Management of Multiple Mandibular Fractures in a Child with Osteogenesis Imperfecta Using Arch Bar Retained Thermoformed Splints: A Novel Technique

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Osteogenesis Imperfecta (OI) is a heterogeneous group of autosomal dominant and recessive inherited disorders of type I collagen metabolism. Clinical features of OI include multiple bone fractures, muscle weakness, joint laxity, skeletal deformities, blue sclerae, hearing loss, and dentinogenesis imperfecta. This report presents a challenging case of multiple mandibular fractures in a five years old child with OI, which was successfully treated with a new, minimally invasive technique of closed reduction with arch bar retained thermoformed splint.

***Key words**: Brittle bone disease, paediatric jaw fracture, closed reduction.*

INTRODUCTION

Osteogenesis imperfecta (OI) also known as “brittle bone disease” or “Lobstein syndrome” is a collective term for a heterogeneous group of connective tissue syndromes characterized by bone fragility and fractures. Incidence of OI ranges from 1:5000 to 1:20000; with no racial or ethnic predilection.[1]

Sillence et al classified OI in four groups [type 1 to 4].[2] *Type 1 (Non-Deforming OI With Blue Sclerae)* is characterized by increased bone fragility, multiple fractures, distinctly blue/gray sclerae, and susceptibility to conductive hearing loss commencing in adolescence and young adult life. *Type 2 (Perinatally Lethal OI Syndromes)* is extremely severe form of OI, characterized by short crumpled long bones, bowing deformities of long bones and marked deficiency of ossification of facial and skull bones. Individuals with *type 3 OI (Progressively Deforming OI)* have bone fragility and multiple fractures leading to progressive deformity of the skeleton. Although the sclerae may be blue at birth, it becomes progressively less blue with age. In *Type 4 OI* the patients have recurrent fractures, osteoporosis and varying degrees of deformity of long bones and spine, but normal sclerae.

Pathogenesis of OI is based on alteration in type I collagen synthesis, which is the major protein of bone, teeth, sclera and ligaments.[2] This quantitative and qualitative defect of structural collagen protein of bones increases their fragility. Bone fracture at minimal trauma and subsequent bone deformities are expected in patients with OI. However fracture of long bones are encountered more frequently, as compared to mandible fracture.[3] Pathological modification of collagen synthesis, also leads to ligament laxity, dentinogenesis imperfecta, blue or gray sclera and more rarely, alterations in the mitral valve and aortic dilation.

This paper reports a rare case of multiple mandible fractures in a five year old child with type 1 OI. The case was successfully managed by a new, minimally invasive technique using arch bar retained thermoformed splint.

Case Report

A five years old male child was referred to department of oral and maxillofacial surgery, Krishna hospital, Karad with a chief complaint of pain over his lower jaw of one day’s duration. His parents gave a history of fall while playing on swing, in which child’s face was hit against the floor. Neurological evaluation of the patient showed absence of any cranial injury. The child had thin built, with normal stature and gait (figure1a). Blue sclera involving both eyes was seen (figure1b, c). There was no loss of hearing on auditory evaluation. Past history revealed multiple long bone fractures, which were treated with closed reduction. The history and clinical features were suggestive of type 1 OI.

Extra-oral examination revealed diffuse swelling over right side of lower face (figure 1d), abrasion at sub-mental region (figure 1e) and restricted mandibular movements. Sublingual hematoma and

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derangement of occlusion with mild anterior open bite was evident on intra-oral examination (Figure 2). Mild occlusal step was present in mandibular anterior region, between 73-74. There was no associated dentinogenesis imperfecta. Haematological and biochemical evaluation including serum calcium, phosphorus and alkaline phosphatase levels were within normal range. Radiographs of both extremities showed mild bowing of long bones with areas of callus formation at sites of previous fractures (figure 3).

Orthopantomogram (OPG) showed three linear radiolucent lines involving mandible, distal to tooth bud of 46, in left parasymphysis region apical to developing tooth bud of 33, and in left mid-ramus region. The fracture lines were also noted on lateral oblique view of mandible (figure 4). Diagnosis of Minimally-displaced fracture of mandible in left parasymphysis, left ramus and right body of mandible in patient with OI was established.

