

# Letters to the Editor

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## **'Proposed new definition of cerebral palsy does not solve any of the problems of existing definitions'**

SIR—The proposed new definition<sup>1</sup> set out in the first sentence (32 words), is no shorter than that of Mutch et al.<sup>2</sup> (31 words) and does not solve any of the problems of existing definitions. The second sentence does not add to the definition, as the comorbidities it describes may or may not be present.

While brevity is a virtue, clarity is far more important, but we do not consider that the terms used in the new definition improve clarity. For example, 'lesions or anomalies' are replaced by 'disturbances' which suggests an active agent – does that exclude genetic anomalies? Furthermore, the new definition states that the disturbances which occurred in the past were non-progressive. What constitutes a non-progressive disturbance? For example, it might be argued that asphyxia is a progressive condition in that, after the initial hypoxic insult there is a biochemical cascade that creates much of the resulting damage. Surely, what the authors meant is that the lesion or anomaly in the brain, once recognized in early childhood, is no longer progressive. Knowledge of this fact, however protracted the development of the lesion or anomaly might have been in the past, is very important to the child, their family and caregivers, and has long been a criterion for belonging to the cerebral palsy (CP) group. With or without the long annotation, the phrase 'developing fetal or infant brain', adds no further clarity to the age range at which CP may be acquired than 'in the early stages of development' and we agree with Blair and Love that 'activity limitation' is too imprecise a term to define the lower limit of severity required to be included in the group, and may, therefore, be incorrect.

In short, none of the aspects of previous definitions that could benefit from clarification appears to have been clarified by this new definition.

The Australian Cerebral Palsy Register (ACPR) is a collaboration between CP registers in each of the States and Territories of Australia. We are committed to pooling the information on people with CP from the whole of our vast continent and are acutely aware both of the need for a valid and reproducible definition of CP and the difficulties in creating one. We are disappointed in this proposed new definition which seems to offer only new wording without additional clarity, and request that more thought be given to its further clarification.

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2. Mutch LW, Alberman E, Hagberg B, Kodama K, Perat MV. (1992) Cerebral palsy epidemiology: where are we now and where are we going? *Dev Med Child Neurol* 34: 547–555.

## **'Absence of reference to progressive musculoskeletal pathology in definition of cerebral palsy'**

SIR—The authors of the Proposed Definition and Classification of Cerebral Palsy<sup>1</sup> are to be congratulated in their development of a carefully worded clear statement which includes the majority of issues currently considered to be important in the definition of cerebral palsy (CP) from the multiple viewpoints and disciplines. However, as the only orthopaedic surgeon who contributed an oral and written submission to the International Workshop on Definition and Classification of Cerebral Palsy, Bethesda, Maryland, 11–13 July 2004, I am very disappointed by the almost complete absence of any reference to progressive musculoskeletal pathology, particularly in the definition statement.

To me the note in parenthesis under: 11 'brain' ('alterations in the neuromuscular and musculoskeletal systems may occur in CP as a consequence of the chronic motor impairment') is manifestly inadequate. Especially so, given the careful listing of associated disorders and impairments under points: 13 'sensation', 14 'cognition', 15 'communication', 16 'perception', 17 'behaviour', 18 'seizure disorder'. As an orthopaedic surgeon, whose professional life is, in large part, dedicated to the management of musculoskeletal problems in children with CP, I find this unbalanced. Population-based studies of children with CP, identified from a state-wide CP register, confirm an extremely high prevalence of musculoskeletal problems including muscle tendon contractures, bony torsion, hip displacement, spinal deformity, etc. Unlike some of the other impairments which accompany CP, such as those listed in the definition, the musculoskeletal problems develop insidiously, often silently, and frequently do not fully manifest until the second decade of life. However, at this stage they can become the most dominant feature affecting a child's mobility and physical well being. Musculoskeletal problems are so pervasive and have such impact on quality of life that they surely deserve to be listed not as a footnote but as an accompanying impairment in children with CP. How can we communicate clearly with each other and educate parents and carers without an explicit mention of one of the most common and most important features of the CP phenotype? I propose the addition of: 19 'Progressive musculoskeletal pathology'.

