# Dentofacial findings in Down's syndrome: report of case

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## Summary

Provision of dental services to the mentally subnormal in our society is relatively neglected. A patient with Down's syndrome who sought dental treatment is reported. Following the assessment of the patient, he needed orthodontic – surgical treatment and scaling and polishing. The former was not given due to the cost, complications and stress of such an involving procedure vis-a-vis the benefit to such a patient. However, patient received thorough scaling and polishing with emphasis on routine 6-monthly dental check-ups given. Handicapped individuals are beginning to seek dental services and the percentage of them doing so is likely to increase as more of such individuals are now staying in the normal home setting. Early preventive treatments for such patients are encouraged. Therefore, the dentists and the rest of the therapy team should be sensitized to face the challenge of providing services to them.

Keywords: Down's syndrome patient; dental treatment

### Résumé

La disposition des services dentaires pour les personnes mentalement retardies dans notre societe est relativement negligee. Un patient souffrant du syndome de Down solicitant un traitement dentaire est reporte. Suivant l'evaluation du patient par le medicin, il devrait subir l'orthodoritique, traitement chirurgical, squarme et cirge. Le cirage n'a pas ete administre a cause du cont, coication et stress de ce dermier et la procedure a suivre vis-à-vis du benefice a un tel patient. Cependant le patient a recu un squamage effectf et cirage avec l'accent mis sur les rendez - vous semestricl. La individus handicappes commencent a visiter les services dentaires et le pourcentage de ces dermiers doit augmenter parce que la plupart de ces individus habitant les maisons normales. Le traitement preventif le plus tot possible de ces maladies est a encourager. Alors, les dentisted et tout le rete du personnel therapentique doivent etre sensibilises pour soulever le defi de render service a ces derniers.

### Introduction

Down's syndrome or mongolism, a case of genetic imbalance (chromosome maldistribution), is usually due to a triplicate set of genes (trisomy 21) instead of the usual paired set of the number 21 chromosomes. The total chromosome complement of 47 is the result instead of the normal 46. It is the most common clinically recognisable type of mental subnormality

The international year of the child, 2001, marks the fortysecond anniversary of the adoption by the united Nation's General Assembly of the Declaration of the Rights of the Child. The fifth of these principles affirms the right of children who are physically, mentally or socially handicapped to the special treatment, education and care required by their particular condition [1]. Dental treatment of the handicapped child lies well within the province of every general practitioner and specialist who is willing to contribute understanding, practice and ingenuity. Even though the handicapped do not constitute

Correspondence: Dr. C.O. Onyeaso, Department of Preventive Dentistry, College of Medicine, University of Ibadan, Ibadan, Nigeria. a large portion of a dentist's practice, it is important that the dentist be psychologically and technically prepared to work with these patients. Good oral health is not only essential to maximise the biological potential of these patients, but often to enhance their self-esteem, dignity and acceptance by society [2].

Though in the last four decades the way in which dental defects can have a contributory effect in the total picture of the intellectually handicapped has become better understood, not much has been reported concerning the mentally handicapped in relation to dentistry in our environment with consequent relative neglect of this group of individuals in the provision of dental care services.

Globally, in most documented studies of malocclusion in the handicapped, the distribution of Angle's classes was related to normative values for the general population. The mentally retarded do not differ widely from these norms except the children with Down's syndrome [3]. None of the reports on mentally handicapped children in relation to dentistry in Nigeria has assessed the occlusion of these patients [4,5,6]. There is no previous report on this common clinically recognisable mental subnormality in our hospital.

This is a report on dentofacial findings in a young man with Down's syndrome who reported for dental treatment. The main objectives of this case report are to affirm that mentally handicapped children can and are seeking dental care including orthodontic services as well as to encourage the need to establish the right initial early contact with such patients for dental preventive procedures. Interdisciplinary team approach in their management is hereby encouraged as in other civilised societies.

## **Clinical observations**

Ten significant deformities or cardinal signs could accompany the Down's syndrome in the new born described by Hall, as follows [7].

