Neuropsychological Changes After Endoscopic Third Ventriculostomy for Long-Standing Overt Ventriculomegaly in Adults
—Case Report—

Hideo HAMADA, Nakamasa HAYASHI, Masanori KURIMOTO, Akiko TAKAIWA, Kunikazu KUROSAKI, and Shunro ENDO

Department of Neurosurgery, Faculty of Medicine, University of Toyama, Toyama

Abstract
A 39-year-old woman presented with long-standing overt ventriculomegaly in adults (LOVA) manifesting as gradually worsening headache. Past history included treatment for myeloschisis at birth. Intelligence quotient (IQ) was 115 and preoperative psychological assessment using the Repeatable Battery for the Assessment of Neuropsychological Status (RBANS) showed normal scores in all domains. However, scores for constructional ability were slightly low. Computed tomography revealed severe ventriculomegaly involving the lateral and third ventricles, and magnetic resonance imaging showed aqueductal stenosis. Endoscopic third ventriculostomy was performed under a diagnosis of LOVA. Postoperatively, the headache resolved and the RBANS showed improvements in memory and constructional ability. Detailed evaluation of cognitive function provides a good indicator in the treatment of hydrocephalus with normal IQ.

Key words: long-standing overt ventriculomegaly in adults, hydrocephalus, cognitive function, endoscopic third ventriculostomy, Repeatable Battery for the Assessment of Neuropsychological Status

Introduction
Long-standing overt ventriculomegaly in adults (LOVA) is a new entity of progressive hydrocephalus characterized by slow and gradual evolution from infant onset, and often progressing without evident clinical symptoms, cognitive or behavioral deficits, in sharp contrast with the marked ventriculomegaly. The main morphological abnormality is the congenital aqueduct stenosis, resulting in unbalanced flow of cerebrospinal fluid (CSF) during the progression of hydrocephalus, which is characterized by active and quiescent states. The clinical symptoms of LOVA include macrocephaly with or without subnormal intelligence quotient (IQ), headache, dementia, gait disturbance, urinary incontinence, vegetative state, akinetic mutism, apathetic consciousness, and parkinsonism. Impairment of cognitive function apparently reflects the increased intracranial pressure (ICP). The therapeutic regimen is based on the specific pathophysiological anatomy of the individual, and ventriculoperitoneal shunting using pressure-programmable valve or endoscopic procedure is generally selected.

The Repeatable Battery for the Assessment of Neuropsychological Status (RBANS) scores 5 functional domains as indexes for the evaluation of neuropsychological function. We previously used the RBANS to show that dysfunction of constructional skill due to impairment of parietal lobe was present in patients with secondary normal pressure hydrocephalus (NPH).

We treated a patient with LOVA by endoscopic third ventriculostomy (ETV) with neuropsychological assessment using RBANS before, and 1 week and 3 months after surgery. We emphasize the necessity and usefulness of detailed evaluation of cognitive function when determining treatment strategies for hydrocephalic patients with normal IQ.

Case Report
A 39-year-old woman presented with gradually worsening headache over the course of a few
months. She had a history of repair of myelomacia at birth. Ventriculomegaly was identified, but CSF diversion was not performed because no symptoms of increased ICP were apparent. Her consciousness was clear. Paraparesis of the lower extremities, and impaired bowel and bladder function were present due to spinal dysraphism. Funduscopic examination revealed no papilledema. Computed tomography (CT) revealed severe ventriculomegaly involving the lateral and third ventricles (Fig. 1). Magnetic resonance (MR) imaging showed aqueductal stenosis (Fig. 2).

Her IQ was 115 and Mini Mental State Examination score was 29. The RBANS showed normal scores in all domains: recent memory, 85; visual spatial and constructional ability, 78; understanding of language, 96; attention, 91; delayed memory, 88; and total score, 83.

Standard ETV was performed using a rigid endoscope under a diagnosis of LOVA. Third ventricle floor was thin. Postoperatively, her headache gradually resolved. The RBANS showed improved scores at one week after the operation: recent memory, 97; visual spatial and constructional ability, 84; understanding of language, 92; attention, 97; delayed memory, 97; and total score, 90; and at 3 months: recent memory, 109; visual spatial and constructional ability, 92; understanding of language, 97; attention, 94; delayed memory, 103; and total score, 98. Both recent and delayed memories, and visual spatial and constructional ability underwent particular improvements after ETV with 90% of confidence level of RBANS. Postoperative CT showed no marked reductions in ventricle size, but MR imaging confirmed the flow void of the stoma (Fig. 3).

**Fig. 1** Computed tomography scans disclosing marked ventriculomegaly involving the lateral and third ventricles.

**Fig. 2** Preoperative sagittal T₁- (left) and T₂- weighted (right) magnetic resonance images revealing severe triventriculomegaly and aqueductal stenosis.

**Fig. 3** Postoperative sagittal T₂-weighted magnetic resonance image disclosing the flow void of the stoma of endoscopic third ventriculostomy on the third ventricle floor (arrow).

**Discussion**

More than half of the previous patients showed dementia or mental retardation, attributed to increased ICP. Such cognitive dysfunction improved after treatment using CSF diversion. However, the pathophysiology of cognitive dysfunctions was not clearly identified in the few reports of LOVA. We previously reported 2 cases of secondary NPH associated with disturbance of constructional skill, which improved markedly after shunt surgery. Patients with NPH generally display dysfunction of the frontal lobe causing impaired attention, memory, and language understanding. The right posterior superior temporal-parietal region processes information on global properties, whereas the corresponding left region processes information on local properties. Therefore, we speculate that
patients suffering functional impairment of the frontal and parietal lobes might lose constructional skills.

We use the RBANS for evaluation of neuropsychological function in various types of disorders. RBANS was developed in 1998 and combines 5 functional domains: recent memory, visual spatial and constructional ability, understanding of language, attention, and delayed memory. The RBANS can be administered in a relatively short time, with no learning effect because 2 different battery components are present, and can be evaluated objectively by the scores in the 5 domains. Cognitive dysfunction cannot be clearly documented by simple psychological testing in patients with normal IQ. Therefore, the RBANS is sensitive and useful for evaluating cognitive functions over time.

The present case report suggests that LOVA with normal IQ impairs both frontal and parietal lobe functions, causing dysfunction in visual spatial and constructional skills. Patients showing ventriculomegaly with normal IQ display relatively fewer signs of increased ICP, so detailed neuropsychological evaluations of cognitive function may offer a good indicator for surgical CSF diversion.

References


Address reprint requests to: Hideo Hamada, M.D., Department of Neurosurgery, Faculty of Medicine, University of Toyama, 2630 Sugitani, Toyama 930-0194, Japan.

e-mail: hideo@med.u-toyama.ac.jp