

Protocole National de Diagnostic et de Soins (PNDS)

Les maladies du spectre de la neuromyéélite optique

Argumentaire

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Centre de Référence Maladies Inflammatoires Rares du
Cerveau Et de la Moelle



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Cet argumentaire a été élaboré par le centre de référence des Maladies Inflammatoires Rares du Cerveau Et de la Moelle (MIRCEM). Il a servi de base à l'élaboration du PNDS des maladies du spectre de la neuromyéélite optique. Le PNDS est téléchargeable sur le site du centre de référence des MIRCEM : www.mircem.fr

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Liste des abréviations

AQP4	Anticorps anti-aquaporine 4
BAFF	B-cell Activating Factor
LCR	Liquide Céphalo-Rachidien
NMOSD	Maladies du spectre de la neuromyéélite optique
NO	Névrite Optique
PNDP	Protocole National de Diagnostic et de Soins
SEP	Sclérose En Plaques

Préambule

Le PNDS sur les maladies du spectre de la neuromyéélite optique a été élaboré selon la « Méthode d'élaboration d'un protocole national de diagnostic et de soins pour les maladies rares » publiée par la Haute Autorité de Santé en 2012 (guide méthodologique disponible sur le site de la HAS : www.has-sante.fr). Le présent argumentaire est présenté sous forme de thématique.

Ce PNDS a été rédigé après une analyse approfondie et une synthèse critique de la littérature nationale et internationale par un groupe de travail pluridisciplinaire. Le document a été soumis à un groupe de relecture pluri-professionnel. Les remarques et suggestions issues des relectures ont été intégrées, discutées et validées par le groupe de rédaction permettant d'aboutir au document final. Le PNDS présenté est le résultat de ce travail collégial.

Argumentaire

Thème	Principales sources	Commentaires
Synthèse à destination du médecin traitant		
	<ul style="list-style-type: none"> • Wingerchuk DM, Lennon VA, Lucchinetti CF, Pittock SJ, Weinshenker BG. The spectrum of neuromyelitis optica. Lancet Neurol. 2007 Sep;6(9):805-15. • Wingerchuk DM, Banwell B, Bennett JL, Cabre P, Carroll W, Chitnis T, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. Neurology. 2015 Jul 14;85(2):177- 89. 	- Les critères internationaux de diagnostic des NMOSD de 2007 et 2015.
1. Introduction		
2. Objectifs du protocole national de diagnostic et de soins		
3. Diagnostic et évaluation initiale		
3.1 Objectifs		
3.2 Professionnels impliqués (et modalités de coordination)		
3.3 Circonstances de découverte / Suspicion du diagnostic		
L'atteinte visuelle : Névrite optique	<ul style="list-style-type: none"> • Wingerchuk DM, Banwell B, Bennett JL, Cabre P, Carroll W, Chitnis T, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. Neurology. 2015 Jul 14;85(2):177- 89. • Toanen V, Vignal-Clermont C. Neuropathies optiques inflammatoires. //www.em-premium.com/data/traites/op/21-71372/ [Internet]. 2016 Nov 23 [cited 2020 Jul 19]; Available from: https://www-em-premium- 	<ul style="list-style-type: none"> - Description des caractéristiques de l'examen clinique de NO dans les NMOSD. - Description de la spécificité de la poussée de NO de NMOSD par rapport aux autres causes de NMOSD. - Description de l'aspect clinique de NO au stade aigu. - Description du risque de récurrence de NO en fonction de la séropositivité des anti-AQP4.

