Case Report

Down Syndrome with Ventricular Septal Defect – A Case Report of Surgical Extraction

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Abstract

Down syndrome (DS) is a chromosomal disorder affecting many children across the globe. This disorder often presents cardiac defects such as ventricular septal defect (VSD) and atrial septal defect, which can be a hindrance in providing safe dental treatment in these patients. Effective treatment planning, which includes taking necessary precautions, can help to deliver a comfortable treatment to such patients. This case report is of a pediatric patient of DS along with unrepaired VSD, requiring extraction of a grossly carious nonrestorable posterior tooth.

Keywords: Down syndrome, extraction, ventricular septal defect

Introduction

Down syndrome (DS) is the most common disorder of chromosomal abnormality, occurring in humans and affecting 1 in 400–1500 neonates born of varied populations, depending upon maternal age and/or prenatal screening routines.[1] The syndrome is caused by the trisomy of the whole or part of chromosome 21 in some or all cells of the body, leading to increment in expression due to gene dosage of the trisomic genes.[1] It manifests as mental retardation, cardiac defects, gastrointestinal anomalies, audiovisual impairment, characteristic facial features,[1] and various intraoral abnormalities such as high-arched palate and skeletal class II malocclusion. Ventricular septal defect (VSD) is a frequently occurring anomaly in patients of DS.[2] A VSD is a cardiac defect manifesting as a hole in the septum which separates the two ventricles of the heart.[3] It is the second most common heart defect (26.6%) in Indian children born with DS.[4] Rate of caries occurrence is usually higher in these kinds of pediatric patients,[5] owing mainly to intellectual disability and subsequent inadequate oral hygiene maintenance. This case report presents a case of DS with VSD having carious permanent tooth, managed under chair-side local anesthesia.

Submitted: 11-Dec-2020 **Accepted:** 21-Dec-2021
Published: 06-May-2022

Access this article online

Quick Response Code:

Website: www.ijpedor.org

DOI: 10.4103/ijpr.ijpr_46_20

Case Report

A 12-year-old male patient reported to our clinic with the chief complaint of pain in the lower back teeth for 4–5 months. Pain was spontaneous in nature and aggravated on being supine. The patient had intellectual disability and was identified as Frankl's type 3 behavioral category. Medical history revealed the presence of unrepaired VSD in the heart and the patient developing dyspnea on exertion. Existing reports of karyotype analysis stated trisomy of chromosome 21, confirming the diagnosis of DS. Nonpharmacological behavior management techniques such as voice control and tell-show-do (TSD) were attempted to gain the patient's cooperation. Extraoral examination revealed typical facial features of DS with upslanting palpebral fissures, flattened nasal bridge [Figure 1], wide short hands and fingers, and palmar surface of both hands showing single palmar creases [Figure 2]. Intraoral examination showed high-arched palate, macroglossia, macrodontia, and grossly carious nonrestorable mandibular left permanent first molar [Figure 3]. Rest of the dentition was found to be in good state. Oral hygiene was satisfactory. Radiographic examination of 36 showed wide periradicular

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How to cite this article: Dev S. Down syndrome with ventricular septal defect - A case report of surgical extraction. Int J Pedod Rehabil 2021;6:60-2.

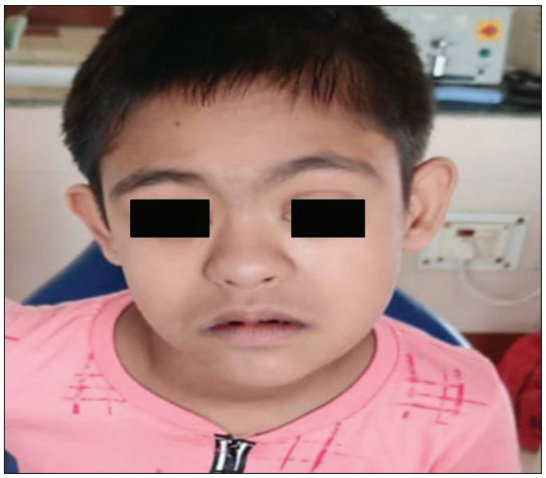

Figure 1: Extraoral facial features typical of Down syndrome.

