NEW RESEARCH

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Development of a Short Sleeper Phenotype after Third Ventriculostomy in a Patient with Ependymal Cysts

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CASE REPORTS

A naturally short sleeper phenotype with a sleep need of less than 6 hours without negative impact on health or performance is rare. We present a case of an acquired short sleeper phenotype after third ventriculostomy.

A 59-year-old patient suffering from chronic hydrocephalus reported an average of 7-8 h of nocturnal sleep. After surgical intervention, the patient noted a strikingly reduced sleep need of 4-5 h without consequent fatigue or excessive daytime sleepiness, but with good daytime performance and well-balanced mood. Short sleep per 24 hours was confirmed by actigraphy. Postoperative imaging revealed decreased pressure around the anterior third ventricle.

The temporal link between development of a short sleeper phenotype and third ventriculostomy is striking. This might suggest that individual short sleep need is not only determined by genetics but can be also be induced by external factors.

Keywords: Short sleeper, sleep regulation, ventriculostomy

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A population-based study of sleep habits in 110,441 non-institutionalized US adults shows an average self-reported sleep duration longer than 6 hours in more than 90%.[1] Habitual sleep shorter than 6 hours per day is associated with unfavorable health and performance status.[2] However, there is a small population of naturally short sleepers with a daily sleep need less than 6 hours without any subjective or objective consequences. Familial accumulation of a short sleeper phenotype led to the discovery of genes involved in the regulation of individual sleep need.[3] We present a case of a patient developing short sleeper phenotype after third ventriculostomy.

REPORT OF CASE

A 59-year-old man with a history of vertigo and gait unsteadiness for about 2 years was diagnosed with chronic internal hydrocephalus. Cranial MRI showed several cystic formations in the thalamic, hypothalamic, and mesencephalic region (**Figure 1A**, **1B**). Lumbar puncture revealed cerebrospinal fluid pressure of 21-22 mm Hg within the upper normal range and increased protein level (680 mg/L), but no signs of inflammation. Detailed serologic analyses including echinococcosis and other parasitic infections were negative. Eventually the MRI pattern was interpreted as congenital ectopic ependymal cysts leading to chronic hydrocephalus and increased protein in the context of a presumably reduced CSF volume turnover in the context of hydrocephalus.

An endoscopic third ventriculostomy was performed due to progression of gait disturbance. Neuronavigation allowed performing a right frontal trajectory to the widened floor of the third ventricle posterior to the hypophysial recess and anterior to the basilar artery (**Figure 1C**). Postoperative imaging showed signs of decreased pressure both supratentorially (**Figure 1C**) and locally around the anterior third ventricle (**Figure 1D**). A biopsy of the cystic formations was not performed to conserve critical structures.

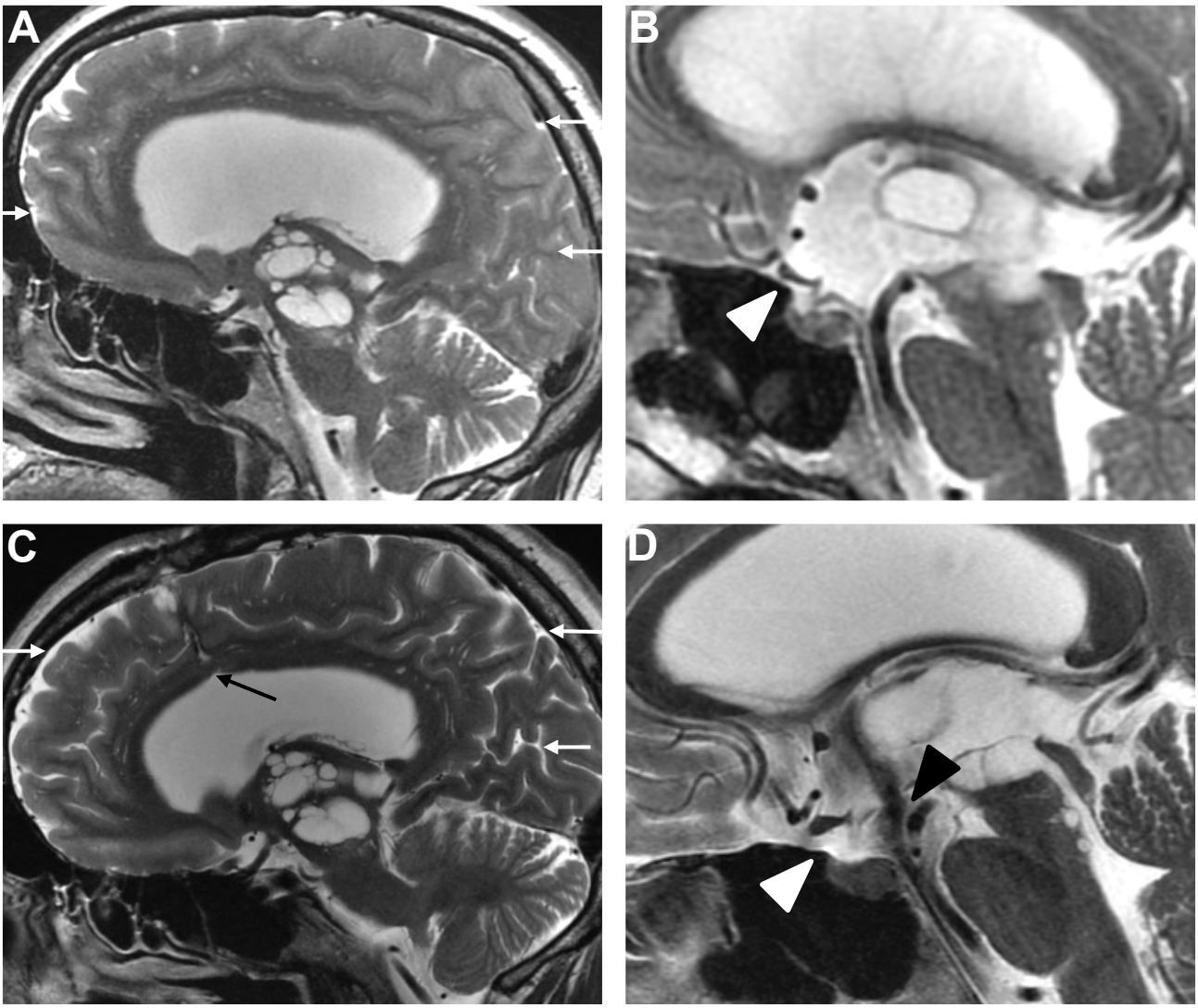
After intervention, vertigo and gait disturbance remitted and neurological examination was normal. However, the patient reported that he was unable to sleep more than 4-5 h per night. In the family, no short sleepers were known. Before third ventriculostomy, he had slept 7-8 h per 24 h since early adulthood without complaining of excessive daytime sleepiness. Interestingly, any negative effects of short sleep on physical or cognitive performance were denied and confirmed by neuropsychological testing. In contrast, a remarkable postoperative improvement of a formerly depressed mood was noted by both the patient and his relatives. Signs of fatigue or excessive daytime sleepiness were absent (fatigue severity scale 2.1/7, Epworth sleepiness scale 9/24). Short time-in-bed of an average 5 h was confirmed by 17-day-actigraphy, with signs of a slightly irregular sleep-wake rhythm. Multiple sleep latency test was normal, with an average sleep latency of 16.5 minutes. In addition, the psychomotor vigilance task showed normal performance regarding reaction time, accuracy, and variability.

