Temporal lobe epilepsy in children: overview of clinical semiology

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ABSTRACT – Temporal lobe epilepsy in children has been less well studied compared to that seen in adults. Whereas hippocampal sclerosis is the most common etiology for the adult temporal lobe epilepsy syndromes, tumors and malformations of cortical development are more commonly seen in children. Differences in seizure semiology are also apparent. Temporal lobe epilepsy in very young infants may exhibit prominent motor manifestations reminiscent of extra-temporal seizures. These motor manifestations however decrease with increasing age and are less abundant in adults. Automatisms, which are commonly seen in temporal lobe epilepsy in all age groups, are simple at a younger age and become increasingly more complex and discrete with age. Several case studies, illustrated on the video are included in this review. They highlight the differences in temporal lobe seizure semiology between children and adults.

[Published with video sequences]

Key words: childhood temporal lobe epilepsy, semiology, hypomotor, automatisms, temporal lobectomy, epilepsy surgery

Although the semiology of temporal lobe epilepsy (TLE) has been studied in detail in adults, relatively few studies have examined TLE in infants and children. The TLE syndrome in adults is a rather homogeneous entity, with mesial temporal (hippocampal) sclerosis, its commonest neuropathological substrate. The distinct neuropathological correlates associated with this entity include neuronal loss in the hippocampus (the CA1 region being the most profoundly affected, with relative sparing of the CA2 region). The imaging correlates of these pathological changes, as demonstrated by high resolution MRI studies, include 1) an increased signal in the hippocampus, best appreciated on FLAIR sequences, 2) hippocampal volume loss, best appreciated on the T1 coronal images, 3) an overall homogeneous appearance of the hippocampus (as opposed to the gray – white demarcation usually found in normal hippocampi). Whether the hippocampal neuron loss precedes the seizure onset or is a result of ongoing seizures, especially prolonged seizures, is a question which has not yet been satisfactorily answered. However, there is some evidence to suggest that this is an ongoing process, which evolves over time, including following the development of seizures (French et al. 1993, Mathern et al. 1995). The relationship between a past history of febrile seizures in early childhood, especially complex febrile seizures, and the development of the mesial temporal...
sclerosis (MTS) syndrome in later life is also a matter of considerable debate and controversy, with contradictory evidence, both in favor of and against this association, presented in several studies (Cendes et al. 1993, French et al. 1993, Hudson et al. 1993, Hufnagel et al. 1994, So et al. 1989, Williamson et al. 1993). As a high proportion of these patients are medically refractory to currently available antiepileptic drug therapy, temporal lobectomy offers them a very good chance for seizure-freedom after resective surgery (Engel et al. 2003).

In contrast to its adult counterpart, childhood-onset TLE probably represents a distinct nosological and probably less homogeneous syndrome, with MTS being relatively less common in this age group. Duchowny et al. (1992) studied 16 children less than 12 years old, with medically refractory seizures of temporal lobe origin who underwent a standard anterior temporal lobectomy. They found prenatally-acquired abnormalities of neurogenesis (malformations of cortical development, migrational disturbances), low-grade neoplasms, tuberous sclerosis (TS) to be much more frequent, whereas MTS was found in only two children. Even in those two cases, MTS was not an isolated finding (coexistent hamartoma in one and cortical dysplasia in the other).

We present a brief review of seizure semiology in the better-studied, adult onset TLE syndrome followed by a detailed discussion of the childhood syndrome highlighting the inherent differences between these two entities.

Semiology of temporal lobe epilepsy in adults

Clinical symptomatology (Kotagal et al. 1995) includes typical auras (the abdominal aura often described as an “epigastric rising sensation” is commonest), fear, déjà vu- or jamais vu-like sensations (King and Ajmone-Marsan 1977, Palmini and Gloor 1992, Van Buren 1963), olfactory or gustatory hallucinations. Auras are extremely common in patients with mesial temporal lobe epilepsy occurring in more than 90% cases and on occasion may be the only manifestation of the seizure. In most cases however, this aura is followed by other manifestations which may include motor and behavioral arrest, the so-called “blank stare” with retraction of the palpebral fissures accompanied by pupillary dilatation, decreased responsiveness and alteration of consciousness.

Automatisms are another common feature of mesial temporal lobe seizures, and are described as semi-purposeful, stereotyped motor activities. Commonly described automatisms are oro-alimentary (lip-smacking and swallowing, chewing, licking) and in the distal upper extremities (picking or fumbling movements).

