

Femoral neck fractures in osteogenesis imperfecta treated with bisphosphonates

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Abstract

Purpose Osteogenesis imperfecta (OI) is a condition characterised by bone fragility and multiple fractures, which cause considerable morbidity in the affected patients. Most cases are associated with mutations in one of the type I collagen genes. Recently, bisphosphonates have been used widely to reduce pain and the incidence of fragility fractures in OI in children, even though there have been concerns raised regarding the long-term complications of it due to their effect on the bone. The fragility fractures involving the neck of the femur in children with intramedullary rods in the femoral shaft are very difficult to treat. Although these fractures are frequently un-displaced, they require optimal internal fixation to achieve fracture union. The aim of this study was to assess the clinical and radiological outcomes of OI patients with intracapsular femoral neck fracture treated with headless compression screws.

Method and results At our institute, we identified seven patients (11 hips) with OI who underwent internal fixation with headless compression screws for a neck of femur fracture between June 2010 and Dec 2012. The time to fractures healing was on average 14 weeks (12 to 16). All patients gained their pre-injury ambulatory status.

Conclusion It is very challenging and technically demanding for orthopaedic surgeons when treating the fragility fracture of the neck of femur in patients with intramedullary rod in the femoral shaft. The published data regarding the management of these complex conditions are very limited. We describe our experience with the technique of percutaneous headless compression screw fixation for treating the femoral neck fractures in OI patients.

Introduction

Osteogenesis imperfecta (OI) is a hereditary disorder with a defect in type I collagen metabolism. The condition is characterised by osteopenia, frequent fractures, progressive deformity and chronic bone pain. Milder forms of the disease have a better prognosis with patients often surviving to adulthood with normal mental function. The Sillence classification is widely used and is based on the inheritance pattern and clinical characteristics.¹ Patients present with painful fractures after trivial injury and often involving the lower limb long bones.¹

The prevalence of OI is estimated to be one in 20000 to 50000 infants, but the incidence is probably higher because, since it is a heterogenous condition, misdiagnosis is frequent.² Most patients with clinical diagnosis of OI are positive for a mutation in one of the two genes COL1 A1 and COL1 A2.³

The medical management of OI with bisphosphonates has been shown to reduce the incidence of fractures and bone pain.⁴⁻⁷ However, the mechanism by which the risk of the fracture may be reduced with bisphosphonates is not fully understood.⁸ Further, there have been increasing concerns about the potential long-term effects of bisphosphonates on bone in adults treated for osteoporosis, where a number of atypical subtrochanteric fractures have occurred in treated patients.¹⁴⁻¹⁷ Also, a recent study comparing a historical group with bisphosphonate-treated OI patients reported a changing pattern of femoral fractures in patients receiving medical therapy.¹⁸

The fragility fractures involving the neck of the femur in children with intramedullary rods in the femoral shaft are very difficult to treat. Although these fractures are frequently un-displaced, they require optimal internal fixation to achieve fracture union (Fig. 1) The evidence in the literature regarding the fixation of femoral neck fractures in OI in either children or adults is limited.^{10,11}

The aim of this study was to assess the clinical and radiological outcomes of OI patients with intracapsular

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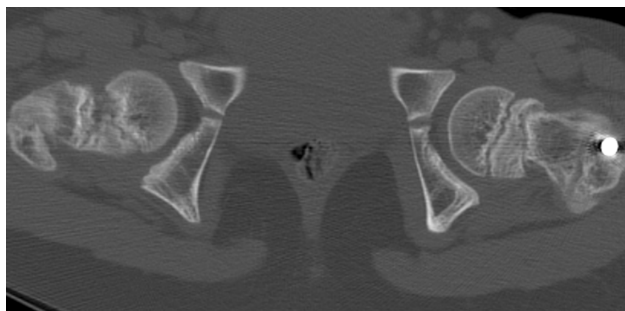


Fig. 1 CT scan showing bilateral femoral neck fractures in an OI patient with an intramedullary rod *in situ* on the right side.

femoral neck fracture treated with headless compression screws.

Further, we describe our operative technique and experience of using headless compression screws in treating femoral neck fractures in children with OI.

