[Research paper]

OFFICIAL For Internal Use Only

Guillain-Barre syndrome diagnosis in children

The content of this document is OFFICIAL.

Please note:

The research and literature reviews collated by our TAB Research Team are not to be shared external to the Branch. These are for internal TAB use only and are intended to assist our advisors with their reasonable and necessary decision-making. Delegates have access to a wide variety of comprehensive guidance material. If Delegates require further information on access or planning matters, they are to call the TAPS line for advice. The Research Team are unable to ensure that the information listed below provides an accurate & up-to-date snapshot of these matters

Research questions:

What are the clinical outcomes expected for children diagnosed with Guillain-Barre syndrome? How does the Guillain-Barre disability score compare to the Hughes scale?

Date: 12/5/2022

Requestor: s47F - personal privacy

Endorsed by (EL1 or above):

Researcher: Stephanie P s47F - personal privacy

Cleared by: Stephanie P s47F - personal privacy

1. Contents

2. Summary

This research is a supplement to RES212 Guillain Barre Syndrome (completed 5/8/2021).

Guillain-Barre syndrome is more common in adults than children. While research in children is limited, information sourced suggests that children are likely to have a better clinical outcome after a diagnosis of Guillain-Barre syndrome than adults. Two subtypes of Guillain-Barre syndrome have been discussed in the literature – AMAN and AIDP (defined below). There is some evidence to suggest that diagnosis of subtype AMAN may lead to greater physical impairment at 6- and 12-months post diagnosis, however similar clinical outcomes are reported at 2 years post diagnosis.

The Guillain-Barre Disability Score appears to be a method used by Hughes to grade symptoms prior to the development of the Hughes Functional Grading Scale. These are discussed below.

3. Guillain-Barre Syndrome in children

Literature regarding the prognosis for Guillain-Barre syndrome for Australian children could not be sourced. However, there are international studies exploring the clinical prognosis of children who are diagnosed with Guillain-Barre syndrome.

Guillain-Barre syndrome predominantly affects adults, however diagnosis in the paediatric population is more common in children under 10 years old (Nasiri et al, 2018). More specifically, diagnosis most often involves children aged between 1 and 5 years and males are affected more than females (Nasiri et al, 2018). Research suggests that children have a more favourable clinical course and outcome compared to adults diagnosed with Guillain-Barre syndrome (Argarwal et al, 2022; Nasiri et al, 2018). Argarwal et al (2022) found that age of presentation and gender did not influence the clinical outcome.

Common paediatric presentations have been identified as distal limb weakness, reduced tendon reflex, pain and paraethesia (Lee et al, 2008; Nasiri et al, 2018). Dysarthria, facial palsy, ophthalmoplegia and urinary problems have also been observed (Lee et al, 2008).

Two main subtypes have been described in the literature depending on the location of neural inflammation: AMAN (acute motor axonal neuropathy) and AIDP (acute inflammatory demyelinating polyradiculoneuropathy). The literature indicates that children diagnosed with AMAN may have greater clinical symptoms evident as a worse Hughes score at nadir compared to children diagnosed with AIDP subtype (Argarwal et al, 2022). Additionally, children diagnosed with AMAN may have a slower recovery phase with poorer outcomes at 6- and 12-months post-onset (Lee et al, 2008). However, Lee et al (2008) also cited research from Japan where 35% of the cohort were diagnosed with AIDP and 48% with AMAN and both groups had comparable favourable recoveries at 12 months post-onset.

Overall, the literature suggests that children diagnosed with Guillain-Barre syndrome have good functional status regardless of whether axons or myelin sheaths are affected (Argarwal et al, 2022; Lee et al, 2008). Retrospective research by Nasiri et al (2018) compared international research data of paediatric outcomes with their study population of Iranian children, unfortunately they did not indicate the time period that had elapsed from onset to when the clinical was outcome measured. In their study, 92% of children recovered fully and there was a 1.8% mortality rate – this was identified as being similar to research data from France, Tabriz and India (Nasiri et al, 2018). Further, 5% reported incomplete recovery which was similar to research data Nasiri et al (2018) obtained from India.

Information on the Better Health Channel website (2014) is not specific to children, but it indicates that full recovery from Guillain-Barre syndrome can be 'two years or more'. Better Health Channel advises that physical therapy is important to facilitate recovery and prevent muscle contractures and associated deformities.

