ABSTRACT

Objective: To evaluate the performance of an extensible nail with hooks, named HIMEX, in osteogenesis imperfecta (OI) deformities. Methods: All child patients were operated on with HIMEX from 1990 to 2004. The number of fractures, reappearance of deformities, improvement of motor development before and after the use of HIMEX, and the incidence of the migration and nail survival were compared. Results: Fourteen patients, with ages from 2 to 18 years, including 8 females, underwent 46 procedures, 39 primary and 7 re-operations. The average age at the first fracture was 148.21 days, and there was an average of 42.6 fractures per patient prior to HIMEX placement. Of the forty-six bones affected, 28 were femurs and 18 were tibias. Average follow-up care lasted 80.21±36.71 months. There was a statistically significant decrease (0.78) in the number of fractures per patient and an improvement in walking in seven of the fourteen patients. Revision occurred in 18% of patients and migration of the nail occurred in 12% (5/39). Eighty percent of the nails remained in situ until 108 months, with femoral procedures lasting significantly longer than tibial procedures. The type of OI and the age at the procedure did not significantly affect the incidence of revision. Conclusion: HIMEX significantly reduced the number of fractures, presenting lower incidence of migration and higher survival rates than those described in literature.

Keywords: Osteogenesis imperfecta. Survival analysis. Implantation, endosseous. Postoperative complications.

INTRODUCTION

The modern era of surgical treatment of OI started in 1948 with the work described by Sofield, presented at the American Academy of Orthopedic Surgery in 1951 and published in 1959. In this paper, the authors concluded that this treatment brought advantages for patients with OI, as it reduced the number of fractures and bone pain, corrected preexisting deformities and prevented their appearance, thus improving the alignment of the limbs and their function. After this pioneer report, other papers were presented using the same technique, with non-extensible nails. It can be observed that the technique of Sofield and Millar, using the non-extensible nail as an implant, although used more frequently in past decades, still has some devotees. The main complications have continued over time, with an emphasis on fracture of the part of the bone that is unprotected by the implant, migration of the nail and relapse of the deformity.

On the other hand, Bailey and Dubow presented the results of their experimental studies initiated in 1958, which gave rise to the creation of an extensible nail capable of accompanying bone growth previously tested on dogs and on only three patients with OI. The Bailey-Dubow nail brought new perspectives for the treatment of patients with OI, with a growing tendency for use, substituting non-extensible implants. Since 1963 it has been possible to observe the predominant use of extensible nails in the treatment of patients with OI, especially as of the 80s, and of the 23 studies selected with significant casuistry, thirteen used only extensible nails, four used extensible and non-extensible nails or conventional implants and three used conventional nails.

Although there is an increasing number of publications that have been using extensible intramedullary nails, the complications with this procedure continue occurring with a high incidence.

All the authors declare that there is no potential conflict of interest referring to this article.
of 27 to 63%. 11,13,17,19 However, the publications of Gamble et al.14, with 69%, and of Porat et al.18, with 72%, are those with the highest rates of complication. It should be emphasized that most studies on the extensible nail in OI were published between 1990 and 2000 and refer to cases treated in the decade prior to that of publication, with the majority of authors having used the classical implant of Bailey-Dubow of 1963, where only one of the “Ts” from the extremities was fixed. However, this nail underwent improvements, one of which aimed at the fixation and enlargement of both “Ts”.

Therefore the goal of this study was to assess the performance of an extensible intramedullary nail, modified by addition of hooks at the extremities (HIMEX), on long bones of the lower limbs of children with OI, as regards:

a) the reduction of the number of fractures in the postoperative period;
b) the change in walking ability;
c) the reduction of the incidence of migration and

d) the increase of the survival rate of nails.

CASUISTRY AND METHODS

There was an investigation and review of all the medical records and radiographies of patients with diagnosis of OI, who had been operated as from 1990 for the correction of bone deformities, using the extensible intramedullary nail modified with hooks (HIMEX). The patients’ classification fulfilled the criteria of Silience26 and Shapiro.27 All the patients were evaluated and treated for possible disorders of the mineral and acid-base metabolism prior to surgical treatment and none had previously used bisphosphonates.

