

MEN Online report 2019 meeting

Purpose

For the 2019 meeting in Drammen, Norway, MEN welcomed 41 participants from 11 countries and a total of 11 organizations: 26 meeting delegates, 11 young adults and 6 external guests.

The member countries had been asked to submit their national reports covering the past two years prior to the conference and presented a brief overview of the main highlights of the past two years during the meeting.

MEN highlights

The main activities since the last meeting have been:

- Establish contact with non-members associations to join MEN
- Getting 3 new members Spain (2018), Luxembourg (2019) and UK (2019)
- Communicate to and with the members by email (official matters) or by newsletter via the website (25 news since the last meeting)
- Make a new website for MEN (www.marfan.eu)
- Use of the Facebook page
- Reply to contact from our members or European Marfan patients concerning medical care and redirection to national associations, VASCERN or known medical specialist centres.
- Work with the Norwegian association on organizing the MEN 2019 meeting in Drammen
- Represent MEN in the Marfan Research Symposium 2018
- Participate in the first European patient day
- Keep in touch with the VASCERN project
- Work on common projects for MEN:
 - Bakoumba children book in European languages
- New projects being considered after the 2019 meeting:
 - Bakoumba video project
 - Compilation of information and experience with surgery
 - Update of the Marfan ICE card
 - Compilation of list of Marfan reference center in all member countries (in collaboration with VASCERN)
 - Collaboration in funding common research projects

Medical presentations

1. Stefan Axelsson, TAKO-Senteret – Lovisenberg Dikonale Sykehus: «*Cardiovascular complications, craniofacial aberrations, impaired breathing during sleep, sleep disruption and fatigue in adults with verified Marfan syndrome.*» Presentation of an interdisciplinary project at Lovisenberg Diaconal Hospital

This interdisciplinary study started in November 2018 and is expected to last until 2021. It aims at establishing a correlation between sleep disruption, impaired breathing during sleep and sleep apnoea and the development of cardiovascular complications and fatigue in Marfan patients. To date there are only some preliminary conclusions: Sleep apnoea is common in Marfan patients, so all of them should be checked via polygraphy or polysomnography and, since it may lead to an increased progression of aorta dilatation, therapy of sleep apnoea may have a positive effect on the aortic disease and the quality of life.

2. Thy Thy Vanem, UiO (University in Oslo), Institute of Clinical Medicine, Faculty of Medicine: “*The Norwegian Marfan Study, part 2 - A 10-year follow-up.*”

The study was carried out in 2014-2015 for the purpose of determining survival and causes of death, changes of organ manifestations during the 10-year follow-up and changes in health related quality of life after 10 years. Conclusions drawn were:

- Ectopia lentis and progression of this can occur in adulthood.
- Dural ectasia and anterior meningocele and progression of this can occur in adulthood
- Life expectancy is still reduced in this MFS-cohort compared to the general Norwegian population.
- Cardiovascular complications develop throughout life, especially aortic pathology, the main cause of death in MFS patients.
- Death and aortic pathology occurs earlier in MFS men compared to MFS women.

Knowledge on new organ manifestations in adults with MFS has implications for diagnosis and follow-up!

3. Gry Velvin and Heidi Johansen, TRS Kompetansesenter: “*Physical function and psychosocial aspect in persons with Marfan syndrome and Loeys Dietz syndrome (and vascular Ehlers Danlos)*”

This presentation focused on the evaluation of the wellbeing of Marfan syndrome or other aortic disease patients.

Having Marfan has a negative influence on the perceived quality of life. In January 2018: questionnaire to 80 adults with questions about QoL. 57 responded. 60% were women. Average age was 42 years (between 18-68). 79% had chronic pain complaints. Average age diagnosis was 34 years. They held a focus group Interviews: the most important aspects that were discussed were psycho-social aspects and psychological complaints. Discuss and share issues with each other in groups. It is a scientific approach. Group of 36 people. 19 worked there, 1 student, 16 retired. Part of the research is that questions have been asked about the physical activities (broad: home garden household and sport). Had they been given guidelines or restrictions in terms of sports etc? All results and answers were discussed.

4. Pr. Michael Jacobs, Maastricht University Medical Center: "International Marfan patients with complex aortic pathologies: the patient flies to the surgeon or vice versa?"

The full presentation of Pr. Jacobs can be found on the MEN website (<https://www.marfan.eu/media/>), and demonstrated full aorta replacement as well as the risks of malperfusion of the spinal chord during surgery.