Original Article

# Results of a bone splint technique for the treatment of lower limb deformities in children with type I osteogenesis imperfecta

*Dasheng Lin, Wenliang Zhai, Kejian Lian, Zhenqi Ding*

## Abstract

**Background:** Children with osteogenesis imperfecta (OI) can suffer from frequent fractures and limb deformities, resulting in impaired ambulation. Osteopenia and thin cortices complicate orthopedic treatment in this group. This study evaluates the clinical results of a bone splint technique for the treatment of lower limb deformities in children with type I OI. The technique consists of internal plating combined with cortical strut allograft fixation.

**Materials and Methods:** We prospectively followed nine children (five boys, four girls) with lower limb deformities due to type I OI, who had been treated with the bone splint technique (11 femurs, four tibias) between 2003 and 2006. The fracture healing time, deformity improvement, ambulation ability and complications were recorded to evaluate treatment effects.

**Results:** At the time of surgery the average age in our study was 7.7 years (range 5-12 years). The average length of followup was 69 months (range 60-84 months). All patients had good fracture healing with an average healing time of 14 weeks (range 12-16 weeks) and none experienced further fractures, deformity, or nonunion. The fixation remained stable throughout the procedure in all cases, with no evidence of loosening or breakage of screws and the deformity and mobility significantly improved after surgery. Of the two children confined to bed before surgery, one was able to walk on crutches and the other needed a wheelchair. The other seven patients could walk without walking aids or support like crutches.

**Conclusions:** These findings suggest that the bone splint technique provides good mechanical support and increases the bone mass. It is an effective treatment for children with OI and lower limb deformities.

**Key words:** Cortical strut allograft, internal fixation, osteogenesis imperfecta

## Introduction

Osteogenesis imperfecta (OI) is a genetically heterogeneous bone disorder caused by defects in the structure and function of type I collagen.[1] OI is clinically characterized by brittle bones, osteopenia, defective dentition, blue sclerae, loose joints and abnormal spinal curvatures.[2] Children with OI can suffer from frequent fractures and limb deformities, resulting in impaired ambulation. Their osteopenia and thin cortices complicate orthopedic treatment. Treatment consisting of plate fixation alone results in high rates of refracture at the ends of the plate due to stresses.[3] Intramedullary fixation is also associated with bone fractures, migration of implants, penetration into the joint and hardware failure.[4-6] Although the cortical strut allograft has been widely used to provide mechanical support and supplement bone healing in revision hip arthroplasty and osseous defects,[7-11] its use in the treatment of lower limb deformities resulting from OI, in children, has not been reported in literature.

This study evaluates clinical results of a bone splint technique consisting of a combined fixation of internal plating and cortical strut allograft, for the management of lower limb deformities in children with type I OI.

## Materials and Methods

Nine children (five boys, four girls) with lower limb

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| Access this article online | |
|---|---|
| Quick Response Code: | Website: www.ijoonline.com |
| | DOI: 10.4103/0019-5413.114922 |

deformities due to type I OI were treated at our center using the bone splint technique between 2003 and 2006. There were eleven femurs and four tibias. The Ethics Committee of our institution approved this study and parents or legal guardians gave informed consent before surgery.

The average age of the children was 7.7 years (range 5-12 years). All children had type I OI, based on the Sillence classification for OI.[12] All the patients had multiple fractures before surgery (average 3.9 times, range 2-6 times). We used a method to measure the magnitude of limb deformity. The lines represent the axes of the proximal and distal parts of the bone on anteroposterior and lateral radiographs and the larger angle is the magnitude of the limb deformity. The femoral deformity in these patients ranged from 50° to 70° (average 59°) and the tibial deformity from 17° to 25° (average 21°). Three children were confined to the bed or wheelchair, four were confined indoors and two were confined to community ambulation. Two children had a family history of OI [Table 1].

Preoperative physical examination was done and radiographs taken to precisely measure the shaft length and the degree of deformity and to map out the exact size of the bone wedge to be removed. The operation was done under an epidural anesthesia or general anesthesia. The patient was positioned on the fracture table to allow use of a C-arm machine. The strut allografts were made with freeze-dried allogeneic long bones (Xinkangchen Medicine Development, Beijing, China).

## Operative procedure

The site of the femoral deformity was approached using an anterolateral incision. The failed implant (if any) was removed. Wedge osteotomies were performed to correct deformity. Coxa vara and limb torsion (if present) were corrected together. The bone of an OI patient often bends anterolaterally. In such a case the shaft needs to be cut completely, not leaving the contralateral cortex, but leaving the periosteum intact where possible, to preserve the blood supply to the bone. A plate is placed on the lateral side of the femur to span the whole extent of the deformity. Two screws were put on each end of the plate. Then a cortical strut allograft (range 9-18 cm) was placed opposite the plate to span the weak part of the shaft. The cortical strut was normally two-thirds the length of the plate. The plate and the strut graft are stabilized with screws [Figure 1].

The surgery for the tibia was similar to that for the femur. With the aid of the C-arm and tourniquet, the site of the tibial deformity was approached using an anterolateral incision. Wedge osteotomies were performed until the deformity was corrected. The plate and the strut graft were placed and stabilized with screws [Figure 2].

