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Pediatric extratemporal epilepsy presenting with a complex auditory aura

Dave F. Clarke[1,2], Frederick A. Boop[2], Amy L. McGregor[1,2], F. Frederick Perkins Jr.[1,2], Vickie R. Brewer[2], James W. Wheless[1,2]

[1] Department of Pediatrics, Division of Pediatric Neurology, University of Tennessee Health Science Center
[2] Le Bonheur Comprehensive Epilepsy Program, Memphis, TN, USA

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ABSTRACT – *Introduction.* Ear plugging (placing fingers in or covering the ears) is a clinical seizure semiology that has been described as a response to an unformed, auditory hallucination localized to the superior temporal neocortex. The localizing value of ear plugging in more complex auditory hallucinations may have more involved circuitry. We report on one child, whose aura was a more complex auditory phenomenon, consisting of a door opening and closing, getting louder as the ictus persisted. *Methods.* This child presented, at four years of age, with brief episodes of ear plugging followed by an acute emotional change that persisted until surgical resection of a left mesial frontal lesion at 11 years of age. Scalp video-EEG, magnetic resource imaging, magnetoencephalography, and invasive video-EEG monitoring were carried out. *Results.* The scalp EEG changes always started after clinical onset. These were not localizing, and encompassed a wide field over the bi-frontal head regions, the left side predominant over the right. Intracranial video-EEG monitoring with subdural electrodes over both frontal and temporal regions localized the seizure-onset to the left mesial frontal lesion. The patient has remained seizure-free since the resection on June 28, 2006, approximately one and a half years ago. *Conclusion.* Ear plugging in response to simple auditory auras localize to the superior temporal gyrus. If the patient has more complex, formed auditory auras, not only may the secondary auditory areas in the temporal lobe be involved, but one has to entertain the possibility of ictal-onset from the frontal cortex. *[Published with video sequences]*

Key words: extratemporal epilepsy, complex auditory auras, ear plugging

Correspondence:
D. F. Clarke, M.D.,
Department of Pediatrics,
Division of Pediatric Neurology,
University of Tennessee Health Science Center,
777 Washington Ave., POB 335,
Memphis, TN 38105, USA
<dclarke3@utmem.edu>

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Frontal lobe seizures might present in a variety of ways and might be difficult to recognize (Jobst and Williamson 2005, Parrino *et al.* 2006, Ryvlin *et al.* 2006, Bonelli *et al.* 2007). The seizures are often nocturnal and frequent. Seizure semiology varies greatly depending on the site of ictal onset or the regions involved. Psychic phenomena, kicking, cycling and other hypermotor phenomena, tonic posturing, focal clonic activity, staring and versive head and eye deviation are some of the EEG features that have been described. Unfortunately, because of the complex anatomy and circuitry of the frontal lobe, it is often difficult to separate the region of seizure-onset from that of seizure-spread.

The temporal lobe, or, even more specifically, Heschl's gyrus or the superior temporal gyrus, is the site identified as the primary auditory area (Penfield and Jasper 1954, Talwar *et al.* 2001, Clarke *et al.* 2003). Ictal auditory semiology often includes unformed sounds such as clicking and buzzing. However, more complex sounds such as hearing a specific song have been described (Penfield and Jasper 1954). Although previous articles describing this seizure semiology have implicated the temporal lobe, scalp EEGs have often revealed discharges in both the temporal and frontal regions (Ottman *et al.* 2004, La Vega-Talbot *et al.* 2006). This is attributed to poor localization as a function of the scalp electrodes. However, even after placement of subdural electrodes, as in three cases in which auditory phenomena presented clinically as ear plugging, there is often rapid spread or involvement at onset, of a region in the frontal lobe (Ottman *et al.* 2004, Clarke *et al.* 2003). We present a child with an auditory aura manifesting as her placing her left and right index fingers in each ear (ear plugging), in whom it is likely that the left mesial frontal lobe was not only the region of ictal onset, but the subdural electrodes over the temporal lobe did not become involved during evolution of the electroencephalographic seizure.

