Complications in the surgical management of femur fractures in children with non-ambulatory cerebral palsy

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Introduction
Children with severe non-ambulatory cerebral palsy (CP) are at high risk of sustaining a pathological femur fracture.1,2 These fractures are rare. In a review of the epidemiology of femur fractures in children from Cape Town, the prevalence of femoral pathological fractures related to cerebral palsy was less than three per cent.3 These patients often have multiple co-morbidities which predispose them to pathological femur fractures.4,5 The gold standard of treatment for these fractures is conservative management with either traction or cast immobilisation. The incidence of complications has been reported as extremely high in a number of studies after surgical management of these fractures.1,7 The objectives of this case report are to discuss the aetiology of these fractures, emphasise the role of conservative management as the gold standard of treatment, and highlight complications of surgical treatment in children with non-ambulatory cerebral palsy.

Key words: cerebral palsy, fracture, bone health

Presentation
ZS is a 16-year-old female with mixed CP, gross motor function classification system (GMFCS) level V and uncontrolled epilepsy. Her parents gave a history of bilateral clavicle fractures and a left femur fracture which was treated surgically with an open reduction and plate fixation in 2012. The current episode occurred after she fitted and fell from her bed. Although verbal communication was impossible it was clear that she was suffering from severe pain in the right thigh. Examination revealed a patient with severe total body CP, poor nutritional status, multiple contractures involving the hip, knee and ankle with an extremely tender right groin and proximal thigh.

Plain X-rays of the pelvis and femurs showed the healed left midshaft femur fracture and the new right completely displaced sub-trochanteric femur fracture as shown in Figures 1 and 2.

The patient was treated surgically, with an open reduction and internal fixation using a blade plate for the right sub-trochanteric femur fracture and removal of the locked plate at the same surgical sitting.

The bone was extremely osteoporotic. She fitted again day 0 post-operatively, causing a re-fracture of her right femur resulting in the blade plate pulling out of her femoral neck (Figure 3). The left femur remained intact after removal of the locked plate at the same surgical sitting.

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She continued to have breakthrough seizures, despite being reviewed by the paediatric neurology team, and being placed on therapeutic doses of an anticonvulsant, sodium valproate. She was subsequently taken back to theatre for a removal of hardware and a new internal fixation of the right sub-trochanteric femur fracture with a locked proximal femur plate (Figure 4). She remained stable in the ward despite occasional breakthrough seizures. Two weeks post revision open reduction and internal fixation with the locking plate she was discharged home in a stable condition.

She returned to hospital two weeks after her discharge with a deep septic surgical wound site and a deteriorated general condition. The wound required four serial debridements and the use of a vacuum dressing in order to get the wound clean enough for secondary closure at the fifth surgery. She was given intravenous antibiotics including cloxacillin and tazobactam. Her in-patient management was complicated by septicaemia from Candida albicans which was cultured from the wound and blood culture. This was treated successfully with intravenous fluconazole, converted to oral medication on discharge. During her second admission a dual energy X-Ray absorptiometry (DEXA) scan was performed. This confirmed that she was severely osteoporotic, with a Z-score of −4.3. After consultation with the paediatric metabolic unit, she was started on calcium and vitamin D. A decision was made not to start bisphosphonates. At six months follow-up her wounds had healed with no sign of infection and the fracture had united.

**Discussion**

Pathological fractures of the femur in adolescents with non-ambulatory CP are rare but well described. Uddenfeldt Wort et al. showed in a population study from 2013 that the risk of pathological fracture was significantly higher in children with non-ambulatory cerebral palsy, GMFCS IV or V compared with ambulatory children (GMFCS I–III). The aetiology of these fractures is multi-factorial. Vitamin D deficiency and overt rickets together with immobility, non-weight bearing and lack of exposure to sunlight have been closely associated. Replacement with vitamin D and calcium have been proposed by a number of studies to normalise levels.
Epilepsy has also been shown to be a risk factor for pathological fractures. Seizures can cause fractures in the setting of osteomalacia and osteoporotic bone, at the time of the seizure, as in the current case. Anticonvulsant therapy (ACT) alters vitamin D metabolism by inducing the hepatic oxidase enzyme, thus further lowering vitamin D levels.