No external fixation was used. The patients were encouraged to perform gentle and protective range-of-motion exercises from the first day after surgery. The patients, without any other problems, were discharged one week after surgery and were called back for followup six weeks, three months, six months, one year and two years after surgery. The patients were rapidly mobilized and were instructed to use toe-touch weight bearing with crutches or a walker from

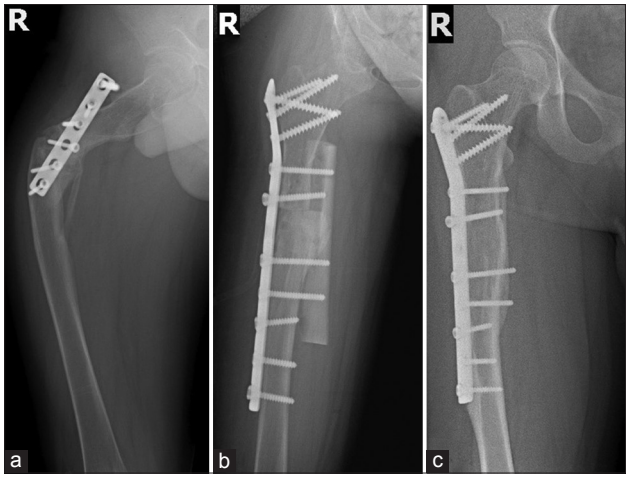


**Figure 1:** X-ray (R) thigh with hip joint anteroposterior view showing (a) A 12-year-old boy with type I OI who had four fractures before he was treated with the bone splint technique. Radiography showed a 65° bending angle of the femur and loosening of screws. (b) Correction of the femoral deformity with the bone splint technique. (c) Good healing of the fracture and bony union between the cortical strut allograft and the host bone, two years after surgery

**Table 1: Clinical details of the patients**

| Patient | Age | Sex | Limb with deformity | Previous fractures | Bending angle (°) | Followup (mo) | Healing time (w) |
|---|---|---|---|---|---|---|---|
| 1 | 6 | F | One femur | 2 | 58 | 69 | 13 |
| 2 | 9 | M | One femur+one tibia | 4+2 | 54+19 | 84 | 14+16 |
| 3 | 6 | F | One tibia | 4 | 25 | 63 | 15 |
| 4 | 5 | M | Two femurs | 2+1 | 68+62 | 72 | 12+12 |
| 5 | 9 | F | Two femurs | 1+3 | 50+56 | 75 | 13+14 |
| 6 | 12 | M | One femur | 4 | 65 | 60 | 16 |
| 7 | 7 | M | One tibia | 2 | 17 | 72 | 15 |
| 8 | 7 | F | Two femurs+one tibia | (3+1) + 2 | (70+52) + 23 | 66 | (15+15) + 15 |
| 9 | 8 | M | Two femurs | 2+2 | 55+61 | 60 | 12+12 |

one week after surgery. The patients progressed to full weight bearing when they had the strength and balance to do so. They were instructed to leave the crutch support when they were able to walk without a substantial limp. Cyclical intravenous pamidronate administration was given to all patients on three consecutive days, every four months. The dose was one milligram per kilogram of body weight per day. The dose was reduced by half during the first day of pamidronate treatment, then full doses 2nd and 3rd day.[13]

### Postoperative evaluation

The clinical and radiological outcomes of all subjects were evaluated.[14] The clinical evaluation covered healing of bone fractures, improvement in deformities, incidence of infection and degree of ambulation (i.e., confined to bed, wheelchair, indoors, community, or independently walking). The radiological evaluation included the callus gray density, position, migration, bending and breakage of internal fixation and refracture rates.

## Results

All the patients had a followup period of at least five years and the mean followup was 69 months (range 60-84 months). All the surgical wounds healed with primary intention. Allograft rejection, neurovascular complications or infections did not occur in any patient. All the patients healed well, with an average healing time of 14 weeks (range 12-16 weeks) and the callus gray density increased with the passing of time. None of the patients had refractures, deformity or nonunion. The fixation remained stable throughout the followup in all cases, with no evidence of loosening or breakage of screws.

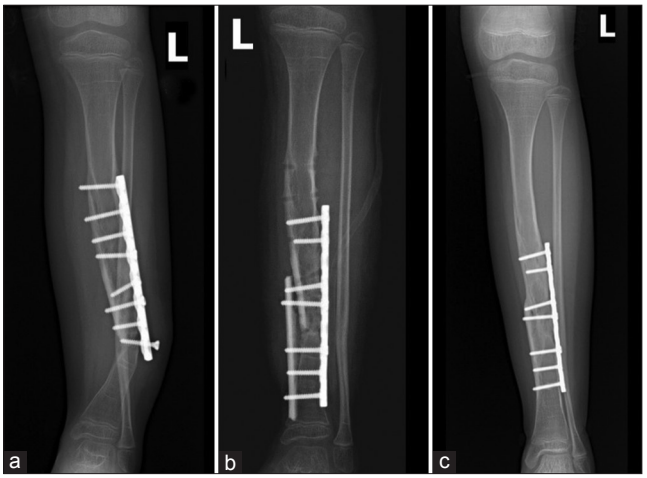


**Figure 2:** X-ray (L) leg bones anteroposterior view showing (a) A six-year-old girl with type I OI who had experienced four fractures before she was treated with the bone splint technique. Radiography showed a 25° bending angle of the tibia, aggravation of the deformity and loosening of screws before surgery. (b) Correction of the tibial deformity with the bone splint technique. (c) The bony union between the cortical strut allograft and the host bone at two years after surgery

