Good news for patients with neuromyelitis optica spectrum disorders with anti-aquaporin 4 antibodies: treatment recommendations established by an international group of recognized experts have just been published!

Neuromyelitis optica spectrum disorders (NMOSD) are rare inflammatory diseases of the brain and spinal cord. Rapid and appropriate care is essential to limit the consequences of flare-ups, which can be very severe and lead to heavy disability.

These rare diseases are poorly understood, and no specific treatment was approved for patients with NMOSD until 2019. The efficacy of the initial pharmaceutical therapies (rituximab, tocilizumab, azathioprine, mycophenolate mofetil, and methotrexate) used to treat NMOSD had been proven in the management of other diseases.

Recent advances in research have led to a better understanding of the mechanisms of these rare diseases. Three new pharmaceutical therapies have now been approved in several countries as maintenance treatments for patients with NMOSD with anti-aquaporin 4 antibodies (anti-AQP4 IgG):

- **Eculizumab** for adults
- **Inebilizumab** for adults
- **Satralizumab** for adults and adolescents (12 years of age and older)

Each of these monoclonal antibodies (also called biologics) has a different mechanism of action (see the figure below). Your neurologist will discuss these treatment options with you.

### Mechanisms of action of monoclonal antibodies prescribed to patients with NMOSD

<table>
<thead>
<tr>
<th>Antibodies</th>
<th>Targets</th>
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<tbody>
<tr>
<td><strong>Eculizumab</strong></td>
<td>Complement component 5 (C5)</td>
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| **Inebilizumab** | CD19, expressed on the surface of B lymphocytes  
**Rituximab targets CD20, which is also expressed on the surface of B lymphocytes** |
| **Satralizumab** | Interleukin 6 (IL-6) receptors  
**Tocilizumab also binds to IL-6 receptors** |

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About 90% of NMOSD patients have anti-AQP4 IgG in their blood. Therefore, the approval of these pharmaceutical therapies for the treatment of NMOSD with anti-AQP4 IgG is very good news; it opens new perspectives in the management of our diseases!

For the first time, standard recommendations for the treatment of NMOSD with anti-AQP4 IgG using these new pharmaceutical therapies have been established by 24 recognized experts from 15 countries*!

These 25 recommendations were established by consensus using the Delphi method:

- 11 relate to the initiation of, or switching between, eculizumab, inebilizumab and satralizumab;
- 3 relate to monotherapy/comboination therapy;
- 7 relate to safety and patient population considerations;
- 3 relate to biomarkers and patient-reported outcomes (PROs);
- 1 addresses current research gaps.

These recommendations provide healthcare professionals with clear guidance on the use of the newly available therapies (eculizumab, inebilizumab and satralizumab) in patients with NMOSD with anti-AQP4 IgG. They were published in the July 2023 volume of the international journal Neurology: Neuroimmunology and NeuroInflammation, and were also presented in early June in Oxford, at the meeting of members of the MOGAD Eugène Devic European Network (MEDEN).

New perspectives

While research is still needed at various levels to improve the treatment strategy for NMOSD, these recommendations are an important step in standardizing care and are a source of hope for patients. They will also allow NMO France to co-build a therapeutic education program with health professionals to best support you.

NMO France thanks the experts for their recommendation regarding the importance of taking patients’ preferences into account in the choice of treatment.

The purpose of this NMO France spotlight on therapeutic news for NMOSD is to inform you about the different treatment options available by passing on the recommendations published by recognized international experts.
Each patient is unique, and this release is not a substitute for medical advice. Your neurologist will advise you on the most suitable management options for your condition, either when starting treatment at the time of NMOSD diagnosis, or when considering switching treatment in case of relapse.

Do not hesitate to discuss your options with your neurologist!

In France, patients with NMOSD can benefit from a consultation either in one of the NMOSD reference centers (coordinating center or constitutive centers) or in an expert center: https://www.nmo-france.org/centres-de-reference/

*International experts who have prepared these recommendations: Friedemann Paul and Ingo Kleiter (Germany); Sudarshini Ramanathan (Australia); Marco Lana-Peixoto and Douglas Kazutoshi Sato (Brazil); Dalia Rotstein (Canada); Chao Quan (China); Nasrin Asgari (Denmark); Romain Marignier and Jérôme De Sèze (France); Lekha Pandit (India); Adi Vaknin-Dembinsky (Israel); Kazuo Fujihara and Satoshi Kuwabara (Japan); Najib Kissani (Morocco); Georgina Arrambide and Albert Saiz (Spain); Ho Jin Kim (South Korea); Jacqueline Palace, Saif Huda and Mr. Isabel Leite (United Kingdom); Jeffrey L. Bennett, Bruce Anthony Campbell Cree and Sean J. Pittock (USA).