	<p>com.sirius.parisdescartes.fr/article/1095381.</p> <ul style="list-style-type: none"> • Matiello M, Lennon VA, Jacob A, Pittock SJ, Lucchinetti CF, Wingerchuk DM, et al. NMO-IgG predicts the outcome of recurrent optic neuritis. <i>Neurology</i>. 2008 Jun 3;70(23):2197–200. • Wingerchuk DM, Hogancamp WF, O'Brien PC, Weinshenker BG. The clinical course of neuromyelitis optica (Devic's syndrome). <i>Neurology</i>. 1999 Sep 22;53(5):1107–14. • Zhou H, Zhao S, Yin D, Chen X, Xu Q, Chen T, et al. Optic neuritis: a 5-year follow-up study of Chinese patients based on aquaporin-4 antibody status and ages. <i>J Neurol</i>. 2016 Jul 1;263(7):1382–9. • Kang H, Chen T, Li H, Xu Q, Cao S, Wei S. Prognostic factors and disease course in aquaporin-4 antibody-positive Chinese patients with acute optic neuritis. <i>J Neurol</i>. 2017 Oct;264(10):2130–40. • Van Nispen RM, Virgili G, Hoeben M, Langelaan M, Klevering J, Keunen JE, et al. Low vision rehabilitation for better quality of life in visually impaired adults. <i>Cochrane Database Syst Rev</i>. 2020 27;1. • Paolilo RB, Hacoen Y, Yazbeck E, Armangue T, Bruijstens A, Lechner C, Apostolos-Pereira SL, Martynenko Y, Breu M, de Medeiros Rimkus C, Wassmer E, Baumann M, Papetti L, Capobianco M, Kornek B, Rostásy K, da Paz JA, Ciccarelli O, Lim M, Saiz A, Neuteboom R, Marignier R, Hemingway C, Sato DK, Deiva K. Treatment and outcome of aquaporin-4 antibody-positive NMOSD. <i>Neurol Neuroimmunol Neuroinflamm</i>. 	
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	2020 Jul 30;7(5):e837.	
L'atteinte médullaire : myélite	<ul style="list-style-type: none"> • Collongues N, Papeix C, Zéphir H, Audoin B, Cotton F, Durand-Dubief F, et al. Nosology and etiologies of acute longitudinally extensive transverse myelitis. Rev Neurol (Paris). janv 2014;170(1):6- 12. • Kim S-M, Go MJ, Sung J-J, Park KS, Lee K-W. Painful tonic spasm in neuromyelitis optica: incidence, diagnostic utility, and clinical characteristics. Arch Neurol. août 2012;69(8):1026- 31. • Bradl M, Kanamori Y, Nakashima I, Misu T, Fujihara K, Lassmann H, et al. Pain in neuromyelitis optica--prevalence, pathogenesis and therapy. Nat Rev Neurol. sept 2014;10(9):529- 36. • Xiao L, Qiu W, Lu Z, Li R, Hu X. Intractable pruritus in neuromyelitis optica. Neurol Sci Off J Ital Neurol Soc Ital Soc Clin Neurophysiol. juin 2016;37(6):949- 54. 	<ul style="list-style-type: none"> - Description du tableau clinique de l'atteinte médullaire. - Description du tableau clinique de l'atteinte motrice et son impact sur la qualité de vie. - Description du tableau clinique de l'atteinte sensitive de la myélite.
Syndrome de l'area postrema	<ul style="list-style-type: none"> • Paolilo RB, Hacoheh Y, Yazbeck E, Armangue T, Bruijstens A, Lechner C, Apostolos-Pereira SL, Martynenko Y, Breu M, de Medeiros Rimkus C, Wassmer E, Baumann M, Papetti L, Capobianco M, Kornek B, Rostásy K, da Paz JA, Ciccarelli O, Lim M, Saiz A, Neuteboom R, Marignier R, Hemingway C, Sato DK, Deiva K. Treatment and outcome of aquaporin-4 antibody-positive NMOSD. Neurol Neuroimmunol Neuroinflamm. 2020 Jul 30;7(5):e837. • Shosha E, Dubey D, Palace J, Nakashima I, Jacob A, Fujihara K, et al. Area postrema syndrome: Frequency, criteria, and severity in AQP4-IgG- positive NMOSD. Neurology. 23 2018;91(17):e1642- 51. 	<ul style="list-style-type: none"> - Description des symptômes de l'atteinte de l'<i>area postrema</i> et leurs importances afin de déterminer le diagnostic de NMOSD. - Description des risques d'extension de l'atteinte de l'<i>area postrema</i> sur la région bulbaire et la moelle cervicale. - Description de l'atteinte de l'<i>area postrema</i> chez l'enfant.

	<ul style="list-style-type: none"> • Dubey D, Pittock SJ, Krecke KN, Flanagan EP. Association of Extension of Cervical Cord Lesion and Area Postrema Syndrome With Neuromyelitis Optica Spectrum Disorder. <i>JAMA Neurol.</i> 01 2017;74(3):359- 61. 	
Autres signes cliniques : Atteintes fréquentes ou classiques du NMOSD	<ul style="list-style-type: none"> • Shinoda K, Matsushita T, Furuta K, Isobe N, Yonekawa T, Ohyagi Y, et al. Wall-eyed bilateral internuclear ophthalmoplegia (WEBINO) syndrome in a patient with neuromyelitis optica spectrum disorder and anti-aquaporin-4 antibody. <i>Mult Scler.</i> 2011 Jul;17(7):885–7. • Kremer L, Mealy M, Jacob A, Nakashima I, Cabre P, Bigi S, et al. Brainstem manifestations in neuromyelitis optica: a multicenter study of 258 patients. <i>Mult Scler.</i> 2014;20(7):843–7. • Beigneux Y, Arnulf I, Guillaume-Jugnot P, Leu-Semenescu S, Maillart E, Lubetzki C, Benveniste O, Papeix C. Secondary Hypersomnia as an Initial Manifestation of Neuromyelitis Optica Spectrum Disorders. <i>Multiple sclerosis and related disorders.</i> 2020 Feb;38 :101869. • Baba T, Nakashima I, Kanbayashi T, Konno M, Takahashi T, Fujihara K, et al. Narcolepsy as an initial manifestation of neuromyelitis optica with anti-aquaporin-4 antibody. <i>J Neurol.</i> 2009 Feb;256(2):287–8. • Poppe AY, Lapierre Y, Melançon D, Lowden D, Wardell L, Fullerton LM, et al. Neuromyelitis optica with hypothalamic involvement. <i>Mult Scler.</i> 2005 Oct;11(5):617–21. • Pu S, Long Y, Yang N, He Y, Shan F, Fan Y, et al. Syndrome of inappropriate antidiuretic hormone 	Description des symptômes classiques des atteintes aiguës du tronc cérébral et du diencephale.

