Narcolepsy (기면증)

기면증의 증상

1) 심한 주간 졸림
거의 모든 환자들은 밤잠을 충분히 잤음에도 낮에 심하게 졸린다고 호소하며 일상생활을 하다가 갑자기 잠에 빠져버리는 수면발작을 종종 경험

2) 탈력발작
웃거나 울거나 화를 냄, 기뻐함 좋아함 등의 감정 변화가 있을 때 몸의 전체 또는 일부의 힘이 갑자기 없어지는 현상으로 서 있다가 쓰러지거나 무릎이 갑자기 풀리거나 턱이나 얼굴 근육의 힘이 빠지는 증상입니다. 지속시간은 수 초에서 수 분 내로 짧으며 곧 완전하게 회복됩니다. 의식은 유지되며 다 기억할 수 있습니다.

3) 수면마비
가위눌림으로 알려진 현상으로 잠이 들 때나 깰 때, 수 초에서 수 분간 움직일 수 없는 상태를 말합니다. 이 때 환자의 정신은 깨어 있지만, 사지를 움직일 수 없게 되어 불안과 공포심을 느끼게 되고 무서운 환상이 보이기도 합니다.

4) 입면/탈면 시 환각
잠이 들려고 할 때 또는 잠에서 깨려고 하는 중간단계에서 꿈이 현실로 이행되거나 환상이 보이거나 환청이 들리며 이상한 감각이 느껴지기도 합니다.

