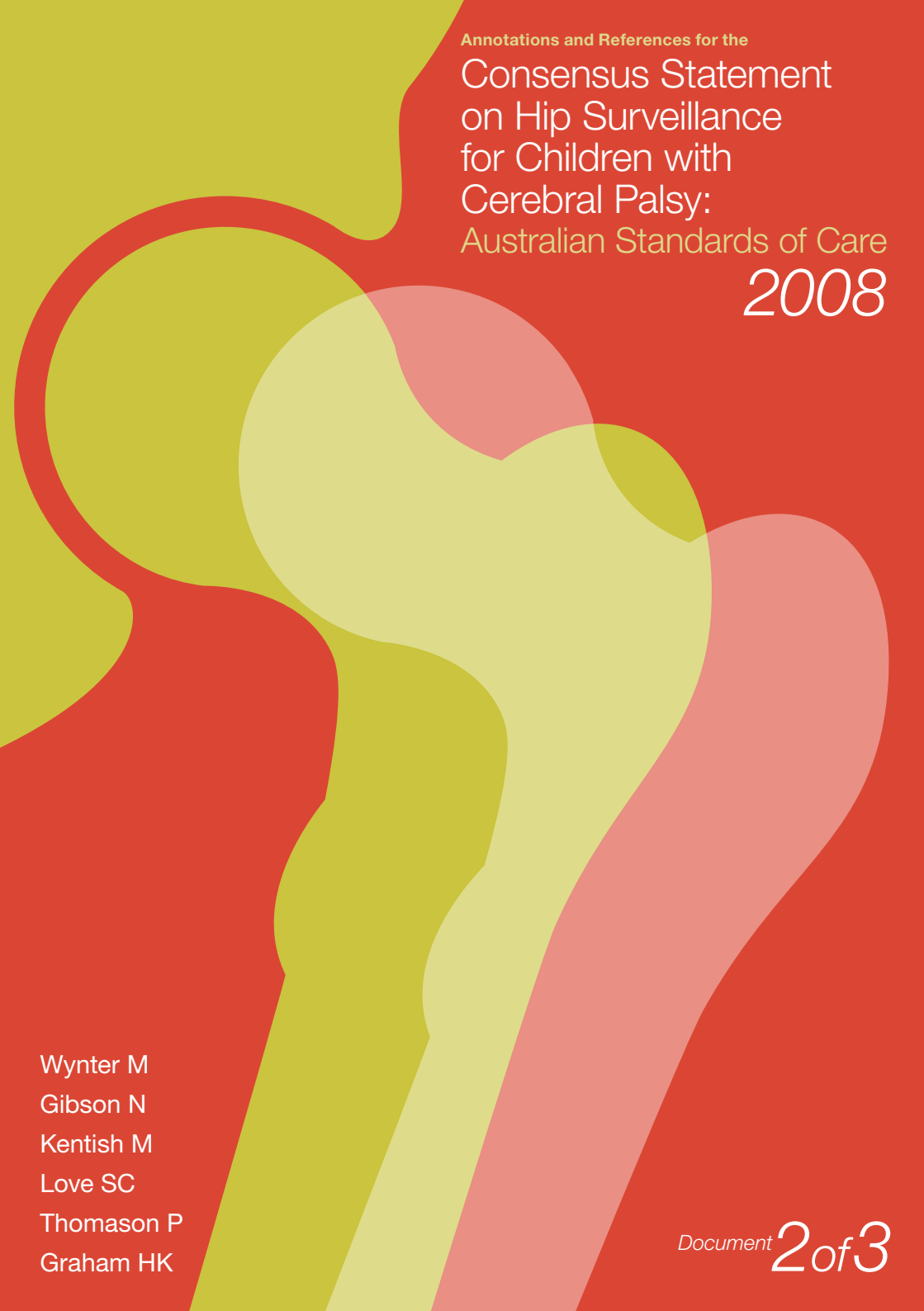
Annotations and References for the

Consensus Statement on Hip Surveillance for Children with Cerebral Palsy: Australian Standards of Care 2008



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Consensus Statement on Hip Surveillance for Children with Cerebral Palsy: Australian Standards of Care *2008*

