

# A review of cerebral palsy and hip surveillance in the UK

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## INTRODUCTION

Cerebral palsy (CP) has many comorbidities including hip dislocation. This morbidity occurs during childhood and mostly affects children with a spastic bilateral subtype of CP. Hip dislocation is often painful and leads to difficulty moving and sitting, pelvic obliquity and poor perineal care. Measuring hip lateralisation of the femoral head at an early stage in children who are most at risk can detect early hip subluxation. It is important that the method of measuring hip lateralisation can accurately measure the severity of the subluxation. It is also vital that children with bilateral CP are screened at the age that most accurately indicates the risk of hip dislocation for the future so that treatment can start at an appropriate time.

The aim of this study is to analyse current National Institute of Health and Clinical Excellence (NICE) guidelines that describe the method of measurement of hip lateralisation and the age at which hip screening begins in children with bilateral CP. To analyse these particular guidelines I will compare them against research that has been published in those fields.

## CEREBRAL PALSY

A recent international working group produced a report that defined CP as 'a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing foetal or infant brain.'<sup>(1)</sup> These disturbances in the infant or foetal brain refer to lesions in the brain that are caused by some sort of injury to the neural tissue.<sup>(2)</sup> The definition refers to CP as a 'group' of disorders demonstrating that CP has subtypes.<sup>(1)</sup> The most common subtype is spastic CP, which is diagnosed 77% of the time in children with CP.<sup>(3)</sup> There are further subtypes of spastic CP. These include unilateral spastic disease (spastic monoplegia and hemiplegia) and bilateral spastic disease (spastic diplegia, tetraplegia and quadriplegia). The other subtypes of CP can be classified into ataxic and dyskinetic (choreoathetoid and dystonic).

The prevalence of CP ranges from 1.5 to 2 for every 1,000 live births,<sup>(4)</sup> and is the most common motor impairment disorder among children.<sup>(5)</sup> Children with CP also have many other clinical manifestations. These include joint problems from the skeletal muscle group that has been affected by spasticity subtypes of CP. These problems include contractures, subluxations, dislocations and pain found at the affected joints. These are all seen in the hips of children.

## CEREBRAL PALSY AND THE HIP

At birth, the hip of a child with CP is no different from a child without CP.<sup>(6)</sup> Therefore, the classical pathological problems (coxa valga antetorsa and femoral anteversion)<sup>(7)</sup> associated with the hip in children with spastic bilateral CP occur during the child's development.

The pathophysiological model for the coxa valga antetorsa and femoral anteversion describes how the reduced activity of gluteus maximus, gluteus minimus and quadriceps femoris found in spastic CP children who are unable to walk increases their risk of both of the common pathologies.<sup>(7)</sup> This model describes how the increase in the contraction of the spastic hip adductors and the reduced activity of the hip abductors causes slower growth of the greater trochanter. This means that the abductors are stretched to a more vertical position and the femoral neck growth plate becomes more horizontal. This leads to coxa valga. This model demonstrated that walking and sitting were major milestones in preventing any hip dislocations in the future and missing these milestones can give the first signs of CP.<sup>(7)</sup>

Spastic contraction of the adductor and flexor muscle groups and a weakness of the abductor muscle groups of the hip joint also causes acetabular deformity.<sup>(8)</sup> Recently it has been found that acetabular is not a prerequisite for hip dislocation as hip lateralisation is rarely preceded by acetabular dysplasia.<sup>(9)</sup>

It has been well documented that walking and sitting prevents hip dislocation.<sup>(7)</sup> In the past, researchers have tried to classify which subtypes of CP are more at risk of hip dislocation due to some subtypes having different ranges of movement than others.<sup>(10)</sup> This has resulted in different levels of risk for different subtypes as shown in Table 1.

Low-risk group	Intermediate-risk group	High-risk group
Ataxic	Spastic diplegia	Spastic tetraplegia
Spastic hemiplegia	Dyskinetic	

*Table 1 This table has grouped together the various subtypes of CP to give the level of risk of hip dislocation that is associated with each subtype<sup>(10)</sup>*

This type of grouping, however, is not useful to clinicians. This is because the universal definitions for the subtypes of CP have not been agreed upon internationally. Another reason is that some of the subtypes are difficult to tell apart, for example spastic hemiplegia and diplegia with asymmetric severity.