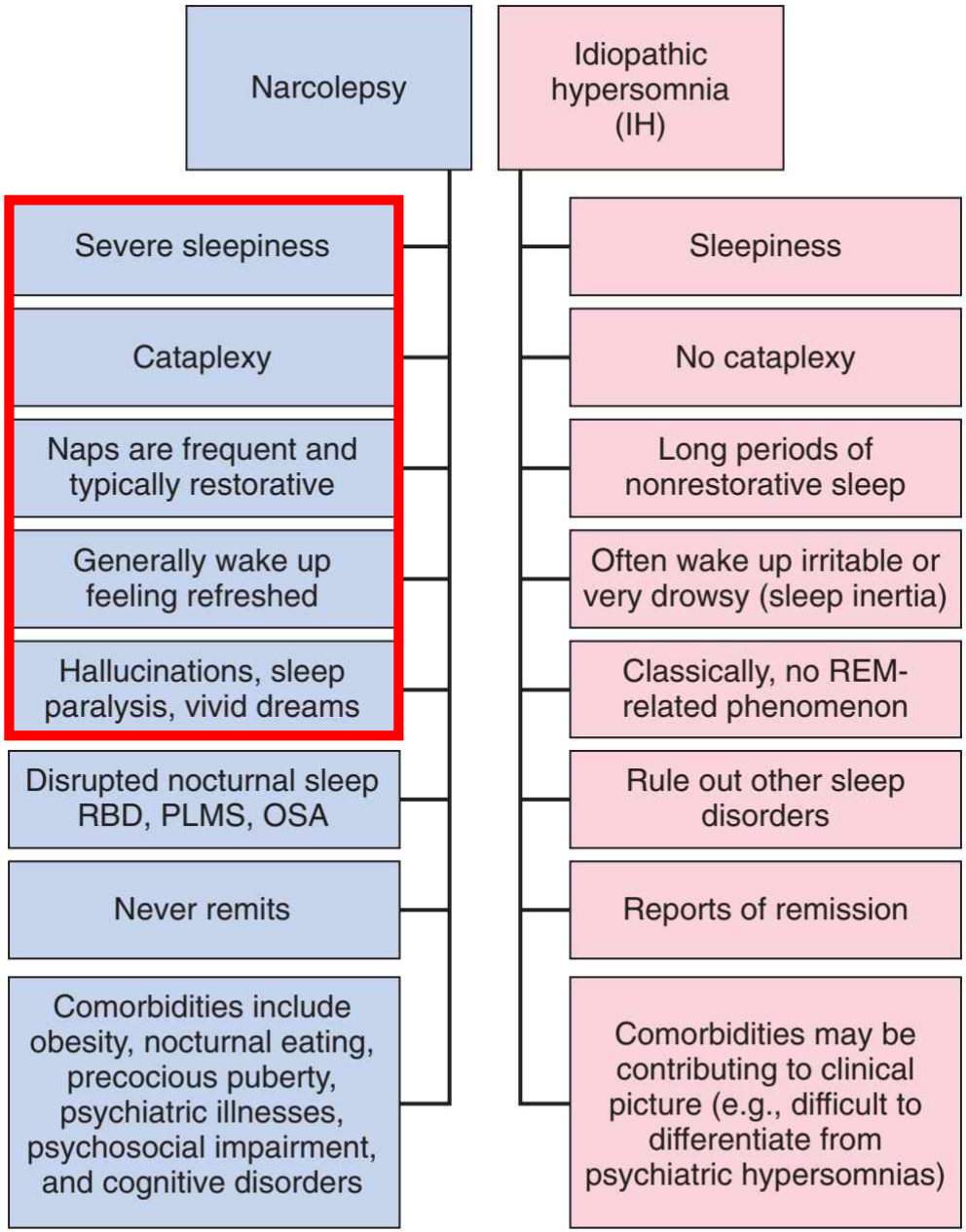
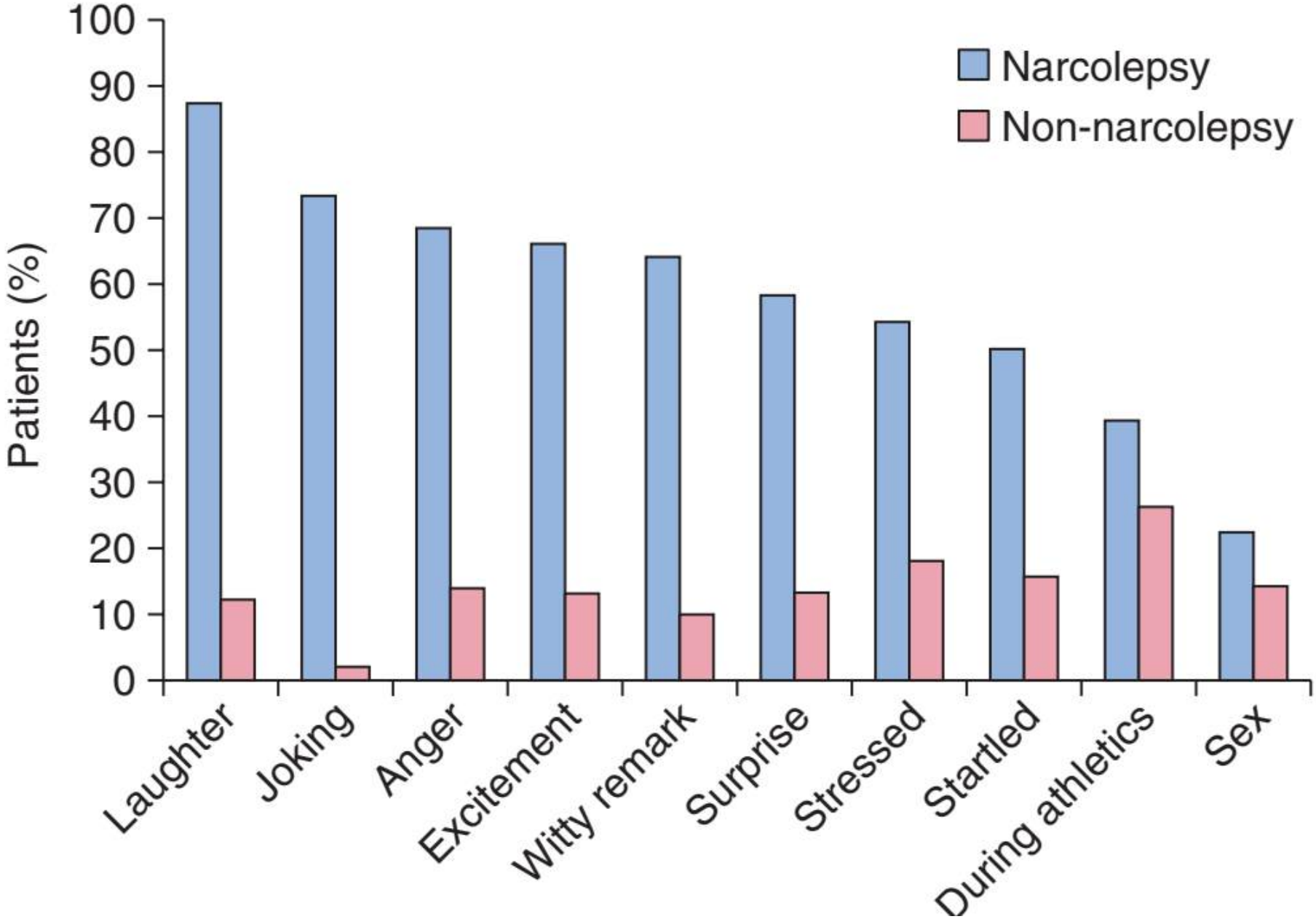


