Practice pointer

Hip dislocation in cerebral palsy

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The first report on treatment of hip contractures in cerebral palsy was published in 1880.[1] Lateral migration of the hip (subluxation or partial dislocation) occurs in 30-60% of children with cerebral palsy who are not walking independently at 5 years.[2–5] Its clinical course has been well documented, including its effect on function (such as pain affecting the ability to sit and hygiene).[w1] We review the literature on current management of hip dislocation in cerebral palsy in terms of its clinical course, measurement, and treatments.

Methods

We searched the major databases including PubMed, AMED, Embase, CINAHL, and the Cochrane Library and focused on the prevalence of hip dislocation, its clinical course, measurement of the condition, and treatment options. Keywords used were: "cerebral palsy", "children", "hip subluxation or hip dislocation", "windswept deformity", "migration percentage", "surgery", "postural management", "botulinum toxin", and "pain". The search found little high quality evidence. Studies were largely cohort or population studies. We found no systematic reviews and only two randomised controlled trials and one evidence report (by the American Academy of Cerebral Palsy and Developmental Medicine).[6–8]

Clinical course of hip dislocation

In children with cerebral palsy the hip is normal at birth but the effects of delayed motor development lead to dysplasia. Asymmetrical activity of the muscles surrounding the hip and lack of load bearing affect bone development and are the main causes of subluxation and dislocation (figs 1 and 2).[w2-w5]

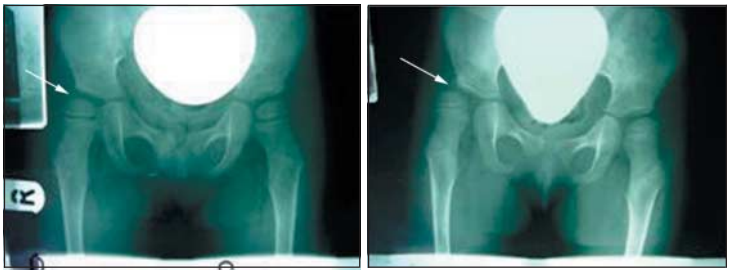

Fig 1 Age 3: early migration of right femoral head (left); age 5: progression to dislocation despite right hip adductor releases (right)

Two large studies (93 and 234 participants) investigated pain in adolescents and adults with cerebral palsy by interview and standardised questionnaires to identify sites and duration of pain. Thirty nine per cent and 47% of interviewees reported hip pain at sites of surgical intervention, which persisted over 20 years.[9 10] Several studies reported that dislocated hips were painful but did not say how this was gauged.[w6, w7] In 29 adults with spastic quadriplegic cerebral palsy, 71% of the 38 dislocated hips were not painful, 11% were intermittently painful, and only 18% were definitely painful.[w8]

Measurement and surveillance

Measuring the percentage of hip migration with an anteroposterior radiograph is the best way to determine the degree of subluxation or dislocation of the hip joint.[1 w1] The rate of migration can help determine risk—an annual migration of 7% or more correlates with a later inability to walk.[w5] The position of the hip and pelvis at radiography must be consistent to ensure that sequential radiographs provide reliable data on changes in hip migration.[1 2] A hip is defined as subluxed if migration is between 33% and 80% and as dislocated if over 80%.[w6] Correlation between hip abduction and migration percentage is weak, and migration percentage does not always reflect a child's functional and clinical picture, both of which contribute to decisions on treatment.[1] The integrity of both hips after intervention should be considered; a successful outcome is one in which both hips are functional and free of pain.

The "paediatric pain profile" is a reliable measure of pain that can be used before, during, and after intervention to determine outcome.[w9] The gross motor function measure and Chailey levels of ability can reliably measure functional outcome.[11 w10]

A hip surveillance programme that developed from a population based study established factors that predicted hip problems at 5 years. These risk factors were migration of 32% or more, had received surgical intervention, or had been prescribed a hip and spinal orthosis by an orthopaedic surgeon.[2] They recommended a radiograph at baseline for all children with cerebral palsy not walking 10 steps at 30 months and

