

Table S1. Second survey.

Main Topic	Background Information	Proposed consensus statement	
Firstline Therapy	<p>ADS</p> <p>The survey data shows a homogeneous approach towards all the ADS entities, using 20-30 mg/kg/die (max 1g) i.v. MP for 5 days.</p> <p>This is in line with the international research community. [1-4]</p>	<p><i>The first line treatment of an acute demyelinating syndrome ADS consists of methylprednisolone i.v. 20-30mg/kg/die for 5 days (max 1000mg/die) *</i></p> <p><i>*if restitutio ad integrum after 3 days in selected cases: consider stopping the first-line treatment after 3 days</i></p>	<input type="checkbox"/> Agree
			<input type="checkbox"/> Disagree
Use of steroid taper	<p>Should steroids in ADS be tapered off? There is no adult or pediatric data supporting the benefit of an oral prednisone taper in ON, RRMS, TM. In ADEM the pediatric data supporting an oral prednisone taper is weak. In NMO-SD, oral prednisone is used as maintenance therapy during the induction phase of a chronic immunosuppression and therefore not considered as a classical taper. We propose the following approach:</p>		
ON	<p>Looking at the survey, 50% use an oral prednisolone taper. The ONTT study of 1992 by Beck RW showed no benefit regarding the final visual outcome in those patients tapered off with steroids [5].</p> <p>Therefore, we propose the following consensus statement for ON not associated with NMO-SD.</p>	<p><i>Pediatric ON (not associated with NMO-SD) does not require an oral prednisone taper in case of (near) complete remission of symptoms at the end of 5 days of iv MP therapy. In case of persisting symptoms, consider second line treatment.</i></p>	<input type="checkbox"/> Agree
			<input type="checkbox"/> Disagree
TM	<p>The survey shows a trend towards an oral prednisone taper, as 50% agree to use one. The other half does not use an oral prednisone taper or decides according to the response of iv steroids. Literature does not show any evidence for or against the oral prednisone taper in pediatric TM. Therefore, we propose the following consensus statement for TM not associated with NMO-SD or SLE.</p>	<p><i>Pediatric TM (not associated with SLE or NMO-SD) does not require an oral prednisone taper in case of (near) complete remission of symptoms at the end of 5 days iv MP therapy. If the patient is still symptomatic after 5 days of iv MP treatment, consider second line treatment.</i></p>	<input type="checkbox"/> Agree
			<input type="checkbox"/> Disagree
ADEM	<p>Looking at the survey, 50% use an oral prednisolone taper. The literature suggests the use of an oral prednisone taper starting at 1mg/kg/die (up to 60mg/die) and then tapered off over 4-6 weeks[3,4,6,7], however the data is not of high level evidence. There are some hints towards an increased risk of relapses, if the steroid treatment lasted <3 weeks, however, the finding was not statistically significant [4].</p>	<p><i>In case of ADEM, we suggest using an oral prednisone taper over 4-6 weeks after high-dose iv MP treatment until more evidence becomes available.</i></p>	<input type="checkbox"/> Agree
			<input type="checkbox"/> Disagree
RRMS	<p>Looking at the survey and the pediatric MS literature, there is no consistency in using a prednisone taper in RRMS. Research in adult RRMS does not recommend an oral prednisone taper.[8]</p> <p>The EDSS is helpful to evaluate the response to i.v. MP. In cases of a persistent EDSS > 2 two weeks after completion of the first course of iv MP, a second course of iv MP can be considered.</p>	<p><i>Pediatric RRMS-attacks do not require an oral prednisone taper in case of (near) complete remission of symptoms at the end of 5 days iv MP therapy. In case of persisting symptoms (EDSS ≥2 after two weeks), consider second line treatment.</i></p>	<input type="checkbox"/> Agree
			<input type="checkbox"/> Disagree
NMO-SD	<p>ADS in the context of NMO-SD require special attention given the high risk for severe disabilities following a relapse. Looking at the survey, the majority uses an oral prednisone taper in ADS due to NMO-SD. Literature on treatment for adult NMO-SD attacks suggest an oral prednisone maintenance therapy for up to 2-6 months [9,10], especially to give time to establish a long-term immunosuppressive treatment.</p>	<p><i>ADS occurring in the context of NMO-SD should be treated with prednisone maintenance therapy of 1mg/kg/die and can be tapered off after long-term immunosuppression is established (2-6 months).</i></p>	<input type="checkbox"/> Agree
			<input type="checkbox"/> Disagree
Second line Therapy	<p>In some cases, the first-line treatment is not sufficient, and the use of second line treatment is required. The options vary between repeating the first line treatment (iv MP), administrating IVIG or make use of plasma exchange.</p> <p>If the single ADS occurs in the context of an established chronic demyelinating disease (i.e. RRMS, NMO-SD), the statement regarding the underlying chronic disease applies!</p>		
	<p>ON</p> <p>RRMS</p>	<p>Looking at the survey, 50% would repeat the first line treatment in cases of ON or ADS in the context of an established diagnosis of RRMS. This is</p> <p><i>In case of insufficient response in an acute optic neuritis or RRMS, repeat the first line treatment with iv MP. If</i></p>	<input type="checkbox"/> Agree
			<input type="checkbox"/> Disagree

	consistent with the literature for pediatric and adult ON/RRMS [1,2,11,12]	<i>further treatment is required, we suggest the use of plasma exchange.</i>	
ADEM	Most patients do not need a second line treatment. If the response to iv MP is poor, the literature suggests plasma exchange in fulminant ADEM (+encephalopathy and/or severe focal neurological deficit requiring ICU management) or IVIG (1-2g/kg over 3-5 days) for the remainder. [4] No recent studies have compared the escalation therapy with IVIG vs the use of plasma exchange. [6]	<i>In case of poor response to iv MP, we suggest using plasma exchange in fulminant forms of ADEM and IVIG in the remainder.</i>	<input type="checkbox"/> Agree <input type="checkbox"/> Disagree
TM NMO-SD	In case of poor response to iv MP, data from the survey as well as the literature recommend the use of plasma exchange [13]. Of note, in cases of severe ADS in the context of an established NMO-SD, early administration of plasma exchange should be considered as adjunctive therapy (limited retrospective data) [10,14]	<i>In case of poor response to iv MP for TM or ADS in NMO-SD, we suggest to use plasma exchange.</i>	<input type="checkbox"/> Agree <input type="checkbox"/> Disagree

List of abbreviations: ADS: Acquired demyelinating syndrome; MP: methylprednisolone; RRMS: relapsing remitting multiple sclerosis; NMO-SD: Neuromyelitis optica spectrum disorders; TM: transverse myelitis; ON: optic neuritis; ADEM: acute disseminated encephalomyelitis; SLE: systemic lupus erythematoses.

References

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