AIRWAY AND ANESTHETIC MANAGEMENT IN ROBINOW SYNDROME PATIENT WITH SEVERE AORTIC STENOSIS FOR DENTAL SURGERY : A CASE REPORT

Ardy Wibowo Cahyono[1*], Adinda Putra Pradhana[2], Tjokorda Gde Agung Senapathi[3]
Anesthesiology and Intensive Care Departement, Medical Faculty of Udayana *University*/Prof Ngoerah *General Hospital*, Denpasar, Bali, Indonesia[1,2,3]
[*]*Corresponding Author* : ardbius2022@gmail.com

ABSTRAK

Robinow Syndrome (RS) adalah kelainan genetik langka yang ditandai dengan fitur kraniofasial dan abnormalitas skeletal yang khas, sering disertai dengan kelainan jantung, yang mempersulit manajemen anestesi. Pasien dengan RS sering mengalami tantangan signifikan dalam manajemen jalan napas karena karakteristik wajah dan skeletal, yang membuat manajemen jalan napas menjadi kompleks. Anomali vertebral juga mempersulit pemberian anestesi neuraksial, dan pemilihan agen induksi harus mempertimbangkan risiko jantung. Laporan ini membahas manajemen jalan napas dan anestesi yang berhasil pada seorang pasien RS berusia 13 tahun yang menjalani ekstraksi gigi akibat kerusakan gigi parah dan stenosis aorta. Pemeriksaan fisik pasien menunjukkan Mallampati IV dan Cormack-Lehane IV pada jalan napas, sehingga diperlukan teknik intubasi khusus seperti penggunaan gum elastic bougie untuk akses jalan napas yang berhasil. Fitur kraniofasial dan skeletal yang terkait dengan RS menimbulkan tantangan dalam manajemen perioperatif, sering kali membutuhkan pembedahan. Intubasi yang efektif pada pasien RS membutuhkan evaluasi praoperatif yang cermat, terutama pada kasus dengan micrognathia dan asimetri wajah. Evaluasi praoperatif yang komprehensif, termasuk penilaian fungsi jantung, ginjal, dan pernapasan, sangat penting untuk mengurangi komplikasi dan meningkatkan hasil. Manajemen anestesi harus memperhitungkan kesulitan jalan napas dan masalah jantung yang unik pada RS, dengan kesiapan menghadapi potensi tantangan intraoperatif. Kasus ini menyoroti pentingnya perencanaan yang teliti dan teknik khusus dalam mengelola anestesi pada pasien RS, khususnya mereka dengan kondisi jantung yang mempersulit.

Kata kunci : intubasi, manjemen anestesi, *robinow syndrome*, stenosis katup aorta, tantangan airway

ABSTRACT

Robinow Syndrome (RS) is a rare genetic disorder marked by distinctive craniofacial and skeletal abnormalities, often accompanied by cardiac anomalies, which complicate anesthetic management. RS patients frequently present significant airway challenges due to facial and skeletal features, making airway management complex. Vertebral anomalies also complicate neuraxial anesthesia, and the selection of induction agents must consider cardiac risks. This report discusses successful airway and anesthetic management in a 13-year-old RS patient undergoing multiple dental extractions due to severe dental decay and aortic stenosis from a restricted right coronary cusp. The patient's physical examination revealed a Mallampati IV and Cormack-Lehane IV airway, necessitating specialized intubation techniques such as a gum elastic bougie for successful airway access. RS-associated craniofacial and skeletal features pose challenges in perioperative management, often requiring surgery. Effective intubation in RS demands careful preoperative evaluation, especially in cases of micrognathia and facial asymmetry. Comprehensive preoperative assessments—including cardiac, renal, and respiratory evaluations—are crucial to reducing complications and improving outcomes. Anesthetic management must account for airway difficulties and cardiac concerns unique to RS, with surgeons prepared for potential intraoperative challenges. This case highlights the importance of meticulous planning and specialized techniques in managing anesthesia for RS patients, particularly those with complicating cardiac conditions.

