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Hypnogenic Paroxysmal Dystonia: Epileptic Seizure or a New Syndrome?

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Summary: Five patients between the ages of 7 and 74 years presented with nocturnal episodes characterized by coarse, often violent movements of the limbs and by a tonic phase of variable duration. Seizures recurred every night or almost every night during slow wave (NREM) sleep and were not associated with electroencephalographic (EEG) abnormalities. Interictal EEGs were normal during both sleep and wakefulness. Carbamazepine treatment was effective in all patients. Uniform clinical behavior and EEG patterns indicate a distinct nosological entity whose pathophysiology is not yet understood. **Key Words:** Sleep—Paroxysmal dystonia.

Boller et al. (1975) reported nightmares that appeared in a 65-year-old man following temporal lobe infarction and which subsided after phenytoin administration. Pedley and Guilleminault (1977) observed an unusual type of somnambulism in 6 patients ranging in age from 17 to 32 years; all experienced episodes characterized by screaming, vocalization, complex automatisms, and ambulation. The episodes ceased after phenytoin or carbamazepine treatment. Four of these patients demonstrated epileptiform abnormalities in their electroencephalograms (EEGs). On the basis of these abnormalities and the favorable response to therapy, these authors suggested that this syndrome represents an atypical form of epilepsy, despite the fact that polygraphic recordings of two abortive attacks failed to correlate with any paroxysmal or other abnormal electrical activity.

We report here on 5 patients who were observed during nocturnal episodes characterized by tonic spasms and violent movements during sleep. These episodes occurred repeatedly almost every night with no concomitant EEG signs of epilepsy.

EXPERIMENTAL PROCEDURES

Observations

The 5 patients all gave a history of attacks of "agitation" during sleep almost

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TABLE 1. *Personal and family histories of the 5 patients*

History variable	Patient 1	2	3	4	5
Sex	F	F	M	M	M
Age	7	10	37	40	74
Family history	—	Somnambulism and unclassified seizures (mother)	—	—	—
Neonatal asphyxia	—	—	—	—	—
Onset of nocturnal seizures (age)	5	7	30	26	40
Frequency of nocturnal seizures	Every night	Every night	Every night	25 out of 30 nights	25 out of 30 nights
Number of seizures per night	4–7	3–4	1–2	4–20	2–3
Number of diurnal seizures during wakefulness	—	1	1 in 3 months	1 in 20 days	—
Previous seizures suggestive of epilepsy	+	—	+	—	—
Neurological examination	—	—	—	—	—
Neuropsychologic testing	Normal	Normal	Normal	Normal	Normal

every night for many years. Only 2 patients reported similar episodes occurring during the day. Personal and family histories are summarized in Table 1.

Methods

Each patient underwent a full daytime EEG evaluation, and an all-night polygraphic monitoring procedure with not only the classic monitoring for sleep-wake scoring, but also with full EEG investigation using the 10–20 International system. A closed-circuit audiovisual system with silicon cameras allowed simultaneous videotape recordings of the patients' behavior and EEGs.

Analysis of the recordings and video tapes prior to and during seizures was carried out by two separate research groups and the results were compared.

TABLE 2. *Summary of polygraphic data*

Polygraphic variable	Patient 1	2	3	4	5
Number of recorded seizures	2	5	2	8	1
Onset	Stage 3–4	Stage 3–4	Stage 2	Stage 2	Stage 3–4
Duration (sec)	15–24	30–35	25–28	30–45	40
EEG during wakefulness	Normal	Normal	Normal	Normal	Normal
EEG during sleep	Normal	Normal	Normal	Normal	Normal
EEG during episodes	Negative	Negative	Negative	Negative	Negative

Note: Seizures were clinically similar in all patients.

RESULTS

Polygraphic data are summarized in Table 2.

Case Observations

Patient 1, 7 years old (Figs. 1 and 2)

While sleeping on her left side during stage 3–4 non-rapid eye movement (NREM) sleep (Fig. 1-1), the patient suddenly turns her head to the right and tries to sit up, propping herself up on her elbows (2), but then falls back and stiffens. The limbs are abducted, the eyes fixed, the head slightly turned to the right (3). Weak repetitive movements of the trunk are observed, as if the girl were trying to free herself from her state of immobilization and stiffness. After 15 sec, the attack subsides abruptly. The patient relaxes and lies in bed, alert and motionless (4). She is unable to give an account of the episode.

