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; Karpinen, 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):
CMS-06. THE NOPHO-EUROPEAN STUDY ON CEREBELLAR MUTISM SYNDROME (CMS)

Morten Wibroe1, Shivaram Avula2, Johan Cappelen3, Charlotte Castor4, Niels Clausen2, Irene Devenney6, Greg Fellows21, Pernilla Grillner7, Ramneek Gupta8, Bengt Gustavsson9, Mats Heyman7, Stefan Holm7, Atte Karppinen9, Rosita Kiudeliene10, Camilla Klausen1, Paivi Lahrenmaki11, Tuula Lonnqvist9, Stephen Lowis21, Conor Mallucci2, René Mathiasen1, Mattias Mattson12, Pelle Nilsson13, Kristina Nordfors14, Per Nyman5, Karsten Nysom9, Karin Persson15, Jouni Pesola20, Barry Pizer2, Olof Rasche4, Magnus Saltel16, Kjeld Schmiegelow12, Astrid Sehested1, Ingrid Tonning-Olsson4, Ingrid Kristin Torvik17, 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; 4Skåne University Hospital, Lund, Sweden; 5University Hospital Aarhus, Skerby, Denmark; 6Linköping University Hospital, Linköping, Sweden; 7Karolinska University Hospital, Stockholm, Sweden; 8Technical University of Denmark, Copenhagen, Denmark; 9Helsinki University Hospital, Helsinki, Finland; 10Hospital of Lithuanian University of Health Sciences Kauno Klinikos, Kaunas, Lithuania; 11Turku University Hospital, Turku, Finland; 12University Hospital of Umeå, Umeå, Sweden; 13Uppsala University Hospital, Uppsala, Sweden; 14Tampere University Hospital, Tampere, Finland; 15BarnReHab Skåne, Lund, Sweden; 16Sahlgrenska University Hospital, Göteborg, Sweden; 17Haukeland University Hospital, Bergen, Norway; 18Radboud University Medical Centre, Nijmegen, The Netherlands; 19Children Brain Tumour Research Centre, Nottingham, UK; 20Kuopio University Hospital, Kuopio, Finland; 21Bristol 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 Rigshospitalet, 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.