Keywords *: anesthetic management, aortic stenosis, difficult airway, intubation, robinow syndrome*

INTRODUCTION

Robinow Syndrome (RS) is a rare genetic disorder characterized by distinct facial features, limb shortening, and genital abnormalities. First described by Dr. Meinhard Robinow and colleagues in 1969, RS has since been recognized in two main forms: autosomal dominant and autosomal recessive. The autosomal dominant form is often milder, while the autosomal recessive form presents more severe phenotypic manifestations. Both forms are linked to mutations in different genes, including ROR2 for the recessive type and WNT5A for the dominant type, underscoring the genetic heterogeneity of the condition (Keçeci et al., 2022). The incidence of RS is difficult to pinpoint precisely due to its rarity, with estimates suggesting an occurrence of approximately 1 in 500,000 to 1 in 1,000,000 live births. This rarity means that many healthcare professionals, including anesthesiologists, may encounter it infrequently, potentially complicating the perioperative management due to unfamiliarity with the syndrome's unique clinical challenges. Patients with RS exhibit a wide range of anomalies, including midface hypoplasia, a short stature, mesomelic limb shortening, and vertebral segmentation defects, which can significantly impact anesthetic planning and delivery (Goitia-Cárdenas et al., 2023).

From an anesthetic perspective, managing patients with RS presents multiple considerations. The craniofacial abnormalities typical of RS, such as midface hypoplasia and a prominent forehead, can complicate airway management. These features may lead to difficulties in mask ventilation and intubation, necessitating careful preoperative assessment and planning for potential difficult airway scenarios. Additionally, vertebral anomalies may influence the choice and administration of neuraxial anesthesia, making regional anesthesia techniques potentially more challenging and requiring a thorough understanding of the patient's spinal anatomy (Chandar et al., 2023; Depal et al., 2020). Cardiac anomalies, which are present in a significant proportion of patients with RS, add another layer of complexity to anesthetic management. Conditions such as ventricular septal defects, atrial septal defects, and tetralogy of Fallot necessitate a detailed preoperative cardiac evaluation and careful intraoperative monitoring. The presence of these cardiac issues can affect anesthetic drug selection and fluid management strategies, highlighting the need for a tailored anesthetic plan that minimizes cardiovascular risk (Chandar et al., 2023; Goitia-Cárdenas et al., 2023).

Lastly, the potential for genitourinary anomalies and skeletal abnormalities in RS patients requires an interdisciplinary approach involving surgeons, anesthesiologists, and other specialists. The preoperative planning must take into account the specific surgical procedure, the patient's overall health status, and the unique anatomical considerations posed by RS. Close postoperative monitoring is essential to promptly address any complications arising from the syndrome's characteristic anomalies and the surgical intervention itself (Gerber et al., 2021). In summary, the anesthetic management of patients with RS demands a comprehensive understanding of the syndrome's diverse manifestations and potential complications. A meticulous preoperative assessment, strategic planning for airway management, vigilant intraoperative monitoring, and a collaborative, interdisciplinary approach are pivotal in ensuring the safe and effective care of these patients. This case report aims to detail the perioperative considerations and anesthetic management strategies employed in a patient with RS, contributing valuable insights to the limited body of literature on this rare condition.

CASE REPORT

A 13-year-old male patient presented to our hospital with multiple dental caries/cavities and residual teeth. This condition resulted in intermittent pain and eating difficulties. From history taking, the patient had been diagnosed with Robinow Syndrome (RS) at the age of 9.

He presented with severe aortic stenosis (AS) due to a restricted right coronary cusp (RCC). The patient was initially scheduled for aortic valve repair surgery at a private hospital. His dental conditions did not allow him to undergo the cardiac surgery, therefore he was scheduled to have a multiple gangrene radices extraction. The patient has a history of cardiac catherization on January 23rd 2020 at our hospital. He is taking several heart medications daily after the catheterization, such as Propranolol 3 x 7.5 mg and Spironolactone 2 x 12.5 mg orally. Additionally, history of hypertension, diabetes, asthma, or other systemic diseases, and food or drug allergies were excluded. The patient is able to engage in moderate to vigorous physical activity without experiencing shortness of breath or chest pain.

