Original Article

Outcome of Shish Kabab Osteotomies Fixed With Sign Naili in Osteogenesis Imperfecta. Our Experience at Ghurki Trust Teaching Hospital, Lahore, Pakistan

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Objective: To evaluate the results of shish kabab osteotomies fixed with SIGN NAIL in long bone deformities of patients suffering from osteogenesis imperfecta.

Methods: This study was carried out on 12 patients who presented in OPD or Emergency Department with deformities of long bones. Detailed history, examination and investigations including X-ray AP and Lateral view of involved bone were done. All these cases were treated with Shish Kabab Osteotomies and SIGN NAIL under spinal or general anaesthesia. These cases were followed up for up to average of 20 months (range 06 months -6 years) and results of the nail were observed in terms of union, rehabilitation and complications.

Results: It was found that 8 (66.7%) were male and 4 (33.3%) were females with a mean age of 14.83±5.24. 5 (41.7%) patients having age 10-15 years and 7 (58.3%) were between 16-20 years. Total 21 surgeries performed. 3(25.0%) had surgery on unilateral femur,2 (16.7%) on unilateral tibia, 3(25.0%) on both unilateral tibia and femur,3 (25.0%) had surgery on bilateral tibia and 1(13.3%) with bilateral tibia and femur. The parents of all patients had a first degree consanguineous marriage. The mean hospital stay were 4.17±1.75 days. All osteotomies sites heal within 06 months, better bone densities, 1 Having recurrence of deformity and no fracture were found. Only two patients using walkers while remaining need no support for walking.

Conclusion: Sign Nail along with shish kabab osteotomies in deformities of osteogenesis imperfecta is the treatment of choice. Patient rehabilitation is early, hospitalization is short, and osteotomies site healing response is good as well as decrease risk of fracture.

Keywords: Osteogenesis imperfecta, SIGN Nail, Shish Kabab, Osteotomy.

Introduction

Osteogenesis imperfecta, also known as "brittle bone disease," is a genetic disorder that affects collagen. Worldwide, osteogenesis imperfecta affects approximately 1 of every 20,000 births. In the United States, it affects between 25,000 and 50,000 individuals and is classified as an orphan disease (ie, a disease that affects fewer than 200,000 individuals nationwide).[1] The first recorded case of osteogenesis imperfecta was in a partially mummified body discovered in ancient Egypt, but it was not until 1835 that the term was coined and the disease was truly understood. Lobstein was the first to correctly identify the pathophysiology of the disease and first termed the disorder Lobstein's disease. The name was later changed to osteogenesis imperfecta, which means "imperfect bone formation.".[2] It is a heritable disorder that is characterized by bone fragility and reduced bone mass. Severity varies widely, ranging from a lethal form with intrauterine fractures to a very mild form with no or few fractures and normal growth. Extraskeletal manifestations include blue sclera, dentinogenesis imperfecta, skin and ligament hyperlaxity, and presence of wormian bones within cranial sutures.[3] Since each person/family with OI has their own unique mutation the management of these children must be individualized. [4]Fractures are common in children, and the commonest cause is osteogenesis imperfecta (OI).[5] Diagnosis is usually clinical. History should include attention to fractures, back pain, motor development and family background. Examination should focus on the skeleton, including the spine, and on identifying other features which support the diagnosis, including scleral hue, teeth and ligamentous laxity. Other investigations including plain radiography, transiliac bone biopsy and genetic analysis which may support the diagnosis.[6] The current classification into four major subgroups (types IIV), based on clinical findings, was proposed by Sillence *et al* in 1979. This was based on clinical and radiological findings of OI.[7] Type I includes patients who have the mild form, almost normal stature, and blue sclera. Type II is considered the most severe form and is lethal in the prenatal period. Type III includes patients with the classic disease manifestation, usually with moderate deformity at birth, and progressively deforming bones. Type IV includes patients with extensive phenotypic variability, including mild to severe forms

of OI .[8] It may also be classified as; Osteogenesis Imperfecta Congenita-usually death occur in utero and Osteogenesis Imperfecta Tarda-manifest in childhood or adolescence. Patients with this disorder suffer from brittle bones that may lead to frequent recurrent fractures.[9] It also occur in two forms. The classic clinical forms of OI comprise Lobstein's type and Vrolik's type. The first has a variable symptomatology, with a greater or lesser degree of deformity and onset of fractures during growth and adulthood. The second is a severe form that is observable from birth, with frequent intrauterine fractures and a high mortality rate[10] Management of OI is multidisciplinary. The standard of care includes pain management, therapy input for muscle strength and range of movement, aids to daily living and mobility, psychologic and social support, and regular monitoring of dentition and hearing.[11]

