HEADACHES, SHUNTS, AND OBSTRUCTIVE SLEEP APNEA:
REPORT OF TWO CASES

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OBJECTIVE: This report describes two shunted patients evaluated with continuous intracranial pressure (ICP) monitors for worsening headaches and subsequently diagnosed with obstructive sleep apnea.

CLINICAL PRESENTATION AND INTERVENTION: ICPs were monitored with strain-gauge sensors inserted into the frontal cortex. After the initial diagnosis of sleep apnea, 8-hour attended polysomnography was performed in each patient. Both patients showed apnea-hypopnea indices greater than 15. Consequently, a "split-night study" was performed to evaluate treatment with titrated nasal continuous positive airway pressure. Patient 1 was a 42-year-old woman (body mass index, 34.1) with a 16-year history of idiopathic intracranial hypertension treated with lumbo-peritoneal and ventriculoperitoneal shunts. Patient 2 was a 20-year-old man (body mass index, 64.4) with the Arnold-Chiari II malformation. The patient had a low-pressure shunt since birth. Neurological examinations were normal or unchanged before evaluation. Neuroophthalmological examinations were normal. Computed tomographic scans failed to show progressive ventriculomegaly. Awake ICPs were less than 15 mm Hg. Nighttime ICPs during rapid eye movement sleep showed multiple Lundberg A waves associated with obstructive sleep apnea and hypoxemia. Blood pressure did not change during these episodes. Polysomnography showed apnea-hypopnea indices of 31 and 41, respectively. Continuous positive airway pressure reduced apnea-hypopnea indices to 17 and 0, respectively; headaches resolved with outpatient therapy.

CONCLUSION: These observations suggest adequate shunting with reduced cerebral compliance in both patients. Altered respiratory mechanics associated with hypoxemia may have triggered cerebral vasodilation and increases in cerebral blood volume, particularly during rapid eye movement sleep. In noncompliant systems, these changes precipitated sustained elevations in ICP and intermittent headaches relieved by continuous positive airway pressure. The clinical patterns also suggest that obstructive sleep apnea should be considered in shunted patients with isolated symptoms of increasing headaches.

KEY WORDS: Hydrocephalus, Shunts, Sleep apnea

The clinical associations of sleep disordered breathing, elevated intracranial pressure (ICP), and various neuropathological conditions have been described previously (1, 3-5, 7, 10, 11, 13, 14, 16, 19, 20), but few investigators have monitored these complex physiological events (2, 8, 12, 18). Only one shunted patient with the Arnold-Chiari II malformation (ACM) has undergone continuous ICP monitoring with polysomnography during sleep apnea (17). This report describes two shunted patients with idiopathic intracranial hypertension (IIH) and the ACM, respectively, who demonstrated periodic elevations in ICP associated with obstructive sleep apnea (OSA).

CASE REPORTS

Patients

Patient 1 was a 42-year-old woman who had undergone lumbo-peritoneal shunting for IIH 16 years previously at another institution. Because of persistent headaches, she underwent insertion of a ventriculoperitoneal shunt several years later, with 13 revisions of the latter shunt. Eight months before this evaluation, a programmable shunt was inserted and set at 140 mm H₂O. The patient was admitted for evaluation of increasing frontal headaches, which she attributed to a shunt malfunction. The headaches were intermittent, were not asso-
Headaches, Shunts, and Sleep Apnea

Figure 1. Tracings showing initial continuous ICP measurements in two shunted patients with OSA. A, ICP elevations in Patient 1 at night. B, ICP elevations in the same patient during daytime naps. Arrows indicate sudden wakening associated with headaches. C, ICP elevations in Patient 2 at night associated with sudden wakening and intermittent headaches. See text for additional clinical details.