	secretion in patients with aquaporin-4 antibody. J Neurol. 2015 Jan;262(1):101–7.	
Autres signes cliniques : Atteintes rares de NMOSD	<ul style="list-style-type: none"> • Jarius S, Lauda F, Wildemann B, Tumani H. Steroid-responsive hearing impairment in NMO-IgG/aquaporin-4-antibody-positive neuromyelitis optica. J Neurol. 2013 Feb;260(2):663–4. • Hage R, Merle H, Jeannin S, Cabre P. Ocular oscillations in the neuromyelitis optica spectrum. J Neuroophthalmol. 2011 Sep;31(3):255–9. • Takai Y, Misu T, Nakashima I, Takahashi T, Itoyama Y, Fujihara K, et al. Two cases of lumbosacral myeloradiculitis with anti-aquaporin-4 antibody. Neurology. 2012 Oct 23;79(17):1826–8. • Eichel R, Meiner Z, Abramsky O, Gotkine M. Acute disseminating encephalomyelitis in neuromyelitis optica: closing the floodgates. Arch Neurol. 2008 Feb;65(2):267–71. • Magaña SM, Matiello M, Pittock SJ, McKeon A, Lennon VA, Rabinstein AA, et al. Posterior reversible encephalopathy syndrome in neuromyelitis optica spectrum disorders. Neurology. 2009 Feb 24;72(8):712–7. • Clardy SL, Lucchinetti CF, Krecke KN, Lennon VA, O'Toole O, Weinshenker BG, et al. Hydrocephalus in neuromyelitis optica. Neurology. 2014 May 20;82(20):1841–3. • Close LN, Zanaty M, Kirby P, Dlouhy BJ. Acute Hydrocephalus Resulting from Neuromyelitis Optica: A Case Report and Review of the Literature. World Neurosurg. 2019 Sep;129:367–71. • Suzuki N, Takahashi T, Aoki M, Misu T, 	Description des autres symptômes rares de NMOSD.

	<p>Konohana S, Okumura T, et al. Neuromyelitis optica preceded by hyperCKemia episode. <i>Neurology</i>. 2010 May 11;74(19):1543–5.</p> <ul style="list-style-type: none"> • Sun H, Ma X, Sun X, Wu L, Huang D. Is transient hyperCKemia a new feature of neuromyelitis optica spectrum disorders? A retrospective study in 439 patients. <i>J Neuroimmunol</i>. 2020 Jun 15;343:577228. 	
Autres signes cliniques : Troubles cognitifs	<ul style="list-style-type: none"> • Oertel FC, Schließeit J, Brandt AU, Paul F. Cognitive Impairment in Neuromyelitis Optica Spectrum Disorders: A Review of Clinical and Neuroradiological Features. <i>Front Neurol</i>. 2019;10:608. • Blanc F, Noblet V, Jung B, Rousseau F, Renard F, Bourre B, et al. White matter atrophy and cognitive dysfunctions in neuromyelitis optica. <i>PLoS ONE</i>. 2012;7(4):e33878. • Eizaguirre MB, Alonso R, Vanotti S, Garcea O. Cognitive impairment in neuromyelitis optica spectrum disorders: What do we know? <i>Mult Scler Relat Disord</i>. 2017 Nov;18:225–9. • Dujardin K, Sockeel P, Cabaret M, De Sèze J, Vermersch P. [BCcogSEP: a French test battery evaluating cognitive functions in multiple sclerosis]. <i>Rev Neurol (Paris)</i>. 2004 Jan;160(1):51–62. 	Description des symptômes des troubles cognitifs, décrits dans les études cliniques récentes.
3.4 Confirmation du diagnostic		
Les anticorps anti-aquaporin 4 (AQP4)	<ul style="list-style-type: none"> • Wingerchuk DM, Banwell B, Bennett JL, Cabre P, Carroll W, Chitnis T, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. <i>Neurology</i>. 2015 Jul 14;85(2):177- 89. 	Description du rôle important joué par les anticorps AQP4 dans le cadre de l'établissement du diagnostic de NMOSD.

