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Dental Management of the Down and Eisenmenger Syndrome Patient

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Abstract

About 40% to 50% of Down syndrome (DS) patients can have significant congenital heart defects such as patent ductus arteriosus, Tetralogy of Fallot, and septal defects. Patients with large septal defects may develop Eisenmenger syndrome (ES), which is defined by the cardiac septal defect and pulmonary hypertension coupled with a reverse right to left shunting of blood flow. DS patients that suffer from this condition require special considerations in the delivery of their dental care to prevent further medical complications or emergencies such as infection, cyanotic episodes, and thromboemboli. Collaboration with the cardiologist is also essential to ensure a complete and comprehensive pre-operative work up. The purpose of this article is to describe the dental management of DS patients with ES under general anesthesia.

Keywords: Down syndrome, septal defect, Eisenmenger syndrome, congenital heart defect, dental management

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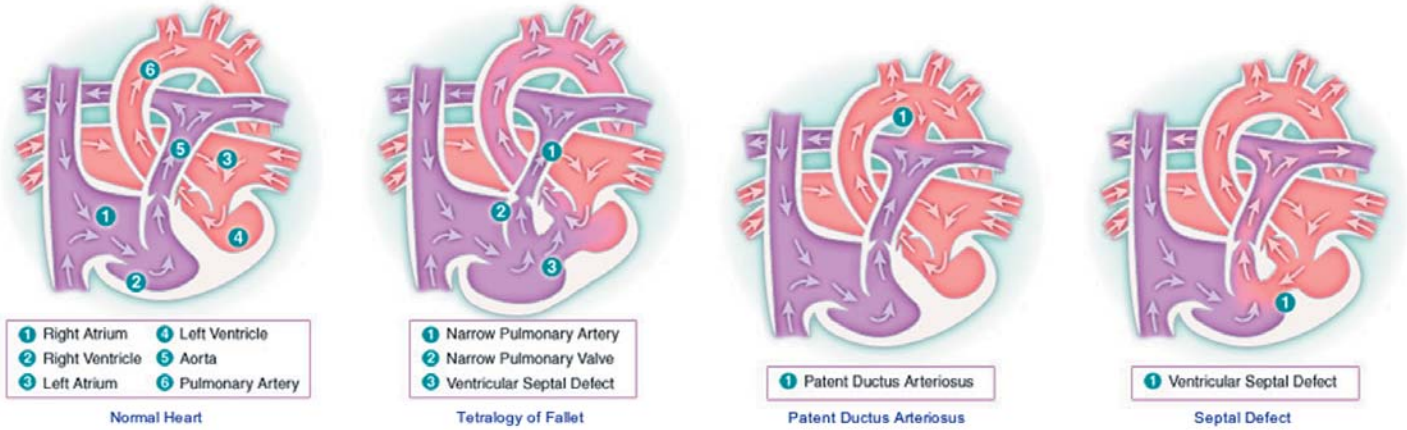
Introduction

Down syndrome (DS) occurs in about one in 700 to 800 births. About 40% to 50% of people with DS have some form of congenital heart disease.[1] Common cardiac abnormalities include Tetralogy of Fallot, patent ductus arteriosus, and septal defects.[2] Tetralogy of Fallot consists of a large ventricular septal defect and pulmonary stenosis. Patent ductus arteriosus is a persistent connection between the aorta and pulmonary artery. A septal defect consists of a hole between the right and left chambers of the heart allowing the blood to flow freely from one side to the other. Since the pressure is usually higher on the left side, the blood flows from the left to the right. Among patients with ventricular septal defects, about half of the patients with a large defect (> 1.5 cm in diameter) develop Eisenmenger syndrome (ES).[3] Patients with significant congenital heart defects require special precautions due to their susceptibility to serious infection. If these patients have both Down and Eisenmenger syndromes, they may require general anesthesia for comprehensive dental treatment and, therefore, require a more extensive work up. The purpose of this article is to describe the dental management of DS patients with ES in the operating room setting and to present two interesting cases.