ICP Measurement

ICPs were monitored with a silicon strain gauge (MicroSensor; Codman, Raynham, MA) inserted into the frontal cortex opposite to the shunt and connected to a recording module (Model 66; Hewlett Packard, Andover, MA). Mean ICPs and chart records were obtained every 8 hours for 3 to 4 days in each patient.

Polysomnography

After the initial diagnosis of sleep apnea, 8-hour attended polysomnography was performed in each patient. Parameters measured continuously included pulse oximetry, a four-channel electroencephalogram, electro-oculograms, chin and leg electromyograms, electrocardiograms, snoring, nasal and oral airflow, respiratory effort, and sleep position. The apnea-hypopnea index (AHI) was determined as the number of apneas and hypopneas per hour. When the AHI exceeded 15 (normal range, 0-5), patients underwent a “split-night study” to evaluate treatment of OSA using titrated nasal continuous positive airway pressure (CPAP) in the second half of the monitoring period.

Results

Initial ICP Measurements

The initial ICP data for both patients are presented in Figure 1. During the first night, as seen in the top left recording, the patient showed slow increases of ICP to 20 mm Hg, followed by sudden elevations of ICP to more than 40 mm Hg. Mean ICPs for the entire study ranged from 10 to 12 mm Hg. The diagnosis of sleep apnea was considered after her daytime study, as demonstrated on the bottom left recording. Frequent naps followed by abrupt wakening (arrows) were associated with headaches and ICP elevations above 25 mm Hg. Nighttime studies for Patient 2 are shown on the right. Mean ICPs ranged from 8 to 12 mm Hg. Abrupt elevations of ICP were associated with apneic spells, sudden wakening, and intermittent headaches. There were no consistent changes in blood pressure during the episodes of ICP elevations for both patients.

Polysomnography

Summaries of the results of the sleep studies in both patients are demonstrated in Table 1. The patients showed baseline elevations of the AHI to 31 and 41, respectively, with

| TABLE 1. Baseline and continuous positive airway pressure titrated apnea-hypopnea indices and pulse oximetry during attended polysomnography in two shunted patients with sleep apnea* |
|---------------------|---------------------|---------------------|
| Patient 1           | Patient 2           |
|---------------------|---------------------|---------------------|
| Baseline            |                      |                      |
| AHI                 | 31                   | 41                   |
| AHI (REM)           | 11.3                 | 46                   |
| SpO₂ (%)            | 85–95                | 71–92                |
| CPAP                | 11 cm H₂O            | 9 cm H₂O             |
| AHI                 | 18                   | 0                    |
| AHI (REM)           | 24.2                 | 0                    |
| SpO₂ (%)            | 95                   | 95                   |

* AHI, apnea-hypopnea index; REM, rapid eye movement; SpO₂, oxygen saturation as measured by pulse oximetry.
AHIs during rapid eye movement (REM) sleep of 11.3 and 46. The arterial desaturation for Patient 1 ranged from 85 to 95%, and that for Patient 2 ranged from 71 to 92%. Spontaneous elevations of ICP to more than 20 mm Hg did not occur. All elevations to more than 20 mm Hg occurred with oxygen desaturation related to apnea, and virtually all elevations occurred during REM sleep.

Both patients showed prolonged sleep latencies with increased wakefulness and decreased REM sleep, particularly Patient 1. More than 95% of the events were hypopneas or obstructive apneas; Patient 2 showed worsened apnea in the supine position. Electrocardiograms showed normal sinus rhythm with occasional bradycardia, but this was not consistently associated with increased ICP. Additional measurements, although consistent with OSA, showed no significant associations with elevated ICP.

The results of CPAP titration to 11 and 9 cm H₂O, respectively, are shown in Table 1. Both patients showed marked reductions in AHIs, whereas Patient 1 showed a compensatory increase in REM sleep and a slight increase in the AHI. ICP elevations to more than 20 mm Hg were eliminated. Both patients reported relief of headaches with CPAP therapy.