Patients and methods

This was a prospective cohort study of seven consecutive patients (11 hips) with OI who underwent internal fixation with headless compression screws for an intracapsular femoral neck fracture at our institute between June 2010 and December 2012. We retrospectively reviewed the prospective data. All fractures were secondary to trivial trauma, except in one patient fracture occurred following a fall. Patients in the study group were all females with a mean age of 11 years (9 to 13) and they were independently mobile.

The patients with OI at our institution are treated by a multidisciplinary team consisting of the metabolic bone disease team, paediatric orthopaedic surgeons and physiotherapists. All the patients in the study had received bisphosphonate treatment and they had an intramedullary rod in the femoral shaft from previous operations. Bisphosphonates were discontinued for six to eight weeks in the post-operative period. The cohort of patients in this study continued with bisphosphonate treatment at six-monthly intervals and bone mineral density checked, according to the local protocol.

OI was classified according to the Sillence classification, with five patients being type 1 and two patients type 4. All operations were performed by the senior authors.

Operative technique

The procedure was performed under general anaesthetic. The patient was positioned supine when bilateral and lateral when unilateral on a standard table without traction. The standard preparation of the surgical site was carried out. Antibiotic prophylaxis (cefuroxime) was given at the time of induction.

Using a minimally invasive approach the guide wire was passed from the lateral femoral cortex of the proximal femur into the centre of the femoral head with the nail technique, posterior to the intramedullary femoral rod under fluoroscopy control. The outer cortex was drilled with a 4.2 mm drill and a 6.0 mm Acutrack plus compression screw (Acumed Innovative Solutions, OR, USA) was passed over the guide wire to achieve maximum compression across the fracture site. In one patient, the whole procedure was technically demanding for the operating surgeon because of the small-sized femur and the presence of the intramedullary rod in the femoral shaft. None of the screws crossed the proximal femoral physis in our study group.

The post-operative regime consisted of wheelchair mobilisation for six weeks followed by partial to full weight bearing. Patients were followed at regular intervals in the outpatient clinic at six weeks, three and six months, and then annually for at least two years. Periodic radiographs of the hip were obtained to assess fracture union and to monitor for avascular necrosis of the femoral head.

Results

There were seven patients with 11 femoral neck fractures; three patients had bilateral involvement and they were treated with headless compression screws. All fractures healed satisfactorily (Fig. 2) with an average time to fracture healing of 14 weeks (12 to 16). All patients had regained their pre-injury ambulatory status and continued to be followed up annually by our multidisciplinary team. There were no cases of avascular necrosis of the femoral head in any of the patients post-operatively to date. However, coxa vara deformity was noted in one patient at the 12-month follow-up.

Discussion

The use of bisphosphonates has brought about a radical change in the medical management of OI.⁹ It is known to reduce the risk of fracture, improve quality of life, and increase bone density and bone pain.^{4-7,12} However, the exact mechanism by which the risk of fracture may be reduced is not fully understood.⁸ Further, there have been increasing concerns regarding the potential long-term effects of bisphosphonate treatment in adults for osteoporosis, with a number of cases reported where atypical subtrochanteric femoral fractures have occurred.¹⁴⁻¹⁷ However, the evidence for long-term results of bisphosphonate treatment in children is insufficient.¹³ There are no published data on the incidence of intracapsular femoral neck fracture or the pattern of proximal femoral fractures in OI patients before the advent of

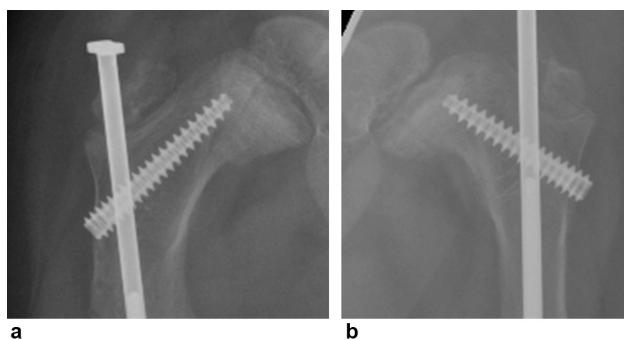


Fig. 2 (a, b) Follow-up images showing healed femoral neck fracture with screw *in situ*.