Additional references w1-w23 are on bmj.com

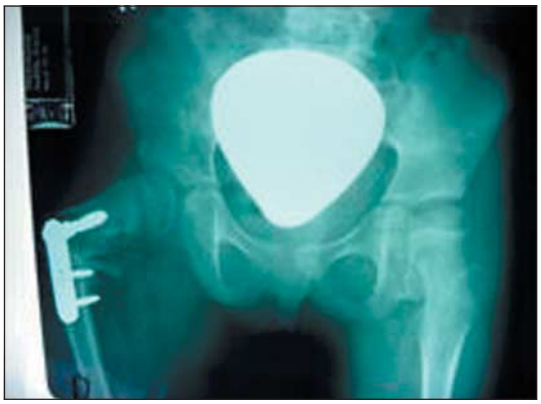

Fig 2 Derotational femoral osteotomy to relocate the femoral head

that children with migration over 15% should be referred to an orthopaedic consultant. Another hip surveillance programme recommends a pelvic radiograph at 18 months and early preventive soft tissue surgery to reduce levels of reconstructive bone surgery.[w11]

Treatment approaches

Soft tissue and bone surgery are the traditional treatments, but newer approaches include 24 hour postural management programmes and injections of botulinum toxin. These complementary treatments can be used early on and may reduce or delay the need for surgery.

The choice of treatment should be based on a clinical assessment that considers the child and the family. Criteria for interventions should be defined according to the child's clinical and functional status, pain levels, hip migration percentage, and long term prognosis, the social and emotional implications of these factors, financial costs, and outcomes.[w12]

The gross motor function classification system predicts later motor ability and together with hip surveillance can identify children at risk of hip problems.[2 w13] This enables conservative approaches such as postural management to be implemented before subluxation occurs. If hip migration continues to worsen, injections of botulinum toxin and surgery may be needed.

Surgery

Unilateral or bilateral soft tissue surgery aims to balance the muscle forces across the hip joint and improve the location of the femoral head in the acetabulum to prevent further displacement.[w1] Soft tissue surgery can be effective when migration is between 30% and 50%,[w14, w15] although several studies report the need for further surgery after soft tissue surgery in over 60% of cases.[1 w7] The American Academy of Cerebral Palsy and Developmental Medicine recently reviewed 27 studies. They found little evidence for the efficacy of adductor releases owing to the "small sample sizes, heterogeneous interventions, poorly defined outcome measures and lack of statistical analysis," and they identified the need for further research.[8]

Unilateral bone surgery and soft tissue surgery cause an alteration in the direction of windsweeping, which causes increased migration of the contralateral hip.[w16, w17]

Bone surgery includes femoral osteotomy, pelvic osteotomy, and open reduction, which restructure the proximal femoral and acetabular anatomy to maintain the position of the hip. Outcomes of bone surgery are generally good when used to prevent progression of hip subluxation.[w1] A long term review of 63 hips found that femoral osteotomy alone was not sufficient to keep the hip centred, and acetabular reconstruction was also advised.[12] Another review supports this finding and indicates that the development of the acetabulum is not affected by femoral osteotomy.[w1]

A prevention programme involving 206 children with cerebral palsy at 5 years of age reported that none had developed hip dislocation.[13] However, many of these children were walking, and many had had surgery (48 soft tissue or bone surgical interventions, 9 selective dorsal rhizotomies, and the insertion of 3 intrathecal baclofen delivery systems).

Many studies on both soft tissue and bone surgery have a short follow-up, and studies cannot be compared because they lack valid functional outcomes, pain is not measured systematically before and after surgery, or they use different combinations of procedures.[8 13]

Long term outcomes of surgery for young adults need to be considered. A cross sectional study found that 54% of patients had a windswept deformity despite hip surgery.[w18] A retrospective study of 60 children and young adults found that those who had not had hip surgery had a significantly increased chance of having both hips undislocated than those who had received surgery.[w19]

Surgery causes upheaval in the lives of children and their families and requires postoperative