Open reduction of mandible fractures could not be undertaken due to risk of damage to the developing tooth buds, possibility of growth disturbance and complication related to OI. Closed reduction with conventional intermaxillary ligation was contraindicated due to mixed dentition, with poor anchorage for arch bar fixation. Conservative management of the mandible fractures with a new, minimally invasive technique using arch bar retained thermoformed splint was planned.

Treatment protocol

After oral prophylaxis, maxillary and mandibular alginate impressions were taken and cast models were prepared. Thermoformed splints were fabricated on both maxillary and mandibular cast models using thermoplastic clear foil [Duran; 1mm X 1.25mm diameter] using pressure molding machine [Biostar,Scheu-Dental, GmbH, 58642, Iserlohn, Germany], at temperature of 220 degree Celsius and 4.8 bars pressure for 30 seconds (figure 5a,b). After its fabrication, the thermoformed splints were cut with the scissors until the depth of labial and lingual vestibule. Small holes were drilled with straight fissure burs on the gingival and occlusal surfaces of the splint for the excess cement to flow out. Arch bars were fixed to the splints over labial aspect with cold cure acrylic resin by sprinkle-on method (figure 5c). The splints were polished and tried in the patient's mouth to check for any overextension. Subsequently the splints were cemented to upper and lower dentition with type I glass ionomer cement. Excess cement squeezed out through the preformed holes was removed using cotton. Intermaxillary elastic ligation (Tru-Force Latex® elastic system, USA; size 3/16", 3.5 ounce) was applied using orthodontic elastics to attain occlusion and immobilise the jaw for healing (Figure 5d). Intermaxillary immobilization was maintained for 2 weeks. After removal of splints, good dento-gingival health and occlusion was achieved. At four months postoperative review patient had satisfactory occlusion (figure 6a) and OPG showed good bone healing with no evidence of fracture lines (figure 6b).

Figure 1: Clinical presentation of the patient with (a) normal stature and gait (b,c) blue sclera involving both eyes. Extra-oral photograph showing (d) swelling over right lower face region, (e) bruise over chin.

Figure 2: Pre-operative occlusion

Figure 3: Radiograph of long bones showing areas of callus formation from previous fractures.

DISCUSSION

Challenges in the management of pediatric mandibular fractures

Various factors which complicate surgical management of jaw fracture in pediatric patients include; small jaw size, existing active bony growth centers and crowded multiple permanent tooth buds in the jaw bone located in close proximity to the mandibular canal and mental nerves.[4]

Open reduction and internal fixation of jaw fracture during mixed dentition causes damage to the permanent tooth buds. This leads to failure of eruption of permanent teeth and narrow atrophic alveolar ridge. Rigid fixation can also cause growth disturbances due to incomplete ossification of jaw bones. Removal of metallic fixation device is therefore recommended after fracture healing is complete, requiring a second surgical procedure. Stress shielding causes weakening of the bone after removal of the implant. Corrosion of metal ions is other indication for removal of the fixation device.[5]

Application of intermaxillary fixation (IMF) for closed reduction of mandible fracture is difficult because of weak tooth anchorage due to physiologic resorption of deciduous tooth roots and incompletely formed permanent tooth roots. These teeth can easily get avulsed due to the pressure exerted onto the tooth during wiring. Retention of dental wiring is further compromised by bell shaped deciduous crowns with little undercut area. The wires also cause discomfort to the child and damage the periodontal tissues as they tend to move apically causing inflammation.

