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

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RATIONALE: 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, reoperation) 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.