Management of hip dislocation

- Children who are unable to sit unsupported at any time or walk more than a few paces with the use of aids (gross motor functional classification system groups IV-V) should start 24 hour postural management programmes in lying as soon as appropriate after birth before hip subluxation occurs, in sitting from 6 months, and in standing from 12 months[11]
- All children who cannot walk more than 10 steps by the age of 30 months should have a hip radiograph to record the percentage of migration.[2] If hip adduction is pronounced, a radiograph at 18 months may be helpful
- Repeat radiographs are recommended every 6-12 months until hip migration is stable[2]
- If migration is greater than 15% at 30 months, positioning equipment to control posture and referral to an orthopaedic surgeon are recommended[2 11]
- An integrated approach to prevent hip problems should include postural management, botulinum toxin injections, orthoses, and surgery[w12]
- The intervention chosen should have a sound clinical basis taking into account the child's clinical and functional status, pain levels, sleep assessment, percentage of hip migration, and long term prognosis, together with the implications of these in social and emotional terms[11]
- Training in postural care should be given to all people directly involved with the child: health professionals, parents, wheelchair services, education services, and respite carers[w23]

rehabilitation. Early intervention programmes, including preventive surgery, have reduced the need for more intensive procedures for hip reconstruction, and salvage surgery has been virtually eliminated.[w10]

Managing posture

Equipment to position lying, sitting, and standing is an established method of maintaining muscle length and joint range.[w20] The widely used kneeblock was first devised in 1978 to control the hips and pelvis when the child is seated.[11 w21]

Twenty four hour approaches to postural management have been developed and are now used routinely; these combine positioning the child during lying, sitting, and standing to encourage active movement and function and prevent deformity. Positioning within the equipment is crucial, and a neutral pelvic position should be maintained in terms of tilt, rotation, and obliquity.[11]

Cohort studies provide limited evidence to support 24 hour postural approaches. A retrospective review of long term outcomes (using percentage of hip migration) showed that children given the treatment before hip subluxation were significantly more likely to have neither hip subluxed than children who received treatment after subluxation or not at all.[14] A pilot study of a sleep system (which placed children in a supine position with hip abduction of 20° bilaterally) on migration percentage, range of motion, and sleep found a significant decrease in percentage of hip migration over one year.[15]

The evidence for postural management programmes is limited and further research is needed.

Botulinum toxin

Despite the increasing use of botulinum toxin in managing migration and reduction of pain in hip subluxation few published studies exist. Three cohort studies have investigated the effect of the toxin on pain and function.[16–18] One study found significant increases in the gross motor function measure and the range of motion and a decrease in the Ashworth scale (towards a more normal muscle tone) one month after injection into the adductor muscles and medial hamstrings. Another study reviewed the effect of the toxin in 27 children aged 4-19 years with complex disability and chronic, severe pain. Dramatic improvements were seen in 25 of the children in tolerating seating and standing supports, sleep, and toileting. Effects lasted an average of 18 weeks. In another study, 11 children received a single injection in the adductor muscle; pain was reduced and function improved, but no effect was seen on hip migration in children with over 40% migration.

A randomised controlled trial investigating the combined effects of injections of botulinum toxin type A and a variable hip abduction orthosis found greater improvement in the intervention group.[6] A randomised controlled trial on botulinum toxin given before surgery for hip dislocation found that it reduced pain after surgery by reducing spasticity.[7]

Summary points

- Reliable measures of pain, hip migration, and function in cerebral palsy are now available
- Options other than surgery, such as postural management and injections of botulinum toxin, offer less invasive approaches to managing hip dislocation but need long term evaluation
- Because of the heterogeneity and complex disability in these children, profiling children rather than using randomised controlled trials might identify which interventions reduce the incidence of hip dislocation
- Combined approaches might be successful in the long term, but they require collaboration between doctors, therapists, and families

Additional educational resources

The American Academy of Developmental Medicine and Child Neurology http://www.aacpdm.org/index?service=page/Home)—a multidisciplinary scientific society devoted to the study of cerebral palsy and other disabilities with childhood onset. It aims to promote professional education for the management of these conditions and to improve the quality of life for people with these disabilities

Information for patients

The National Service Framework for Children, Young People and Maternity Services (www.dh.gov.uk/PolicyAndGuidance/HealthAndSocialCareTopics/ChildrenServices/ChildrenServicesInformation/fs/en)—establishes clear standards for promoting the health and wellbeing of children and young people with disabilities and complex health needs and for providing high quality services that meet their needs