The majority of children with CP develop some definable impairment of form or function in the musculoskeletal system. The most common impairments are contractures (e.g. equinus, flexion deformities at the hips and knees), torsional changes in long bones (e.g. femoral and tibial torsion) and joint instability (e.g. hip displacement and breakdown of the mid foot).

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#### 'The authors reply'

We thank the Australian Cerebral Palsy Register (ACPR) and Prof. Graham for their comments on the definition of CP that we and others proposed recently.<sup>1</sup>

The ACPR views our definition as essentially a rewording of previous definitions, including the one suggested by Mutch et al.<sup>2</sup> However, although the basic concept of CP as a clinical descriptive term has been retained, the choice and order of words is deliberately different from the usage in previous definitions. Our goal was to increase the accuracy of the CP construct, and to promote its more uniform use across concerned disciplines. Whereas Mutch et al. designed their definition for use in epidemiological studies,<sup>2</sup> we intend our revised definition to serve the needs of clinicians, researchers, and health officials, and especially to improve the consistency and clarity of communication among these different disciplines.

For this reason, we provided an annotation that expounds in detail on the critical words used in the definition, and our definition cannot be fully appreciated without this annotation. For example, we preferred the term 'disorders' over 'syndromes', because we describe disturbed neurobiological processes, and not distinctive clinical patterns, as implied in the term 'syndromes'. Similarly, we preferred 'disturbances' to 'lesions or anomalies', as the former term encompasses chemical and genetic processes as well as structural abnormalities. Our stress on lack of progressiveness was not intended to address clinical manifestations, which Mac Keith et al.<sup>3</sup> rendered as 'persisting but not unchanging' and Mutch et al. as 'non-progressive but often changing',<sup>2</sup> but to refer to the lack of progression of the underlying pathologic processes. ACPR is concerned that asphyxia could be regarded as progressive, and, thus, its sequelae would be excluded from the CP concept. However, the term 'non-progressive disturbances' refers to processes that do not progress or whose progression has stopped while the fetal or infant brain is developing. These non-progressive disturbances include asphyxia, infection, inflammation, and other initially progressive, but distinctly time-limited, pathological processes.

Additional differences from the Mutch et al. definition include avoiding the term 'stages of development' as it lacks biological precision, and adding a requirement for 'activity limitation' to set clinically and societally-relevant boundaries to the CP concept, following the model of the World Health Organization's International Classification of Functioning, Disability and Health model.<sup>4</sup>

Frequently occurring comorbidities, with particular emphasis on additional neurodevelopmental features, are explicitly cited in the definition to underscore the observation that motor issues are rarely the only features of CP. As Prof. Graham rightly points out, the prevalence, severity, and natural history of musculoskeletal problems seen in people with CP may well justify the inclusion of musculoskeletal pathology in the list of associated disorders and impairments. In the format of the definition, they could be included as 'progressive musculoskeletal pathology'. The writing group will consider this emendation when it meets in February 2006 to complete these documents with full consideration of all feedback received.

We urgently need to increase our understanding of the pathophysiology, and the prevention and management of disabilities in people described as having CP. We also need to improve the organization of services for people with CP and their families. These goals require a clear definition of the CP concept. We are grateful for the responses to our proposal, which will contribute to achieving this objective.

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If you would like to make comments either on the original documentation or on the correspondence published here please respond to the Castang Foundation website ([www.castangfoundation.net](http://www.castangfoundation.net)).

