Shropshire Community Health NHS
NHS Trust

Policies, Procedures, Guidelines and Protocols

Document Details	
Title	**Fracture Risk Management in Children and Young People with Cerebral Palsy**
Trust Ref No	2155
Local Ref (optional)	
Main points the document covers	To prevent fractures in Children and Young People with Cerebral Palsy
Who is the document aimed at?	Community Paediatricians Community Physiotherapists
Author	Dr Diane Short and Dr Sam Danaher
Approval process	
Approved by (Committee/Director)	Clinical Policy Group
Approval Date	09/09/2024
Initial Equality Impact Screening	Y
Full Equality Impact Assessment	N
Lead Director	Director of Nursing and Operations
Category	Clinical
Sub Category	
Review date	09/09/2027
Distribution	
Who the policy will be distributed to	Community paediatricians Community Physiotherapists
Method	Email
Document Links	
Required by CQC	No
Required by NHSLA	No
Keywords	Cerebral Palsy, Fractures

Amendments History		
No	Date	Amendment
1	12.09.16	Guideline approved
2	16.09.20	Guideline reviewed and retitled
3	V3 Sep24	Guideline Reviewed
4		
5		

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1 Introduction

1.1 Fractures in Children and Young people with Cerebral Palsy

Children with moderate to severe cerebral palsy are at increased risk of sustaining fracture following minimal trauma. Such fractures predominantly occur in lower limb bones and are associated with low bone mineral density.

Risk factors for fracture in this group include non-ambulatory status, previous fractures, post surgery, malnutrition, low vitamin D levels and antiepileptic drugs.

2 Purpose

The aims of this guideline are to describe the prevalence and pathogenesis of fractures in non- ambulant children with cerebral palsy. It provides guidance on interventions and treatments that improve low bone mineral density and which may help to reduce the fracture risk in this population.

3 Definitions

3.1 Cerebral Palsy (CP)

A term used to describe a group of non-progressive disorders of movement and posture, resulting from an insult to the developing brain.

3.2 The Gross Motor Function Classification System (GMFCS)

A widely used five level clinical standardized system to classify the gross motor function of children with CP, with emphasis on function in sitting and walking.

3.3 Children and Young People (C&YP)

This guideline is for children and young people up to age 18 years old.

4 Duties

4.1 Chief Executive

Should ensure that all clinical staff working with children and young people have access to this guideline. Should ensure that appropriate training and updates are provided to all relevant staff groups. Should ensure that staff have access to appropriate equipment that complies with safety and maintenance requirements.

4.2 Managers

Managers should ensure that staff are aware of and have access to policy guidelines. Staff training needs should be highlighted and addressed. Appropriate education, supervision and mechanisms are in place to ensure good practice.

4.3 All clinicians working with children and young people

To be aware of the guideline and follow appropriately.

5.0 Cerebral Palsy (CP)

Cerebral palsy is a term used to describe a group of non-progressive disorders of movement and posture, resulting from an insult to the developing brain.

It is one of the commonest chronic disabling conditions of childhood, with prevalence of 2.0–3.5 per 1000 live births. The most common motor abnormality is spasticity, which may be categorized into diplegia, hemiplegia, and quadriplegia. Other forms of CP include dystonia, choreo-athetosis, ataxic or a mixture of these disorders.

The Gross Motor Function Classification System (GMFCS) is a widely used five level clinical standardized system to classify the gross motor function of children with CP, with emphasis on function in sitting and walking.

Children with CP often have other disabilities, which may affect their quality of life and life expectancy. These include intellectual impairment, behavioural problems, hearing and visual problems, feeding difficulties, poor growth, recurrent respiratory infections, and epilepsy. Secondary musculoskeletal problems include joint contractures, kyphoscoliosis and hip subluxation.

6.0 Fractures in C&YP with Cerebral Palsy

Children and young people with CP are prone to low trauma fractures, which occur during normal activities, such as dressing and transferring. Such fractures are more common in non-ambulant children who are at the severe end of the spectrum of CP, defined as level IV or V according to the GMFCS. Fractures not only cause pain but they further limit the mobility of children with CP leading to muscle wasting through disuse, hospitalization, and missed schooling.