To address this problem, a five-level classification system called gross motor function classification system (GMFCS) has been developed.<sup>(11)</sup> Each level has a different set of criteria that describes a child's motor function which is demonstrated in Table 2.

Levels	Criteria for the GMFCS for children aged 6-12
I	Walks without restrictions, limitations in more advanced gross motor skills
II	Walks without restrictions, limitations walking outdoors and in the community
III	Walks with assistive mobility devices, limitations walking outdoors and in the community
IV	Self-mobility with limitations, children are transported or use power mobility outdoors and in the community
V	Self-mobility is severely limited, even with the use of assistive technology

Table 2 This is only a summary of the criteria used for the GMFCS for children aged 6-12 years. Other tables with adapted GMFCS criteria are used for children of different ages. This is carried out to standardise results across a large population of children with different ages<sup>(11)</sup>

Two studies have successfully used GMFCS to show that children in levels III, IV and V were more at risk of hip dislocation than others.<sup>(9,12)</sup> This system removed the problems that were associated with designating subtypes of CP with different levels of risk.

The natural age of hip dislocation for children with CP varies depending on which study you read. The most recent and reliable study showed that the mode age at which dislocation occurred was seven years, but there was a group of patients that didn't dislocate until they were 12-17 years.<sup>(9)</sup>

The incidence of hip dislocation among children with CP ranges from 10-20% if not treated,<sup>(13,14)</sup> but it is mostly associated with children with bilateral types of CP. Hip dislocation/subluxation is associated with pain in over 50% of cases.<sup>(15)</sup> Hip pain can cause difficulty sitting and moving, and comorbidities such as pelvic obliquity and scoliosis can occur.<sup>(14,16)</sup> Perineal care is also made difficult, increasing the chances of infection to the patient.<sup>(9)</sup> Therefore, hip subluxation causes a reduction in the child's quality of life and it is vital to prevent this.

## MEASUREMENT OF HIP LATERALISATION AND THE NICE GUIDELINES

Radiological imaging is considered the best way to gain a picture of the position of the femoral head in the acetabulum in children with CP. A study in 1997 described the standardised position that is used for radiological imaging of the hip.<sup>(17)</sup> This included the child lying flat on the bed with their legs parallel to each other with the patella facing upwards. Some children will have tight hip flexors which will give them a lordotic pelvis and obscure the important horizontal view of the pelvis needed for assessment. To prevent this problem, the pillow can be placed under the child's legs to flatten the lower spine.

Once a pelvic X-ray is taken, two measurements are usually calculated. These are the migration percentage (MP),<sup>(18)</sup> and the acetabular index (AI).<sup>(19)</sup> Figure 1 uses a sketch of a hip X-ray to construe how MP and AI are measured.

A study in 1989, that investigated the natural history and predictive factors of hip dislocation in children with CP, found that MP and AI were the best ways to attempt to predict hip dislocation.<sup>(14)</sup> It was discovered for MP there was a lot of overlap between the MPs of children that did not dislocate their hip compared to those that did. When looking at AI,

there was a pronounced decrease in the amount of overlap between the two groups that did and did not dislocate. From these findings, it was recommended that AI would be the best method of screening for hip subluxation in children. A key limitation of this study is that they didn't see children at regular intervals and they only saw them when they presented to their clinic with clinical signs with hip subluxation.

