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Functional outcome of correction of skeletal deformities due to Osteogenesis Imperfecta using Intramedullary devices: A Retrospective study

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Abstracts
Introduction: The surgical correction of skeletal deformities associated with Osteogenesis Imperfecta is sometimes a challenge as a significant number of non-union cases have been reported after surgical correction of deformities by various techniques. We conducted a retrospective study of these cases in which specifically Intramedullary devices were used to fix the fracture, to know the outcome after surgery
Methodology: A retrospective study was carried out in our Institution to analyse the results of 21 children operated using Intramedullary implants for correction of skeletal deformities due to Osteogenesis Imperfecta between January 2010 and December 2018. Patients were followed up to analyse the functional outcome and know the incidence of requirement of revision surgery in these cases
Results: A total of 43 segments were operated with the Femur being operated in 55.8% of cases while Tibia were operated in 44.2% of cases. In 61.9% of children one segment was operated while in 38.1% of children 2 or more segments were operated. It was observed that 19 segments operated required revision and the mean period of revision was 36 months. It was observed that in 63.1% of cases which required revision, Rush Nails were used for the corrective surgery while in only 15.7% of cases which required revision, Telescoping nails were used for the corrective surgery initially.
Conclusion: Based on our analysis of correction of skeletal deformities associated with Osteogenesis Imperfecta using Intramedullary devices, Telescoping nails were found to have the best functional outcome.

Keywords: deformity correction, osteogenesis imperfecta, intramedullary implants, zoledronic acid

Introduction

Osteogenesis Imperfecta is a genetic disease with increased bone fragility and low bone mass. Most patients of Osteogenesis Imperfecta have a mutation in one of the two genes that encode the α chains of collagen type I (COL1A) and (COL1A2) [1]. These two genes are located in chromosomes 7 and 17 [2]. Ty Figal extra-skeletal manifestations associated with this disorder include hyperlaxity of ligaments and skin, dentinogenesis imperfecta, hearing impairment and presence of Wormian bones on skull radiographs [3].

Correction of skeletal deformities associated with this condition is usually done with the help of Intramedullary rods. Though it is generally assumed that the fractures of long bones in Osteogenesis Imperfecta rarely fail to unite, few reports describing non-union of fractures and osteotomy sites have appeared in literature [4]. Hence, we decided to conduct a retrospective study at our Institution to analyse the functional outcome after correction of skeletal deformities in Osteogenesis Imperfecta using Intramedullary rods.

Aim

This study aims to analyse the functional outcome of correction of skeletal deformities associated with Osteogenesis Imperfecta using Intramedullary rods.

Methodology

This is a retrospective study conducted in our Institution. 21 patients operated for correction of deformities associated with Osteogenesis Imperfecta using Intramedullary rods between January 2010 and December 2018 were followed up to analyse the functional outcome.

All patients were on regular cycles of Bisphosphonates. Bisphosphonates are effective in reducing the fracture rate and also in improving the bone mineral density [5]. We used 3rd generation nitrogen compound, Zoledronic acid. Patients between the age group 1 – 3 years of age were given 0.025mg/kg of Zoledronic acid through a 30 minute infusion every 3 months. Patients between the age group 3 and 17 years were given 0.05mg/kg of Zoledronic acid through a 30 minute infusion every 4 – 6 months. A cycle was skipped before the Intramedullary rod insertion and the cycle was delayed after the rodding also.

All patients were operated by the same surgeon. After surgery all patients were immobilised in a cast. 6 weeks after surgery the cast was removed and Non- weight bearing gait training was given. Full weight bearing was allowed after the Osteotomy completely healed which was around 3 months after surgery. All patients were followed up for a minimum of 3 years.

Results

A total of 21 chidren were included in the study. 14 were boys and 7 were girls (Table 1). The age of the children ranged between 3years and 16 years with average age at

presentation being 7.4 years. As per the Silence classification [6] 14.3% of patients were Osteogenesis Imperfecta Type 1, 28.6% were Osteogenesis Imperfecta type 3 & 57.1% were Osteogenesis Imperfecta Type 4(Table2). In 61.9% of children only one segment (Femur/Tibia) was operated, while in 38.1% of children 2 or more segments were operated (Table 3).

Total of 43 segments were operated out of which the Femoral segment was operated in 55.8% of cases while Tibial segment was operated in 44.2% of cases (Table 4).

6 segments were stabilised with TENS, 18 segments were stabilised with Rush Nails while 19 segments were stabilised with Telescoping nails (Table5).

At the end of 3 years 15 patients were able to walk with or without walking aids while the rest 6 patients were wheel chair mobilised. Patients were mobilised after union was seen on the X-rays (Fig1 &2)

The mean Hip flexion was 95 degrees and mean knee flexion was 125 degrees.