Contralateral dystonic posturing (most commonly seen in the hand, but occasionally seen in the face and leg), has been long recognized as a manifestation of temporal lobe seizures, but was only relatively recently recognized as an important lateralizing sign (Kotagal et al. 1989). Figures ranging between 15-70% have been quoted regarding the frequency of this sign in mesial temporal epilepsy, however, the lateralizing specificity of this sign is extremely high (90-100%). Studies utilizing ictal SPECT have concluded that this unilateral, dystonic posturing is the clinical correlate of increased perfusion in the basal ganglia ipsilateral to the side of seizure onset.

A summary of features seen in this subtype of TLE is shown in Table 1.

TLE in children

Epilepsies (both focal and generalized at onset) in infants and young children have a limited repertoire of ictal manifestations. Duchowny (1987) studied 187 seizures in 14 infants less than 2 years old, with suspected partial epilepsy (of temporal/extratemporal origin) and reported that behavioral arrest and tonic, bilaterally symmetric extensor stiffening were the commonest clinical manifestations seen in complex partial seizures during infancy. Similar observations were reported by Acharya et al. (1997). They analyzed 125 seizures in 23 infants aged 2-24 months, with “localization-related epilepsy” (again of temporal or extratemporal origin), with seizure-free outcome after resective surgery. Localization-related epilepsy was defined as either a localized ictal EEG or a localized lesion on neuroimaging. They concluded that seizure symptomatology could be divided into the following broad categories: the first characterized by behavioral arrest and decreased motor activity (hypomotor), which arose from temporal, temporo-pial or occipital regions (7 patients), and the other with predominantly motor (clonic, tonic or atonic) manifestations, which arose predominantly from frontal, fronto-parietal, central or fronto-central regions (12 patients). They also classified a 3rd major category, i.e. epileptic spasms, which could arise from either location.

In a more detailed, subsequent study (Hamer et al. 1999), 296 videotaped seizures from 76 patients aged 1-35 months, with either generalized or partial epilepsy were reviewed. The authors reported that four clinical patterns accounted for 81% of all seizure types. These were epileptic spasms (24%), clonic seizures (20%), tonic seizures (17%) and hypomotor seizures (20%; defined as behavioral arrest, cessation or significant decrease in motor activity with an indeterminate level of consciousness). The Cleveland Clinic epilepsy group (Acharya et al. 1997, Hamer et al. 1999) prefer the use of the term “hypomotor”, since it emphasizes the cessation, or the significant reduction of behavioral and/or motor activity without committing to the level of consciousness, since this is difficult to ascertain in very young children.
Other studies of TLE seizure in children revealed similar observations. Fogarasi et al. (2002) analyzed 83 seizures in 15 children aged 11-70 months, selected by post-temporal lobectomy, seizure-free outcome and found that there was a linear and inverse correlation between the ratio of motor components with increasing age. All children younger than 42 months had seizures with early and prominent motor manifestations; tonic, clonic, myoclonic which may be bilateral and symmetric. These motor components decreased with increasing age; simple gross motor and oro-alimentary automatisms became increasingly complex with age. Whereas only simple automatisms such as oro-alimentary, gestural and blinking were seen in preschool children, more complex automatisms such as hand clapping, beating hands on a blanket or card shuffling were only seen in children older than eight. None of the pre-school children (1-6 years) in their series exhibited any complex automatisms compared to 4/4 children in the 13-16 years age group who had complex automatisms. Jayakar and Duchowny (1990) examined 126 seizures in 26 children younger than 12 years, with unilateral TLE (in this case defined by clear electrographic documentation of seizures of unilateral temporal lobe onset on the basis of ictal EEG), and found motionless staring, behavioral automatisms and motor activity to be the commonest manifestations throughout childhood. While initial motor activity was significantly more common in infants (28%), staring at seizure onset was more frequently (90%) seen in the school-age child. Automatisms were frequent (71%-88%) at all ages, but their nature and complexity changed with age. Simple gross motor and oro-alimentary automatisms were seen in infants and pre-schoolers, fine motor acts, complex postures and axial automatisms were more frequent in the school age child (75%).
Limitations of ictal semiology

Oller-Daurella and Oller (1989) emphasized the importance of recognizing patients with partial (temporal and extratemporal) epilepsy presenting initially with a non-partial ictal semiology. They identified 154 cases of partial epilepsy in the first 3 years of life. Of these, only 35 (23%) patients had partial seizures (simple or complex partial or secondarily generalized tonic-clonic seizures) as their initial seizure manifestation, while non-partial seizures were the initial manifestation in 119 (77%) patients. Generalized (tonic, clonic, tonic-clonic, myoclonic or atonic) seizures were the initial manifestation in 62 (40%) patients. Also, of these 119 patients with non-partial initial manifestations, 31 patients showed a seizure-free interval of several years between the initial presentation and the subsequent occurrence of partial epilepsy, raising the possibility of misdiagnosis based on the initial seizure type alone. Their conclusion was that partial epilepsy may be misdiagnosed in the first 3 years of life, either because of the lack of partial features in the ictal semiology or the young age of the child precluding reporting of subjective phenomena during the initial part of the ictus (e.g. aura). Other studies (Dravet et al. 1989, Yamamoto et al. 1987) emphasized the limitations of the role played by ictal semiology in infants, in determining the localization-related nature of an epileptic seizure.

