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Cognitive development in children with Prader–Willi syndrome

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Abstract

© 2018 International Strategic Management Association. All rights reserved. Aim: The article is devoted to the specificity of cognitive development in three children with Prader–Willi syndrome (PWS) in the period up to 6-year-old. The PWS is a rare hereditary disease caused by the absence of the father's copy of the 15q11-13 chromosome. Methods: Genomic imprinting is involved into the regulation of the genes in this area of chromosome 15. Diagnostic signs of this syndrome are muscular hypotension, hypogonadism, obesity, excessive adiposity, respiratory complications, mental retardation, small brushes and feet, dysplasia of the hip joints, and stigma of disembryogenesis. Currently, specific ways of treating people with this syndrome have not been developed. It is considered that children with PWS suffer from the retardation of cognitive development; however, there is a lack of scientific information about it. The study of PWS requires an interdisciplinary approach and the detailed description of cognitive development. Results: The results showed that in the period up to 6 years in three children with this rare syndrome the most obvious decline is registered predominantly in the development of impressive speech, and other functions do not suffer significantly in spite of the presence of impairments in motor development. Conclusion: The research was conducted at A. Yu. Ratner Pediatic Clinical Hospital NO. 8 (Kazan, Russia) by the group of clinical linguists, neurologists, and speech therapists.

Keywords

Assessment of cognitive functions, Clinical linguistics, Developmental disorders, Prader–Willi syndrome

References

[1] Šepec SG, Sabolić LL. Prader-Willi syndrome–clinical and endocrinological characteristics and treatment options. Paediatr Croatica 2016;60:153-60.

[2] Bodrova E, Leong DJ, Akhutina TV. When everything new is well-forgotten old: Vygotsky/Luria insights in the development of executive functions. New Dir Child Adolesc Dev 2011;2011:11-28.

[3] Whittington J, Holland A. Cognition in people with prader-willi syndrome: Insights into genetic influences on cognitive and social development. Neurosci Biobehav Rev 2017;72:153-67.

[4] Cimolin V, Cau N, Galli M, Santovito C, Grugni G, Capodaglio P. Gait initiation and termination strategies in patients with Prader-Willi syndrome. J Neuro Eng Rehab 2017;14:44.

[5] Pasiński M. Motor system disturbances in overweight children with genetically determined syndromes. Pediatria Polska 2008;83:549-52.

[6] Tan HL, Urquhart DS. Respiratory complications in children with prader willi syndrome. Paediatr Respir Rev 2017;22:52-9.

[7] Smith A, Hung D. The dilemma of diagnostic testing for Prader-Willi syndrome. Trans Pediatr 2017;6:46-56.

[8] Bakker NE, Lindberg A, Heissler J, Wollmann HA, Camacho-Hübner C, Hokken-Koelega AC, et al. Growth hormone treatment in children with prader-willi syndrome: Three years of longitudinal data in prepubertal children and adult height data from the KIGS database. J Clin Endocrinol Metab 2017;102:1702-11.

[9] Dykens EM, Roof E, Hunt-Hawkins H. Cognitive and adaptive advantages of growth hormone treatment in children with prader-willi syndrome. J Child Psychol Psychiatry 2017;58:64-74.

[10] MacIver NJ. Oxytocin treatment may improve infant feeding and social skills in Prader-Willi syndrome. Pediatrics 2017;139:e20163833.

[11] Miller JL, Tamura R, Butler MG, Kimonis V, Sulsona C, Gold JA, et al. Oxytocin treatment in children with prader-willi syndrome: A double-blind, placebo-controlled, crossover study. Am J Med Genet A 2017;173:1243-50.

[12] Wilson KS, Wiersma LD, Rubin DA. Quality of life in children with prader willi syndrome: Parent and child reports. Res Dev Disabil 2016;57:149-57.

[13] Avrahamy H, Pollak Y, Shriki-Tal L, Genstil L, Hirsch HJ, Gross-Tsur V, et al. Prader-Willi syndrome mental health research strategy workshop proceedings: The state of the science and future directions. Orphanet J Rare Dis 2016;11:1-7.

[14] Dykens EM, Maxwell MA, Pantino E, Kossler R, Roof E. Assessment of hyperphagia in prader-willi syndrome. Obesity (Silver Spring) 2007;15:1816-26.

[15] Eliseeva MB, Vershinina EA, Ryskina VL. The MacArthur Inventories: Russian version. Assessment of Speech and Communication Development in Children of Early Age. The Norms of Development. The Samples of Analysis. Comments. Ivanovo: LISTOS; 2016. p. 76.

[16] Fenson L, Dale PS, Reznick JS, Thal D, Bates E, Hartung JP, et al. The MacArthur Communicative Development Inventories: User's Guide and Technical Manual. Baltimore: Paul H. Brokes Publishing Co.; 1993.

[17] Akhutina TV. The Methods of Neuropsychological Examination of Children from 6 to 9 Year Old. Moscow: V. Sekachev; 2016. p. 280.

[18] Gorobets EA, Kulsharipova RE, Lotfullina NZ. Semantico-phonological disorders in patients with Wernicke's aphasia. Soc Sci 2015;10:566-70.