HIP PATHOLOGIES THAT BEDEVIL

Surgical challenges and clinical outcomes of total hip replacement in patients with Down’s syndrome

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Down’s syndrome is associated with a number of musculoskeletal abnormalities, some of which predispose patients to early symptomatic arthritis of the hip. The purpose of the present study was to review the general and hip-specific factors potentially compromising total hip replacement (THR) in patients with Down’s syndrome, as well as to summarise both the surgical techniques that may anticipate the potential adverse impact of these factors and the clinical results reported to date. A search of the literature was performed, and the findings further informed by the authors’ clinical experience, as well as that of the hip replacement in Down Syndrome study group. The general factors identified include a high incidence of ligamentous laxity, as well as associated muscle hypotonia and gait abnormalities. Hip-specific factors include: a high incidence of hip dysplasia, as well as a number of other acetabular, femoral and combined femoroacetabular anatomical variations. Four studies encompassing 42 hips, which reported the clinical outcomes of THR in patients with Down’s syndrome, were identified. All patients were successfully treated with standard acetabular and femoral components. The use of supplementary acetabular screw fixation to enhance component stability was frequently reported. The use of constrained liners to treat intra-operative instability occurred in eight hips. Survival rates of between 81% and 100% at a mean follow-up of 105 months (6 to 292) are encouraging. Overall, while THR in patients with Down’s syndrome does present some unique challenges, the overall clinical results are good, providing these patients with reliable pain relief and good function.

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In addition to the well-recognised phenotypic characteristics associated with Down’s syndrome, this condition is associated with a number of musculoskeletal abnormalities that can lead to symptomatic, degenerative hip pathology. It is associated with muscle hypotonia and generalised ligamentous laxity,8–10 and has also been linked to an increased incidence of slipped capital femoral epiphysis, Perthes’ disease and posterior acetabular insufficiency.11–13 These factors can be expected to result in a high incidence of symptomatic degenerative hip pathology at a relatively young age. Given the continuing increased life expectancy of these patients, it is likely that orthopaedic surgeons can expect to see an increasing number with end-stage arthritis of the hip. Recently, Gill et al14 described a patient who had to seek opinions from four orthopaedic surgeons before total hip replacement (THR) was suggested, which highlights the potential barriers to surgical treatment faced by patients with Down’s syndrome.14 While there are a number of both anatomical and patient comorbidities that make THR in these patients more challenging when compared with the standard procedure, several authors have reported good outcomes in this group with the use of appropriate pre-operative
planning and intra-operative techniques.\textsuperscript{15-17} Coppus et al\textsuperscript{6} reported that mobility restrictions are among the disorders most related to mortality in older patients with Down’s syndrome. The purpose of the present study is to review the general and hip-specific factors potentially compromising THR in patients with Down’s syndrome, and how to deal with them as well as the reported clinical outcomes.

Patients and Methods

This review is based on a search of publications using the PubMed indexing service for terms including Down’s syndrome, trisomy 21, hip abnormalities, and hip replacement. Results were reviewed to identify commonly reported general, as well as hip-specific clinical and anatomical factors potentially influencing THR in patients with Down’s syndrome. All original English language studies reporting on the clinical outcomes of THR in patients with Down’s syndrome published until November 2012 were identified, and the results were reviewed. Finally, the authors’ own clinical experiences in the Replacement Arthroplasty in Down Syndrome (RADS) study group, which were recently reported in the orthopaedic literature,\textsuperscript{17} were used to inform the present review.