FIGURE 11.1-1. Comparison of narcolepsy and idiopathic hypersomnia. Narcolepsy can be described as a "pentad of symptoms" that includes sleepiness, cataplexy, sleep paralysis, hallucinations, and disrupted nocturnal sleep; it is uncommon for a patient to manifest all symptoms. RBD, rapid eye movement (REM) sleep behavior disorder; PLMS, periodic limb movements in sleep; OSA, obstructive sleep apnea.

Cataplexy가 발생되는 상황




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Narcolepsy with Cataplexy (탄력발작)



Narcolepsy with Cataplexy (탄력발작)



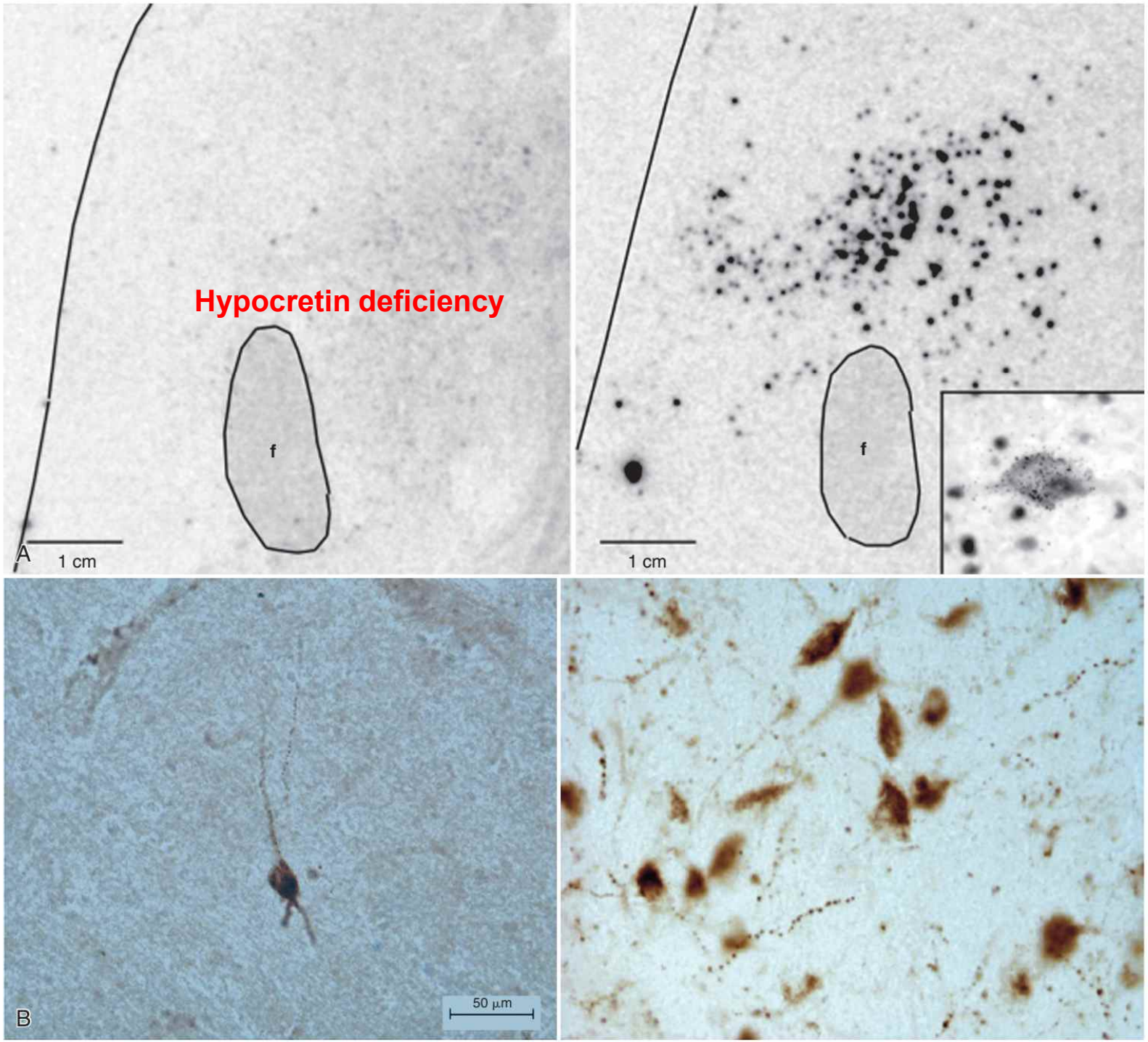


FIGURE 11.1-5. Hypocretin peptides and neurons in the lateral hypothalamus of narcoleptics versus controls and cerebrospinal fluid hypocretin levels in central nervous system hypersomnias and other conditions. A, Dramatic reduction of hypocretin mRNA (gene expression) in the lateral hypothalamus in a narcoleptic (*left*) versus a control brain (*right*). **B,** Dramatic reduction of hypocretin-stained peptides (hypocretin cells) in the lateral hypothalamus in a narcoleptic (*left*) versus a control brain (*right*). Narcoleptics have an 85% to 95% reduction in the number of hypocretin neurons. f, fornix.

