

Dental management of a 6-year-old boy with ventricular septal defect and down syndrome under general anesthesia: A case report

Anak Agung Sagung Dyah Karuniadewi ¹, Dimas Surya Saputra ¹, Rosiana Dewi Prayogo ¹, Lely Indriasari ² and Sindy Cornelia Nelwan ^{1,*}

¹ Department of Pediatric Dentistry, Faculty of Dental Medicine, Universitas Airlangga, Surabaya, East Java, Indonesia.

² Dr. Saiful Anwar Regional General Hospital, Malang, East Java, Indonesia.

World Journal of Advanced Research and Reviews, 2024, 22(03), 2173–2178

Publication history: Received on 20 May 2024; revised on 28 June 2024; accepted on 30 June 2024

Article DOI: <https://doi.org/10.30574/wjarr.2024.22.3.1955>

Abstract

Introduction: Down Syndrome (DS) is a genetic disorder caused by the presence of an extra copy of chromosome 21, characterized by physical and intellectual disabilities. DS has a significant risk factor for congenital heart diseases (CHD), which are abnormalities in the structure or function of the heart present from birth. Ventricular Septal Defect (VSD) is a septal defect in the ventricular wall, where small defects are usually asymptomatic and large defects can cause shortness of breath, feeding difficulties, and poor growth.

Case History: A 6-year-old boy patient was referred to the Pediatric Dentistry Clinic at Dr. Saiful Anwar Regional General Hospital, Malang, by his pediatric cardiologist to investigate focal infection prior to heart surgery. The patient has been diagnosed with Ventricular Septal Defect (VSD) and Down Syndrome (DS). During intraoral examination, multiple caries and multiple gangrene radix were found. The patient was given prophylactic antibiotics previously and scheduled to undergo tooth fillings and extractions under general anesthesia.

Discussion: Children with VSD have an increased risk of bacterial endocarditis. Dental procedures that cause trauma to soft tissue or bone may lead to bleeding, potentially leading to transient bacteremia. Prophylactic antibiotics are used in an attempt to prevent bacterial endocarditis. Due to these considerations, it was decided to undergo comprehensive dental management under general anesthesia (GA). With a comprehensive dental management approach under GA, all necessary procedures can be completed in a single visit.

Conclusion: Dental management in children with VSD and DS under general anesthesia is a good option.

Keywords: Ventricular Septal Defect; Down Syndrome; Special Needs Child; General Anesthesia; Quality of life

1. Introduction

Down Syndrome (DS), also known as trisomy 21, is a genetic disorder caused by the presence of an extra copy of chromosome 21. This condition is characterized by physical and intellectual disabilities and is the most common chromosomal abnormality [1]. The exact cause of Down Syndrome is unknown, but it is believed to occur randomly without behavioral or environmental factors affecting its probability. However, the risk of having a baby with DS increases with maternal age, especially after 35 years. Additionally, parents who have one child with DS have a higher risk of having another child with the condition [2].

* Corresponding author: Sindy Cornelia Nelwan

There are three types of Down Syndrome: Trisomy 21, Translocation Down Syndrome, and Mosaic Down Syndrome. Trisomy 21 is the most common type, representing about 95% of cases. Individuals with Down Syndrome often have distinct physical features, such as a flat face, round eyes, short neck, and small ears, hands, and feet [1,2]. They may also experience developmental challenges, including delayed speech compared to other children. This condition is usually associated with mild to moderate intellectual disability, with the mental abilities of adults with Down Syndrome typically comparable to those of an 8 or 9-year-old child [2].

Down Syndrome is a significant risk factor for congenital heart diseases (CHD), which are the most common type of birth defect affecting the heart. Approximately 40 to 60% of children with Down Syndrome are born with CHD [3,4]. The prevalence of CHD in Down Syndrome is higher compared to the general population, where CHD affects about 1 in 100 births. Early detection of CHD can be done with an echocardiogram, a test that uses ultrasound to see if the heart muscles and valves are functioning properly, typically performed within the first two or three months of life [3,5].