This document is endorsed by:

Australasian Academy of Cerebral Palsy and Developmental Medicine

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The **Royal Children's Hospital** Melbourne

Queensland Government
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Government of **Western Australia**
Department of **Health**
Child and Adolescent Health Service
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1. Hip surveillance

Hip surveillance is the process of monitoring and identifying the critical early indicators of progressive hip displacement[3]. These early indicators include the Gross Motor Function Classification System (GMFCS)[4], age[6], gait classification (Winters Gage and Hicks, Group IV)[12] and migration percentage (MP)[9]. The information gathered from the clinical assessment[8] and radiological review[11] are vital components of hip surveillance and are required to capture often silent displacement of the hip while minimising radiation exposure. Hip surveillance cannot be based on clinical assessment[8] alone.

Hip surveillance will assist identification of prognosis; inform planning for ongoing hip management; support education and assist clear communication. Protocols or recommendations for hip management are beyond the scope of this document.

Hip surveillance is an ongoing process that continues for every child until discharge[14] or skeletal maturity[17]. Hip surveillance should always be resumed following the perioperative period for any child who has undergone surgery for hip management, or following an unplanned break in surveillance for any other medical reason. All children with CP[2] or like conditions should be referred for hip surveillance even if classification and determination of GMFCS[4] are not yet confirmed[13].

A body of evidence supports the implementation of hip surveillance as an effective means towards prevention of hip dislocation[3]. A systematic review of the evidence for children with CP[2] (Gordon and Simkiss 2006) identified 6 studies where results showed support for hip surveillance programs. All studies used radiological measures[7] to monitor hip displacement[3], with MP[9] (Reimers 1980) most frequently used.

Dobson et al. (2002) and Hagglund et al. (2005) have demonstrated that hip surveillance programs are an effective step in the prevention of hip dislocation[3]. The monitoring of MPs[9] enabled identification of children for surgery at a younger age,[6] thus reducing the need for later salvage surgery.

The recommendation by Scrutton and Baird (2001) that all children with bilateral CP[2] should have a pelvic radiograph[11] prior to 30 months of age has been cited as a guideline for hip surveillance. Others have reported early radiographic changes as young as 12 months for some children (Vidal et al. 1985), hence the recommendations for an early starting age in this document.

There have been no previously published recommendations outlining the commencement and frequency of hip surveillance, where surveillance is based on risk relative to GMFCS[4] level. Frequencies of 6 to 12 monthly are considered in some studies (Dobson et al. 2002) whilst others had yearly radiographs[11] from diagnosis until 8 years of age, with follow-up beyond this on an individualised basis[29] (Hagglund et al. 2005). This consensus statement takes into account the more recent data (Soo et al. 2006) on risk of hip displacement[3] relative to GMFCS[4] levels and the need to minimise radiation exposure, when recommending frequency for repeated surveillance.

2. Cerebral Palsy

The term Cerebral Palsy (CP) refers to CP and like conditions, where clinical signs or descriptions are most relevant, not aetiology.

In line with the decision made by the Surveillance of Cerebral Palsy in Europe (SCPE 2000) and methodology adopted in 2003 by the Australian Cerebral Palsy Register Group (Blair et al. 2007), for the purposes of this document the definition of CP that is acceptable includes the following 5 key elements (Mutch et al. 1992):

- CP is a group of disorders that is, it is an umbrella term
- It involves a disorder of movement and/or posture and of motor function
- It is due to a non-progressive interference/lesion/abnormality
- This interference/lesion/abnormality is in the developing/immature brain
- It is permanent but not unchanging

In conditions other than CP, where there is no evidence for the natural history of hip displacement, the risk seems likely to also relate to functional ability[5]. In all probability, the more clinically similar a child's condition is to CP, the more likely that these guidelines will be effective in identifying at risk hips.

For the purposes of these guidelines, like conditions refers to those conditions where motor dysfunction results from genetic and metabolic aetiologies, including clearly recognised syndromes or progressive brain disorders (Badawi et al. 1998), or from brain injury acquired in childhood within the first 2 to 3 years of life.

