Case Report

DOI: 10.7860/JCDR/2023/66005.18481

Anaesthesia Section

Anaesthetic Management of a Down's Syndrome Patient Acyanotic Congenital Cardiac Anomalies and Hypothyroidism: A Case Report

SHYAMOLIMA BHUYAN[1], DEEPJIT BHUYAN[2]

ABSTRACT

Down Syndrome (DS) is a common chromosomal disorder that is associated with multiple anomalies in different organ systems. Cardiac anomalies are frequently observed in individuals with DS, and as patients with Congenital Heart Disease (CHD) are at an increased risk of developing complications related to anaesthesia during the perioperative and postoperative periods, it is crucial to pay attention to the anaesthetic management of DS patients undergoing corrective surgery for cardiac anomalies. Complete Atrioventricular Septal Defects (CAVSD) are the most prevalent cardiac defects in DS patients, followed by isolated ventricular septal defects, atrial septal defects, patent ductus arteriosus, and tetralogy of Fallot. This case report focuses on the anaesthetic management of a one-year-old female with DS who was diagnosed with a combination of Atrioventricular Septal Defects (AVSDs) and patent ductus arteriosus. Due to the concurrent diagnosis of hypothyroidism, the case required a thorough preanaesthetic examination and meticulous attention to perioperative anaesthetic management, considering factors such as airway difficulty, cervical spine instability, ligament laxity, and susceptibility to infections. The patient underwent cardiac surgery following a standard anaesthetic protocol, and the procedure was well tolerated. After the surgery, she was transferred to the Intensive Care Unit (ICU). The postoperative period was uneventful, and the patient was discharged on the eighth postoperative day.

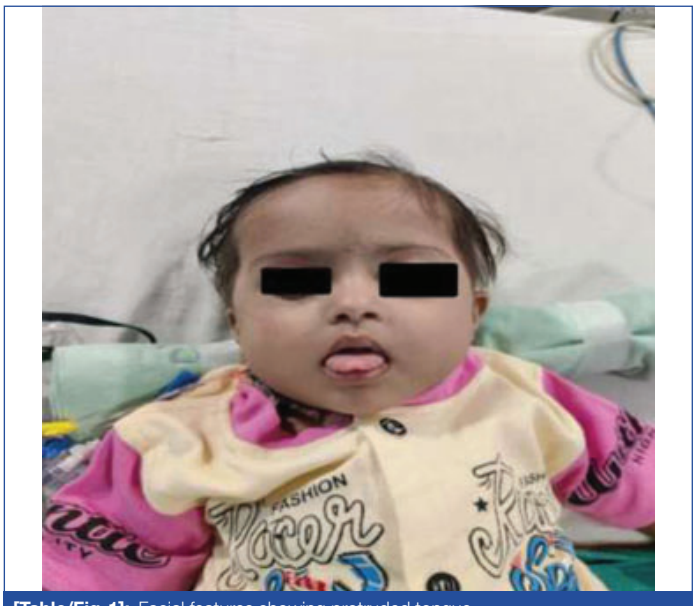
Keywords: Anaesthetics, Cardiac, Down's syndrome, Hypothyroidism, Infant

CASE REPORT

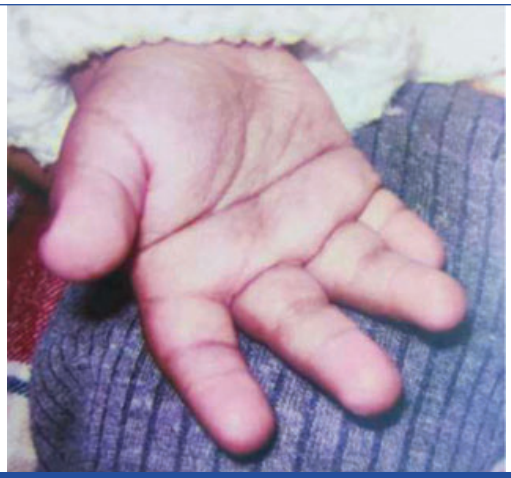
A one-year-old female child with Down's Syndrome (DS), hypothyroidism, and multiple acyanotic heart diseases (ventricular septal defect, atrial septal defect, and patent ductus arteriosus) was admitted to the hospital for surgical correction of the heart defects. DS was diagnosed at birth through karyotype analysis, while the heart defects were identified 15 days after birth, and hypothyroidism was diagnosed at 10 months. The child was prescribed diuretics (Furosemide drops 5 mg) twice daily, a vasodilator (enalapril 2.5 mg tablet) once daily, and a daily dose of 25 micrograms of thyroxine following the respective diagnoses. The child had a history of breathing difficulty, forehead sweating during feeding, recurrent episodes of cough, and failure to gain weight. In terms of the birth history, the child was born prematurely at eight months and two days of gestation through normal vaginal delivery. There was no immediate cry after birth, and the birth weight was 2.3 kg. The family history revealed a second-degree consanguineous marriage between the parents.

