Journal of Orthodontics, Vol. 30, 2003, 291–296

CLINICAL SECTION

Orthodontic and orthognathic management of a patient with osteogenesis imperfecta and dentinogenesis imperfecta: a case report

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Abstract

This case report describes a patient's severe Class III malocclusion, managed with a combination of orthodontic and orthognathic treatment. The medical history was complicated by osteogenesis imperfecta and dentinogenesis imperfecta. In addition the patient was a Jehovah's Witness.

Patients with osteogenesis imperfecta carry an increased risk of perioperative haemorrhage, and this led to bimaxillary surgery being carried out as two discrete surgical episodes for the patient described. In addition, the risk of enamel fracture led to orthodontic bands being cemented on all teeth. In spite of the increased risks a successful outcome was achieved.

Index words:
Orthognathic, osteogenesis imperfecta

Received 1 February 2001; accepted 9 September 2002

Introduction

Osteogenesis imperfecta (OI) is a genetically determined disorder of connective tissue, also known as 'brittle bone disease'.[1] It results from mutations in the genes COL1A1 and COL1A2 that encode for either chain of type 1 collagen.[2] All tissues rich in type 1 collagen can thus be affected. Patients can therefore present with a combination of features, including multiple long bone fractures and deformities, growth deficiency, joint laxity, hearing loss, blue sclera, and dentinogenesis imperfecta (DI).

Some patients with OI display no clinical or radiographic abnormalities in the dentition, whereas others manifest significant dentinal involvement. The primary dentition is generally more severely affected and, clinically, the teeth appear opalescent grey, brown, or yellow. Radiographically, there is a marked cervical constriction, the crowns are bulbous, the roots short, and the pulp chambers and canals become increasingly obliterated with time. Loss of enamel results from a weakness within the dentine itself, rather than from an abnormality in the dentino-enamel junction.[3]

Distinctive facial traits may also be apparent. These include a triangularly shaped face, a broad bossed forehead,[2] and an overhanging occiput.[4] Studies demonstrate that approximately 75% of adult patients with OI exhibit Class III malocclusions.[2,3,5] Ectopic eruption of first and second molars has also been found to be more common.[2]

There are a number of important issues relating to the surgical and anaesthetic management of these patients:

(1) the ease of fracture of bone and teeth;
(2) increased tendency to bleed secondary to platelet and possible vascular disorders;
(3) increased tendency to develop malignant hyperthermia;
(4) difficulty in intubation as many patients may have short necks, large tongues and thoracic deformity.

One helpful feature enabling segmental osteotomy may be the favourable configuration of the roots.[4]

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Case history

A female aged 17 years, 7 months was referred by her general dental practitioner complaining of a prominent mandible (Figure 1a–f). The severity of the malocclusion caused great difficulty obtaining photographs with the teeth in occlusion. Her medical history indicated that she had osteogenesis imperfecta together with dentinogenesis imperfecta. The patient was a Jehovah witness and, consequently, refused blood transfusions.

She presented with a severe Class III malocclusion on a skeletal Class III base (Figure 2a; ANB –10 degrees) with a reduced maxillary/mandibular plane angle (15 degrees). The aetiology of the Class III malocclusion was a combination of maxillary retrusion (SNA 76 degrees) and mandibular protrusion (SNB 86 degrees). There was

