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Dental management of 9- yr old child with congenital heart disease: A case report

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Abstract

Case report: A 9-year-old boy with his mother reported to the Outpatient department of Paediatric and Preventive dentistry who was referred by Paediatrician on account of dental caries. The patient was a known case of congenital heart disease having ejection systolic murmur. The patient had pulmonary stenosis at birth which got resolved without medications. The patient complained of pain in the upper left back region of jaw since 3-4 months which had aggrevated in past 3-4 days. The pain lingered on having sweet foodstuffs.

Clinical findings (dental): Maxillary left first molar had deep caries with a history of nocturnal pain. The intra-oral periapical radiogragh (IOPA) revealed severe caries with 64 and no periradicular involvement. The mandibular right first molar (84) was submerged and had reverse overjet of 5 mm in anterior region. IOPA with 84 showed infraocclusion, drifting of 85 mesially and presence of tooth bud of permanent successor. Space analysis was

performed by Huckaba's method and it showed surplus space by 0.5mm. The lateral cephalogram of the patient revealed a micrognathic and posteriorly placed maxilla in relation to cranial base, and vertical growth pattern of mandible. The coagulation tests were suggested by the Paediatrician and were normal.

Management: Treatment plan to extract the 64 followed by placement of band and loop space maintainer, evaluation of submerged tooth and restoration for pit and fissure caries was planned

Conclusion: Comprehensive dental care of children with CHD requires careful consideration of their medical condition, and dental care delivery should be coordinated with the paediatric cardiologist.

Keywords: case report; management; congenital heart disease; dental

Introduction

Congenital heart defects (CHD) are abnormal cardiocirculatory structure or function present at birth, and often not detected until later in life. They are the most frequent lethal malformation, affecting about 1% of new-borns and causing significant morbidity and mortality in infants.[1] Most prevalence studies reported from India, are on prevalence at birth and the data is based on Paediatric patients reporting to hospitals, it varies from 1.3 to 9.2/1000 population. Meshram and Gajimwar,(2018) who examined total of 42,423 patients availing outpatient and inpatient facilities at the Paediatric department of a tertiary referral centre in Nagpur reported the prevalence as 0.13/1000 live births, with the male:female ratio of 1.3:1 for congenital heart disease.

CHD can be classified in a variety of ways depending upon the site of anomaly, pathophysiology, obstructive or cyanotic and acyanotic defects. Acyanotic defects as present in this case are those defects which causes blood movement from the left to the right side of the heart (oxygenated to deoxygenated).Meshram and Gajimwar,(2018)reported that in Nagpur about 66.74% of patients were diagnosed with acyanotic and 33.26% with cyanotic type. CHD has a range of aetiologies including; chromosomal abnormalities, maternal illness, maternal drug use, in addition to multifactorial and isolated causes. Alcohol ingestion during pregnancy can also lead to congenital heart disease. It is a common feature in a number of chromosomal abnormalities such as 22q11 chromosome deletion, Noonan syndrome, Williams syndrome, trisomy 21, and Marfan syndrome.[4]

Noonan syndrome is a developmental disorder characterized by facial dysmorphia, short stature, cardiac defects and skeletal malformations. It may be sporadic or inherited as an autosomal dominant or recessive trait and occurs, one in 1000-2500 children.[5]

The case report describes the novel occurence of accessory occlusal cusp and submerged tooth in a child with CHD. Short stature, pulmonary stenosis, down slanting palpebral fissures, low set of ears, hypertelorism are some the features which bear resemblance to Noonan's syndrome. Hence, this case discusses the management of dental caries using antibiotic prophylaxis in a case with suspected Noonan's syndrome.

Case report

Chief complaint: A 9-year-old boy and his mother reported to the Out-patient Department of Paediatric and Preventive dentistry who was referred by his Paediatrician. The patient complained of pain in the upper left back region of jaw since 3-4 months which lingered on intake of sweet foodstuffs since 3-4 days and had a history of nocturnal pain.

Medical history: History revealed that the patient was a known case of acynotic congenital heart disease, discovered at 6th week postnatally. The patient was not under any medications, had no prosthetic valves/patches

and hypertension. The parents did not report any drug or environmental allergies.