Developmental delay was observed, with gross motor milestones of neck holding achieved at six months, ability to roll over at eight months, but inability to sit without support. Language milestones included vocalising meaningless monosyllables at six months. Fine motor milestones involved bidextrous grasp at six months, unidextrous age at nine months, and immature pincer grasp at 11 months. The child did not achieve social milestones of feeding herself, and stranger anxiety developed at eight months of age. There was a history of two hospital admissions for pneumonia at the ages of eight and 10 months, respectively. The second episode required a 12-day stay in the intensive care unit.

On examination, the patient weighed 4.8 kg, measured a length of 66 cm, and had a body surface area of 0.2918 m^2. The patient exhibited upward-slanted eyes, a flattened facial profile, a small mouth with a protruding tongue, a flat nasal bridge, and a unilateral simian crease (refer to [Table/Fig-1,2]). The pulse rate was 110 beats per minute, blood pressure was 62/44 mmHg, respiratory rate was 26 per minute, and room air saturation was 97% in all four extremities. During systemic cardiovascular examination, no remarkable findings were observed upon inspection and palpation. However, a diastolic murmur was detected upon auscultation, which was loudest in the mitral area. The murmur was accompanied by a split-second heart sound (S2) with a loud accentuated pulmonary component (P2). The musculoskeletal system examination revealed generalised hypotonia and laxity in the hip and knee joints. Clinical examination of the respiratory system and abdomen revealed normal findings. An airway examination revealed macroglossia, a small mouth, and the absence of dentition.

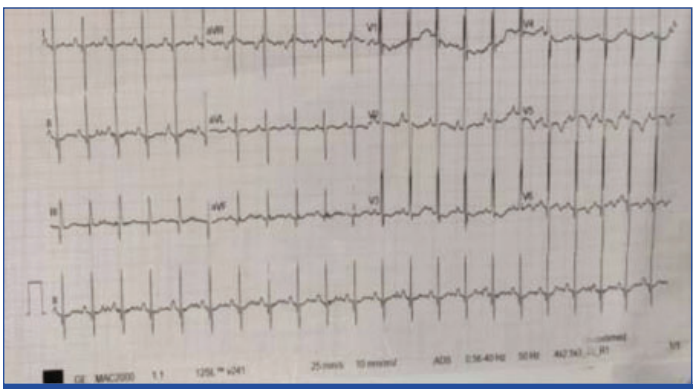


[Table/Fig-1]: Facial features showing protruded tongue.

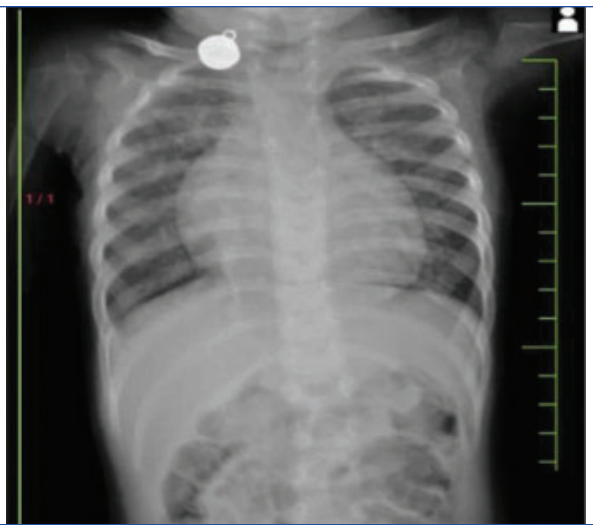


[Table/Fig-2]: Simian crease.