Congenital Heart Diseases (CHD) are abnormalities in the structure or function of the heart that are present from birth. CHD is one of the most common congenital anomalies found in children and also one of the types of medically compromised patients often seen by pediatric dental specialists. These anomalies can affect the heart walls, heart valves, or the blood vessels entering or leaving the heart. CHD is associated with structural abnormalities of the heart and can be one of the symptoms or syndromes of chromosomal abnormalities. CHD occurs in 8-10 cases per 1000 live births with a balanced gender distribution [6,7].

Most cases indicate no specific genetic factors as the cause, but high-risk factors for CHD include viral infections during pregnancy, such as rubella, maternal diabetes, alcohol and drug consumption during pregnancy, such as phenytoin and warfarin [6,7]. The severity of the disease depends on the hemodynamic lesions. Blood flow disorders are caused by structural abnormalities or obstructive defects that result in blood flow shunting. Types of CHD include Ventricular Septal Defect (VSD), Atrial Septal Defect (ASD), Atrioventricular Septal Defect (AVSD), Patent Ductus Arteriosus (PDA), and Tetralogy of Fallot (TOF). The most common types of CHD in children with Down Syndrome include Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD), and Patent Ductus Arteriosus (PDA) [8,9].

Ventricular Septal Defect (VSD) is the most common septal defect in the ventricular wall. Small defects are usually asymptomatic and detected during routine examinations [8,10]. Large defects can cause shortness of breath, feeding difficulties, and poor growth. 30%-50% of small defects can close on their own within the first year, while large defects typically require surgical closure. Double-Outlet Right Ventricle (DORV) is a type of abnormal ventriculoarterial connection where both great arteries are fully or mostly connected to the right ventricle. Hemodynamically, DORV can resemble VSD, TOF, Transposition of the Great Arteries (TGA), and functional single ventricle. Patients with DORV often have a history of VSD. Cardiac MRI has been widely used to depict the anatomy of DORV with VSD [5,10].

2. Case History

A 6-year-old boy presented with his mother to the pediatric dental clinic at Dr. Saiful Anwar Regional General Hospital, referred by a pediatric specialist for dental treatment of his cavities before undergoing heart surgery. The patient has a medical history of Ventricular Septal Defect (VSD) and Down Syndrome (DS). His general condition is stable, and there is no history of drug allergies. The patient is 113 cm tall and weighs 16 kg. The parents want the child's teeth treated to prevent bacterial infection from the teeth and to prepare for heart surgery.

Upon extraoral examination, everything appears normal, with no swelling or redness observed. However, intraoral examination, revealed moderate caries in teeth 73, 82, and 83; deep caries with perforation in teeth 65 and 74; and multiple gangrenous radices in teeth 55, 54, 53, 52, 51, 61, 62, 63, 64, 75, 84, and 85. There was no intraoral swelling or redness. In addition, since the first visit, the patient had shown very uncooperative behavior. Based on the Frankl behavioral rating scale, the patient showed a rating scale of 1, which is definitely negative and considering the patient's medical condition. Because of these conditions, it was decided to undergo comprehensive dental treatment under GA.



Figure 1 Intraoral Examination

GA was carried out after an examination by a pediatrician and anesthesiologist. Several examinations of systemic conditions were carried out including complete blood cell test, thorax examination. All examinations showed normal and controlled conditions. Before the procedure, the patient was instructed to fast for 6 hours. GA was performed with an endotracheal tube through the mouth and Midazolam 1 mg, ketamine 1 mg, fentanyl 150 mcg, atracurium 15mg was induced also inhalation with midazolam.