Sofield and Millar introduced the concept of multiple osteotomies and IM rodding to realign bones for children with OI in 1959.[12] This so called 'shish kebab' technique reduced the frequency of fractures and achieved an acceptable correction of deformity of the extremities. Nonexpendable rods such as Rush nails, William's and Kuntscher's rods were used for the 'shish kebab' technique. These rods can be outgrown resulting in angulation and fracture in areas of the bone that are no longer splinted beyond the extremities of the rod. [12] In an effort to solve this problem, Bailey and Dubow designed the first expandable telescopic rod in 1963[13] . As anticipated, the time interval between the initial operation and revision has increased remarkably with the use of this device.[13]

Our goal was early mobilisation of the patient without external support. We therefore planned intramedullary fixation with a SIGN nail and upto best of our knowledge it is the first study in which the SIGN NAIL had been used .

Methods

This prospective study was carried out at Orthopaedics department of Ghurki Trust Teaching Hospital, Lahore from Jan 2009 to July 2016 after the approval by institutional medical ethics committee. A total of 12 patients of either sex and upto age of 20 years with Osteogenesis Imperfecta with lower limb deformities were included in the study while children with upper limbs, spinal, and other extra osseous deformities ,in whom other implants used were excluded from the study. After history, examination , investigations ,pre operative anesthesia fitness and written informed consent ,the patients underwent surgery. The patients were followed on regular basis with mean follow up of 20 months. The longest follow up was of 6 years and the minimum follow up of 6 months and in each follow up, they were clinically and radiologically assessed for fracture healing, joint movements and implant failure. According to the criteria the results are graded as excellent when the fractures unites within 16 weeks. without any complications, good when union occurs within 24 weeks with treatable complications like superficial infection and knee stiffness and poor when union occurs before or after 24 weeks with one or more permanent complications like infection (osteomyelitis), implant failure, non-union, limb shortening and permanent knee stiffness. Delayed union was recorded when the fracture united between three to six months while nonunion was noted when union had not occurred after eight months of treatment. All the patients were given proper regimen of bisphonates according to their weight and duration as advised by the paediatrician. The preoperative and postoperative gait ability was assessed by dividing the patients into two groups of ambulating and no ambulating. All the patients were non ambulatory preoperatively, but after the operation they were being ambulated with support like walker with full weight bearing. The surgical procedure was carried out by using the same technique as described by Sofield and Millar, which consists of exposing long bones and preserving periosteum, performing multiple bone osteotomies between proximal and distal metaphyses using an electric saw, achieving bone straightening. Proper size SIGN Nail were passed with proximal and distal static locking done. The data were initially entered on a pre formed proforma and later on SPSS 17.0 version.

Results

It was found that 8 (66.7%) were male and 4 (33.3%) were females with a mean age of 14.83±5.24. 5 (41.7%) patients having age 10 - 15 years and 7 (58.3%) were between 16-20 years. Total 21 surgeries were performed. 3(25.0%) had surgery on unilateral femur,2 (16.7%) on unilateral tibia, 3(25.0%) on both unilateral tibia and femur,3 (25.0%) had surgery on bilateral tibia and 1(13.3%) with bilateral tibia and femur. The parents of all patients had a first degree consanguineous marriage. The mean hospital stay The average follow up was 20 months (6 months 6 years).

All osteotomies sites heal within 06 months, better bone densities. Overall the results were excellent in 9 patients and good in 3 patients while considering the union. One patient having recurrence of deformity with bending of nail after 3 years but no fracture were found. Only two patients now using walkers because of having generalized weakness and at last follow up ,there ambulation without support were started at physiotherapy department. **(Table-1)** limb.