The electrocardiogram showed biventricular hypertrophy (refer to [Table/Fig-3]). A 2D echocardiography revealed a large inlet ventricular septal defect (7 mm), a large os atrial septal defect (9 mm), and a small patent ductus arteriosus (1 mm). All four chambers of the heart were dilated, and there was evidence of severe pulmonary artery hypertension and moderate atrioventricular regurgitation. The chest radiograph displayed increased pulmonary vascular markings with mild cardiomegaly (refer to [Table/Fig-4]). Haematological investigations, including complete blood count, renal function, liver function, coagulation profile, and TSH value (4.7 µIU/mL within the normal range of 0.465-4.68 µIU/mL), were all within normal limits. The T3 value was 1.5 ng/mL (within the range of 0.970-1.69 ng/mL), and the T4 value was 5.2 µg/dL (within the range of 5.53-11 µg/dL). At the time of presenting for cardiac surgery, the patient was in a euthyroid state as she had been taking a daily dose of thyroxine 25 micrograms since the age of 10 months. The patient was scheduled for surgery under general anaesthesia according to the



[Table/Fig-3]: Preanaesthetic electrocardiogram.



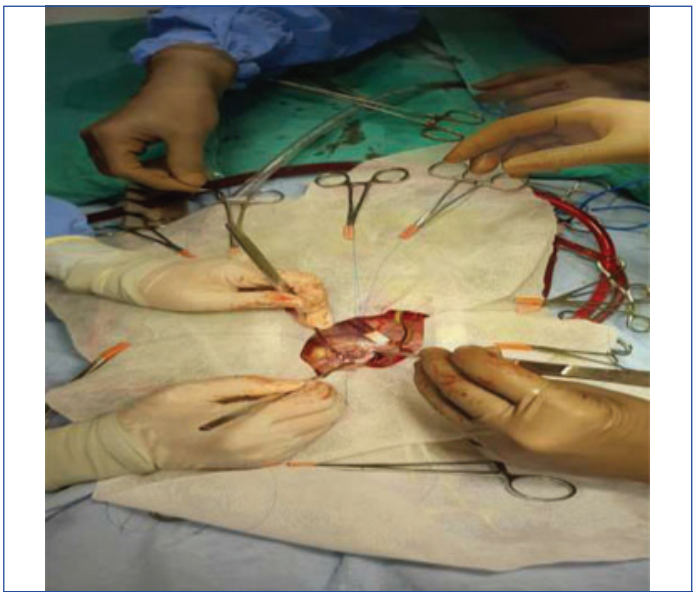
[Table/Fig-4]: Preanaesthetic chest radiograph.

American Society of Anesthesiologists (ASA) III classification. She followed the fasting guidelines and was kept nil per oral. A 24-gauge intravenous cannula was secured on the right wrist, and intravenous fluids (dextrose normal saline) were administered at a maintenance rate of 19.2 mL per hour, determined by the Holliday Segar formula [1]. Blood and blood products were appropriately cross-matched and reserved. Informed written consent was obtained from the parents. In the operating room, standard multipara monitors were attached.

The induction was performed using single vital capacity breath induction with 8% sevoflurane, 50% nitrous oxide, and 100% oxygen fresh gas flow. This was followed by the administration of intravenous midazolam (0.05 mg/kg body weight), fentanyl (0.05 mg/kg), and thiopental (5 mg/kg). The muscle relaxant vecuronium (0.1 mg/kg) was administered. Endotracheal intubation was performed using laryngoscopy with a Macintosh blade size 1. A 3.0 mm internal diameter uncuffed endotracheal tube was placed, confirmed by five-point auscultation, fogging of the endotracheal tube, and capnography. Care was taken to avoid excessive neck extension.

A 4.5 French triple-lumen catheter was inserted into the right internal jugular vein to measure central venous pressure and administer ionotropic drugs and vasodilators. Additionally, a 20-gauge arterial line was secured in the right femoral artery for invasive blood pressure monitoring, arterial blood gas analyses, and serum electrolyte assessment. The concentration of sevoflurane was adjusted according to the patient's haemodynamic parameters. Normothermia and normocapnia were maintained throughout the procedure. Anaesthesia was sustained using a combination of oxygen, nitrous oxide, sevoflurane, intravenous fentanyl, and the muscle relaxant vecuronium.