The cause of injury may not be clear in over 50% of cases, and delay in diagnosis is not uncommon. In the literature, the term ‘spontaneous fracture’ has been used to describe such fractures as they apparently occur without any known external cause. The lack of a clear history of the injury causing the fracture, difficulties in communication, and delay in presentation sometimes lead to suspicion of non – accidental injury.

6.1 Epidemiology of Fractures in C&YP with Cerebral Palsy

There have been a few large epidemiologic studies of fragility fractures in children with CP. One study[9] reported a fracture prevalence of 12 % and another study[10] of reported a fracture prevalence of 6 %. Another study[11] reported 1 year and lifetime fracture prevalence of 3.6 % and 9.7 %.

In children with moderate to severe CP (GMFCS III to V) another study[12] reported a fracture incidence of 4 % per year.

6.2 Types of Fractures in Children with Cerebral Palsy

One study found that the majority (74%) were in the femoral shaft and the supracondylar region.

In a population-based study over 70 % of fractures occurred in lower limb bones.

In another study 80% of fractures occurred in the lower limbs.

6.3 Complications after Fractures C&YP with Cerebral Palsy

Over 10% of CP children developed complications after a fracture, which included further fractures, malunion, non-union, and infections, including pneumonia.

6.4 Risk Factors for Fractures in C&YP with Cerebral Palsy

6.4.1 Non-ambulatory Children

Children with CP who are non-ambulatory and classified as GMFC level V have the highest risk. Contractures and stiffness of the major joints create long lever arms also predisposed to fracture.

Development of bone is dependent on an inherited genetic 'template' that is modified by mechanical loading, and nutritional and endocrine environments. Postnatal bone development is modulated by the mechanical forces to which it is subjected to, which primarily arise from muscle contractions. The skeleton of a healthy growing child continuously adapts to increasing mechanical loading from bigger and stronger muscles by increasing bone mass and altering bone geometry.

By contrast, in a non-ambulant child with CP, muscle weakness and habitual inability to participate in normal load-bearing activities results in reduced periosteal bone expansion. This leads to development of slender long bones, which have increased propensity to fracture.

Radiographs of the bones of CP children often show signs of impaired bone development, such as slender or 'gracile' long bones. They may also show thinning of cortices, prominent trabecular pattern due to osteopenia, or the "washed-out appearance".

Increased fragility of a long bone, such as the femur, in subjects with CP arises because of slower rate of bone mass accrual, smaller periosteal diameters, and thinner cortices. Low areal bone mineral density in the distal lateral femur is associated with fracture risk in children with CP.

Children and young people with disabilities are more likely to suffer all types be abused including physical abuse[14]. If any form of abuse is suspected then follow local safeguarding policies and procedures.

6.4.2 Previous Fractures

Previous fracture is associated with increased fracture risk as the fracture rate increases more than threefold after a previous fracture

6.4.3 Post Surgery

Fractures after lower limb surgery, particularly after hip osteotomy and surgery related to a hip Spica cast, were observed in several studies.

6.4.4 Malnutrition

Oro-motor difficulty is a common co-morbidity in CP. Poor lip closure, inadequate jaw control and a delay in swallowing and sucking are common oro-motor difficulties.

Gastrostomy is used in providing enteral feeding when faced with severe feeding difficulty. Gastrostomy use is associated with an increased fracture risk. However, even mild feeding difficulty can result in malnutrition.

6.4.5 Low Vitamin D levels

Low vitamin D status is common in children with CP due to inadequate exposure to sunlight.

6.4.6 Antiepileptic Drugs

The use of antiepileptic drugs (AED) also contributes to low vitamin D levels and osteomalacia.
The most well-known are phenytoin and phenobarbital, which are inducers of cytochrome P450 enzymes leading to the catabolism of vitamins. Treatment with phenytoin and phenobarbital can be associated with rickets.
More recently established AEDs such as sodium valproate, carbamazepine and oxacarbazepine have been shown to be associated with decreased BMD.

7.0 Bone mineral density in C&YP with Cerebral Palsy

As children have not yet reached peak bone mass, it is not appropriate to use the t-score. Instead, the z-score adjusted for age and sex should be used. When the z-score is less than or equal to −2.0, there is 'low bone mass for chronological age'.

