Cerebral palsy and the birth process

CT Lau, TH Lao

Objective. To review the relationship between cerebral palsy and the birth process.

Data sources. *Medline* and non-*Medline* literature search and personal experience.

Study selection. Articles that commented on the routinely used markers of foetal distress, such as abnormal foetal heart rate, meconium-stained liquor, and foetal acidosis.

Data extraction. Data were extracted and reviewed independently by both authors.

Data synthesis. The use of meconium alone as a predictor of cerebral palsy has a high false-positive rate of up to 99.6%. No specific foetal heart rate pattern can accurately predict subsequent neurological impairment, and a low Apgar score is not by itself an indication of intrapartum asphyxia. The presence of encephalopathy in a neonate after birth and the association of multi-organ system dysfunction are important clues to the prior occurrence of foetal asphyxia.

Conclusion. Cerebral palsy can be caused by asphyxia associated with the birth process. To be able to attribute cerebral palsy to peripartum asphyxia, there should be a sequence of signs during labour, delivery, and the perinatal period. Honest and sympathetic discussion between the obstetrician, paediatrician, and parents is critical throughout the counselling process.

Introduction

It was William John Little, a London orthopaedic surgeon, who in 1862 first reported the association of the conditions known today as cerebral palsy and mental retardation with the birth process. Cerebral palsy is a chronic non-progressive disorder of movement and posture resulting from damage to the developing brain at an early stage of life. It is not a disease but a category of disability. It manifests in early life, usually during the first 2 to 3 years after birth. Although cerebral palsy is a motor system dysfunction, it can be associated with other handicaps that may be more devastating. Moreover, mental retardation occurs in 50% to 60% of children who have cerebral palsy.[1,2] Other neurological impairments include sensory deficits (ie hearing or vision defects) and epilepsy, with the latter occurring in up to 30% to 50% of affected children.[1,2] The motor dysfunction

may be severe enough to affect essential activities such as dressing, eating, and mobility.

The more commonly adopted classification of cerebral palsy is a clinical system based on the extremities involved, the type of tonal dysfunction, and features of involuntary movement. Spastic cerebral palsy is the most common type and accounts for 70% to 80% of all cases[2]; it may present as spastic diplegia, quadriplegia, or hemiplegia. Dyskinetic cerebral palsy occurs in 10% to 15% of cases and can be further classified by type as dystonic, choreoathetoid, ballismus, or tremulous.[2] The athetoid type of cerebral palsy is classically associated with bilirubin encephalopathy. With the advent of phototherapy, the incidence of athetoid-type cerebral palsy has decreased significantly.[3] There is increasing concern, however, about a possible rise in incidence, as more babies are now discharged early from hospital, especially in the first 12 to 48 hours of life.[3] Rigid and ataxic cerebral palsy are relatively rare.

The incidence and prevalence of cerebral palsy

Prevalence studies of cerebral palsy are difficult to conduct, as the condition represents a heterogeneous

Department of Obstetrics and Gynaecology, The University of Hong Kong, Queen Mary Hospital, Pokfulam, Hong Kong
CT Lau, MRCOG, FHKAM (Obstetrics and Gynaecology)
TH Lao, FRCOG, FHKAM (Obstetrics and Gynaecology)

Correspondence to: Dr CT Lau

one. Furthermore, there are multiple aetiologies and the resulting handicap is extremely variable. Since 1960, the incidence of cerebral palsy has been quoted as 2 to 4 cases per 1000 live births. Population studies in the United Kingdom, Australia, Ireland, and Sweden over the past few decades have revealed little change in the overall incidence and little difference between populations.[4] However, a sharp increase in the prevalence of cerebral palsy among babies weighing less than 1500 g at birth has been noted; these preterm babies are now more likely to survive.[5] Preterm babies have at least a 30-fold increased risk of the development of cerebral palsy, compared with babies born at term.

Between 17% and 60% of infants with cerebral palsy have experienced no recognisable adverse event.[1,2] Many studies have suggested a causal relationship between cerebral palsy and birth asphyxia. However, these studies have been influenced by the presence of bias, small sample size, and inadequate control for confounding factors. A large population study is needed and should analyse all affected children and their well-documented perinatal histories; so far, two large-scale studies have been published in the literature. The first was a large cohort study performed in North America called The Collaborative Perinatal Project.[6] More than 41 000 children born between 1959 and 1966 were followed up and examined until the age of 7 years. There were 189 children with cerebral palsy, but 3% to 13% of cases were associated with intrapartum asphyxia. A large proportion of cases were unexplained.[6] The other study was a case-control study of a complete population-based cerebral palsy register in western Australia.[7] There were 183 children with cerebral palsy born between 1975 and 1980. Only approximately 8% of the children with spastic cerebral palsy could be related to intrapartum asphyxia. These two studies found a similar estimate of the proportion of cerebral palsy that could be related to intrapartum asphyxia, despite the difference in geographic area, time of survey, and perinatal care.