After sternotomy, partial excision of the thymus, and pericardiotomy, 300 IU of heparin were administered for anticoagulation. Ascending aorta and bi-caval cannulation were performed, initiating Cardiopulmonary Bypass (CBP). The patient was cooled to 32°C (nasopharyngeal temperature), and del Nido cardioplegia was delivered. Right atriotomy revealed a large os Atrial Septal Defect (ASD) with all pulmonary veins draining into the left atrium, a single atrioventricular valve, a large Ventricular Septal Defect (VSD), and a small Patent Ductus Arteriosus (PDA). The PDA was tied off, the ASD was closed with a pericardial patch, the anterior mitral valve leaflet cleft and the VSD were closed, and the atrioventricular valves were tied over a pericardial patch (refer to [Table/Fig-5]). The adequacy of perfusion during CBP was evaluated by monitoring arterial pressure, urine output, and arterial blood gases (refer to [Table/Fig-6]).



[Table/Fig-5]: Pericardial patch.

Parameters	Measured 37° Celsius
pH	7.36 (7.35-7.45)
pCO_2	41 mmHg (75-100)
pO_2	129 mmHg (35-45)
Na^+	135 mmol/L (135-145)
K^+	4.4 mmol/L (3.5-4.5)
Ca^{++}	1.08 mmol/L (1.02-1.04)
Glucose	165 mg/dL (70-100)
Lactate	2.1 mmol/L (<2)
Haematocrit	25%
Derived parameters	
Ca^{++} at PH-7.4	1.06 mmol/L
HCO_3^-	23.2 mmol/L (22-26)
Base excess	-2.1 mmol/L
sO_2	99%
Haemoglobin	7.8 g/dL

[Table/Fig-6]: Arterial blood gas analysis report on CPB.

Following the completion of the repair, the patient's body temperature was gradually increased to 37°C by rewarming. Milrinone, dobutamine, and nitroglycerine were used during the weaning process. De-airing maneuvers were conducted under transesophageal echocardiography monitoring to prevent air embolism. The cross-clamp was removed to restore coronary perfusion. Subsequently, the patient was successfully weaned off CBP, and protamine was administered to neutralise residual heparin. Paracetamol (15 mg/kg) and ondansetron (0.1 mg/kg) were administered before skin closure. After completion of the surgery, the patient was shifted to the ICU with the endotracheal tube in situ and electively ventilated on pressure-regulated volume control ventilator mode. The patient was successfully extubated after one hour. Dexmedetomidine infusion at a dose of 0.5 µg/kg per hour was used for sedation and pain relief on day zero and the first postoperative days. The rest of the postoperative period was uneventful, and the patient was discharged on the eighth postoperative day.

DISCUSSION

The DS is a genetic disorder that affects multiple body systems due to an additional copy of chromosome 21 in the karyotype, which can occur in three ways: simple trisomy, mosaic trisomy, and translocation trisomy [2]. The incidence of DS is estimated to be one in 1000 to one in 1100 live births worldwide [3]. Upto 60% of individuals with DS have Congenital Heart Disease (CHD), with 12% experiencing clinical symptoms [4,5]. Consequently, there is a high likelihood of encountering DS patients with cardiac anomalies, making paediatric cardiac surgery a complex process which is further complicated by the presence of DS [6]. Since DS patients have multiple system involvement, those with cardiac defects will also have various other conditions. Therefore, a systematic approach and proper knowledge regarding associated risks, needs, and complications are required for appropriate anaesthetic management. The most commonly seen cardiac defect associated with DS is CAVSD (40%). The second most common is VSD, followed by PDA, and then ASD [7].

Cardiovascular system: The DS patients commonly have acyanotic cardiovascular defects. There is a greater propensity for these patients to develop Pulmonary Hypertension (PH) earlier than children without DS. PH can occur secondary to the cardiac defect or due to chronic airway obstruction and intrinsic anomalies in the pulmonary vasculature. The cardiac cause is the increased pulmonary blood flow via the left-to-right shunt. PH may result in a postoperative crisis. Therefore, it is prudent to set an anaesthetic goal of avoiding exacerbations of PH by preventing elevations in peripheral vascular resistance and avoiding high systemic vascular resistance to prevent Eisenmengerisation of the left to right cardiac lesion [7]. In this study, this was achieved in the patient by using inodilators (milrinone and dobutamine) and pulmonary dilators (nitroglycerin). Fentanyl, midazolam, and muscle relaxant-based anaesthesia are suitable for patients with PH, and nitric oxide is beneficial in reducing PH [8].

