A queduct stenosis is a well recognised cause of hydrocephalus.\(^{17}\) In the adult, the idiopathic variant with a late onset has recently been termed LIAS (late onset idiopathic aqueductal stenosis).\(^{18-21}\) With the reinvention of cerebral endoscopy during the latter decades, the treatment of LIAS has been changed from shunt implantation to endoscopic third ventriculostomy. Mixter undertook the first successful endoscopic third ventriculostomy in 1923.\(^{22}\) Today, based on the development of modern neuroimaging\(^{23-25}\) and endoscope technologies,\(^{26,27}\) endoscopic third ventriculostomy has become the procedure of choice for the treatment of non-communicating forms of hydrocephalus in many centres.\(^{28,29}\) Hopf et al reported a success rate of 83% in patients with aqueduct stenosis, without mortality or permanent morbidity, in a series of 100 consecutive endoscopic third ventriculostomies.\(^{30}\) However, although the clinical features, magnetic resonance imaging (MRI) findings, and clinical response to endoscopic treatment have been reported recently,\(^{31}\) little is known about the cognitive state of LIAS patients.\(^{32}\) Our primary aim in this study was thus to evaluate neuropsychological function in LIAS patients before and after endoscopic third ventriculostomy.

**METHODS**

Between July 1997 and April 2000, 14 patients were diagnosed as having aqueduct stenosis at the Neurosurgery Clinic, University of Innsbruck, Austria. From this group, six subjects (table 1) fulfilled the inclusion criteria for this study, which were as follows:

- no other intracranial pathology apart from hydrocephalus;
- lateral and third ventricles enlarged, with a comparatively small fourth ventricle;
- decreased or absent flow in the aqueductal canal on cross sectional phase contrast MRI for CSF flow, together with morphological signs of obstruction of the aqueduct on sagittal, high resolution gradient echo imaging (3D-MPRAGE, CISS-3D).

None of the patients had had previous surgical treatment for hydrocephalus, and none had a history of meningitis, stroke, epilepsy, or substance abuse. Endoscopic third ventriculostomy was done as described by Hopf et al.\(^{33}\) The operation was carried out with rod lens ventriculosopes (Wolf, Knittlingen, Germany). The outer diameters of the ventriculosopes (elliptical ventriculosope-shaft configuration) were 6 mm and 5.8 x 4.8 mm, respectively. They were equipped with a 2.3 mm optical probe and three or four channels for irrigation, suction, and instrument insertion.

**Abbreviations:** LIAS, late onset idiopathic aqueduct stenosis; MGT, Münchner Gedächtnistest

### Table 1 Patient data, preoperative clinical, and magnetic resonance imaging findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Headache</th>
<th>Gait disturbance</th>
<th>Endocrinological dysfunction</th>
<th>Average duration of symptoms before diagnosis</th>
<th>Papilloedema on fundoscopy</th>
<th>Preoperative TFA</th>
</tr>
</thead>
<tbody>
<tr>
<td>WA</td>
<td>M</td>
<td>25</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>1 month</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>HE</td>
<td>M</td>
<td>28</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>3 months</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>NE</td>
<td>F</td>
<td>37</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>3 months</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>SB</td>
<td>F</td>
<td>34</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>6 months</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>GR</td>
<td>M</td>
<td>58</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>12 months</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>MW</td>
<td>F</td>
<td>60</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>9 months</td>
<td>No</td>
<td>Yes</td>
</tr>
</tbody>
</table>

F, female; M, male; TFA, preoperative transependymal fluid absorption as found on magnetic resonance imaging.
In all patients, MRI was undertaken before endoscopic third ventriculostomy, and postoperatively within the first week and then at 12, and 24 months. Preoperative and postoperative MRI was done on a 1.5 T scanner (Magnetom Vision, Siemens, Erlangen, Germany) using a standard CP head coil. The examination protocol included standard spin echo sequences, cardiac gated cross sectional phase contrast studies and a three dimensional flash sequence (3D-MPRAGE; time of echo (TE) = 4 ms, time of repetition (TR) = 9.7 ms, flip angle (FA) = 40°, slice thickness (SL) = 1 mm) after intravenous administration of contrast agent (Gd-BMA, Nycomed, Vienna, Austria). High resolution CISS-3D imaging was not routinely done. A comparison of preoperative and postoperative MRI was undertaken by an independent neuroradiologist. The preoperative and postoperative clinical status of the patients was assessed by a surgeon who was not involved in the operation (KT). Successful endoscopic third ventriculostomy was defined as:

- evidence for a clear improvement of preoperatively assessed clinical deficits;
- follow up MRI showing flow through the stoma at the third ventricular floor on cross sectional phase contrast studies and diminished ventricular size.

