Elisa Bonaiuti 23/11/2021

PROVA ESTRATTA

12_CONCORSO PUBBLICO, PER TITOLI ED ESAMI, PER LA COPERTURA A TEMPO DETERMINATO, DELLA DURATA DI CINQUE ANNI PER N. 2 POSTI DI COLLABORATORE PROFESSIONALE DI RICERCA SANITARIA - CAT. D, DA ASSEGNARE A: - UOC NEUROLOGIA DELLO SVILUPPO - UOC NEUROPSICHIATRIA INFANTILE

PROVA 1

1. Come valutare dal punto di vista cognitivo un bambino affetto da SMA?

2. A cosa serve il programma Microsoft Excel?

   a. A gestire spool di stampa
   b. A realizzare fogli elettronici per analisi di dati
   c. A realizzare presentazioni

3. Leggere e tradurre il testo sul retro

PERGAMON

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Intelligence and cognitive function in children and adolescents with spinal muscular atrophy

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Abstract

Spinal muscular atrophy is a chronic disease characterised by loss of motor function. The aim of the study was to analyse cognitive functions in a large group of patients with spinal muscular atrophy. It was hypothesised that their intelligence is comparable to controls, but not above average as previously postulated. Ninety-six children and adolescents with spinal muscular atrophy I–III, aged 6.0–18.11 years, 45 non-affected siblings and 59 healthy, matched controls were examined with one- (CPM/SPM), as well as multi-dimensional intelligence tests (Kaufman-ABC; Wechsler tests). The mean IQ measured with the CPM/SPM tests was 109.6 for the spinal muscular atrophy group, 107.3 for the sibs and 104.1 for the healthy controls (no significant difference). In the older children and adolescents (SPM only) the mean IQ was significantly higher for the spinal muscular atrophy patients (109.6) than for the controls (95.4). The standard score in the 'mental processing composite' scale of the Kaufman-ABC was identical in the spinal muscular atrophy group and controls (103.8). The cognitive profile was relatively homogeneous. However, the older children and adolescents did have a significantly higher verbal IQ (113.8) than controls (104.6) in the Wechsler tests. There were no significant differences in any of the tests among different grades of severity (spinal muscular atrophy types I–III). It can be concluded that children and adolescents with spinal muscular atrophy have a general intelligence in the normal range. By adolescence, environmentally mediated aspects of intelligence are higher in patients with spinal muscular atrophy. It could be speculated that the development of cognitive skills and knowledge is a creative way to compensate the many restrictions due to their physical handicap.

Keywords: Spinal muscular atrophy – intelligence; Spinal muscular atrophy – cognitive functions; Spinal muscular atrophy – children and adolescents; Chronic illness; Kaufman-ABC; Raven tests; Wechsler tests

1. Introduction

Proximal spinal muscular atrophy is a genetically heterogeneous group of disorders characterised by loss of motor function, muscular atrophy and degeneration of anterior horn cells. With an incidence exceeding 1:10000 in the population, it is one of the most common autosomal recessive diseases of childhood and adolescence. The gene responsible for autosomal recessive SMA, the survival motor neuron (SMN) gene, has been mapped to the chromosome interval 5q11.2–13.3 and shows homozygous deletions of the telomeric copy in about 90% of patients, leading to a reduction of SMN-protein in the anterior horn cells of the spinal cord (for review, see [1]).

According to the criteria of the SMA Consortium [2] and Zerres and Rudnik-Schöneborn [3] three subtypes can be differentiated according to age of onset, severity of symptoms and motor milestones: SMA type I (severe) with early onset usually in the first 6 months (patients who are never able to sit without support [3], with life span not exceeding infancy in most cases– therefore only the selected subgroup of children surviving beyond the age of 6.0 years could be tested); SMA type II (intermediate) with onset before 18 months of age (patients who are unable to stand or walk unaided, with death occurring usually in adulthood); and SMA type III (mild) with onset mostly after 18 months of age (patients who are able to stand and walk, with near normal life expectancy). Type III has further been subdivided into type IIIa (onset before the age of 3 years) and type IIIb (onset after the age of 3 years) due to the better prognosis of the latter form [3].