One year postsurgery, the allografts were incorporated into the host bones. The medullary cavity junction between the allograft and the host bone was recognized [Figures 1 and 2]. After surgery, the deformity and mobility significantly improved ($P < 0.05$, Table 2). Of the two children confined to bed before surgery, one was able to walk on crutches and the other needed a wheelchair. The other seven patients could walk without support.

## Discussion

Osteopenia and local deformities are common in children with OI and often associated with multiple fractures. Reposition and plaster cast fixation can result in satisfactory results temporarily. However, long term immobilization results in further reduction in bone mass and poor bone stock, thereby increasing the risk of further fractures and worsening deformities. Thus, a vicious cycle of fracture, immobilization and refracture set in.[15]

Saldanha *et al.*[16] have reported the use of external fixation to correct limb deformity. However, children with OI often have associated osteopenia and thin cortices and fixator screws do not have enough purchase in such bones. Implant loosening, fracture displacement or more severely, recurrent fractures frequently occur in these cases. Similar problems exist when internal bone plating is used alone. In addition, the stress shielding effect associated with bone plating further leads to reduction in bone density, aggravating the osteopenia.[3]

All the patients in our study had undergone unsuccessful orthopedic operations before they had been referred to our institution. Four had been treated with plating (four femurs, two tibias) and two with external fixation (three femurs). Refractures occurred shortly after these unsuccessful operations and the deformities were aggravated.

The mainstay of treatment for deformity in the lower limb caused by OI are multiple osteotomies and intramedullary nailing, which can correct the deformity, increase limb strength, reduce the risk of further fractures and increase limb length. There is wide variability in the types of intramedullary

**Table 2: Preoperative and postoperative walking ability of patients**

| No. of patients | Confined to bed or wheelchair | Indoors | Community | Independently walking |
|---|---|---|---|---|
| Preoperative | 3 | 4 | 2 | 0 |
| Postoperative | 1 | 0 | 1 | 7 |

rods and their clinical results.[4-6,17-20] With bone growth, the rod becomes relatively shorter and require revisions when the patient outgrows the rod. The complications of intramedullary fixation include fractures, migration of implants, joint penetration and hardware loosening or disengagement.

In our study, lower limb deformities in children caused by type I OI were treated with osteotomies and internal plating, combined with cortical strut allografts. This technique effectively rebuilt the thin cortex and helped the screws gain better purchase on the bone, thus preventing the fixation from loosening.[21] The screws were inserted through three layers of cortices to achieve a so-called bone splint technique composed of the steel plate, screws and allograft. The bone splint technique shared the load with the bone and decreased the bending and torsion stress on the plate. As a result, the entire mechanical performance of the fixation was increased and the risks of fracture at the ends of the implant or internal fixation failures were reduced. Due to the stable fixation, the creeping substitution of the allograft and bone healing proceeded smoothly. After the union occurred, the allograft underwent adaptive remodeling secondary to physiological load bearing.[22] In addition, the bone splint technique blocked the cartilage from intruding into the fracture section, favoring the healing of the bone fracture. In our study, during the 69-month followup, no patient had failure of internal fixation. Radiography showed that 12-16 weeks after surgery, the bone healed well. One year after surgery, the allograft was mostly incorporated into the host bone. The bony union of the allograft and the host bone greatly increased the bone mass and improved the strength of the bone.

It has been reported that Pamidronate increases bone density and reduces the incidence of fractures and complications.[23-26] In our study, cyclical intravenous Pamidronate administration was concomitantly used to improve bone mineral density, facilitate greater activity of patients in the early stage and reduce the risks of complications.

Limitations of this study include the small cohort of patients and the lack of a control group. Although many problems associated with cortical strut allografts (e.g., immune rejection and infection) have been solved, there is still a controversy and limitations to their use, especially regarding the potential donor shortage.

The bone splint technique described herein can provide additional support to the host bone, decrease stress on the implant and reduce the risks of recurring fractures or fixation failures. After bony union of the allograft and the host bone occurs, the bone mass increases and its biomechanical strength is improved. In conclusion, this bone splint technique is an effective treatment for lower limb deformities in children with type I OI.

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**How to cite this article:** Lin D, Zhai W, Lian K, Ding Z. Results of a bone splint technique for the treatment of lower limb deformities in children with type I osteogenesis imperfecta. Indian J Orthop 2013;47:377-81.

**Source of Support:** This study was supported by the key projects from Nanjing Military Region during the 11th Five Year Plan Period (No. 06MA97)., **Conflict of Interest:** None.