	<ul style="list-style-type: none"> • Lennon VA , Kryzer TJ, Pittock SJ, Verkman AS, Hinson SR. IgG marker of optic-spinal multiple sclerosis binds to the aquaporin-4 water channel. J Exp Med. 2005 Aug 15;202(4):473-7. • Jarius S , Wildemann B. Aquaporin-4 antibodies (NMO-IgG) as a serological marker of neuromyelitis optica: a critical review of the literature. Brain Pathol. 2013 Nov;23(6):661-83. 	
L'imagerie	Wingerchuk DM, Lennon VA, Lucchinetti CF, Pittock SJ, Weinshenker BG. The spectrum of neuromyelitis optica. Lancet Neurol. 2007 Sep;6(9):805-15.	Description de l'imagerie dans les NMOSD.
L'étude du liquide céphalo-rachidien	<ul style="list-style-type: none"> • Wingerchuk DM, Lennon VA, Lucchinetti CF, Pittock SJ, Weinshenker BG. The spectrum of neuromyelitis optica. Lancet Neurol. 2007 Sep;6(9):805-15. • Sellner J, Boggild M, Clanet M, Hintzen RQ, Illes Z, Montalban X, Du Pasquier RA, C H Polman CH. EFNS guidelines on diagnosis and management of neuromyelitis optica. Eur J Neurol. 2010 Aug;17(8):1019-32. 	Description des caractéristiques de référence du LCR dans les NMOSD.
3.5 Les diagnostics différentiels		
	<ul style="list-style-type: none"> • Pache F, Zimmermann H, Mikolajczak J, Schumacher S, Lacheta A, Oertel FC, et al. MOG-IgG in NMO and related disorders: a multicenter study of 50 patients. Part 4: Afferent visual system damage after optic neuritis in MOG-IgG-seropositive versus AQP4-IgG-seropositive patients. J Neuroinflammation. 2016 01;13(1):282. • Ramanathan S, Prelog K, Barnes EH, Tantsis EM, Reddel SW, Henderson APD, et al. Radiological differentiation of optic neuritis with 	Les caractéristiques de la NO ischémique et la NO associée aux anticorps anti-MOG permettant de différencier le diagnostic de NMOSD.

	<p>myelin oligodendrocyte glycoprotein antibodies, aquaporin-4 antibodies, and multiple sclerosis. <i>Mult Scler.</i> 2016 Apr;22(4):470–82.</p> <ul style="list-style-type: none"> • Tournaire-Marques E. Neuropathies optiques ischémiques. //www.em-premium.com/data/traites/op/21-88803/ [Internet]. 2019 Apr 19 [cited 2020 Jul 19]; Available from: https://www-em-premium-com.sirius.parisdescartes.fr/article/1288254. 	
3.6 Recherche de comorbidités		
	<ul style="list-style-type: none"> • Pittock SJ, Lennon VA, De Seze J, Vermersch P, Homburger HA, Wingerchuk DM, Lucchinetti CF, Zéphir H, Moder K, Weinschenker BG. Neuromyelitis optica and non organ-specific autoimmunity. <i>Arch Neurol.</i> 2008 Jan;65(1):78-83. • Shahmohammadi S, Doosti R, Shahmohammadi A, Mohammadianinejad SE, Sahraian MA, et al. Autoimmune diseases associated with Neuromyelitis Optica Spectrum Disorders: A literature review. <i>Mult Scler Relat Disord.</i> 2019 Jan;27:350-363. • Iyer A, Elson L, Appleton R, Jacob A. A review of the current literature and a guide to the early diagnosis of autoimmune disorders associated with neuromyelitis optica. <i>Autoimmunity.</i> 2014 May;47(3):154-61. 	Description de l'association de NMOSD aux autres pathologies auto-immunes.
3.7 Recherche de contre-indications au traitement		
3.8 Annonce du diagnostic et information du patient		
4. Prise en charge thérapeutique :		
4.1 Objectifs		
4.2 Professionnels impliqués (et modalités de coordination)		
4.3 Prise en charge thérapeutique (pharmacologique et autre)		

