Childhood dementia: quantifying the burden on patients, carers, the healthcare system and our society

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Introduction

- Childhood dementia is a recognised group of disorders comprised of >70 individual rare and ultra-rare genetic conditions; many are inborn errors of metabolism.
- Characterised by progressive neurocognitive decline, severely impacted quality of life and shortened life expectancy.
- Childhood dementia as a collective has received little recognition, instead, disorders are often considered individually.
- Children with dementia commonly suffer co-morbid disease; e.g. seizures, motor and sensory deficits, visual loss.
- It places a tremendous emotional, physical and financial burden on those involved in care and support.
- The direct healthcare and indirect societal costs of the collective has not been assessed.

The objectives of this study were to:
- define and identify the population affected by childhood dementia
- quantify the current burden of childhood dementia within the Australian population
- estimate the projected financial costs of childhood dementia to the Australian economy.

Methods

- Desktop research defined and identified the current and forecasted number of cases and costs associated with childhood dementia in Australia from 2021 to 2030. Case definition was restricted to childhood dementia disorders of monogenic genetic etiology.
- Incidence and life expectancy estimates were derived for each individual condition through extensive literature research. These estimates were used to calculate an Years of Life Lost (YLL) estimate for each condition in addition to a prevalence and life expectancy estimate for childhood dementia as a whole.
- A Disability Adjusted Life Year (DALY) was assigned to each condition based on an average health state weight (0.312) from the Global Health Estimate study (2015).
- An average annual healthcare cost estimate of AUD$27,900 based on three studies was applied to all individual conditions (Wyatt et al. 2012, Imrie et al. 2009 and Hendrie et al. 2011).
- Annual indirect costs, derived from comparator literature (Imrie et al., 2009), equate to AUD$27,433 and were equally applied to all conditions.
- An estimate of AUD$213,000 per year for the Value of a Statistical Life Year (VSLY); as recommended by the Australian Government’s Office of Best Practice Regulation was applied to the YLL to measure the opportunity cost of a life year lost due to premature mortality.
- An average annual cost to the National Disability Insurance Scheme (Australia) of AUD$97,000 is applied using age-specific costs to estimate the spend over an individual’s lifetime, depending on life expectancy for each condition.
Defining the population

- Case definition: restricted to childhood neurocognitive decline (dementia) of monogenic genetic aetiology. Episodic encephalopathies and epileptic encephalopathies were excluded.
- >70 individual genetic conditions were identified, including Batten disease, Sanfilippo syndrome, Niemann-Pick disease, Tay-Sachs disease, metachromatic leukodystrophy, Rett syndrome and some mitochondrial disorders.
- The true number of conditions is >>>70 as conditions were grouped under broad disease categories, & the number of ultra-rare and novel disorders satisfying inclusion is constantly expanding.
- Identified disorders, based on their cause and characteristics, were grouped into 11 broad categories. A large proportion of the disorders are inborn errors of metabolism. Lysosomal disease was the largest category (21%) followed by mitochondrial disorders (20%). Third most frequent is the group of “other rare neurodegenerative conditions” a disparate group of conditions such as Rett syndrome and Juvenile Huntington’s disease (19%).
- < 5% of the disorders have widely available treatments with a close-to-normal life expectancy (assuming timely diagnosis). Symptom management and palliative care are the only options for most.

Key findings

- The collective incidence of disorders that cause childhood dementia is 36 per 100,000 births (1 in 2,800 births).
- The average life expectancy for childhood dementia is 28 years.

Globally the impact of childhood dementia is:

- 50,000 births every year
- 700,000 individuals currently living with childhood dementia
- 48,300 premature deaths annually

The cost to the Australian economy each year is $389 million and will be at least $3.9 billion in the next decade.
Table 1: Childhood dementia in Australia: Incidence, prevalence and costs

<table>
<thead>
<tr>
<th>OUTCOME</th>
<th>ANNUALLY</th>
<th>2021 to 2030</th>
</tr>
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<tbody>
<tr>
<td>Australian birth cohort</td>
<td>363,074</td>
<td>3,630,736</td>
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<tr>
<td>Total childhood dementia births</td>
<td>129</td>
<td>1,295</td>
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<tr>
<td>Incidence per 100,000</td>
<td>35.67</td>
<td>35.67</td>
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**PREVALENCE**

