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Atlantoaxial Instability And Cardiopathies In Down Syndrome -A Dental Perspective

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Abstract
Background*:Down's syndrome (trisomy 21) is the most common chromosomal abnormality that is compatible with life. These patients are of special concern because of their associated problems with regard to congenital heart diseases, gastrointestinal conditions, respiratory and other health issues including oral health problems like dental caries, periodontal diseases, tooth anomalies, facial trauma and injury.*
Aim: *Aim of the study was to find out the prevalence of Atlantoaxial instability and cardiopathies among children diagnosed with Down syndrome so as to be cautious while extending the head while working on the maxillary arch, intubation and the need for antibiotic prophylaxis to prevent infective endocarditis.*
Methodology: *46 children belonging to both the genders who were diagnosed with Down syndrome and referred from the Department of Pediatrics, Institute of Child Health, Medical College, Kottayam to the Department of Pedodontics, Government Dental College, Kottayam were included in the study. Roentgenographic study and Echocardiogram was done at Government Medical College, Kottayam to screen for Atlantoaxial instability and Cardiopathy respectively.*
Results*: The results showed 12 out of 46 children with Atlantoaxial instability(P=<0.01). The incidence of cardiopathies among these children were 20, which is again highly significant.(P=0.01)*
Conclusion*: The provision of dental services to the individual with Down Syndrome presents unique challenges to the dental staff. A thorough knowledge of the unusual medical and dental implications of this syndrome and an innovative problem-solving approach to treatment planning and preventive procedures will do much to alleviate the dental effects of this handicapping condition.*
Key words*: Down's syndrome, Cardiopathies, Atlantoaxial Instability, Dental Management*

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I. Introduction

Down syndrome (DS), first described by Esquirol in 1838, and later in 1866, was described by John Langdon Down and named mongolism.[1] It appears in about 1 in 700 newborns depending on the different population.[2]

These patients are of special concern because of their associated medical problems with regard to congenital heart diseases, hematopoietic disorders, hypotonia (weak neuromuscular tone),recurrent infections, hypothyroidism, gastrointestinal conditions(including digestive and celiac diseases),epilepsy, visual and audiovestibular disability, behavioural and emotional problems like depression, anxiety, attention deficit hyperactivity, respiratory problems with other oral health problems including dental caries and associated infections, periodontal diseases, tooth anomalies, facial trauma and injury.[3]These patients may at times require comprehensive dental care under general anaesthesia

Atlantoaxial (C-1, C-2) instability in Down Syndrome has not attracted general attention as the clinical manifestations are rare and the condition is limited to a small portion of the population.Atlantoaxial instability poses serious challenges to the dental surgeon and anaesthesiologist. Subluxation, dislocation of C-1and C-2 causing spinal cord injury may result in quadriplegia, hemiplegia or even death. As a responsible health professional we have to be prudent in our approach in these patients while flexing their head, to work on maxillary arch and for intubation to give anesthesia.

Another common condition in down syndrome which we tend to overlook is the underlying congenital heart disease. Congenital heart disease (CHD) is the leading cause of mortality and morbidity (about 40 to 63.5%) in DS patients.[4] The high incidence of CHD mandates systematic screening in all children with DS. This is to avoid any complications including bacterial endocarditis as a consequence of the bacteraemia that may occur during the dental procedure.

These issues are often overlooked by the wider dental and medical community. However, in a given context, it is important to be familiar with the incidence and anatomical characteristics of Atlantoaxial instability and Congenital heart disease in DS, as well as the associated complications.Health professionals should be familiar with the common causes of morbidity and mortality in Down syndrome , in order to apply preventive measures and to improve the patient's quality of life.

II. Materials and Methods

Ethical clearance was taken from the Institutional Ethics Committee at Government Dental College, Kottayam and prior permission was obtained from parents / caretakers. A convenient sample of 46 children belonging to both genders who were diagnosed and referred from the Department of Pediatrics, Institute of Child Health, Medical College, Kottayam to Department of Pedodontics, Government Dental College, Kottayam were included in the study for a period of one year. Roentgenographic study and Echocardiogram was done at Department of Radiology and Department of Cardiology,Government Medical College Kottayam to diagnose Atlantoaxial instability and cardiopathy respectively in these children.

Patients with an anterior atlanto odontoid distance of 3 mm or more on lateral cervical X-ray in flexion were diagnosed to have radiological evidence of Atlantoaxial instability[5]. This was done by taking lateral cervical X-rays with the neck held in the neutral position and in flexion.[5] One single faulty has done the study to avoid any bias.