Neuropsychological assessment was done one week preoperatively and on two follow up examinations (mean (SD), 7.5 (9.2) weeks and 81.2 (9.2) weeks after the operation) using standardised psychometric testing procedures for various aspects of memory, attention, visuoconstructive abilities, and executive function (table 2). Testing and evaluation were undertaken by an experienced neuropsychologist. Orientation was tested with the orientation procedures for various aspects of memory, attention, visuoconstructive abilities, and executive function (table 2). Testing and evaluation were undertaken by an experienced neuropsychologist. Orientation was tested with the orientation procedures for various aspects of memory, attention, visuoconstructive abilities, and executive function (table 2). Testing and evaluation were undertaken by an experienced neuropsychologist.

### RESULTS

#### Clinical and MRI findings

Patient data and relevant clinical findings are summarised in table 1.
Frequent preoperative abnormalities were chronic headache (five patients) and gait disturbance (three patients). No patient complained of urinary incontinence. Only two patients (GR and MM) reported problems at work and during everyday activities related to headache, poor memory, or fluctuating attention. Papilloedema was found in one patient, and endocrinological dysfunction in two female patients.

Endoscopic third ventriculostomy was undertaken in all patients without complication. On postoperative MRI, ventricular size diminished in all cases but did not reach normal size, and transependymal cerebrospinal fluid absorption disappeared where previously present. All patients recovered from their preoperative neurological deficits (table 1). After the third ventriculostomy, the two patients who had suffered from amenorrhea-dysmenorrhea syndrome showed normal endocrinological function, resulting in pregnancy six months after surgery in one (NE) and a regular menstrual cycle in the other (SB). With the exception of GR, who continued to experience stress induced headaches, all subjects recovered from chronic headache. Hypersomnia, psychiatric complications, eating disorders, and personality disorders were not observed after the intervention. From MRI and clinical criteria, the endoscopic third ventriculostomy was rated successful in all six subjects.

Neuropsychological findings

<table>
<thead>
<tr>
<th>Cognitive function</th>
<th>Preoperative</th>
<th>Postoperative 1</th>
<th>Postoperative 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>MMSE orientation</td>
<td>10.0</td>
<td>10.0</td>
<td>10.0</td>
</tr>
<tr>
<td>Digit span forward</td>
<td>5.0</td>
<td>6.0</td>
<td>6.0</td>
</tr>
<tr>
<td>Digit span backward</td>
<td>3.3</td>
<td>4.0</td>
<td>4.2</td>
</tr>
<tr>
<td>Münchner Gedächtnistest</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Encoding trial 1</td>
<td>5.3</td>
<td>4.3</td>
<td>7.2</td>
</tr>
<tr>
<td>Encoding trial 5</td>
<td>9.8</td>
<td>13.2</td>
<td>13.4</td>
</tr>
<tr>
<td>Short delay free recall</td>
<td>7.0</td>
<td>11.0</td>
<td>10.2</td>
</tr>
<tr>
<td>Long delay free recall</td>
<td>9.0</td>
<td>11.2</td>
<td>11.0</td>
</tr>
<tr>
<td>Recognition correct</td>
<td>15.0</td>
<td>15.8</td>
<td>14.6</td>
</tr>
<tr>
<td>False positive</td>
<td>1.5</td>
<td>0.3</td>
<td>1.0</td>
</tr>
<tr>
<td>Confabulations</td>
<td>1.8</td>
<td>1.2</td>
<td>1.6</td>
</tr>
<tr>
<td>Intrusions</td>
<td>0.0</td>
<td>0.2</td>
<td>0.2</td>
</tr>
<tr>
<td>Perseverations</td>
<td>3.5</td>
<td>2.5</td>
<td>2.2</td>
</tr>
<tr>
<td>Rey Osterrieth C figure</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Copy</td>
<td>32.2</td>
<td>34.0</td>
<td>34.6</td>
</tr>
<tr>
<td>Short delay free recall</td>
<td>14.6</td>
<td>19.9</td>
<td>24.2</td>
</tr>
<tr>
<td>Long delay free recall</td>
<td>15.0</td>
<td>20.1</td>
<td>22.7</td>
</tr>
<tr>
<td>Category word fluency</td>
<td>18.5</td>
<td>24.6</td>
<td>25.2</td>
</tr>
<tr>
<td>Trail making test A (s)</td>
<td>51.7</td>
<td>32.3</td>
<td>34.8</td>
</tr>
<tr>
<td>Trail making test B (s)</td>
<td>154.8</td>
<td>19.6</td>
<td>103.4</td>
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<tr>
<td>Stroop test</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Reading of colour words (s)</td>
<td>29.6</td>
<td>36.2</td>
<td>31.8</td>
</tr>
<tr>
<td>Naming of colours (s)</td>
<td>45.2</td>
<td>46.8</td>
<td>44.0</td>
</tr>
<tr>
<td>Interference</td>
<td>14.5</td>
<td>73.2</td>
<td>71.8</td>
</tr>
</tbody>
</table>