Until there is natural history data for children with acquired brain injury, early and frequent surveillance is recommended, as clinical experience indicates a high prevalence of hip displacement in this group.

Motor disorders of spinal, peripheral nerve, muscular or mechanical origin are not considered as like conditions. Disorders of impaired cognition but no motor signs are not considered as like conditions.

3. Progressive hip displacement, dislocation and sequelae

Progressive hip displacement refers to the gradual displacement of the femoral head laterally from under the acetabulum. This displacement is expressed as a migration percentage (MP)[9].

Hip subluxation defines the state of the hip joint and can be used interchangeably with hip displacement where MP[9] is between 10% and 99%.

Hip dislocation is defined when the femoral head is completely displaced laterally from under the acetabulum (MP[9] = 100%).

The sequelae of progressive hip displacement are variable (Cornell 1995). Progressive displacement can result in asymmetric pressure that may deform the femoral head and/or acetabulum (also termed acetabular dysplasia). Hip dysplasia may lead to degeneration of articular cartilage and pain[25]. Problems with limited range of movement[21] and pain[25] can interfere with function, ability to be positioned and hygiene and personal care. In a large subset of children the progressive displacement can develop into dislocation of one or both hips (Cooke et al. 1989).

4. Gross Motor Function Classification System

The Gross Motor Function Classification System (GMFCS) classifies the gross motor function[5] of children and youth with CP[2] on the basis of their self-initiated movement with particular emphasis on sitting, walking, and wheeled mobility (Palisano et al. 1997, Palisano et al. 2008).

The GMFCS has 5 levels for describing differences in severity of motor abilities[5]. Distinctions between levels are based on functional limitations, the need for hand-held mobility devices or wheeled mobility, and to a much lesser extent, quality of movement. Since classification of motor function[5] is dependent on age[6], separate descriptions are provided for several age[6] bands within each level. The age[6] ranges described are as follows: before 2nd birthday, from age 2nd to 4th birthday, from age 4th to 6th birthday and from 6th to 12th birthday, and from 12th to 18th birthday. There is a tendency for children classified prior to 6 years of age to be reclassified after 6 years of age (Palisano et al. 2006) hence the need to confirm GMFCS level at each occasion of clinical presentation.

The distinctions between levels I and II are not as pronounced as the distinctions between the other levels, particularly for infants less than 2 years of age[6]. Emphasis is on what they do (usual performance in home, school, and community settings), rather than what they are known to be able to do at their best (capability). It is therefore important to classify current performance in gross motor function[5] and not to include judgments about the quality of movement or prognosis for improvement. Generally it takes only a few minutes to assign a GMFCS classification.

Both the original GMFCS (Palisano et al. 1997) and the new GMFCS: Expanded and Revised (GMFCS-E & R) (Palisano et al. 2008) can be downloaded free of charge from the website www.canchild.ca

5. Gross motor functional ability

This refers to the gross motor activities that the child is able to accomplish in his/her own environment (performance) rather than what he/she maybe able to achieve in a testing situation (capability), including the achievement of developmental milestones.

6. Corrected age

Assessment for hip surveillance[1] takes into consideration corrected age for prematurity up to 2 years of age. Pre term or premature is defined as a gestational age less than 36 weeks. To calculate corrected age subtract the expected date of birth (i.e. not actual date of birth) from the date of evaluation.

7. Radiological measures

These are reproducible measures taken manually or electronically from a standard radiograph[11]. For hip surveillance[1] the standard radiograph[11] required is a frontal antero-posterior plain film radiograph of the pelvis (AP pelvis) (Reimers1980). Radiological measures maybe less accurate in the very young and will not be accurate below 12 months of age[6].

8. Clinical assessment

The essential elements of clinical assessment undertaken for hip surveillance[1] are only a part of the overall assessment required by a child with CP[2]. For the purpose of hip surveillance[1], clinical assessment should include both subjective and objective aspects to identify and document concerns, care and comfort, pain[25], any change in gross motor function[5] including gait[20] and assessment of the child's spine[18], pelvis[19] and lower limb musculoskeletal system[21]. The assessor should be able to classify the child's GMFCS[4] level and gait pattern if WGH IV[12].

