Aetiology and epidemiology of cerebral palsy

Paul Eunson

Abstract

Cerebral palsy is a common and significant disorder of motor development, with an incidence of 2–2.5 per 1000 live births. Despite improvements in antenatal and perinatal care, there has been little change in the overall numbers of children developing cerebral palsy in the last 40 years. More extremely premature infants are surviving and have more severe forms of cerebral palsy. The common risk factors are prematurity, small-for-gestational age, multiple pregnancy and maternal genitourinary infections. Many children have more than one risk factor for developing cerebral palsy and it is useful to consider causal pathways to cerebral palsy rather than singe causal events. Only by understanding the aetiology and epidemiology of cerebral palsy can programmes be developed to prevent cerebral palsy and plan health services to meet the needs of the affected children.

Keywords aetiology; asphyxia; cerebral palsy; children; epidemiology; maternal health; neonatal encephalopathy; prematurity

Introduction

Cerebral palsy is one of the commonest disorders of child development and potentially it has a major impact on quality of life for the child, acting as a barrier to participation in society. Quality of life for the child's family is also affected with implications for parents being able to work, housing and care of a disabled young person through childhood into adult life. The condition also has a major impact on health, social and education services with increasing survival of more severely affected children.

Cerebral palsy (CP) is defined as follows:

> "Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitations that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems."

Therefore, the term CP can be used to describe a range of motor problems from the child who has a mild impairment in one limb that interferes with sporting activities to the motor difficulties of a child in a wheelchair who has no voluntary movement including speech. The second part of the definition is intended to draw our attention to the accompanying developmental and neurological disorders that may have a greater effect on quality of life than the motor problem. For example, the presence of epilepsy in a child with mild hemiplegic cerebral palsy may significantly affect quality of life and limit independence in later childhood.

Understanding the aetiology of CP is of course important for developing strategies to prevent CP but I believe is also crucial in communicating with parents and children. Parents wish to know what went wrong with brain development or growth, and was there any preventable risk factor. Knowing the cause and, in particular, knowing the neuroanatomy from Magnetic Resonance Imaging of the brain allows an informed discussion with the family. Although the correlation between imaging abnormalities and clinical features is not always strong, it does permit some prediction of future progress and risk of accompanying disorders in the young child.

Understanding the epidemiology of CP is important for planning services for children and young people as well as informing preventative strategies through identification of risk factors and trends in prevalence and severity over time.

Epidemiology

Frequency of cerebral palsy

Prevalence is the number of children with cerebral palsy in a defined population at a given time, usually assessed by population surveys. Studies from various countries quote prevalence of 1.5–2.7 per 1000 children. Incidence is usually calculated as the number of children who develop cerebral palsy in a defined region divided by the number of neonatal survivors in that region.

Variation in frequency of CP between studies is due to a number of factors, including methods of ascertainment. It has been proposed that a severity measure of disability – the Lifestyle Assessment Score – be included in any population study so that only children with a significant developmental impairment are included. It tends to be children at the mild end of the spectrum (Gross Motor Function Classification System level I) who are missed in studies.

Some studies exclude children who have CP of postnatal aetiology, CP as part of a genetic or malformation syndrome, or children with CP who move in or out of the study area. Thus, study results may not be directly comparable.

Improvements in maternal health, better management of premature and difficult deliveries and improvements in neonatal care would have been expected to improve outcomes of pregnancy. Although there have been some changes in patterns of cerebral palsy in the last four decades in developed countries, there has been a disappointing lack of significant decrease in frequency of CP. The numbers of children with more severe forms of CP are increasing, mainly in the group born prematurely as a result of greater survival of these children to an age when CP can be diagnosed. The outcome for children born with lesser degrees of prematurity e.g. born at 28–32 weeks gestation is improving.

Improved survival

Even in the most severely affected young person – unable to lift their head up from the lying position, need for tube feeding, and profound learning difficulties – 50% will survive into the middle

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of their third decade. This is a result of better nutrition, more aggressive prevention and management of chest infections, and more recently, surgery for scoliosis. This is welcome as long as the increased years of survival are good quality of life years. It does mean that there are an increasing number of profoundly disabled young adults reliant on ageing parents, or a poorly funded community care service, and adult services who may not yet able to meet their needs.

Variations between countries

CP registers run under the auspices of European Collaboration of Cerebral Play Registers (SCPE) using similar methodology have similar rates of CP. Very detailed population studies where all children are regularly reviewed produce higher rates of up to 2.7 per 1000.

There is a relative lack of information from developing countries. Where health services are not well developed, it is likely that infants in poor condition at birth, severely premature or small for gestation age (SGA) will not survive and that there may be more postnatal cases caused by conditions such as meningitis, cerebral malaria or severe neonatal jaundice from G6PD deficiency or maternal rhesus isoimmunisation.

