

Oral Health Management of a Patient with 47 XYY Syndrome



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Introduction

The 47XYY syndrome is an aneuploidy (abnormal number) of sex chromosomes, where a human male receives an extra Y-chromosome making a total of 47 chromosomes instead of the usual 46. This produces a 47XYY karyotype individual that occurs in about 1 in 1000 male live births. These patients have an increased growth velocity from childhood. Physical phenotype is normal, with tall stature by adolescence. Some cases have been reported with learning disability although majority of them have IQ comparable to the general population. Most 47XYY males have normal sexual development and usually normal fertility.

T. Laine & L. Alvesalo (1985) reported that an extra Y chromosome in 47XYY men is associated with changes in palatal and mandibular arch dimensions, more often an increase than a decrease. Studies on individuals with sex chromosome anomalies have demonstrated the promoting effect of the Y chromosome on tooth crown enamel and dentin growth (Alvesalo et al, 1993).

R. Lähdesmäki & L. Alvesalo (2004) investigated permanent tooth root lengths in 47XYY males that indicate longer tooth roots in 47XYY males compared with those in controls.

Alvesalo et al (1985) indicated, that the thickness of enamel and that of dentin is increased in the teeth of 47XYY males as compared with controls.

Case Report



Fig 1 & 2

The Patient is an eighteen year old male (DOB - 30/6/1994) student of Yemini origin with normal physical structure and development. He was diagnosed with 47XYY syndrome in 1999 at King Khalid University Hospital, King Saud University by high resolution chromosomal analysis. The patient has been on record since 1999 at College of Dentistry, King Saud University. According to the report from the Genetics clinic, IQ was assessed to be 91.

Medical History

Previous Medical History;

The patient was on treatment and follow-up for the following conditions;

- Congenital Heart Disease, Right bundle branch block (RBBB)
- Atrial Septal Defect (ASD) which was surgically closed. Later, Echo Cardiograph was normal in 2003
- Bronchial Asthma; presently using Ventolin inhaler

Drug History;

The patient was on Ventolin inhaler SOS

Allergies;

Patient is allergic to penicillin (Amoxycillin); that was noted during the previous antibiotic prophylaxis for dental procedures.

Dental History

Date of first visit for current treatment; May 2012

Presenting Complaint; The patient desires to have replacement for missing teeth.

History of Present Complaint; Patient has numerous unerupted permanent teeth and many retained deciduous teeth. Patient desires to have good aesthetics as he is being bullied at school.

Past Dental History ;

The patient has undergone various dental procedures including surgical extractions of teeth # 51 , # 71 and # 81 in Dec 1999 and multiple amalgam restorations for all primary molars and fluoride application in 2002 under general anaesthesia and removable partial dentures and other dental procedures under local anaesthesia with antibiotic prophylaxis (for his childhood cardiac condition). He usually brushes his teeth twice daily with a fluoridated tooth paste. Upper primary incisors were indicated for extraction but were not removed.

Social History;

Non Smoker

Examination

Physical Examination;

Height: 177 cm Weight: 55 Kg

Extra-Oral Examination;

Some dysmorphic features not explained by the 47XYY syndrome such as hypertelorism, depressed nasal bridge, epicanthal folds and low-set small ears were also present (Fig 1 & 2).

Intra-Oral Examination;

Tongue is macroglossic and protruding between the teeth. Anterior open bite with reverse overjet was present.

Teeth Present

6 E D C 2 1 2 C D E 6
7 6 5 E 4 C 1 1 2 4 E

Previous Radiographic Record

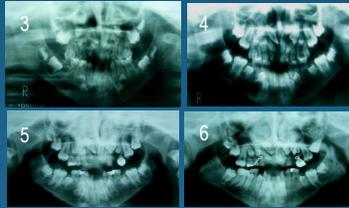


Fig 3-6 Series of Dental Panoramic Tomographs (DPT) showing progression of dentition since 1999. Multiple unerupted permanent teeth and multiple retained deciduous teeth can be seen.

Clinical Presentation



Fig 7-10 The peg lateral in the upper right quadrant is almost present in the horizontal plane (Fig 7). Multiple decayed teeth and undesirable aesthetics due to an overused partial denture (Fig 8). The DPT confirms the impact of right maxillary incisor (Fig 9). Lateral cephalometric radiograph shows a skeletal class III relationship (Fig 10). The patient possessed an anterior reverse overjet with an open bite.

Treatment plan;

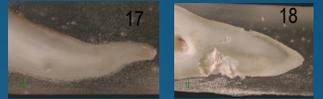
- Extraction of malposed and non-restorable teeth
- Caries control by Oral Hygiene Instructions, high fluoride tooth paste and mouthwash
- Restoration of permanent teeth
- Extraction of deciduous teeth to aid eruption of permanent teeth
- Prosthodontic rehabilitation by over-denture & single crowns to restore aesthetics
- Orthodontic extrusion of permanent teeth.

Intervention

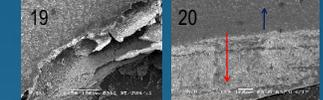


Extraction of deciduous teeth and peg lateral to aid the eruption of succeeding teeth (Fig 11 & 12). Patient was treated under sedation (Fig 13). Caries control and stabilizing the condition. Extraction of multiple retained deciduous teeth (Fig 14). Rubber base impression and bite registration for construction of immediate denture after extraction (Fig 15 & 16). All procedures were done without any antibiotic prophylaxis as per the NICE guidelines, 2008 for prevention of Infective Endocarditis during dental procedures.

Results



Photomicrographs— Fig 17 showing thin cemental layer on root surface (in peg lateral tooth) Fig 18 showing thin cemental layer on sides of root with dental sclerosis



Scanning Electron Microscope (SEM) Images Fig 19 showing thin irregularly arranged enamel rods and more inter-rod substance Fig -20 SEM showing thin enamel layer with haphazardly arranged enamel rods (red arrow) and normal dentinal layer (blue arrow) in the crown of the tooth. Note the straight Dentino-enamel junction.



Fig 21-23 shows restoration of acceptable patient aesthetics. Maxillary removable over-denture and mandibular temporary crowns.

Orthodontic treatment plan

- Facilitating the eruption of permanent teeth by extraction of retained primary teeth
- Distalisation of lower left first premolar to provide more space for the eruption of lower anterior segment
- Disimpaction of impacted teeth by orthodontic traction through the use of upper mini-implants.
- Upper transpalatal arch and lingual holding arch to anchor the disimpaction of the anterior teeth.
- Leveling and alignment will be performed to allow for space creation and final occlusal detailing.

Acknowledgment

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