

## Cerebral Palsy Life Expectancy: Discrepancies between Literature and Community Data

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### ABSTRACT

Cerebral palsy patients and their families need to predict patients' length of survival for emotional, medical, and financial planning reasons. Providing these estimations is wrought with challenges, some of which are specific to the significant variations in survival that are observed amongst this group of patients. The statistical models that are used to assess life expectancy are plagued by mathematical limitations, faulty assumptions, and the exclusion of factors that are critical to prognosis. In this commentary, we provide evidence that the medical community generally underestimates life expectancy in cerebral palsy. With medical innovations extending lives, some of the literature on life expectancy is outdated, but old data does not explain the extent of the discrepancies we see between what we observe in our communities and what is espoused in the literature. Herein, we offer potential explanations for these discrepancies and call on the medical community to improve predictions of survival in cerebral palsy patients so that they can get the care they need. The harms and dangers of biased life expectancy data cannot be overstated, and cerebral palsy patients are consistently living longer than the current literature would suggest. We demonstrate here why life expectancy models underestimate cerebral palsy survival in the community.

**Keywords:** Cerebral palsy; Life expectancy; Prognosis

## INTRODUCTION

Estimating cerebral palsy patients' survival is critical, not only for the emotional well-being of the patients and their families but also for optimizing medical and financial planning to ensure that the patients receive appropriate care and resources over the course of their lives [1]. Unfortunately, predicting an individual patient's lifespan is problematic in any context, and cerebral palsy presents specific challenges that make survival prognosis particularly complex.

Given the wide range of cerebral cortical and subcortical clinical presentations and different symptoms in cerebral palsy, some experts question if the condition should be a diagnosis or instead simply a description of pathology [2-4]. The heterogeneous nature of cerebral palsy makes it even more unwieldy to predict survival in those with this condition than in those whose condition is associated with narrower clinical variability [3].

Not only is there an enormous range in outcomes across cerebral palsy patients - survival in adults with the condition can differ by

more than 40 years - but many of the relevant factors are lacking in life expectancy models or are considered in an outdated way [5]. These insufficiencies render these models incomplete and unable to reliably predict how long an individual cerebral palsy patient will live.

## LITERATURE REVIEW

### Statistical models have limited predictive value

Certain factors can help to predict outcomes in cerebral palsy patients. For example, those with higher functioning tend to have much more favorable prognosis [6]. Given the importance and the specific implications for neuroplasticity in improving functioning related to cerebral palsy, timing of intervention is also a factor [6]. Nonetheless, as these factors represent a small part of the picture for any individual patient, they are limited in their practical use when estimating the lifespan of any given patient.

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Even with more precise information on cerebral palsy and its prognostic factors, predicting how long a given patient will live is riddled with challenges. Statisticians have long recognized that for even the most clearly defined populations, group data tend to offer little information on individual cases [7-10]. As they point out, mean distributions, which are used to calculate patients' life expectancies, simultaneously provide information about everyone and no one [11].

Predictive models are limited in their value partly because they depend on the imprecise practice of extrapolation. Calculating mean life expectancy requires that each member of the specified cohort has died [12]. Given that meeting this requirement for accurate life expectancy calculation is impossible, statisticians instead extrapolate survival curves and specify the assumptions that accompany them.

We are restricted in our ability to estimate the life span of any given cerebral palsy patient because of mathematical limitations. However, there are several other factors unique to cerebral palsy and its relevant data that make it clear that the estimates we do generate are based on faulty assumptions. These assumptions include that the care each patient receives is equivalent in quality, that predictive factors remain stably related to survival, and that we have enough information on patients to produce reliable information on life expectancy.

#### *Quality of care is not properly accounted for in models of life expectancy*

Though quality of care significantly impacts outcomes, group statistics on cerebral palsy tend to neglect differences in the quality of care that patients receive. Cerebral palsy care has been shown to be affected by several factors that are independent of the clinical realities of the patient, including where the patient lives, the provider they see, and how their care is financed [13].

In some places, cerebral palsy patients receive outdated approaches to care, presumably with less beneficial outcomes than in cases where the most advanced and evidence-based interventions are deployed [13]. Any model for life expectancy that fails to take the quality of an individual patient's care into account is inherently incomplete.

