

Absence Epilepsy and Panayiotopoulos Syndrome

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Valorisation

Valorisation addendum

The aim of clinical research is to ultimately improve patient care. Research questions often arise from health care itself. Physicians hope that these patients will benefit the most from new research findings. Nevertheless, clinical research does also benefit others in society. This valorisation addendum will discuss ways to capitalize on the scientific knowledge discussed in this thesis.

Valorisation as defined by the The National Valorisation Committee:

“the process of creating value from knowledge, by making knowledge suitable and/or available for social and/or economic use and by making knowledge suitable for translation into competitive products, services, processes and new activities”

(Waardevol - Indicatoren voor Valorisatie 2011(1))

Relevance

Epilepsy affects about 41-187/100.000 children per year (2). Being diagnosed with epilepsy is accompanied by many uncertainties, such as, seizure reoccurrence, treatment success and possible therapeutic side effects. In addition, psychosocial and socio-economic factors also play a role. For example, fears regarding the unpredictability of seizures, developing a low self-esteem, the need for special education and problems in family adjustment. Furthermore, in some children, the question arises whether possible effects on cognition and/or behavior may impact academic achievement as cognitive deficits, educational underachievement and behavioral problems are more common among some patients with epilepsy (3, 4).

To be able to improve care in patients with epilepsy it is necessary to better understand cognitive abilities and brain development in patients with epilepsy. This thesis focused on two epilepsy syndromes, namely absence epilepsy and Panayiotopoulos syndrome.

Findings

Neurodevelopment and neurocognitive co-morbidities in allegedly benign epilepsy syndromes, such as absence epilepsy and Panayiotopoulos syndrome, are

far less researched, but may have an impact on academic, behavioral and overall development. In this thesis we have shown that both AE and PS show neurocognitive weaknesses in a wide variety of domains, although as a group average performance mostly falls within the average range. In AE, attentional deficits are the most pronounced cognitive weakness, but up to half show an improvement over time. On the other hand, cognitive weaknesses in other cognitive domains seem to persist. A lower myelin content in the frontal lobe of children with AE may suggest differences in the neurodevelopment of children with epilepsy. This thesis, contributes to the knowledge on the neurocognitive aspects of epilepsy, which is helpful to improve counseling and treatment strategies.

Target groups

The results of this thesis are relevant for people involved in the care of children with absence epilepsy or Panayiotopoulos syndrome such as, (child)neurologists, epileptologist, (neuro)psychologists, paediatricians, and general practitioners. Awareness of cognitive problems in children with epilepsy enables health care professionals to watch out for a possible neurocognitive co-morbidity or academic underachievement, so that children can be referred as soon as possible. For neuropsychologists, knowledge on the cognitive functions most likely affected is valuable to be able to perform the necessary neuropsychological tests. Furthermore, this thesis may be relevant to those involved in the academic careers of these children, such as teachers, orthopedagogues or people providing special education for children with epilepsy. In the Netherlands educational counsellors of the LWOE (Landelijk Werkverband Onderwijs en Epilepsy) are involved in the guidance of children with epilepsy and their teachers to optimize school achievements.

Next, our findings are useful for people involved in epilepsy research, such as people writing on consensus articles, such as guidelines for epilepsy syndrome classification. The research described in this thesis contradicts that absence epilepsy and Panayiotopoulos syndrome are benign epilepsy syndromes at least partly due to the neurocognitive co-morbidity as discussed throughout this thesis. It has led to a better understanding of the neurocognitive development of

children with absence epilepsy and Panayiotopoulos syndrome. As discussed in chapter 6, the International League Against Epilepsy (ILAE) has proposed to replace the term benign by “self-limited” and “pharmacoresponsive” (5). However, the impact on cognitive performance and possible consequences on academic achievement should also be formally recognized.

It may be useful for institutions involved in funding scientific research. Additionally, it may help policy makers and health insurances to justify funds for adequate neuropsychological follow-up and academic support programs (such as the LWOE). Parents and patients with absence epilepsy or Panayiotopoulos syndrome may want to know to what extent it may have an impact on cognitive performance. For parents, it is important to not simply dismiss absence epilepsy and Panayiotopoulos as a benign epilepsy type “which they will outgrow”, as this may lead to false expectations in at least a proportion of children.

Knowledge translation

A prerequisite for the implementation of new scientific knowledge is communicating this knowledge to the scientific community and making it available for the public. Therefore, the chapters in this thesis were published in international peer-reviewed journals. Furthermore, the results were discussed during (poster)presentations and at (inter)national symposia (such as a yearly international symposium at epilepsy centre Kempenhaeghe). Moreover, a case presentation and discussion was published in a Dutch journal (*Epilepsie Periodiek voor professionals*). In addition, we have given participating children and parents information on the results of the study.

Implementation and future work

Although the used methods and neuropsychological tests are not routine care for every child with absence epilepsy or Panayiotopoulos syndrome, and a cost-effectiveness study has not been performed, it does provide necessary insight for further development of follow-up protocols. Therefore, our findings can be used

to improve clinical practice and neuropsychological follow-up in these children. Other clinical implications have been discussed in chapter 6.

This thesis may inspire new research and businesses (for example involved in the development of neuropsychological tools) to develop new diagnostic measures and treatments specifically made for children with epilepsy. Myelin imaging may be used as a prognostic tool in the follow-up of children with epilepsy. As myelin imaging is a very novel imaging technique in epilepsy research, our methodology and findings warrant further imaging studies focusing on myelin and epilepsy. Nevertheless, it is still too early to predict the relevance of these findings on a larger scale such as how this will improve the care of children with epilepsy. In the future, more research on myelinisation patterns of children with epilepsy is needed to explore how this relates to neurodevelopmental problems and if this may be a core pathophysiological process to which treatment could be developed. More recommendations for future research are described in chapter 6.

References

1. Van Drooge L, Vandeberg R, Zuijdam F, Mostert B, van der Meulen B, Bruins E. Waardevol-
Indicatoren voor Valorisatie. Rathenau Instituut. 2011.
2. Camfield P, Camfield C. Incidence, prevalence and aetiology of seizures and epilepsy in
children. *Epileptic disorders : international epilepsy journal with videotape*. 2015;17(2):117-23.
3. Berg AT. Epilepsy, cognition, and behavior: The clinical picture. *Epilepsia*. 2011;52 Suppl 1:7-
12.
4. Fastenau PS, Jianzhao S, Dunn DW, Austin JK. Academic underachievement among children
with epilepsy: proportion exceeding psychometric criteria for learning disability and
associated risk factors. *Journal of learning disabilities*. 2008;41(3):195-207.
5. Scheffer IE, Berkovic S, Capovilla G, Connolly MB, French J, Guilhoto L, et al. ILAE
classification of the epilepsies: Position paper of the ILAE Commission for Classification and
Terminology. *Epilepsia*. 2017;58(4):512-21.