Patient 2, 10 years old (Figs. 3 and 4)

During stage 3–4 sleep, the patient appears to wake up and makes small automatic movements (Fig. 3-1). After a few seconds, her trunk begins to turn slightly. The legs are abducted and rhythmically lifted, the right arm extended, and the left forearm and wrist flexed. The right shoulder is rotated and follows the movements of the trunk and legs (2–4). Eight seconds later, the patient stiffens in a tonic contraction. Both legs are rigid and extended, the right arm is also extended, and the left arm flexed. Opisthotonus is present and the head is slightly rotated to the right. The eyes are wide open and fixed in a stare that does not betray loss of consciousness (5). After 25 sec, the tonic contraction disappears and the patient tries to sit up and wrap the sheets about her. She is fully conscious and remembers having "a fit" but is unable to give details. Tachycardia is present at the onset of the episode; the tonic phase is accompanied by bradycardia and apnea.

Patient 3, 37 years old (Figs. 5 and 6)

The episode occurs during stage 2 sleep. The patient starts, moves under the sheets, turns his body slightly to the right, and then stiffens. The right arm is flexed and lifted behind the head, the left arm extended, and the legs slightly abducted. After 8 sec, he utters a prolonged cry and makes several coarse, slow movements of the limbs. The attack subsides 25 sec later; the patient relaxes and swears twice. He is lucid, excited, and panting as after an effort.

Patient 4, 40 years old

Just before the episode, the EEG shows a lightening of stage 2 sleep lasting 4–5 sec. The patient suddenly tries to sit up in bed, but is restrained by the collodion electrodes. He falls back and begins to toss about with coarse, spasmodic movements of all extremities accompanied by grimacing and head rotation in various directions. After 40 sec, he relaxes. He appears exhausted and confused and fumbles automatically with his own body, beating his chest with his right hand and touching his genitalia. He then crosses his legs and is ready to fall asleep again.

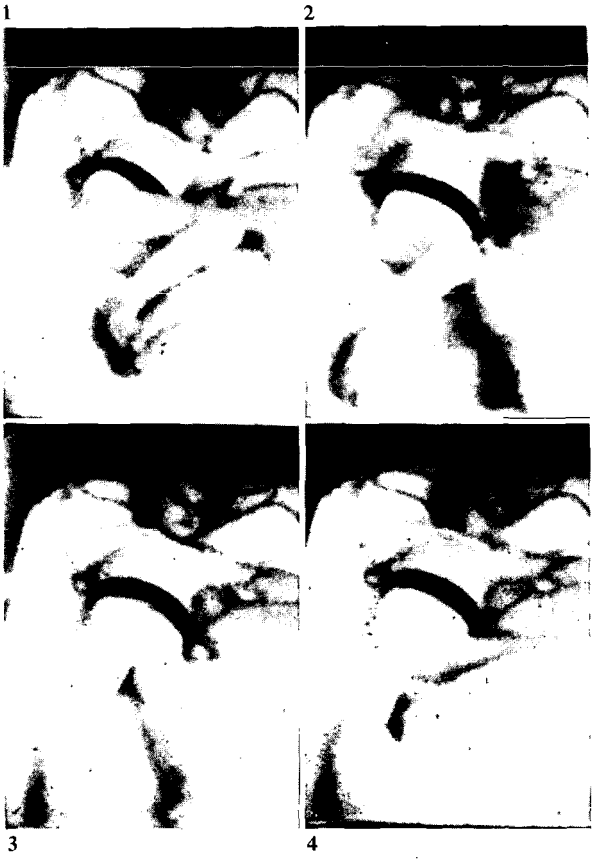

FIG. 1. Videotape pictures of a nocturnal episode in Patient 1.

Patient 5, 74 years old

The patient wakes suddenly from a slow wave sleep (stage 3–4) and attempts to sit up but is restrained by his wife. He is staring and appears to be frightened. Weak movements of the legs and automatic activity of the arms are observed. After 10 sec, he falls back and becomes rigid. The tonic contraction is maintained for approximately 15 sec, after which the patient relaxes suddenly. After making a few additional movements of the arms, the patient reestablishes contact with his environment and motions for the lights to be turned off. The tonic phase is associated with tachycardia lasting for approximately 1 min and thus continuing after the episode subsides.