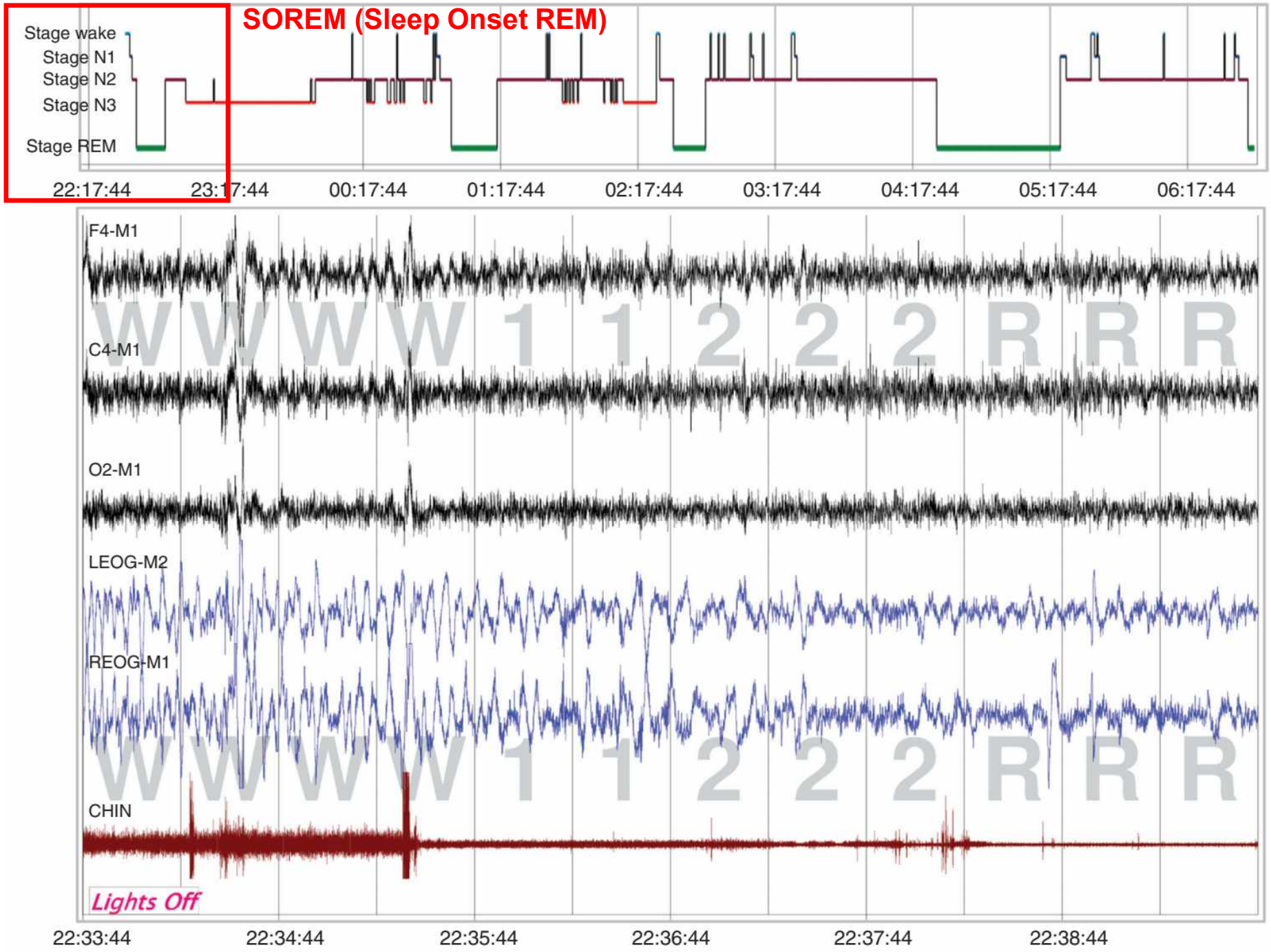