The criteria of Hoffer and Bullock28 were modified to classify the patients according to walking ability in the pre- and postoperative periods. For the analysis of this casuistry the above criteria were simplified and adapted as follows: ambulatory without assistance, ambulatory with assistance and functional ambulatory (A) versus non-ambulatory, in this case dependent on wheelchair (NA).

Description of the implant

HIMEX nails are currently manufactured by the company Engimplan with head offices in the city of Rio Claro, Sao Paulo. The raw material used in the production of this implant is ASTM F138 stainless steel, which complies with NBR-ISO-5832-1/ISO5832-1 standards. The tubular part of the nail is manufactured with an external diameter of 5mm, internal diameter of 3.2mm and solid part with external diameter of 3mm. The tubular and solid components of HIMEX are diagrammatically presented in Figure 1.

Description of the surgical procedure for the femur and tibia.

Definition of the size of the nails in the preoperative period

Radiographies were taken in the preoperative period, with a metallic pattern, with known dimensions, fixed to the side of the limb, to allow the estimation of the bone length. A simple rule of three was used to obtain the actual length, in which the actual and apparent measurement of the radiograph pattern and the apparent measurement of the bone were available. Thus they calculated the actual measurement of the bone to be operated. The surgical procedure was carried out under general anesthesia, with prior reserve of blood and prophylactic antibiotic.

Treatment of femoral deformities

The patient was placed in horizontal supine (HS), with a cushion under the lumbar region. The femur was approached laterally, and the extension of the incision depended on the location and extent of the deformities. The vastus lateralis muscle was separated from the intermuscular septum and the periosteum was incised lengthwise and repaired with suture threads. The deformed bone segment was withdrawn from the periosteal sheath. The medullary channel of the bone fragment as well as that of the proximal and distal segments of the femur were opened with drills of progressive diameter until they surpassed the nail diameter by at least 0.5 millimeter.

With the proximal fragment of the femur in adduction and flexion, a guide wire measuring 3mm in diameter was introduced in retrograde fashion, and emerged medially to the greater trochanter through the skin. The tubular portion of the nail was introduced from this guide wire in an anterograde manner, using rotating movements until they reached the proximal focus of the osteotomy. After this the knee was approached through an anterior longitudinal incision below the patella to expose the intercondylar region of the femur. The pin was introduced from the center of this region, under direct vision, seeking the marrow cavity of the distal fragment up to its externalization in the distal focus. The limb was aligned under traction and the bone defect created by the removal of the deformed bone segment was filled by the bone fragments previously sectioned and perforated until the maximum possible length of the femur was reestablished. At this point, the pin was introduced under direct vision in the tube and slid in cranial direction until the hook became anchored in the surface of the articular cartilage of the intercondylar region of the femur without penetrating it. An evaluation was conducted of the tube position, which was supposed to have the hook anchored at the end of the greater trochanter, and rotational alignment of the femur. The periosteum was carefully closed around the bone segment and the vastus lateralis was reinserted in the muscular septum with 000 nylon non-absorbable thread. Aspiration drainage tubes were left in the muscular plane and in the subcutaneous cell tissue. An occlusive dressing was applied, followed by a pelvic-podalic plaster cast for four weeks as the nail does not offer rotational stability.
Treatment of tibial deformities