The diagnosis of osteoporosis in children, as defined by the International Society for Clinical Densitometry, includes a bone mineral density (BMD) z-score of less than −2.0 adjusted for age, gender and body size, plus a clinically significant fracture history: either (1) two upper extremity fractures, or (2) a vertebral compression fracture, or (3) a single lower limb fracture. Indeed, CP is the most prevalent childhood condition associated with osteoporosis.

In children with CP, the rate of bone mineral acquisition is diminished relative to normal; thus, the BMD and bone mineral content (BMC) are lower than age-matched normal values. With growth, BMD falls further below normal with increasing age. In spite of an average of 2–5% per year increase in BMD in the distal femur, the BMD z-score decreases further with increasing age.

Poor mobility status predicts a low BMD in children with CP: 97% of non-ambulatory children older than 9 years with moderate-to-severe CP had a distal femur z-score of less than −2.0. Significantly lower z-scores on lumbar spine BMD were found in patients with a history of fracture.

Bone density scans are hard to do on children and unreliable so not routinely indicated except on specialist advice.

8.0 Prevention of bone fragility and fractures in C&YP with Cerebral Palsy

Prevention of bone fragility and fractures is the best strategy in order to avoid pain and suffering, muscle wasting and disuse osteoporosis, increased disability, complications from fractures and missed school time in children with CP. All known risk factors should be minimized.

8.1 Physiotherapy Management

Physical activity and standing weight-bearing should be encouraged. The use of supported standing using a Standing frame is recommended as part of a Postural management programme.

Any stiffness of the major joints and extended periods of immobilization should be avoided. Orthopaedic operations to correct lower limb joint deformities in order to provide plantigrade feet and straight knees will allow standing weight-bearing in children with severe CP.

8.2 Feeding

A multidisciplinary feeding team approach is ideal during assessment and planning for the management of feeding problems.

8.3 Adequate vitamin D and calcium intake

Adequate vitamin D and calcium intake should be assured. A period of 10–15 minutes of exposure to the sun three times a week provides most of the vitamin D needed. Another convenient source of vitamin D is vitamin D-fortified milk and food.

Vitamin D and calcium supplementation should be considered in children with CP who have an insufficient dietary intake and are on chronic AED therapy.

Abidec 0.6mls once a day contains 400 IU ergocalciferol solution

See Shropshire Community NHS Trust Vitamin D guideline -Shropshire Community Health (shropcom.nhs.uk)

9.0 Treatment of bone fragility in C&YP with Cerebral Palsy

9.1 Investigations

Determination of vitamin D status is desirable in children with CP who sustain fractures. A study showed that all 20 institutionalized children and young adults with quadriplegic CP and a history of long-bone fractures had radiological and biochemical evidence of rickets or osteomalacia, and vitamin D treatment resulted in marked clinical improvement with no recurrence of fracture during the treatment period.

Consider additional investigations should a child develop a fracture:

1. Blood: Calcium, Phosphate, PTH, Alkaline phosphatase, 25 OH-D, Urea and Creatinine, FBC and ferritin.

2. Urine: Ca / Osmolality ratio.

3. X-rays: Consider need for lateral spine x- ray to assess for the presence of vertebral compression fractures.

4. X-ray of wrist for bone age or rachitic changes.

9.2 Referral to Consultant with Special Interest in Bone Health

Bisphosphonates are a group of drugs that inhibit osteoclastic bone resorption resulting is an increase in bone mass, improvement in bone strength and reduction in the risk of fragility fractures.

Cyclical intravenous pamidronate therapy in non ambulant C&YP with CP results in increase in BMD and reduction in fracture rate. It is given 3 monthly.

In one study pamidronate treatment lowered the rate of fracture, and a one year course appears to provide a protective effect after treatment ends.

After a child has sustained one fracture discuss with their family if they would like a referral to Prof Shaw, Consultant with special Interest in bone health at Birmingham Children's Hospital, for consideration of DXA scan and bisphosphonate treatment.

If a child has sustained more than 1 fracture then will need to be referred to Prof Shaw at BCH.

The referral can be made by either the child's Paediatrician or GP.

