

CEREBRAL PALSY IN ADOLESCENTS

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Cerebral palsy is a lifelong condition but little information is available on the evolving clinical picture in adolescents. The existing evidence supports the view that cerebral palsy does not remain static once growth is complete, but is subject to key periods of deterioration depending on environmental factors and classification.

SURVIVAL RATE AND LIFE EXPECTANCY

A survival rate in cerebral palsy of 87% at 30 years has been reported by Crichton and co-workers.¹ The same study showed that factors such as total body involvement, associated impairments and learning disability were all associated with shorter life expectancy. Similar conclusions were drawn in an extensive survey by Strauss and Shavelle involving close to 25,000 adults with cerebral palsy.² They showed life expectancy in high functioning adults to be close to that of the general population; these authors identified the key predictors of early mortality as poor basic skills, especially those related to mobility and feeding; this study, however, excluded death in childhood. When considering the overall survival rate it should be remembered that only 85% of children with severe cerebral palsy involving all four limbs survive into adult life.³

CHANGING CLINICAL PRESENTATION

Epidemiological studies have shown that the ratios of the different classifications of cerebral palsy have changed over last 5 decades; these changes are mainly due to changes in the quality of medical care. Since the late 1960s there has been an increase in the survival of preterm infants, and more recently a small rise in survival of full term infants with asphyxia.⁴

These changes in classification mean that different age groups will present with different problems, and experience currently being gained cannot be directly

applied to the problems of future generations. With the increasing number of survivors of preterm birth, the problems of primary and secondary perceptual problems are likely to have a considerable impact on participation in society in adult life, and the survival of more severely affected individuals may in the future result in an increase in early mortality.

EVIDENCE OF DETERIORATION IN ADOLESCENTS

Adequate understanding of cerebral palsy in adolescents is hampered by the lack of consistent ongoing services. However, current evidence shows a changing clinical picture throughout adolescence.

Studies looking at the period of 'transition' from childhood to adult life have reported a decline in both health and functional ability.⁵⁻⁸ Bax and co-workers cited loss of specialist therapy intervention as one of the reasons for this decline.⁵

Several authors have questioned whether there are degenerative changes in the central nervous system; the most convincing of these is the work by Nagashima and co-workers, who studied ten subjects with mild spasticity or dyskinesia.⁹

However, secondary changes in non-neural tissues are the most likely mechanism for deterioration in cerebral palsy. These include changes in the visco-elastic properties of muscle and connective tissue, and poor postural alignment and deformity leading to abnormal 'wear and tear' on the joint surfaces and skeletal system and severe degeneration of femoral heads in adult life as a result of early hip migration. Similarly increased incidence in adult life of spondylolysis of the 5th vertebra in patients with diplegia are examples of this 'wear and tear'.¹⁰

It is useful to divide patients into two groups based on severity of disability. Individuals with moderate to severe

disability are functionally dependent, needing support for everyday activities and health issues; they are therefore more likely to receive ongoing care. Surprisingly, Bax et al showed that school leavers with motor disorder and learning impairment are less likely to get the assistance they need.⁵ This group with profound disability with or without associated learning impairment continues to require therapeutic management to maintain functional skills achieved in childhood. Special attention needs to be paid to communication skills, eating and drinking difficulties, participation in regulation of bodily functions, ease of maintenance of personal hygiene, and ability to transfer.

Patients with mild to moderate disability are functionally independent. By the time they reach early adulthood, they are typically living and working independently. These previously high functioning individuals often show signs of deterioration from the fourth to fifth decades of life. There may be a gradual decline in key functional activities or increasing incidence of discomfort and pain.

DETERIORATION ASSOCIATED WITH SPASTICITY

There is also evidence to suggest that some individuals with a predominant picture of spasticity have a history of sudden deterioration of functional skills, the onset of which may be precipitated by a minor injury, increasing physical demands, or emotional stress. This sudden decline has been described as 'physiological burnout' and attributed to long-term stress and energy depletion. Physiological burnout seems to be most commonly associated with subjects who have dyskinetic symptoms.

Clinical observation suggests that for many patients there is a slow and difficult to perceive decline in ability with increasing use of compensatory strategies. A crisis point may be reached where the patient can no longer compensate.

DETERIORATION ASSOCIATED WITH ATHETOSIS

The most common pattern of deterioration in this group is one of increasing pain, breathing difficulties and urinary tract dysfunction. These symptoms have been attributed to 'wear and tear' on the cervical spine causing narrowing of the upper and midsections of the spinal canal and there may also be compression of the cervical nerve roots.⁹ It has been suggested that spinal cord compression may precipitate ventricular dilatation and further cortical damage. Maruishi and co-workers have reported associated diffuse brain atrophy.⁹

DETERIORATION IN ADULT LIFE

The Gross Motor Function Classification System (GMFCS) has been used as a tool for gauging peak childhood functional level and current status.¹¹ Overall 89% of respondents reported increasing or additional problems including pain, weakness, increased spasticity, reduced endurance, and reduced balance. Outcomes varied according to the GMF level achieved in childhood; overall 6% showed improvement, 48.3% stayed unchanged, and 45.7% declined by one or more levels. These findings are vital to our understanding of cerebral palsy in adult life as they give insight into the magnitude of this problem.

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