



Adults with Dravet syndrome

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Dravet Syndrome in adults

Most children with Dravet Syndrome become adults with Dravet Syndrome

Sounds obvious, but somehow often missed in planning or awareness

The original diagnostic label of 'severe myoclonic epilepsy of infancy' was misleading

Probably less recognition of diagnosis and less resource in adult services

Can lead to difficulties at transition

Limited information on what happens in adulthood, and in general not the same provision as for children

We need to gather more information and consider development of recommendations and guidance

Dravet Syndrome in adults: case history

The oldest person I see: 70 years

Diagnosis made at 59 years of age

Life before and life after the diagnosis

Speech, seizures, admissions, behaviour

Quality of life

Dravet Syndrome in adults: case histories

Other individuals – ages from 18 upwards, seven people over 40

A number of similar results after diagnosis made

Not a formal study, just observation

Mutation type does not explain outcome in this small sample of people

Dravet Syndrome in adults: case histories

All have intellectual disability

Four people had further progression in adulthood

Many people did not, and eight had improvement in speech after diagnosis and medication changes

Eleven people developed swallowing difficulties: five needed dietary changes; six had PEG

Six people experienced motor deterioration

Dravet Syndrome in adults: case histories

Seizures

Types

Timing

Responses to medication

Dravet Syndrome in adults: case histories

New diagnoses still being made in adulthood

‘Myoclonic epilepsy’

‘Epilepsy, hyperactivity, mental retardation’

‘Spastic quadraparesis, cognitive impairment, epilepsy’

‘Generalised tonic-clonic seizures, spastic quadraparesis, severe learning disability’

Dravet Syndrome in adults

We do not number of people with Dravet who are adults

We do not know much about course in adulthood

We do know that improvement in seizure control may be associated with gains, but no formal trial

We know that some people continue to have treatment-resistant seizures

We need to know more