

Progressive course in cerebral palsy?

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Non-progressiveness has been a fundamental constituent of the cerebral palsy (CP) concept across many of its formulations, including the recent definition.¹ This emphasis on distinguishing CP from metabolic and other degenerative conditions is justified by essential differences in pathophysiology, diagnostic work-up, counselling, and (sometimes) management.

In CP, the notion of a lack of progression applies to the underlying pathological processes and not to clinical manifestations,¹ which have been described as 'persisting but not unchanging'.² In a minority of cases, clinical course may even include some loss of skills in late childhood or later life. This may pose serious diagnostic difficulties, which might be lessened by better documentation of the natural history of patients with CP. It also opens the question of secondary prevention.

Over the past 50 years, walking has remained a primary focus, and indeed the most studied motor skill in CP, be it to reflect motor control strategies, evaluate gross motor function, or guide management. It is also a most prominent preoccupation from the time of diagnosis: 'Will my child walk?' Wu et al.³ recently provided probability curves that help predict walking outcome in 2-year-old children with CP based on the presence of basic skills such as rolling, sitting, or pulling to stand. They conducted this population-based study on more than 5000 children sampled from the richly informative database of the Californian Department of Developmental Services.

In this issue of *DMCN*, Day et al. present estimates of maintenance of ambulation capacity in CP based on questionnaire data obtained in 7550 children aged around 10 years and 5721 adults aged around 25 years identified from the same database. They found that a large proportion of individuals with CP are likely to remain within the same functional class of ambulation through adolescence and early adulthood. A significant proportion of children walking with some difficulty at 10 years of age are expected to improve their ambulatory capacity.

Despite the absence of reference to management, the presented figures offer a useful, mostly encouraging picture. They update and refine Crothers' and Paine's observation in their seminal work on the natural history of CP⁴ that among 289 patients assessed for walking ability, only four 'formerly walked but quit'. Day et al. also suggest that more than 25% of children who walk at 10 years are expected to lose this ability by age 25. In particular, children who can walk with more or less difficulty but use a wheelchair around the age of 10 years are more likely to decline in their walking ability. More than one-third of them are expected to completely lose the ability to walk by age 25. This part of the study appears as a negative corollary to the general trend of stability in level of walking ability.

The mechanisms leading to deterioration or loss of

ambulatory capacity in patients with CP were reviewed by the late Michele Bottos and Christina Gericke.⁵ They include a variety of progressive musculoskeletal problems, which may develop throughout life and are related to physical growth, spasticity-related muscle alterations,⁶ and other factors. Some orthopaedic operations may also lead to deterioration of walking ability.

Another suggested group of mechanisms has been termed 'physiological burn-out' (www.geocities.com/pierremal/cp_pages/burnout.htm). This condition is conjectured to correspond to a decline in motor function in relation to demands placed upon the physiological system, resulting in fatigue, reduced muscle power, and deteriorated dexterity and mobility. Psychological factors are also likely to be important. Depressive disorder, in particular, has been increasingly recognized in adolescents and adults with CP. Finally, activity-related brain plasticity, which has been the focus of much recent research, might be responsible for restriction of cortical motor representation of affected body segments.

All these aspects need to be specifically evaluated in order to design effective prevention strategies. In this context, the significance of occasional wheelchair use should be carefully assessed. In the cohorts studied by Day et al., it appeared to reflect severity. However, periodic wheelchair use might be hypothesized to contribute to prevention of walking deterioration by minimizing biomechanical stress on the musculoskeletal system and the imbalance incriminated in 'physiological burn-out'.

In addition to providing valuable information on the natural history of walking ability, further exploitation of the database used by Day et al. is likely to expand our knowledge of other aspects of motor functioning in CP.

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References

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