Anthropometric measurements showed the patient's weight was 23 kg, height was 123 cm, and body mass index (BMI) was 15.2 kg/m^2. Based on the measurement, the patient was concluded to have moderate protein energy malnutrition (PEM). General examination was done, with numeric rating score (NRS) for pain was 0 out of 10, the Ebenhart score was 2 out of 4, and his metabolic equivalent of tasks (METs) was 5-6. The patient appeared to be compos mentis with a Glasgow Coma Scale (GCS) score of E4V5M6. The facial features were peculiar to RS presenting with midface hypoplasia (Figure 1). During the physical examination, it was found that the patient had a palpable pulse, warm extremities, a heart rate of 75 beats per minute, and a blood pressure of 110/70 mmHg. Her capillary refill period was less than two seconds, and she had a respiratory rate of 20 breaths per minute. The patient's temperature was 36.5°C, and her oxygen saturation level was 98% without oxygen supplementation. Upon examination of the eyes, it was determined that the sclera was normal and there was no conjunctival anemia. A chest examination revealed no palpable tumors and an asymmetry check showed no intercostal retraction or distortion. From auscultation, mumurs were present. Examination of the abdomen revealed normal skin turgor and no venous dilatation. There were no signs of clubbing of the extremities, edema, or dampness. The musculoskeletal examination indicated normal flexion and deflexion of the neck, Mallampati IV, Cormack-Lehane IV, macroglossia (+), teeth were not intact, even though there was no loose teeth and dentures, there were slightly sharp teeth that can complicate the intubating procedure (Figure 2A). Brachydactyly was also found on the extremities (Figure 2B).