The examination protocol during echocardiographic assessment was as follows: subxiphoid imaging followed by a segmental approach for description of the major cardiovascular structures in sequence, with the image apex at the bottom of the video. All cases underwent detailed review by a single paediatric cardiologist to classify CHD according to standard nomenclature used by the Society of Thoracic Surgery congenital heart surgery database [1]

III Results

We found 12 children out of 46 children with Atlantoaxial instability. ($P<0.01$) , (Significant Value :6.49). Statistical analysis has been done with Z-test Proportion test. There were 5 females and 7 males who were diagnosed with Atlantoaxial Instability. We have done Chi-Square test for testing the statistical significance among genders and found no significance. Chi- square Value-0. 755.P=0.05(Insignificant).

In our study, the prevalence of cardiopathy among these children were 20, which is again highly significant. ($P<0.01$) (9males and 11Females). There was no significant difference among both the genders.

IV.Discussion

Atlantoaxial instability is characterized by excessive movement at the junction between the atlas (C1) and axis (C2) vertebrae, due to either bony and or ligamentous laxity. Neurological symptoms may occur if spinal cord is involved.[7]

The prevalence of radiologically evident atlantoaxial instability in this study was 12 out of 46 children(26.09%)diagnosed with Atlantoaxial instability. The prevalence of AAI varies widely; according to studies done by Bhattathiri et al with roentgenograms in Down syndrome (DS), an incidence of 20%, were noticed while studies with plain films and CT images showed a prevalence of 50% with only 1-2% symptomatic[6]

According to the study done by William C, it is estimated that two percent of children with Downs's syndrome have cord compression producing symptoms, whereas about 20 percent have laxity without any symptoms, which is also known as asymptomatic atlantoaxial instability[7]. Positioning of head and neck during dental treatment and anesthetic management may place the spinal cord at risk if ligamentous laxity is present, so the patients are recommended to undergo radiological evaluation of cervical spine before dental procedures and general anesthesia.[8]The signs and symptoms of atlantoaxial instability include easy fatigability, difficulty in walking, abnormal gait, neck pain limiting neck mobility, torticollis, incoordination and clumsiness, sensory deficits, spasticity, hyperreflexia. These signs and symptoms remain stable for months or years; occasionally they progress and may result in hemiplegia, quadriplegia and death[9].

Cardiopathies in DS is reported to be as high as 40 to 63% and is a major cause of morbidity and early mortality in these patients[10.] Spicer RL in his study on Cardiovascular disease in Down Syndrome found out 40% have congenital heart disease[11]

In our study, patients with cardiopathy who needs antibiotic prophylaxis before dental procedures were 20 out of 46 children (43.48%) with Down Syndrome. According to the study done by Bhattathiri et al, the incidence of cardiac anomalies in Downs's syndrome ranges up to 40 percent[8].Cohort study done by Wright LK et al on 1671 Down syndrome individuals 764 individuals were found out to have congenital heart diseases.[11] Patients do present with Atrial Septal defect, Ventricular Septal defect,Atrioventricular Septal defect,Atrioventricular septal defects ,Patent Ductus Arteriosis,Tetralogy of Fallot, Pulmonary arterial hypertension (PAH) and Eisenmeiger syndrome which mandates antibiotic prophylaxis to prevent endocarditis.[10]

Individuals with CHD should be reviewed thoroughly before any invasive procedures in dentistry as it can cause bacteraemia leading to infective endocarditis and later death. We found no significant difference among both the genders 9 males and 11 females (P<0.01). In the study done by Sanaa Benhaourech among 156 patients with CHD, 128 were identified with Down syndrome. The genders were equally represented (gender ratio 1) and the overall mortality rate was 14.1%.[4]

V Conclusion

Downs syndrome continues to be the most common congenital anomaly. Its incidence is growing due to increasing age of the mothers in developed and developing countries. The provision of dental services to the person with Down Syndrome presents unique challenges to the dental staff. A thorough knowledge of the unusual medical and dental implications of this syndrome and an innovative problem-solving approach to treatment planning and preventive procedures will do much to alleviate the dental effects of this handicapping condition. All these considerations are to be taken care and extra vigilance have to be maintained in these patients during the intraoperative and postoperative period. All specialists and non-specialist clinicians providing care for people with Down syndrome should be aware of the best clinical practice in all aspects of care of this distinct population. The high incidence of Atlanto axial instability and CHD mandates systematic screening in all children with DS, including clinical examination, Roentgenographic examination, Electrocardiography and Echocardiography.

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