In the present study, the procedure has been shown not to interfere with physis, because we had no patient with physeal changes.

We didn't find growth disorders inherent to the adopted procedure. length discrepancy were not found in any patient. The children as well as parents were satisfied with their overall progress and improvement in ambulatory status. were 4.17±1.75 days.

Table-1: Summary of the study;

Mean Age		14.83±5.24 years
Average Follow up		20 month (6months-6 years)
Sex	Male n (%)	8 (66.7%)
	Female n (%)	4 (33.3%)
Mean Hospital stay		4.17±1.75 days
Outcome of surgery	Excellent	9 (75.0%)
In term of union	Good	3 (25.0%)
	Poor	0
Ambulatory status	Ambulatory without support	10 (83.33%)
	Ambulation with support	2 (16.67%)

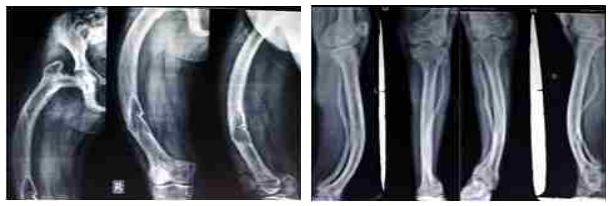

Fig-1a,b: Pre Opp xrays of young female suffering from bilateral tibia and femur deformity.

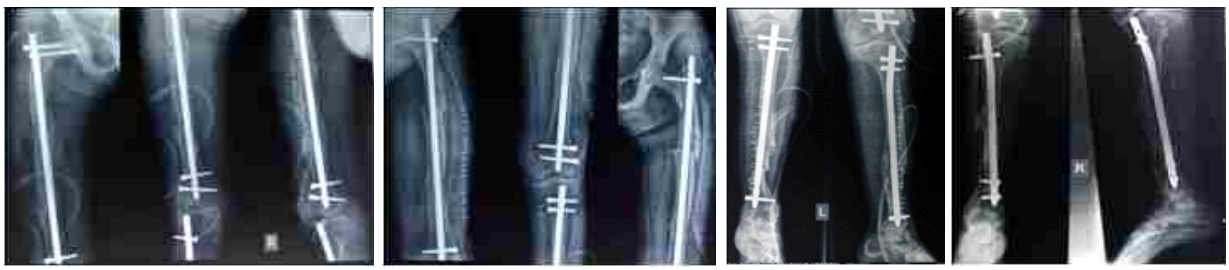

Fig-2 a,b,c; Immediate Post opp x-rays of young female with Shish kabab Osteotomies with SIGN nail in bilateral tibia and femur.