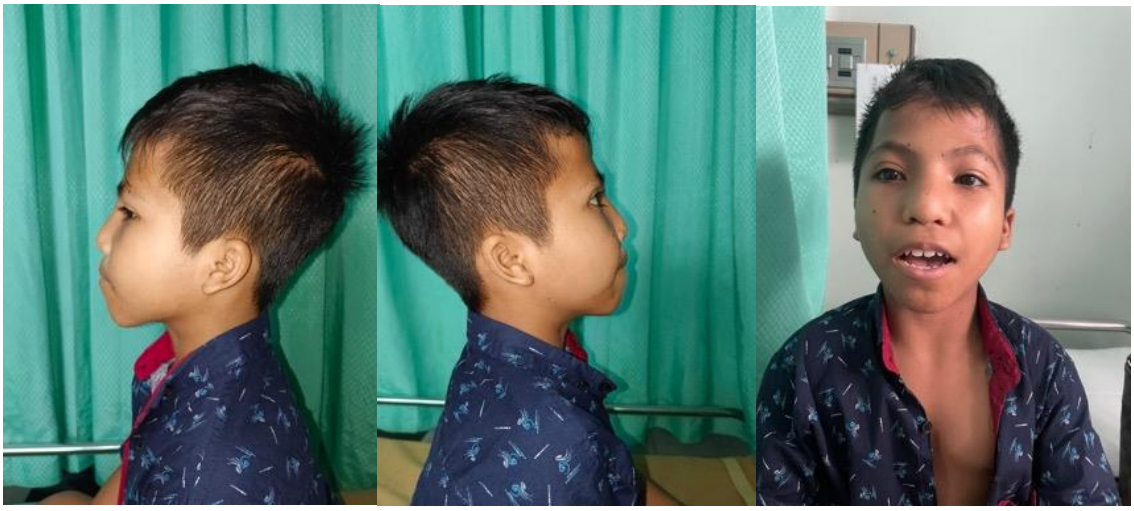


Figure 1. Facial Profile of The Patient

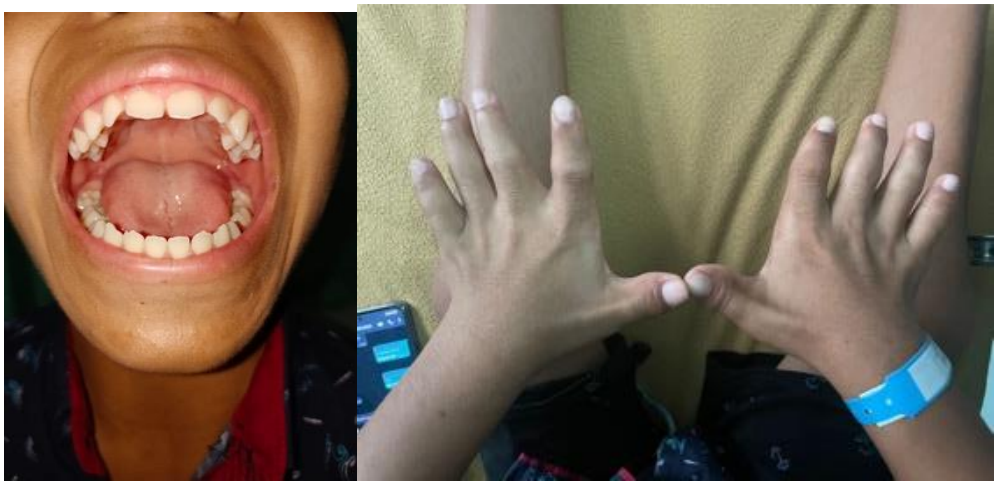


Figure 2. Facial Profile of The Patient

Table 1. Laboratory Examination

October 9th 2023	**Result**	**Normal Value**
WBC	7.01 x 10^3 /uL	4.1 - 11 x 10^3 /uL
Hb	14 g/dL	12 – 16 g/dL
Hematocrit	39 %	36 – 46 %
Platelet	264 x 10^3 /uL	140 – 440 x 10^3 /uL
MCV	83.7 fL	80 – 100 fL
MCH	30 pg	26 – 34 pg
MCHC	35.9 g/dL	31 – 36 g/dL
October 9th 2023	**Result**	**Normal Value**
SGOT	29 U/L	5 - 34 U/L
SGPT	7 U/L	<55 U/L
BUN	13.3 mg/dL	7 – 18.7 mg/dL
Creatinine	0.63 mg/dL	0.57 – 1.11 mg/dL
Potassium (K)	3.73 mmol/L	3.5 – 5.1 mmol/L
Sodium (Na)	143 mmol/L	136 -145 mmol/L
Chloride (Cl)	110 mmol/L	94 – 110 mmol/L
Calcium (Ca)	9.1 mg/dL	9.2 – 11 mg/dL
PTT	11 s	10.8 – 14.4
aPTT	35.7 s	24-36
INR	0.96	0.9 – 1.1

Other supporting examination was also done to the patient. Thorax x-ray from anterior-posterior approach (10/09/2023) showed CTR of 55% with left atrium enlargement (LAE) and left vetrivular hypertrophy (LVH) (Figure 3). Unerupted teeth (no. 35 and 48) and multiple carious teeth were also found from panoramic examination (10/05/2023), requiring extractions and restorations (Figure 4). Echocardiography (03/03/2023) was also done to the patient which showed minimal release of severe subvalvar dan valvular aortic stenosis with tyshak ballon no 12x20 mm (01/23/2020) with mild aortic regurgitation (AR), mild pulmonic regurgitation (PR), and LVH. To have a more detailed and clear picture, transesophageal echocardiogram (TEE) was conducted on April 4th 2023 which indicated severe AS caused by restricted RCC caused by suspected syndrome related, Mild AR, Mild mitral regurgitation (MR), Mild tricuspid regurgitation (TR), low probability of pulmonary hypertension (PH) with left ventricle (LV) concentric hypertrophy, Normal LV and right ventricle (RV) systolic function, Grade I LV diastolic dysfunction, ejection fraction of 79.4%, tricuspid annular plane systolic excursion (TAPSE) of 24mm, and estimated right atrial pressure (eRAP) of 3 mmHg.