Treatment

All subjects had received various different treatments previously but had experienced no beneficial effects. In particular, tranquilizers, antidepressant drugs, hydantoin, and barbiturates had been of little value.

Carbamazepine was administered to all patients (15 mg/kg daily). The frequency of seizures was distinctly reduced in patients 2 and 3; in patients 1 and 4 the

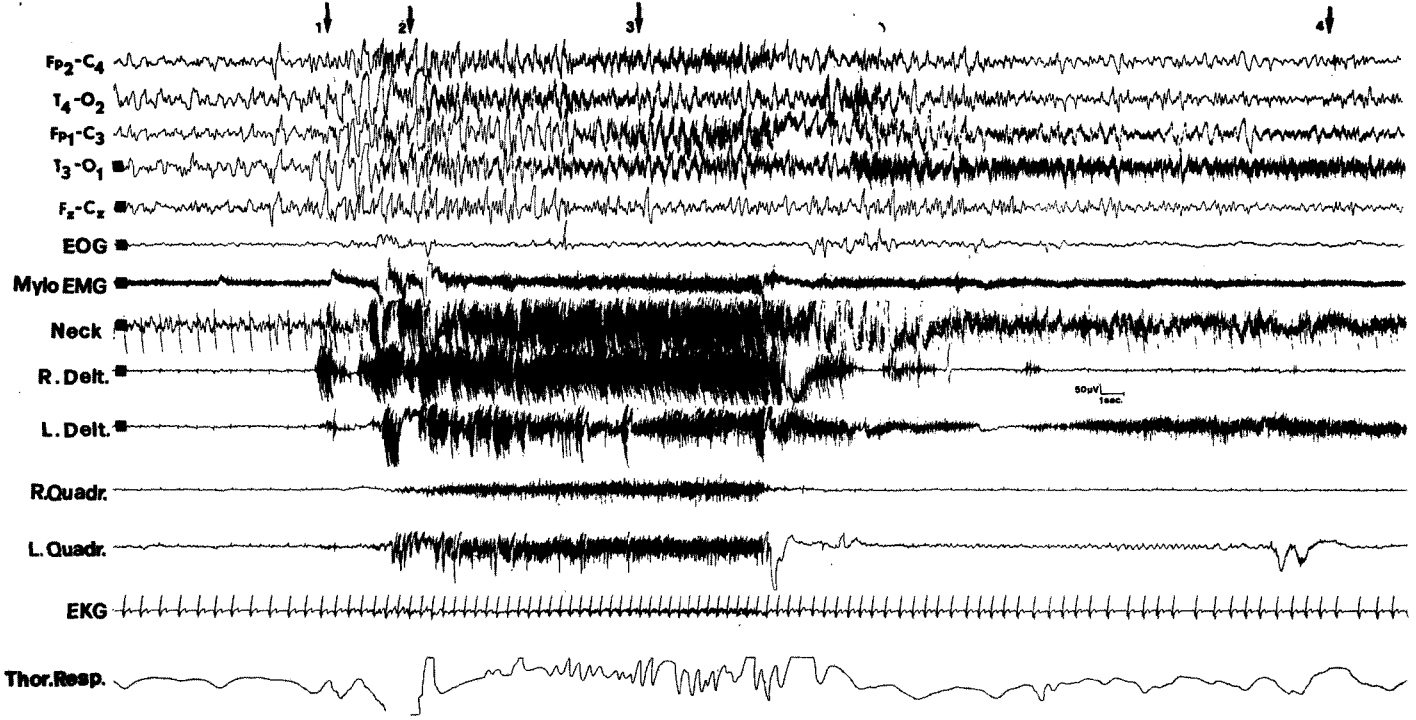

FIG. 2. Polygraphic recording corresponding to the episode shown in Fig. 1. Numbered arrows indicate the moments represented in the videotapes.

FIG. 3. Videotape pictures of a nocturnal episode in patient 2.

episodes completely disappeared for at least 4 months after initiation of therapy. We have no news of patient 5.

