

# Stan narządu słuchu u dzieci z zespołem Brachmann-de Lange

## Auditory function in children with Brachmann-de Lange syndrom

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### Summary

**The Aim.** The aim of the research work is the evaluation of auditory function in children with rare, genetically determined Brachmann-de Lange syndrome. **Methods.** Test material came from 18 children (7 girls and 11 boys) between 11 months and 18 years of age with Brachmann-de Lange syndrome who have been diagnosed and treated at ENT Department and Department of Paediatrics, Haematology, Oncology and Endocrinology Medical University of Gdansk with support of Cornelia de Lange Association — Poland. In all children examinations of brainstem auditory evoked potential have been carried out as well as tympanometric examination in case of finding hearing loss. All these examinations were carried out in ENT Department of Medical University of Gdansk, using Racia-Alvar Centor C apparatus and Madsen-Zodiak 901. **Results.** 9 (50%) of patients demonstrated hearing loss. In 3 (16.7%) cases the conduction hearing loss was connected with the chronic diseases of middle ear which required medical treatment. In remaining 6 (33.3%) cases due to sensorineural hearing loss children had hearing aids applied and underwent rehabilitation. **Conclusions.** The results indicate that all children with Brachmann-de Lange syndrome should undergo examinations of brainstem auditory evoked potential. It enables to detect hypoacusis and initiate proper treatment. Lack of the opportunity of having a hearing aid applied and further rehabilitation deepens the social isolation of the little patients and inhibits their natural progress in communication. In such cases mental retardation may subsequently occur.