#### *The specific relationship between factors and survival changes over time*

The need for gastrostomy feeding has been identified as an indicator of poorer prognosis in cerebral palsy, so lifespan estimates have tended to be lower for those using feeding tubes [6]. However, in recent years gastrostomy feeding has increased significantly, with cerebral palsy patients being offered feeding tubes much sooner than they once would have been. It is thus likely no longer appropriate in many instances for life expectancy models to consider gastrostomy feeding as a marker for shorter survival. On the contrary, earlier use of feeding tubes may instead now indicate better quality of care, which could extend survival rather than shorten it. Indeed today, those using feeding tubes have a lower average degree of disability than they once did, and their survival has improved across recent decades [14].

#### *We do not fully understand the impact of certain factors on survival*

In addition to higher quality of care, there are also nonclinical factors that have been deemed important for survival in cerebral palsy patients. Researchers point to socioeconomic factors, for instance, including access to better care, appropriate housing, superior schooling, occupational opportunities, and proper nutrition as important for promoting healthier and longer lives in cerebral palsy patients [6]. However, how exactly to factor these things into an individual case is not clear.

Though cerebral palsy registers could help us improve our understanding of these factors, many of these registers are not complete with respect to the information they provide. For instance, some may focus specifically on educating those with cognitive deficits and thus not include reliable information on those with good cognitive functioning [15].

New cerebral palsy registers are being developed to comprehensively cover all the cases within given geographical areas and to stratify them based on both clinical and nonclinical factors such as: dates of birth, sex, birth weight, and severity of functional impairments, but these registers have yet to provide significant information on longevity [15,16].

### **The medical community underestimates life expectancy in cerebral palsy**

Given the importance of insight into life expectancy for cerebral palsy patients and their families, we cannot shy away from the challenge of optimizing our tools for providing such information. Clearly the current models suffer several limitations and need to be updated and improved. Specifically, there is evidence that the medical literature on life expectancy in cerebral palsy patients significantly underestimates how much time these patients have left.

#### *Medical innovations are leading to longer lives in cerebral palsy patients*

High quality cerebral palsy research in the 21<sup>st</sup> century has led to significant improvements in both the safety and efficacy of cerebral palsy interventions and growth in clinical trials aimed at identifying more and better ways to improve and extend the lives of those with the condition [13-17]. There is little debate that cerebral palsy survival has improved since the middle of the 20<sup>th</sup> century, particularly for those with severe disease [6].

One study showed that the life expectancy for a group of males with cerebral palsy increased from 14 years based on data from 1983 to 1995 to 20 years based on new data available in 2002 [6]. Life expectancy among people with cerebral palsy has now been shown in studies across several countries to be largely similar to that of the general population, with more than 90% of children with the condition having normal life expectancy [13-17].

Even in the tube-fed patients who are considered at higher risk for poor prognosis, improvements are being observed. A study on more than 50,000 cerebral palsy patients found that between

1983 and 2010, adolescent and adult life expectancies increased by 1 to 3 years for tube-fed patients [1].

There are a multitude of factors that help explain the improvements we have seen in cerebral palsy longevity and care over the past 25 years, including better anti-seizure medications, use of the ketogenic diet and other evidence-based nutritive strategies, prevention and treatment of fractures and osteoporosis, and medications for spasticity including intrathecal baclofen pumps and botulinum toxin [6,18-23]. We can also better prevent aspiration and pneumonia through salivary gland injections of botulinum toxin and salivary gland or duct ligation duct ligation, which helps prevent pneumonia [24-28].

*Discrepancy exists between what is observed in the community and what is seen in the literature*

Despite its complexity, life expectancy estimation for cerebral palsy patients is based largely on a small number of relevant publications. Unfortunately, there are critical flaws in the data represented in these pieces, and the results are inconsistent with what we have found while investigating the lifespan of cerebral palsy patients across several local clinics.

Interestingly, we found not only that age of death of individual cerebral palsy patients differed significantly from statistical models for life expectancy may predict but that the difference tended heavily in one direction. Rather than simply deviating randomly from what might be expected, the actual lifespan of the cerebral palsy patients we followed was invariably longer than what would be predicted based on the current medical literature.

An investigation into the details of the many publications on life expectancy in cerebral palsy paints a clearer picture for the discrepancy between what is seen in the literature versus what is seen in our communities. According to sworn deposition testimony by Dr. Herbert Grossman, he did not gather the information on patients that was included in his *New England Journal of Medicine* article "Life Expectancy in Profoundly Handicapped People with Mental Retardation" from August 1990 that described nearly 130,000 people within the California Department of Developmental Services between 1980 and 1991[29,30]. In addition to the data collection issues and resulting unreliability of these data, the study was lacking critical information with serious implications for life expectancy, including the type of medical care the patients were receiving, their specific cause of death, and whether patients died in institutions.