DISCUSSION

The clinical and polygraphic features shared by all our patients can be outlined as follows:

All patients, who were of both sexes and ranged in age from 7 to 74 years, gave a history of 2–34 years duration of isolated or repeated nocturnal episodes of 15–40 sec duration. "Seizures" recurred every night or nearly every night a few seconds after an arousal from NREM sleep stage; typically, they consisted of twisting of the trunk and coarse movements of the limbs, and they were occasionally associated with a tonic phase of variable duration and severity.

The patients were aware of the episodes, but their recollections were vague. EEG recordings during both sleep and wakefulness and during the course of the "seizure" were normal in all cases. Of the 4 patients we were able to examine on follow-up visits, all responded well to carbamazepine treatment. In all cases, hydantoin and barbiturates had been ineffective.

The uniform clinical behavior and EEG pattern suggest that this syndrome

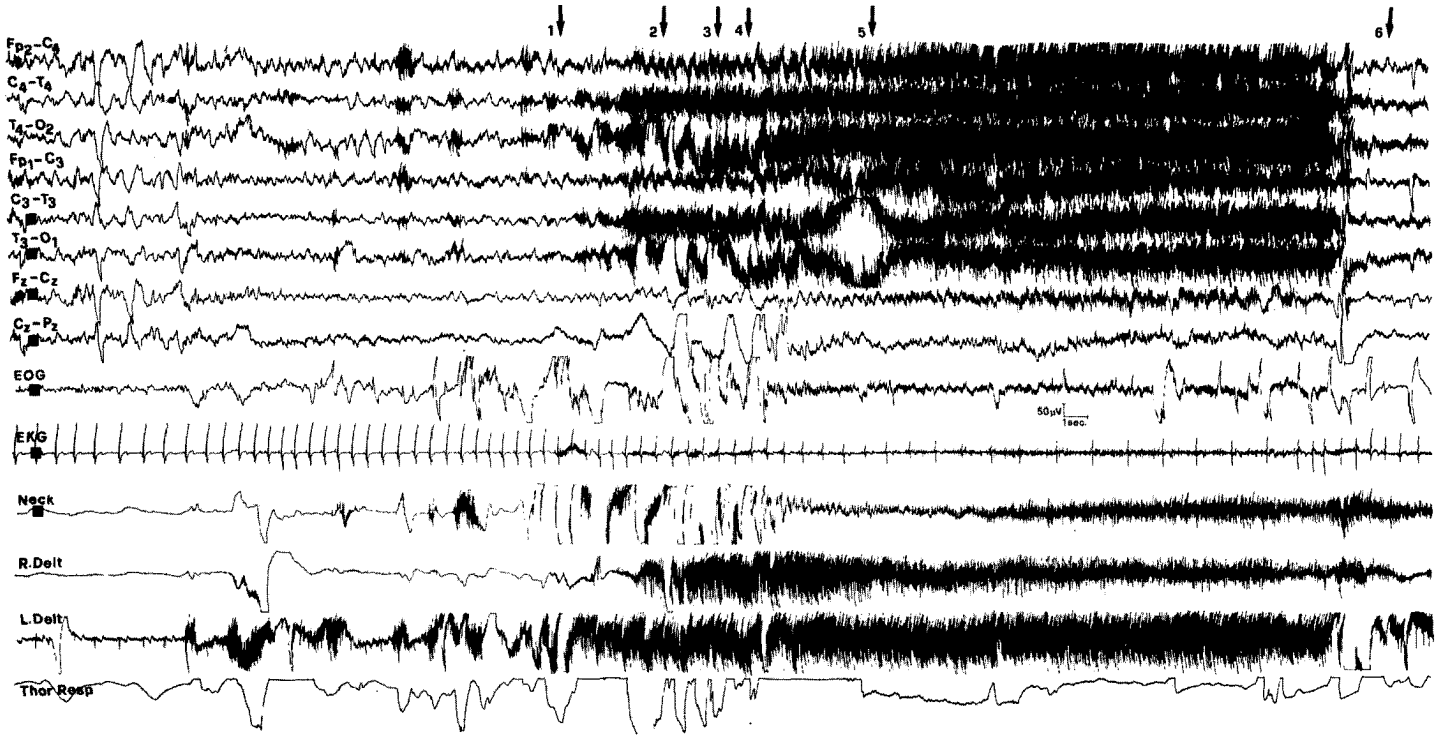

FIG. 4. Polygraphic recording corresponding to the episode shown in Fig. 3. Numbered arrows indicate the moments represented in the videotapes.

