**Pathologic Hip Morphology in Cerebral Palsy**

**Epidemiology:** Cerebral palsy is a non-progressive disorder of the central nervous system with variable impairment of motor function. Hip dysplasia in CP is the second most common musculoskeletal deformity, second only to equinus.[1] The incidence and severity of hip dysplasia disease is directly proportional to the extent of neuromuscular impairment most often quantified by gross motor classification system (GMFCS). For example, 3-7% of CP patients who walk (GMFCS I-III) develop hip dysplasia as opposed to 70% of CP patients who do not walk (GMFCS IV-V).[2] Classification of hip dysplasia in CP patients combines acetabular and femoral morphologic changes in association with migration of the femoral head from the acetabulum and is described cumulatively in the cerebral palsy hip classification system (CPHCS).[3]

**Natural History:** Early improper development of the proximal femur and the acetabulum results in early instability (4-12 years of age). In milder cases, increased tone in later stages of development (often associated with hydrocephalus or shunt dysfunction) can also result in late subluxation or dislocation.[1] As the majority of hip pathology occurs in the more impaired CP patients (GMFCS IV-V) the report of long term outcomes has been infrequently cited as this patient population is more likely to sustain early death and less likely to have the capacity to localize pain.[4, 5] From reports available on long term outcome it is estimated that approximately 50% of patients with CP related hip subluxation or dislocation of the hip have hip pain after skeletal maturity.

**Soft Tissue Pathology:** Pathologic muscular forces (tone and balance) across the hip during development are thought to be the primary contributing factors to pathomorphology of the femur and acetabulum leading to hip instability. Increased tone in the hip flexors and adductors overpower the relatively weaker hip extensors and abductors leading eventually in a shift of the center of rotation of the hip from the femoral head to the lesser trochanter.[1, 2, 6, 7] Secondary to this characteristic muscle imbalance in combination with abnormal proximal femoral and acetabular development the majority of instability occurs in the posterior, lateral and superior direction.

**Proximal Femoral pathomorphology:** Change in femoral geometry is directly proportional to severity of neuromuscular disease (GMFCS level). The pathologic morphology has been reported by multiple observers and most commonly consists of increased femoral neck valgus and anteverision.[1, 2, 6, 7] Other pathologies may include deformation of the femoral head secondary to persistent pressure by the labrum, rim of the acetabulum or the abductors in the subluxed or dislocated hip and coxa valga from growth disturbances of the proximal femoral physis.

**Acetabular Morphologic Changes:** Pathologic development of the acetabulum is directly proportional to pathologic morphology of the proximal femur and therefore the severity of the neuromuscular disease (GMFCS).[1-3, 6, 7] Acetabular dysplasia occur secondary to alteration of muscular forces and migration of the center of rotation of the femoral head from the center of rotation of the acetabulum. These changes cumulatively result in primarily posterior and lateral deficiencies.
**Pathologic Hip Morphology in Down syndrome**

**Epidemiology:** Trisomy 21, first described in 1866, is the most common chromosomal abnormality with an incidence of 1 in 795 live births. Life expectancy of a 1 year old child with Down syndrome is between 43-55 years. Hip instability reported to occur in 1.3% to 7.0% during childhood. Hip pathology increases to 28% of adult patients.[8]

**Natural History:** Down syndrome patients who develop hip dysplasia initially have stable hips before walking age. Progressive instability develops typically around 7-8 years old with subluxation potentially leading to fixed dislocation and/or osteoarthritis. Down syndrome patients with problematic hip pathology are more likely to become non-ambulatory with use of wheelchair.[8]

**Soft Tissue Pathology:** Capsular insufficiency, ligamentous laxity and hypotonia are the main contributing factors in Down syndrome hip dysplasia.[8] These connective tissue pathologies lead to altered dynamics of hip motion during development resulting primarily in a deficient posterior wall of the acetabulum (relative acetabular retroversion).[9-11]

**Proximal Femoral Morphology:** The pathomorphology of the proximal femur in Down syndrome remains a debate. The proximal femur in Down’s syndrome has been described as having only a slight increase of anteversion and near-normal neck-shaft angle[12] whereas other studies report an more significant increase proximal femoral valgus[13].

**Acetabular Morphology:** In Down syndrome patients with hip instability the morphology of the acetabulum has most often described is a deficient posterior wall which results in a shallow, relatively retroverted acetabulum.[8, 9, 11] In a study of Downs syndrome age and sex matched controls, the average acetabular version of the Down syndrome patient with subluxation or dislocation was significantly more retroverted than non-dysplastic normal children and patients with idiopathic hip dysplasia.[10]
## Pathologic Hip Morphology in Cerebral Palsy

<table>
<thead>
<tr>
<th></th>
<th>GMFCS I</th>
<th>GMFCS II</th>
<th>GMFCS III</th>
<th>GMFCS IV</th>
<th>GMFCS V</th>
</tr>
</thead>
<tbody>
<tr>
<td>FNA</td>
<td>FNA = 30°</td>
<td>FNA = 38°</td>
<td>FNA = 40°</td>
<td>FNA = 40°</td>
<td>FNA = 40°</td>
</tr>
<tr>
<td>NSA</td>
<td>NSA = 136°</td>
<td>NSA = 141°</td>
<td>NSA = 149°</td>
<td>NSA = 155°</td>
<td>NSA = 163°</td>
</tr>
<tr>
<td>MP</td>
<td>MP = 8%</td>
<td>MP = 13%</td>
<td>MP = 25%</td>
<td>MP = 37%</td>
<td>MP = 46%</td>
</tr>
</tbody>
</table>

Prevailing morphologic changes in hips of patients with cerebral palsy: Severity of disease is predicted by gross motor function classification system (GMFCS). The combination of muscular forces results in increased femoral anteversion (FNA) and valgus femoral neck shaft angle (NSA) resulting in an increased migration percentage (MP). Reproduced with permission and copyright © of the British Editorial Society of Bone and Joint Surgery [2].

## Pathologic Hip Morphology in Down syndrome

Prevailing morphologic changes in hips of patients with Down’s syndrome: Severity of disease is dependent on the combination of ligamentous and capsular laxity and hypotonia with changes in the acetabular morphology. Deficient posterior wall results in relative retroversion and posterior instability.
References