<table>
<thead>
<tr>
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<th>2021 to 2030</th>
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<tbody>
<tr>
<td>Children living with childhood dementia: 2021 to 2030</td>
<td></td>
<td>1,396 to 1,545</td>
</tr>
<tr>
<td>Persons living with childhood dementia: 2021 to 2030</td>
<td></td>
<td>2,273 to 2,516</td>
</tr>
</tbody>
</table>

<table>
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<tr>
<th></th>
<th>ANNUALLY</th>
<th>2021 to 2030</th>
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<tbody>
<tr>
<td>Years of life lost</td>
<td>1,096</td>
<td>10,961</td>
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<tr>
<td>Life expectancy</td>
<td>27.81</td>
<td>27.81</td>
</tr>
<tr>
<td>Years of life lost due to disability</td>
<td>451</td>
<td>4,513</td>
</tr>
<tr>
<td>Costs to the healthcare system</td>
<td>$40,391,688</td>
<td>$403,916,877</td>
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<tr>
<td>Indirect costs</td>
<td>$39,715,290</td>
<td>$397,152,904</td>
</tr>
<tr>
<td>Costs of life year lost</td>
<td>$233,479,553</td>
<td>$2,334,795,527</td>
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<td>Cost to National Disability Insurance Scheme</td>
<td>$75,022,926</td>
<td>$750,229,262</td>
</tr>
<tr>
<td><strong>TOTAL COST OF CHILDHOOD DEMENTIA</strong></td>
<td>$388,609,457</td>
<td>$3,886,094,570</td>
</tr>
</tbody>
</table>

Figure 1: Costs of childhood dementia

**Note:** Of the 70 identified disorders, 32 were excluded from the analysis due to non-availability of incidence and life expectancy data. There is limited robust data on the costs attributed to each condition in this list, therefore, the costs attributed to childhood dementia as a whole are expected to be higher than those provided in this analysis.
Angelina was like most other teenagers. She had no signs or symptoms or any abnormalities. She was extremely social, self-motivated, goal-driven, academic and had big dreams for her future. She was in school musicals and attending acting classes and wanted to become a makeup artist and business owner.

In September 2018, 14-year-old Angelina, was found unconscious in a corridor at her school. One year later, Angelina was diagnosed with childhood dementia caused by a rare genetic disorder called Lafora disease. Symptoms include seizures, muscle spasms, difficulty walking, behavioural changes, confusion and cognitive decline. Within just a few years from the onset of symptoms, children typically find it hard to complete daily activities. Most only live for around 10 years from those first symptoms.

Sadly, Angelina’s condition declined in the year following her diagnosis. At times Angelina started to find it difficult to speak, swallow or walk unassisted. By June 2020, Angelina’s behaviour started to change. For Angelina, this made her irritable and impacted her mental health. She started to refuse to eat, co-operate with self-care, get out of bed, or take her medications. Angelina’s short and long-term memory and cognitive abilities significantly declined too.
Conclusions & Acknowledgements

- Dementia is typically assumed to be an aging adult disease, however, tragically, children are affected too.

- This study demonstrated for the first time the collective burden associated with childhood dementia: the tremendous negative impact it has on affected children, families, and the community, and the resulting health and productivity costs.

- The individually rare disorders which result in childhood dementia are underrepresented and often neglected in research funding and health system planning and resourcing.

- The Childhood Dementia Initiative is a driving research and advocacy to urgently disrupt the impact of childhood dementia on children and families across the world.

- By considering the >70 disorders that cause childhood dementia as a collective, the Initiative aims to challenge the world to think differently about childhood dementia.

- We will enable global collaborative research across multiple childhood dementia disorders: achieving economies of scale and gains in efficiency in all aspects of research to accelerate therapeutic development.

Thank you to:

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- the clinicians who gave much valued input
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Read the full report here: [www.childhooddementia.org/burdenstudy](http://www.childhooddementia.org/burdenstudy).