Cerebellar mutism syndrome in children with brain tumours of the posterior fossa - DTU Orbit (07/01/2019)

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Background: Central nervous system tumours constitute 25% of all childhood cancers; more than half are located in the posterior fossa and surgery is usually part of therapy. One of the most disabling late effects of posterior fossa tumour surgery is the cerebellar mutism syndrome (CMS) which has been reported in up to 39% of the patients but the exact incidence is uncertain since milder cases may be unrecognized. Recovery is usually incomplete. Reported risk factors are tumour type, midline location and brainstem involvement, but the exact aetiology, surgical and other risk factors, the clinical course and strategies for prevention and treatment are yet to be determined.

Methods: This observational, prospective, multicentre study will include 500 children with posterior fossa tumours. It opened late 2014 with participation from 20 Nordic and Baltic centres. From 2016, five British centres and four Dutch centres will join with a total annual accrual of 130 patients. Three other major European centres are invited to join from 2016/17. Follow-up will run for 12 months after inclusion of the last patient. All patients are treated according to local practice. Clinical data are collected through standardized online registration at pre-determined time points pre- and postoperatively. Neurological status and speech functions are examined pre-operatively and postoperatively at 1-4 weeks, 2 and 12 months. Pre- and postoperative speech samples are recorded and analysed. Imaging will be reviewed centrally. Pathology is classified according to the 2007 WHO system. Germline DNA will be collected from all patients for associations between CMS characteristics and host genome variants including pathway profiles.

Discussion: Through prospective and detailed collection of information on 1) differences in incidence and clinical course of CMS for different patient and tumour characteristics, 2) standardized surgical data and their association with CMS, 3) diversities and results of other therapeutic interventions, and 4) the role of host genome variants, we aim to achieve a better understanding of risk factors for and the clinical course of CMS - with the ultimate goal of defining strategies for prevention and treatment of this severely disabling condition.
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