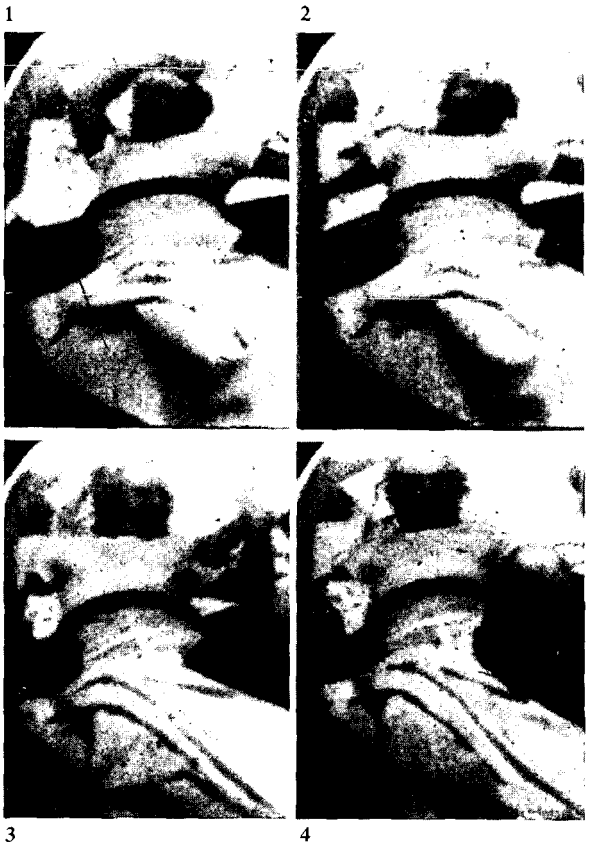

FIG. 5. Videotape pictures of a nocturnal episode in patient 3.

represents a distinct nosological entity; however, the underlying pathophysiology remains uncertain. Three possible interpretations can be entertained, namely that the syndrome represents

1. a particular form of pavor nocturnus,
2. epileptic seizures whose discharges occur in deep or mesial regions and are therefore not recorded, or
3. a distinct form of paroxysmal dystonia initiated during sleep and triggered by an arousal.

The suggestion that the episodes might be triggered by nightmares or pavor nocturnus is supported by their occurrence during sleep, often following an arousal, in the absence of EEG abnormalities. However, the hypothesis seems unlikely in light of the high frequency and very short duration of the seizures, their persistence (patient 5 presented with a 34-year history), and their occasional occurrence during wakefulness.

The view that the seizures represent a peculiar form of epilepsy is substantiated by the occurrence of attacks during the day (in patients 2–4) and by the fact that there were past symptoms suggestive of epilepsy in 2 cases. Patient 1 had, between the ages of 2 and 4 years, suffered from sporadic, unclassified spells during