The Scottish Seating and Wheelchair Group (www.sswg.scot.nhs.uk/PubDocs/)—a multidisciplinary organisation that aims to represent all the people who providing and use wheelchairs and seating within Scotland

The Association of Paediatric Chartered Physiotherapists www.csp.org.uk/director/groupandnetworks/ciogs/clientgroups/paediatrics.cfm offers information and advice and has several publications related to cerebral palsy

Conclusion

Randomised controlled trials may not be the best way to assess the treatment options for hip dislocation in cerebral palsy. Instead, studies that profile children to determine the most appropriate interventions to reduce the incidence of hip dislocation might provide more valuable information.[w22] We should seek the views of children and their families on these interventions. Combinations of these approaches might achieve a long term successful outcome, but this requires a commitment to collaborative working between doctors, therapists, and families.

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Lesson of the week

Secondary drowning in a patient with epilepsy

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Drowning is defined as death by suffocation after submersion in a liquid; near drowning is survival, at least temporarily, after aspiration of fluid into the lungs.[1 2] Secondary drowning is death or serious clinical deterioration following near drowning after a period of relative wellbeing; it is not due to neurological causes, respiratory sequelae of inhaled foreign material, or secondary infection. Secondary drowning is caused by inadequate alveolar gas exchange, probably due to primary alveolar membrane dysfunction and loss of surfactant. It can happen after immersion in fresh water or salt water.[3–5] Secondary drowning occurs in 2-5% of all submersion incidents.[6 7]

We report a patient who seemed to recover fully from near drowning. The diagnosis of secondary drowning was not made until several hours after admission with acute respiratory distress. We highlight the need to consider secondary drowning and describe its immediate care, diagnosis, and management.

Case report

A 44 year old woman with a history of hypertension and epilepsy had a tonic-clonic seizure while swimming unaccompanied in her local pool. She was submerged for about one minute then taken to the side of the pool, where the seizure continued for two minutes before stopping spontaneously. She seemed to recover fully and refused offers to be taken to the hospital, preferring to go home.

About an hour and a half after returning home she developed shortness of breath, chest pain, and a cough that produced pink frothy sputum. Her general practitioner was called. Her oxygen saturation was 80% and she was given 50 mg of furosemide. An ambulance was called and she had a second tonic-clonic seizure in the ambulance. On arrival in the accident and emergency department she was deeply unconscious with a Glasgow Coma Scale of 3/15 and a rigid decorticate posture. She had a further tonic-clonic seizure, which was treated with lorazepam 4 mg intravenously. Vital signs were respiratory rate of 26 breaths/min, temperature of 38.5°C, blood pressure of 177/118 mm Hg, and pulse rate of 124 beats/min. On auscultation of the chest, bilateral coarse crepitations were audible throughout both lung fields.

Because of her depressed level of consciousness and the risk of obstruction to ventilation or aspiration of stomach contents, she was given intravenous thiopentone 200 mg and suxamethonium 100 mg and intubated using an 8.0 mm oral tracheal tube. After intubation, pink frothy sputum was aspirated from the endotracheal tube. A nasogastric tube was also placed. The patient was subsequently ventilated with 80% oxygen and 8 cm H_2O PEEP (positive end expiratory pressure). Her oxygen saturation rose to 92%. She was given a further 100 mg of furosemide, which increased oxygen saturation to 100%.

Initially the pulmonary oedema was thought to have a cardiogenic or neurogenic cause. A chest radiograph was consistent with pulmonary oedema, and an echocardiogram performed in the accident and emergency department showed concentric left ventricular hypertrophy with good left ventricular function and slight septal dysfunction. A computed tomogram of the brain was normal. Serum chemistry and haematology were within normal ranges except for mild hyponatraemia (sodium 133 mmol/l) and leucocytosis (13.98×10^9/l). The patient was transferred to theatre recovery before being admitted to intensive care as no bed was immediately available. She was initially sedated

Even after apparent full recovery, near drowning victims are at risk of delayed major complications

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