Periacetabular osteotomy in patients with Down’s syndrome

We treated eight dysplastic acetabula in six skeletally mature patients with Down’s syndrome by a modified Bernese periacetabular osteotomy. The mean age at the time of surgery was 16.5 years (12.8 to 28.5). Mean length of follow-up was five years (2 to 10.4).

Pre-operatively the mean (Tönnis) acetabular angle was 28˚, the centre-edge angle was -9˚, and the extrusion index was 60%; post-operatively they were 3˚, 37˚, and 17%, respectively. Two patients with post-operative (Tönnis) acetabular angles > 10˚ developed subluxation post-operatively and required secondary varus derotation femoral osteotomies. Another patient developed a late labral tear which was treated arthroscopically. All eight hips remain clinically stable, and are either asymptomatic or symptomatically improved.

These results suggest that the modified Bernese periacetabular osteotomy can be used successfully in the treatment of acetabular dysplasia in patients with Down’s syndrome.

The existing literature suggests that the prevalence of acetabular dysplasia in skeletally mature patients with Down’s syndrome is between 10% and 25%. Acetabular dysplasia may lead to pain and decreased walking ability. Treatment may be complicated by generalised hypotonia and ligamentous and capsular laxity. Capsular plication, femoral and/or pelvic osteotomies have been suggested as methods of treatment. The results of acetabular shelf procedures, innominate, Sutherland, and Chiari osteotomies have been variable.

We report our experience with the modified Bernese periacetabular osteotomy in the treatment of acetabular dysplasia of eight hips in six patients with Down’s syndrome.

Patients and Methods
We describe six skeletally mature patients (eight hips) with Down’s syndrome who have undergone a modified Bernese periacetabular osteotomy for acetabular dysplasia. There were three girls and three boys. The mean age at the time of surgery was 16.5 years (12.8 to 28.5). The mean follow-up was 67 months (24 to 125). Two patients (three hips 1L, 5L, 5R) had hip pain, one had thigh pain (hip: 6L), and three had knee pain prior to the time of surgery (hips: 2R, 3R, 4L). One patient (case 5) required a wheelchair for long distances and one (2R) had a noticeable limp. One hip (1R) was not painful but showed progressive subluxation on radiographs. One patient (case 5) had had previous bilateral varus intertrochanteric femoral osteotomies and Pemberton osteotomies at four years of age. The lateral centre-edge angle, the acetabular angle of Tönnis, and the extrusion index were measured pre-operatively, immediately post-operatively and at the latest follow-up. Two of the femoral heads (hips: 3R and 4L) were aspherical and did not show concentric reduction on radiographs taken in abduction and internal rotation, but there was no evidence of hinging. Despite this, treatment by the modified Bernese periacetabular osteotomy was considered appropriate.

The modified Bernese periacetabular osteotomies were performed by the senior author (MBM) as described by Murphy and Millis. One hip (1R) had an intertrochanteric varus femoral osteotomy at the same time as the periacetabular osteotomy.

Results
All patients showed marked radiographic improvement of their dysplasia (Table I). The mean lateral centre-edge angle was -9˚ (-28 to +8) pre-operatively; 37˚ (15 to 49) post-operatively; and 37˚ (23 to 44) at the latest review. The mean acetabular angle of Tönnis was 28˚ (15 to 37) pre-operatively; 5˚ (-2 to +15˚) post-operatively; and 3˚ (-8 to +13) at the latest review. The mean extrusion index was 60%
(42 to 90) pre-operatively; 12% (0 to 33) post-operatively; and 17% (10 to 26) at the latest review (Fig. 1).

**Complications.** Further subluxation was noted approximately six months post-operatively in two patients (hips 3R and 4L). These hips had the highest Tönnis acetabular angles, 13˚ and 15˚. They were treated by varus derotation femoral intertrochanteric osteotomy with no recurrence of the subluxation (Fig. 2).