<p>Gestion de la poussée chez un patient dont le diagnostic d'une NMOSD est déjà posé</p>	<ul style="list-style-type: none"> • Watanabe S, Misu T, Miyazawa I, Nakashima I, Shiga Y, Fujihara K, Itoyama Y. Low-dose corticosteroids reduce relapses in neuromyelitis optica: a retrospective analysis. <i>Mult Scler.</i> 2007 Sep;13(8):968-74. • Bonnan M, Valentino R, Olindo S, Mehdaoui H, Smadja D, Cabre P. Plasma exchange in severe spinal attacks associated with neuromyelitis optica spectrum disorder. <i>Mult Scler.</i> 2009 Apr;15(4):487-92. • Merle H , Olindo S, Jeannin S, Valentino R, Mehdaoui H, Cabot F, Donnio A, Hage R, Richer R, Smadja D, Cabre P. Treatment of optic neuritis by plasma exchange (add-on) in neuromyelitis optica. <i>Arch Ophthalmol.</i> 2012 Jul;130(7):858-62. • Kleiter I , Gahlen A , Borisow N , Fischer K, et al. Neuromyelitis optica: Evaluation of 871 attacks and 1,153 treatment courses. <i>Ann Neurol.</i> 2016 Feb;79(2):206-16. • Bonnan M , Valentino R , Debeugny S , Merle H, Fergé JL , Mehdaoui H , Cabre P. Short delay to initiate plasma exchange is the strongest predictor of outcome in severe attacks of NMO spectrum disorders. <i>J Neurol Neurosurg Psychiatry.</i> 2018 Apr;89(4):346-351. • Trebst C, Jarius S, Berthele A, et al. Update on the diagnosis and treatment of neuromyelitis optica: recommendations of the neuromyelitis optica study group (NEMOS). <i>J Neurol</i> 2014;261:1–16. • Ipe TS, Raval JS, Fernando LP, et al. Therapeutic plasma exchange for neuromyelitis optica spectrum disorder: A multicenter 	<ul style="list-style-type: none"> - L'initiation des échanges plasmatiques, comme techniques thérapeutiques à suggérer, dans la prise en charge du patient adulte après la survenue d'une poussée. - Recommandations concernant l'utilisation des corticoïdes chez l'enfant et l'adulte. - Suggestions concernant l'utilisation d'autres traitements thérapeutiques dans la gestion de la poussée chez l'enfant.
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	<p>retrospective study by the ASFA neurologic diseases subcommittee. J Clin Apher. 2020;35(1):25-32.</p> <ul style="list-style-type: none"> • Kleiter I, Gahlen A, Borisow N, et al. Apheresis therapies for NMOSD attacks: A retrospective study of 207 therapeutic interventions. Neurol Neuroimmunol Neuroinflamm. 2018;5(6):e504. 	
Cas particulier de la prise en charge thérapeutique de la première poussée de neuromyéélite optique		
Traitement de fond de la maladie : Les anti-lymphocytes B (anti-CD20 et anti-CD19)	<ul style="list-style-type: none"> • Paolilo RB, Hachohen Y, Yazbeck E, Armangue T, Bruijstens A, Lechner C, Apostolos-Pereira SL, Martynenko Y, Breu M, de Medeiros Rimkus C, Wassmer E, Baumann M, Papetti L, Capobianco M, Kornek B, Rostásy K, da Paz JA, Ciccarelli O, Lim M, Saiz A, Neuteboom R, Marignier R, Hemingway C, Sato DK, Deiva K. Treatment and outcome of aquaporin-4 antibody-positive NMOSD. Neurol Neuroimmunol Neuroinflamm. 2020 Jul 30;7(5):e837. • Kim SH, Huh SY, Lee SJ, Joung A, Kim HJ. A 5-year follow-up of rituximab treatment in patients with neuromyelitis optica spectrum disorder. JAMA Neurol. 2013 Sep 1;70(9):1110-7. • Zéphir H, Bernard-Valnet R, Lebrun C, Outteryck O, Audoin B, et al. Rituximab as first-line therapy in neuromyelitis optica: efficiency and tolerability. J Neurol. 2015 Oct;262(10):2329-35. • Mealy MA, Wingerchuk DM, Palace J, Greenberg BM, Levy M. Comparison of relapse and treatment failure rates among patients with neuromyelitis optica: multicenter study of treatment efficacy. JAMA Neurol. 2014 Mar;71(3):324-30. • Torres J, Pruitt A, Balcer L, et al. Analysis of the 	<ul style="list-style-type: none"> - Description de l'efficacité du rituximab par rapport à d'autres immunosuppresseurs. - Description d'une élévation du taux d'anticorps anti-AQP4, associée à une augmentation de la cytokine BAFF, chez plusieurs patients, 2 semaines après la première injection de rituximab, pouvant parfois expliquer l'aggravation sous rituximab. - Description de l'efficacité de l'inébilizumab dans la survenue de nouvelles poussées de NMOSD. - Description de l'efficacité de l'ofatumumab dans le traitement de la NMOSD associée à des anticorps anti-AQP4 en cas d'intolérance au rituximab. - Recommandation concernant l'utilisation du rituximab comme traitement de première intention ou de recours.