For the treatment of tibial deformities the calculation of the nail size was performed in the same way. The access route was anteromedial, accompanying the tibial deformity and centered at the deformity apex. The deformed bone segment was sectioned in its proximal and distal portion and, in general, withdrawn from the periosteal bed to be fragmented. The medullary canal was opened with drills of progressive diameter until it surpassed the diameter of the nail by 0.5 millimeter. The most distal portion in the distal tibial epiphysis was identified for introduction of the guide wire. If the deformity to be corrected was in varus, access was accomplished in the anterior medial region and if in valgus, in the anterior lateral region. To facilitate the introduction of the guide wire by the distal tibial epiphysis, the ankle was kept in maximum planter flexion, as was the knee. The tubular portion of the nail was then introduced through this guide wire, from distal to proximal, up to the focus of the osteotomy. The proximal tibial epiphysis was approached through the same incision made to expose the femur. The pin was introduced in the proximal epiphysis on the articular surface of the proximal tibial epiphysis in the direction of the center of the marrow cavity, with the knee in maximum flexion. After this the bone fragments removed were put back on the nail, until the bone defect was corrected. The pin was introduced into the tube and both were slid in the opposite direction, reestablishing the length and the realignment of the tibia. The rotational alignment and the position of the hooks at the proximal and distal epiphyses were evaluated by the image intensifier. The periosteum was sutured with 000 nylon and an aspiration drainage tube was installed. An occlusive dressing and inguinai-podalic plaster cast were applied for four weeks.

When the bones were very fragile, their fragmentation occurred at the time of the osteotomy, or during the opening of the medullary canal. Under these circumstances, these fragments were reinserted around the nail and kept in place by the periosteal suture. The rotational alignment, as well as the length, were reestablished and maintained by the plaster cast. The aspiration drainage tubes were removed after 24 hours and the stitches together with the cast after four weeks.

ANALYSIS OF RESULTS

To appraise the performance of HIMEX there was a comparison of the number of fractures, the change of ambulatory ability, the reappearance of deformities, the number of complications and the survival of the nail before and after its insertion. Complications were classified as greater and lesser with the former defined as those that could compromise the efficacy of the surgery and that needed a surgical procedure for their correction and the latter as those that did not influence the efficacy of the implant and did not need new surgical procedures. There was an analysis of the survival of the nails, considering re-operation as an undesirable final event. The risk factors that were considered possible interferers in the implant performance were: bone operated, age of patient and type of osteogenesis according to Sillence, and year of performance of the surgery.

Statistical analysis

Non-parametric tests, respectively Wilcoxon, Mann-Whitney U test and McNemar, were used for the comparison of the number of fractures, age of the patients at the time of the procedure and performance in relation to ambulatory ability in the pre- and postoperative periods. Survival curves were performed using the Wilcoxon-Gehan test. A value of p equal to 0.05 was used for all the analyses.

RESULTS

The study involved fourteen patients, with mean age at time of surgery of 9.17±3.42 years, eight of whom were female. Before the placement of the Himex, the mean number of fractures was 42.6±25 per patient (ranging from 5 to 187), with mean age upon first fracture of 148.21±311.14 days (ranging from zero to 900 days). According to the classification of Sillence, seven patients were type III, six were type IV and one type I. According to the classification of Shapiro, ten were CB (congenita B), two were TB (tarda B) and two TA (tarda A). Nine were non-ambulatory. After a mean follow-up time of 80.21 ± 36.71 months, there was significant reduction in the number of fractures after the placement of HIMEX (10 fractures/14 patients; p=0.002), and of these, eight were after a fall by the patient. As regards the degree of walking activity, five patients remained in the same mode as in the preoperative period and seven improved their performance, with no significant difference in relation to the distribution of classes according to the modified criteria of Hoffer and Bullock (p=0.25).

Forty-six surgical procedures were executed on 14 patients. Of the 28 femurs operated, 25 were primary surgeries and 3 reoperations, and of the 18 tibias, 14 were primary and 4 were reoperations. The follow-up time was significantly longer for the implants positioned in the femur (92.29±7.85 months) than in the tibia (61.62±6.23 months). The patients' age at the time of the primary procedures was significantly lower for the implants positioned in the femur 7.64±4.20 years, than those of the tibia 11.56±2.64 years. The mean number of procedures per patient was 3.5, ranging from 1 to 6.