Signs		Incidence (Percentage)
(1)	Hypotonia	80
(2)	Poor mororeflex	85
(3)	Hyperflexibility of joints	80
(4)	Excess skin on back of neck	80
(5)	Flat facial profile	90
(6)	Slanted palpebral fissures	80
(7)	Ear anomalies	60
(8)	Dysplasia of pelvis	70
(9)	Clinodactyly of fifth finger	60
(10)	Simian crease	45

There is a total retardation of appositional and endochondral growth leading to small stature with an awkward, waddling gait. The head is brachycephalic with a greater reduction in size in the posterior aspect, producing a relatively flat occiput. The skin could be dry, eczematous, and frequently hyperkeratotic. Kisling [8] described hypoplasia of the frontal sinuses and a small nasal bridge, with complete aplasia of the nasal bone in 15% of the cases. The eyes are characterised by an oblique slant, narrow palpebral fissures, and, in the younger patients, epicanthal folds. There is usually a convergent or divergent strabismus. Speckling of the iris (Brush field's sports) is frequently present in the infant but disappears with age. The neck is usually short and thick, presenting anatomic problems in the administration of general anaesthesia, and is frequently webbed (pterygium colli).

Cardiac anomalies are present in approximately 40% of individuals with Down's syndrome and together with a tendency to leukemia, constitute the greatest hazards to long life [2].

The oral manifestations in Down's syndrome include the following: Scrotal tongue, macroglossia with protrusion and openbite, palatal anomalies (prominent anterior rugae, thickened lateral processes in maxilla, two furrows on palate), hypoplastic maxilla, oblique gonial angles (class III tendency), occlusal disharmonies (anterior crossbite, posterior crossbite, rotations), microdontia, over-retained deciduous teeth, aberrant patterns of eruption, cone-shaped teeth, hypoplastic enamel, anodontia, supernumerary teeth and everted, thick, dry and crusted lips.

The incidence of caries in patients with trisomy 21 syndrome has been studied extensively and has been found to be lower than in controlled subjects, both institutionalized and non-institutionalized [2]. The reasons for this are speculative, ranging from theories of morphologic characteristics to cultural factors, such as diminished access to high carbohydrate foods and high bicarbonate content and PH of the saliva. Numerous studies agree that there is a higher rate of periodontal disorders in these individuals [2]. Plaque accumulation and poor oral hygiene remain some of the major problems of dental management due to patients disabilities and made worse by mouth breathing.

Most of the children are affectionate, happy, and friendly with their social performance frequently exceeding the mental age. Their mental age rarely rises above 50. However, they may present problems in dental management if they suddenly become stubborn.

## Case Report

S. A., a Nigerian young man, first presented in March 2001, when aged 22 years, having been referred from the plastic surgery clinic because of inability to close his mouth. Information was obtained from his Aunt (the Guardian) and his mother confirmed the history. The patient had not been able to close his mouth since childhood with consequent problem with incision of his food. Patient was the first born of his mother who is presently a 42-year-old woman. The mother gave no relevant drug history during pregnancy and claimed she had an uneventful 9-month gestation period, which ended in normal delivery. He had squinted eyes as a baby and hearing difficulty was noted at about 5 years of age. The father who had divorced the mother long time ago has other wives. No other sibling has similar condition.

The patient could neither walk nor talk until 3 years of age. He could not cope with his schoolwork and only managed to get to primary 5 before he dropped out. The intelligent quotient (I Q) was subjectively assessed to be about 45.

## Mental age x 100

### Chronological age

Extra oral examination showed patient's inability to close his mouth with severe skeletal anterior open bite. (Fig. 1)



Fig. 1: Facial view of the patient showing the skeletal open bite and grossly incompetent lips

There was mid-face retrusion and Mongoloid slant (upward sloping of palpebral fissure). The lips were thick, everted and dry.

The intra-oral evaluation of the patient revealed the following:

- Caries:
  - No carious lesion was detected.
  - Periodontal diseases
     There was generalised chronic marginal
     gingivitis
     The gingival index was 0.67 according to
     Loe [9].
- Plaque and calculus deposition. There was plaque and calculus accumulation mainly on the right upper and lower quadrants. The plaque index was 1.0 according to Silness and Loe [10]. The calculus index was 1.67 according to Greene and Vermillion [11]. There wasn't any severe periodontal disease except for the chronic marginal gingivitis
   Congenitally Missing Teeth
- The patient had all the permanent teeth present including the third molars
- Occlusion The patient had Angle's class III malocclusion on the right side and class II on the left side. Skeletal anterior open bite measuring 7mm was noted.

Anterior crossbites of 12 and 22 were observed

Posterior crossbites involving 16, 15, 14 and 24 were recorded too

- Palate anomaly. The palate was high and narrow
  - Tongue anomaly There was macroglossia with protrusion. (Fig. 2).
- Habit
  - Tongue thrusting was evident. (Fig. 2)



Fig. 2: Frontal view of the patient's occlusion (Intraoral)



Fig. 3: Side view of the patient's occlusion showing the heavy dental calculus and large protruding tonque.



