case of acyanotic CHD and 2D-echo suggested mild perimembranous VSD with left to right shunt and trivial TR.

At the time of presentation in our hospital, patient's vital signs were within normal Limits. All investigations were within normal limits with HB- 10 gm%. MRI brain was insignificant.

Fitness was given by the cardiologist.

### Case 9

A 5-y old male, weighing 6.5 kg presented with deafness and mutism since birth was posted for right side cochlear implant. Patient was born preterm with normal vaginal delivery without any complications, weighing 1.8 kg. There was a history of jaundice after birth which was managed conservatively. There was no history of breathlessness, convulsions, or NICU admission. Patient was fully vaccinated till date and achieved developmental milestones at normal age.

Patient was a known case of acyanotic CHD, and his 2D-echo suggested mild ostium secundum type of ASD with right ventricular systolic pressure (RVSP) of 38 mmHg.

At the time of presentation to our hospital, patient's vital signs were within normal limits. MRI brain was insignificant. All investigations were within normal limits with HB 10.2 gm%.

Fitness given by the cardiologist.

### Case 10

A 4-y old male child, weighing 7 kg presented with deafness and mutism since birth was posted for right side cochlear implant. Patient was born at full term with cesarean section for meconium stained liquor and CPD without any complications, weighing 2.5 kg. There was no history of jaundice, breathlessness, convulsions, or NICU admission. Patient was fully vaccinated till date and achieved developmental milestones at normal age.

Patient was a known case of CHD and his 2D-echo suggested mild AR, mild MR. RVSP was 35 mmHg.

Patients vital signs were within normal limits at the time of presentation in our hospital. MRI brain was insignificant. All investigations were within normal limits with a HB of 11 gm%.

Fitness was given by the cardiologist.

### Peri-operative Management

We conducted 10 pediatric cases posted for cochlear implant with history of congenital cardiac anomalies. Standardized protocol was followed in all cases peri-operatively. All patients were pre-operatively assessed. A detailed history was sought and complete physical examination was done to assess the kind of cardiac lesion and stratify the risk associated with surgery and anesthesia. Additional non-cardiac anomalies were ruled out. Upper and lower respiratory tract infections were ruled out and optimized if any before surgery.

History of previous surgery and medication intake was taken. Out of ten, one patient had been operated for moderate perimembranous VSD with left to right shunt. He did not have any medication history.

Routine laboratory investigations were done like complete blood count, renal function, coagulation profile and serum electrolytes. Chest radiographs were ordered. A 12 lead ECG and 2-D echo was done in all patients to identify the type of CHD, status of shunt, cardiac function and status of pulmonary hypertension. None of our patients had complex CHD.

All patients were planned for the cochlear implant surgery and fasting orders were given according to recent fasting guidelines for elective surgery. The risk associated with the surgery and anesthesia was explained to the parents of patients and all patients were labelled as ASA-III.

On the day of surgery venous access was obtained and patients were premedicated by inj. midazolam 0.2 mg/kg and inj. fentanyl 2 µg/kg. Etomidate 0.3 mg/kg was used as an induction agent along with sevoflurane in oxygen. Inj. lignocaine (preservative free) 1 mg/kg was used 90

sec before laryngoscopy to attenuate hemodynamic response to laryngoscopy and intubation. Muscle relaxation was achieved with inj. atracurium 0.5 mg/kg and the patients were intubated with appropriate-sized micro-cuffed endotracheal tube after application of lignocaine jelly. Intubation was done by an experienced anesthesiologist to reduce the time of laryngoscopy. All patients were ventilated by pressure-controlled mode. Maintenance of anesthesia was done with sevoflurane in 50% oxygen and 50% nitrous oxide, along with bolus doses of inj. atracurium as and when required. Inj. paracetamol 15 mg/kg was given intraoperatively as an analgesic. Inj. methylprednisolone was injected for surgical requirement. At the end of the procedure, smooth extubation was achieved by inj. lignocaine 1 mg/kg and reversal agent inj. neostigmine 0.05mg/kg with inj. glycopyrrolate 0.008 mg/kg. Patients were extubated after complete return of motor power, tone and reflexes. All patients were observed in the post anesthesia care unit (PACU) for six hours and no immediate or early post-operative complications were noted.