10.0 Consultation

This clinical guideline has been developed with input from with community paediatricians:

Dr Unsworth - Consultant Community Paediatrician
Dr Buch - Consultant Community Paediatrician
Dr Mushtaq - Consultant Community Paediatrician
Dr Minnaar - Consultant Community Paediatrician
Dr Postings – Associate Specialist Community Paediatrics
Dr Ogilvie - Specialty Doctor in Community Paediatrics
Dr Raveendran – Speciality Doctor in Community Paediatrics
Sara Butler – Paediatric Physiotherapist

11.0 Dissemination and Implementation

This clinical guideline will be distributed to relevant staff groups by managers and published on the Trust website.

These guidelines will be disseminated by the following methods:

- Managers Informed via DATIX system who then confirm they have disseminated to staff as appropriate.
- Staff via Team Brief.
- Published to the staff zone of the trust website.

12.0 Monitoring Compliance

Compliance will be monitored by review of any concerns raised about the service by staff or patients.

13.0 References

1. Development and reliability of a system to classify gross motor function in children with cerebral palsy.Robert Palisano,, Peter Rosenbaum, Stephen Walter, Dianne Russell, Ellen Wood5 and Barbara Galuppi. Dev Medicine & Child Neurol 1997;39 (4):214–223
2. Informing evidence-based clinical practice guidelines for children with cerebral palsy at risk of osteoporosis: a systematic review Darcy Fehlings, Lauren Switzer,Payal Agarwal, Charles Wong, Etienne Sochett, Richard Stevenson, Lyn Sonnenberg, Sharon Smile, Elizabeth Young, Joelene Huber, Golda Milo-Manson, Ghassan Abu Kuwaik, Deborah Gaebler. Developmental Medicine & Child Neurology 2012, 54: 106–116
3. Fractures in Children with Cerebral Palsy. M. Zulf Mughal. Curr Osteoporos Rep (2014) 12:313–318
4. Review of fractures and low bone mass in children with cerebral palsy. Ho S-T. J Orthop Trauma Rehabil. 2012;16(2):45–50.
5. Epidemiology of low bone mineral density and fractures in children with severe cerebral palsy: a systematic review. Mergler S, Evenhuis HM, BootAM, DeMan SA, Bindels-De Heus KG, HuijbersWA, et al. Dev Med Child Neurol. 2009;51:773–8.
6. Informing evidence-based clinical practice guidelines for children with cerebral palsy at risk of osteoporosis: a systematic review. Fehlings D, Switzer L, Agarwal P, Wong C, Sochett E, Stevenson R, et al. Dev Med Child Neurol. 2012;54(2):106–16.
7. Fractures in Childen with cerebral palsy: a total population study. Ulrika Uddenfeldt, Eva Nordmark, Phillippe Wagner, Henrik Duppe, Lena Westbom. Dev Med Child Neurol. 2013;55821–827.
8. Decreased fracture incidence after 1 year of pamidronate treatment in children with spastic quadriplegic cerebral palsy. Bachrach SJ. Dev Med Child Neurol. 2010,52:837-842
9. Leet AI, Mesfin A, Pichard C, Launay F, Brintzenhofeszoc K, Levey EB, et al. Fractures in children with cerebral palsy. J Pediatr Orthop. 2006;26:624–7.
10. Presedo A, Dabney KW, Miller F. Fractures in patients with cerebral palsy. J Pediatr Orthop. 2007;27:147–53.
11. Maruyama K, Nakamura K, NashimotoM, Kitamoto F, Oyama M,Tsuchiya Y, et al. Bone fracture in physically disabled children attending schools for handicapped children in Japan. EnvironHealth Prev Med. 2010;15(3):135–40.
12. Stevenson RD, ConawayM, Barrington JW, Cuthill SL,Worley G, Henderson RC. Fracture rate in children with cerebral palsy. Pediatr Rehabil. 2006;9:396–403
13. NICE guideline. Cerebral palsy in under 25s: assessment and management
    Published: 25 January 2017. www.nice.org.uk/guidance/ng62
14. Fifteen-minute consultation: Fractures in non-ambulant children with cerebral palsy | ADC Education & Practice Edition (bmj.com)
    Debelle G, et al. Arch Dis Child Educ Pract Ed 2022;107:15–20. doi:10.1136/archdischild-2020-319882

14.0 Appendices

Appendix 1 - Association of Paediatric Chartered Physiotherapists Standing Frames Patient Leaflet

APCP standing_frames_5[1

Appendix 2- Bone Mineral density Risk Identifier

Bone Mineral Density Risk Identifier.doc