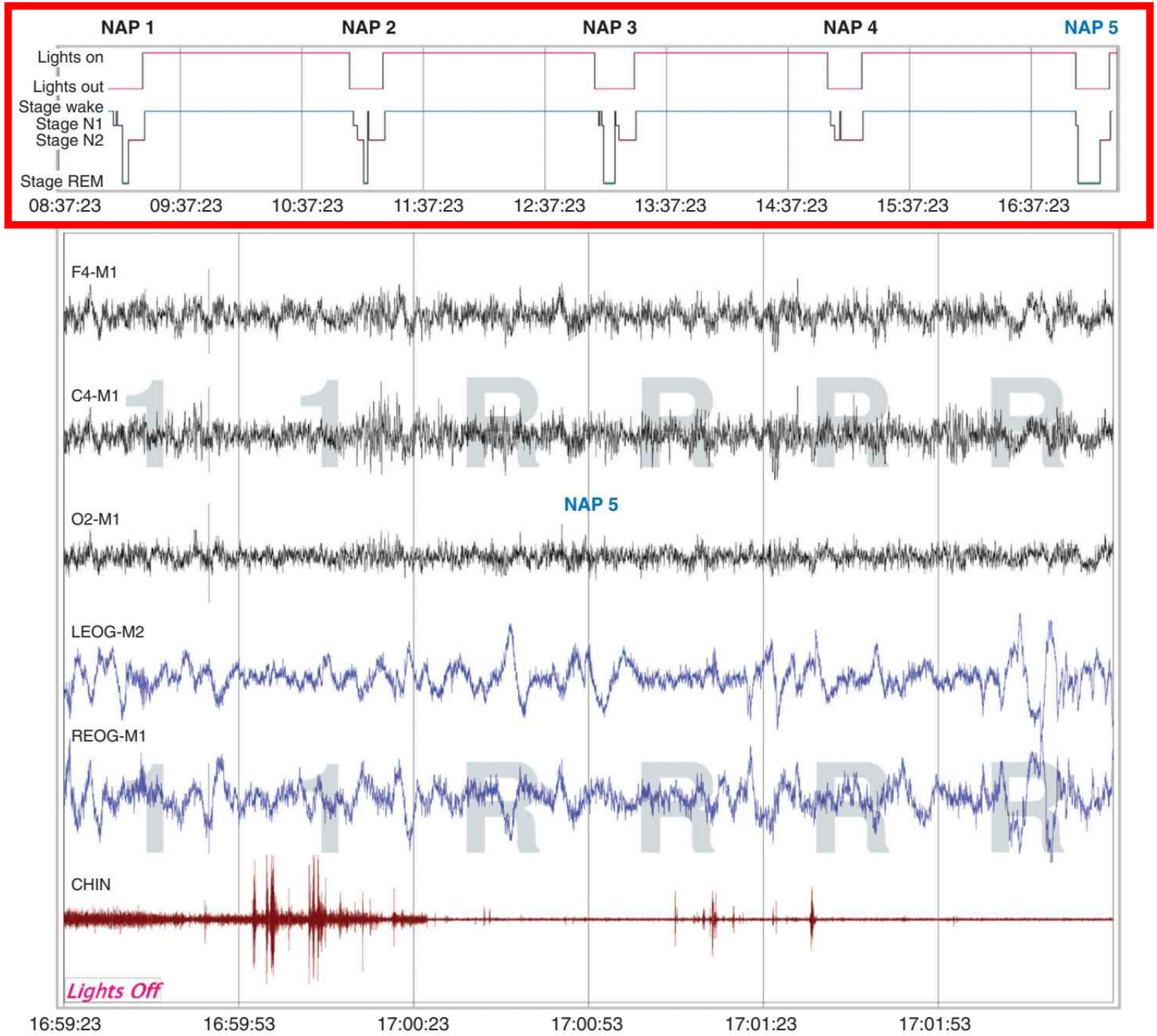


