Leukodystrophies: clinical and therapeutic aspects

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Résumé en anglais	This chapter reviews that leukodystrophies is a genetic diseases that occur in early childhood. It is a possibility that leads to screening for leukodystrophy patients with early-onset dementia of unknown origin or atypical psychiatric symptoms. The chapter focuses on genetic cases of white matter involvement presenting with deficits in cognitive functions or dementia. Leukodystrophies are rare causes of dementia in the adult. They may cause a dementia of the frontal type or psychiatric symptoms than can mimic schizophrenia but are rapidly associated with long tract involvement. The presence of other cases in the family is a crucial step in the diagnosis but apparently isolated cases are frequent. Homochrony and homotypy are the rule but with many exceptions they have to be considered for genetic counseling. MRI is also critical, showing abnormal increased signals of the white matter on T2-weighted and FLAIR sequences, with a frontal predominance. Normal MRI imaging, at least at the beginning of the disease, does not rule out the diagnosis. The study of the pathogenetic mechanisms of leukodystrophies has been improved by the development and the analysis of animal models. The chapter states that no curative treatment is yet available. New perspectives have opened with the development of cell and gene therapies, even in adult forms, where the demyelination can at least be stabilized.
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