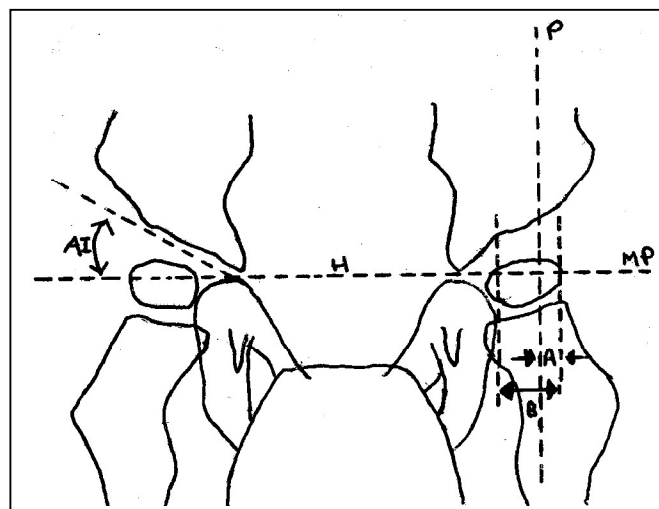


Figure 1 (H = Hilgenreiner's line. P = Perkin's line. MP = migration percentage. AI = acetabular index) MP represents the percentage of the ossified femoral head that is positioned laterally past Perkin's line.<sup>(20)</sup> It detects lateral subluxation. A high MP value means a high amount of lateral subluxation. The equation used to work out MP is  $MP = A/B \times 100$ .<sup>(20)</sup> AI indicates the angle at which the acetabular slopes and it is measured in degrees. AI can also help determine the amount of acetabular dysplasia there is. It is suggested that the higher the AI the more acetabular dysplasia has taken place. MP and AI are the two measurements that are known to change during hip subluxation of children with CP<sup>(19)</sup>

In a separate study conducted in 2007, with the aim to evaluate the use of different radiological measurement and threshold values for hip screening in children with CP, it was found that lateral displacement often occurred before acetabular dysplasia.<sup>(9)</sup> In this study, the authors conducted a hip monitoring programme and measured the AI and MP on all the radiographs taken of 272 children with CP who were all born between 1992 and 1998. Radiographs were taken at regular intervals for those subtypes of CP that were more at risk of hip dislocation, while other subtypes were screened, but not as regularly. The methodology of this study is far more robust as they screened their patients far more regularly.

Another large study, carried out in 2005, compared the effects of a prevention programme of hip dislocation in a study group only containing children with CP against a control group of children with CP who were not in the prevention programme.<sup>(21)</sup> Screening at regular intervals depending on the child's subtype was established in the study group as well as always measuring the AI and MP on every radiograph. From the study's results, they proved that MP is the only measurement that should be used in a prevention programme. This study showed that the majority of high MP values had normal AI values. All of their results pointed towards femoral head lateralisation occurring before acetabular dysplasia. This means MP is the more appropriate measurement of subluxation of the hip. A weakness of the study above is that they published the results before the children had stopped growing. This means that most of the children have not passed

the ages when they are most likely to dislocate. Apart from that weakness, this study was carried out with robust methodology.

Non-radiological techniques, such as the combined hip abduction angle (CHAA), have been suggested as a measurement for hip subluxation.<sup>(22)</sup> This method is described as follows. The investigators would measure the angle made between the axes of both hips whilst abducted using a goniometer. The child would first be made to lie in a supine position and passive stretching of the hamstrings and psoas muscles would take place for five minutes before the measurement to allow maximum hip abduction. With both hips and knees flexed as much as possible and the hips abducted as much as possible the measurement would be taken with the child remaining in the supine position.

Research into non-radiological methods of hip surveillance have not been so widely conducted, but a recent study carried out at an orthopaedic clinic investigated the use of CHAA for hip surveillance in children.<sup>(22)</sup> Every child with CP who attended the orthopaedic clinic with a GMFCS of IV or V had a pelvic X-ray taken to compare the MP measurement of the child with their CHAA measurement. Their results showed that there is a correlation between high MP values and low CHAA values. The major significance of this study was that the authors were offering an alternative to radiological imaging which carries a high level of exposure to harmful radiation.

There are, however, many potential problems with the method of CHAA. One of these problems was that CHAA may mask a fixed infrapelvic obliquity and a false high CHAA will be acquired. Another problem that has been identified is the difference in the strength of passive stretching between the examiners of CHAA. This can affect the CHAA value, hence inter-observer variability increases.

