Pediatric Onset Chronic Inflammatory Demyelinating Polyneuropathy: A Multicenter Study from Türkiye

Gamze Sarıkaya Uzan [1], Deniz Yüksel [2], Erhan Aksoy [2], Ülkühan Öztoprak [2], Mehmet Canpolat [3], Selcan Öztürk [3], Çelebi Yıldırım [3], Ayten Güleç [3], Hüseyin Per [3], Hakan Gümüş [3], Çetin Okuyaz[4], Meltem Çobanoğulları Direk[4], Mustafa Kömür[4], Aycan Ünalp[5], Ünsal Yılmaz[5], Ömer Bektaş[6], Serap Teber[6], Nargiz Aliyeva[7], Nihal Olgaç Dündar[8], Pınar Gençpınar[8], Esra Gürkaş[9], Sanem Keskin Yılmaz[10], Seda Kanmaz[10], Hasan Tekgül[10], Ayşe Aksoy[11], Gökçen Öz Tuncer[11], Elif Acar Arslan[12], Ayşe Tosun[13], Müge Ayanoğlu[13], Muhittin Bodur[14], Bülent Ünay[15], Semra Hız Kurul[1], Uluç Yiş[1]

1:Division of Child Neurology, Department of Pediatrics, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey 2:Department of Pediatric Neurology, Faculty of Medicine, Dr Sami Ulus Maternity Child Health and Diseases Training and Research Hospital, University of Turkey 3:Division of Child Neurology, Department of Pediatrics, Erciyes University Faculty of Medicine, Kayseri, Turkey 4:Division of Child Neurology, Department of Pediatrics, Mersin University Faculty of Medicine, Mersin, Turkey 5:Division of Pediatric Neurology, Department of Pediatrics, University of Health Sciences Dr. Behçet Uz Child Disease and Pediatric Surgery Training an Turkey 6:Division of Child Neurology, Department of Pediatrics, Ankara University Faculty of Medicine, Ankara, Turkey 7:Division of Pediatric Neurology, Department of Pediatrics, University of Health Sciences Tepecik Research and Training Hospital, Izmir, Turkey 8:Division of Child Neurology, Department of Pediatrics, İzmir Katip Çelebi University Faculty of Medicine, İzmir, Turkey 9:Department of Pediatric Neurology, Children's Hospital, Ankara City Hospital, Ankara, Turkey 10:Division of Child Neurology, Department of Pediatrics, Ege University Faculty of Medicine, İzmir 11:Division of Child Neurology, Department of Pediatrics, On Dokuz Mayıs University Faculty of Medicine, Samsun, Turkey 12:Division of Child Neurology, Department of Pediatrics, Karadeniz Technical University Faculty of Medicine, Trabzon, Turkey 13:Division of Child Neurology, Department of Pediatrics, Adnan Menderes University Faculty of Medicine, Aydın, Turkey 14:Division of Child Neurology, Department of Pediatrics, Uludağ University Faculty of Medicine, Bursa, Turkey 15:University of Health Sciences Turkey, Gülhane Faculty of Medicine, Department of Pediatric Neurology, Ankara, Turkey

INTRODUCTION and OBJECTIVE

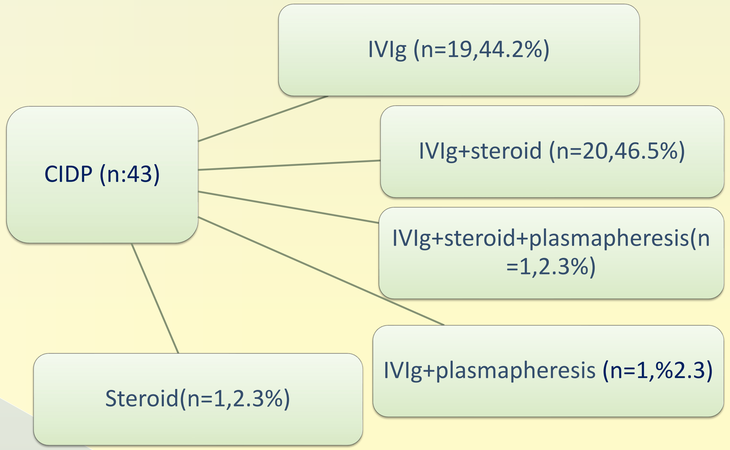
Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare immune-mediated neuropathy in children (1). Children typically present with proximal and distal upper and lower extremity weakness, sensory loss and/or paresthesias, and depressed tendon reflexes (2). We aimed to evaluate the clinical, demographic, electrophysiological and treatment response of pediatric onset chronic inflammatory demyelinating polyneuropathy (CIDP) in Türkiye.

METHODS

The clinical data and electrophysiological results of patients with CIDP between January 2010-December 2021 from 14 centers were reviewed retrospectively. The patients were classified according to Joint Task Force of the European Federation Neurological of Societies and the Peripheral Nerve Society Guideline on management of CIDP (2021).

RESULTS

A total of 43 patients, 22 (%51.2) male and 21 (%48.8) female, were included in the study. The median age was 14.5 years (4.3-23) and the median age of onset of symptoms was 11 years (3 month-17 y). Thirty-eight patients were diagnosed with typical CIDP (88.4%) and 5 patients (11.6%) with possible CIDP. The most common electrophysiological finding of demyelination was ≥30 percent reduction of motor conduction velocity below the lower limit in two nerves (78.1%n= 25). All patients except one received immunotherapy either alone or in combination. There was a significant difference between pre and post-treatment Modified Rankin Scales (MRS) ($p<0.05$) of all patients. First-line treatments were IVIg (n=19,44.2%), IVIg+steroid (n=20,46.5%), steroid(n=1,2.3%), IVIg+steroid+plasmapheresis(n=1,2.3%) and IVIg+plasmapheresis (n=1,%2.3). Second-line treatment was given to 7 (16.6%) patients (n=3,azathioprine; n=2,rituximab; n=1,azathioprine+rituximab; n=1, azathioprine+micofenolate mofetil+methotrexate). The patients were separated into two different groups according to first-line treatment modalities (group-1:only IVIg, group-2:IVIg+steroid). There was no difference between pre and post treatment MRS of both groups.



CONCLUSIONS

This multicenter study showed that first-line immunotherapy modalities (IVIg versus IVIg+steroid) had equal efficacy for treatment CIDP patients in childhood.

Keywords: Chronic Inflammatory Demyelinating Polyneuropathy, immunotherapy, IVIg, steroid

REFERENCES

1.McLeod JG, Pollard JD, Macaskill P, Mohamed A, Spring P, Khurana V. Prevalence of chronic inflammatory demyelinating polyneuropathy in New South Wales, Australia. *Ann Neurol*. 1999; **46**(6): 910- 913.

2. Connolly AM. Chronic inflammatory demyelinating polyneuropathy in childhood. *Pediatr Neurol*. 2001; **24**(3): 177- 182.

Contact: gamzeuzan36@gmail.com