Antibiotic prophylaxis was administered 1 hour pre-operation (intravenous injection of ampicillin 1000 mg). After the patient was anesthetized, asepsis was performed on the area around the oral cavity. Dental treatment began with the placement of GIC fillings in teeth 73, 71, 81, 82, and 83, followed by the extraction of teeth 55, 54, 53, 52, 51, 61, 62, 63, 64, 65, 74, 75, 84, and 85. Spongostan was placed in the extraction sockets, and suturing was performed in regions 54, 55, 64, 65, 74, 75, 84, and 85 using Vicryl 4.0 (absorbable silk). Finally, topical fluoride application was performed on the lower anterior teeth. Six hours post-operation, an intravenous injection of 500 mg ampicillin was administered.

After all procedures had been completed, the patient's parents were given instructions, including allowing the patient to drink once consciousness had recovered well, avoiding hot or warm foods and drinks, refraining from playing with scars, instructions for maintaining oral hygiene and scheduling routine follow-up visits.



Figure 2 The extracted teeth



Figure 3 Intraoral photograph after 1 week

One week later, a follow-up appointment was conducted. The patient's parents confirmed that the patient had no complaints. There was no bleeding or swelling at the extraction site, and the sutures in regions 54, 55, 64, 65, 74, 75, 84, and 85 were intact. The glass ionomer cement (GIC) fillings in teeth 73, 71, 81, 82, and 83 were in good condition (Figure 3). Finally, the patient was provided with follow-up instructions, including maintaining oral hygiene, dietary control, brushing teeth twice daily with fluoride-containing toothpaste, scheduling routine dental examinations, and applying topical fluoride every 3 months.

3. Discussion

Children with CHD are known to have low nutritional levels in their early years. Frequent feeding and nighttime feeding patterns are sometimes necessary for children with CHD to maintain adequate caloric intake. Additionally, some medications containing sugar and diuretics are known to cause xerostomia. These factors increase the risk of dental caries in children with CHD. Untreated dental caries can lead to dental infections, increasing the risk of bacteremia and potentially causing endocarditis in patients with congenital heart disease [11,12].

Endocarditis is a microbial infection of the endocardial (endothelial) surface of the heart. Endothelial damage occurs due to structural abnormalities of the heart or great vessels, resulting in blood flow turbulence due to significant pressure differences. This endothelial damage triggers the formation of sterile thrombi and platelet and fibrin deposits (nonbacterial thrombotic endocarditis), which provide a site for bacterial adherence, leading to the formation of infected vegetations. The platelet and fibrin deposits encase the organisms, causing the vegetation to grow larger. Therefore, dental health and the prevention of invasive dental treatment are crucial for children with CHD [11,13].

Children with severe CHD have low tolerance for stress, including anxiety during dental treatment. Hence, dental treatment should be provided in a stress-free condition to prevent cyanosis. For uncooperative pediatric patients with CHD, dental treatment can be performed under sedation anesthesia or general anesthesia (GA). This procedure requires good cooperation with an anesthesiologist due to various conditions associated with CHD, such as chronic hypoxemia, risk of brain abscess, pulmonary hypertension, and bacterial endocarditis. The main factors to consider are the anesthesia techniques used and perioperative medications that can affect the patient's physiological condition during the surgical procedure [5,11,13].

Given the various risks of GA, dental treatment planning under GA should aim for radical treatment to reduce the risk of complications and the need for repeated dental treatment under GA. Teeth that cannot be restored or have a poor prognosis should be extracted. Extraction of necrotic teeth is preferred over root canal treatment. Complex dental treatments, such as root canal treatment on teeth with periapical lesions or pulp necrosis, should be avoided to prevent the need for retreatment under GA. Repeated dental treatment under GA carries greater risks, including morbidity and mortality [12,14].

4. Conclusion

Dental care for children with Congenital Heart Diseases (CHD) is crucial to prevent bacterial endocarditis. Comprehensive dental treatment under general anesthesia is a viable option for children who are medically

compromised. In this case, the pediatric patient was uncooperative and had a medical history of Ventricular Septal Defect (VSD) and Down syndrome with multiple root gangrene and pulpitis. Therefore, comprehensive dental management under general anesthesia can help eliminate infection risk factors in a single visit, providing comfort for pediatric patients who cannot undergo regular outpatient care.