Table 3 Neuropsychological findings

Postoperative 1 and 2, first and second postoperative examination.

DISCUSSION

Previous studies of LIAS have investigated the long term clinical outcome of patients given shunts during childhood.26 27 28 or the MRI characteristics before and after surgical intervention.29 Endoscopic third ventriculostomy has become a standard treatment for LIAS.27 However, the assessment of neuropsychological function as an indicator of outcome after endoscopic surgery has not been reported previously. In this study we evaluated the clinical, radiological, and cognitive outcome of the endoscopic third ventriculostomy procedure in a small but carefully selected sample of LIAS patients. Endoscopic third ventriculostomy was undertaken following the standard, minimally invasive procedure, from a coronary burr hole (Kocher’s point) through the foramen of Monro,27 without surgical complications. According to the chosen outcome criteria,
Endoscopic third ventriculostomy in aqueduct stenosis

the procedure was considered effective in all the patients. Clinical recovery correlated well with the reduction in ventricular size and the flow through the stoma. Thus postoperative neuropsychological improvement was based on a highly homogeneous group in relation to neurosurgical outcome.

There is a lack of systematic studies of the cognitive impairments found in LIAS patients treated by endoscopic third ventriculostomy. In this study, all LIAS patients showed a preoperative cognitive impairment on standard neuropsychological tests, some of them ranging into the lowest centile scores. The most common problem was an impairment of anterograde memory in combination with frontal-executive and other minor cognitive deficits. As far as can be judged from a relatively small patient group, our investigation suggests that neuropsychological deficits attributable to compression in LIAS patients. The deficit pattern is similar to that found in other hydrocephalic disorders, but is very dissimilar to hydrocephalic dementia, which commonly presents with severe cognitive alterations such as apathy, decreased speed of information processing, severe memory impairment, and impaired ability to manipulate acquired knowledge. Follow up examination after endoscopic third ventriculostomy showed a good recovery of memory and other impairments in five patients, and moderate recovery in one. Because the target sample size was too small to have satisfactory statistical power, this conclusion is based on a subject by subject analysis and a comparison of descriptive statistics. Although care was taken to minimize the impact of practice effects in, for example, the MGT, by using alternate test forms, and despite the fact that follow up intervals were long, test-retest effects cannot be ruled out. However, on the basis of the available information we conclude that endoscopic third ventriculostomy may result in substantial and stable neuropsychological recovery in LIAS patients. This finding needs further confirmation in studies with larger numbers of patients and in case–control studies.

Interestingly, three patients (SB, HE, NE) reported no symptoms apart from headache, and they were unaware of any cognitive deterioration. Preoperative MRI did not show signs of transepidual CSF absorption in these subjects. Thus one could have falsely diagnosed their enlarged ventricles as “arrested hydrocephalus,” a diagnosis for which a surgical intervention may not be considered necessary. However, all three patients were found to have significant improvement in their test scores after surgery. We can therefore conclude that a subgroup of LIAS patients is unaware of any cognitive deficit, and that patients with MRI findings typical for LIAS (although without signs of transepidual CSF absorption) should be routinely screened for memory and executive functions. Overall, endoscopic third ventriculostomy has proved to be an effective, safe, and minimally invasive surgical procedure for the treatment of LIAS patients.

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