Jaw fractures in patients with OI

Multiple fractures occurring spontaneously or secondary to minor injury is the most common feature of OI. Greater mechanical load on long bones of the body make them prone to fracture. The incidence of long bone fracture in OI is as high as 80%. However fractures of the jaw bones are less frequent, with reported incidence of 1-2% .[6]

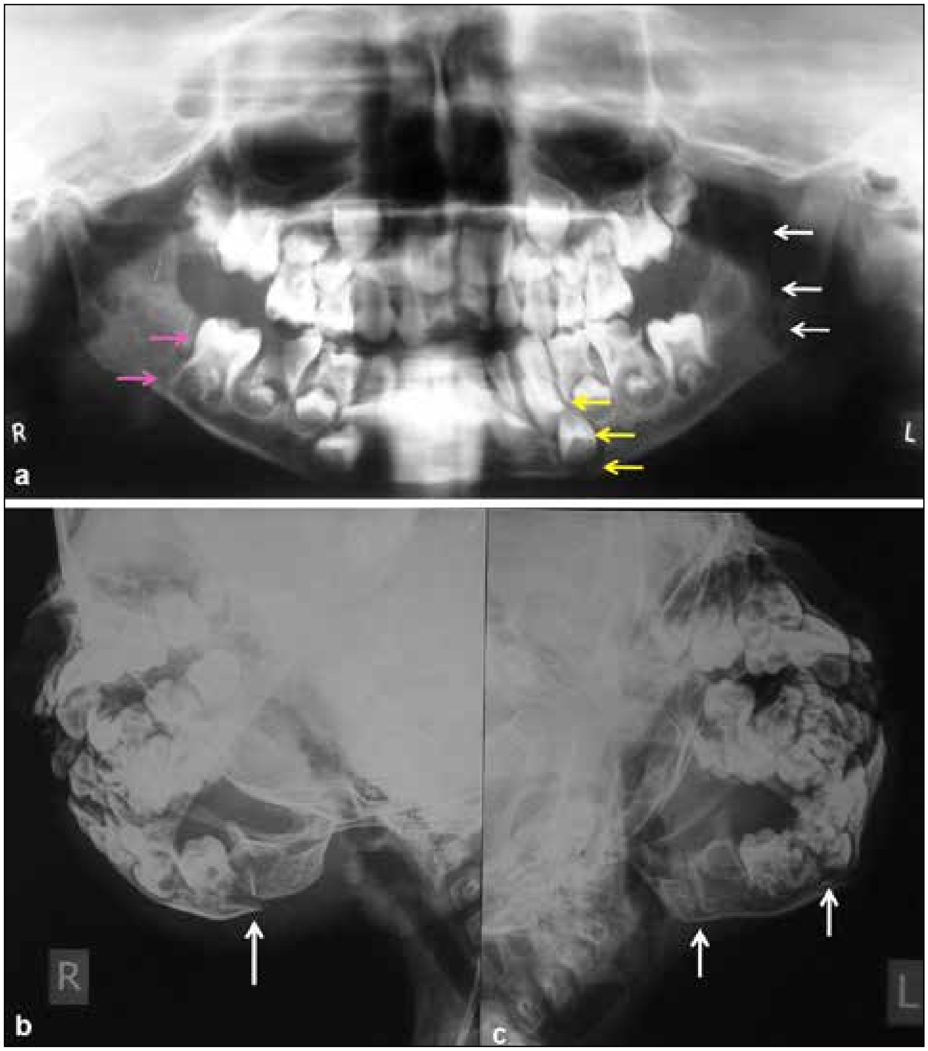
Poor quality, mass and geometry of osseous tissue in OI adversely affect the bone strength. The relatively immature, disorganised and hypermineralized bone makes it prone to fracture. Fracture healing in OI is characterised by formation of new bone which is often osteoporotic in nature with poor trabeculated spongiosa. The newly formed bone is deficient in osteoid material with tendency of multiple micro-fractures.[7]

Special considerations for surgical management of jaw fracture in patients with OI include; compromised bone healing due to altered periosteal microvasculature, increased risk of bleeding caused by qualitative platelet disorders and microvascular endothelial defects, and anesthetic complications.[7] Poor quality of newly formed jaw bone after fracture fixation, increases the risk of non-union or mal-union. Healing can be delayed and scars are often poor. Increased capillary fragility and functional platelet abnormality increase the risk of bleeding during the surgery. [8, 9] General anesthesia in these patients with OI is complicated by difficult airway management, due to existence of spinal deformity and respiratory disorders. Changing patient position during intubation increases the intra-operative risk of mandible and cervical fractures. Co-existing cardiac anomalies, thrombocyte function disorder, risk of malignant hyperthermia, and metabolic disorders further increase the risk of anesthesia in these patients.