Case report

The patient is an eleven-year-old, right-handed female who was first diagnosed with seizures at the age of four years. Her initial events consisted of ear plugging (index fingers placed in both ears), followed by laughter or crying. These were initially thought to be behavioral events. At six years of age, she was better able to describe what she felt. She described an auditory aura of a door opening and closing, becoming closer, thus getting louder as the ictus persisted. After hearing the sound, a "scared feeling" was described, which she attributed to annoyance with the sound. Clinically, she consistently turned from supine to prone and to her right when lying down, placing her fingers in both ears and often curled up into a fetal position, while having a fearful expression on her face. If requested, she could remain supine, but it was more distressing. She had never had versive head or eye deviation. Occasionally, the seizures were accompanied by tachypnea associated with the frightened expression described, but both she and her family members denied any alteration of consciousness during the event, or post-ictal fatigue. She could answer questions during the seizure but disliked doing so, and was able to call out during and speak clearly after the event. Her receptive and expressive language therefore, remaining intact *(see the video sequence)*. Each seizure lasted ten to forty-five seconds. Previous medications that had not controlled her seizures included valproic acid, topiramate, and levetiracetam. When seen initially she was on oxcarbazepine.

The patient experienced a gradual increase in seizure clusters. These clusters consisted of five to 60 seizures per day occurring for periods of four to six weeks with intervening seizure-free intervals of one to three months. During these clusters she could not function in school, and they were psychosocially devastating, with her not eating or sleeping adequately, being depressed and not socializing with others. Her initial neuropsychological evaluation at six years of age revealed a verbal IQ of 121 and performance IQ of 106. Two years later, just prior to resection, repeat testing found her to have a verbal IQ of 94 and no change in her performance IQ of 106. Pertinent family history includes febrile seizures in an older brother who also had learning difficulties.

Magnetic resonance imaging (MRI) revealed a left mesial frontal lesion. Because of several ash leaf spots on her trunk and the MRI finding described, she underwent genetic testing for tuberous sclerosis complex, but both the *TSC1* and *TSC2* genes were absent. Other investigations (abdominal ultrasound, heart ultrasound and ophthalmologic examination) were negative.

The interictal scalp video-EEG abnormalities revealed a wide irritative zone encompassing the left frontal area with a field involving the left temporal leads (Fp1, F3 > F7, T3). Bifrontal discharges were also seen (F3, F2, F4). During video-EEG monitoring, scalp ictal electroencephalogram-onset encompassed a wide field with monomorphic slowing seen predominantly over the left frontocentral region, and some involvement of the contralateral frontal area. This scalp-EEG onset was consistently seen 6 - 23 seconds after onset of her clinical symptoms in 12 seizures captured over 24 hours. Magnetoencephalography (MEG) lateralized, but did not definitively localize, epileptiform dipoles to the lesion in question, with dipoles seen in several areas over the left frontal lobe.

Subdural electrodes were placed because of the non-localizing nature of the MEG and EEG, and a clinical semiology possibly suggestive of seizures of temporal lobe origin, (prominent auditory phenomena), rather than mesial frontal. Subdural strips were placed anterior and posterior to the lesion, and a depth electrode was placed along its lateral border *(figures 1A* and *1B)*. A left orbitofrontal strip and a left frontal 32-channel grid were placed *(figures 1A* and *1B)*. Also placed were two subdural longitudinal strips over the lateral surface of the left temporal lobe, covering the mid- to anterior and posterior aspect of the left superior, middle and a small portion of the inferior temporal gyrus *(figures 1A* and *1C)*.

Six seizures were captured in eight hours. They were all clinically identical to the events described above. The electroencephalographic seizure-onset, unlike that seen from the scalp, localized to the left frontal interhemispheric strips and depth electrode only at seizure-onset *(figures 2A* and *2B)*. The electrodes placed deeper, approximating the base of the lesion, became involved first (D1-2, AIH 1-2, and PIH 1-2/2-3). The temporal electrodes