9. Migration percentage

This is a radiographic measure[7] of the amount of ossified femoral head which is not covered by the ossified acetabular roof (Reimers 1980). It is the percentage of the femoral head which is lateral to Perkin's line on a frontal view radiograph[11] **(Figure 3)**.

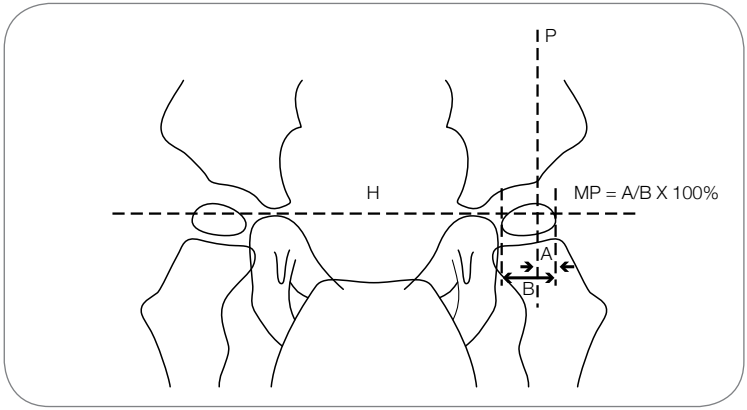

Figure 3: Migration percentage

MP is measured by drawing a horizontal line through the most superior medial point of each triradiate cartilage (Hilgenreiner's or H-line) and a vertical line (Perkin's or P-line) drawn perpendicular to it at the lateral margin of the acetabulum **(Figure 3)**.

MP = A/B X 100%.

10. Stability of migration percentage

In children with CP[2] the majority of hips are normal at birth (Bleck 1987, Laplaza et al. 1993, Vidal et al. 1985). In the absence of treatment, the MP[9] increases progressively from an early age[6] at an average rate of about 5.5% per year. A change of greater than 8% in repeated measurement by one experienced measurer is required to be 95% confident of true change (Parrott et al. 2002, Faraj et al. 2004). For the purpose of this document, stability of MP[9] is progression of not more than 10% in a 12 month period (Gordon and Simkiss 2006) over a period of 2 to 3 years.

An unstable MP[9] is when the progression is greater than or equal to 10% over a 12 month period.

11. Antero-posterior pelvic radiograph

A frontal plain film radiograph (AP pelvis) within certain positioning limits is required to enable MP[9] to be accurately measured. The MP[9] is to a large extent dependent on the abduction or adduction of the leg, so the leg should be in neutral abduction/adduction **(Figure 4a)**. Acceptable range of adduction/abduction is ±6°. The effect of rotation of the leg is small (when in the range of acceptable abduction/adduction). The MP can be measured only if the Hilgenreiner's line can be plotted accurately: i.e. the triradiate cartilages need to be clearly visible and the pelvis not in forward or backward pelvic tilt. This tilt needs to be corrected in children who have a fixed flexion deformity of the hip(s) [21] or a significant lumbarlordosis **(Figure 4b)**.

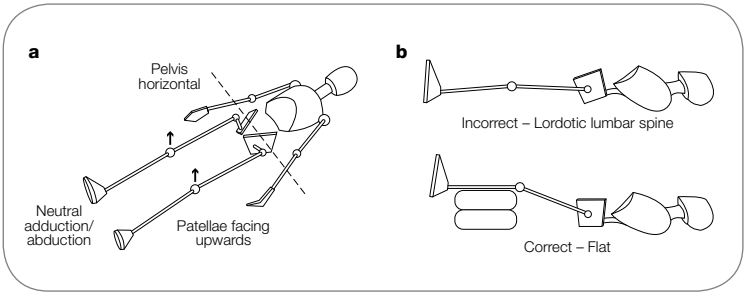

Figure 4: Positioning for antero-posterior pelvic radiograph

12. Winters, Gage and Hicks classification

Winters, Gage and Hicks (WGH) classification of hemiplegic gait[20] describes four types of gait[20] patterns based on the sagittal plane kinematics of the ankle, knee, hip and pelvis (Winters et al. 1987). The characteristic of each group is as follows:

Group I – foot drop in the swing phase of gait[20], normal dorsiflexion range in stance phase of gait

Group II – excessive plantarflexion of the ankle in both stance and swing phase of gait[20]

Group III – Group II deviations as above plus limited flexion /extension range of motion at the knee during stance and swing phases of gait[20]

Group IV – Group III deviations as above plus limited flexion/extension range of motion at the hip during stance and swing phases of gait[20]

This is represented diagrammatically in **Figure 2, Document 1**.