Advantages of the new thermoformed split technique

Thermoformed splints are made of same material and system which is used for fabricating periodontal splints, night guards, bleaching trays and bite splints. Use of thermoformed splint in the management of mandible fracture was first reported by Lloyed et al in 2001.[10] The procedure could be carried entirely in out-patient unit with minimal discomfort. Modification of the thermoformed splint by joining upper and lower units to attain IMF was later reported by Terai in 15 patients with unilateral condylar fracture.[11] Transparency of the splint permitted easy checking of occlusion. Unlike dental wiring which increases the risk of puncturing operator's skin and injuring the patient's periodontal tissue, the smooth surface of the splint allowed easy handling and was comfortable to wear.

Figure 4: (a) Orthopantomogram showing fracture at right angle, left parasymphysis and ramus of mandible (b) fracture lines evident on lateral oblique view of mandible



However, the fixation strength of thermoformed splint was found to be less compared to wiring, contraindicating its use in significantly displaced fractures. In our case the splint was modified by incorporating Erichs arch bar to the labial aspect of the splint. The hooks of the arch bar allowed application of rubber elastics for achieving traction as well as for IMF. The direction and magnitude of fixation could be simply varied and quickly done at every visit as required. Patient compliance was good. The flexibility and transparency of splints provided comfort and esthetics during the fixation period. No local anesthesia was required for splint placement and IMF, making it completely atraumatic and non-invasive procedure. The labio-lingual extension of the splint with excellent tooth-gingival adaptation stabilizes the fracture and prevents any vertical or horizontal displacement.

The even occlusal thickness of splints increases the joint space partially, thus preventing chances of ankylosis.[12] Glass ionomer luting cement (GIC) used for splint cementation provides adequate strength for its retention. The fluoride released from GIC is known to prevent decalcification, thus providing an added benefit of reducing the risk of dental caries. The smooth and clear surface of splint also decreases plaque accumulation, when compared to arch bar and dental wiring. In the present case intermaxillary fixation was maintained using thermoformed splints for two weeks and stable occlusion was achieved. The panoramic radiograph taken four months after treatment showed complete healing of mandible bone at fracture areas.

CONCLUSION

Management of mandibular fractures in pediatric patients with medical conditions like OI contraindicate invasive surgical procedures and intermaxillary fixation with dental wiring. The clinical outcome of the present case indicates that using intermaxillary ligation with thermoforming splint is safe, easy, effective and reliable treatment modality in such situations. The technique is non-invasive and improves child patient comfort and compliance.

Figure 5: (a) Thermoplastic clear foil, (b) Biostar pressure molding machine used to fabricate thermoformed splint. Fabricated splint lower arch and upper arch (c), splint cemented to the dentition with elastic traction applied (d).