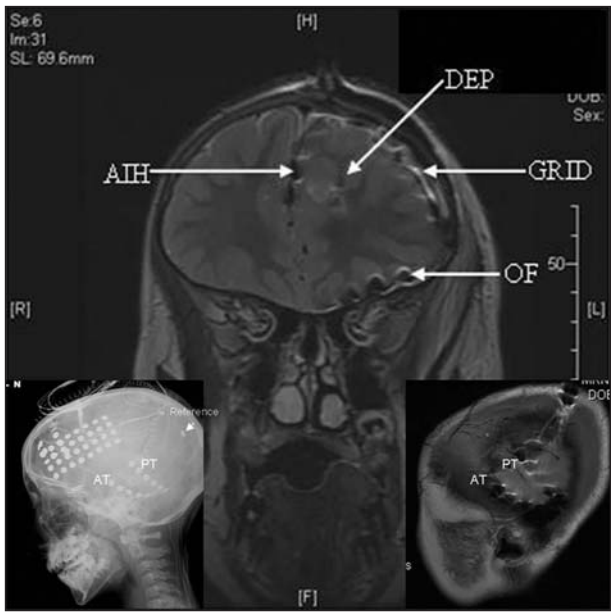

Figure 1A. Coronal T1 MRI revealing anterior, interhemispheric (AIH) 4-channel strip, 4-channel depth (DEP) lateral to the lesion, 4-channel orbito-frontal (OF) strip and 32-channel lateral grid. Anterior temporal (AT) and posterior temporal (PT) subdural strips are also shown on lateral X-ray and a sagittal MRI.

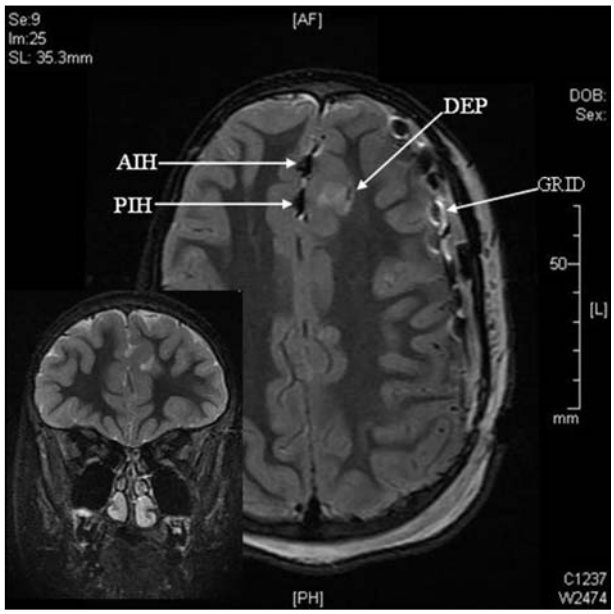

Figure 1B. Axial view revealing anterior and posterior interhemispheric (AIH, PIH) strips and depth (DEP) surrounding the lesion, as well as the lateral 32-channel grid. A coronal image depicting the lesion prior to subdural electrode placement is also shown.

did not become involved and the lateral grid revealed delta-slowing several seconds after onset, which explains the late electroencephalographic onset seen on the scalp

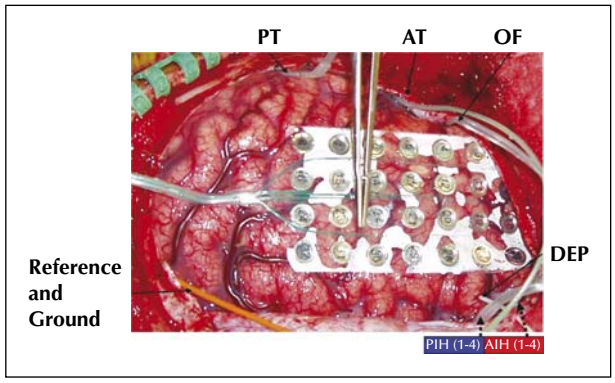

Figure 1C. Digital photo with labeling of the grid wires of the interhemispheric strips, orbitofrontal (OF) strip, and depth (DEP) and anterior and posterior temporal (AT, PT) strips elevated to reveal the portion of the superior temporal gyrus that they cover.

EEG *(figures 2C* and *2D)*. The lesion was resected two days after subdural electrode placement, with no complications. Pathology revealed balloon cell cortical dysplasia. The patient was discharged after four days.

The patient has remained seizure-free since the surgical resection on June 28, 2006 to the present, approximately one and a half years. She is doing better in school, and, at her neuropsychological testing six months after resection, subtle improvements were seen in verbal and performance tasks.