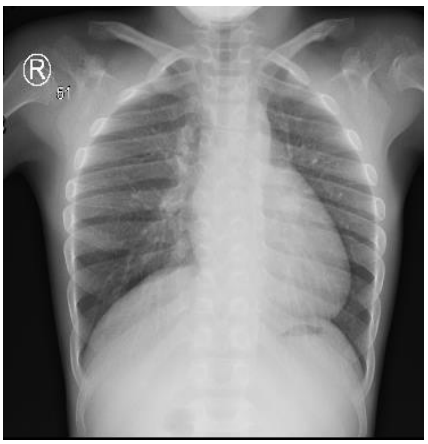


Figure 3. Thorax X-ray from Anterior-Posterior Approach

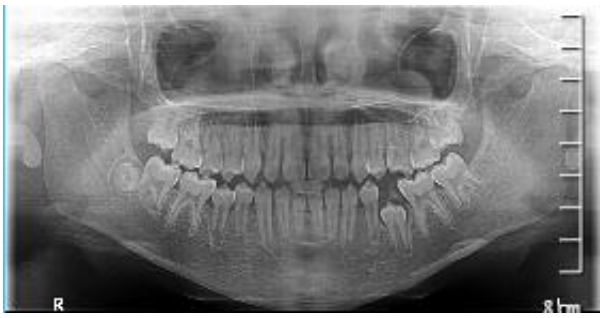
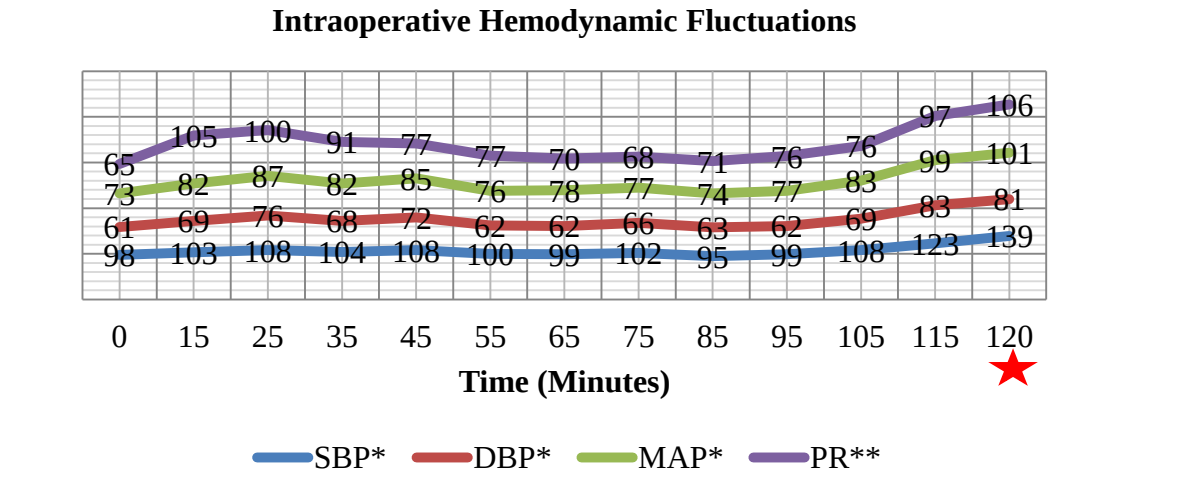


Figure 4. Panoramic Examination

Preparation for the procedure includes a thorough medical history and physical examination, as well as additional diagnostic tests. The patient was scheduled for elective multiple gangrene radix extraction surgery with ASA III based on physical, laboratory, and other supporting examinations. The patient' family had been informed about the general anesthesia procedure and had provided informed consent. The family had also been informed about the patient's condition and the potential risks associated with anesthesia and post-operative care in the pediatric intensive care unit (PICU). The patient was then planned for surgery with general anesthesia and endotracheal tube inserted (GA-OTT). The patient was positioned in supine position.