ES develops due to the increased blood flow from the left ventricle to the right ventricle secondary to the septal defect. This results in pulmonary hypertension. Sustained exposure of the pulmonary vasculature to the higher systemic arterial pressure leads to progressive fibrosis, capillary, and arteriolar occlusion.[4] The obliteration of the pulmonary arterioles and capillaries results in an increase in vascular resistance.[5] Pressure builds in the right ventricle until it exceeds the left ventricular pressure reversing the flow through the septal defect from the right to the left ventricle. Venous blood then mixes with the oxygenated blood in the left ventricle, bypassing the lungs, and re-entering the peripheral circulation. The congenital septal defect, pulmonary hypertension coupled with a reverse right to left shunting defines a person with ES.[6]

These patients may suffer from the following complications: erythrocytosis, bleeding disorders, hyperuricemia/gout, arrhythmias, valve regurgitation, right sided heart failure, paradoxical emboli, thromboses, infective endocarditis, and brain abscesses.[6] Frequent causes of death include sudden cardiac death, congestive heart failure, thromboembolism, and complications from noncardiac surgery.[4,7]

DS patients demonstrate an increased susceptibility to infection. The mechanism for this is unknown, but certainly there are defective and short-lived neutrophil leukocytes. There also appears to be impaired cell mediated immunity, disturbed serum immunoglobulins, and a corresponding decrease in the number of T cells.[8] This could contribute to a higher rate of infection as well as a higher incidence of periodontal disease in this patient population. Due to the congenital heart defect, and increased susceptibility to infections, DS patients with ES can be at an increased risk for bacterial endocarditis and need antibiotic prophylaxis as recommended by the American Heart Association



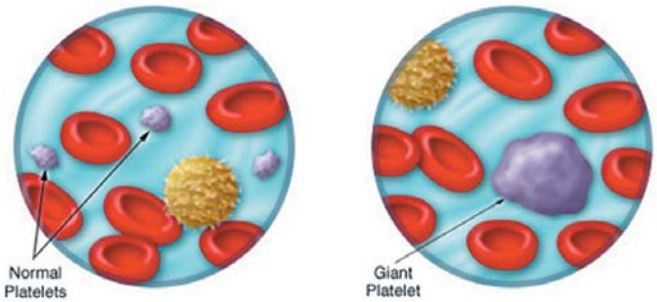
prior to dental treatment. The standard regimen for adults is two grams of Amoxicillin orally one hour or two grams of Ampicillin intravenously or intramuscularly thirty minutes before the dental procedure.[9]

Case History #1

A 29-year old female with a history of ES, a complete atrioventricular septal defect, pulmonary hypertension, seizures, and DS was referred by her cardiologist for comprehensive dental care due to the possible risk of endocarditis. The patient also has a seizure disorder and right hemiparesis secondary to a brain abscess. The seizure disorder is currently controlled with phenobarbital 60 mg once in the morning and then 90 mg once at night. The patient has no known food or drug allergies.

The patient was four feet and eleven inches in height and weighed 109.8 pounds. (49.9 kilograms). She was pale and moderately cyanotic. There was positive clubbing of the extremities as well as perioral cyanosis. Her blood pressure was 100/70 mmHg, heart rate 70 bpm and regular, and oxygen saturation using a pulse oximeter was 74% on room air. A 2/6 early systolic murmur could be heard at the upper left sternal border.

Laboratory data showed her white cell count to be within normal range. Hemoglobin was elevated significantly at 22.6 g/dL, with an elevated hematocrit at 65.3%, mean corpuscular volume (MCV) of 105 fL, mean corpuscular hemoglobin (MCH) of 36.4 pg, and a MCH concentration (MCHC) of 34.6 g/dL. Her platelet count was significantly low at 34,000, but the platelets were described as "huge" as seen by a smear. Her basic metabolic panel was found to be relatively within normal limits. Her phenobarbital levels were elevated at 53 mcg/mL.



The dental examination was deferred until the operating room appointment to minimize the stress for the patient since she was uncooperative. The patient was admitted to the hospital's pre-operative anesthesia unit and given two grams of Amoxicillin orally for antibiotic prophylaxis for bacterial endocarditis. A cardiac anesthesiologist managed the patient's anesthesia.

Dental examination revealed moderately severe generalized periodontal disease and widespread dental caries. Most of the teeth had Class I to Class II mobility. Gross calculus and heavy plaque were found around most of the teeth. Given the poor long-term prognosis of the remaining teeth, lack of proper oral hygiene due to poor patient compliance, and the risk of bacterial infection, the teeth were removed. Primary closure was obtained using 3.0 chromic gut sutures in a continuous loop. Adequate hemostasis was obtained. No blood transfusions were necessary. The patient was extubated without incident and recovered well from the anesthesia. She was discharged later that day.