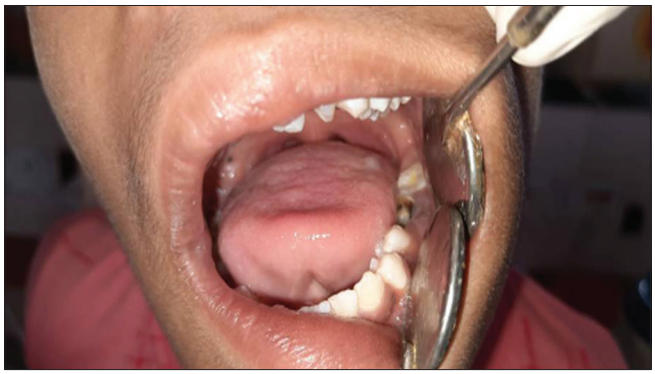

Figure 3: Nonrestorable grossly carious left permanent mandibular first molar.

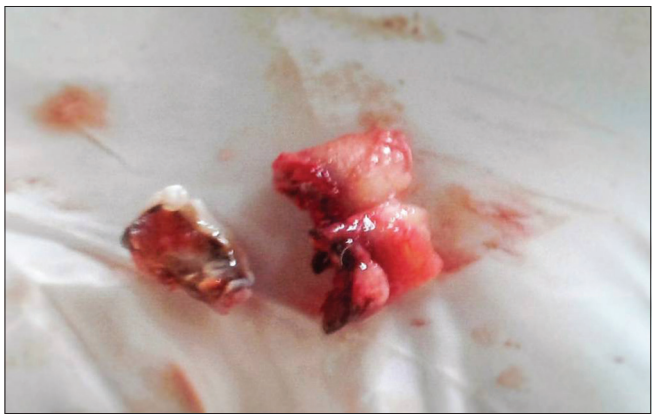

Figure 5: Extracted 36 along with granulation tissue.

radiolucency, with furcation involvement along with taurodontism of 35 and 37 [Figure 4]. Extraction of 36 was planned. Since the patient was of ASA Type 4 category and carried a high risk of mortality if treated under general anesthesia, cardiologist opinion was sought after. The final treatment plan was to perform extraction of 36 under antibiotic prophylaxis using local anesthetic without vasoconstrictor. Amoxicillin 50 mg/kg was administered orally preoperatively as per the American Heart Association recommendations. The

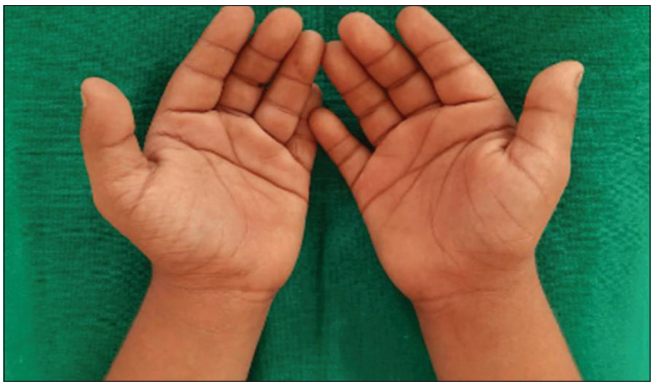

Figure 2: Single palmar crease on both hands.

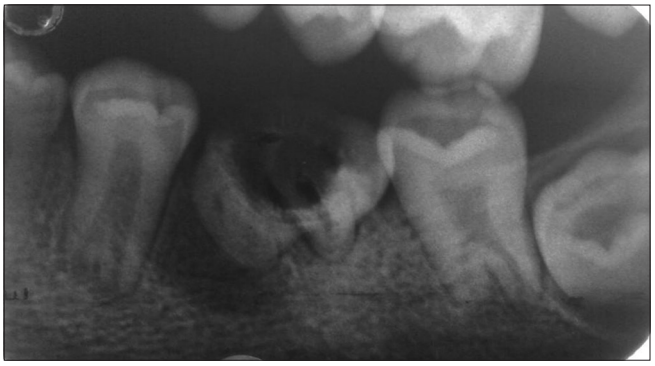

Figure 4: Intraoral perioral radiograph showing grossly carious left permanent mandibular first molar along with taurodontism of 35 and 37.

