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CASE REPORT

Oral and systemic manifestations, and dental management of a pediatric patient with Tetralogy of Fallot. A case report.

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Abstract: Introduction: Tetralogy of Fallot is a congenital heart disease and the most common cyanotic heart defect in children. It is clinically characterized by a ventricular septal defect, pulmonary stenosis, overriding aorta over ventricular septal defect and right ventricular hypertrophy. There is little or no information about the oral manifestations in patients with this pathology. A report and discussion of a pediatric patient diagnosed with Tetralogy of Fallot, its clinical manifestations, oral findings and dental management are presented. Case Report: A four-year-old male patient diagnosed with Tetralogy of Fallot and epileptic attacks. The patient has deciduous teeth with many severe early childhood caries, stomatitis and cyanotic mucous membranes, root fragments, periapical abscess and noticeable enamel hypoplasia. Conclusions: In agreement with other authors, children with systemic diseases such as Tetralogy of Fallot have a higher rate of caries, poor oral hygiene, high susceptibility to other infections and bacterial endocarditis, cyanotic mucous membranes and enamel hypoplasia. Primary prevention is critical, proper dental hygiene, regular dental check-ups and the use of antibiotic prophylaxis are particularly important, especially in high-risk patients.

Keywords: *Tetralogy of Fallot, Dental Caries, Heart Disease, Antibiotic Prophylaxis.*

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INTRODUCTION.

Cardiovascular diseases are a wide range of conditions with significant morbidity and mortality. Tetralogy of Fallot (TF), was described in 1673 by Niels Stenson[1], and subsequently by Etienne-Louis Fallot in 1888, who reported the clinical-pathological correlation of the "blue disease"[2]. The term "tetralogy" refers to the four anatomical elements initially described by Fallot: interventricular septal defect (VSD), pulmonary stenosis, overriding of the aorta over the VSD, and finally, hypertrophy or thickening of the right ventricle, whose description is attributed to the Canadian physician Maude Abbott in 1924[3]. TF accounts for 10% of all forms of congenital heart disease worldwide; with a higher prevalence in males[4].

In Mexico congenital heart diseases are the third leading cause of death in children of one year and the sixth cause of death in three-year-olds. TF is a common congenital malformation, with an incidence of 0.1/1000 live births, accounting for 11-13% of all clinical congenital heart diseases (1/8500 live births). The evolution of medicine and technology has increased the survival rate in children with congenital heart disease. However, oral implications are virtually unknown, since attention has been focused on systemic disorders[5].

The etiology of TF is unknown and is related to defects in embryogenesis occurring between the 3rd and the 8th week of gestation. Some of the described risk factors are: maternal infection; rubeola; the use of thalidomide, phenytoin, warfarin, as well as alcoholism and smoking during pregnancy. TF has been considered as a malformation associated with different genotypes, however, most patients are non syndro-

mic. It is related to the use of teratogens and trimethadione, influencing on triggering genetic factors[6]. Gene mutations that may be responsible for up to 4% of all cases of TF have recently been identified[7].

Clinical manifestations of TF are variable and sometimes asymptomatic; among the most frequent are: cyanosis, acropachy, hypoxemic crisis, cardiovascular events, hematological, immunological and respiratory disorders, polycythemia in tissue formation, growth retardation, oral manifestations (stomatitis, cyanotic mucous membranes and tongue, increased risk of early childhood caries, delayed tooth eruption, abnormalities of tooth position and enamel hypoplasia)[8].

The lack of information in the literature on dental management of patients with this type of systemic disease makes this case report valuable.

The aim of this report is to present the systemic and oral manifestations and the dental management of a patient diagnosed with Tetralogy of Fallot.

CASE REPORT.

Socio-demographic data: A four-year-old male patient, full-term eutocic delivery, cyanotic at birth, referred by the Mexican Social Security Institute to the Pediatric Dentistry Clinic at Universidad Autónoma de Zacatecas, for being edentulous.

Medical diagnosis.

Tetralogy of Fallot and epilepsy (Fig. 1), hypersensitive to erythromycin, penicillin and metoclopramide. Receiving treatment with Omeprazole every 12h and Phenytoin/5ml every 8h. Well fed, with uncooperative behavior. Corrective surgeries of cardiac abnormalities and tracheotomy at the age of one year and eleven months were performed. The patient suffers from frequent seizures.

Orofacial diagnosis.