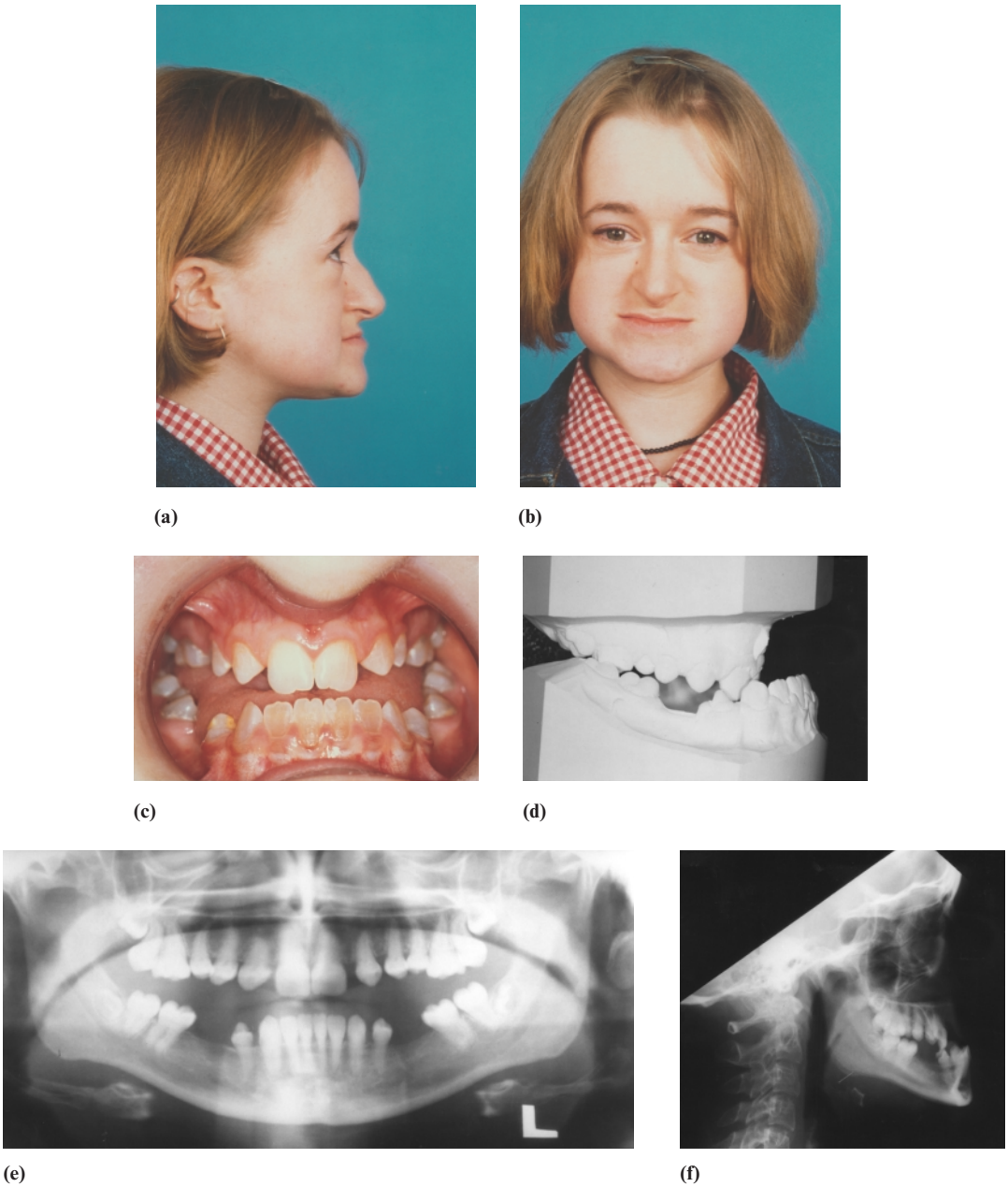

Fig. 1 (a–f) Pre-treatment photographs, study models, orthopantomograph and lateral cephalograph. Intra-oral records and lateral cephalograph taken with teeth in occlusion.