ICP Patterns

A reconstruction of a typical ICP pattern in both patients is shown in Figure 2. Concurrent with oxygen desaturation (top line), periodic pressure waves (solid line) increased to 40 to 50 mm Hg, with a decrease to baseline as the desaturation improved. The waves corresponded to Lundberg B waves and lasted approximately 1 to 2 minutes; however, approximately 75% of the waves above 20 mm Hg continued beyond the period of desaturation (dotted line and arrow) for 7 to 20 minutes and were best described as Lundberg A waves. These longer waves occurred during REM sleep and did not correlate with the extent of oxygen desaturation.

Follow-up

The patients have been followed up for 20 and 15 months, respectively. Patient 1 has been compliant with the use of CPAP at home and has reported no further headaches, uninterrupted nighttime sleep, and infrequent daytime napping. Patient 2 has not been compliant but reported significantly fewer headaches when CPAP was used. Both patients have continued activities of daily life without shunt revisions.

DISCUSSION

Five investigations in 28 patients have monitored ICP and polysomnography in patients with isolated OSA or associated with various neurological conditions (Table 2). Nine of these patients had isolated OSA; 17 adults were monitored for evaluation of IHH or hydrocephalus after trauma, subarachnoid hemorrhage, or meningitis; 1 unshunted patient had cutaneous hemangiomas with ventriculomegaly; and 1 patient had been shunted for the ACM. Given the diverse pathogeneses and technical methods and reportage, data are difficult to extrapolate; nevertheless, some patterns are distinguishable.

The mean ICP was less than 15 mm Hg in most patients, although three patients reported by Jensen and Bergersen (8) using an epidural catheter had morning ICPs between 20 and 30 mm Hg, with evening elevations to 22 mm Hg. In the 17 patients reported by Kuchiwaki et al. (12) using an epidural catheter, 15 had transient ICPs greater than 15 mm Hg, although only 5 showed baseline levels greater than 15 mm Hg when B waves occurred. Most studies report increased prevalence of B waves during REM sleep; most of these waves were similar to "ramp"-type B waves described by Krauss et al. (10) in 14 shunted adult patients with normal-pressure hydrocephalus but without sleep apnea. A waves were described infrequently but usually appeared during REM sleep. All studies emphasized that pressure waves were associated with oxygen desaturation and rarely occurred spontaneously. The shunted patient described by Pastelekamp et al. (17) showed elevation of the mean arterial pressure associated with OSA and increased ICP, whereas Jensen and Bergersen (8) described increases in systolic and diastolic blood pressure variations correlated with variations in ICP elevations.

The patients in our series conformed to a pattern seen in the majority of patients in the previous studies with baseline ICPs lower than 15 mm Hg and short "ramp"-type B waves predominantly during REM sleep, occasionally higher than 20 mm Hg, associated with oxygen desaturation and minimal autonomic changes. However, the predominance of longer A waves during REM sleep in our two shunted patients was not a common pattern in the previous studies. These observations may be explained by assuming functioning shunts in noncompliant systems. OSA, particularly during periods of altered
TABLE 2: Investigations of intracranial pressure and polysomnography in patients with obstructive sleep apnea