Two authors of several of the publications on life expectancy in cerebral palsy - Drs. Strauss and Shavelle - also used data from the State of California and specifically employed the Client Development Evaluation Report (CDER, which formed the basis of their life expectancy data and relevant claims in their publications [5,6,31-35]. Unfortunately, medical experts were not involved in providing the information required for this complex tool marring the data foundational to much of the literature on cerebral palsy life expectancy.

Any resulting statistical analysis conducted on these unreliable data cannot therefore be viewed as valid. One egregious problem

that has since been revealed is that approximately 30% of the people whose data were analyzed did not have the appropriate diagnosis for this type of analysis. Critical information on the care received is thus completely lacking in all published literature based on CDER.

*Investigations have revealed underlying drivers for the discrepancy*

More troubling than statistical limitations and lack of comprehensive data is corruption to the medical literature due to specific monetary incentives. Under oath, an author of the *New England Journal of Medicine* study mentioned above admitted that the intention behind the publication of the article was to defend medical malpractice lawsuits [29,30]. In other words, the motivation for the article was to provide evidence for shorter life expectancies and thus lower perceived lifetime costs for cerebral palsy patients so that payouts could be justifiably lowered.

In addition to the lack of reliability of the CDER data, it has also come to light that Drs. Strauss and Shavelle have not cooperated in providing access to the relevant database so that other experts could have the opportunity to analyze the raw data to determine their validity and the ability to draw any meaningful conclusions from them [36]. When considering the reasons for their refusal to share the database, it is important to note that Dr. Shavelle has conceded that in his expert witness testimonies, approximately 90% of his depositions are on behalf of the defendant, meaning that the value his testimony serves is to minimize payouts for cerebral palsy patients and their families by providing apparent evidence for lower economic damages.

It has also come to light that these statisticians conducted complex but seemingly arbitrary statistical analyses that further drove down life expectancy estimates in their publication [37]. In addition, it is worth noting that literature in support of plaintiffs - i.e. that suggests longer survival in cerebral palsy patients - was once available on Strauss and Shavelle's website but has since been taken down [36]. While we cannot draw firm conclusions for the reasons for any of these pieces of information, we must consider that when taken together in context, Drs. Strauss and Shavelle may have had some motivation to underestimate survival in those with cerebral palsy.

## CONCLUSION

It is impossible to predict with great accuracy and precision how long an individual cerebral palsy patient will live. Our current strategies for these estimations are limited by mathematical realities as well as a lack of relevant data on how clinical and nonclinical factors affect longevity in these patients. With new medical innovations, even the most severely disabled cerebral palsy patients are living longer than ever before, and what we are observing in our communities does not match the medical literature on cerebral palsy life expectancy. Specifically, the medical literature - and often medical professionals themselves - underestimate how long cerebral palsy patients will survive.

Though more research is needed to help us improve our ability to predict life expectancy so that patients and their families can plan accordingly, we know that the best care a cerebral palsy patient can receive is individualized and includes a combination of treatments [1,6]. Common sense dictates that more resources are associated with more access to better care and more opportunity to leverage whatever medical innovations become available in the future - the realities of which we simply cannot incorporate into any model of life expectancy or any model for financial planning.

The complexity of life expectancy estimation in cerebral palsy makes it perhaps unsurprising how a case for a shorter lifespan could be built in almost any individual case. Nonetheless, as clinicians, we know that even highly compromised cerebral palsy patients, who are spastic, severely mentally retarded, and fed through a gastrostomy tube, can live a long life and that their care should not be undermined by cold statistics that do not produce predictions consistent with what we see in practice. Several aspects of each patient's condition and fragility, the care they receive, and the environment in which they live play into their survival and cannot be accounted for by current models.

Our goal as a medical community must be to provide the best possible information at hand. The current medical literature on life expectancy in cerebral palsy is flawed and cannot be used to accurately address individual cerebral palsy patients. Instead, as physicians who care for cerebral palsy patients, our approach to predicting survival should embrace the strengths of the literature and incorporate our knowledge, training, and clinical practice experience. By supplementing the credible data on life expectancy in cerebral palsy with our expertise on the influence of fragility, stability, environmental factors, and access to appropriate medical care, we can significantly improve upon the predictions offered through statistical models alone.

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