bisphosphonate therapy. However, in their study, Nicolaou et al¹⁸ reported the varying trends of femoral fracture pattern in OI patients treated with bisphosphonates compared with the historical group. Their findings of the radiographic review were a frequent subtrochanteric location of the femoral fracture in children treated with bisphosphonates compared with the more diffuse anatomical location of the fractures in the historical group. All fractures were short oblique or transverse in configuration. Two femoral neck and four distal fractures were present in the group and the majority were in the mid-diaphyseal region. In total, 73% of the fractures occurred in patients already treated with intramedullary rodding of the femur. Further, the majority of these fractures were due to trivial trauma in the ambulatory group.¹⁸ All patients in our study group were treated with bisphosphonate therapy and had sustained un-displaced intracapsular femoral neck fractures secondary to trivial trauma except in one unilateral case. The evidence for the long-term effects of bisphosphonates on the bones in patients with this skeletal dysplasia is insufficient.

At our institute, all but one patient in the study group were treated with Sheffield telescoping intramedullary rods and the main indications for rodding were to reduce pain and promote walking in patients with recurrent fractures or deformity. The unilateral case had previous elastic nail stabilisation. These rods shield the femoral shaft and provide stability, but because of the weaker mechanical properties of the bone in OI and the unsupported neck-shaft junction could lead to coxa vara. The altered biomechanics thus created around the hip joint could increase the risk of femoral neck fractures. Aarabi et al has reported increased incidence of coxa vara in patients with severe OI treated with femoral rodding.¹⁹ They conclude that regular clinical and radiologic follow-up is indicated in children with previous femoral rodding and in severely affected children, particularly those with OI type III.¹⁹

We have shown that a single 6.0 mm compression screw inserted with miss-the-nail technique can

provide satisfactory fixation of the femoral neck fracture in patients with OI. Further, in these patients, internal fixation was technically demanding for the operating surgeon because of the fragile bone, small femur and the presence of an intramedullary growing rod in the femoral shaft.

There are only a few published reports looking at internal fixation of femoral neck fractures in OI patients.^{10,11} Tsang et al,¹⁰ in their report, highlighted the difficulties faced while performing internal fixation for femoral neck fractures with an intramedullary rod being present in the femoral shaft in cases of OI patients. To our knowledge, there are no published data pertaining to the incidence of the neck of femur in OI patients.

With recent advances in methods of internal fixation of complex femoral fractures in children and the introduction of lateral entry multifunctional reconstruction femoral intramedullary nails (Pedinail, Ortho paediatrics, IN, USA), the risk of femoral neck fractures in patients with OI may be reduced.

To overcome the inadequacy of the proximal fixation with Sheffield telescoping rods, in our unit we currently use Pedi nail to treat complex proximal femoral fractures and for stabilisation of proximal femoral corrective osteotomy in children with OI in the older child with adequate canal size. Preliminary results are awaited.

There is some evidence to suggest that bisphosphonates affect the quality of the organic matrix, leading to bone with weaker mechanical properties.^{20,21} Further, the proximal femur fracture pattern may be altered due to the above-mentioned factors and the tensile forces that would warrant further investigation. In addition, there are no published data on the pre-bisphosphonate era incidence of femoral neck fractures in OI patients. Since the advent of bisphosphonates treatment for OI patients, at our centre we have noted changing patterns of femoral fractures in patients treated with bisphosphonates compared with the historical group.¹⁸ Senior authors propose that rods shield the femoral shaft, but the forces are transferred to the femoral neck and because of the weaker mechanical properties of the bone in OI, the unsupported neck-shaft junction could increase the risk of femoral neck fractures.

There are limitations to this retrospective study. It does not compare fracture incidence in bisphosphonate-treated patients with untreated patients.

To conclude, there has been no mention in previous literature specifically on intracapsular fracture neck of femur in OI and our experience with compression screws for internal fixation of femoral neck fractures has been satisfactory with no complications. Also, this method of fixation is a safe and viable option for treating these fractures with femoral intramedullary rods *in situ* in the younger child.

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COMPLIANCE WITH ETHICAL STANDARDS

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ETHICAL STATEMENT

Ethical approval: Since this was a retrospective outcome study, hospital audit department approval was obtained and ethical approval was not needed. Informed consent: Informed consent was obtained from all individual participants included in the study.

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