Discussion

The child described, clearly and consistently placed her fingers in her ears secondary to an auditory phenomenon during her seizures. The scalp video-EEG failed to adequately localize and lateralize the ictal-onset zone. The lesion could have been removed without really defining the epileptogenic zone. It is however known, that the epileptogenic zone does not always involve solely the tumor (Awad *et al.* 1991). Therefore, it was decided to monitor the child and then carry out a more definitive surgery. It should be emphasized that even with subdural electrodes, polymorphic theta activity over the lateral grid was not seen until several seconds after ictal-onset. This was likely because of the very focal, restrictive nature of the epileptogenic zone. Electrodes were not placed directly within the sylvian fissure on top of the superior surface of Heschl's gyrus, but strips were placed over the lateral aspect of the temporal lobe *(figure 1A)*. Although we cannot definitely state that a small, isolated region within the sylvian fissure was not involved during the seizure without spread to the surface, the lateral region approximating the primary or secondary auditory areas of the superior and middle temporal gyri did not become involved during the event. Also, removing the lesion alone has rendered the patient seizure-free.

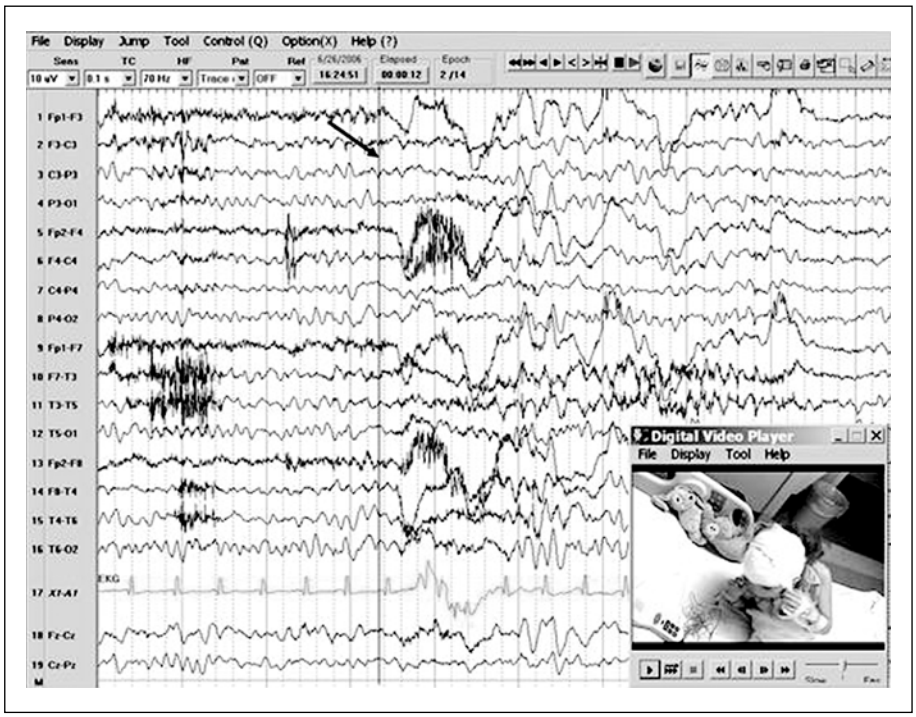

Figure 2A. Clinical onset of seizure is depicted by the line and arrow. Scalp-EEG revealed theta intrusions into the background and bi-temporal slowing preceding the seizure suggestive of drowsiness, followed by movement artifact and bi-temporal theta wave that was poorly sustained after clinical onset, but no definitive focal changes until that shown in *figure 2C* (During scalp EEG monitoring, similar epochs of diffuse slowing and temporal theta was seen during drowsiness).