The patient has a slightly convex profile, leptoprosopic type, with increased lower third. At the age of three he underwent extractions and dental restorations. The intraoral examination showed: dentition with multiple severe early childhood caries in 53, 54, 55, 63, 64, 71, 72, 73, 81, 82, 83 and 85, restorations with glass ionomer crowns in 74, 75 and 84, stomatitis and cyanotic mucous membranes, root fragments, periapical abscess and noticeable enamel hypoplasia. (Fig. 2)

Dental treatment and interconsultation with cardiologist.

Initial treatment, prior written consent of the mother, was controlling caries disease. Hygiene and diet counseling was done, as well as regular applications of fluoride. In interconsultation antibiotic prophylaxis was prescribed in agreement with the cardiologist, using Clindamycin in doses of 20mg/kg intravenously within 30 minutes before the invasive procedure to remove the root fragments in teeth 81, 82, 83, 71, 72, 73, pulpotomy and glass ionomer crowns placement in teeth 64 and 65. Finally, rehabilitation treatment was provided by installing a fixed partial denture with a Nance appliance with two orthodontic bands to restore the esthetics of teeth (Fig. 3).

Monitoring the patient.

The mother and the patient were informed about the importance of proper and thorough oral hygiene, especially in the retention area of the prosthesis, thereby avoiding accumulation of plaque, which can lead to infection and subsequent bacterial endocarditis.

Figure 1. Patient with Tetralogy of Fallot.

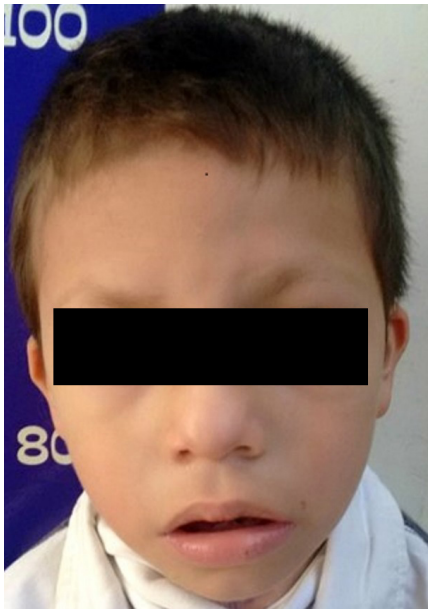


Figure 2. Severe early childhood caries with root fragments.

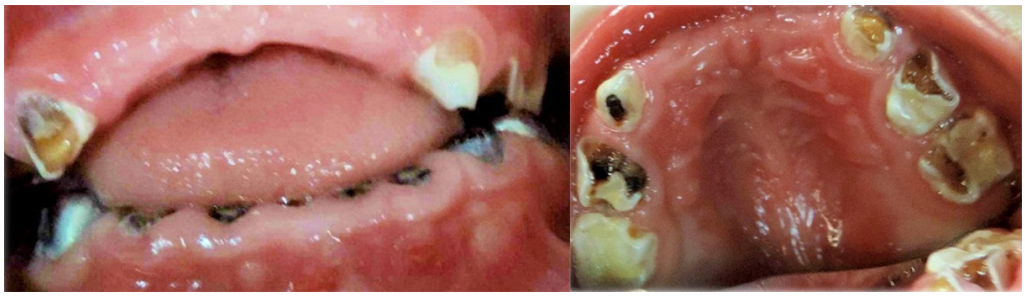
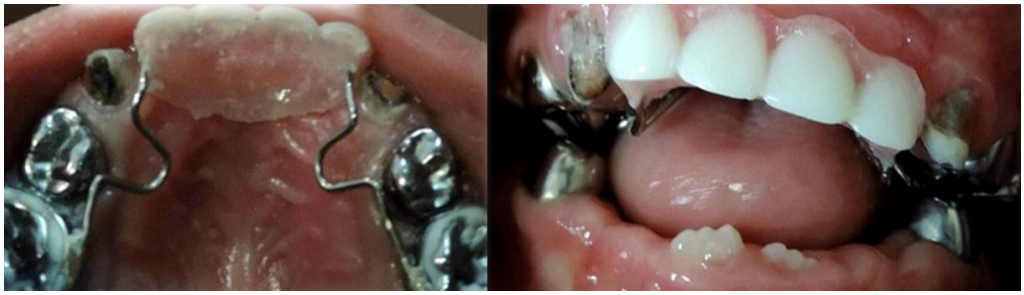


Figure 3. Rehabilitation treatment.



DISCUSSION.