![Fig. 1a](image1a.png) ![Fig. 1b](image1b.png) ![Fig. 1c](image1c.png)  
Figure 1a – Dysplasia of the left hip in a 16-year-old female with Down’s syndrome with a pre-operative centre-edge angle of 8˚, Tönnis acetabular angle of 22˚ and the extrusion index of 47%. Figure 1b – After left periacetabular osteotomy the centre-edge angle was 47˚, the Tönnis acetabular angle 2˚ and the extrusion index of 0%. Figure 1c – At follow-up 24 months post-operatively the centre-edge angle was 44˚, the Tönnis acetabular angle 5˚ and the extrusion index of 10%. This patient developed an inferior ramus stress fracture that has been asymptomatic but shows signs of healing.

![Fig. 2a](image2a.png) ![Fig. 2b](image2b.png)  
Dysplasia of the right hip in a 15-year-old male with Down’s syndrome with a pre-operative centre-edge angle of -22˚, Tönnis acetabular angle of 33˚ and the extrusion index of 90%. Figure 2b – After right periacetabular osteotomy the centre-edge angle was 15˚, the Tönnis acetabular angle 15˚ and the extrusion index of 33%. Figure 2c – Recurrent subluxation approximately six months post-operatively. Figure 2d – Restoration of femoral head cover following varus derotation intertrochanteric femoral osteotomy. Figure 2e – At latest follow-up the centre-edge angle was 31˚, Tönnis acetabular angle 11˚ and the extrusion index of 20%.

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### Table I. Radiographic indices

<table>
<thead>
<tr>
<th>Patient/hip</th>
<th>Age at surgery (yrs)</th>
<th>Follow-up (yrs)</th>
<th>Pre-operative</th>
<th>Final follow-up</th>
<th>Complications/comment</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>CE˚</td>
<td>Tonnis acetabular angle (˚)</td>
<td>Extrusion index (%)</td>
</tr>
<tr>
<td>1/R</td>
<td>14.2</td>
<td>10.4</td>
<td>0</td>
<td>35</td>
<td>53</td>
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<tr>
<td>1/L</td>
<td>15</td>
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<td>5</td>
<td>15</td>
<td>42</td>
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<tr>
<td>2/R</td>
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<td>32</td>
<td>74</td>
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<tr>
<td>3/R</td>
<td>15.6</td>
<td>4.2</td>
<td>-22</td>
<td>33</td>
<td>90</td>
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<tr>
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<td>-27</td>
<td>37</td>
<td>62</td>
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<td>4.1</td>
<td>-10</td>
<td>30</td>
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<tr>
<td>5/L</td>
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<td>3.2</td>
<td>-2</td>
<td>22</td>
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<tr>
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<td>16.3</td>
<td>2</td>
<td>8</td>
<td>22</td>
<td>47</td>
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</table>

* CE, centre-edge  
† ITO – intertrochanteric femoral osteotomy, PAO – periacetabular osteotomy, I+D – incision and drainage
One patient (hip: 5L) developed a labral tear which became symptomatic 20 months post-operatively. At that time radiographs showed a centre-edge angle of 43°, a Tönnis acetabular angle of -8° and an extrusion index of 18%. The labral tear was treated by arthroscopic debridement with relief of pain. However, she subsequently developed recurrence of mild mechanical symptoms.

One patient (hip: 2R) had an ischial nonunion which persists but is asymptomatic at seven years follow-up. Another patient (hip: 6L) had a stress fracture of the inferior pubic ramus which remains asymptomatic and shows signs of healing two years post-operatively. Another (hip: 5R) developed a sterile post-operative wound haematoma requiring operative drainage two weeks post-operatively.

Discussion
Acetabular dysplasia in infancy in Down’s syndrome is rare. In skeletally mature patients with Down’s syndrome, the bony acetabulum is often deeper and more horizontally placed than usual.1,11-14 However, approximately 5% of patients with Down’s syndrome develop dislocation of the hip before the age of ten years.3 Untreated, habitual dislocators typically progress to fixed subluxation, dislocation, progressive dysplasia with difficulty in walking, and often painful osteoarthritis.5,15 As the lifespan of patients with Down’s syndrome has increased6 the need for increased attention to the orthopaedic manifestations in adult life has emerged.

Hresko et al3 found that in an institutionalised population of 65 adults with Down’s syndrome, 28% had radiographic abnormalities of one or both hips. Sixteen of the 18 patients with abnormal radiographs had dysplasia and/or osteoarthritis. One patient had a unilateral slipped capital femoral epiphysis and one had unilateral protrusio acetabuli. Treatment of these abnormalities was not discussed. This study also indicated that acetabular dysplasia was associated with a deterioration in mobility.

