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Natural course and long-term prognosis in idiopathic Normal Pressure Hydrocephalus

Akademisk avhandling

Som för avläggande av medicine doktorsexamen vid Sahlgrenska akademin, Göteborgs universitet kommer att offentligen försvaras i hörsal Carl Kylberg, Medicinaregatan 7, den 29 maj 2020, klockan 13.00

av Kerstin Andrén

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Avhandlingen baseras på följande delarbeten

I. Andrén K, Wikkelsö C, Tisell M, Hellström P.
Natural course of idiopathic normal pressure hydrocephalus.
Journal of Neurology, Neurosurgery and Psychiatry 2014 Jul; 85: 806-810.

II. Andrén K, Wikkelsö C, Hellström P, Tullberg M, Jaraj D.
Early shunt surgery improves survival in idiopathic Normal Pressure Hydrocephalus. Submitted.

III. Andrén K, Wikkelsö C, Sundström N, Agerskov S, Israelsson H, Laurell K, Hellström P, Tullberg M.
Long-term effects of complications and vascular comorbidity in idiopathic normal pressure hydrocephalus: a quality registry study. Journal of Neurology 2018 Jan; 265: 178-186

IV. Andrén K, Wikkelsö C, Sundström N, Agerskov S, Israelsson H, Laurell K, Hellström P, Tullberg M.
Survival in treated idiopathic normal pressure hydrocephalus. Journal of Neurology 2020 Mar;267(3):640-648.

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Natural course and long-term prognosis in idiopathic Normal Pressure Hydrocephalus

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Abstract

Idiopathic Normal Pressure Hydrocephalus, iNPH, causes gait and balance difficulties, urinary incontinence and cognitive decline in mainly older persons and is treatable by insertion of a cerebrospinal fluid diverting shunt. The effects of postponing treatment in these patients have have been largely unknown and the benefits of treatment in the long-term, mortality and causes of death have not been reported in any large cohort of patients. The aims of this thesis were to study the natural course in untreated iNPH patients, and the effect of postponed treatment, with regard to outcome and survival. Moreover, the aim was to study the long-term outcome and survival in a large unselected cohort of iNPH patients treated all over Sweden, registered in the Swedish Hydrocephalus Quality Registry, SHQR.

A group of patients diagnosed with iNPH who due to capacity problems had to wait median 13 months for shunt surgery, was studied and compared to a group of patients operated without delay. Symptoms progressed during the wait. Once treated, these patients improved, but outcome was less beneficial than in the patients operated without delay (*paper I*). Their mortality was more than two-fold increased (*paper II*). In 979 iNPH patients from the SHQR, around 60% stated being improved 2 to 6 years after shunt surgery. Re-operations were necessary in 26% but did not influence the long-term outcome, and vascular comorbidity had only minor effects (*paper III*). Survival was reduced compared to the general population, and shorter in patients with more pronounced symptoms or with heart diseases. Patients with the most beneficial treatment effects, survived similarly as the general population. Death due to cerebrovascular diseases was more common in iNPH patients, while death due to malignancy was less common, than in the general population (*paper IV*).

This thesis indicates that the natural course of iNPH is progression of symptoms which are only partially reversible and in order to optimize treatment benefits and survival, surgery should be performed without delay. The majority of this aged patient group, also those with vascular comorbidities, have favourable long-term effects and should also be offered treatment. Complications are common, but do not seem to hamper the long-term results. Treatment improves the symptoms and increases survival in iNPH.

Keywords: Normal pressure hydrocephalus, Gait disorders, Cognitive disorders, Natural history, Prognosis

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