Dental pediatric patients with TF require adjustments in their treatment. First, stress should be minimized during dental procedure, thereby preventing cyanotic events[6,7], which can induce hypoxemia, hyperpnoea and irritability. These can become out of control and lead to loss of consciousness, metabolic acidosis, convulsions and death.

Teeth with enamel defects are common in children with congenital heart diseases such as TF developed during the neonatal period[8]. Hypoplastic and/or hypocalcified teeth are more susceptible to a greater accumulation of plaque resulting in a more rapid progression of caries. These teeth are more sensitive, which results in a greater difficulty to perform oral hygiene, situation that is made even worse by the food that sticks to the teeth. Patients with congenital heart diseases have a higher rate of untreated carious lesions and a higher percentage of enamel defects[4], showing high vulnerability and a higher risk for the development of caries disease[5,9].

Among these children, those treated with Digoxina® showed a greater number of carious lesions[10]. In the case of this patient, the presence of severe early childhood caries (S-ECC) was evident. This can lead to bacterial proliferation and dissemination. S-ECC is related to a child's diet high in carbohydrates, excessive use of bottle and frequent consumption of sweetened liquids. Similarly, chronic use of medications containing sugar increases the number of germs in the mouth and thus the risk of bacteremia after invasive dental treatments[11].

Systemic problems during the amelogenesis stage irreversibly affect ameloblasts, causing defects in enamel secretion or mineralization of enamel matrix[12-15]. The process of mineralization of the dental crown of the first permanent molars begins during the first month of life, taking place between two and four years of age[13]. In the reported case, this period coincides with the uncorrected heart disease. Once the surgery was performed, when the child was a year and eleven months old, it could have adversely affected the formation of this mineralized tissue. Interventions must be performed at an early age in patients suffering from systemic diseases, at the time of the eruption of permanent dentition. Thus, if hypocalcification or other defects in the enamel development appear, the occurrence of carious lesions and loss of hypocalcified enamel can be prevented[15,16].

Children with TF have an increased risk of bacterial endocarditis. The presence of plaque and gingivitis can increase the risk for bacterial endocarditis in children with congenital heart disease[4,17]. It is very necessary to emphasize the importance of preventive measures in health education, through motivation, information, and instructions for oral hygiene, proper diet and active involvement of family members[17]. This will contribute to avoid invasive procedures. If these procedures are mandatory in the case in the case of patients with TF, they will always be preceded by antibiotic prophylaxis[12,18,19].

In the case of this patient, the placement of a fixed rehabilitating prosthesis involves an adequate and thorough oral hygiene that should always be supervised by an adult. The latter in order to prevent plaque build-up, especially in the retention area (bands), thereby preventing future infection, which may result in bacterial endocarditis[20].

Consistent with the literature, in this case a treatment protocol with detailed medical and dental history was followed, achieving a proper diagnosis and treatment. Interconsultation with the cardiologist reduced the patient's risk of developing serious systemic complications.

Características sistémicas, orales y manejo dental de paciente pediátrico con Tetralogía de Fallot. Reporte de caso.

Resumen: Introducción. La tetralogía de Fallot es la patología cardíaca congénita y defecto cardiaco cianótico más común en niños. Clínicamente se caracteriza por una comunicación interventricular, estenosis pulmonar, cabalgamiento de la aorta sobre la comunicación interventricular e hipertrofia del ventrículo derecho. Existe poca o nula información sobre las manifestaciones orales de pacientes con esta patología. Se relata el informe y discusión de un paciente pediátrico diagnosticado con tetralogía de Fallot, sus manifestaciones clínicas, hallazgos orales y manejo dental. Refiriendo además la condición de salud oral del paciente antes y después de los procedimientos dentales. Reporte del Caso. Paciente masculino de 4 años, diagnosticado con Tetralogía de Fallot y crisis epilépticas. Presenta dentición temporal con múltiples caries de la infancia temprana severa, estomatitis y mucosas cianóticas, restos radiculares, absceso periapical y una evidente hipoplasia del esmalte. Conclusiones. En concordancia con otros autores, los niños con enfermedades sistémicas como tetralogía de Fallot tienen un mayor índice de caries, higiene oral deficiente, alta susceptibilidad a otras infecciones y a endocarditis bacteriana, mucosas cianóticas e hipoplasia del esmalte. La prevención primaria es decisiva, higiene dental apropiada, revisiones odontológicas habituales y uso de antibioprofilaxis son significativos sobre todo en pacientes de alto riego.