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## 'Epilepsy responds to vagus nerve stimulation in ring chromosome 20 syndrome'

SIR—With reference to the report of ring chromosome 20 (r20) syndrome with intractable epilepsy by Alpman et al.<sup>1</sup>, we report a 9-year-old male with r20 who has had a good response to vagus nerve stimulation (VNS). Our patient was born at 38 weeks' gestation and the perinatal period was uncomplicated. Development was within normal limits and there was no family history of neurological conditions. Seizures began at age 5 years, with upward deviation of the eyes, associated with episodes of confusion lasting up to 1 hour. Other seizures took the form of arrest of activity with upward deviation of the eyes, eyelid flickering, and loss of posture, followed by postictal sleep. An ambulatory electroencephalogram (EEG) in the wake state showed variable frequency waveforms in the bifrontotemporal regions, and periods of generalized rhythmic sharp and slow wave activity were demonstrated from both the prefrontal and anterior temporal electrodes. In sleep, there were periods of rhythmic, fast activity followed by diffuse sharp and slow wave bursts from both frontal leads. Our patient had no dysmorphic features; however, based on clinical history, seizure semiology, and EEG findings, karyotyping was requested particularly with reference to r20. This revealed a mosaic pattern with two cell lines – one normal (20% cells) and one ring chromosome 20 (80% cells).

Treatment with several antiepileptic medications was unsuccessful, including sodium valproate, lamotrigine (as mono and combination therapy), clobazam, prednisolone, and leviteracetam. In addition, whilst on clobazam, rapid regression of skills occurred, resulting in complete loss of language and ambulation. Video telemetry confirmed the presence of numerous absence seizures, nocturnal tonic-clonic seizures, and periods of non-convulsive status epilepticus.

In view of the case report by Chawla et al.<sup>2</sup> suggesting some benefit of VNS in a r20 patient, he was referred for the procedure aged 8 years. A Cyberonics model 102 vagus nerve stimulator was implanted in the upper left chest and connected to the left vagus nerve. Implantation was followed by cycles starting at 30 seconds of stimulation followed by 5 minutes off, with a starting current of 0.25mA, increasing to 2.25mA over a 6-month period. An initial good response was observed, with reduction in seizure frequency and the reacquisition of some previously lost skills, including ambulation. Overall affect improved, and eye contact and social smiling returned. Although the generalized tonic-clonic seizures abated, absence seizures continued, although at a significantly decreased frequency, thus the cycling frequency was increased to 30 seconds of stimulation followed by periods of 3 minutes off. A subsequent increase of the stimulation current to 2.5mA led to all seizure types worsening, and prolonged episodes of non-convulsive status were observed. Reduction of current to 2.25mA led to a gradual improvement, with a return to the previous improved state.

In contrast to the findings of Alpman et al.<sup>1</sup> our experience of VNS in r20 was similar to that previously reported by Chawla et al.<sup>2</sup> At present, on a combination of leviteracetam monotherapy and VNS, our patient remains ambulant and seizures are now confined to occasional nocturnal episodes. Treatment of seizures with a benzodiazepine in this patient

was associated with worsening seizures and regression of skills, although it is unclear whether this was the natural course of the encephalopathy or a side effect of the medication. We, therefore, recommend that benzodiazepines be used with caution in r20. Vagus nerve stimulation may be a useful clinical treatment and should be considered early in the management of intractable epilepsy associated with r20. Considering our experience, there may be a critical level of stimulation that is most effective, above which the clinical benefit may be lost or seizures may even be worsened.

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### 'Alpman replies'

The authors reported that the seizures began at age 5 and vagal nerve stimulation (VNS) was implanted 3 years after. An initial good response was observed in that patient with a current of 2.25mA. In our case, seizures began very early at age of 15 months. VNS was implanted at age of 12. The duration period between onset of seizures and VNS implantation was long, but the seizures were intractable both to medication and to VNS. The frequency of seizures reduced at the beginning of VNS implantation but this was temporary. Although the current was increased to 2.5mA over a 8-month period, no subsequent benefit was achieved at any current. In our opinion the follow-up time is also important. Our patient was followed up for 13 years with antiepileptic drugs and for 4 years with VNS as well as antiepileptic drugs. The reduction of seizures may also be temporary in that case within time.

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