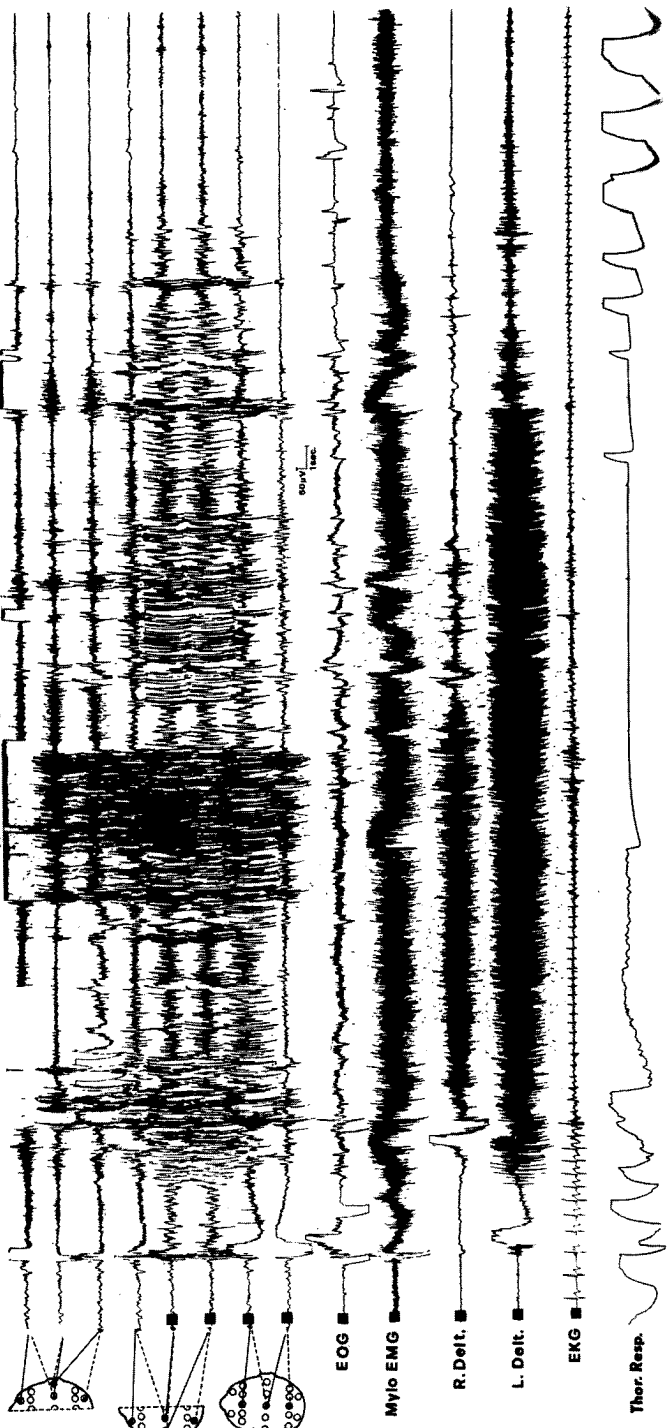

FIG. 6. Polygraphic recording corresponding to the episode shown in Fig. 5. Numbered arrows indicate the moments represented in the videotapes.

which she would suddenly fall to the ground and display automatic behavior. Patient 3, when 26–28 years old, had experienced occasional episodes during sleep, lasting 30–60 sec and involving loss of consciousness, stiffness, and "diffuse tremor."

Seizures were always very short (15–45 sec), with protracted, stereotyped opisthotonic contraction and motor activity strongly resembling certain seizures of undoubted epileptic nature. We have observed a patient with sporadic episodes during sleep with a temporal spike focus and diurnal psychomotor seizures (unpublished data). Such episodes, whose epileptic nature clearly emerged from EEG ictal discharges, were clinically similar to the present cases.

In addition, the hypothesis that we are dealing with a peculiar form of epilepsy is further supported by the favorable response of our patients to carbamazepine treatment. Nonetheless, this interpretation is challenged by their previous lack of response to hydantoin treatment and by the ictal and interictal EEG findings, which were consistently normal during both sleep and wakefulness.

Finally, the episodes could be related to cases of paroxysmal kinesigenic choreoathetosis (PKC) (Lance, 1977). Common features of PKC are the short duration, the stereotypic, tonicodystonic movements, and the possible incidence of episodes during nocturnal sleep. In his discussion of the possible determinants of PKC, Lance suggests that occasional dystonic and choreoathetosic symptoms may be due to an "hereditary sensitivity to dopamine or some related transmitter and that stress, excitement and any situation in which norepinephrine is liberated allows the build-up of its precursor, dopamine, in the basal ganglia in a concentration sufficient to cause paroxysmal dystonia."

By analogy, it might be that in our patients, sudden neurophysiological and neurochemical changes—induced, for example, by arousal—cause a paroxysmal disturbance of the dynamic balance between the cortex and the basal ganglia, thus explaining the occurrence of dystonic episodes. The restlessness associated with the seizures might be due to a psychological reaction to dystonia.

Whatever the pathophysiological substratum of these episodes, they can be recognized easily as abrupt sleep disturbances with peculiar clinical and EEG features that respond favorably to carbamazepine treatment, but not to other anticonvulsant drugs. This hypothesis is not contradicted by the effectiveness of carbamazepine. In fact, carbamazepine in particular among anticonvulsant drugs can relieve PKC (Kato and Araki, 1969).

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