Palabras clave: *Tetralogía de Fallot, Caries, Enfermedad cardiaca, Profilaxis antibiótica.*

REFERENCES.

1. Silverman NH. The essential echocardiographic features of tetralogy of Fallot. Cardiol Young. 2013;23(6):871–82.
2. Loukas M, Cesmebasi A, Le D, Etienne D, Tubbs RS, Anderson RH. Etienne-Arthur Louis Fallot and his tetralogy. Clin Anat. 2014;27(7):958–63.
3. Swamy P, Bharadwaj A, Varadarajan P, Pai RG. Echocardiographic evaluation of tetralogy of Fallot. Echocardiography. 2015;32 Suppl 2:S148–56.
4. Sawardekar KP. Profile of major congenital malformations at Nizwa Hospital, Oman: 10-year review. J Paediatr Child Health. 2005;41(7):323–30.
5. Cornejo KBZ, Brener IV, Bouchan DR. Manejo estomatológico del paciente pediátrico con cardiopatía congénita. Revisión de la literatura. Univ Odontol. 2011;30(64):57–66.
6. Ajitha N, Chunawalla Y, Morawala A. Dental considerations and management of a pediatric patient with tetralogy of Fallot. Guident. 2014;8(1):74–6.
7. Montero JV, Nieto EM, Vallejo IR, Montero SV. Intranasal midazolam for the emergency management of hypercyanotic spells in tetralogy of Fallot. Pediatr Emerg Care. 2015;31(4):269–71.
8. Nosrati E, Eckert GJ, Kowolik MJ, Ho JG, Schamberger MS, Kowolik JE. Gingival evaluation of the pediatric cardiac patient. Pediatr Dent. 2013;35(5):456–62.
9. Cruvinel VR, Gravina DB, Azevedo TD, Rezende CS, Bezerra AC, Toledo OA. Prevalence of enamel defects and associated risk factors in both dentitions in preterm and full term born children. J Appl Oral Sci. 2012;20(3):310–7.

10. Alva C. Tetralogía de Fallot: Actualización del diagnóstico y tratamiento. Rev Mex Cardiol. 2013;24(2):87–93.

11. Kato T, Yorifuji T, Yamakawa M, Inoue S, Saito K, Doi H, Kawachi I. Association of breast feeding with early childhood dental caries: Japanese population-based study. BMJ Open. 2015;5(3):e006982.

12. Breibart RE, Fyler RE. Tetralogy of Fallot. In Keane JF, Lock JE, Fyler DC (Eds.), Keane: Nadas' Pediatric Cardiology. 2º ed. Philadelphia: Saunders; 2011. pp. 559–79.

13. Babaji P. Tetralogy of Fallot: a case report and dental considerations; Tetralogia de Fallot: relato de caso com considerações dentárias. Rev Clín Pesq Odontol. 2009;5(3):289–92.

14. Corrêa-Faria P, Martins-Júnior PA, Vieira-Andrade RG, Marques LS, Ramos-Jorge ML. Perinatal factors associated with developmental defects of enamel in primary teeth: a case-control study. Braz Oral Res. 2013;27(4):363–8.

15. Denysova EG, Sokolova II. Caries and enamel hypoplasia temporary teeth at children of early age: prevention and treatment. J Health Sci. 2014;4(11):283–91.

16. Seow WK. Developmental defects of enamel and dentine: challenges for basic science research and clinical management. Aust Dent J. 2014;59 Suppl 1:143–54.

17. Paolucci M. Microbiological and epidemiological characterization of the oral cavity microbiota using cultural and molecular techniques.[Dissertation thesis] Bologna: Alma Mater Studiorum Università di Bologna; 2014.

18. Gutiérrez JL, Bagán JV, Bascones A, Llamas R, Llena J, Morales A, Noguerol B, Planells P, Prieto J, Salmerón JI. Documento de consenso sobre la utilización de profilaxis antibiótica en cirugía y procedimientos dentales. Rev Esp Cirug Oral y Maxilofac. 2006;28(3):151–81.

19. Roberts GJ, Holzel HS, Sury MR, Simmons NA, Gardner P, Longhurst P. Dental bacteremia in children. Pediatr Cardiol. 1997;18(1):24–7.

20. Falces C, Miró JM. Prevention of infective endocarditis: between progress in scientific knowledge and the lack of randomized trials. Rev Esp Cardiol (Engl Ed) 2012;65(12):1072–4.