Clinical features: On general examination, the patient had short stature and his weight was deficient as per the 95th percentile of WHO chart.[6] The general examination did not show any pallor, lymphadenopathy, clubbing or icterus. On extra-oral examination(fig.1), the patient had triangular facies with low set of ears, hypertelorism, down slanting palpebral fissures. He had symmetric face with no facial divergence, and straight profile, an increased lower anterior face height, with everted lip and altered mentolabial sulcus. The patient was suspected case of Noonan's syndrome and was advised to undergo molecular genetic testing for confirmation but was unwilling due to financial constraints.

Intra-oral examination:- Patient was in mixed dentition stage, had lingually erupted permanent central incisors(fig.2) due to over retained primary central incisors. The occlusal evaluation detected a reverse overjet with maxillary anterior teeth (of approximately 5 mm), Angle's class III molar relationship on both sides(fig.2) In maxillary arch there was disto-occlusal caries with 64, pit and fissure caries with primary maxillary first molar on left side(54), and smooth surface caries with maxillary primary central incisor(51,61). Interestingly, accessory occlusal cusp was present on maxillary primary second molars(55,65)(fig.3). In the mandibular arch, pit and fissure caries were present with primary second molars[(75,85)(fig.2)]. The right primary first molar(84) was found to be submerged (fig.3). In addition, noticeable plaque accumulation was observed on plaque score index by Loe and Sillness.

Radiographic and other investigations: Intra-oral periapical radiograph (IOPA) was advised with 64, which showed severe caries and no periradicular changes. Lateral cephalogram was obtained for reverse overjet following which Tweed's, Steiner's and Down's cephalometric analysis was performed and the values are as presented in table 1. It was inferred that the patient had retrognathic maxilla and mandible in relation to cranial base, reverse overjet, Angle's class 3 molar relation and vertical growth pattern of mandible. On space analysis using Huckaba's radiographic method the space was surplus by 0.5 mm and was adequate for eruption of permanent successor. Arch length analysis was not performed due to submergence of 84 and mesial drifting of 85 which would have given an impression of deficient space. Congenital heart disease is associated with systemic conditions such as Down syndrome, Turner syndrome, 22q11 deletion syndrome, Williams syndrome, and Noonan syndrome. Noonan syndrome is the second most common genetic syndrome of congenital heart disease (70-80%), followed by Down syndrome and can be diagnosed by DNA Sequencing Analysis. The most frequent anomalies are pulmonary stenosis. It is an autosomal dominant disorder showing variable phenotypes including short stature, congenital heart disease, and typical facial features. Other manifestations include a bleeding tendency. Opinion from the Paediatrician was obtained to determine the risk of developing infective endocarditis (IE) and to ascertain the fitness for the dental procedure. Blood investigations were suggested such as complete blood count, activated partial thromboplastin time (aPTT) and Prothrombin time(PT). The results of coagulation test were normal.

Diagnosis: The patient had skeletal class 3 with retrognathic maxilla and mandible and Angle class III with Dewey's mod. 3 alongwith posterior crossbite.[7] He also had chronic irreversible pulpitis with 64, ankylosis with 84 on IOPA.

Treatment: The child had moderate to low risk of infective endocarditis (IE)and therefore antibiotic prophylaxis was prescribed 1 hr before with 50mg/kg

amoxicillin and after the invasive procedures. A treatment plan to extract the 64 followed by placement of band and loop space maintainer, and restoration for pit and fissure caries was planned. In the first visit the restorations were done (75,85) followed by pit and fissure placement in permanent molars. In second visit, the band adaptation was done for 65 under prophylactic antibiotic coverage and in the third visit extraction was done under local anaesthesia [lignocaine and adrenaline 1:2,00,000]. (fig.7), following by placement of space maintainer. The third visit was planned within the duration of completion of antibiotic coverage of 5 days which was started after band placement and hence did not require prophylaxis before the procedure. The submerged tooth was kept under observation. Correction of reverse overjet and orthodontic alignment of teeth was planned with the Department of Orthodontics.

Discussion

Providing safe and appropriate oral care to children affected with CHD represents a particular challenge for the paediatric dentist and essentially requires a well-coordinated multidisciplinary approach involving other health professionals, mainly the paediatric cardiologist.