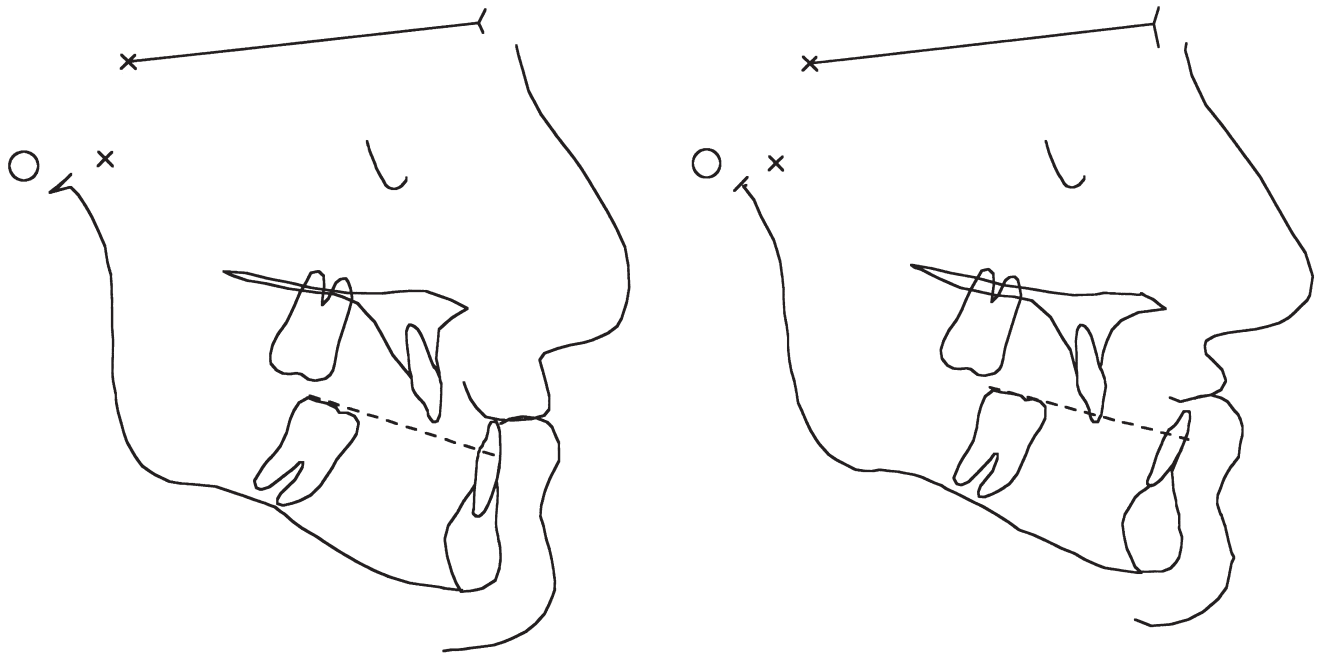

Fig. 2 (a,b) Cephalometric tracings: pre-treatment, and post-decompensation.

no apparent facial asymmetry. Intra-orally the upper permanent lateral incisors (12,22) and the lower second premolars (35,45) were developmentally absent. Otherwise all other permanent teeth were present with the third molars unerupted. The upper incisor inclination was within normal range at 106.5 degrees with no incisal display with the lips at rest. The lower incisors were retroclined at 79 degrees compensating for the skeletal discrepancy, and there were 1-cm spaces mesial to the lower first molars. The overjet was –11 mm and the molar relationships were Class III bilaterally in the intercuspal position. The overbite was increased and incomplete. There was a complete crossbite of the lower arch, with no mandibular displacement on closure. Radiographically, features of DI were noted, such as short roots, marked cervical constriction, and pulpal obliteration.

Treatment plan

A joint orthodontic/orthognathic approach was decided upon with the following aims of treatment:

1. Orthodontic levelling, alignment and decompensation of the incisors. The spaces in the lower premolar regions were to be maintained. The lower incisors were to be proclined approximately 14 degrees. Significant upper arch expansion was required for correlation of arch form. The expansion was to be carried out orthodontically as most of the discrepancy related to a constriction in arch form in the premolar region.
2. Body ostectomy for set back of the mandible to reduce the mandibular prognathism and close the spacing in the lower buccal segments.
3. Maxillary Le Fort 1 advancement and inferior repositioning of the maxilla (with bone grafting) to correct the maxillary retrusion and increase upper incisor display.
4. Close all spaces accepting the upper permanent canines as lateral incisors by recontouring the incisal edges and restorative build-ups. Additionally, this would lead to a reduced overbite due to canine crown morphology.

Concerns were raised over the risks of enamel fracture, perioperative bleeding, and bone fracture. The risk of

Table 1. Cephalometric values

Angle	Pre-treatment	Post-decompensation	1 year after maxillary surgery
SNA	76°	76°	82°
SNB	86°	86°	77°
ANB	–10°	–10°	+5°
MMPA	14°	12°	17°
UI/Max.Pl	113°	108°	108°
LI/Man.Pl	79°	97°	93°

haemorrhage was further complicated by the patient's religious convictions regarding blood transfusion.

Full records were obtained including study models, facebow record, dental panoramic tomogram, lateral cephalograph (tracing shown Figure 2a), and photographs (Figure 1a–f). A preliminary prediction tracing indicated that a body osteotomy of 10 mm utilizing the spaces mesial to the first molars, a maxillary advancement of 8 mm with an inferior movement of 3 mm was required. It was thought likely that an iliac crest bone graft for the maxilla might be necessary. A joint consultation with the maxillofacial surgeons was arranged at which time the treatment plan was agreed and the patient gave informed consent. In view of the possibility of excessive perioperative haemorrhage, the surgeons planned the mandibular and maxillary osteotomies as separate surgical episodes. It was proposed to reduce the risk of enamel fracture by using bands on all teeth cemented with glass ionomer cement, rather than bonded brackets.