Pediatric dentists need to be knowledgeable about Congenital Heart Diseases (CHD) and Down syndrome, and a good multidisciplinary collaboration between dentists, pediatricians, anesthesiologists, nurses, nutritionists, and various related professions is required to improve the patient's quality of life. The role of the patient and parents in improving oral hygiene is also crucial to enhance the patient's quality of life.

Compliance with ethical standards

Acknowledgments

The authors thank the reviewers for their insightful suggestions.

Disclosure of conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this document.

Statement of informed consent

Informed consent was obtained from patient included in the study.

References

- [1] Bull MJ. Down Syndrome. *New England Journal of Medicine* 2020;382:2344–52. <https://doi.org/10.1056/NEJMra1706537>.
- [2] Kazemi M, Salehi M, Kheirollahi M. I IJ JM MC CM M Down Syndrome: Current Status, Challenges and Future Perspectives. n.d.
- [3] Versacci P, Di Carlo D, Digilio MC, Marino B. Cardiovascular disease in Down syndrome. *Curr Opin Pediatr* 2018;30:616–22. <https://doi.org/10.1097/MOP.0000000000000661>.
- [4] Arias-Lobo R, Lupinta-Paredes E, Calderón-Colmenero J, Cervantes-Salazar JL, García-Montes JA, Patiño-Bahena EJ, et al. Congenital heart disease in Down's syndrome. *Archivos de Cardiología de Mexico (English Ed Internet)* 2023;93. <https://doi.org/10.24875/ACME.M22000409>.
- [5] Delany DR, Gaydos SS, Romeo DA, Henderson HT, Fogg KL, McKeta AS, et al. Down syndrome and congenital heart disease: perioperative planning and management. *Journal of Congenital Cardiology* 2021;5. <https://doi.org/10.1186/s40949-021-00061-3>.
- [6] Cameron AC, Widmer RP. *Handbook of Pediatric Dentistry*. 4th ed. Mosby Elsevier Ltd; 2013.
- [7] Dean JA (Jeffrey A, Jones JE (James E, Sanders BJ, Vinson LAW, Yepes JF, Scully AC. *McDonald and Avery's dentistry for the child and adolescent*. n.d.
- [8] Hassan M, Hasan K, Quader S, Sarker R, Ahmed F, Salam A, et al. Ventricular Septal Defect in Children with Down Syndrome-Two Case Reports. vol. 3. 2010.
- [9] Dimopoulos K, Constantine A, Clift P, Condliffe R, Moledina S, Jansen K, et al. Cardiovascular Complications of Down Syndrome: Scoping Review and Expert Consensus. *Circulation* 2023;147:425–41. <https://doi.org/10.1161/CIRCULATIONAHA.122.059706>.
- [10] Dakkak W, Alahmadi MH, Oliver TI. Ventricular Septal Defect. [Updated 2024 Apr 14]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK470330/>
- [11] Koerdt S, Hartz J, Hollatz S, Heiland M, Neckel N, Ewert P, et al. Prevalence of dental caries in children with congenital heart disease. *BMC Pediatr* 2022;22. <https://doi.org/10.1186/s12887-022-03769-2>.

- [12] Garrocho-Rangel A, Echavarría-García AC, Rosales-Bérber MÁ, Flores-Velázquez J, Pozos-Guillén A. Dental management of pediatric patients affected by pulmonary atresia with ventricular septal defect: A scoping review. *Med Oral Patol Oral Cir Bucal* 2017;22:e458–66. <https://doi.org/10.4317/medoral.21864>.
- [13] Pourmoghaddas Z, Meskin M, Sabri M, Norousali Tehrani M, Najafi T. Dental caries and gingival evaluation in children with congenital heart disease. *Int J Prev Med* 2018;9. https://doi.org/10.4103/ijpvm.IJPVM_401_15.
- [14] Ramazani N. Different aspects of general anesthesia in pediatric dentistry: A review. *Iran J Pediatr* 2016;26. <https://doi.org/10.5812/ijp.2613>