

Childhood dementia: key facts and statistics

A fact sheet for health professionals

childhood
dementia
INITIATIVE

| Statistics

- One in every 2,900¹ babies is born with a condition that causes childhood dementia. That equates to an estimated 50,000 babies born each year worldwide with a condition that causes childhood dementia².
- An estimated 1,400 people in Australia are living with childhood dementia (over 70% of which are children)¹. Based on Australian figures, it's estimated that 420,000 people are living with childhood dementia globally¹.
- Annually in Australia, there are more than 90 deaths in people with childhood dementia, compared with 92 deaths per year from cancer in children aged 0–14¹. Globally, 48,300² children and young people with childhood dementia die annually.
- Half of the children with dementia die by the age of 10. Less than a third of those born with a childhood dementia condition reach the age of 18¹.
- There are no cures and childhood dementia is terminal. Research is making progress and showing great promise for children in the future.
- The incidence of childhood dementia is similar to that of well-known disorders like cystic fibrosis in Australia¹.

| Definition

Childhood dementia is an umbrella term representing a diverse group of progressive childhood conditions. Defined as global neurocognitive decline with multiple developmental skill loss following a period of developmental progress³, the hallmark of childhood dementia is enduring and progressive loss of previously acquired developmental skills^{3,4}. Moreover, childhood dementia can be distinguished from static and transient intellectual disabilities without ongoing cognitive decline^{3,5}. (Figure 1)

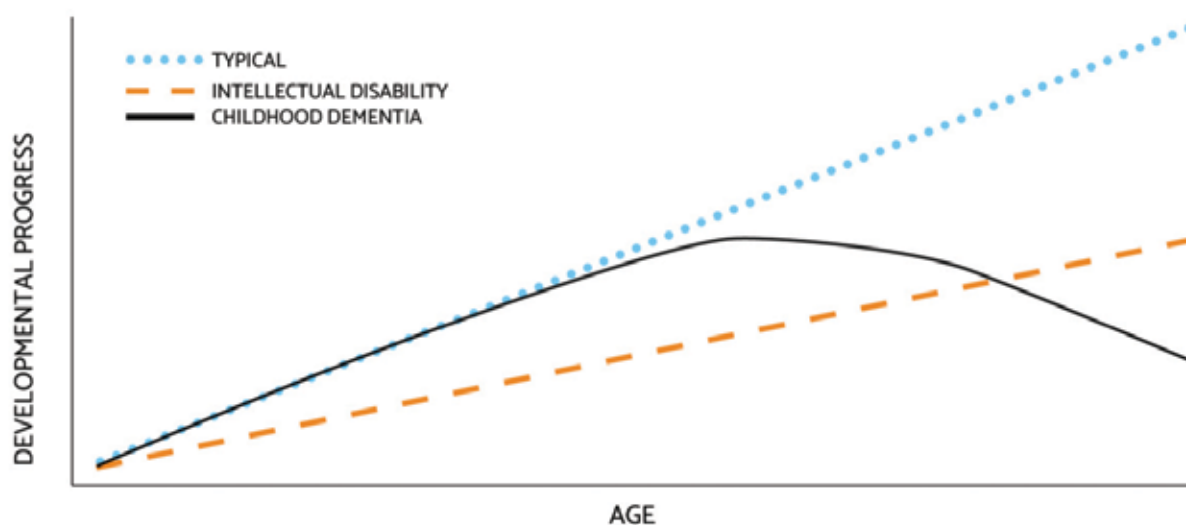


Figure 1. Childhood dementia disease trajectory compared with intellectual disability.

| Causes of childhood dementia

Childhood dementia is an umbrella term for more than 145 individual genetic conditions which cause progressive brain damage.

Broadly, there are four different categories of childhood dementia causing conditions^{1,6,7} including:

Inborn errors of metabolism

- ➔ Lysosomal disorders (e.g. mucopolysaccharidosis types I, II, III and VII, Tay Sachs disease, Niemann-Pick disease type C, Batten disease)
- ➔ Other disorders of lipid metabolism and transport
- ➔ Disorders of amino acid and other organic acid metabolism
- ➔ Vitamin-responsive inborn errors of metabolism
- ➔ Disorders of mineral absorption and transport
- ➔ Mitochondrial disorders (e.g. Leigh syndrome)
- ➔ Peroxisomal disease (e.g. X-linked adrenoleukodystrophy)
- ➔ Other inborn errors of metabolism (e.g. Canavan disease, cobalamin C disease, Menkes disease, Lafora disease)

Neurodegeneration with brain iron accumulation (NBIA)

- E.g. pantothenate kinase-associated neurodegeneration (PKAN), beta-propeller protein-associated neurodegeneration (BPAN)

Other leukodystrophies

- E.g. Alexander disease type I, vanishing white matter disease, Aicardi-Goutières syndrome

Diseases not otherwise categorised

- E.g. Rett syndrome, juvenile Huntington's disease, Cockayne syndrome

This list is regularly reviewed and expanded as new evidence emerges. Notably, the list is expected to grow significantly following the completion of a global expert consensus process on the definition of childhood dementia, currently in preparation for publication. This consensus affirms that any condition causing progressive cognitive decline, with onset before the age of 18, may qualify as a cause of childhood dementia – including conditions not yet named on this list, and developmental and epileptic encephalopathies (DEEs) where progressive cognitive decline is present. Importantly, this decline does not need to be continuous; periods of plateau or stability before further decline are recognised as part of the condition. A condition's absence from this list should not be taken as exclusion.

For a full list of conditions, go to our website:

<https://www.childhooddementia.org/childhood-dementia-disorders>

| Symptoms

The symptoms of childhood dementia vary from condition to condition and also change over time. The common symptoms are^{1,2,6}.

Cognitive symptoms

- Decline in cognitive ability
- Problems with attention and concentration
- Memory loss and learning difficulties
- Problems with thinking and reasoning
- Confusion and disorientation
- Loss of communication and language skills

Physical and motor symptoms

- Movement disorders such as muscle spasms and ataxia
- Loss of vision and hearing
- Incontinence
- Seizures

Behavioural and psychological symptoms

- Changes in behaviour, which may include aggression, irritability, hyperactivity and social disinhibition
- Difficulties with emotional regulation, including anxiety, fear and panic attacks
- Wandering and restlessness
- Psychosis which may include hallucinations
- Sleep disturbance, which is often severe
- Deterioration of social skills^{1,2,6}

In contrast to adult-onset dementia, and in addition to the symptoms above, childhood dementia is commonly associated with the involvement of other organs and physiological systems such as the liver, gastrointestinal system, bones and joints, and the heart^{1,6}. This varies depending on the underlying cause of childhood dementia.

| Prognosis

Some types of childhood dementia present in infancy, progressing rapidly and leading to death in the first year of life. For other conditions, initial symptoms may not present until later in childhood and progress relatively slowly, with survival typically into the teens or early adulthood. The cause of death in childhood dementia disorders is usually attributed to respiratory complications of end-stage disease (such as pneumonia), neurological complications (for example, intractable epilepsy), or cardiac events².

| Childhood Dementia Initiative

Childhood Dementia Initiative was established in 2020. It is the world's first organisation to represent conditions that lead to childhood dementia. Our purpose is to transform research, care and quality of life for children with dementia so they may live long and fulfilling lives. To find out more, go to <https://www.childhooddementia.org/>

| References

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