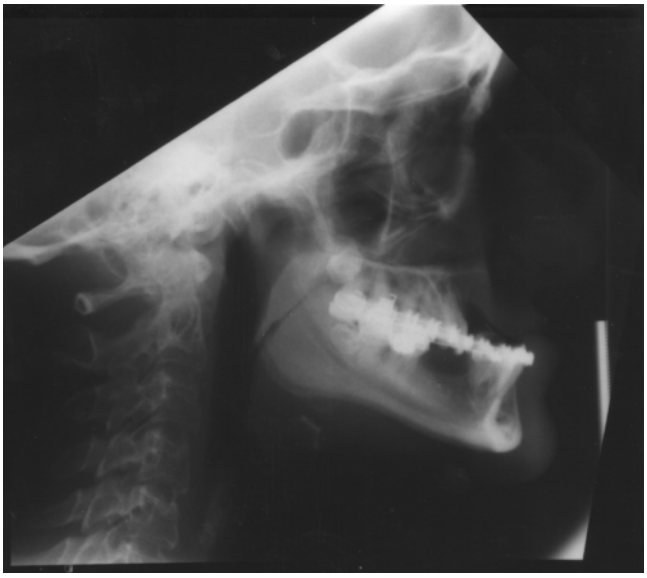

Fig. 3 Post-decompensation lateral cephalograph.

Pre-surgical orthodontics lasted 13 months at which time further study casts, face bow recording and radiographs were taken for the final surgical planning (Figure 3). Surgery to the mandible only was then undertaken. It was felt that the amount of mandibular movement was not entirely predictable due to surgical difficulties relating to a body ostectomy. The secondary maxillary surgery could more predictably fit the new mandibular position. After a 7-month period, the second phase of surgery was carried out with maxillary advancement and inferior repositioning. Bone grafting was not carried out due to the severity of perioperative haemorrhage. Post-surgical orthodontics lasted 7 months, but full interdigitation of the buccal occlusion proved impossible to achieve. The

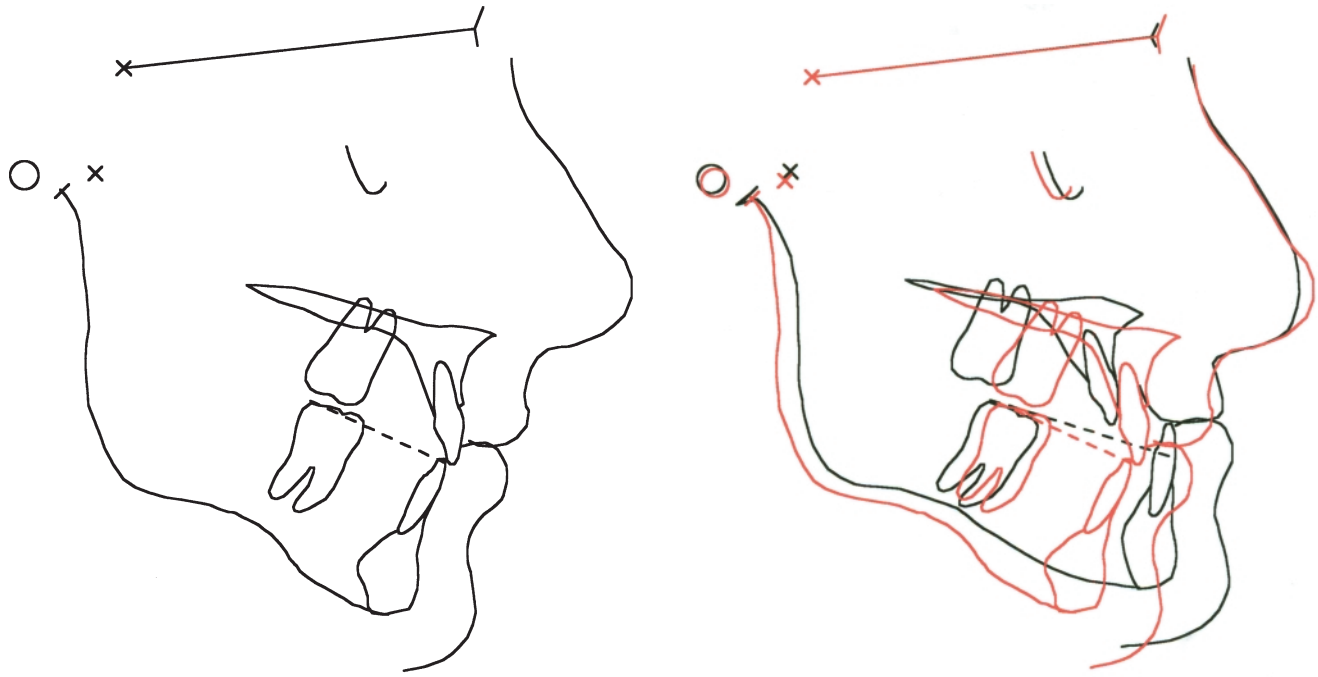

Fig. 4(a,b) Cephalometric tracings: 1 year after maxillary surgery and superimposition of post-treatment on pre-treatment using sella–nasion (registered at sella).