Table 2. Result

Premedication	**Ketamine 5 mg IV, Midazolam 1 mg IV**
Analgesic	Fentanyl 50 mcg IV
5"	Preoxygenation with 100% O2
3" Induction	Propofol 25 mg IV
1" Muscle Relaxant	Atracurium 15 mg IV
0"	Sellick maneuver was performed to facilitate the intubation process. Due to macroglossia, finding of Cormack-Lehane IV, and Mallammpati IV, patient was difficult to be intubated (difficult airway). Bougie was then used. The Bougie was inserted into the glottis during direct laryngoscopy, facilitating insertion of the ETT into the trachea. Subsequently, Atracurium at 15 mg IV was given, and the patient was intubated with a cuffed ETT no. 6, positioned at a lip level of 19 cm. Confirmation of the ETT location and bilaterally symmetrical breath sounds were obtained.
Maintenance	The patient received oxygen-compressed air, Sevoflurane, and intermittent Fentanyl at 0.25 – 0.5 mcg/kgBW IV every 45 – 60 minutes.
Other medication	Phenylephrine titration on stand by but was not used at the end.



: After intubation; : End of Surgery

SBP : Systolic Blood Pressure, DBP : Diastolic Blood Pressure, MAP : Mean Arterial Pressure, PR : Peripheral Resistance.

Figure 5. Intraoperative Hemodynamic Fluctuations

DISCUSSION

Robinow syndrome (RS) is a rare genetic disorder that manifests in either autosomal recessive or autosomal dominant forms. Among these two overlapping types, the recessive form typically displays more pronounced skeletal abnormalities. The genetic basis for the autosomal recessive form has been pinpointed to the long arm of chromosome 9, implicating the ROR2 tyrosine kinase as a key enzyme in its development. ROR is part of the tyrosine kinase family located at the cell surface, and it plays a role in the formation of the skeleton, the cardiovascular and the genital system. Clusters of recessive RS have been documented in regions with high rates of consanguinity, such as Oman, Turkey, and Chechnya (Keçeci et al., 2022; Soman & Lingappa, 2015).

RS is a complex developmental disorder that affects multiple systems. It is characterized by limb shortening (mesomelic or acromesomelic), facial anomalies including hypertelorism, midfacial underdevelopment, a short and upturned nose, a broad forehead and nasal root, a ‘tented’ upper lip, cleft lips, and an appearance of pseudo-exophthalmus due to lower eyelid deficiencies. Patients often exhibit spinal deformities, underdeveloped genitalia, kidney disorders, and congenital heart defects. These distinct anomalies and potential related conditions require careful consideration during the perioperative management of patients with RS (Soman & Lingappa, 2015).

Patients with RS often require surgical interventions to address vertebral, orofacial, and dental abnormalities . The unique facial and dental deformities, along with their short neck and overall small body structure, can present significant challenges in ensuring adequate ventilation and maintaining airway control with a mask. Given these facial abnormalities, careful planning is essential to prepare for the possibility of difficult intubation. Additionally, congenital airway anomalies, such as extrathoracic and functional airway obstructions, may be present in these patients, sometimes necessitating the use of a tracheostomy (Chandar et al., 2023; Depal et al., 2020; Saraç et al., 2023).

In our case, due to dental and cardiac condition, a surgical operation was planned and undertaken. The patient has midfacial hypoplasia and macroglossia, accompanied with big and

sharp shaped teeth which made the intubation process more difficult. The patient had Mallampati IV and Cormack-Lehane IV from the preoperative examination. Difficult intubation is a challenging situation, especially in very young children, as desaturation can occur very quickly. In this case, the use of a laryngeal mask airway (LMA) was not feasible because the patient was scheduled for dental surgery, which necessitated shared access to the airway. During dental procedures, the surgical field often requires unobstructed access to the mouth and upper airway, making the placement of an LMA impractical. The LMA's presence would obstruct the surgeon's view and impede the surgical instruments, creating a significant challenge in performing the extractions of multiple gangrenous dental roots. Therefore, an alternative approach to airway management had to be considered to accommodate the surgical requirements and ensure patient safety.