FIGURE 11.1-11. A sleep-onset rapid eye movement period during an overnight polysomnogram performed in a patient with excessive daytime sleepiness and cataplexy. The sleep hypnogram (*top*) and selected fragments from epochs of the recording (*bottom*), starting from "lights out," demonstrate the rapid onset of rapid eye movement (REM) sleep. A normal REM latency is typically defined as 90 to 120 minutes; in this recording, it is 2.5 minutes. A REM latency of 15 minutes or less is diagnostic for narcolepsy.



MSLT test
- SOREM이 2회 이상
- Mean Latency <8 min

FIGURE 11.1-12. A multiple sleep latency test (MSLT) diagnostic for narcolepsy from the patient shown in Figure 11.1-11. The sleep hypnogram (*top*) shows that the patient attained rapid eye movement (REM) sleep on four of the five naps. Selected epochs from the NAP 5 recording (*bottom*), starting from "lights out," show the patient immediately falling asleep (sleep latency, 0 min) and entering REM sleep 1 minute later (REM latency, 1 min). The calculated mean sleep latency of all five naps was 1.6 minutes. Given the MSLT findings and sleep onset REM period (SOREMP) on the polysomnogram, the patient was diagnosed with narcolepsy with cataplexy.

- **<u>기면증의 진단</u>**

  → 주간의 졸리움

  → 탄력발작 (Cataplexy)

  → Hypocretin 저하 (뇌척수액)

  → 야간 수면검사 (SOREM <15 min, 또는 MSLT 평균 latency < 8 min 과 2회 이상의 SOREMs