The hip in cerebral palsy

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Abstract

Cerebral Palsy (CP) is a common cause of progressive musculo—skeletal deformity and loss of function in children; with an incidence of around 3/1000 live births. The majority of even the most severely involved children survive to adulthood.

Children with CP have complex needs and need to be regularly assessed by a multidisciplinary team in order to target appropriate interventions. These do not alter the underlying chronic condition but can provide improvement in function and comfort, and reduce the burden of care if considered within the context of the overall needs of the child.

Hip displacement is seen in up to 60—80% of severely affected children. Hip surgery is therefore unsurprisingly the second most common orthopaedic intervention in children with CP. Surveillance of at risk populations is now well established.

Setting functional goals and assessing outcomes of any intervention is necessary, with medical, anaesthetic, nursing and therapy components all contributing significantly to the positive outcome if surgery is deemed necessary. Any surgical intervention in a child with CP should always be a process rather than a discrete event.

Keywords cerebral palsy; hip development; multidisciplinary team interventions; paediatric orthopaedic surgery; screening/surveillance programmes

Introduction

Cerebral palsy, though a descriptive not a diagnostic term, is the commonest form of movement disorder encountered in childhood. At birth, the rates of congenital dislocation of the hip in children with CP are very similar to the wider population. With growth, the altered biomechanical forces working across the hip joint can lead to progressive subluxation, dislocation and deformity.

With the introduction of comprehensive screening programmes, the risk of hip dislocation can be dramatically altered. How and why the hip migrates out of joint in children with cerebral palsy continues to be the focus of much debate; as with most things a basic review of normal and abnormal development is a starting point.

Normal hip development

Embryogenesis of hip

We are all built up of three basic embryological cell types. At our earliest stage, the spherical blastocyst develops into an internal endoderm, a middle mesoderm and outer ectoderm. As this flattens and rolls up an inner neuro-ectoderm differentiates into the central nervous system. The limb buds start to develop from the mesodermal layer as a lateral projection off the embryo at around 4 weeks of gestational age. By 6 weeks there is a primitive cartilaginous femur, and soon afterwards an early hip joint forms by cellular degeneration in the area between the two opposing articular surfaces. By 5 months the ball-and-socket hip joint, capsule and surrounding muscles are all fully developed.

Postnatal hip development

At birth the femoral head is completely made of cartilage. Proximal femoral head growth occurs disparately in four separate regions resulting in elongation and angulation. Ossification starts at around 2—6 months. At this stage the acetabulum faces anteriorly (is anteverted) by approximately 40° and the femoral neck by about 35°.

The acetabulum does not change its orientation but by 10 years the femoral neck is positioned at the adult level of anteversion — around 15°. Further femoral growth and remodelling beyond the neonatal stage is mediated by the trochanteric and femoral head physeal plates. Normal biomechanical forces working across the joint stimulate linear and circumferential growth of the femur, ensuring the ball of the femoral head stays snugly in the socket of the acetabulum.

The overlying muscles also contribute greatly to the hip stability together with the capsule, ligaments and fibrous acetabular brim. Subsequent normal joint development is reliant on a stable and concentric relationship between the femoral head and acetabulum.

Abnormal hip development in cerebral palsy

Cerebral palsy

As stated, Cerebral Palsy (CP) is a descriptive term, not a diagnostic one. It encapsulates a 'group of permanent disorders of the development of movement and posture, causing activity limitation that is attributed to non-progressive disturbances that occurred in the developing foetal or infant brain'. There is massive variance in clinical presentation and the range of disabilities and co-morbidities observed in individuals with 'cerebral palsy'. The impact of the lesion generally depends on its site and size within the brain.

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If there is predominant cortical damage, this causes an 'upper motor neuron lesion' with a pattern of positive and negative clinical features observed.

Spasticity – a *velocity dependent increase in tonic stretch reflexes causing hypertonia* – is associated with the 'positive' side. Simplistically, the descending cortico-spinal tracts normally stimulate release of the inhibitory neurotransmitter GABA at the spinal level. Lesions in this pathway therefore lead to dysinhibition of the spinal reflex arc and muscle over-activation, rigidity and hypertonia. Whilst we focus on damage in these tracts, there is considerable interplay with other pathways of descending motor control. In the long term this muscle over-activation leads to altered muscle physiology, reduced muscle growth, an increase in non-contractile connective tissue and development of joint contractures.