There are limitations in using this classification as it is based only on sagittal plane kinematics (Dobson et al. 2007). Many children with hemiplegia will present with coronal and transverse plane gait[20] deviations which may predispose them to a higher risk of hip displacement[3] than those with only sagittal plane deviations. Hence children with coronal or transverse plane abnormalities particularly at the hip level should also be considered in this group for the purposes of hip surveillance[1]. While this classification is based on three dimensional gait analysis kinematic data, visual observation of gait and classification is sufficient for the purpose of hip surveillance. Children classified as

WGH IV are those at risk of progressive hip displacement[3]. They develop displacement[3] later than children with bilateral CP[2] and progress slowly until puberty[16]. Presentation at puberty[16] may be characterised by rapid increasing leg length discrepancy[19], apparent shortening and pelvic obliquity.[19]

13. Confirmed

For the purpose of this document confirmed is defined as the GMFCS[4] level which best fits on today's assessment. GMFCS[4] levels may not always be distinct or easily apparent, particularly for the younger child and between the higher levels (Palisano et al. 1997). It is important to reassess for the correct GMFCS[4] level on each occasion of hip surveillance[1].

14. Discharge

Discharge is the cessation or release from continuing hip surveillance[1]. Children will most often be involved with other management programs including spasticity[23] management or orthopaedic gait[20] corrective surgery according to best practise and evidence based medicine. Gait corrective surgery may simultaneously address displacement[3] of the femoral head whilst correcting other bony alignment.

15. Normal/abnormal migration percentage

A normal migration percentage[9] is considered to be zero or even negative as displacement should not occur in a normal hip (Perkins 1928). Reimers (1980) found that among children with normal motor development, the 90th centile for hip migration at 4 years of age[6] was 10%. For the purpose of this document, normal MP[3] is less than 10% after the corrected age[6] of 4 years.

MP[9] above 30% are high and should be considered at risk/abnormal.

16. Puberty

Puberty can be recognised by a combination of growth acceleration, development of secondary sexual characteristics, chronological age[6] and bone age. Bone age can be assessed with a range of radiological investigations of which X-ray of the wrist or elbow are the most widely used. In normally developing children, girls will experience the onset of puberty at 11 years (bone age) and boys at 13 years (bone age) but there is wide variation in both normally developing children and even more so in children with CP[2]. In normally developing children, about 50% have a bone age which is significantly different from their chronological age and in CP[2] the percentage is even higher (Dimeglio 2006). Delayed bone age is particularly common in severe CP[2] (GMFCS[4] IV and V) and it is probable that the pattern of skeletal maturation varies by GMFCS[4] level. Although hip displacement[3] may occur in children with CP[2] from early childhood, the pubertal growth spurt is a period of particular risk for both progression of existing hip displacement[3], the development of hip displacement[3] in previously stable[10] hips, as well as the development of pelvic obliquity[19] and scoliosis[18].

17. Skeletal maturity

A number of definitions of skeletal maturity have been employed using radiographic parameters. The earliest of these is closure of the triradiate cartilage (Dimeglio 2006) followed by closure of the growth plates around the elbow and then progression of the Risser sign (Risser 1958) from I to V. Although the Risser sign is probably the most commonly used mark of skeletal maturation, especially in the management of scoliosis[18], it is somewhat easier to measure skeletal maturity from the closure of the triradiate as the entire iliac crest may not be fully visible in all pelvic radiographs[11]. For the purposes of this document the closure of the triradiate cartilage will be used as the prime indicator of skeletal maturity for hip surveillance[1] (Acheson 1957).

