The Czech National Registry of Myasthenia Gravis MyReg

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PURPOSE AND BACKGROUND

Myasthenia gravis belongs to a group of rare autoimmune neuromuscular diseases. Its prevalence varies from 14 to 20 per million in Europe. Therefore, the expected prevalence in Czech Republic is estimated to be 1500-2000. The rareness of the disease and the fluctuating symptoms contribute to several difficulties and challenges for patients and their caregivers, stressing the need of patient-registry.

In 2015, the Czech National Registry of Myasthenia Gravis (MyReg) was founded. The registry collects data from 10 neuromuscular centres, two of which are members of the European Reference Network. The structure of the registry was created according to EuroMyasthenia recommendations that allow the registry to be opened for cooperation with other European registries. The scales MGC (Myasthenia Gravis Composite scale) and MG-QOL15 (Myasthenia Gravis Quality of life) are being used to quantify the clinical symptoms and quality of life. All data in the registry, but MG-QOL15, are professional-reported, however, patients can see their own data using their account login. In the future, patients will also be able to fill out the MG-QOL15 form online. The technical background, storage and backup of data are provided by the Institute of Biostatistics and Analyses (IBA).

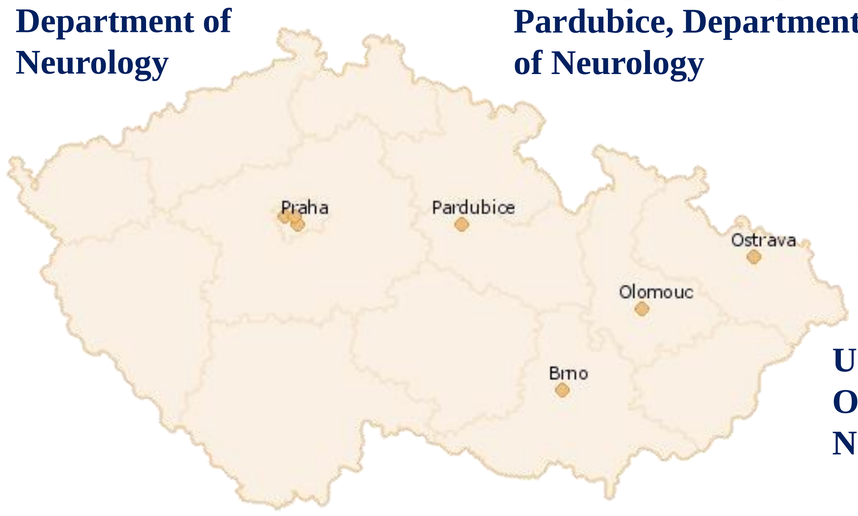
European Reference Network for rare or low prevalence complex diseases
Network Neuromuscular Diseases (ERN EURO-NMD)
Member Motol University Hospital — Czechia

Motol University Hospital in Prague, Department of Neurology and Department of Paediatric Neurology

The General University Hospital in Prague, Department of Neurology

Thomayer Hospital in Prague, Department of Neurology

The Regional Hospital in Pardubice, Department of Neurology



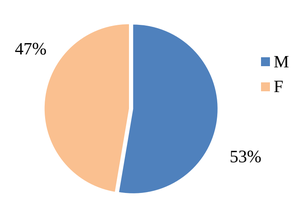
University Hospital Ostrava, Department of Neurology and Department of Paediatric Neurology

University Hospital Olomouc, Department of Neurology

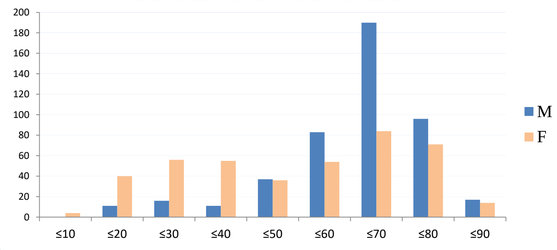
The University Hospital Brno, Neuromuscular Centre

European Reference Network for rare or low prevalence complex diseases
Network Neuromuscular Diseases (ERN EURO-NMD)
Member Fakultní nemocnice Brno — Česká republika