### 3. DISCUSSION

Cochlear implant is an electronic prosthesis which partially replaces the functions of cochlea in patients with severe sensorineural hearing loss.<sup>4</sup> Of all the patients with hearing loss, 50% cases are hereditary.<sup>4</sup> Among these 30% also have additional abnormalities, like craniofacial, skeletal, ocular, neurologic, renal and cardiovascular system defects as a part of a syndrome.<sup>4</sup> The most important of these anomalies are cardiac anomalies which pose a significant challenge to the anesthetist. Various syndromes have been described pertaining to cardiac anomalies such as the Jervell Lange-Nielsen syndrome, Goldenhar syndrome, congenital rubella syndrome, Pierre Robin syndrome, CHARGE (coloboma, heart defects, atresia choanae, growth retardation, genital abnormalities, and ear abnormalities) syndrome, Treacher Collins syndrome, branchio-oto-renal syndrome, stickler, Aperts, Crouzon, Velocardiofacial, mitochondrial disorders, etc.<sup>3</sup> Most of the pediatric patients appear asymptomatic when they present to pre-anesthesia clinic. For the purpose of risk stratification, patients with CHD are classified into 3 categories: normal circulation, balanced circulation and single ventricle circulation.<sup>1</sup> Therefore, thorough knowledge about diagnosis and management of congenital cardiac and other anomalies is required on the part of peri-operative physician in order to successfully manage such patients and avoid major mishaps.

We successfully managed 10 pediatric cases posted for cochlear implant surgery with history of congenital cardiac anomalies. In addition to the risk associated with

extremes of age, there are additional risks associated with cardiac anomalies.

Upper and lower respiratory tract infections have to be ruled out as they may cause changes in airway reactivity and pulmonary vascular resistance (PVR) which may be poorly tolerated in children with reduced pulmonary compliance or patients with pre-existing pulmonary hypertension (PHT). PHT has been seen to increase perioperative morbidity.<sup>9,10</sup>

History of associated non-cardiac congenital anomalies, like musculoskeletal abnormalities, neurological defects and genitourinary irregularities should also be ruled out as they may impose additional risk for anesthesia, for example a patient with Down's syndrome may have atlanto-occipital subluxation, that will warrant airway management precautions.<sup>3</sup> History of previous surgery and medication is needed and past documents will provide true information about the type of CHD, type of intra-cardiac shunt and help us categorize the cardiac risk that an elective non-cardiac surgery might pose.

Peri-operative anesthetic management should include a strategy that maintains balance between PVR and SVR. Sudden changes in heart rate and sympathetic stimulation must be avoided as these patients have reduced cardiac reserve.<sup>11</sup>

Premedication is important in such patients to avoid distress and anxiety and thereby minimizing oxygen consumption. It also reduces the amount of induction agent and thus minimizes reductions in SVR.

Etomidate was used as an induction agent along with sevoflurane. Etomidate provides smooth induction and stable hemodynamics throughout surgery, which might not be possible with other induction agents like propofol. Propofol significantly reduces SVR and MAP and may cause increase in shunt flow in patients with right to left shunt. Inhalational agents should also be used in titrated manner as they sensitize myocardium to catecholamines. We used sevoflurane in less than 1.5% concentration for induction and less than 1% for maintenance.

These patients might not tolerate sudden hemodynamic responses to laryngoscopy and intubation. Tachycardia during laryngoscopy will increase myocardial oxygen demand and might cause myocardial ischemia and life-threatening arrhythmias. To attenuate this stress response, lignocaine 1% was used during intubation and extubation. Smooth intubation was achieved by using atracurium instead of succinylcholine.

The hemodynamic parameters remained stable throughout the perioperative period. We could achieve this by using above measures and adequate plane of anesthesia. The balance between PVR and SVR was

maintained by using the described measures and adequate analgesia intraoperatively.