While the clinical phenotype and the natural history is

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PROVA 2

1. Quali scale cognitive e di valutazione della qualità di vita adottare per un bambino con tetraplegia?

2. Con il termine "Base di dati" si intende
   a. una collezione di dati, inerenti una specifica attività, opportunamente strutturati e accessibili tramite un software di gestione
   b. un linguaggio di programmazione
   c. un insieme di dati distribuiti sulla rete e accessibili solo tramite un browser

3. Leggere e tradurre il testo sul retro

Current thinking in the health care management of children with cerebral palsy

David Graham[1,2], Simon P Paget[3], Neil Wimalasundera[4]

Summary

- Cerebral palsy is a developmental disorder of movement and posture which is often associated with comorbidities.
- While there is currently a limited range of evidence-based treatments that change the underlying pathology of cerebral palsy, there are many areas in which health care professionals can change the natural history of cerebral palsy and improve participation and quality of life for children with this condition.
- Early identification has become of paramount importance in the management of cerebral palsy, and it is hoped that it will allow earlier access to cerebral palsy interventions that may improve the natural history of the condition.
- Common challenges in the management of cerebral palsy include spasticity and dystonia, management of pain, hip surveillance, sleep and feeding, swallowing and nutrition.
- The six Fs framework (function, family, fitness, fun, friends and future) provides a guide to developing shared goals with families in the management of cerebral palsy.

Cerebral palsy is a movement disorder that encompasses a wide range of non-progressive neurological disabilities;[1] it is the most common cause of physical disability in childhood.[2] At present, cerebral palsy affects about 2.1 per 1000 live births in Australia,[3] a rate similar to other developed nations.[4] A recent article has suggested that the incidence and severity of cerebral palsy in Australia is decreasing, with the authors postulating that improved neuroprotective strategies may account for some of this change.[5] These strategies include the use of maternal magnesium sulphate during labour,[6] the use of maternal antenatal steroids,[5] and better ventilation protocols in pre-term neonates.[7]

The causal pathways to cerebral palsy are multifactorial and are incompletely understood. Events that result in cerebral palsy can occur prenatally, perinatally or postnatally[1,4] — about 70–80% of all cases originate prenatally.[4] A meta-analysis of the use of antenatal magnesium sulfate significantly reduced the risk of cerebral palsy.[6] A recent systematic review found that, since the short term benefit on lung function does not sufficiently outweigh the long term risk of neurodevelopmental impairment, the use of postnatal steroids for ventilation weaning should be abandoned.[7] Less than 10% of all cases are now believed to occur due to acute intrapartum hypoxia-ischaemia.[8] A recent Cochrane review found that brain cooling for term infants with hypoxic ischaemic injury improves outcomes by reducing the level of ongoing neuronal loss after the initial insult.[9] While prematurity remains a common risk factor, about 60% of children with cerebral palsy are born at term.[10] Common risk factors (Box 1)[1,4] are potential targets for prevention of cerebral palsy.

It is increasingly recognised that genetic and epigenetic factors may contribute to cerebral palsy.[11] Mutations in single genes have been found to be associated with ataxic (eg, *KCNC3*, *ITPR1*, *SPTBN2*) and spastic (eg, *KANK1*, *ADD3*, *AP4M1*) forms of cerebral palsy, and genomic copy number variants were found in 20% of cases in a recent study.[12] Gene products that are involved in thrombosis and in the response to cell injury may also be implicated in cerebral palsy causal pathways. Knowledge in this area is likely to expand rapidly over the next decade, providing an increased understanding of causal pathways and offering potential avenues for prevention and/or treatment.

This review provides current thinking in cerebral palsy and summarises common challenges associated with managing the condition. Recently published guidelines have been highlighted to help guide evidence-based practice. We searched Medline and EMBASE using the search terms "cerebral palsy" and "treatment" in order to identify Australian and landmark reviews and studies over the past 20 years, as well as articles that emphasise emerging treatment for cerebral palsy.