18. Scoliosis

In CP[2] most spinal deformities involve neuromuscular scoliosis although sagittal plane deformities such as kyphosis (dorsal spine) and lordosis (lumbar spine) are also common. Spinal deformities in children with CP[2] are related to the severity of involvement and are most common in

GMFCS[4] IV and V. Initially the problems are postural but tend to progress rapidly and become fixed[24] during puberty[17]. During the pubertal[17] growth spurt, scoliosis may increase at a rate of 2–4 degrees per month with the curve reaching magnitudes of 60–90 degrees very quickly and then becoming increasingly stiff. Even when the individual reaches skeletal maturity[16], curve progression may continue at between 1 and 4 degrees per year. In children with neuromuscular scoliosis, the pelvis is often part of the curve and the incidence of pelvic obliquity[19] is very high (Miller 2005).

19. Pelvic obliquity, real and apparent leg length discrepancy

Pelvic obliquity may occur in younger children with CP[2] as the result of neuromuscular imbalances around the trunk, pelvis and hips. Pelvic obliquity may be secondary to influences above the pelvis (scoliosis[18]) or below the pelvis (leg length inequality, hip displacement/dislocation[3] or asymmetric contractures of the hip adductors or hip flexors[21]), or from a combination of supra-pelvic and infra-pelvic influences.

It is important to determine the contributions of both real and apparent shortening in the evaluation of leg length discrepancy as well as the contribution of supra-pelvic and infra-pelvic factors. This is done by careful clinical examination of real and apparent leg length with interpretation of this information with radiographs of the pelvis and/or spine. Although unilateral hip subluxation[3] and dislocation[3] may result in a real leg length discrepancy, there is frequently a combination of real and apparent discrepancy.

20. Gait

Gait describes the particular manner or way of moving on foot. It is the description of locomotion style. Alterations in gait that may necessitate increased frequency of hip surveillance[1] may include increasing asymmetry of the pelvis with retraction or pelvic obliquity[19], increased hip adduction or internal rotation, changes or increased asymmetry of step length. This is by no means inclusive of all possible gait deviations.

21. Musculoskeletal measures relating to the hip

Musculoskeletal measures relating to the hip should include assessment of the spine[18], pelvis[19], leg discrepancy[19] and physical examination of the lower limbs including passive and dynamic range of movement (Boyd and Graham 1999), muscle strength, and measures of spasticity[23].

Assessment of musculoskeletal measures around the hip should include:

- Passive range of movement
  - Hip abduction with hips at 90 degrees of flexion
  - Hip abduction with hips at 0 degrees of flexion
  - Thomas test
  - Hip flexion
  - Hip extension (Staheli)
  - Hip internal rotation
  - Hip external rotation
  - Femoral neck angle
  - Popliteal angle
- Dynamic contracture as measured by Modified Tardieu Scale[23] (Boyd and Graham 1999)
  - Hip adductors
  - Hamstrings
- Modified Ashworth Score[23] (Bohannon and Smith 1987)
  - Hip adductors
  - Hamstrings
  - Hip flexors

22. Muscle tone

Muscle tone refers to the normal resting tension or the change in the resistance of the muscle to passive movement or muscle lengthening. It excludes resistance as a result of joint, ligament, or skeletal properties such as those that may occur with fixed deformities, including contracture (Sanger et al. 2003). An abnormal increase in resistance to passive movement is termed hypertonia. Hypertonia may be the result of a number of factors, one of which is spasticity[23].

23. Spasticity

Spasticity is a disorder of the sensorimotor system characterised by a velocity-dependant increase in muscle tone with exaggerated tendon jerks, resulting from hyperexcitability of the stretch reflex. It is one component of the upper motor neuron syndrome, along with released flexor reflexes, weakness, and loss of dexterity (Mayer 2002). When spasticity is present, the resistance to externally imposed movement rises rapidly above a threshold speed or joint angle (Delgado and Albright 2003, Sanger et al. 2003). Spasticity does not worsen with age but its manifestation of movement such as the paucity of variety of movement may result in worsening secondary effects e.g. contractures (Delgado and Albright 2003).

