The Nopho-European Study on Cerebellar Mutism Syndrome (CMS)

Wibroe, Morten; Avula, Shivaram; Cappelen, Johan; Castor, Charlotte; Clausen, Niels; Devenney, Irene; Fellows, Greg; Grillner, Pernilla; Gupta, Ramneek; Gustavsson, Bengt; Heyman, Mats; Holm, Stefan; Karpipinen, Atte; Kiudeliene, Rosita; Klausen, Camilla; Lahteenmaki, Paivi; Lönnqvist, Tuula; Lowis, Stephen; Mallucci, Conor; Mathiasen, Rene; Mattson, Mattias; Nilsson, Pelle; Nordfors, Kristiina; Nyman, Per; Nysom, Karsten; Persson, Karin; Pesola, Jouni; Pizer, Barry; Rask, Olof; Sabel, Magnus; Schmiegelow, Kjeld; Sehested, Astrid; Tonning-Olsson, Ingrid; Torsvik, Ingrid Kristin; van Baarsen, Kirsten; Walker, David; Westerholm-Ormio, Mia; Zetterqvist, Barbara; Juhler, Marianne

Published in:
Neuro-Oncology

Link to article, DOI:
10.1093/neuonc/now066.5

Publication date:
2016

Document Version
Publisher's PDF, also known as Version of record

Link back to DTU Orbit

Citation (APA):

General rights
Copyright and moral rights for the publications made accessible in the public portal are retained by the authors and/or other copyright owners and it is a condition of accessing publications that users recognise and abide by the legal requirements associated with these rights.

- Users may download and print one copy of any publication from the public portal for the purpose of private study or research.
- You may not further distribute the material or use it for any profit-making activity or commercial gain.
- You may freely distribute the URL identifying the publication in the public portal.

If you believe that this document breaches copyright please contact us providing details, and we will remove access to the work immediately and investigate your claim.
CMS-06. THE NOPHO-EUROPEAN STUDY ON CEREBELLAR MUTISM SYNDROME (CMS)
Morten Wibroe1, Shivaram Avula1, Johan Cappelen1, Charlotte Castor2, Niels Clausen3, Irene Devenney4, Greg Fellows5, Pernilla Grillner1, Ranmeek Gupta2, Bengt Gustavsson2, Mats Heyman2, Stefan Holm2, Atte Karpinnen2, Rosita Kueckelhoven6, Camilla Klausen7, Paivi Lahrenmaa11, Tuula Lönnyqvist4, Stephen Lowis21, Conor Mallucci2, René Mathiassen1, Mattias Mattson2, Pelle Nilsson3, Kristina Nordfors1, Per Nyman8, Karsten Nysom1, Karin Persson6, Joumi Pesola10, Barry Pizer2, Olof Rask4, Magnus Salvel16, Kjeld Schmiegelow1, Astrid Sehested5, Ingrid Tenning-Olsson4, Ingrid Kristin Torsvik17, Kirsten van Baarsen18, David Walker19, Mia Westerholm-Ormio9, Barbara Zetterqvist7, and Marianne Juhler1; 1University Hospital Rigshospitalet, Copenhagen, Denmark; 2Alder Hey Children’s Hospital, Liverpool, UK; 3St Olavs University Hospital, Trondheim, Norway; 4Ska˚ne University Hospital, Lund, Sweden; 5Linköping University Hospital, Linköping, Sweden; 6Karolinska University Hospital, Stockholm, Sweden; 7Technical University of Denmark, Copenhagen, Denmark; 8Helsinki University Hospital, Helsinki, Finland; 9Hospital of Lithuanian University of Health Sciences Kauno Klinikos, Kaunas, Lithuania; 10Turku University Hospital, Turku, Finland; 11University Hospital of Umeå, Umeå, Sweden; 12Uppsala University Hospital, Uppsala, Sweden; 13Tampere University Hospital, Tampere, Finland; 14BarnReHab Skåne, Lund, Sweden; 15Sahlgrenska University Hospital, Göteborg, Sweden; 16Haukeland University Hospital, Bergen, Norway; 17Radboud University Medical Centre, Nijmegen, The Netherlands; 18Children Brain Tumour Research Centre, Nottingham, UK; 19Kuopio University Hospital, Kuopio, Finland; 20Bristol Royal Children’s Hospital, Bristol, UK

BACKGROUND: The cerebellar mutism syndrome (CMS) is one of the most disabling late effects after neurosurgery for a posterior fossa tumour in childhood. The reported incidences vary substantially in previous studies. AIMS: Pathophysiology is unknown, but damage to cerebello-thalamo-cerebral circuits is likely. The study focuses on the risk factors for development and severity of CMS including surgery (approaches, techniques and tissue and vascular damage, re-operation) and host genome variants. METHODS: Multicentre study developed as a NOPHO collaborative study coordinated from Righospitalet, Copenhagen with online data registration and database management at Karolinska, Stockholm and quarterly online participant meetings. Registration includes clinical data and speech samples collected preoperatively and at four defined postoperative points for the subsequent 12 months. Therapy, including neurosurgery, is by local standards. A blood sample for genetic analysis is collected from all patients. Imaging is collected and reviewed centrally. RESULTS: The study aims to recruit 550 children. It opened in five Nordic and Baltic countries during 2014/2015; in the Netherlands in February 2016 and will open in the UK during 2016. Two German centres will join in 2017. The target accrual of 550 patients will be reached by the end of 2018. As of February 2016, 67 patients have been included from 12 centres. Mutism has occurred in 7 cases. CONCLUSION: The study will be the largest prospective international study on CMS to date, and the first one to 1) systematically register surgery, use of steroids, standardized speech samples and 2) to investigate the influence of host genome.