**The NICE guidelines for measuring hip lateralisation<sup>(23)</sup>**

Consider repeating the hip X-ray after six months in children and young people where the initial hip migration is greater than 30%, and then consider repeating the hip X-ray every six months after this if the hip migration is increasing by more than ten percentage points per year. [1.1.19]

*This describes how children should be monitored if they have a migration percentage of more than 30%. These hip surveillance guidelines were published by NICE in July 2012. They clearly state here that the measurement of choice for hip displacement is migration percentage (MP)*

The aim of this discussion was to decide whether the NICE guidelines<sup>(23)</sup> had used a measurement that was accurate at measuring the severity of the pathology of hip dislocation. Figure 2 shows that MP is categorically recommended by the NICE guidelines. I agree with this choice because it has been proven in severe large reliable studies that MP is the only measurement used that can accurately measure hip subluxation.<sup>(9,21)</sup> CHAA and other non-radiological methods need to be researched further before they can be considered for hip subluxation surveillance.

**HIP SURVEILLANCE AND THE NICE GUIDELINES**

The age to start hip surveillance in children with bilateral CP is a contentious subject as researchers have been debating this point as soon as it became apparent that hip dislocation could be prevented.

A study conducted in 1972, with the target of understanding the natural history of hip dislocation, discovered that hip subluxation can occur as early as 18 months old.<sup>(24)</sup> This significant finding has been the main reason why researchers believe screening of children with bilateral CP needs to start as early as 18 months.<sup>(21)</sup> The results from this study are debatable, due to the study being a retrospective case review of 272 children where the authors looked at pelvic X-rays taken without a standardised method of patient positioning.

In a more recent and methodical study, a tertiary referral centre was created which screened the groups of children which were most at risk of hip dislocation.<sup>(20)</sup> In three years, they reviewed children ranging from 1-15 years. They noticed that subluxation occurred even earlier than 18 months and they suggested a screening for hip subluxation should begin before 18 months so treatment can begin as soon as possible. They did not detect a high rate of subluxation before 18 months but believe that delayed detection of subluxation can be disastrous for the child and if detected too late the best treatments are not possible. Detection this early can be impractical because a proportion of patients will not be diagnosed by this age, therefore, the screening test will miss some of those at risk from hip dislocation.

A major study published in 1997 looked into ascertaining the best age to first screen the hips of children with bilateral CP who are most at risk of hip dislocation so that they can predict future subluxation.<sup>(17)</sup> The authors collected data between 1989 and 1992 of all the children born with bilateral CP in a specified area: 262 children had standardised pelvic X-rays taken at 18, 24, 30, 48 and 60 months. The results indicated that the 30-month MP measurement correlated better with the four-year hip state than at any other time. Therefore, 30 months is the best age to be referred to an orthopaedic surgeon for closer monitoring programme.

A major strength of this study is that its method was extremely robust and the results are considered to be very reliable. In their results, it was found that five hips had an extremely high MP and again it was shown that subluxation must have occurred at an earlier stage than 18 months. This provides more evidence to the experts who believe screening should be conducted at 18 months or before. However, it was shown that in this study a scan at 30 months would still allow enough time for effective treatment before hip dysplasia is well developed.

**Current NICE guidelines that are related to this debate<sup>(23)</sup>**

Offer a hip X-ray to assess for hip displacement: if there are clinical concerns about possible hip displacement at 24 months in children with bilateral cerebral palsy. [1.1.17]

*Twenty-four months is the age at which children with bilateral CP are recommended to have a hip X-ray (NICE, July 2012)*

NICE currently recommends that a hip surveillance programme starts at 24 months for children diagnosed with bilateral CP.<sup>(23)</sup> This guideline is contradictory to the evidence I have put forward. The major study published in 1997 found that there was a poor correlation with MP measurements taken at 24 months and the hip state at four years.<sup>(17)</sup> It seems that the guideline has been chosen at 24 months just because it is between 18 months and 30 months. I believe that 30 months is the best age to start hip screening. This is because it correlates well with the future risk of hip subluxation, and the study from 1997 showed that even if the hip has started to subluxate at an earlier age treatment is still effective.<sup>(17)</sup>

## SUMMARY

Hip dislocation in children with CP is disastrous for that child and it is imperative that this event be prevented. In the UK, hip surveillance programmes vary between the different Clinical Commissioning Groups (CCGs) but usually follow the guidelines set out by NICE. These guidelines state the MP measured at 24 months is the best way to measure and start screening. From my review, I believe I have shown a different conclusion. From the evidence I have provided, I think that measuring MP at 30 months of age is the most appropriate method and age to detect any subluxation and future dislocations in children with bilateral CP.

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