There were seven reoperations in 39 primary procedures, which corresponds to a frequency of 18%, with one case of infection case, three cases of migration of the nail from the tibia, two cases of migration of the nail from the femur and one case of nail fracture. Therefore, the incidence of nail migration was 12%. It should be observed that of the complications observed, the case of infection occurred 31 months after the surgery, with the need for removal of the implant; the case of proximal cortical migration of the nail in the tibia was caused by the placement of the implant outside its proximal epiphysis; the case of proximal cortical migration in the femur occurred 10 years after surgery due to placement of the implant on the lateral side of the greater trochanter; the fracture of the nail in the femur occurred 8 months after manipulation of the nail, performed to correct its curvature; the cranial migration of the distal nail from the femur in the direction of the metaphysis was diagnosed 4 years after the surgery and was caused by exaggerated introduction of the hook in the epiphysis; two migrations of the nail in the tibia were favored by the exclusive placement of the solid part of the HIMEX.

Analysis of the risk factors for reoperation

In Figure 2 it can be noted that close to 80% of the implants were well positioned up to 108 months. Upon the evaluation of the bone treated (femur or tibia), it was observed that nails implanted in the femur had a significantly lower risk of reoperation than those positioned in the tibia (p=0.04). The final observation time of 80 months was considered for this analysis. (Figure 3)

In the evaluation of the effect of age, the group of patients was divided up into patients 6 years of age or under and patients...
Figure 2 – Accumulated survival curve of all the nails inserted.

Figure 3 – Accumulated survival curve of the nails placed in the femurs and in the tibias.

Figure 4 – Patient aged 4 years and five months with deformities on the two femurs and history of 37 fractures of the lower limbs.

Figure 5 – Previous patient in the fourth postoperative week with the telescopic nail.

over 6 years of age. In this manner, there were no significant differences in the time the implants remained, evaluated up to 80 months of follow-up (p=0.08). However, evaluating just the nails placed in the femur, the effect of the age bracket showed a tendency for shorter stay of the implant in the lower age bracket (p=0.06). The type of osteogenesis did not significantly influence the removal of the nails either, excluding the only case of type I (p=0.60).

We present the evolution of a clinical case in Figures 4, 5 and 6.

DISCUSSION

In OI, tibial and femoral deformities are produced by the traction of the muscles during bone growth, the reason why they follow a pattern that can be identified in almost all children, even if they are not yet ambulatory. For the surgical treatment to be efficient, it is necessary for the implant to act as a reinforcement, transforming the bone segment into a more resistant structure, particularly to efforts of flexion and shearing. This goal will only be reached with the rigorous and correct performance of the surgical technique, since even after the placement of the implant the bone fragility and the deformity perpetuating factors will continue acting as undesirable vectors for a good result. In literature, the result of surgical treatment with intramedullary nail has reduced pain besides the number of fractures and corrected
deformities, from the reports of Sofield s and Milla1, although countless complications continue to occur, mostly originating from bone growth.2,7 Therefore, in the fashion it was idealized, the extensible nail of Bailey-Dubow6 appeared to be the solution to this problem. However, they continued to occur, making its advantages dubious.14,18

Analyzing the complications associated with the extensible nail, it is possible to identify with decreasing frequency: loosening of the T-bolt, migration of the implant (to the metaphysis, to outside the cortical bone and to the joint), lack of lengthening, bending and fracture of the nail.

The loosening and the migration of the T-bolt are considered common incidents with the Bailey-Dubow nail, and we can mention, for example, the work of several authors11-13,16,17,19 that attempted to fix the T with a steel thread loop, but even so observed detachment of the T in 20% of the cases and the study by Karbowski et al.24, who encountered loosening of the T-bolt in 7.9% in the knee region.

On the other hand, nail migration deserves special emphasis for the frequency at which it occurs in literature (7 to 45%)12-14,16,17,19,23,24 and because it is the main cause of reoperation. Nail migration arises when the nail is not optimally positioned in the diaphysis and in the epiphysis. So much so that, even eliminating the possibility of detachment of the T-bolt, migrations continued to be reported in literature.16,23,25

In this casuistry we did not observe cranial or caudal migration of the nail at the proximal end of the femur. Although we should impute this fact to the correct placement of the nail, the presence of the hook introduced in the greater trochanter may have contributed to this end, as with this structure the nail can be more firmly anchored in the bone. On the other hand, the nail was not properly impacted at the proximal end in two patients, and was positioned just above the greater trochanter (without clinical re-percussion so far), and in another two there was varus deformity of the neck, with the need for re-intervention in one of them. This technical error tends to be more common when there is deformity in varus in the preoperative period. In this set of circumstances, the entrance point of the nail should be the more medial to the greater trochanter as the deformity is more in varus.