In order to intubate the patient, a Bougie was used. Given the patient's need for unobstructed access to the oral cavity during dental surgery, the bougie provided an effective solution. The bougie, a flexible, thin device, was instrumental in guiding the endotracheal tube through the narrow and complex airway without occupying the oral cavity. By using the bougie, the anesthesiologist was able to secure the airway efficiently while allowing the surgeon full access to the patient's mouth for the necessary dental procedures.To enhance the laryngoscopic view, external manipulation of the larynx was employed. If the laryngeal view remained limited at grade III or IV after manipulation, a paraglossal approach was used to facilitate orotracheal intubation with the assistance of a gum elastic bougie. This approach, as described by Henderson, involves inserting a size 0 or 1 Miller blade into the right corner of the mouth and guiding it along the groove between the tongue and the tonsils. The technique requires applying leftward and anterior pressure to move the tongue to the left of the laryngoscope, maintaining its position away from the blade. The blade is then advanced until the epiglottis is located, and its tip is positioned behind the epiglottis. By lifting the blade anteriorly, the epiglottis is elevated to reveal the glottis (Bonnette et al., 2021; Semjen et al., 2008).

Although this method generally provides a clear view of the glottis, it can be more challenging in infants due to the reduced space, often causing the anesthetist's right hand to block the view during intubation attempts. Therefore, a gum elastic bougie was used, as its greater length compared to a stylet or tracheal tube allows for easier manipulation without obstructing the view of the glottis. The proper placement of the tracheal tube was verified through capnography and chest auscultation. Two intubation attempts were permitted; if both were unsuccessful, fiberoptic intubation was then performed. In our case, intubation attempt was succeeded after conducting Bougie-assisted intubation (Bonnette et al., 2021).

Atrial septal defect, aortic coarctation, tetralogy of Fallot, severe pulmonary stenosis or atresia and tricuspid atresia are congenital heart defects being reported with a prevalence of 15% in RS patients. Due to pulmonary and cardiac complications, nearly 5-10% of the cases die in early childhood. In our case, tachycardia was not present but cardiac auscultation, ECG and echo showed several findings, particularly indicating valvular disease, highlighting severe aortic stenosis. Severe aortic stenosis can occur in children with RS due to underlying genetic mutations affecting embryonic development, particularly involving genes like ROR2 and WNT5A. These mutations disrupt normal heart development, leading to structural abnormalities such as a malformed or narrowed aortic valve. The presence of severe aortic stenosis in these patients reflects the complex interplay of genetic factors impacting cardiovascular development, necessitating careful monitoring and management to address potential complications related to cardiac function (Keçeci et al., 2022).

The patient's severe aortic stenosis necessitated careful selection of induction and anesthetic agents. Aortic stenosis is a critical condition that affects hemodynamic stability, making it imperative to avoid drugs that could exacerbate the patient's cardiac condition. Agents that significantly lower blood pressure or alter heart rate can be particularly dangerous

in patients with this type of cardiac disease, especially induction agents and agents used to maintain the sedation throughout the procedure. Our patient received Propofol for induction. Propofol is a commonly used induction agent known for its rapid onset and smooth sedation. However, in patients with severe aortic stenosis, it is administered cautiously due to its potential to cause hypotension. In this patient, a reduced dose of 25 mg was used to induce anesthesia gently, minimizing the risk of hemodynamic instability. By using a lower dose, the anesthesiologist could achieve the necessary depth of anesthesia for intubation while avoiding significant drops in blood pressure. Propofol's quick action also allows for precise control over the level of sedation, which is beneficial in managing patients with complex cardiac conditions (Oktavia, 2020).