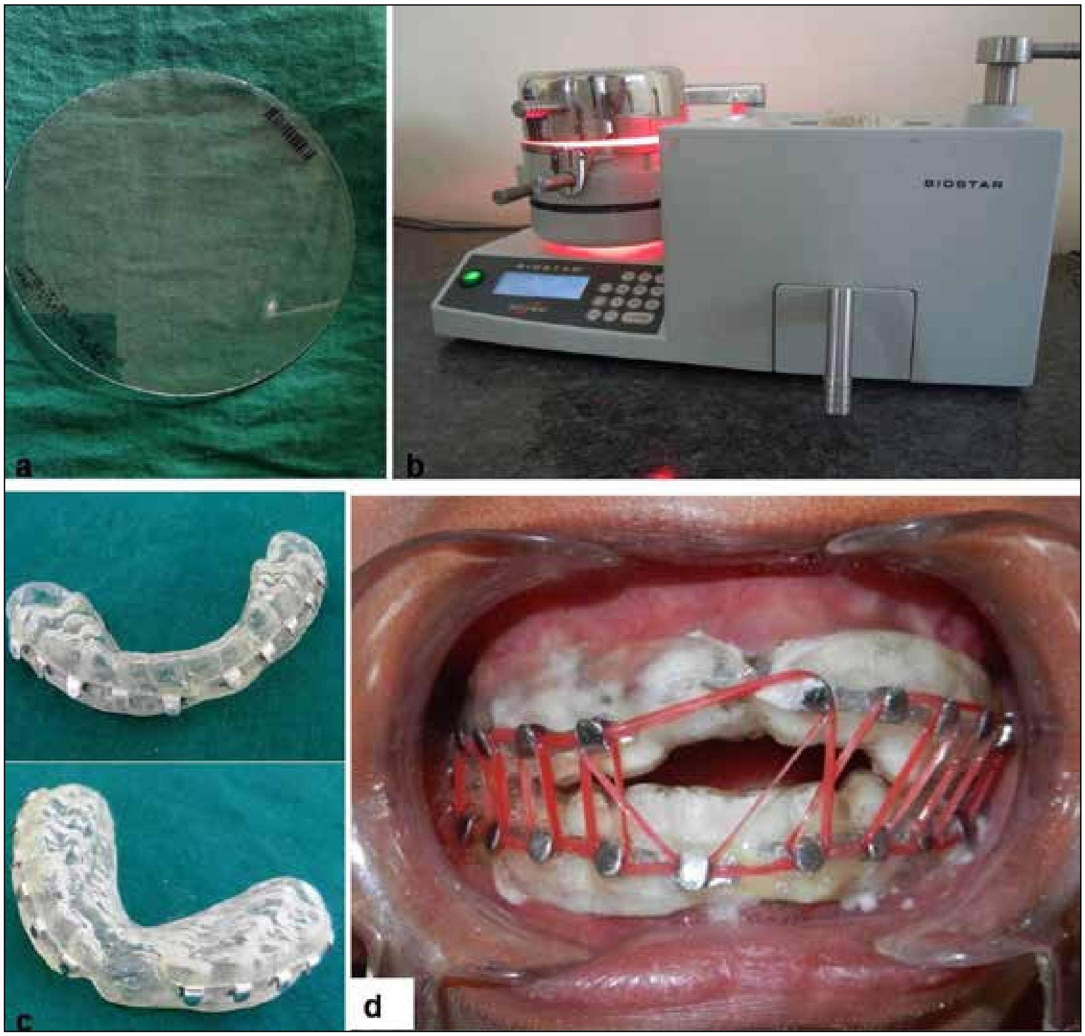
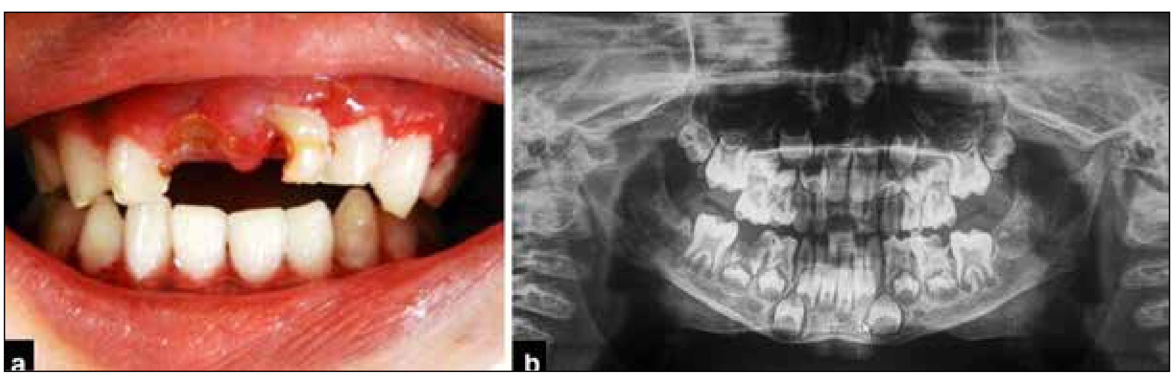


Figure 6: Intraoral view showing stable occlusion (a) and OPG showing good healing at four months postoperative period



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