	<p>treatment of neuromyelitis optica. <i>J Neurol Sci.</i> 2015;351:31-35.</p> <ul style="list-style-type: none"> • Jeong IH, Kim SH, Hyun JW, Joung A, Cho HJ, Kim HJ. Tumefactive demyelinating lesions as a first clinical event: Clinical, imaging, and follow-up observations. <i>J Neurol Sci.</i> 2015 Nov 15;358(1-2):118-24. • Nikoo Z, Badihian S, Shaygannejad V, Asgari N, Ashtari F. Comparison of the efficacy of azathioprine and rituximab in neuromyelitis optica spectrum disorder: a randomized clinical trial. <i>J Neurol.</i> 2017;264:2003-2009. • Zhang M, Zhang C, Bai P, Xue H, Wang G. Effectiveness of Low Dose of Rituximab Compared With Azathioprine in Chinese Patients With Neuromyelitis Optica: An Over 2-year Follow-Up Study. <i>Acta Neurol Belg.</i> 2017 Sep;117(3):695-702. • Chen H, Qiu W, Zhang Q, Wang J, et al. Comparisons of the Efficacy and Tolerability of Mycophenolate Mofetil and Azathioprine as Treatments for Neuromyelitis Optica and Neuromyelitis Optica Spectrum Disorder. <i>Eur J Neurol.</i> 2017 Jan;24(1):219-226. • Poupart J, Giovannelli J, Deschamps R, Audoin B, Ciron J, Maillart E, Papeix C, Collongues N, Bourre B, Cohen M, Wiertlewski S, Outteryck O, Laplaud D, Vukusic S, Marignier R, Zephir H; NOMADMUS study group. Evaluation of efficacy and tolerability of first-line therapies in NMOSD. <i>Neurology.</i> 2020 Apr 14;94(15):e1645-e1656. • Tahara M, Oeda T, Okada K, Kiriya T, Ochi K, Maruyama H, Fukaura H, Nomura K, Shimizu Y, 	
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	<p>Mori M, Nakashima I, Misu T, Umemura A, Yamamoto K, Sawada H. Safety and efficacy of rituximab in neuromyelitis optica spectrum disorders (RIN-1 study): a multicentre, randomised, double-blind, placebo-controlled trial <i>Lancet Neurol.</i> 2020 Apr;19(4):298-306.</p> <ul style="list-style-type: none"> • Zhou Y, Zhong X, Shu Y, et al. Clinical course, treatment responses and outcomes in Chinese paediatric neuromyelitis optica spectrum disorder. <i>Mult Scler Relat Disord.</i> 2019 Feb;28:213-220. • Ciron J, Audoin B, Bourre B, Brassat D, Durand-Dubief F, Laplaud D, Maillart E, Papeix C, Vukusic S, Zephir H, Marignier R, Collongues N, NOMADMUS group, under the aegis of OFSEP, SFSEP. Recommendations for the use of Rituximab in neuromyelitis optica spectrum disorders. <i>Rev Neurol (Paris).</i> 2018 Apr;174(4):255-264. • Nakashima I, Takahashi T, Cree BA, Kim HJ, Suzuki C, Genain CP, Vincent T, Fujihara K, Itoyama Y, Bar-Or A. Transient increases in anti-aquaporin-4 antibody titers following rituximab treatment in neuromyelitis optica, in association with elevated serum BAFF levels. <i>J Clin Neurosci.</i> 2011 Jul;18(7):997-8. • Kim SH, Jeong IH, Hyun JW, Joung A, Jo HJ, Hwang SH, Yun S, Joo J, Kim HJ. Treatment Outcomes With Rituximab in 100 Patients With Neuromyelitis Optica: Influence of FCGR3A Polymorphisms on the Therapeutic Response to Rituximab. <i>JAMA Neurol.</i> 2015 Sep;72(9):989-95. • Kim SH, Kim W, Li XF, Jung IJ, Kim HJ. Repeated treatment with rituximab based on the 	
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	<p>assessment of peripheral circulating memory B cells in patients with relapsing neuromyelitis optica over 2 years. Arch Neurol. 2011 Nov;68(11):1412-20.</p> <ul style="list-style-type: none"> • Cho EB, Cho HJ, Seok JM, Min JH, Kang ES, Kim BJ. The IL-10-producing regulatory B cells (B10 cells) and regulatory T cell subsets in neuromyelitis optica spectrum disorder. Neurol Sci. 2018 Mar;39(3):543-549. • Cohen M, Romero G, Bas J, Ticchioni M, Rosenthal M, Lacroix R, Brunet C, Rico A, Pelletier J, Audoin B, Lebrun C. Monitoring CD27+ memory B-cells in neuromyelitis optica spectrum disorders patients treated with rituximab: Results from a bicentric study. J Neurol Sci. 2017 Feb 15;373:335-338. • Durozard P, Rico A, Boutiere C, Maarouf A, Lacroix R, Cointe S, Fritz S, Brunet C, Pelletier J, Marignier R, Audoin B. Comparison of the Response to Rituximab between Myelin Oligodendrocyte Glycoprotein and Aquaporin-4 Antibody Diseases. Ann Neurol. 2020 Feb;87(2):256-266. • Cree BAC, Bennett JL, Kim HJ, Weinshenker BG, Pittock SJ, Wingerchuk DM, Fujihara K, Paul F, Cutter GR, Marignier R, Green AJ, Aktas O, Hartung HP, Lublin FD, Drappa J, Barron G, Madani S, Ratchford JN, She D, Cimbora D, Katz E; N-MOmentum study investigators. Inebilizumab for the treatment of neuromyelitis optica spectrum disorder (N-MOmentum): a double-blind, randomised placebo-controlled phase 2/3 trial. Lancet. 2019 Oct 12;394(10206):1352-1363. 	
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<p>Traitement de fond de la maladie : Les autres traitements</p>	<ul style="list-style-type: none"> • Mealy MA, Wingerchuk DM, Palace J, Greenberg BM, Levy M. Comparison of relapse and treatment failure rates among patients with neuromyelitis optica: multicenter study of treatment efficacy. <i>JAMA Neurol.</i> 2014 Mar;71(3):324-30. • Nikoo Z, Badihian S, Shaygannejad V, Asgari N, Ashtari F. Comparison of the efficacy of azathioprine and rituximab in neuromyelitis optica spectrum disorder: a randomized clinical trial. <i>J Neurol.</i> 2017;264:2003-2009. • Paolilo RB, Hachohen Y, Yazbeck E, Armangue T, Bruijstens A, Lechner C, Apostolos-Pereira SL, Martynenko Y, Breu M, de Medeiros Rimkus C, Wassmer E, Baumann M, Papetti L, Capobianco M, Kornek B, Rostásy K, da Paz JA, Ciccarelli O, Lim M, Saiz A, Neuteboom R, Marignier R, Hemingway C, Sato DK, Deiva K. Treatment and outcome of aquaporin-4 antibody-positive NMOSD. <i>Neurol Neuroimmunol Neuroinflamm.</i> 2020 Jul 30;7(5):e837. • Zhou Y, Zhong X, Shu Y, et al. Clinical course, treatment responses and outcomes in Chinese paediatric neuromyelitis optica spectrum disorder. <i>Mult Scler Relat Disord.</i> 2019 Feb;28:213-220. • Costanzi C, Matiello M, Lucchinetti CF, Weinshenker BG, Pittock SJ, Mandrekar J, Thapa P, McKeon A. Azathioprine: tolerability, efficacy, and predictors of benefit in neuromyelitis optica. <i>Neurology.</i> 2011 Aug 16;77(7):659-66. • Elson L, Kitley J, Luppe S, Lythgoe D, Mutch K, et al. Long-term efficacy, tolerability and retention rate of azathioprine in 103 aquaporin-4 antibody-positive neuromyelitis optica spectrum disorder 	<ul style="list-style-type: none"> - Efficacité de l'azathioprine et le mycophenolate mofétil dans les NMOSD. - Efficacité de l'azathioprine et le mycophenolate mofétil chez l'enfant dans les NMOSD. - Efficacité de la mitoxantrone dans les NMOSD.
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5. Suivi		
5.1 Objectifs		
5.2 Professionnels impliqués (et modalités de coordination)		
5.3 Rythme et contenu des consultations		
5.4 Examens complémentaires		