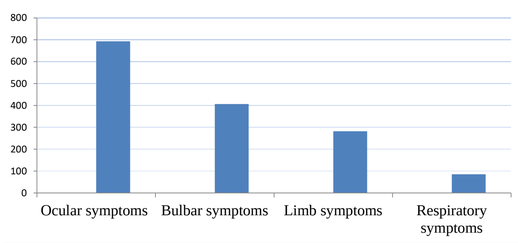
GENDER



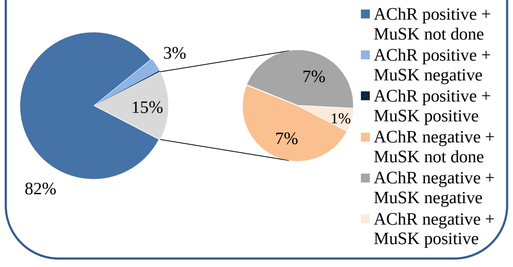
AGE OF ONSET



FIRST SYMPTOMS



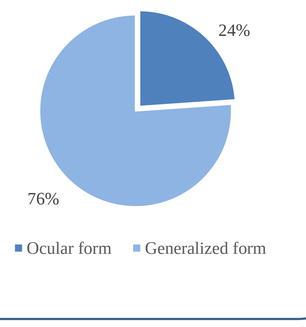
ANTIBODIES



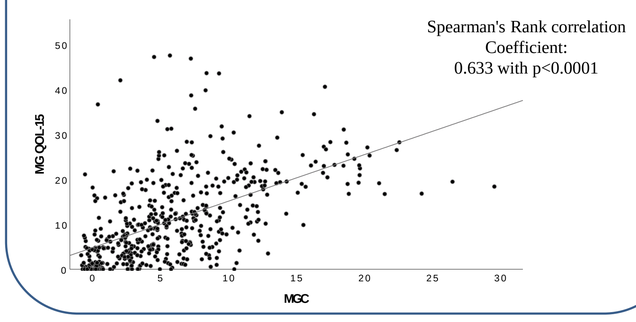
THE POPULATION OF MyReg

As of October 2019, there are 932 patients in the registry, 53% of which are males and 47% are females. The average age of onset is 62 years in males and 50 years in females. The most frequent first symptoms are ocular (79%), bulbar symptoms (46%), followed by limb (32%) and respiratory symptoms (10%). In agreement with the other studies, about a quarter of patients (23%) developed pure ocular myasthenia, while the rest, 77%, is classified as generalized form. Similarly, 15% of patients are sero-negative, 85 % anti-AChR positive and 1,3% anti-MuSK positive. The average MGC and MG-QOL15 is 5 and 11 respectively. The correlation between MGC and MG-QOL15 is 0.633 with $p<0.0001$.

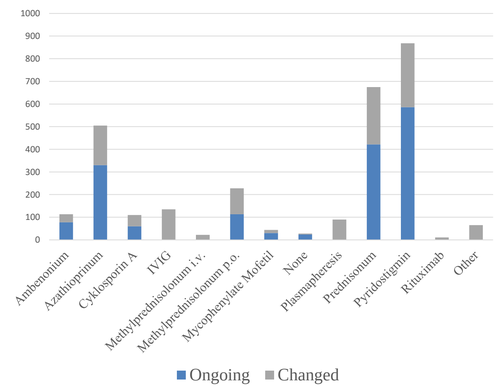
CLINICAL FORM



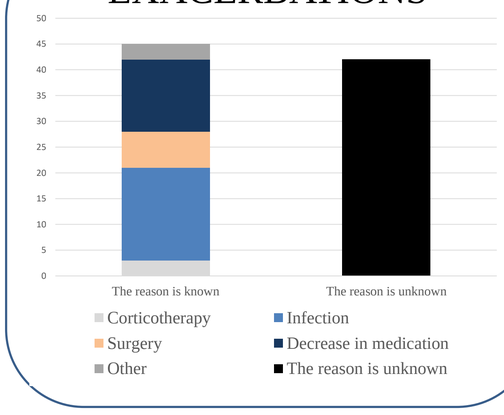
THE CORRELATION BETWEEN MGC AND MG-QOL15



THERAPY



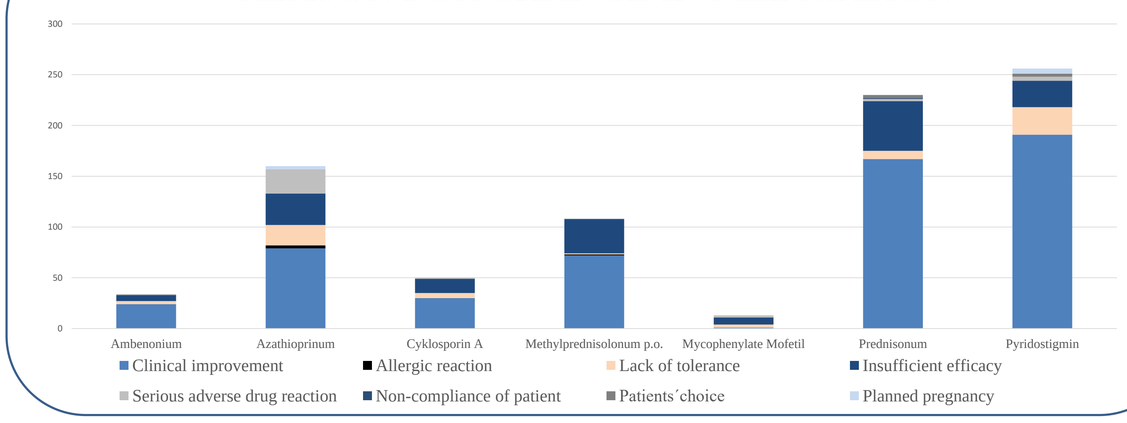
EXACERBATIONS



TREATEMENT AND EXACERBATIONS

The exacerbations accounted for 9% of hospital visits. Those were mostly caused by infections, decrease of doses and surgeries. The most frequently used medication is a combination of pyridostigmine with prednisone or azathioprine. The reasons for changes in treatment were clinical improvement in 65% of cases, insufficient efficacy in 19% and lack of tolerance in 8% of cases. The azathioprine accounted for majority (66%) of adverse reactions.

REASON FOR CHANGE IN MEDICATION



CONCLUSION

The registry MyReg serves us as a source of data for improvement in ongoing care and therapy strategies and it is also used for patient-recruitment for clinical trials. The registry is opened for international cooperation. The future data will be essential for the healthcare planning in patients with myasthenia in Czech Republic.

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