The existing literature offers little guidance regarding the treatment of acetabular dysplasia in the mature patient with Down’s syndrome. Operative treatment has consisted of soft-tissue and bony procedures, either separately or combined. The primary soft-tissue procedure recommended has been capsular plication. Gore17 supplemented his capsular plication with a tether to the acetabulum. He divided the superior and inferior gemelli and the obturator internus in their mid portion and sutured the cut edge of the distal segment to the posterior acetabulum.17 Capsular plication without osteotomy has been successful in the treatment of dislocation of the hip in patients under ten years of age with Down’s syndrome, but in these series there was no mention of the degree of acetabular dysplasia.5,17

Various osteotomies have been used in the treatment of acetabular dysplasia in patients with Down’s syndrome, with variable results.4,6 Aprin et al6 described the management of six children with Down’s syndrome and dislocation of the hip. The three younger patients, aged 3 to 5 years, had bilateral dislocatable hips and normal acetabula. The three older patients, aged 9 to 13 years, had unilateral or bilateral dislocation with dysplastic acetabula. All three younger patients were treated by varus femoral osteotomy and one also had a capsular plication. The three older patients were treated by open reduction, capsular plication, varus intertrochanteric femoral osteotomy, and innominate or Chiari osteotomy. Two of the younger patients had an excellent result and one a good result. In the older patients, there was one good result, one fair, and one poor.

In our series of six patients (eight hips), the modified Bernese periacetabular osteotomy has been an effective treatment for acetabular dysplasia in skeletally mature patients with Down’s syndrome. However, secondary varus intertrochanteric femoral osteotomy was necessary for recurrent subluxation in two hips. All patients showed marked improvement in clinical function and radiographic indices. All are community walkers and some participate in athletics. Five of the eight hips are asymptomatic, but three (hips: 1L, 4L, 5L) have mild symptoms on strenuous activity.

Two patients (with Tönnis acetabular angles of 13° and 15°) developed recurrent subluxation approximately six months post-operatively. It has been suggested previously that a Tönnis acetabular angle outside the normal range (-10° to 10°) may be an indication for proximal femoral osteotomy.18 Pre-operatively, both of these patients were noted to have aspherical femoral heads without congruous reduction on abduction and internal rotation radiographs, although no hinging was seen on these views. These patients, therefore, would not be considered ideal candidates for this procedure. Both these hips required secondary varus femoral derotation intertrochanteric osteotomy to correct the recurrent subluxation. At final follow-up both remained stable, showed clinical improvement and no deterioration of arthrosis. As a result we believe that the modified Bernese periacetabular osteotomy was preferable to alternative procedures such as Chiari osteotomy, hip arthrodesis and total hip arthroplasty although the radiographic criteria were not ideal. In future perhaps varus intertrochanteric femoral osteotomy should be carried out at the same time or the hip monitored carefully for subluxation after periacetabular osteotomy.

One patient developed acute groin pain while running 20 months after periacetabular osteotomy. A magnetic resonance arthrogram suggested a labral tear. Radiographs at that time showed a centre-edge angle of 43° and a Tönnis acetabular index of -8° (i.e. varus source) which may have led to impingement and the resultant labral tear.19 Arthroscopic labral debridement resulted in relief of the acute symptoms, but mild mechanical symptoms recurred 18 months after arthroscopy.

Management of acetabular dysplasia in the skeletally mature patient with Down’s syndrome is challenging. Periacetabular osteotomy allows major dramatic correction of the acetabular orientation. This small series suggests that
correction of the radiographic parameters to normal in these patients is important in order to maintain hip stability. A post-operative Tönnis acetabular angle of > 10˚ may predispose to recurrent subluxation. Conversely, the patient with the greatest correction of the sourcil beyond neutral to -8˚ developed a labral tear which may have been secondary to impingement. We recognise that optimal radiographic correction remains uncertain and that longer follow-up is important in this difficult group of patients. However, we have found the modified Bernese periacetabular osteotomy to be an effective treatment for acetabular dysplasia in skeletally mature patients with Down’s syndrome.

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