4. Guillain-Barre Disability Score

A 2014 study by Fokke et al suggests the Guillain-Barre syndrome disability score, as described in research by Hughes et al (1978), is a widely accepted scale of disability for individuals with Guillain-Barre syndrome. The research by Hughes et al (1978) uses the score for neurological assessment of enduring symptoms of Guillain-Barre syndrome 1 year after diagnosis. The 'Guillain-Barre Disability Score' is a 6-point rating scale as follows (Hughes et al, 1978):

0 = healthy

1 = minor signs or symptoms of neuropathy but capable of manual work

2 = able to walk without support of a stick but incapable of manual work

3 = able to walk with a stick, appliance, or support

4 = confined to bed or chairbound

5 = requiring assisted ventilation

6 = dead

A reference to the Guillain-Barre Disability Scale was found in van Koningsveld et al (2007) who aimed to develop a clinical scoring system for clinical prognosis and used the Guillain-Barre Disability score in their modelling. These researchers defined a 'poor outcome' as a score of 3 or more at 6 months which corresponded to the inability to walk 10m independently. A 'fairly good outcome' was considered a score of 2 or less at 6 months. Data was taken at 6 months as this was considered the time period where most of the recovery process would have occurred (van Koningsveld et al, 2007).

Validity and reliability data for the Guillain-Barre Disability Score could not be sourced.

5. Hughes Functional Grading Scale

The Hughes Functional Grading Scale provides a measure of disability and is used to rate clinical performance (Shi et al, 2019). The scale is a 6-point rating scale and appears to be based on the Guillain-Barre Disability Score but has more specific criteria for ratings 2 and 3:

0 = normal

1 = slight clinical symptoms

2 = able to walk 5m or more without assistance but unable to run

3 = able to walk 5m with help

4 = bedridden or chairbound

5 = ventilator-assisted breathing

6 = death

6. References

Agarwal, E., Bhagat, A., Srivastava, K., Thakore, B., Jagtap, S., Kalane, U., & Rajadhyaksha, S. (2022). Clinical and electrophysiological factors predicting prolonged recovery in children with Guillain-Barre syndrome. *Indian journal of Pediatrics, 89*(5), 452-458. https://doi.org/10.1007/s12098-021-03804-7

Better Health Channel. (2014). Guillain-Barre syndrome [website]. Accessed from https://www.betterhealth.vic.gov.au/health/conditionsandtreatments/guillain-barre-syndrome

Fokke, C., van den Berg, B., Drenthen, J., Walgaard, C., van Doorn, P. A., & Jacobs, B. C. (2014). Diagnosis of Guillain-Barre syndrome and validation of Brighton Criteria. *Brain, 173*(1), 33-43. Accessed from https://www.medscape.com/viewarticle/819343_2#:~:text=Clinical%20course%20was%20described%20by%20using%20the%20Guillain-Barr%C3%A9,%28normal%29%20to%206%20%28death%29%20%28Hughes%20et%20al.%2C%201978%29.

Hughes, R. A. C., Newsom-Davis, J. M., Perkin, G. D., & Pierce, J. M. (1978). Controlled trial of prednisolone in acute polyneuropathy. *The Lancet, 312*, 750-753. Accessed from https://www-sciencedirect-com.ezproxy-b.deakin.edu.au/science/article/pii/S0140673678926442/pdfft?md5=6c43282de955f404e8ffa7bb6ef3bdbb&pid=1-s2.0-S0140673678926442-main.pdf

Lee, J. H., Sung, I. Y., & Rew, I. S. (2008). Clinical presentation and prognosis of childhood guillain-Barre syndrome. *Journal of Paediatrics and Child Health, 44*, 449-454. doi:10.1111/j.1440-1754.2008.01325.x

Nasiri, J., Ghazavi, M., Yaghini, O., & Chaldavi, M. (2018). Clinical Features and Outcome of Guillain-Barré Syndrome in Children. *Iran J Child Neurol, 12*(2):49-57.

Shi, M., Zhu, J., & Deng, H. (2019). Clinical characteristics of intravenous injection of monosialotetrahexosyl ganglioside sodium-related Guillain-Barre syndrome. *Frontiers in Neurology, 10*(225). https://doi.org/10.3389/fneur.2019.00225

van Koningsveld, R., Steyerberg, E. W., Hughes, R. A. C., Swan, A. V., van Doorn, P. A., & Jacobs, B. C. (2007). A clinical prognostic scoring system for Guillain-Barre syndrome. *Lancet Neurology, 6*, 589-94. DOI:10.1016/S1474- 4422(07)70130-8

7. Version control

Version	Amended by	Brief Description of Change	Status	Date
0.1				

Version	Amended by	Brief Description of Change	Status	Date
1.0	47F - personal privacy	Document creation	Cleared	15/5/2022