Airway and respiratory system: A detailed history should be taken to identify symptoms of sleep apnea, such as snoring, excessive daytime sleepiness, and restless sleep, as central nervous system depressant drugs can exacerbate upper airway obstruction. A history of croup, stridor, wheezing, pneumonia, or cyanosis may indicate subglottic or tracheal stenosis [9]. A meticulous evaluation of the head, neck, and oropharynx is paramount to predict a difficult airway [9]. Acute infections in the preoperative period increase the likelihood of developing acute respiratory distress syndrome. Lower airway anomalies, such as stenotic lesions, branching anomalies, and tracheoesophageal fistulae, are linked to congenital cardiac anomalies and require examination [9]. The combination of a large tongue, small mouth, and macrognathia might result in difficulty maintaining the airway, necessitating the use of an oropharyngeal airway. Assistance may be provided with gentle jaw thrust, considering the probable laxity of the temporomandibular joint. A smaller endotracheal tube size than what would typically be chosen based on the patient's age should be used due to the possibility of airway anomalies, shorter stature, and narrower airways. It is important to have a wide range of endotracheal tubes, emergency airway modalities such as a flexible bougie, fibreoptic equipment, laryngeal mask airways, and surgical airway equipment available. Opting for an "awake" extubation approach is preferable when planning for extubation [9].

Spinal disease: There is a notable prevalence of Atlantoaxial Instability (AAI) in individuals with DS [10]. Approximately 10 to 20% of patients are asymptomatic, while 15% experience symptoms [11]. Causative factors include laxity of the transverse ligament between the anterior arch of the atlas and the odontoid peg, bony anomalies of the odontoid process, and odontoid impingement. A correlation has been observed between AAI and laxity of other joints, such as the fingers, thumbs, elbows, and knees [11]. Precautions should be taken during anaesthetic management to avoid forceful neck flexion or extension during laryngoscopy, and care should be taken during intraoperative positioning [11].

Other organ systems: Gastrointestinal system involvement in DS includes gastroesophageal reflux, which increases the risk of perioperative aspiration. This highlights the importance of appropriate aspiration prophylaxis [11]. DS patients have a relative immune deficiency, making them more susceptible to infections [12]. Strict aseptic practices should be followed in the operating room and postoperatively in the ICU, and prophylactic antibiotics should be administered prior to induction.

Endocrine system: There is a well-established link between DS and endocrine diseases, particularly thyroid disorders. Hypothyroidism is seen in 15 to 20% of patients [13]. Hypothyroidism increases the risk of hypothermia and delays recovery [13]. Preoperative assessment should include evaluation of TSH values.

Induction and maintenance of anaesthesia: Sevoflurane is the preferred agent for induction, but it can cause bradycardia [9]. This can be managed with anticholinergic premedication. Accurate dosing of muscle relaxants and close monitoring of neuromuscular blockade are important due to the general hypotonia seen in DS patients. Individuals with DS may exhibit a lower minimum alveolar concentration due to reduced central catecholamine concentrations.

Postoperative pain: Pain management can be challenging in DS patients due to their reduced verbal and behavioral expressions, as well as, the difficulty for parents in recognising their child's pain levels. These patients may require higher doses of pain medication and can be challenging to sedate. Therefore, careful planning and supervision are necessary for postoperative pain management [14].

Atropine: There have been reports of an exaggerated increase in heart rate as a response to parenteral atropine in DS patients, but the consensus is that these reports are erroneous. Atropine or glycopyrrolate can be used to counter the effect of lowered catecholamine levels and act as an antisialagogue [14].