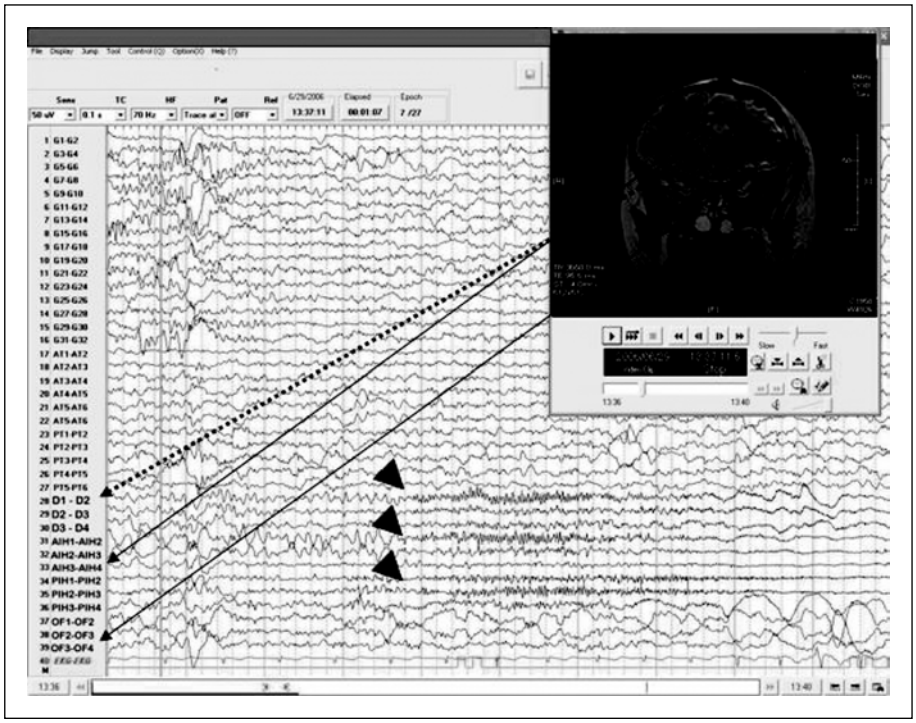

Figure 2B. Intracranial EEG with ictal onset involving the depth as well as the anterior and posterior interhemispheric strips (solid arrow heads). Unlike the scalp EEG, it occurs concurrently with the clinical event.

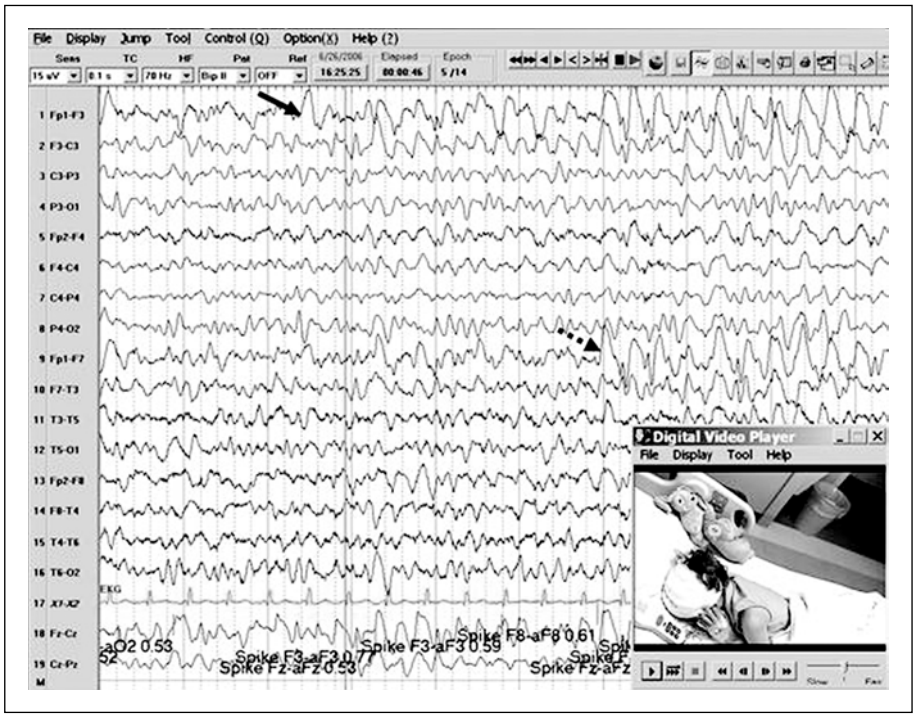

Figure 2C. Rhythmic, left frontal electroencephalographic ictal activity (solid arrow) over 30 seconds after clinical onset, with more left hemispheric involvement some three seconds later (discontinuous arrow).