5.5 Grossesse

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- Shosha E, Pittock SJ, Flanagan E, Weinshenker

- Etude des risques de l'utilisation des traitements NMOSD au cours de la grossesse et l'allaitement et recommandations concernant l'utilisation de ces thérapies durant la période de grossesse.
- Effet de la NMOSD sur la fertilité et les complications au cours de la grossesse et à l'issue de la grossesse.
- Effet de la grossesse sur l'évolution de la maladie.
- Risque de fausses couches chez les femmes NMOSD séropositives pour l'anticorps anti-AQP4.
- Risque de pré-éclampsie chez les patientes NMOSD par rapport au groupe témoin.
- Risque de rechute du spectre de NMOSD après l'accouchement.
- Augmentation de risque de poussées après l'interruption de l'immunosuppresseur pendant la grossesse.

	<p>BG. Neuromyelitis optica spectrum disorders and pregnancy: Interactions and management. <i>Mult Scler.</i> 2017 Dec;23(14):1808-1817.</p> <ul style="list-style-type: none"> • Klawiter EC, Bove R, Elson L, Alvarez E, Borisow N, Cortez M, Mateen F, Mealy MA, Sorum J, Mutch K, Topyne SM, Ruprecht K, Buckle G, Levy M, Wingerchuk D, Paul F, Cross AH, Jacobs A, Chitnis T, Weinshenker B. High risk of postpartum relapses in neuromyelitis optica spectrum disorder. <i>Neurology.</i> 2017 Nov 28;89(22):2238-2244. • Bove R, Elson L, Alvarez E, Borisow N, Cortez MM, Mateen FJ, Mealy MA, Mutch K, Topyne S, Ruprecht K, Buckle G, Levy M, Wingerchuk DM, Paul F, Cross AH, Weinshenker B, Jacob A, Klawiter EC, Chitnis T. Female hormonal exposures and neuromyelitis optica symptom onset in a multicenter study. <i>Neurol Neuroimmunol Neuroinflamm.</i> 2017 Mar 24;4(3):e339. • Davoudi V, Keyhanian K, Bove RM, Chitnis T. Immunology of neuromyelitis optica during pregnancy. <i>Neurol Neuroimmunol Neuroinflamm.</i> 2016 Oct 7;3(6):e288. • Nour MM, Nakashima I, Coutinho E, Woodhall M, Sousa F, Revis J, Takai Y, George J, Kitley J, Santos ME, Nour JM, Cheng F, Kuroda H, Misu T, Martins-da-Silva A, DeLuca GC, Vincent A, Palace J, Waters P, Fujihara K, Leite MI. Pregnancy outcomes in aquaporin-4-positive neuromyelitis optica spectrum disorder. <i>Neurology.</i> 2016 Jan 5;86(1):79-87. • Shimizu Y, Fujihara K, Ohashi T, Nakashima I, 	
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<u>5.6 Transition</u>		
6. Accompagnement médico-social		
6.1 <u>Accès aux soins et aux droits</u>		
6.2 <u>Les aides et l'accompagnement spécifiques des enfants</u>		
6.3 <u>Les aides et l'accompagnement spécifiques des étudiants</u>		
6.4 <u>Les aides et l'accompagnement spécifiques des adultes</u>		
6.5 <u>Recommandations particulières à destination des structures sociales, médico-sociales et scolaires</u>		
6.6 <u>Recommandations pour le remplissage des dossiers destinés à l'évaluation du handicap ou de la perte d'autonomie de la personne atteinte de NMO</u>		
6.7 <u>Contacts et autres informations utiles</u>		

