Leukodystrophy with Oligodontia

**Alternative Names**
- Dentoleukoencephalopathy
- Dentoleukoencephalopathy

**WHO International Classification of Diseases**
Diseases of the nervous system

**OMIM Number**
607694

**Mode of Inheritance**
Autosomal recessive

**Description**
Leukodystrophies are rare degenerative disorders that involve mainly the white matter of the brain. It is associated specific clinical features including neurological deterioration following a period of normal development, predominant involvement of the motor function at least initially, and absence of convulsions or myoclonus. Leukodystrophy with oligodontia is characterized by progressive ataxia beginning during infancy, a pyramidal syndrome (spasticity, hyperreflexia), and dental agenesis.

**Molecular Genetics**
The mode of transmission of leukodystrophy with oligodontia is suggested to be autosomal recessive. It has been suggested that the disorder is caused by two independent mutations or a deletion of very closely linked genes. Also, the disorder may be the result of a mutation in a single gene with pleiotropic effects.

**Epidemiology in the Arab World**

**Lebanon**
See also: Syria > Atrouni et al., 2003

**Syria**
Atrouni et al. (2003) studied four Syrian sibs of consanguineous parents with oligodontia, a neurodegenerative disease appearing at around age 12 years with abnormalities of the white matter, and cortical atrophy. The patients were one brother and three sisters. The brother was 22 years old and he had signs of abnormal gait at 13 years which worsened gradually. His speech was severely dysarthric and he had a slight enophthalmia, positive Romberg sign, diffuse spasticity, increased tendinous reflexes, symmetrically positive abdominal reflexes, slightly diminished muscle strength, vibration and position sense at the lower limbs, bilateral foot clonus, and bilateral Babinski sign. He had also few permanent teeth and two abnormal teeth. Chromosomal karyotype was normal (46,XY). Cervico-dorsal MRI revealed diffuse alterations of his medullary white matter. His eldest sister was 17 years old. Her clinical and neurological features considered to be similar to her affected brother, except that she could speak more clearly and had mild intentional tremor of the left hand. All her permanent teeth were absent and her chromosomal karyotype was normal (46,XX). Other two sisters of ages 11 and 12 years were found to have enophthalmia and oligodontia. The 11-year-old sister had brisk tendinous reflexes and no permanent teeth. Neurological examination of the 12-year-old sister revealed an unstable gait, exaggerated tendinous reflexes, slightly diminished vibratory sense at lower limbs, a right Babinski sign, and a mild bilateral intentional tremor. She had few permanent teeth and two abnormal teeth. Brain MRI for the four patients disclosed degeneration of the deep white matter of the centrum semi-ovale in both hemispheres. Cortical brain atrophy was detected in all patients. The parents and other four sibs were normal. Two double cousins aged six and eight years were presumably affected by the same disease as they had oligodontia. Atrouni et al. (2003) suggested
an autosomal recessive inheritance of the disease due to the presence of consanguinity.

References

Contributors
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