The strongest risk factors for development of cerebral palsy are prematurity, low birth weight, twins or higher multiple births, and perinatal infection. Infants who are in poor condition at birth are also at higher risk although this is more likely to indicate that the child is ill rather than that this is the damaging event. These risk factors operate both independently and in conjunction with each other.

Variation with gestational age

Prematurity is a risk factor for developing cerebral palsy and the rate rises considerably with decreasing gestation age to odds risk of 70 if born before 32 weeks. If born before 26 weeks, 16–28% of children will develop CP. Even children born between 32 and 36 weeks gestation still have a higher odds risk of CP than those born after 36 weeks.

Birth weight

Birth weight is a risk factor for cerebral palsy independent of gestation age. The small for gestation age (SGA) infant is more likely to be damaged by hypoxic-ischaemic events in labour. The symmetrically growth retarded neonate (underweight, short height, and low head circumference) is also at risk of developmental delay even in the absence of birth asphyxia. The large for gestational age infant also has a higher risk related to maternal diabetes and obstructed labour. The graph that illustrates the relationship between birth weight and subsequent development of cerebral palsy is J-shaped (Figure 1).

Socio-economic factors

Recent research from a large population study in Sweden shows that mothers in less affluent socio-economic groups have children more at risk of CP and the risk is 50% higher in least affluent group than in highest affluence group. Some but not all the increased risk is associated with perinatal variables such as prematurity and low Apgar scores. Other aetiological factors may be nutritional deficiencies and infections that cause damage to the placenta and predispose towards hypoxia in labour.

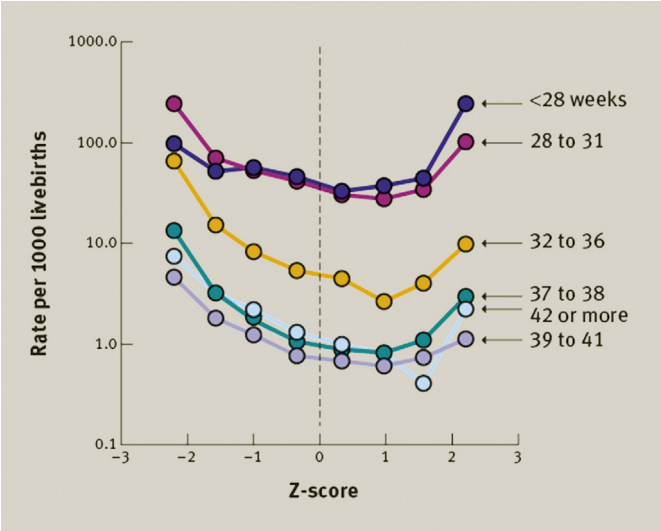

Figure 1 Prevalence of cerebral palsy by Z score of birth weight at different gestation ages (from Jarvis S, Glinianaia S, Blair E. Cerebral Palsy and Intrauterine Growth. Clin Perinatal 33 (2006) 285–300 with permission).

Multiple births

Twin and higher multiple births are strong risk factors for cerebral palsy. The risks rise with increasing number of infants and are higher in identical twins. The risk are related to factors such as twin-to-twin transfusions, vascular anomalies of the placenta, death-in utero of a twin, prematurity, SGA, premature rupture of membranes and hypoxia during labour.

Twin and triplet pregnancies are more common in In-vitro Fertilisation (IVF) programmes and there is some concern that certain techniques to improve IVF outcomes may predispose the infant to CP and other developmental disorders. The disappearing twin syndrome as a cause of cerebral palsy undoubtedly occurs but it is uncertain how common it is.

Classification of cerebral palsy

Severity of cerebral palsy

Severity is determined by the degree of functional motor impairment most often using the Gross Motor Function Classification System, derived from the child's score on the Gross Motor Function Measure. A number of different assessments of upper limb function have been developed, e.g. Manual Ability Classification Score, Adaptive Hand Skills. Alternatively a more global assessment of impact of CP and associated difficulties can be made using measures such as Lifetime Assessment Questionnaire LAQ.

Topographical distribution of motor impairment

The traditional method of describing the topographical type of CP is to use hemiplegia, diplegia and quadriplegia. Clinicians had difficulty agreeing what is severe diplegia and what is mild quadriplegia and differentiating between hemiplegia and asymmetric diplegia. Therefore it has been proposed by SCPE to use the terms symmetrical and asymmetrical CP, to describe which limbs are predominantly affected and to describe if trunk and face are affected. With more children with cerebral palsy being