The patient was given a prescription to use 500 mg of Amoxicillin every eight hours for one week and Tylenol #3 every six hours as needed for pain. The parents were instructed in the post-operative management of the patient and to return to the dental office for a one-week post-operative appointment.

The patient returned to the clinic one week later with no report of complications, bleeding, or difficulty in eating. A limited oral examination revealed no bleeding, and good healing in the extraction sites. Due to the patient's inability to cooperate and lack of coordination, the dentist and parents decided complete dentures would not be tolerated by the patient.

Case History #2

A 36-year old female with DS, blindness, hypothyroidism, an atrioventricular defect with pulmonary vascular disease, and ES underwent general anesthesia for comprehensive dental care.

The patient was uncooperative and did not respond to verbal commands. She was being cared for by her mother. She could only be examined in the sitting position with her mother holding

her. She does not have any known drug allergies and takes 0.75 mg of Synthroid a day.

The patient was small in stature and weight for her stated age. She appeared somewhat pale and moderately cyanotic in her extremities. Her blood pressure was 118/80 mmHg, heart rate was 74 beats per minute and regular, and the oxygen saturation level was 75% on room air. Her lungs were clear to auscultation. Her cardiac examination revealed a palpable right ventricle with normal first heart sound, a loud single second heart sound, and no gallops. She had a 2/6 early systolic murmur along the left sternal edge, but no diastolic murmurs were heard.

Laboratory data at the time revealed a normal white blood cell count but an elevated RBC of 7.45 X 10^6 /mL, elevated hemoglobin of 19.1 g/dL, and a raised hematocrit of 60.1%. She also had slightly decreased MCH of 25.6 pg. The patient's platelet count was also below the normal range at 108 X 10^3, and the platelets were also described as "giant" platelets. The electrolyte panel was within normal limits.

The patient was given two grams of Ampicillin intravenously 30 minutes before the start of her dental procedures for prophylaxis for bacterial endocarditis. She was nasally intubated without complications by the cardiac anesthesiologist.

Her dental examination revealed partially edentulous maxilla and mandible. The maxillary right third molar was fully impacted without any associated pathology. The gingiva was quite red and erythematous with loss of stippling. There was considerable plaque and calculus found around the remaining teeth. Teeth #12 and #17 demonstrated Class III mobility and severe bone loss. The radiographs revealed mild to moderate generalized horizontal bone loss throughout the rest of the mouth. No caries were found by either clinical or radiographic examination. A thorough prophylaxis was performed using hand and ultrasonic scalers.

Tooth #12 was anesthetized by local infiltration with 0.9 cc of 0.5% Marcaine with 1:200000 epinephrine. The remaining 0.9 cc of the carpule was administered as a left inferior alveolar nerve block for post-operative pain management due to its extended anesthetic properties. Teeth #12 and #17 were then elevated and extracted with forceps. The extraction sites were gently irrigated with chlorhexidine and primary closure was obtained using 4.0 chromic gut sutures.

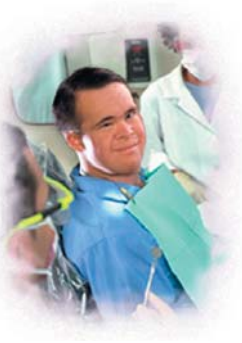
The patient was extubated without incident and recovered uneventfully in the post-anesthesia care unit. She was later discharged that day and went home. The patient was given prescriptions for 500 mg of Amoxicillin in suspension every eight hours for one week and Tylenol with codeine elixir for pain management. She returned to the dental clinic one week later. Her mother reported minimal bleeding following the extractions and no other sequelae. The patient is to return for annual dental exams.

Discussion

Since 40-50% of patients with DS present with congenital cardiac defects, ES can be commonly associated with this patient population. These patients require special considerations in the delivery of their dental care.