Alongside the careful selection of induction agents to maintain hemodynamic stability, the preparation of phenylephrine for titration was a critical preventive measure in managing anesthesia for this patient. This approach provided a rapid and effective means to counteract hypotension, ensuring the patient's safety and stability during the dental surgery. This readiness to manage hemodynamic changes reflects a comprehensive and vigilant anesthetic plan for this patient (Oktavia, 2020). For patients with RS who exhibit renal cysts or obstructive uropathies along with genital hypoplasia, it is crucial to conduct preoperative evaluations of baseline blood urea nitrogen, creatinine, and electrolytes. If these tests reveal any abnormalities, further diagnostic procedures, such as ultrasonography, should be performed to investigate the underlying issues (Chandar et al., 2023; Depal et al., 2020; Yeter et al., 2017). In our particular case, the laboratory results were within normal ranges. RS patients may experience compromised chest wall movements and respiratory function due to conditions such as pectus excavatum, scoliosis, and rib anomalies. Severe cases can impair their ability to cough and clear secretions, heightening the risk of respiratory infections (Soman & Lingappa, 2015; Yeter et al., 2017). In our patient, tachypnea and wheezing were not present. A posterior-anterior chest x-ray revealed cardiomegaly without lung abnormalities.

In general, patients with RS typically exhibit normal mental and motor development. However, about 20% of patients may experience mental or developmental delays. While macrocephaly is commonly seen in RS, it does not contribute to an increased risk of developmental delays. Structural anomalies of the central nervous system are rare, with only one reported case of cortical dysplasia (Yeter et al., 2017). In our patient, both mental and motor development were normal. In summary, when managing anesthesia in RS patients, particular attention should be given to potential airway challenges, especially in the presence of facial asymmetry and micrognathia. Comprehensive preoperative assessments should include evaluations of the airway, as well as respiratory, renal, and cardiac functions. During surgery, it's crucial to be prepared for complications, not only from difficult airway management but also due to potential cardiac anomalies.

CONCLUSION

Robinow Syndrome (RS) is a rare genetic disorder characterized by distinct facial features, limb shortening, and genital abnormalities that may complicate anesthetic management of the patient requiring surgery. The distinct craniofacial abnormalities associated with RS, including midface hypoplasia and a prominent forehead, pose significant challenges for airway management. These anatomical features can make mask ventilation and intubation particularly difficult, necessitating meticulous preoperative evaluation and strategic planning for managing potentially complicated airway scenarios. Moreover, the vertebral anomalies commonly seen in RS can impact the selection and administration of neuraxial anesthesia, complicating regional anesthesia procedures and requiring a deep understanding of the patient's specific spinal anatomy. This case report highlights the successful management of a challenging airway

in a patient with RS using a gum elastic bougie.

Due to the distinctive craniofacial features associated with RS, such as midface hypoplasia, a short neck, and potential micrognathia, airway management can be particularly complex. In our patient, these anatomical factors contributed to a difficult intubation scenario. Despite these challenges, the use of a gum elastic bougie facilitated successful orotracheal intubation after initial attempts with direct laryngoscopy were unsuccessful. This approach proved effective in navigating the anatomical constraints posed by RS, ensuring secure airway access and minimizing the risk of complications. The bougie's flexibility and length allowed for precise guidance of the endotracheal tube without obstructing the view of the glottis, a critical advantage in the context of RS's unique anatomical considerations. Careful selection of induction agents is crucial in managing patients with associated cardiac diseases like severe aortic stenosis. Agents such as low dose Propofol was chosen in our case due to their favorable hemodynamic profiles, minimizing risks of exacerbating cardiovascular instability. Avoiding agents like Thiopental and high-dose Propofol, which can cause significant myocardial depression and hypotension, is paramount in maintaining cardiac function during induction. This tailored approach acknowledges the need for meticulous perioperative planning and emphasizes the importance of adapting anesthesia strategies to mitigate potential complications in RS patients with complex cardiac conditions.

This case underscores the importance of thorough preoperative planning and the availability of advanced airway management tools when anesthetizing patients with RS. It also highlights the utility of the gum elastic bougie as a valuable aid in managing difficult airways. The successful intubation and subsequent uneventful surgical course in this patient demonstrate that, with appropriate preparation and technique, the challenges of airway management and cardiac conditions in RS can be effectively addressed. The experience from this case can serve as a guide for anesthesiologists encountering similar scenarios, emphasizing the need for vigilance and adaptability in the face of potential airway and cardiac difficulties associated with RS.

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