Results

General factor: ligamentous laxity. This is frequently cited as a characteristic of these patients. Mik et al\textsuperscript{18} observed that the genes that encode for type VI collagen are located on chromosome 21, and are believed to be responsible for laxity. Semine et al\textsuperscript{8} reported that 65 (76.4\%) of 85 children with Down’s syndrome had generalised joint laxity using the criteria of Carter and Wilkinson.\textsuperscript{19} However, Livingstone and Hirst\textsuperscript{20} evaluated all children with Down’s syndrome living in the authors’ health district for ligamentous laxity, and found that while 23 (59\%) of 39 children had one or more lax joints according to the Carter and Wilkinson criteria,\textsuperscript{19} only two of 39 (5\%) met the criteria for generalised joint laxity (three of the five tests positive, including both upper and lower joints, bilaterally). This was similar to the baseline incidence of 7\% for all children reported by Carter and Wilkinson.\textsuperscript{19} The true incidence of ligamentous laxity in persons with Down’s syndrome remains unclear. Several authors have demonstrated a high incidence of cervical spine hypermobility, reaching as high as 79\% for the atlanto-occipital joint.\textsuperscript{21,22} Similarly, Shaw and Beals\textsuperscript{23} reported that the range of movement of the hip was greater in all planes than would be expected in the general population. However, it remains unclear whether this is specifically the result of ligamentous laxity, and if so, what contribution if any it makes to the development of hip joint pathology, or to instability following THR in these patients.

General factor: muscle hypotonia and gait abnormalities. Downs syndrome has been associated with generalised muscular hypotonia. Morris et al\textsuperscript{9} evaluated 28 children with Down’s syndrome and 22 normal children, and found decreased muscle tone and lower grip strength in the children with Down’s syndrome. Pitetti et al\textsuperscript{10} compared isokinetic arm and leg strength in individuals with Down’s syndrome, with those with learning difficulties not associated with Down’s syndrome, and normal sedentary young adults. The authors found significantly lower leg strength in individuals with Down’s syndrome compared with both unaffected groups, suggesting that muscular hypotonia is associated with the syndrome itself, and not simply as a result of a less active lifestyle.

Several authors have reported that patients with Down’s syndrome have a more unstable gait pattern when compared with the general population.\textsuperscript{24,25} They have increased medial-lateral motion of their centre of mass, and both decreased and more variable step length and speed when compared with control subjects.\textsuperscript{24} While patients with Down’s syndrome have been reported to have generally increased passive range of movement of the hip joint, both Cimolin et al\textsuperscript{26} and Galli et al\textsuperscript{27} found increased individual hip and knee stiffness in patients with Down’s syndrome, suggesting a decreased effectiveness of dynamic joint stabilisation during the gait cycle.

These findings have two-fold implications for patients undergoing THR. Firstly, the generalised muscle hypotonia suggests that these patients may have greater difficulty with rehabilitation after surgery, especially, if preceded by a prolonged period of immobilisation. Secondly, because of impaired dynamic hip joint stabilisation, it is important to maximise the intrinsic stability of the replacement hip. Strategies for maximising implant stability include: the use of large femoral heads, constrained acetabular liners, or post-operative hip bracing. Given the mobility restrictions associated with bracing, as well as potential difficulties with compliance, the use of higher-stability implants is preferable.

Hip-specific factor: dysplasia. While many patients may have anatomical abnormalities of the hip joint, only a certain subset can be expected to develop potentially symptomatic hip joint pathology. Hresko, McCarthy and Goldberg\textsuperscript{28} reviewed hip radiographs for a subset of 65 mostly adult patients with Down’s syndrome in a single institution in Massachusetts, and found radiological evidence of hip pathology in 22\% of patients. Additionally, the authors found that radiological hip pathology was associated with impaired mobility.

Dysplasia and/or subluxation of the hip joint appear to be among the most commonly detected radiological hip pathologies. Hresko et al\textsuperscript{28} reported on these abnormalities in 16 hips in nine patients, of a total of 65 primarily adult patients reviewed (12\% of hips, 14\% of patients). Dysplasia of the hip appears to be the most common indication for THR based on the published results to date, being found in between 54\% and 100\% of hips in each reported series.\textsuperscript{15-17} Hresko et al\textsuperscript{28} identified five dislocated hips in three patients out of the total of 65 primarily adult patients with Down’s syndrome (4\% of hips, 5\% of patients), while Bennet et al\textsuperscript{29} identified ten pediatric and adult patients with dislocated or dislocatable hips out of a total of
acetabular components. If there are concerns about cœur bony contact to allow the implantation of standard components, many cases of dysplastic acetabuli, there is frequently sufficient superolateral coverage and/or posterior wall bone mass in the typical safe zone for all hip replacements regardless of the pre-operative deformity, namely 40° (±10°) of abduction, and 15° (±10°) of anteversion. Surgeons should try to reproduce the native centre of rotation of the femoral head, and avoid excessive medialisation of the acetabular component, which could result in instability of the joint due to reduced offset. While there may be reduced superolateral coverage and/or posterior wall bone mass in many cases of dysplastic acetabuli, there is frequently sufficient bony contact to allow the implantation of standard acetabular components. If there are concerns about acetabular component fixation, surgeons can additionally use a highly porous metal acetabular shell and/or bone grafting of the superior acetabulum. Surgeons should have a low threshold for the use of supplementary screw fixation to enhance acetabular stability. In cases of high riding fixed dislocation of the hip, it may be necessary to perform a subtrochanteric shortening osteotomy of the femur to obtain adequate positioning of the acetabular components, while minimising the risk of sciatic nerve traction injury.