A complete and thorough review of the patient's medical history, pertinent laboratory and clinical data, and communication with the cardiologist are essential to prevent complications. Diagnostic workup should include a thorough clinical examination, electrocardiogram, chest x-ray, echocardiogram, pulse oximetry at rest, and blood work (CBC, clotting profile, ferritin, and uric acid). Since these patients are at risk for bacterial endocarditis, they should also be given antibiotics according to the American Heart Association guidelines for the prevention of subacute bacterial endocarditis (SBE).[10] Consideration should be given to prevent any cyanotic episodes due to increase in stress[11] as well as possible dehydration. Maintenance of an adequate circulating volume and the avoidance of pre-operative dehydration or post-operative nausea and vomiting minimize this risk.[12]

Hypovolemia may lead to hypotension, hypoxemia, and sometimes hemo-concentration and thromboemboli. Blood volume expansion should be provided if needed.[13] Both patients in this report were provided adequate intravenous hydration during the surgery.



Good oral hygiene needs to be maintained to minimize the possibility of infections, especially since there is considerable risk involved each time the patient undergoes general anesthesia for routine dental treatment.

Summary

DS is an easily recognized congenital, autosomal anomaly characterized by generalized growth deficiency and mental deficiency. It is likely dental healthcare providers may encounter DS patients with cardiac abnormalities in their clinical practice. Ventricular septal defects are the most common of these abnormalities. Most of these defects are repaired within the first few years of life. However, if the defect is significant enough and remains unrepaired, the patient can develop the cardiopulmonary disorder referred to as ES. Proper oral health maintenance is important in the overall health prognosis for these patients. Early preventive dental treatment will aid greatly in the long-term dental success of these patients by reducing the need to perform invasive or extensive dental treatment that may require general anesthesia. Regular periodic examinations and even a dental prophylaxis may be performed in the dental office setting. If general anesthesia is needed, pertinent diagnostic tests and workup, communication between the dental and medical teams, and appropriate treatment planning are essential for successful long-term care.

References

1. Goodman R, Gorlin R. The Malformed Infant And Child: an illustrated guide. New York: Oxford University; 1983:122-3.s
2. Pastore E, Marino B, Calzolari A, et. al. Clinical and cardiorespiratory assessment in children with Down syndrome without congenital heart disease. Arch Pediatr Adolesc Med. 2000 Apr;154(4):408-10.
3. Kidd L, Driscoll DJ, Gersony WM, et. al. Second natural history study of congenital heart defects. Results of treatment of patients with ventricular septal defects. Circulation. 1993 Feb;87(2 Suppl):I38-51.
4. Vongpatanasin W, Brickner ME, Hillis LD, et. al. The Eisenmenger syndrome in adults. Ann Intern Med. 1998 May 1;128(9):745-55. Review.
5. Heath D, Edwards JE. The pathology of hypertensive pulmonary vascular disease; a description of six grades of structural changes in the pulmonary arteries with special reference to congenital cardiac septal defects. Circulation. 1958 Oct;18(4 Part 1):533-47. No abstract available.
6. Wood P. The Eisenmenger syndrome or pulmonary hypertension with reversed central shunt. Br Med J. 1958 Sep 27;46(5099):755-62. No abstract available.
7. Young D, Mark H. Fate of the patient with the Eisenmenger syndrome. Am J Cardiol. 1971 Dec;28(6): 658-69. No abstract available.
8. Scully C. Down's syndrome and dentistry. Dent Update. 1976 Jul-Aug;3(4):193-6. No abstract available.
9. Dajani AS, Bisno AL, Chung KJ, et. al. Prevention of bacterial endocarditis. Recommendations by the American Heart Association. JAMA. 1990 Dec 12;264(22):2919-22. No abstract available.
10. Dajani AS, Taubert KA, Wilson W, et. al. Prevention of bacterial endocarditis. Recommendations by the American Heart Association. JAMA. 1997 Jun 11;277(22):1794-801. Review.
11. Spivack, E. Tetralogy of Fallot: an overview, case report, and discussion of dental implications. Spec Care Dentist. 2001 Sep-Oct;21(5):172-5.
12. Bird TM, Strunin L. Anaesthesia for a patient with Down's syndrome and Eisenmenger's complex. Anaesthesia. 1984 Jan;39(1):48-50.
13. Newman JH, Sinclair-Smith BC. Risk of high-altitude travel in a patient with Eisenmenger's syndrome. South Med J. 1984 Aug;77(8):1057-8.

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