<table>
<thead>
<tr>
<th>Series (ref. no.)</th>
<th>No. of patients</th>
<th>Patient characteristics</th>
<th>Baseline ICP</th>
<th>B waves</th>
<th>A waves</th>
<th>Correlation with O₂ desaturation</th>
<th>Autonomic changes</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sugita et al., 1985 (18)</td>
<td>3</td>
<td>SAHS</td>
<td>~10 mm Hg</td>
<td>REM &gt; +nREM</td>
<td>−</td>
<td>+</td>
<td>−</td>
<td></td>
</tr>
<tr>
<td>Kuchiwacki et al., 1988 (12)</td>
<td>17</td>
<td>Ventriculomegaly</td>
<td>&gt;15 mm Hg in 5 patients</td>
<td>nREM</td>
<td>Occasionally in 3 patients</td>
<td>+</td>
<td>−</td>
<td></td>
</tr>
<tr>
<td>Jernum and Borgesen, 1989 (8)</td>
<td>6</td>
<td>Isolated OSA</td>
<td>&gt;20 mm Hg in 3 patients</td>
<td>nREM + REM</td>
<td>REM</td>
<td>+</td>
<td>↑ Systolic and diastolic pressure variations</td>
<td></td>
</tr>
<tr>
<td>Pasterkamp et al., 1989 (17)</td>
<td>1</td>
<td>Shunted ACM</td>
<td>&lt;10 mm Hg Entire study</td>
<td>?</td>
<td>REM</td>
<td>+</td>
<td>↑ Mean arterial pressure</td>
<td></td>
</tr>
<tr>
<td>Becker et al., 1994 (21)</td>
<td>1</td>
<td>Cutaneous hemangioma + ventriculomegaly</td>
<td>&lt;10 mm Hg</td>
<td>?</td>
<td>REM</td>
<td>+</td>
<td>−</td>
<td></td>
</tr>
<tr>
<td>Hanigan and Zallek, present work</td>
<td>2</td>
<td>Shunted ACM and IIIH</td>
<td>8–12 mm Hg</td>
<td>Usually REM</td>
<td>REM</td>
<td>+</td>
<td>−</td>
<td></td>
</tr>
</tbody>
</table>

*ICP, intracranial pressure; SAHS, sleep apnea hypoventilation syndrome; OSA, obstructive sleep apnea; ACM, Arnold-Chiari II malformation; IIIH, idiopathic intracranial hypertension; REM, rapid eye movement; nREM, no REM; CPAP, continuous positive airway pressure; +, positive; −, negative; f, data not given.

cerebral blood flow such as REM sleep (4, 5, 10, 20), result in varying degrees of hypoxemia, possible CO₂ retention with cerebrovascular vasodilation, increases in cerebral blood volume, and elevations in ICP. Compliant systems seen in patients with isolated OSA or without long-term shunts show Lundberg B waves that are short in duration and correlated with the duration of the apneic events. Noncompliant systems show a higher percentage of A waves that are not correlated with the severity of the event. Functioning shunts act to reduce the ICP and restrict an A wave, thus obviating the need for reflex autonomic compensation to increase the cerebral perfusion pressure.

Our explanation for these observations is speculative and cannot be corroborated by the earlier heterogeneous investigations. Precise clinical relationships among sleep disorders, elevated ICP, and neurological conditions such as normal-pressure hydrocephalus (11, 12, 16) or, as in our Patient 1 with IIIH (13, 14), are not clear, whereas sleep disordered breathing in patients with the ACM, as in our Patient 2, has been attributed to several mechanisms related to central nervous system structural abnormalities and treated by various interventions (1, 3, 7, 17, 19). Descriptive physiological data such as the presence of Lundberg A waves are generally accepted as manifestations of intracranial pathology, but their association with normal sleep or sleep disorders remains to be explained (4–6). Similarly, the pathogenesis of Lundberg B waves in patients with sleep disorders and elevated ICP has been related to apnea (8), hypoxemia or hypercarbia (18), altered brainstem function (12), or normal sleep (15). This lack of clinical and physiological concordance reinforces the successful treatment in both our patients, indicating that OSA can occur independently of any preexisting intracranial abnormalities. Seventeen patients with ventriculomegaly caused by idiopathic adult hydrocephalus and various sleep disorders reported by Kristensen et al. (11) failed to improve after lumbar drainage, whereas the patient reported by Pasterkamp et al. (17) with a functioning shunt and the patient described by Becker et al. (2) with ventriculomegaly improved after measures to alleviate OSA. The beneficial effect of CPAP on restorative sleep may have improved the patients’ symptoms, but the lack of spontaneous ICP elevations and the direct association among headache, elevated ICP, and OSA throughout the monitoring periods suggest that specific treatment prevented the clinical and physiological consequences of OSA. Thus, long-term shunted patients may or may not develop OSA. If they do, they can develop headaches associated with significant pressure elevations as seen by Lundberg A waves and require a functioning shunt system to control the ICP until the OSA is treated appropriately.

