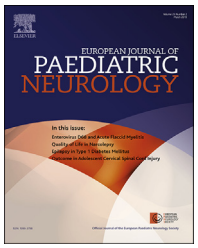
ELSEVIER

Official Journal of the European Paediatric Neurology Society



Editorial

Attention measures of patients with Rett Syndrome need to overcome the challenges in evaluating the oculomotor function using electronystagmography

Rett Syndrome (RTT) is a genetic neurodevelopmental disorder characterized by normal early development followed by loss of purposeful use of the hands and of muscle tone, and acquisition of distinctive hand movements, slowed brain and head growth, gait abnormalities, seizures, and intellectual disability. It almost exclusively affects females. Another symptom, apraxia — the inability to perform motor functions — is perhaps the most severely disabling feature of RTT, interfering with every body movement, including eye gaze and speech.

Due to the severity of symptoms it is very hard to apply common or standardized instruments for the measurement of cognitive and communication abilities in individuals with RTT.[1,2] There are many factors that influence attention measures such as the stereotypic behaviours, epilepsy and seizure frequency.

Many parents report that their child is far more cognitively able than can be demonstrated on assessment or is suggested by current research. This mismatch may be answered, at least in part, by the apraxia which limits the ability to respond easily on command. For this reason, the use of eye tracking technology offers a potentially effective method of communication. Most used eye tracker parameters are: Fixation Length (FL) that is the amount of time spent by the girl in looking at the target, Fixation Count (FC) that the time spent by the girl in looking at the target, Time to First Fixation (TFF) that is the time employed by the girl before looking at the target for the first time.[3,4]

At the current time, although eye gaze and eye tracking technologies appear to offer the most appropriate access to assessments of attention and communication, there is a condition that must be met: to have an intact oculomotor function.

In this issue De Breet et al.[5] rightly focus on this condition and investigated oculomotor function in patients with Rett Syndrome using electronystagmography (ENG). Their aim was to improve oculomotor examination in individuals with RTT by evaluating the challenges encountered during ENG examination in females with RTT when compared to healthy, typically developing children, and to propose possible solutions for obtaining more reliable and reproducible oculomotor results when assessing individuals with RTT. Their results indicated that challenges in examination were mainly related to quality of attention and quality of signals. They suggested some adaptations that include reducing the number of electrodes, changing the picture stimuli and bringing them closer, performing observational assessments rather than ENG, and using virtual reality goggles.

I think that this article addresses an important issue and it can be useful to overcome the main problems that researchers have in working with RTT with Eye tracking technology.

REFERENCES

1. Antonietti A, Fabio RA, Martinazzoli C. Costruzione e standardizzazione dello strumento RARS (rett assessment rating scale). *Life Span and Disabil* 2005;**8**(2).
2. Fabio RA, Magaudda C, Caprì T, Towey GE, Martino G. Choice behavior in rett syndrome: the consistency parameter. *Life Span and Disabil* 2018;**21**(1):47–62.
3. Fabio RA, Billeci L, Crifaci G, Troise E, Tortorella G, Pioggia G. Cognitive training modifies frequency EEG bands and neuropsychological measures in rett syndrome. *Res Dev Disabil* 2016;**53**–**54**:73–85. https://doi.org/10.1016/j.ridd.2016.01.009.
4. Fabio RA, Giannatiempo S, Oliva P, Murdaca AM. The increase of attention in Rett syndrome: a pre-Test/Post-test research design. *J Dev Phys Disabil* 2011;**23**(2):99–111. https://doi.org/10.1007/s10882-010-9207-z.
5. De Breet LHM, Townend GS, Curfs LMG, Kingma H, Smeets EEJ, Lucieer F, Widdershoven J, van de Berg R. Challenges in evaluating the oculomotor function in individuals with rett syndrome using electronystagmography. *Eur J Paediatr Neurol* 2019;**23**(2):262–9. https://doi.org/10.1016/j.ejpn.2018.12.003.

Rosa Angela Fabio
Department of Clinical and Experimental Medicine, University of Messina, Via Bivona, Messina, Italy
E-mail address: rafabio@unime.it

https://doi.org/10.1016/j.ejpn.2019.02.008
1090-3798/© 2019 European Paediatric Neurology Society. Published by Elsevier Ltd. All rights reserved.