Implant migration at the distal end of the femur occurs more frequently in the cranial direction.24,25 In this casuistry only one case of cranial migration of the nail was observed in the patient TCR due to exaggerated introduction of the hook in the epiphyseal. We consider that the characteristics of the implant may also have added favorable factors to the non-migration of the nail at this site. Unlike the technique proposed by Bailey-Dubow6, with HIMEX the hook should not be kept below the articular cartilage, but rather impacted against its surface, and visible in the joint.

Moreover, this modification can facilitate removal of the nail when necessary, as such a task with Bailey-Dubow nails is a difficult one, so much so that Mulpuri and Joseph26, in two cases, reported a severe lesion of the cartilage and of the subchondral bone in the removal of this nail in this region. Thus, we could conclude that the optimal placement of the nails and the addition of the hooks on the extensible nail were responsible for the long survival of these in the femur. The finding of more than 90% of continuity of nails in 60 months is an uncommon result in literature.

In the tibia, migration may occur at the proximal end in the direction of the knee joint, or at the distal extremity in the cranial direction. Now in the distal region access is usually more difficult and the incorrect introduction of the screw may facilitate its migration, as observed by Karbowski et al.24, who actually reported 38% of cranial migration of the nail in this region, in 60 tibias.

In this casuistry it was observed that the survival of nails in the femur was significantly higher than that of the tibia, although there were only four reoperations (three due to migration) in 14 primary procedures in the tibia. However, this result can be partly justified by the fact that in one of the patients, the placement of the nails was incomplete in the two tibia, which undoubtedly foretold an undesirable outcome of the procedure. As there were a total of four tibial reoperations, these two had significant statistical weight, suggesting that the results in the tibia can also be considered good on account of the addition of the hooks.

Although not significant, there was a tendency for age to reduce the survival of the HIMEX in the femur, corroborating the idea that the greater the growth potential, the greater the risk of reoperation. It is also worth emphasizing that although important, the technical difficulties expected for the lower age bracket did not appear to influence the results since the reoperations occurred at 23, 51 and 113 months after the initial procedure.

In this casuistry, even after improving the mechanical and anatomical alignment of the bones of the lower limbs and increasing their resistance, the patients did not significantly improve their performance in terms of ambulatory ability. It is known that the walking ability is influenced by the type and intensity of disease, by the degree of previous neuromuscular impairment, and by the rehabilitation program established.29,30 This program involves postural correction, muscular strengthening, aerobic conditioning and the use of orthoses, when necessary. However, the influence that the family and socioeconomic conditions have played should not be disregarded. 30 In this casuistry, most of the patient treated had, besides intense bone fragility, unfavorable socioeconomic conditions and lived long distances from the institution, which greatly
hindered the maintenance of a minimum rehabilitation program. However, if we take into consideration Shapiro’s classification27, of the cases of type B congenital OI that correspond to 70% (10/14) of the patients from this casuistry, there was a reduction of wheelchair dependence from 80% to 50% at the end of the evaluation, a better percentage than that foreseen by this author.

In spite of the good results, there are still problems that deserve to be improved, such as the size and the curvature of the hooks and studies targeting the possibility of performance of the procedure without arthroty.

CONCLUSIONS

The use of HIMEX in the treatment of 14 children with OI, mainly Sillence type III and IV, with a mean follow-up time of 50 months, showed the following performance:

- It significantly reduced the number of fractures;
- It did not significantly influence the walking ability;
- It presented lower incidence of migration than that referred to in literature;
- It presented a higher survival than that referred to in literature.

REFERENCES