The 'negative' symptoms of the upper motor syndrome – reduced motor activity leading to weakness and poor selective motor control can be even more disabling for the individual and are generally more difficult for us to treat.

If, however, the damage is primarily to the deeper basal ganglia then patterns of sustained disturbed muscle contraction causing abnormal postures that are frequently associated with involuntary movements are observed. These patterns are exacerbated by sensory 'overload' such as anxiety, noise or discomfort ~ dystonic cerebral palsy. These strange, non-sustained movements can of course be further subdivided e.g. chorea and athetosis, but the over-riding term of **dystonia** is simple and useful.

Further description of the motor disorder outlines the laterality of involvement: unilateral or bilateral (though there is usually a degree of asymmetry of movement) and severity of the functional disability. With regards to movement, the **Gross Motor Functional Classification System (GMFCS)** delineates individuals with CP into five groups dependent on their level of mobility: level one being completely independent with minimal restriction in activity and five totally dependent for their needs (Figure 1). Generally the higher the group, the greater the changes of muscle tone, weakness and increased risk of developing muscle contractures.

Problems of the hip and CP

Any alteration in the normal forces applied across the hip joint, caused by abnormal neurological control, primarily leads to deformity and in particular femoral dysplasia. These biomechanical changes also reduce the secondary stimulus for acetabular development, leading to a shallow cup with less inherent bony stability.

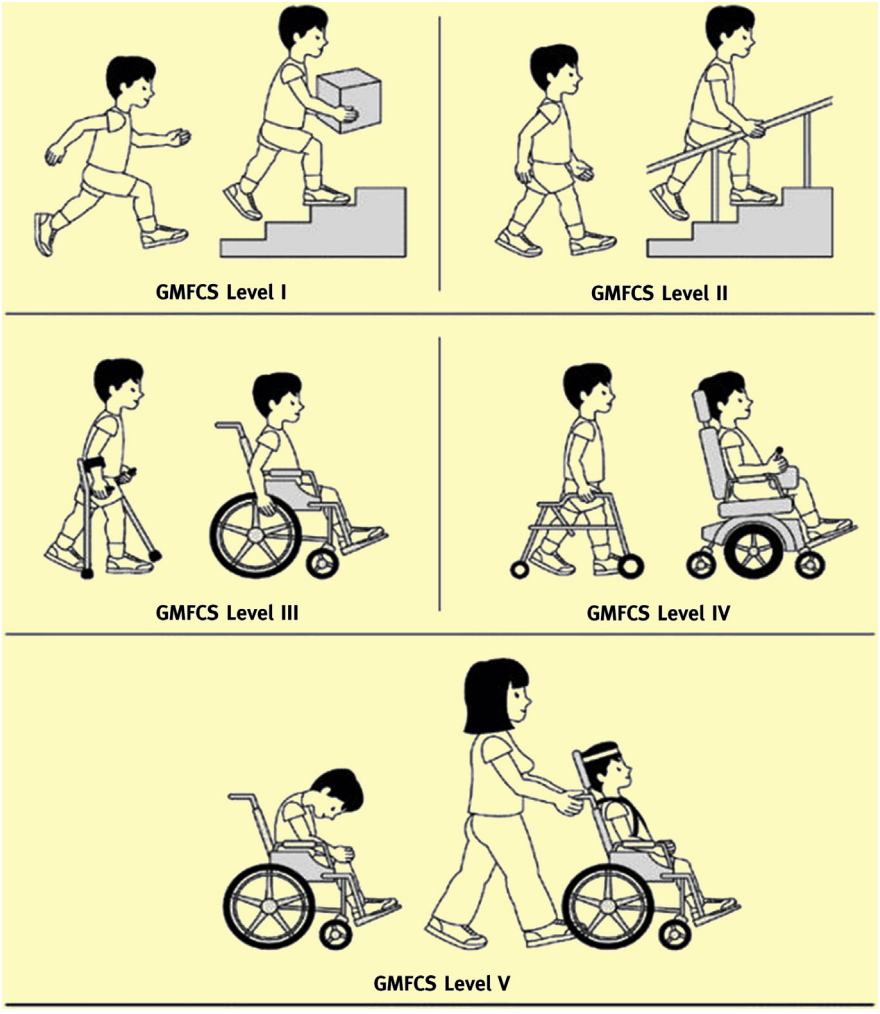

Figure 1 Gross motor functional classification system.