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Case Report
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Probably the Longest Lived Patient on Record with Alexander's Disease

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Introduction

Alexander's Disease or Alexander's Leukodystrophy (AL) was named after W.S Alexander, an Australian pathologist working in the United Kingdom on a fellowship. He described a 15-month-old boy with megalencephaly, hydrocephalus, and psychomotor delays together with progressive degeneration of white matter in the brain and intellectual decline [1]. AL is essentially a childhood dementia. It is a rare genetic disorder characterised by abnormal protein deposits. These deposits are found mainly in astrocyte cells. The disease is inherited in an autosomal dominant pattern. The affected gene is the glial fibrillary acidic protein or GFAP gene. There are three main subtypes of AL the infantile form with onset between birth and two years of age; a juvenile form with an onset between 2 and 12 years; and an adult form with an onset after 12 years of age. The infantile form is the most common and is usually fatal within the first decade of life. Onset in adulthood is the least common [2].

In 2018, Wilson, Vargha-Khadem and Florschutz published a paper on a woman aged 38 years who had been diagnosed with Alexander's Disease at the age of 5 years. The woman was followed up for a further two years until she died at the age of 40 years. She is believed to be the longest-lived person reported with this condition. The second longest would appear to be a patient from Japan. The authors report the long term survival of a patient with the infantile form of the disease who was still alive at the age of 25 years and 7 months [3].

Case Study

The [4] patient was diagnosed with AL at Great Ormond Street Hospital (GOSH), London, United Kingdo at the age of 5 years. Thus, she had the juvenile form. Genetic testing in July 2016 confirmed that the diagnosis was correct. We believe her to be the longest-lived patient with AL on record. She was assessed neuropsychologically over a number of years. Between the ages of five and 16 years she was assessed at GOSH and found to have mild special educational needs. Between the ages of 35 and 40 years she was assessed once a year. Initially she had some speech, smiled frequently and could write her name. Over the years she steadily declined but remained physically well until she died at the age of 40. Although a more detailed paper about LD was published when she was 38 years old [4], she was monitored for a further two years. The highest ranked score on the Wessex Head Injury Matrix [2] (Table 1).

Table 1: A Summary of Assessments Carried Out Between 2012 and 2017.

A Summary of the Wessex Head Injury Matrix Assessments		
Date	**Highest Score**	**Total Behaviors**
2012	52 (1 or 2 gestures)	37
2013	43 (smiles)	24
2014	43 (smiles)	21
2015	14 (mechanical vocalization)	8
2016	14 (mechanical vocalization)	7
2017	14 (mechanical vocalization)	5

Discussion

Why did this patient live so long? Good physical care probably contributed to her survival. In addition, the juvenile form shows a slower rate of decline than the infant onset form. Perhaps the expected survival of patients diagnosed with the juvenile form of the disease should be revised?

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