CHD is a common feature in a number of chromosomal abnormalities such as 22q11 chromosome deletion, Noonan's syndrome, Williams syndrome, trisomy 21, and Marfan syndrome.(Hughes, 2019). The incidence of Noonan syndrome is not well characterized, but is estimated at 1/1000 to 1/2500, which makes this disorder one of most common syndromes associated with congenital heart disease. Bertola et al found that short stature (71%); cardiac anomalies (65%) short or webbed neck (87%); and fetal pads in fingers and toes (70%) were most common findings. Some of the other findings were high arched palate (43%) dental malocclusion (37%) low-set ears (19%), down-slanting palpebral fissures 14/31 (45%).[9] Noonan's syndrome was suspected in this case as the patient presented with features like short stature, pulmonary stenosis, hypertelorism, down-slanting of the palpebral fissures, high arched palate and dental malocclusion. The patient did not show any tendency for bleeding diathesis but had delayed eruption of permanent teeth. Though it is seen in Noonan's syndrome, there could be a possibility that underweight of child have could have had an effect on eruption of permanent teeth.[10]

Early intensive prevention and diagnosis of oral disease is vital to reduce the need for dental treatment under general anaesthetic(hughes,2019). Fitzgerald, (2014) stated that children with cardiac disease are often more challenging to manage due to their medical history, possible comorbidities and increased dental anxiety. They may have psychosocial issues and a child's fear of medical treatment may prevent parents from bringing their child for dental care. The patient was precoperative and had little to no dental anxiety. However, behaviour management was carried out using tell-show-do technique and contingency management during dental treatment.

Hallett et al. (1992), Pollard et al. (1992), and Rosén and Stecksén-Blicks (2007) detected higher caries prevalence in children with CHD.[8] Pimental et al(2013)found that children with cyanotic cardiac disease showed a statistically higher caries index, in comparison to those with acyanotic conditions and reasoned that blood supply being an additional variable in caries pathophysiology is an important aspect of host defence and has a risk of enamel hypoplasia.[10,16]Because the oral cavity is highly susceptible to the effects of the disease, early and definitive oral interventions, including toothbrushing and flossing, fissure sealants, topical fluoride therapies, and nutritional measures, need to be implemented to reduce the risk of local and systemic complications (Franco et al. 1996;AAPD2015).

When treating children with congenital heart disease, the paediatric dentist must always consider that transient bacteraemia usually occurs after performing invasive dental procedures; and could be a potential cause of infective endocarditis. Therefore, the patient should be administered antimicrobial prophylaxis prior to dental or surgical treatments that are likely to cause bacteraemia, according to updated guidelines.[6]In the present case, the patient had ejection systolic murmur at the time of presentation and was deemed to be having low- moderate risk of developing infective endocarditis by the Paediatrician and hence, antibiotic prophylaxis was recommended for 2 appointment: band adaptation for space maintainer and extraction of tooth. The dose was as per given in the AAPD policy guideline(2019) i.e. 50mg/kg amoxicillin.[12]

Fitzgerald, 2014[13] stated that while making a treatment plan for the patients with congenital heart disease, identification of potential foci of infection intraorally and risk of developing IE should be taken into account and sometimes a timely decision to extract the tooth might be beneficial. The 64 was the potential source of infection and even after pulpectomy the risk of infection in the coming years cannot be completely nullified and hence needed to be extracted, whereas the restorations were conservative and preventive resin restoration was done.

Infraocclusion is a condition where teeth are found with their occlusal surface below the adjacent teeth, long after they should have reached occlusion. Many terms have been used to describe this condition; the most commonly used being submergence. Radiographically, an obliteration of the periodontal ligament can be seen using conventional methods and the CT-method can also be used to reveal greater detail regarding ankylosis. A treatment decision must take into account based on the presence of tooth bud of permanent successor. In the present case, patient monitoring was recommended, because the primary teeth might exfoliate normally.as the patient had a successor tooth.[14]

The accessory occlusal cusp on primary molar is a rare entity and is situated between the cusp tips of the premolars and molars and on the lingual surface of the incisors and canine. Leigh in 1975, first reported the central cusp as an enamel tubercle on the maxillary right third molar of an Eskimo's skull. The Paired box(PAX) and Msh homeobox (MSX) genes are responsible for this and during the bell stage the altered proliferation and folding of a portion of the inner enamel epithelium and subjacent ectomesenchymal cells of the dental papilla forms a supplemental solid elevation on the crown surface. Chandra and Das(2014) reported a case with accessory occlusal cusp and suggested that they could lead to early involvement of pulp and hence should diagnosed and managed as early as possible to maintain the integrity of primary arch.[15]

Conclusion

While managing a case of congenital heart disease, a Paediatric dentist should have a thorough knowledge about the syndromes associated with it and take into account prenatal and natal history. The detailed history of patient should emphasize on the nature of the disease, previous hospitalisations, current medications, the presence of prosthetic valves/patches, exercise tolerance, and the presence of hypertension. There are considerable challenges associated with providing dental care for these children, hence excellent communication between all healthcare professionals is essential.