Early identification and diagnosis

Early identification has become a key theme in the management of cerebral palsy around the world. Cerebral palsy is often diagnosed between 12 and 24 months of age, but it is hoped that earlier identification will allow earlier access to cerebral palsy interventions that may improve the natural history of the condition.[13]

Clinical assessment remains the main diagnostic tool in establishing whether a child has cerebral palsy or is at high risk of developing cerebral palsy.[13] Delayed motor milestones, asymmetry of movement or abnormal muscle tone are all indicators of possible cerebral palsy diagnosis, but historically identifying children through these methods alone can be slow.

A number of tools have been validated for the early identification of cerebral palsy in high risk populations. The General Movements Assessment has a high sensitivity and specificity in the prediction of cerebral palsy and can be used in children from birth to 20 weeks of age (corrected for prematurity).[14] The best age to perform the assessment is 12–14 weeks after the baby's due date, and is therefore best suited to high risk neonatal follow-up. The assessment is made from a 3–5-minute video of the child's movements taken in a standardised method. Baby Moves is a current trial in Victoria using a phone-based app to record movements as part of screening for extremely pre-term babies.[15] If this study finds a high predictive ability for the app, it could improve access to early identification assessments, as they will not be limited to patients in metropolitan areas with specialist trained therapeutic and medical staff.

The Hammersmith Infant Neurological Examination is not a new tool nor is it specific for cerebral palsy, but it has recently been promoted for use in high risk infant follow-up programs.[16] It can be performed from age 2 months to 2 years and is fast to perform and score, but some training in its use is recommended. Using this standardised neurological assessment provides a framework for monitoring and allows early identification of deviation from normal development, facilitating faster referral for diagnostic assessment and treatment.[16] It has been shown to have

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PROVA 3

1. Cosa si intende per Comunicazione Aumentativa Alternativa e quali sono le sue applicazioni nelle patologie neurologiche dell'età evolutiva?

2. Il termine "Open Source" indica

   a. un software che può essere modificato da chiunque a patto di corrispondere all'autore una offerta libera

   b. un software protetto da diritti d'autore che non può essere modificato da nessuno tranne da chi ne detiene i diritti

   c. un software i cui autori ne permettono e favoriscono il libero studio e l'apporto di modifiche da parte di altri programmatori

3. Leggere e tradurre il testo sul retro

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STUDY PROTOCOL Open Access

Cognition and bimanual performance in children with unilateral cerebral palsy: protocol for a multicentre, cross-sectional study

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Abstract

Background: Motor outcomes of children with unilateral cerebral palsy are clearly documented and well understood, yet few studies describe the cognitive functioning in this population, and the associations between the two is poorly understood. Using two hands together in daily life involves complex motor and cognitive processes. Impairment in either domain may contribute to difficulties with bimanual performance. Research is yet to derive whether, and how, cognition affects a child's ability to use their two hands to perform bimanual tasks.

Methods/Design: This study will use a prospective, cross-sectional multi-centre observational design. Children (aged 6–12 years) with unilateral cerebral palsy will be recruited from one of five Australian treatment centres. We will examine associations between cognition, bimanual performance and brain neuropathology (lesion type and severity) in a sample of 131 children. The primary outcomes are: Motor - the Assisting Hand Assessment; Cognitive - Executive Function; and Brain – lesion location on structural MRI. Secondary data collected will include: Motor - Box and Blocks, ABILHAND- Kids, Sword Test; Cognitive – standard neuropsychological measures of intelligence. We will use generalized linear modelling and structural equation modelling techniques to investigate relationships between bimanual performance, executive function and brain lesion location.

Discussion: This large multi-centre study will examine how cognition affects bimanual performance in children with unilateral cerebral palsy. First, it is anticipated that distinct relationships between bimanual performance and cognition (executive function) will be identified. Second, it is anticipated that interrelationships between bimanual performance and cognition will be associated with common underlying neuropathology. Findings have the potential to improve the specificity of existing upper limb interventions by providing more targeted treatments and influence the development of novel methods to improve both cognitive and motor outcomes in children with unilateral cerebral palsy.

Trial registration: ACTRN12614000631606; Date of retrospective registration 29/05/2014.

Keywords: Cerebral palsy, Children, Cognition, Upper limb, Bimanual performance, Occupational therapy, Neuropsychology

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