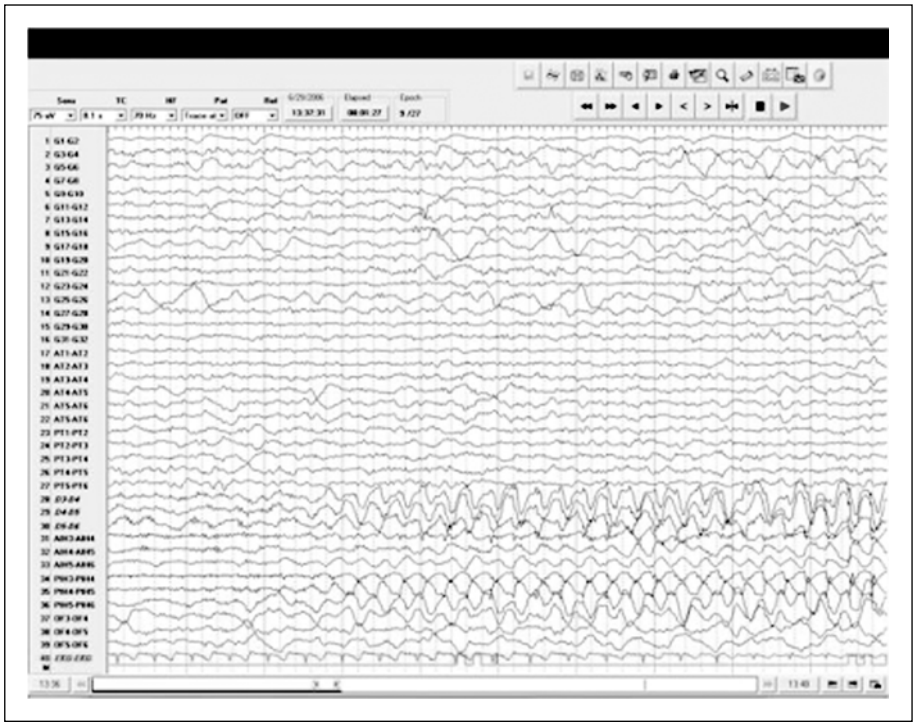

Figure 2D. With evolution of the seizure, an intracranial EEG slowing is seen over the lateral grid over 20 seconds after ictal onset. This likely represents the delayed onset seen on the scalp EEG.

Visual and auditory hallucinations associated with psychosis have been described as evolving from the right orbital frontal cortex (La Vega-Talbot *et al.* 2006). The authors suggested that similar symptoms are seen in schizophrenia, a condition in which frontal lobe dysfunction has been implicated. In the case reported, electrode coverage of the temporal lobe was not described, hence it is unclear if there was spread to the primary auditory cortex. They also did not report if the medial frontal cortex was involved, therefore, it is again unclear if these complex hallucinations involved a similar site of ictal-onset as our patient. Although our patient had ictal fear after the onset of her auditory aura, no other vivid hallucinations or psychoses were ever described.

The patient's auditory hallucinations were seemingly of the complex type, as she was able to identify a specific type of sound getting louder and closer as the ictus persisted. This could have been the perception of a child who heard a simple auditory aura, but in view of her clearly defined explanation, this, as in other subjective descriptions of an aura, has to be respected and believed. Although if requested she could remain prone, she consistently turned towards the right when lying down. She did not however, have versive movement, and it is unclear if she was attempting to lateralize or turn away from the sound. Using functional MRI (fMRI), the processing of sound coming from different directions has been found to activate complex neuronal pathways, which not only included Heschl's gyrus and the superior temporal lobe but also the inferior and middle frontal gyri (Brunetti *et al.* 2005). Positron emission tomography (PET) and MEG have shown a similar complexity of the auditory pathway, with involvement of both the frontal and temporal lobe in individuals with no identified brain pathology (Brunetti *et al.* 2005, Goycoolea *et al.* 2005).

Conclusion

Patients with repetitive stereotyped auditory phenomenon should be investigated for seizures. If it is a simple auditory aura such as buzzing, the primary auditory area along the superior temporal gyrus is likely the site of ictal-onset. If it is a complex or atypical auditory aura, although the temporal lobe is still most often implicated, one has to determine if this ictal phenomenon might be evolving from or involving a more complex pathway, including the frontal lobe. □

Legend for video sequence

Clinical seizure semiology characterized by placing index fingers in both ears and turning over towards the right. The patient retains consciousness and can return to the supine position during the seizure if requested.

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