It was observed that 19 segments required revision surgery. The mean period of revision surgery was 36 months.

Out of the 19 cases which required revision surgery it was observed that Rush nails were used in 12 cases during the initial surgery, TENS in 4 cases and Telescoping nails in 3 cases (Table 6).

Bending of the rod was observed in 5 segments, Cut through of Implant in 5 segments, Migration of Pins/Rods in 4 segments, In-situ fractures observed in 3 segments and Infection in 2 segments which were operated (Table7)

A failure rate of 66.6% was observed in those cases stabilised with Rush nails, 66.6% failure rate in those stabilised with TENS while only 15.8% failure rate was observed in those cases stabilised with Telescoping Nails (Table 8)

Table 1: Patient demographics and variables

***n* = 21(children)**	**Frequency (%)**
Gender	
Female	7(33.3%)
Male	14 (66.7%)

Table 2: Type of Osteogenesis Imperfecta as per Sillence Classification

Type I	**3 (14.25)**
Type III	6(28.6%)
Type IV	12 (57.1%)

Table 3: Number of Segments operated

One Segment	**13(61.9%) children**
Two or more segments	8 (38.1%) children

Table 4: Segment Operated- Femur/Tibia

***n* = 43(segments)**	**Frequency (%)**
Segment operated	
Femur	24(55.8%)
Tibia	19 (44.2%)

Table 5: Intramedullary device used

TENS	**6 (14%)**
Rush Nail	18(41.9%)
Telescoping nail	19 (44.1%)

Table 6: Revision Surgeries. (n = 19). Intramedullary device used

Rush Nails	**12(63.2%)**
TENS	4 (21%)
Telescoping nail	3(15.8%)

Table 7: Complication observed

Bending of rod	**5 (26.3%)**
Cut through of impant	5(26.3%)
Migration of rods/pins	4 (21.1%)
Insitu fractures	3(15.6%)
Infection	2(10.5%)

Table 8: Comparison of failure rates of the Intramedullary devices

Implant Used	**No. of cases**	**No. of failures**	**Failure rate**
TENS	6	4	66.7%
Rush Nails	18	12	66.7%
Telescoping Nails	19	3	15.7%

Table 9: Comparison of Failure rates of Rush Nails and Telescoping Nails

	Failure	**Success**	**Total**
Rush Nail	12	6	18
Telescoping Nail	3	16	19
Total	15	22	37

Using the Chi-square test it was found that there was significant more failures when Rush Nails were used compared to Telescoping Nails (p-value = 0.002)

Fig 1: Pre-operative X-rays of Patient with Osteogenesis Imperfecta

Fig 2: Post-operative X-rays of patient

Discussion

Correction of skeletal deformities associated with Osteogenesis Imperfecta is challenging and associated with several complications including non union at the osteotomy sites [4]. Zoledronic acid has been shown to be associated with clinical and radiological benefits in form of reduction of number of fractures and increase in bone density on Dexa scan [5]. Hence Zoledronic acid was given for all the patients of Osteogenesis Imperfecta at our centre.

Studies have been carried out to find out the ideal Intramedullary device for the femur and the tibia for correction of deformities in Osteogenesis Imperfecta. The results of the survival analysis showed that both in terms of rod survival and the interval between initial surgery and reoperations, Telescoping rods fared significantly better than single rush nails [7]. The greater longevity of the implants may be attributed to the fact that telescoping rods elongates as the bone grows.

Our study also showed that Telescoping rods fared much better than Rush nails or TENS. There was a significant (Chi square test, p value= 0.002) lesser failure rate with Telescoping rod than those cases operated with Rush Nails (Table 9). The findings would have been much more significant if there were at least 30 cases operated by each technique, which was a limitation of our study. Another limitation of the study was that we could not use a scoring system like the Gross Motor Function Measure Score sheet [8] to assess the functional outcome of the cases as this was a retrospective study.

Use of dual Rush nails have been reported to have the same success rate like Telescoping nails though in some cases it led to bending of the bones [7]. This technique was not used at our centre.

Conclusion

The ideal choice of Intramedullary device for correction of skeletal deformities in patients suffering from Osteogenesis Imperfecta appears to be Telescoping rods as it was found to be associated with the least number of revision surgeries and complications as it could account for the growth of the bones.

Clinical Message

Telescoping rods seems to be the ideal implant for stabilisation after correction of deformities in Osteogenesis Imperfecta as it has been found to be associated with fewer complications, has a sufficiently long interval between insertion and revision and is easy to insert and revise.

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