Asphyxia studies

Asphyxia implies suffocation with anoxia and carbon dioxide accumulation. In animal studies, the degree of hypoxia that is necessary to produce permanent brain damage is close to that which is lethal.[8] The relative resistance of the newborn mammal to oxygen deprivation is impressive. Animal studies have also suggested that asphyxia may be a subacute or chronic process beginning in pregnancy, rather than an acute insult occurring during labour.[8] The evidence of asphyxia can be manifested during labour, at the time of delivery, or in the early neonatal period.

During labour, abnormal foetal heart rate patterns, meconium-stained liquor, and foetal acidosis are markers of 'foetal distress' and may be related to intrauterine asphyxia. Although continuous foetal heart rate monitoring is commonly adopted in modern obstetric practice, the more frequent detection of an abnormal foetal heart rate pattern and the more frequent intervention as a result of this surveillance has not been shown to decrease mortality or improve long-term outcome.[9,10] There is no specific foetal heart rate pattern that can accurately predict subsequent neurological impairment. Cases of neurological handicap are often preceded by an intrapartum foetal heart rate tracing that is indistinguishable from those obtained from babies that have a normal outcome.[11] The consensus is that although foetal monitoring may provide early evidence of intrauterine asphyxia, such evidence alone is far from adequate to prove that the hypoxia has been of sufficient duration or severity to produce irreversible brain damage. The use of meconium alone as a predictor of cerebral palsy has a high-false positive rate of up to 99.6% and does not prove that a term infant has experienced a degree of asphyxia sufficient to account for later neurological abnormality.[12] Foetal scalp or cord pH are believed to be the best measure of the degree of intrauterine asphyxia. However, there is little correlation between severe acidosis and Apgar score or with the infant's neurological status in the neonatal period.[13,14] The incidence of neonatal neurological complications is increased significantly only in infants with an umbilical arterial pH below 7.00.[15]

Apgar scores

Apgar scores are routinely used by obstetricians and paediatricians. Although asphyxiated babies have low 1- and 5-minute Apgar scores, the majority of babies with scores of <3 at 1 and 5 minutes do not have cerebral palsy later on. The risks of cerebral palsy occurring in a full-term infant is approximately 17% if the Apgar score is ≤3 at 10 minutes; 36% if the score is ≤3 at 15 minutes; and up to 57 % if the score is ≤3 at 20 minutes.[16] Hence, the presence of a low Apgar score does not by itself indicate that intrapartum asphyxia has occurred. Rapid improvement in scores by 5 to 10 minutes indicates that the prior insult was unlikely to have been sufficiently severe to result in neurological deficit.

Presence of encephalopathy

Significant asphyxia that results in subsequent brain damage essentially always produces manifestations of neurological dysfunction in the early neonatal period. The important predictors of later cerebral palsy are a collection of signs that are termed hypoxic-ischaemic encephalopathy (HIE). Infants with mild HIE are usually jittery and hyperalert. Those with moderate HIE have hypotonia, lethargy, and decreased Moro's embrace and sucking reflexes, whereas infants with severe HIE present with stupor and seizures, and lack primitive reflexes. In one study, all infants with mild HIE had a normal neurological outcome; 80% of those with moderate HIE were normal but all of those with severe HIE either died or had neurological sequelae.[17] As neonatal seizures are a hallmark of severe HIE, it has become an important prognostic indicator of later cerebral palsy. Seizures due to peripartum asphyxia most commonly begin during the first 48 to 72 hours of life. The earlier the onset and the more difficult they are to control, the more likely they are to be associated with death or subsequent cerebral palsy.[18] However, 70% of infants with neonatal seizures who survive do not have cerebral palsy[19] and there are conditions other than asphyxia that can produce encephalopathy. It is fair to say that the absence of encephalopathy is strong evidence that substantial prior asphyxia did not occur.

Refining the diagnostic criteria

The American Association of Pediatrics has emphasised that the diagnosis of asphyxia should include evidence of multi-organ system dysfunction that involves the cardiac system, respiratory system, and the renal system.[20] In one study, death occurred in 5% and neurological sequelae developed in 10% of infants with a normal urine output, while in infants with persisting oliguria, 33% died and 67% developed neurological impairment.[21]

It is important that one must avoid using terms such as 'birth asphyxia' or 'birth damage' without carefully reviewing the peripartum and the early neonatal course of a baby that has neurological dysfunction after birth. Once these terms are known to the parents, it may be difficult to convince them that the adverse outcome may not be birth-related. A parent usually wants to know "what exactly has gone wrong" during labour. Obstetricians and paediatricians should jointly examine the records of the labour and neonatal period and explain to the parents what happened and why various decisions were made. Frank discussion and explanation are usually welcomed by parents. Another motive for litigation is anger with the staff. Unkind remarks by nursing or medical staff, failure to listen to the mother's concern around the time of labour and any suggestion that the mother herself is to blame for an adverse outcome are the common complaints from parents.