Hip-specific factor: proximal femoral and combined femoral and-acetabular deformities. Roberts et al reviewed pelvic radiographs for 66 adults with Down’s syndrome residing in the typical safe zone for all hip replacements regardless of the pre-operative deformity, namely 40° (±10°) of abduction, and 15° (±10°) of anteversion. Surgeons should try to reproduce the native centre of rotation of the femoral head, and avoid excessive medialisation of the acetabular component, which could result in instability of the joint due to reduced offset. While there may be reduced superolateral coverage and/or posterior wall bone mass in many cases of dysplastic acetabuli, there is frequently sufficient bony contact to allow the implantation of standard acetabular components. If there are concerns about acetabular component fixation, surgeons can additionally use a highly porous metal acetabular shell and/or bone grafting of the superior acetabulum. Surgeons should have a low threshold for the use of supplementary screw fixation to enhance acetabular stability. In cases of high riding fixed dislocation of the hip, it may be necessary to perform a subtrochanteric shortening osteotomy of the femur to obtain adequate positioning of the acetabular components, while minimising the risk of sciatic nerve traction injury.

Surgical techniques and clinical outcomes

The senior author (AEG) recently reported the clinical outcomes of THR in patients with Down’s syndrome as part of a multi-centre study encompassing 26 hips in 21 patients. A subset of these patients had previously been reported elsewhere. To the best of our knowledge there have only been three other studies to date specifically examining this patient population, evaluating nine hips in six patients,
eight hips in five patients, and two hips in one patient, respectively.\textsuperscript{15,16,36} The results of these studies will be reported in aggregate below.

**Acetabular fixation and stability.** The large majority of THRs reported to date in this patient population have been performed using cementless acetabular components (Table I). While Kioschos et al\textsuperscript{15} and Skoff and Keggi\textsuperscript{16} did not specify whether supplementary screw fixation was used in their studies, acetabular screws were used to enhance stability in all but one of the remaining 28 hips. Eight hips in six patients reported by Gross et al\textsuperscript{17} additionally received constrained liners to address instability identified intraoperatively. Of the 27 hips for which the use of ancillary fixation was specified, there were no cases of aseptic acetabular loosening reported at follow-up times ranging from six to 292 months. There was one case of acetabular loosening at seven-year follow-up reported by Kioschos et al\textsuperscript{15} although it is unclear whether any supplementary acetabular fixation was used. One patient treated with an unconstrained liner suffered recurrent dislocations in the early post-operative period, and underwent revision of the acetabular component using a constrained liner, with no further dislocations at four year follow-up.

**Femoral components.** The large majority of THRs reported to date in patients with Down’s syndrome were performed using cementless femoral components, with cemented fixation used in only two of 33 hips (Table I). Solayar et al\textsuperscript{36} reported using modular neck cementless femoral components for both hips in the case report. The remaining procedures reported to date were performed with standard cementless femoral components, with no authors reporting the use of custom stems. Two cases of symptomatic aseptic femoral component loosening requiring revision have been reported to date at approximately six and 16 years respectively following the index procedures. Both of these occurred in the series reported by the RADS study group.\textsuperscript{17}

**Clinical outcomes.** Gross et al\textsuperscript{17} reported an 81% survival rate (21 hips) at a mean follow-up time of 106 months (25 to 292). One hip had undergone two-stage revision for peri-prosthetic infection approximately six months following primary surgery, yielding a survival rate of 85% (22 hips) with aseptic failure as the endpoint. All patients had improvements in their Harris hip scores\textsuperscript{37} at final follow-up when compared with pre-operative values, with a mean score in surviving hips of 83 (55 to 100) at final follow-up. Consistent results were reported by Kioschos et al\textsuperscript{15} who evaluated nine hips in seven patients treated with either THR or bipolar hip replacement. The authors reported a survival rate of 89% at a mean follow-up of 95 months (24 to 168), with all patients pain-free and walking well. Both Solayar et al\textsuperscript{36} and Skoff and Keggi\textsuperscript{16} reported no revisions at mean follow-ups of six months and four years respectively.