Finally, our patients remind clinicians to maintain a high index of diagnostic suspicion in shunted patients with headaches. Recently, Kirk et al. (9) showed that sleep-disordered breathing occurs in 20% of children with the ACM, but fewer than half of this group had undergone polysomnography. Both our patients, like the patient described by Pasterkamp et al. (17), were undiagnosed and admitted for evaluation of shunt function. In retrospect, Patient 1 related complaints of sleep disruption and frequent daytime naps that suggested
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OSA, whereas Patient 2 had a family history of obesity and OSA. With an unambiguous history, shunted patients may not require continuous ICP monitoring. However, if patients undergo monitoring, polysomnography including continuous measurements of ICP and blood pressure will solve the clinical dilemma. Although not essential to the diagnosis, monitoring of the carbon dioxide levels and cerebral blood flows may help to explain the physiological mechanisms of this interesting and perhaps underdiagnosed problem.

REFERENCES


Acknowledgment

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COMMENTS

Headaches that occur after shunting are often a considerable challenge for patient and neurosurgeon alike. The authors provide a helpful reminder of one of the causes of such headaches, namely, obstructive sleep apnea. It would be useful if the pattern of headaches in both patients were described in more detail, particularly with regard to provocative factors. With regard to the intracranial pressure (ICP) wave patterns, by looking more carefully at the relative time courses, is it possible to say whether the oxygen desaturation preceded, exactly coincided with, or slightly followed a change in ICP? Lundberg A waves have conventionally been described as exceeding 50 mm Hg for more than 5 minutes.

This careful study highlights yet again the need to consider multimodality monitoring to accompany ICP monitoring (1). Intracranial recordings have to be presented with the appropriate time scale if the clinician is to recognize and distinguish artifact from genuine ICP waves. There is still no good way to annotate the record to take account of clinical events.

John D. Pickard
Cambridge, England


This is an interesting report of two patients with ventriculoperitoneal shunts and small cerebral ventricles who were evaluated for worsening headaches by means of simultaneous ICP and polysomnographic monitoring. The workup revealed awake ICPs below 15 mm Hg. During rapid eye movement sleep, abrupt elevations of ICP above 25 mm Hg were observed in association with Lundberg A waves, apneic spells, and sudden wakening. Polysomnography confirmed the presence of obstructive sleep apnea with apnea-hypopnea indices that responded dramatically to continuous positive airway pressure. Headaches in both patients were managed effectively with therapy on an outpatient basis.

The value of this article is that it calls attention to the putative association of obstructive sleep apnea and idiopathic headache in patients with functioning ventriculoperitoneal shunts. More work will be necessary to confirm this connection and to better understand the underlying physiological mechanisms.

Thomas H. Milhorat
Manhasset, New York
This is an interesting case report describing two patients with the combination of shunted hydrocephalus and obstructive sleep apnea. Continuous ICP monitoring and polysomnography revealed elevations in ICP occurring primarily in association with obstructive, apneic episodes. Elevations in ICP and headaches were reportedly eliminated with continuous positive airway pressure therapy. Diagnosing and managing headaches in shunted patients can be challenging and frustrating. As the authors suggest, risk factors for or indications of obstructive sleep apnea should be part of the routine questioning of shunted hydrocephalic patients with refractory headaches.

Marvin Bergsneider
Daniel F. Kelly
Los Angeles, California

The android Maria and the mad scientist Rotwang in Fritz Lang's film Metropolis (1926). The machine assumes a discernibly human shape and form. This film provided an early forum for contemplating the possible replacement of humans by robots.