Case studies

We present some illustrative cases of children with a diagnosis of TLE as defined by clinical semiology, EEG findings, neuroimaging, as well as seizure-freedom/significant improvement after temporal lobectomy.

Case 1

A 4-year-old, right handed male with medically intractable epilepsy whose seizures began at 2 years of age. In the video, after the child’s face is turned towards the camera, he lies still without much response or movement. This seizure would be classified as hypomotor in the Cleveland Clinic Classification scheme. Facial automatisms are mentioned by the nurses, although they are not clearly seen on the video. The prominent motor manifestations seen in infants are not seen with increasing age, especially in school-age children. The EEG showed focal spikes, and seizure origin from the left temporal region. The MRI of this patient (figure 1) showed a left temporal lobe lesion (tumor versus dysplasia) and a PET scan showed left temporal hypometabolism. He had a left temporal lobectomy and a follow-up of 3 years during which he remained seizure-free. Pathology revealed cortical dysplasia.

Figure 1. Coronal T1 image of brain MRI of patient described in Case 1. Note the asymmetry between the two temporal lobes. Left temporal lobe shows poorly defined sulcation pattern as compared to the right, suggestive of cortical dysplasia. Differential diagnoses included a low grade tumor in the left temporal lobe.

Case 2

A 2 1/2-year-old male who started having seizures when he was one year old. In the video, note the extensive motor manifestations that the child exhibits that are more reminiscent of frontal lobe seizures; this is often the case in infants and pre-school children with TLE. Contrast this seizure with the relative paucity of motor symptomatology in Cases 1, 3 and 4. This patient’s MRI showed a right temporal lobe tumor. He underwent resection of this tumor and remains seizure-free 12 years after surgery. Pathology suggested a protoplasmic astrocytoma (with coexistence of meningioma, in a single piece of the resection).

Case 3

A 13-month-old female who started having seizures at nine months old. In the video, after the child puts down the bottle, notice the paucity of motor manifestations. This is another example of a hypomotor seizure where it is difficult to establish any alteration of consciousness. Therefore, it becomes difficult to classify the seizure as simple or complex partial using the ILAE seizure classification system. In the later part of the seizure, notice the simple nature of the oral automatisms, which are characteristic of this age group. With increasing age these become more elaborate and complex. The MRI of this patient (figure 2)
showed a left temporal lobe tumor, which had been partially resected at another institution (without benefit). The patient underwent further resection of the tumor and has since remained seizure-free and tumor-free 7 years after surgery. Pathology showed a ganglioglioma.

Case 4
A 3-year-old boy who started having seizures at the age of 13 months. Initially, seizures were reported as myoclonic jerks of the limbs with occasional head drops. However, over the course of time they evolved to the current semiology as shown in the video. As in the previous case, note the behavioral arrest with a paucity of motor features during the initial part of the seizure, followed by the simple mouth automatisms seen later in the video. The MRI (figure 3) was suggestive of right temporal lesion (dysplasia versus tumor). PET showed hypometabolism of right temporal region. He underwent resection of the lesion, along with right temporal lobectomy. Pathology was consistent with cortical dysplasia. He was seizure-free at last follow-up, 6 months after surgery.

Case 5
The salient features in the video of this 6-year-old, left-handed girl who started having seizures at the age of two...
years include the initial cry, which probably represents an aura of fear probably in response to a visual hallucination (she reported seeing monsters on some occasions and robbers on other occasions). She displays fine distal automatisms of the left upper extremity, along with dystonic posturing of the right upper extremity (similar to TLE semiology characteristic of adults and older children), with lateralization and localization to the left temporal lobe. EEG showed left temporal spikes and seizure onset. The MRI scan (figure 4) showed a cystic lesion in the left mesial temporal region. She underwent left temporal lobectomy along with tumor resection. Pathology was suggestive of ganglioglioma. She was seizure-free at last follow-up, six months after surgery.