The short sleeper phenotype persisted in follow-up visits up to at least 17 months when the patient had his last follow-up consultation.

DISCUSSION

The phenotype of short sleepers is commonly described either as natural congenital state without impairment of health or performance status or as a habitual state with unfavorable subjective and objective health consequences. In comparison to

Figure 1—Cranial MRI imaging before and after the third ventriculostomy.



T2-weighted images before **(A, B)** and after surgery **(C, D)** showing cystic formations in the region of thalamus, hypothalamus and mesencephalon as well as the path of the right frontal ventriculostomy **(C)** (black arrow). The subarachnoid space in general (white arrows) **(A, C)** and especially between skull base and third ventricular margins, e.g., chiasmatic or hypophysial recess (white arrowheads) **(B, D)** is widened postoperatively **(C)**. Cerebrospinal fluid flow from the third ventricle to the prepontine cistern can be visualized by black jet phenomenon at the floor of the third ventricle **(D)** (black arrowhead).

long sleepers, short sleepers exhibit enhanced slow wave sleep activity without differences in homeostatic sleep regulatory mechanisms.[4] Further, melatonin and cortisol levels as well as body temperature in short sleepers indicate a shorter biological duration of nocturnal sleep in contrast to long sleepers.[5] Others found that a mutation in a transcriptional repressor (hDEC2-P385R) is correlated with short sleeper phenotype in humans and mice.[3]

To our knowledge, development of a short sleeper phenotype by external circumstances or a surgical intervention has not been described before. In this line, the timely association with third ventriculostomy in this clinical case is highly suggestive of a causal link. The alleviation of the preexisting condition of hydrocephalus might explain a reversion of symptomatic hypersomnia to normal sleep duration of 6 to 8 hours, but not to a sleep length less than 5 hours. Similarly, a hypomaniac psychiatric condition could be considered to transiently reduce sleep length. However, the very low sleep duration as well the long and stable course of both reduced sleep need and the persistent well-balanced mood favors the hypothesis of a causal link between the intervention and the short sleeper phenotype. Interestingly, a hypomaniac phenotype was found in naturally short sleepers compared to respective control subjects in matched-control study.[6]

Still, the underlying mechanisms remain highly speculative. Beyond presumably lowered overall supratentorial brain pressure, thalamic and hypothalamic structures near the intervention site might be responsible for the changed regulation of sleep homeostasis and circadian regulation. Due to their exposed location and the proximity to the third ventricle, decompression of the wake-promoting histaminergic neurons in the tuberomammillary nucleus might be involved or alternatively altered expression of genes regulating circadian rhythm and sleep homeostasis. Further studies in animal models and

human cohorts are warranted to validate the correlation of third ventriculostomy with a short sleeper phenotype.

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