CASE REPORT

Fractures of the acetabulum in osteogenesis imperfecta

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We describe two patients aged 16 and 25 years with osteogenesis imperfecta who sustained displaced fractures of the acetabulum following minor trauma. The femoral heads were deformed by impact against the acetabular margin and both cases underwent surgical reconstruction. The quality of the bone and soft tissues made the operations challenging. There were potential complications specific to osteogenesis imperfecta, including bleeding, the creation of secondary fracture lines and shredding of the soft-tissue. The cases provide useful guidelines for addressing these difficulties.

In a healthy adolescent or adult, a displaced fracture of the acetabulum is usually treated by open reduction and fixation. Such management may be difficult in osteogenesis imperfecta because of possible deformities, such as protrusio acetabuli, osteopenia, a bleeding diathesis and friable tissues. We have treated displaced fractures of the acetabulum by operation in two patients with osteogenesis imperfecta and believe this to be the first instance of such patients to be published.

Case reports

A male patient, aged 16 years (weight 46.4 kg; height 159.6 cm) sustained a posterior dislocation of the right hip with a concomitant incomplete T-shaped fracture of the acetabulum while playing football (Figs 1a and 1c). In addition, a dimple-sign on the femoral head was observed on plain pelvic radiographs (Fig. 1c). He had osteogenesis imperfecta (Sillence Type IVa) and had previously sustained fractures of both femora. A syrinx of the thoracic cord (T5-T8) had been decompressed and he had generalised osteopenia. There was no neurovascular complication. The dislocation of the hip was reduced three hours after the accident and reconstruction was carried out six days later. A Kocher-Langenbeck approach was used with a digastric-slide osteotomy of the greater trochanter to allow better access to the anterior column. The soft tissues were thin and friable and the bone was markedly osteopenic. The posterior column was reduced and fixed with a 3.5 mm reconstruction plate (Fig. 1b). Due to marked protrusio acetabuli, the quadrilateral plate was very thin, so a screw could not pass through it to the anterior column without penetrating the hip joint. Therefore, a screw was passed transversely from the ilium to the anterior column. There was a total blood loss of 1500 ml. The post-operative period was uneventful. Touch weight-bearing began on the affected side from the second post-operative day and progressed slowly to partial weight-bearing by the end of the sixth week. Full weight-bearing was achieved by the end of the third post-operative month. At three months, there was no loss of position of the fracture.

A female patient, aged 25 years (weight 46 kg; height 142 cm), slipped as she was walking and sustained a displaced T-shaped right-sided fracture of the right acetabulum (Fig. 2a). She had osteogenesis imperfecta (Sillence Type IVb) and had sustained undisplaced fractures of the acetabulum at the age of 16. Skeletal traction was applied through the femur, and subsequently she was transferred to our hospital, where she underwent surgery ten days after the injury. The plain pelvic radiographs showed protrusio acetabuli (Fig. 2a) and a dimple-sign on the femoral head (Fig. 2c). An ilioinguinal approach was used. As in the previous case, a screw could not pass through the quadrilateral plate, so a four-hole plate was placed over the large pelvic brim plate to support the quadrilateral plate, as well as the posterior column fracture (Fig. 2b). The soft tissues were frail and consequently, the patient bled easily. She lost 1800 ml of blood and a cell saver was used. The post-operative period was uneventful. She was mobilised on crutches with touch weight-bearing for six weeks and partial weight-bearing for another six weeks.
Discussion
The fractures in these two cases were T-shaped, in accordance with the Judet-Letournel classification. This demanding pattern often requires two surgical approaches, or at the least, the passage of screws through the quadrilateral plate from one column to the other. We encountered protrusio acetabuli or a very thin quadrilateral plate, neither of which permits screw fixation.

In both cases, the femoral heads had a dimple-sign, due to impact with the edge of the acetabulum at the time of injury. If this defect had been of greater magnitude, it might have justified reconstruction or partial resurfacing, as has been described by Siguier et al. We did not undertake this because in case 1 the defect was small and in the second case the anterior approach did not allow clear visualisation of the depressed fragment. The chances of a good outcome with reconstruction of the acetabulum must be balanced against the benefits of early or late arthroplasty.

In patients with osteogenesis imperfecta, there is an incidence of bleeding diathesis in 10% to 30%. This coagulation defect is related to decreased capillary strength, as well as to the effect of abnormal collagen on platelet-endothelial cell interactions. Laboratory investigations in patients with osteogenesis imperfecta demonstrate increased capillary fragility, decreased platelet retention, decreased levels of Factor VIII and deficient collagen-induced platelet aggregation. Bleeding may still occur despite normal coagulation studies, thereby making predictions concerning intra-operative haemorrhage more difficult. The collagen defect may result in small blood vessels being unable to constrict adequately, a problem which is compounded by defective platelet aggregation around exposed subendothelium and generally friable tissues. In case 2, the iliopectineal fascia was hypoplastic and therefore very difficult to dissect without affecting the external iliac artery.

In most patients with osteogenesis imperfecta, protrusio acetabuli does not permit the passage of a screw through the
quadrilateral plate without penetrating the joint. If the fracture pattern requires the fixation of both columns, it is more appropriate to use two approaches. If the Kocher-Langenbeck approach is used, a digastric-slide trochanteric osteotomy might be beneficial in screw fixation of the anterior column. The surgical technique must be gentle because bony fragments can easily become comminuted. In both patients there was significant intra-operative haemorrhage. Use of a cell-saver is therefore recommended. The iliopectineal fascia and neighbouring tissues were hypoplastic and required special care during dissection in the ‘middle window’ of the ilioinguinal approach. The defect in the femoral head caused by its impaction against the acetabular edge (Bircher’s sign) may cause a predisposition to early degenerative changes.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References