Conclusion

Cerebral palsy can be caused by asphyxia associated with the birth process; in these cases, asphyxia has to be severe and prolonged. Evidence of such a degree of asphyxia may manifest itself during labour, delivery, and the neonatal course. Therefore, to be able to attribute cerebral palsy to peripartum asphyxia, there should be a sequence of signs during labour, delivery, and the perinatal period. The process of asphyxia is complex and may not result in the foetus showing signs of distress that can be recognised with currently available methods. Even when foetal distress is apparent and delivery is expedited, the damage may already have been done. When an infant is found to have cerebral palsy, it is unwise to assume that more prompt action would have avoided it. It is worth reiterating a British group statement that "professionals' ideas about the obstetric antecedents of cerebral palsy are, at present, dangerously simplistic; those of the general public are even more so."[22] Adverse events in the peripartum period and cerebral palsy are not a common association and may not represent a cause-effect relationship. It is important to complete the clinical work-up to exclude other conditions that may be more pertinent to the development of cerebral palsy. The diagnosis of cerebral palsy may only be apparent when the infant reaches the age of 1 to 2 years. Causes such as trauma, postnatal infection, or metabolic abnormalities must also be considered and excluded.

References

1. Eicher PS, Batshaw ML. Cerebral palsy. Pediatr Clin North Am 1993;40:537-49.
2. Taft LT. Cerebral palsy. Pediatr Rev 1995;16:411-8.
3. MacDonald MG. Hidden risks: early discharge and bilirubin toxicity due to G6PD deficiency. Pediatrics 1995;96:734-8.
4. Murphy DJ, Johnson A. Antenatal and intrapartum risk factors for cerebral palsy in preterm babies. Contemp Rev Obstet Gynaecol 1996;8:20-4.
5. Pharoah P, Cooke T, Cooke RW, Rosenbloom L. Birthweight-specific trends in cerebral palsy. Arch Dis Child 1990;65:602-6.
6. Nelson KB, Ellenberg JH. Antecedents of cerebral palsy: multi-variate analysis of risk. N Engl J Med 1986;315:81-6.

7. Blair E, Stanley FJ. Intrapartum asphyxia: a rare cause of cerebral palsy. J Pediatr 1988;112:515-9.

8. Winn K. Similarities between lethal asphyxia in postneonatal rats and the terminal episode in SIDS. Pediatr Pathol 1986;5:325-35.

9. Leveno KJ, Cunningham FG, Nelson S, et al. A prospective comparison of selective and universal electronic fetal monitoring in 34 995 pregnancies. N Engl J Med 1986;315:615-24.

10. MacDonald D, Grant A, Sheridan-Pereira M, et al. The Dublin randomized controlled trial of intrapartum fetal heart rate monitoring. Am J Obstet Gynecol 1985;152:524-39.

11. Rosen GM, Dickinson JC. The paradox of electronic fetal monitoring: more data may not enable us to predict or prevent infant neurologic morbidity. Am J Obstet Gynecol 1993;168: 745-51.

12. Nelson KB, Ellenberg JH. Obstetric complications as risk factors for cerebral or seizure disorders. JAMA 1984;251: 1843-8.

13. Dijxhoorn MJ, Visser GH, Fidler VJ, et al. Apgar score, meconium and acidemia at birth in relation to neonatal neurological morbidity in term infants. Br J Obstet Gynaecol 1986;93:217-22.

14. Sykes GS, Johnson P, Ashworth F, et al. Do Apgar scores indicate asphyxia? Lancet 1982;1:494-6.

15. Hauth JC. Fetal monitoring: utility and interpretation of umbilical cord gases and fetal scalp sampling. In: Acute perinatal asphyxia in term infants. Bethesda, MD: National Institutes of Health, 1996:63-72.

16. Nelson KB, Ellenberg JH. Apgar scores as predictors of chronic neurologic disability. Pediatrics 1981;68:36-44.

17. Robertson C, Finer N. Term infants with hypoxic-ischemic encephalopathy: outcome at 3.5 years. Dev Med Child Neurol 1985;27:473-84.

18. Minchom P, Niswander K, Chalmers I, et al. Antecedents and outcome of very early neonatal seizures in infants born at or after term. Br J Obstet Gynaecol 1987;94:431-9.

19. Holden KR, Mellits ED, Freeman JM. Neonatal seizures: I. Correlation of prenatal and perinatal events with outcomes. Pediatrics 1982;70:165-76.

20. American Academy of Pediatrics. American College of Obstetricians and Gynecologists: Use and abuse of the Apgar score. Pediatrics 1996;98:141-2.

21. Perlman JM, Tack ED. Renal injury in the asphyxiated newborn infant: relationship to neurologic outcome. J Pediatr 1988;113:875-9.

22. Hey E. Foetal hypoxia and subsequent handicap: the problem of establishing a causal link. In: Chamberlain GV, Orr CJ, Sharp F, editors. Litigation and obstetrics and gynecology. London: Royal College of Obstetricians and Gynaecologists; 1985;233-42.