Annexe 1. Recherche documentaire et sélection des articles

Recherche documentaire

Sources consultées	Bases de données : PUBMED Sites internet : PUBMED
Période de recherche	Non limitée dans le temps
Langues retenues	Anglais et français
Mots clés utilisés	Neuromyelitis, neuromyelitis optica, NMOSD
Nombre d'études recensées	> 300
Nombre d'études retenues	106

Critères de sélection des articles

Selon le type de la publication et le thème traité.

Annexe 2. Liste des participants

Ce travail a été coordonné par le Pr Kumaran Deiva, coordonnateur du Centre de Référence des Maladies Inflammatoires Rares du Cerveau Et de la Moelle (MIRCEM) et le Pr Romain Marignier, responsable du site constitutif MIRCEM de Lyon.

Ont participé à l'élaboration du PNDS :

Groupe multidisciplinaire rédactionnel

- Pr Kumaran Deiva, neuropédiatre, CHU Kremlin Bicêtre
- Pr Romain Marignier, neurologue, Hospices Civils de Lyon
- D^r Caroline Papeix, neurologue, CHU Pitié-Salpêtrière
- D^r Hélène Maurey, neuropédiatre, CHU Kremlin Bicêtre
- D^r Jonathan Ciron, neurologue, CHU Toulouse
- D^r Nicolas Collongues, neurologue, CHRU Strasbourg
- D^r Emmanuel Cheuret, neuropédiatre, CHU Toulouse
- Pr Bertrand Audoin, neurologue, Hôpital de la Timone
- Pr Hélène Zephir, neurologue, CHU Lille
- D^r Elisabeth Maillart, neurologue, CHU Pitié-Salpêtrière
- D^r Pierre Meyer, neuropédiatrie, CHU de Montpellier
- Pr Sandra Vukusic, neurologue, Hospices Civils de Lyon
- Pr Muriel Doret-Dion, gynécologue, obstétricien, Hospices Civils de Lyon
- D^r Julie Pique, neurologue, Hospices Civils de Lyon
- M^{me} Evelyne Yver, assistante sociale, CHU Kremlin Bicêtre
- M^{me} Carole Lattaud, assistante sociale, CHU Pitié Salpêtrière
- M. Ala-Eddine Allouche, chef de projet MIRCEM, CHU Kremlin Bicêtre

Groupe de relecture

- Pr Sylvie Nguyen The Tich, neuropédiatre, CHU Lille
- D^r Nafissa Mamoudjy, neuropédiatre de ville, Saint-Maurice
- D^r Marie Thérèse Abi Warde, neuropédiatrie, CHU Strasbourg
- D^r Bertrand Bourre, neurologue, CHU Rouen
- D^r Marie-Caroline Pouget, médecin physique réadaptateur, Hospices Civils de Lyon
- Pr Caroline Froment Tilikete, neurologue, neuro-ophtalmologue, Hospices Civils de Lyon
- Pr Jérôme De Sèze, neurologue, CHU Strasbourg
- M^{me} Anne-Colombe Debrouse, psychologue, CHU Kremlin Bicêtre
- M^{me} Marine Gelé, infirmière, Hospices Civils de Lyon
- M^{me} Christelle Berthier-Maillard, patiente et membre de l'association NMO France
- M^{me} Souad Mazari, responsable de l'association NMO France

Gestion des intérêts déclarés

Tous les participants à l'élaboration du PNDS sur les maladies du spectre de la neuromyéélite optique ont rempli une déclaration d'intérêt. Les déclarations d'intérêt sont en ligne et consultables sur le site internet du centre de référence des maladies inflammatoires rares du cerveau et de la moelle (www.mircem.fr) et sur le site internet de la filière de santé maladies rares BRAIN-TEAM (www.brain-team.fr).

Les déclarations d'intérêt ont été analysées et prises en compte, en vue d'éviter les conflits d'intérêts, conformément au guide HAS « Guide des déclarations d'intérêts et de gestion des conflits d'intérêts » (HAS, 2010).

Modalités de concertation du groupe de travail multidisciplinaire

Réunions par visioconférence : 15/09/2020, 13/10/2020 et 03/12/2020.

Réunion téléphonique : 02/10/2020.

Nombreux échanges par e-mails.

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