**Discussion**

Patients with Down’s syndrome experience a number of different musculoskeletal abnormalities. As the life expectancy for these patients continues to increase, more are presenting with advanced degenerative disease of the hip suitable for treatment with THR. Because of the increased risk of decreased bone density and muscle hypotonia in these patients and the association between reduced mobility and mortality, THR can contribute to improving the quality of life in these patients. Orthopaedic surgeons can expect some of these patients to present with a degree of hip joint deformity and anatomical abnormalities, an awareness of which is relevant to ensuring the best results with hip replacement in this group. Good clinical outcomes at follow-up approaching a decade or more can be achieved for THR in patients with Down’s syndrome, although in the authors’ experience as well as that previously reported by other surgeons, re-revision rates are somewhat higher than would be expected in the general population.

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**Table I. Summary of results of total hip replacement in patients with Down’s syndrome reported to date (n/a, not available)**

<table>
<thead>
<tr>
<th>Author/s</th>
<th>Patients/hips (n)</th>
<th>Mean age (yrs) (range)</th>
<th>Associated diagnoses*</th>
<th>Femoral component</th>
<th>Acetabular component</th>
<th>Mean clinical score (range)†</th>
<th>Hips surviving at final follow-up (%)</th>
<th>Revisions‡</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gross et al\textsuperscript{17}</td>
<td>21/26</td>
<td>35 (17 to 54)</td>
<td>DDH (14), DJD (6), SCFE (4), LCP (1), PTA (1)</td>
<td>Uncemented (25), Standard (28), cemented (1)</td>
<td>Screws (26) 8</td>
<td>105 (25 to 292)</td>
<td>HHS: 62 (19 to 83)</td>
<td>21 (47)</td>
</tr>
<tr>
<td>Solayar et al\textsuperscript{36}</td>
<td>1/2</td>
<td>19</td>
<td>DDH (3)</td>
<td>Uncemented (2), Modular (2), cemented (1)</td>
<td>Screws (1) 0</td>
<td>6</td>
<td>WOMAC: 98, VAS 19, VAS 1</td>
<td>2 (100)</td>
</tr>
<tr>
<td>Kioschos et al\textsuperscript{15}</td>
<td>6/9</td>
<td>36 (22 to 47)</td>
<td>DDH (4), DJD ON (3), SCFE (1), ON (1)</td>
<td>Uncemented (6), Standard (9), cemented (1), bipolar (3)</td>
<td>n/a</td>
<td>n/a</td>
<td>95 (24 to 108)</td>
<td>n/a</td>
</tr>
<tr>
<td>Skoff and Keggi\textsuperscript{16}</td>
<td>5/8</td>
<td>46 (27 to 64)</td>
<td>DDH (8)</td>
<td>n/a</td>
<td>Standard (8), custom (0)</td>
<td>n/a</td>
<td>n/a</td>
<td>52 (25 to 108)</td>
</tr>
</tbody>
</table>

* DDH, developmental dysplasia of the hip; DJD, degenerative joint disease; SCFE, slipped capital femoral epiphysis; LCP, Legg–Calvé–Perthes’ disease; PTA, post-traumatic arthritis; ON, osteonecrosis
† HHS, Harris hip score; WOMAC, Western Ontario and McMaster Universities osteoarthritis index; VAS, visual analogue scale for pain
‡ POD, post-operative day
population. Nevertheless, with awareness of the unique aspects of musculoskeletal disease in this patient population, good pre-operative planning, excellent surgical technique, and appropriate implant choice with selective use of acetabular constraint, THR can provide these patients with reliable pain relief and improved function.

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References