Malnutrition due to a high catabolic rate and difficult feeding, together with central neurological causes, have also been cited as aetiological associations. Poor nutrition and stunted growth is well described in children with non-ambulatory CP.

Biomechanical abnormalities such as long lever arms and multiple joint contractures also play a contributory role in the aetiology of these fractures.

The assessment of low bone mineral density (BMD) in these patients is also associated with an increased risk of fragility fractures. Children with non-ambulatory CP should be assessed for osteoporosis. This can be done with the use of a DEXA scan using the distal femur as shown by Henderson et al. This method proved easier than the traditional hip and spine measurements and showed strong correlation with low bone mineral density (BMD) and pathological fractures in this patient population. Lower BMD Z scores, as in our patient, have been attributed to more severe GMFCS levels, difficult feeding and use of anticonvulsants. Sheridan found that in a large adult population of adults with disabilities the incidence of low bone density was 50 per cent. They also highlighted the need for better understanding of the ‘muscle-bone unit’ physiology as a means to improving the outcome of fragility fractures in this population.

Treatment with bisphosphonates is a relatively recent addition to the management of osteoporosis in these patients but has been shown to increase BMD and decrease fracture rates. There have, however, not been any studies comparing the effectiveness of bisphosphonates alone in comparison to sunlight, increased dietary calcium, vitamin D supplementation or adjustment of anti-convulsant therapy. Our patient did not receive bisphosphonates.

The patient in this case report had all of the above risk factors – contractures, immobility, resistant epilepsy, malnutrition, calcium and vitamin D deficiency and a low general physiological state.

The preferred orthopaedic treatment of femur fractures in children with non-ambulatory CP is conservative. Malunion in these patients is generally well tolerated as long as it does not interfere with perineal care and sitting balance. Conservative management in severe cerebral palsy has been described with good outcomes. McIvor et al. in 1966 reviewed 134 fractures in 92 patients with cerebral palsy. Over two-thirds of these fractures occurred in the femur and all were treated non-operatively. Although 85% of these patients healed in a malunion, they found that none of the malunions interfered with the function of the patients. Leet et al. also showed that fractures in non-ambulatory patients with CP can be treated conservatively. Of 32 femur fractures treated conservatively, one-third healed in a malunion and five resulted in cast-related pressure sores. They emphasised that special attention should be paid to prevent high rates of significant malunion as shown in the article by McIvor et al. Significant malunion was defined as resulting in discomfort, seating or caring difficulties. The risk of non-union in this population was negligible in both studies. Conservative treatment should be regarded as the gold standard but careful attention must be paid to addressing causes for osteoporosis, controlling epilepsy and preventing complications, specifically pressure sores (either related to cast or bed immobility), malunion and pneumonia (either related to immobility or aspiration).

Surgical management in non-ambulatory children with CP has also been described. In a study by Leet et al., six femur fractures in non-ambulators were treated operatively. One resulted in a malunion, one developed a cast-related pressure sore, one developed pneumonia and one developed a bacteraemia giving an overall complication rate of 67%. Treating femur fractures in non-ambulatory CP children with surgery is associated with a much higher complication rate and can be extremely challenging, as seen in our patient. Our initial primary goal with this patient was to alleviate her pain and allow her to be mobilised into a chair as soon as possible following the initial fracture. Surgical management in this case resulted in a high complication rate. Her seizure resulting in re-fracture requiring revision of her surgical fixation in extremely osteoporotic bone, and subsequent bacteraemia and deep wound sepsis highlights the major risk of complications in children with non-ambulatory cerebral palsy. Medical co-morbidities must be considered as well as low ambulatory potential, spasticity and osteopaenia. Leet et al. concluded that surgical management should be reserved for ambulatory CP children with unstable femur fractures.

Conclusion

This case study highlights the extremely high complication rate with the surgical management of femoral fractures in children with non-ambulatory cerebral palsy.
Although the conservative treatment of femoral fractures in these children is not without its complications, it should always be considered the gold standard. Epilepsy control, calcium and vitamin D supplementation have been shown to improve the bone health of these patients and should be considered as part of the management of femur fractures in children with non-ambulatory cerebral palsy.

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References

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