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Legend Figures

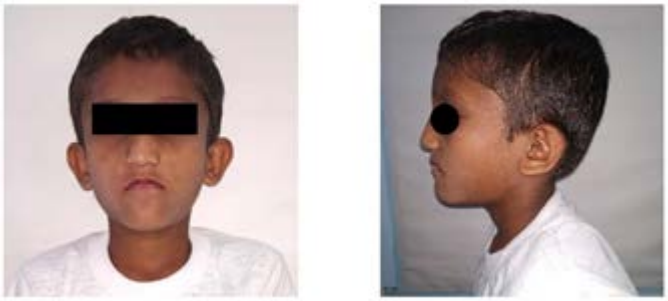


Fig.1: extra-oral features (front and lateral profile of the patient)

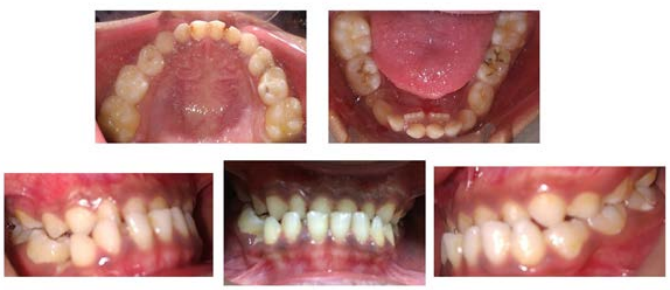


Fig.2: maxillary arch, mandibular arch and occlusal views

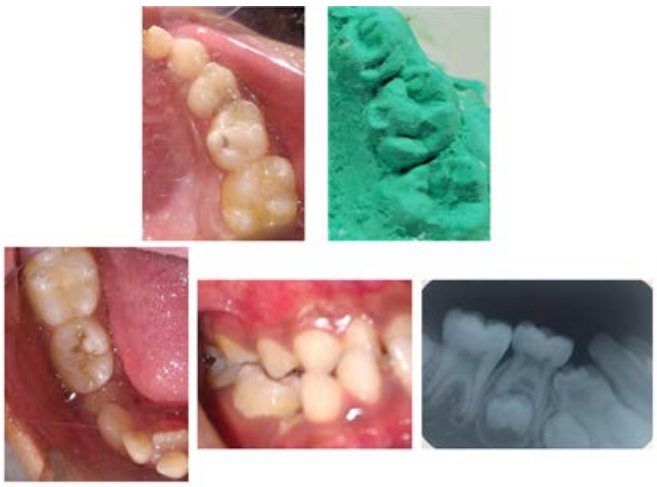
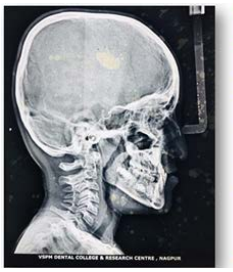


Fig.3: intra-oral findings- accessory occlusal cusp and submerged tooth



Angles	Values	inference
S-N-A angle	68°	Posteriorly placed maxilla in relation to cranial base
S-N-B angle	76°	
ANB angle	-8°	Reverse overjet
occlusal plane- lower incisor	9.2	no proclination of lower incisors
mandibular plane- lower incisor	-7.8	no proclination of lower incisors
Y-axis [FH plane- SGn]	65	Vertical growth pattern of mandible

Table 1: values of cephalometric analysis

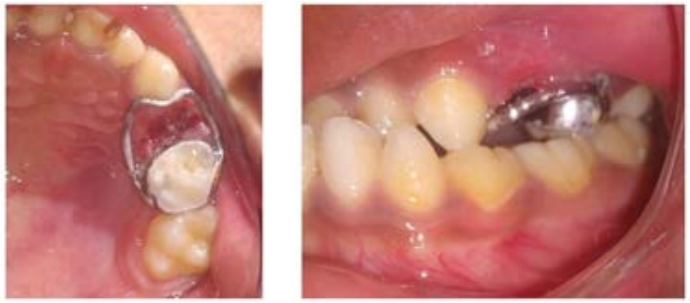


Fig.4: placement of band and loop space maintainer