Case 6
This 5 1/2-year-old, right-handed girl presented with an episode of status epilepticus at 15 months of age. Her current seizure type began at 3½ years of age and these would occur 2-4 times per month. After arousing from sleep, in this video she displays discrete automatisms of the distal right upper extremity, including tumbling around with the blanket. This is in contrast to the coarse and simple automatisms exhibited by the younger patients in Cases 3 and 4. EEG showed spikes and seizures from the left temporal region. The MRI (figure 5) showed volume loss and increased signal in the left hippocampus. PET showed marked left temporal hypometabolism. She underwent a left temporal lobectomy and remains seizure-free 3½ years after surgery. Pathology was suggestive of hippocampal sclerosis

Case 7
This 4 1/2-year-old, right-handed boy presented with status epilepticus at the age of nine months. His present seizure type (see video sequence) begins with an abdominal aura. The abdominal aura is followed by dystonic posturing of the right hand, behavioral arrest, inability to speak and bilateral, fine upper extremity automatisms. These features suggest a left temporal lobe seizure-onset, as confirmed by focal spikes and seizures noted on EEG from this region. The MRI of the brain revealed evidence of volume loss as well as a signal abnormality in the left hippocampus. PET scan showed hypometabolism of the left temporal region. He had a left temporal lobectomy and remains seizure-free 3½ years post-surgery. Pathology revealed evidence of hippocampal sclerosis as well as a cortical dysplasia.
Case 8
An 18-year-old, left-handed male who started having seizures at the age of 10 months. In the video he wakes up from sleep and tries to press the seizure button. This is followed by a phase during which he just lies in bed experiencing some subtle, fine automatisms. Next we see tonic pulling of the right side of the face along with head deviation to the left followed within seconds by a generalized tonic-clonic seizure. This type of sustained, involuntary, unnatural and forceful head deviation is classified as a versive seizure and when closely followed (within 10 seconds) by a generalized tonic-clonic seizure, lateralizes seizure focus to the contralateral hemisphere (Ochs et al. 1984, Wyllie et al. 1986, Kernan et al. 1993, Fakhoury and Abou Khalil 1995, Marks and Laxer 1998), in this case the right. The patient's EEG showed evidence of slowing, and seizure onset from the right temporal region. The MRI revealed a right temporal malformation of cortical development (figure 6). The patient underwent a right temporal resection and has been seizure-free for 16 months. Pathology was suggestive of focal cortical dysplasia.

Case 9
This 16-year-old, right-handed female started having seizures at the age of 12 years. She has an aura of fear and déjà vu followed by profound oral and hand automatisms

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**Figure 5.** A) Coronal T1 image of brain MRI of Case 6 showing smaller left hippocampus compared to the right. Also note the increased size of the temporal horn of the left lateral ventricle; a marker of volume loss in the left hippocampus. B) Coronal T2 weighted images of brain MRI of patient described in Case 6. The increased signal over the left mesial temporal region is suggestive of left mesial temporal (hippocampal) sclerosis.

**Figure 6.** Coronal FLAIR images of brain MRI of patient described in Case 8. Increased signal is seen in the right temporal region suggestive of possible malformation.
along with some difficulty with speech, although she is able to produce purposeful speech. The EEG localized seizure onset to the left temporal region. The MRI (figure 7) revealed a possible hamartoma in the left temporal region. Interestingly, the WADA test lateralized speech to the left hemisphere. She had invasive monitoring followed by left temporal lobectomy and remains seizure-free one year after surgery. Pathology revealed focal cortical dysplasia.

Case 10
This video is illustrative of how temporal lobe seizures may occasionally present with atypical features. This 8-year-old boy had gelastic seizures (usually associated with hypothalamic hamartomas), with spikes, and seizure-onset (after invasive recordings with subdural electrodes) from the left temporal region. MRI revealed possible subtle malformation in the left anterior temporal region. He remains seizure-free seven months after left temporal resection. Pathology revealed focal cortical dysplasia.

Conclusion
To summarize, TLE in infants and young children is often due to etiologies other than mesial temporal sclerosis, namely developmental low-grade tumors and malformations of cortical development. Differences in seizure semiology are also noticeable. In infants, motor manifestations are prominent; with increasing age, these become less evident. Although automatisms are also seen in young children with TLE, they are simpler and less elaborate. Oral automatisms are more common compared to manual automatisms. With age, automatisms tend to become more discrete and complex. It is also important to remember that very young children with partial epilepsy (including TLE) may present with initial manifestations which are non-partial or generalized in character (such as epileptic spasms) and could be misdiagnosed as generalized epilepsy. This has profound implications for the pre-surgical evaluation since epilepsy surgery may render these hitherto “non-surgical candidates” seizure-free. The salient features of TLE syndromes presenting at different ages are summarized in table 1, highlighting the differences between the childhood and the adult syndromes.

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References