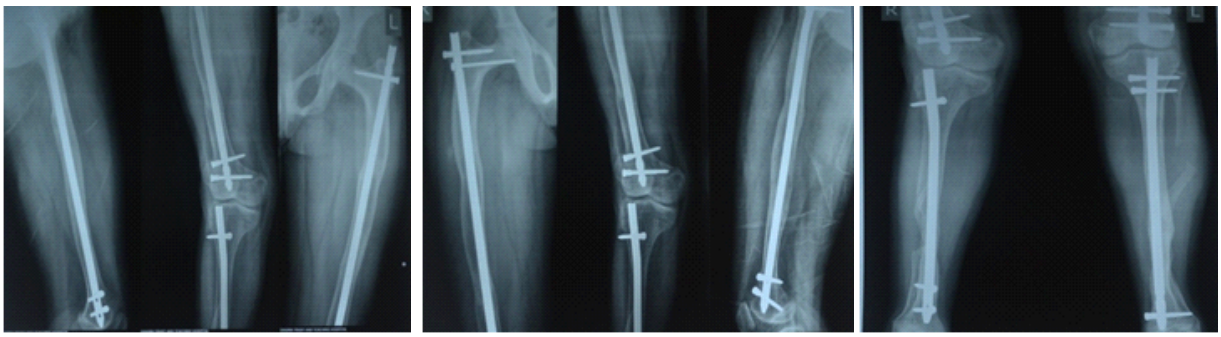

Fig 3 a,b,c; Post opp xrays after 3 months of follow up

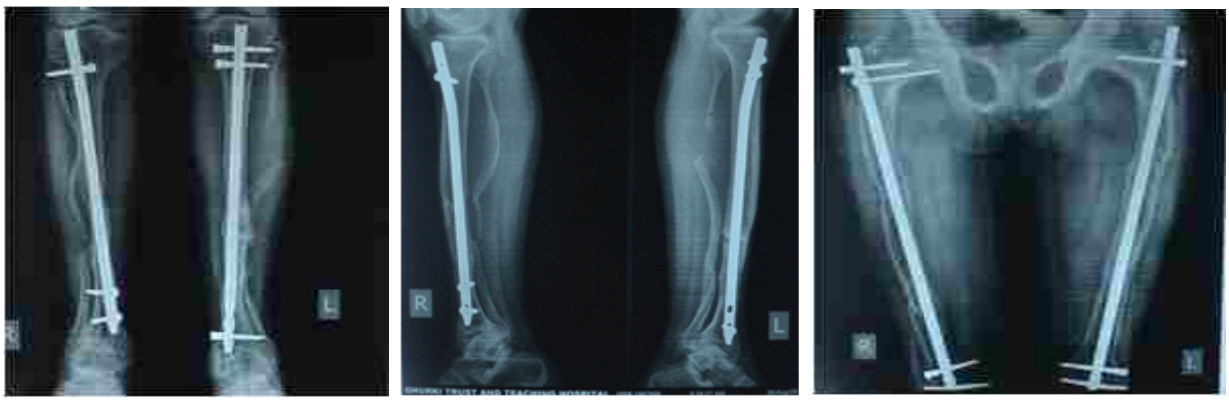

Fig 4 a,b,c: Post opp x-rays after 6 months.

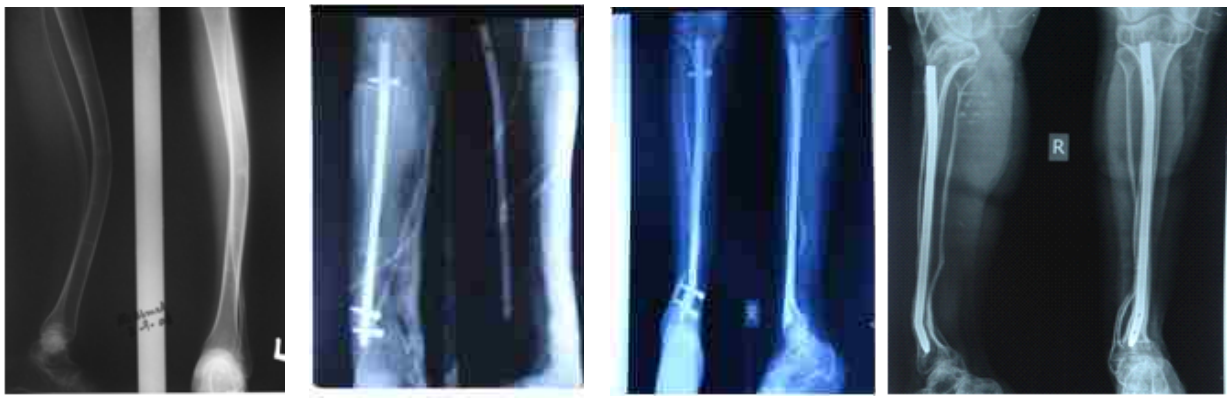

Fig 6 a,b,c ,d;19 years old male with pre ,immediate post opp, after 3 years and after 6 years xray.Note the bending of SIGN Nail.

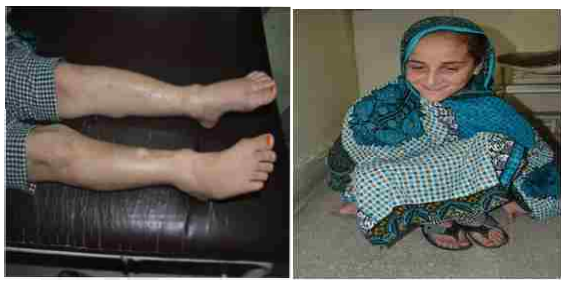

Fig-5 a,b: Post opp follow up images.