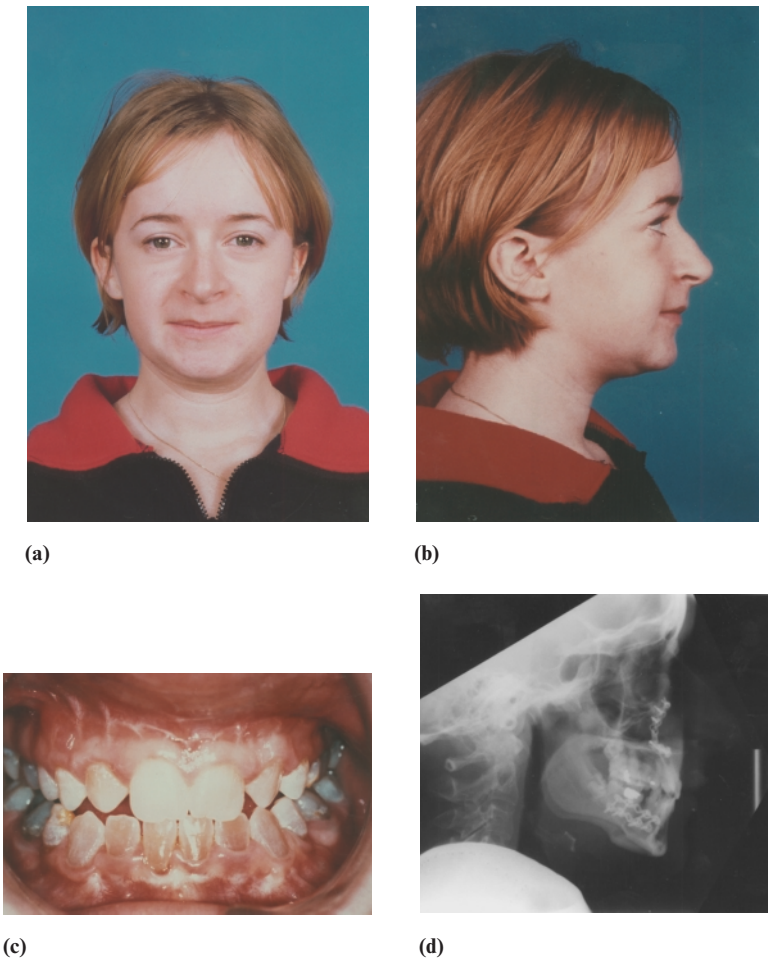

Fig. 5 (a–d) Post-debond photographs and lateral cephalograph (1 year after maxillary surgery).

patient was then debonded and removable retainers fitted. A cephalometric tracing 1 year after maxillary surgery is displayed in Figure 4a. A superimposition of 1 year post-surgery on pre-treatment tracing is shown in Figure 4b. The retention period lasted for 1 year, compliance, however, was unfortunately poor. A review 1 year out of all retention showed that the overjet and overbite remained stable, and that the patient was happy with her appearance (Figure 5a–d). However, a bilateral posterior crossbite had re-established and some spaces had reopened mesial to the lower first molars.

Discussion

This case report highlights a number of issues related to osteogenesis imperfecta that are of relevance to the orthodontist. First, it is likely that such patients will present for treatment of dentofacial deformity typically with a Class III malocclusion often requiring a joint orthodontic/surgical approach. Dentinogenesis imperfecta may be found with varying severity. According to O'Connell and Marini, adhesive dentistry is not contraindicated in such patients and successful bonding of orthodontic brackets can be accomplished.[2] However, although it may be accomplished, the authors did not report whether there was a problem with subsequent enamel fracture.

Surgical treatment, as well as its inherent risks, poses special risks for such patients related to excessive haemorrhage, bone fragility, difficulty intubating and increased risk of developing malignant hyperthermia. As demonstrated by this case report such patients, despite having a

very severe malocclusion, can be successfully treated. This patient exhibited some relapse of upper arch expansion, which detracted from the occlusal result. This relapse was largely due to poor retainer wear and detracted minimally from the overall result. Close collaboration between the orthodontist, surgeon, anaesthetist and the patient's general medical practitioner or hospital physician are essential to establish the severity of the OI and to minimize the risks. Finally, it is important that the patient fully understands the risks and provides informed consent.

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