The Modified Tardieu Score (MTS) is a rating of spasticity which measures the intensity of muscle reaction at maximal velocity movement through range (Boyd and Graham 1999). The quality of the muscle response is noted if there is a “catch” in motion and the angle at which the catch occurs is measured. The “catch” is sometimes referred to as R1, the first resistance to rapid passive movement. It is described as the clinical estimate of the threshold angle of spasticity (Boyd and Graham 1999). A lowering of the “threshold” for R1 (i.e. an earlier catch), may be an indication that there is increasing spasticity. Spasticity can be graded using the Modified Ashworth Scale (MAS) (Bohannon and Smith 1987).

24. Fixed posture and asymmetry

Fixed posture describes structural changes to the posture/mobility of the trunk and/or limbs which cannot be voluntarily, passively or forcibly corrected. This can be assessed clinically[8] and/or radiologically and is differentiated from non-structural postural changes which may be fully corrected.

Asymmetry is dissimilarity in corresponding parts on opposite sides of the body which are normally alike. Fixed asymmetry describes structural changes to the trunk[18], pelvis[19] and/or limbs characterised by the lack or absence of symmetry which cannot be voluntarily, passively or forcibly corrected. This can be assessed clinically and/or radiologically and is differentiated from non-structural postural changes which may be fully corrected.

Newly developed is a clinical sign or measure of recent onset which was not apparent at the previous assessment[8], or is subjectively described by the patient/caregiver as having recently appeared.

25. Pain

Pain in the hip region for children with CP[2] is variably reported in the literature and may or may not be associated with hip displacement[3] or dislocation[3] (of one or both hips). In some cases pain may be clinically expressed in the knee or leg but be referred from the hip. The relationship between hip pain and displacement[3] or dislocation[3] remains elusive in both children and adults. Chronic musculoskeletal pain is a complaint in up to 67% of adults with CP[2], most commonly in the low back, hip, and leg (Engel et al. 2003). Hodgkinson et al. (2001) found that the prevalence of pain was 47.2% of 234 non ambulatory adolescents with CP[2].

Pain may be observed at rest, with certain positions, or with such movements as passive abduction[21] (Hodgkinson et al. 2001). Identifying the source of pain in the region of the hip remains a challenge. In children with limited communication, the clinician must rely on the perception of the parents or caregivers to help identify the source. Pain may originate in the skin or subcutaneous tissues, the musculature surrounding the hip, the osteoarticular structures, or may be referred from another location (Spiegel and Flynn 2006).

26. Other orthopaedic conditions

Other orthopaedic conditions include, but are not limited to developmental dysplasia of the hip, muscle contracture that is not able to be managed conservatively, an inflammatory reaction, such as transient or toxic synovitis, a slipped capital femoral epiphysis, Perthes Disease, excessive femoral anteversion, juvenile idiopathic arthritis, septic arthritis or bursitis, osteomyelitis, other unusual bone or joint anomalies and in rare cases, bone tumours.

27. Individualised management plan

Individualised management plan is the adaptation of a standard management plan in response to individual clinical presentation and need. This management plan may include ongoing hip surveillance[1], altered frequency of surveillance[1] from 'standards of care', and/or intervention including surgical intervention.

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Australasian Academy of Cerebral Palsy and Developmental Medicine

These hip surveillance standards of care for children with cerebral palsy were endorsed by the Australasian Academy of Cerebral Palsy and Developmental Medicine (AusACPDM) on 28th October 2008. Endorsement by AusACPDM is granted for a period not exceeding five years, at which date the approval expires. The AusACPDM expects that these standards of care will be reviewed no less than once every five years.

These Standards of Care are due for review by 28/10/2011

This document is one of three:

1. Consensus Statement on Hip Surveillance for Children with Cerebral Palsy: Australian Standards of Care

2. Annotations and References for the Consensus Statement on Hip Surveillance for Children with Cerebral Palsy: Australian Standards of Care

3. Explanatory Statement to Accompany the Consensus Statement on Hip Surveillance for Children with Cerebral Palsy: Australian Standards of Care

Disclaimer

This document is endorsed as a general outline of appropriate clinical practice, based on a review of the best evidence available at the time of publication, and is to be followed subject to the clinician's judgment and the patient's preference in each individual case. The AusACPDM takes no responsibility for evidence or information published subsequent to this review.

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