CONCLUSION(S)

The authors present a case report of a paediatric patient with DS and multiple cardiac defects accompanied by PH. A thorough understanding of the pathophysiology, hemodynamics, and potential complications of the complex cardiac lesion is crucial for a successful perioperative course in the hospital. This case emphasises the significance of proper knowledge, appropriate planning, and treatment of congenital heart defects in children with DS. PH was effectively managed while adhering to all standard protocols, ultimately leading to the successful correction of the cardiac defects.

Acknowledgement

The authors would like to express their gratitude to the participants for their valuable contribution to this study.

REFERENCES

[1] Mathew A, Rai E. Pediatric perioperative fluid management. Saudi J Anaesth. 2021;15(4):435-40.
[2] Kadakol GS, Bagoji I, Patil SV, Bulagouda RS. Cytogenetic analysis of down syndrome. Int J Clin Biomed Res. 2019;5(1):37-40.
[3] Nations U. United Nations. United Nations; [cited 2023 Jun 2]. World Down Syndrome Day. Available from: https://www.un.org/en/observances/down-syndrome-day.
[4] Delany DR, Gaydos SS, Romeo DA, Henderson HT, Fogg KL, McKeta AS. Down syndrome and congenital heart disease: Perioperative planning and management. J Congenit Cardiol. 2021;5(1):7.
[5] Santamaria LB, Di Paola C, Mafrica F, Fodale V. Preanesthetic evaluation and assessment of children with down's syndrome. Sci World J. 2007;7:242-51.
[6] Matsuishi Y, Sakuramoto H, Hoshino H, Shimojo N, Enomoto Y, Mathis BJ, et al. Down syndrome reduces the sedative effect of midazolam in pediatric cardiovascular surgical patients. Sci Rep. 2020;10(1):2148.
[7] Meitzner MC, Skurnowicz JA. Anesthetic considerations for patients with Down syndrome. AANA J. 2005;73(2):103-07.
[8] Price LC, Martinez G, Brame A, Pickworth T, Samaranayake C, Alexander D, et al. Perioperative management of patients with pulmonary hypertension undergoing non-cardiothoracic, non-obstetric surgery: A systematic review and expert consensus statement. Br J Anaesth. 2021;126(4):774-90.
[9] Steward DJ. Anesthesia considerations in children with down syndrome. Semin Anesth Perioper Med Pain. 2006;25(3):136-41.
[10] Santha N, Upadya M, Vishwanatham S. Anaesthetic management of a case of down's syndrome with achalasia cardia. J Clin Diagn Res. 2016;10(10):UD03-05.
[11] Allt JE, Howell CJ. Down's syndrome. BJA CEPD Rev. 2003;3(3):83-86.
[12] Haque MI. Anesthesia for patients with Down syndrome. Int J Health Sci Res. 2019;9(5):381-85.
[13] Amr NH. Thyroid disorders in subjects with Down syndrome: An update. Acta Bio Medica Atenei Parm. 2018;89(1):132-39.
[14] Bhattarai B, Kulkarni A, Kalingarayar S, Upadya M. Anesthetic management of a child with Down's Syndrome having atlanto axial instability. J Nepal Med Assoc [Internet]. 2009 Jan 1 [cited 2023 May 28];48(173). Available from: https://www.jnma.com.np/jnma/index.php/jnma/article/view/208.

PARTICULARS OF CONTRIBUTORS:
1. Postgraduate, Depatment of Anaesthesia, Jawaharlal Nehru Medical College, Dattta Meghe Institute of Higher Education and Research, Sawangi (Meghe), Wardha, Maharashtra, India.
2. Senior Resident, Depatment of Anaesthesia, Jawaharlal Nehru Medical College, Dattta Meghe Institute of Higher Education and Research, Sawangi (Meghe), Wardha, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Shyamolima Bhuyan,
Postgraduate, Department of Anaesthesiology, Jawaharlal Nehru Medical College, Dattta Meghe Institute of Higher Education and Research, Sawangi (Meghe), Wardha-442004, Maharashtra, India,
E-mail: shybhuyan@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Jun 15, 2023
- Manual Googling: Jul 25, 2023
- iThenticate Software: Aug 12, 2023 (6%)

ETYMOLOGY: Author Origin

EMENDATIONS: 7

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Jun 13, 2023**
Date of Peer Review: **Jul 20, 2023**
Date of Acceptance: **Aug 14, 2023**
Date of Publishing: **Sep 01, 2023**