Discussion

Conventional management for patients with OI consisted of supplementation with vitamin D and minerals, or administration of bisphosphonates However, these methods did not ameliorate the symptoms of OI .Moreover, in young teenage patients the bisphosphonates cannot be used continuously, because they slow down bone turnover and as consequence child's growth and healing of eventual bone fractures .Another option of supportive treatment of OI patients is surgical and orthopedic intervention or protection with the use of braces, steel rods and other orthopedic equipment .[14] Children and adults with OI will also benefit from maintaining a healthy weight, eating a balanced diet and avoiding activities like smoking or excessive caffeine intake or alcohol consumption or steroid medication. Such things deplete the bone mass making them more fragile and thereby susceptible to fractures.[15] Regarding the surgical management,Intramedullary fixation rather than plating is preferred, and allowing early protected weight bearing and rehabilitation of children with ambulatory potential is the ideal goal.[16] The IM devices are broadly classified into two major categories: the Static rods (Williams' rods and Rush nails) and the telescoping or elongating rods (Sheffield, Baily-Dubow, and Fassier Duval). The choice of IM device is influenced by the clinical features, efficacy of the implant in preventing fractures, the frequency of complications, the technical details of surgery and the cost of the implant. Healing of fractures and osteotomies usually is satisfactory in Osteogenesis imperfecta, although the healed bone may be no stronger than the original.[17] The male to female ratio of OI in our study were 2:1 (8 male and 4 female), indicating that there is gender predilection in OI. Lin et al reported a male to female ratio of 1:2 (15 male and 33 female) [18]. Patel et al [19] showed a 1:1.3 male to female ratio and Plotkin et al [20] reported a 1:0.9 male to female ratio. 4 (33.33%)had a positive family history of a first degree relative with Osteogenesis Imperfecta. Greeley et al reported the presence of family history in 46% of patients OI [21]. Patel et al reported a family history of 40.3% across all types of OI in a 47 cross-sectional multi-centre study [19]. The reason for the reported lower percentage of a positive family history could be attributed to lower level of community awareness and community health care amongst the study population.

The only complication seen in Using SIGN NAIL was bending of nail after 3 years in 1 patient without fracture.However the patient was kept on conservative and after 6 years ,the deformity didn't increase and the nail didn't bend any more.Georgescu et all found the different complications after the implant of the Sheffield telescopic rods i-e 7 external migrations of the obdurate rod, 7 pseudarthroses, 3 fractures with the bending of the rod, 2 fractures with the breaking of the rod, 1 fracture on disengaged rod, 1 case of osteitis of the femur, 1 case of cellulitis, 1 tibia fracture below Dall-Miles plate with hematoma and 1 case with wound .dehiscence.[22]Watzl et all found the following complications with the use of non extensible nails i-e deformity (67%) and fracture (33%).[23] SATVINDER KAUR et all in their mean follow-up of 30 months Found that the using of

intramedullary rods having no recurrence of deformities and better bone densities .[24] Saldanha et all used the monolateral external fixator and illizarov frame over previously inserted intramedullary rods for lengthening and correction of angular deformity but no significant results found.[25] Nicolas Nicolaou et all used the Shieldfield telescoping rods and the complications rate was almost 50%.ten rods (15%) were exchanged because of rod disengagement due to growth, thirteen rods (20%) were exchanged because of complications, and ten rods (15%) required further surgery other than exchange because of complications.[26] G.EL.ADL et all did a comparative study of using telescoping vs non telescoping rod insertion. The reoperation rate was high with non telescoping rods as compared to telescoping group.[27] Similarly Alzahrani et all in their study found the migration of the FD rod whiIe managing the osteogenesis Imperfecta.[28] There are some limitations in our study.First of all the number of patients are less though the follow up is sufficient.Moreover the SIGN Nail is not an extensible nail .We didn't discuss the exchange of nail needed with the growth.So,further study needed which can evaluate more .

Conclusion

The use of SIGN NAIL for the correction of deformity in Osteogenesis Imperfecta is excellent choice.It prevents the implant associated complications like displacement, breakage etc as compared to other intramedullary nailing system. It should always be considered while treating the patients with Osteogenesis Imperfecta**.** Patient rehabilitation is early, hospitalization is short, and osteotomies site healing response is good .

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