Fig 4a: The photographs of the study models of the patient - frontal view (4a) and lateral views (4b and 4c)



Fig. 4b: Note the crossbites of 16,15, 14 and class III molar relationship on the side (4b)



Fig. 4c: Note the crossbite of 24 and class II molar relationship on the left side (4c)

### Records

Study casts of the patient's occlusion were made along with the clinical photographs. Patient already had taken radiographs such as postero anterior and oblique lateral views before presenting in the orthodontic clinic but cephalometric radiograph was not prescribed when it was obvious that patient could not have orthodontic treatment/orthognathic surgery considering the cost, complications and stress of such involving procedure visa-vis the benefit of the procedure to the patient.

## Treatment

Patient was referred to the periodontology clinic where he had thorough scaling and polishing of all the teeth done. Himself and the guardian were further motivated on the need for good oral hygiene and 6-monthly dental check-up visits since prevention is the hallmark of treatment in such a patient.

#### Discussion

Previous studies have reported a decreased caries activity among Mongoloid individuals [12-15]. This patient had no carious lesion in any of the teeth.

The finding regarding gingivitis (chronic generalised marginal gingivitis) in this patient appears to agree with the findings in previous investigations [12, 16]. This condition is said to be more prevalent among the Mongoloid retardates [12].

The oral hygiene of this patient, though poor, was not too bad as would have been expected from such a patient and this could be due to supervision of the oral hygiene procedure at home by the guardian. A study [6], which compared the oral hygiene status of the mentally retarded children and normal children of comparable ages in Lagos, showed that the normal children needed more scaling and polishing procedures than the retardates.

This patient had all the teeth present. However, missing permanent teeth, particularly the maxillary lateral incisors, appear to be a part of the Down's syndrome in a significant number of such patients as noted by other investigators [12,13] Class III type of malocclusion, anterior open bite and crossbites found in this patient were found in a significant percentage of such patients studied 12]. These findings are documented in the literature [2,12,16,17]. However, it is interesting to note class II molar relationship on the left side suggesting basic skeletal disproportion in this patient.

Usually, prolonged retention of deciduous teeth causes malposition of teeth and malocclusion. The reported [2] high incidence of this condition (retention of deciduous teeth) in Down's syndrome patients suggests that early dental monitoring of such patients and prompt extractions of the retained deciduous teeth is likely to reduce the prevalence of malposition of the permanent teeth [2]. This is likely to reduce the prevalence of crossbites which are usually more likely to occur in class III malocclusion cases. Also, further studies will be necessary to determine whether this class III malocclusion is due to a small palate or protrusive mandible.

The high palatal vault noted in this Mongoloid patient agrees with some previous reports [2,12,16,17] However, Shapiro [8] reported no significant difference in measured vault height in a group of Mongoloid children. He, however, reported of narrower and shorter palates. Thus, there may not be an actual difference in measured height of the palatal vault, but merely a different palatal form for Mongoloid retardates. Further studies are indicated in regard to palatal form and vault height in our environment when more cases are found.

The finding of a large and protruding tongue in this patient appears to agree with the findings of other investigators, although this is only a subjective judgment [2,12,16]. Tongue thrusting habit found in this patient is likely to have been giving an accentuating effect on the anterior open bite. Macroglossia with protrusion is documented in the literature in Down's syndrome cases [2,3,12,13,16,17,18].

The incompetent, thick, everted and dry lips noted in this patient could be related to abnormal development of oral musculature documented in Down's syndrome children [19].

It is important to emphasis here that orthodontic management or dental care generally of the mentally subnormal patients should be handled in an interdisciplinary team approach. The following should be involved so as to give the mentally retarded patients the best available and effective care [2]: dental team (orthodontist, paedodontist, general dentist, oral surgeon); psychiatric team (psychiatrist, psychologist); plastic surgeon; parent (guardian, nursing staff); educational team (principal, teacher); social service worker (sociologist); speech therapy (speech pathologist, audiologist); physical medicine team (physician, orthopaedic surgeon, physiotherapist) and paediatric team (paediatrician, paediatric neurologist, allergist).

In conclusion, it is worthwhile to affirm that mental retardates are showing up for treatment and the number is likely to increase as more of them are being kept in the natural home setting in our environment. As oral health awareness increases in our environment, the parents/guardians of the affected are likely to seek dental care for them including orthodontic services. Therefore, not only that the dental profession should be more prepared to offer these services but the rest of the therapy team should be sensitized to their roles in giving the best of the required treatments to the mentally subnormal in our society.

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