## 4. CONCLUSION

Children with CHD who present for non-cardiac surgery are at increased risk of perioperative morbidity. Factors that contribute to morbidity are pre-existing pulmonary hypertension, reduced cardiac reserve, right to left shunt, cardiac failure, history of cyanosis and arrhythmias. Anesthesia for children with CHD needs complete understanding of the anatomy, physiology, and risk factors associated with perioperative morbidity and mortality. Low risk and intermediate risk cases can be successfully managed during elective procedures with minimal complications by maintaining balance between pulmonary and systemic vascular resistance, avoiding undue sympathetic stimulation and adequate analgesia in perioperative period.

## 5. Conflicts of interest

The authors declare no conflicts of interest.

## 6. Authors' contribution

HNG, NDJ: Conduct of study, Manuscript editing

MMK: Conduct of study, Data collection

VMS: Conduct of study, Literature search, manuscript editing

PDT: Conduct of study

## 7. REFERENCES

1. White MC. Approach to managing children with heart disease for noncardiac surgery. *Paediatr Anaesth.* 2011;21(5):522-9. [PubMed] DOI: [10.1111/j.1460-9592.2010.03416.x](https://doi.org/10.1111/j.1460-9592.2010.03416.x)
2. Phillips M. Genetics of hearing loss. *Medsurg Nurs.* 2003;12(6):386-90, 411. [PubMed]
3. Shahani JM. Anaesthetic considerations in children with congenital heart disease undergoing non-cardiac surgery. *Indian J Anaesth.* 2012;56:491-5. [PubMed] DOI: [10.4103/0019-5049.103969](https://doi.org/10.4103/0019-5049.103969)
4. Jafra A, Panda NB. Anesthetic implications for cochlear implant in a child with cardiac anomaly. *Anaesth pain intensive care.* 2017;21(2):260-2. [Free Full Text]
5. Greenwood RD, Rosenthal A, Parisi L, Fyler DC, Nadas AS. Extracardiac abnormalities in infants with congenital heart disease. *Pediatrics.* 1975;55(4):485-92. [PubMed]
6. Ramamoorthy C, Haberkern CM, Bhananker SM, Domino KB, Posner KL, Campos JS, et al. Anesthesia-related cardiac arrest in children with heart disease: data from the Pediatric Perioperative Cardiac Arrest (POCA) registry. *Anesth Analg.* 2010;110(5):1376-82. [PubMed] DOI: [10.1213/ANE.0b013e3181c9f927](https://doi.org/10.1213/ANE.0b013e3181c9f927)
7. Warner MA, Lunn RJ, O'Leary PW, Schroeder DR. Outcomes of noncardiac surgical procedures in children and adults with congenital heart disease. *Mayo Perioperative Outcomes Group.* *Mayo Clin Proc.* 1998;73(8):728-34. [PubMed] DOI: [10.4065/73.8.728](https://doi.org/10.4065/73.8.728)

8. Taylor CJ, Derrick G, McEwan A, Haworth SG, Sury MR. Risk of cardiac catheterization under anaesthesia in children with pulmonary hypertension. *Br J Anaesth.* 2007;98(5):657-61. [PubMed] DOI: [10.1093/bja/aem059](https://doi.org/10.1093/bja/aem059)
9. Carmosino MJ, Friesen RH, Doran A, Ivy DD. Perioperative complications in children with pulmonary hypertension undergoing noncardiac surgery or cardiac catheterization. *Anesth Analg.* 2007;104(3):521-7. [PubMed] DOI: [10.1213/01.ane.0000255732.16057.1c](https://doi.org/10.1213/01.ane.0000255732.16057.1c)
10. Karmakar CS, Hannan MA, Islam MS, Bhowmik DK, Akhtaruzzaman AKM. Pulmonary hypertension following severe COVID-19: a case report. *Anaesth. pain intensive care* 2021;25(4):539–543.. DOI [10.35975/apicare.v25i4.1554](https://doi.org/10.35975/apicare.v25i4.1554)
11. Chari VRR, Suryawanshi CM, Sahu P. Anesthetic management of a case of a large atrial septal defect with mild pulmonary hypertension for emergency cesarean section. *Anaesth pain intensive care.* 2013;17(2):182-4. [Free Full Text]