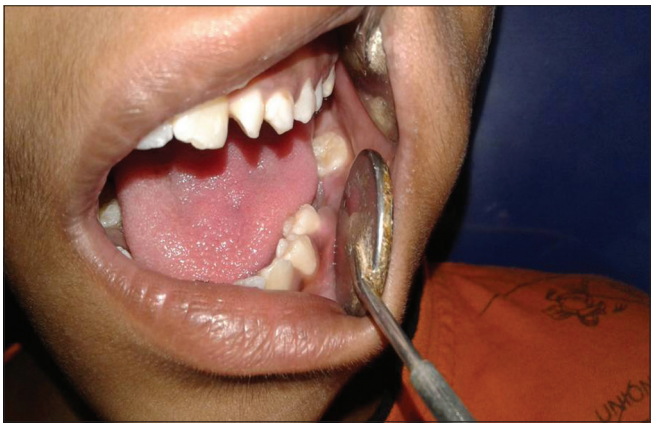

Figure 6: Healed socket of 36 on follow-up.

patient was made comfortable on dental chair using repeated TSD techniques and voice alteration. On achieving satisfactory cooperation, inferior alveolar nerve and lingual nerve blocks along with buccal infiltration adjacent to 36 were performed by administering 2% plain lidocaine. Elevators and forceps were used for the extraction [Figure 5]. Granulation tissue was curetted out from socket, and thorough irrigation of the socket with povidone-iodine was done. The patient was advised to bite on a gauze piece for 1 h and discharged after issuing postoperative instructions. Follow-up appointment was scheduled after 1 week following extraction. Uneventful healing of the socket was seen both clinically [Figure 6] and

radiographically on follow-up. Prosthetic replacement of 36 was also suggested to the parents.

Discussion

VSD is a congenital heart defect which may occur during the first 8 weeks of fetal development. In normal cardiac circulation, oxygen-poor blood returns to the right atrium passes to the right ventricle, which is then pumped into the lungs where it receives oxygen. Oxygen-rich blood returns to the left atrium from the lungs, travels into the left ventricle, and is then pumped out to the systemic circulation through the aorta. A VSD allows oxygen-rich blood to pass from the left ventricle, through the opening in the interventricular septum, and then mix with oxygen-deficient blood in the right ventricle.[6] Thus, less proportion of oxygenated blood in the systemic circulation leads to lesser delivery of oxygen to the different tissues of the body, leading to their hypoxia and disrupted cellular metabolism, and also decrease in respiratory rate, leading to dyspnea, i.e., difficulty in breathing upon any strenuous physical exercise as recorded in the present case. Treating a pediatric patient with a VSD under general anesthesia may present with perioperative complications such as sudden increase in pulmonary vascular resistance (PVR) and rapid deterioration. Manipulations in the oral cavity or any other part of the body may increase PVR in these patients, leading to hypoxia, hypercapnia, acidosis, hypothermia, hyperinflation of the lungs, atelectasis, sympathetic stimulation, and polycythemia.[7] Hence, the decision was made to treat the present case under local anesthesia. Nonpharmacologic management such as TSD has been proved to be an effective psychological intervention in pediatric patients of DS[8] and hence is used to relieve anxiety during dental treatments in such patients. Voice control was also used to gain the patient's attention when needed. Antibiotic prophylaxis before extraction prevents bacteremia in pediatric patients of unrepaired congenital cardiac defect. Although it has been stated in numerous studies that the amount of epinephrine present in local anesthetic used for dental procedures[9] is well below limits to cause any perioperative cardiac complications, lignocaine without adrenaline was used in the present case to provide local anesthesia, merely out of safety concerns. Taurodontism is quite prevalent in DS,[10] as evident in 35 and 37 of this patient. It usually results from delayed growth and fusion of developing epithelial root sheaths due to decreased mitotic activity in the tooth germ cells.[10] Prosthetic replacement of 36 with fixed partial denture has been planned after 1 year, when 37 would be completely erupted and its maximum clinical crown length would be available.

Conclusion

VSD is one of the most common congenital cardiac defects in patients of DS. As these patients often have coexisting intellectual disabilities, dental diseases are also quite prevalent in them due to inadequate practice of oral hygiene measures. Consequently, carious teeth are a common site, sometimes requiring extraction. However, the presence of cardiac defect often poses a treatment dilemma. Hence, appropriate application of nonpharmacological behavior management techniques in these child patients along with necessary antibiotic prophylaxis can avoid the need of general anesthesia and at the same time can provide successful treatment at chair-side practice.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given consent for images and other clinical information to be reported in the journal. The patient's parents understand that the names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Acknowledgments

Department of Pedodontics and Preventive Dentistry, Kolkata, India, is acknowledged.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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