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Lateralizing Value of Asymmetric Tonic Limb Posturing Observed in Secondarily Generalized Tonic–Clonic Seizures

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Summary: *Purpose:* A striking asymmetry of limb posture occurs during secondarily generalized tonic–clonic (GTC) seizures wherein one elbow is extended while the other is flexed during the tonic phase of the GTC seizure. We have named this phenomenon asymmetric tonic limb posturing (ATLP) or the "Figure 4 Sign."

Methods: Fifty-nine secondarily GTC seizures from 31 patients with partial epilepsy who underwent successful epilepsy surgery were analyzed, in addition to another group of 64 GTC and generalized clonic seizures from 26 patients collected prospectively over a 7-month period. Three observers reviewed these seizures blinded to the side of ictal EEG onset and other clinical data.

Results: The extended elbow was contralateral to the side of ictal onset in 35 of 39 patients who had ATLP during their seizures. The kappa index, a measure of interobserver agreement, was calculated, and ATLP was found to have very good agreement between observers.

Conclusions: In secondarily generalized tonic–clonic seizures, ATLP (Figure 4 Sign) may sometimes be only available lateralizing sign. **Key Words:** Asymmetric limb posturing—Figure 4—Generalized tonic–clonic seizures.

In a previous study looking at the lateralizing value of various limb motor signs in partial-onset seizures of temporal and extratemporal origin, we had noticed asymmetric tonic limb posturing (ATLP) occurring during partial-onset seizures progressing to a secondary generalized tonic–clonic (GTC) seizure (1). We have analyzed the lateralizing significance of this phenomenon in this patient group as well as a second population of patients collected prospectively over a 7-month period who had GTC or generalized clonic seizures recorded in the adult and pediatric epilepsy monitoring units.

PATIENTS AND METHODS

Two patient groups were studied: Group I comprised 54 patients with partial epilepsy successfully treated with surgical resection of the temporal lobe (n = 34) or extratemporal regions (14 frontal, three parietal, and three occipital). These patients had a total of 238 seizures recorded, of which 59 seizures from 31 patients became secondarily generalized. Success was defined as complete seizure freedom or >90% seizure reduction measured at 1 year after surgery. Group II consisted of 28 patients who had 70 GTC seizures or generalized clonic seizures collected prospectively over a 7-month period (2). Six seizures and two patients had to be excluded because of suboptimal quality of videotape recordings that did not show both upper limbs clearly. Thus 64 seizures from 26 patients were available for analysis. Of these, 23 patients had focal epilepsy, whereas three patients were available for analysis. Of these, 23 patients had focal epilepsy, whereas three patients had symptomatic generalized epilepsy. Patients with psychogenic seizures also were excluded. Three observers (P.K., A.F.B., E.G.) analyzed the seizure patients independently, blinded to all clinical and electrographic information that was collected by two other observes (P.K.W., B.I.M.).

We defined ATLP as a striking asymmetry of limb posture during the tonic phase of a GTC. One arm is

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FIG. 1. Asymmetric limb posturing during a secondarily generalized tonic–clonic (GTC) seizure that began in the right occipital region. After version of the head and eyes to the left, the head returns to the midline, the left arm is tonically extended, while the right arm is flexed at the elbow. This posture resembles the figure 4. Thereafter both arms became tonically extended, followed by clonic jerking of the extremities and facial muscles.

rigidly extended at the elbow, often with the fist clenched tightly and flexed at the wrist, whereas the opposite extremity is flexed at the elbow (Figs. 1 and 2). Usually this is followed by tonic posturing of both upper extremities, which may give way to bilateral clonic jerking of the extremities and face during the second clonic phase. ATLP may be preceded by version and clonic movements of the face and extremities that occur during the first clonic phase.

We also examined version, defined as unquestionably forced, sustained, and unnatural eye and/or head deviation to one side, to see how well it compares with ATLP. Version is a well-recognized lateralizing sign that has been shown in have good interobserver agreement and high positive predictive value for the side of ictal onset (1,3–5). In addition, the principal author also noted the time of onset of version and ATLP in seizures of group II patients. Statistical analysis was performed by using the kappa index to quantify interobserver agreement. A kappa value of <0.40 is indicative of poor agreement beyond chance, a value between 0.40 and 0.75 indicates fair agreement, and a value >0.75 indicates excellent agreement beyond chance. The Wilcoxon rank-sum test was used to compare the time of onset and duration of ATLP between temporal (TLE) and extratemporal (XTLE) seizure patients.

RESULTS

In group I patients, ATLP occurred in 12 (75%) of 16 TLE patients and eight (53.3%) of 15 XTLE patients in whom seizures progressed to secondary generalization.

FIG. 2. Similar asymmetric limb posturing with the right arm tonically extended at the elbow with clenching of the right fist; the left arm is flexed at the elbow. This patient's seizures were obscured by extensive electromyogram (EMG) artifact on scalp EEG; clinically he would experience a nonlateralized visual aura followed by right head deviation, evolving into a secondarily generalized tonic–clonic seizure. During visual auras, a focal seizure pattern was seen in the left occipital region.

TABLE 1. *Patient characteristics*

	Group I temporal	Group I extratemporal	Group II
Total number of patients	34	20	26[a]
Patients with GTC seizures	16	15	26
GTC seizures analyzed	27	32	64
Number of patients with ATLP in GTC seizures	12/16 (75%)	8/15 (53.3%)	18/26 (69.2%)
Number of seizures showing ATLP	19/27 (70.3%)	10/32 (31.2%)	22/64 (34.3%)
Patients who had ATLP in every GTC seizure	10/12 (83.3%)	6/8 (75%)	2/26 (7.6%)
Version and ATLP	8/16	4/15	10/26
Version, no ATLP	1/16	4/15	12/26
ATLP, no version	4/16	4/15	9/26
No ATLP, no version	3/16	3/15	0
Kappa index for ATLP	0.68	0.71	0.711
Patients with ATLP contralateral to focus	10/12 (83.3%)	7/8 (87.5%)	17/18 (94.4%)
Patients with version	9/16 (56.3%)	8/15 (53.3%)	13/26 (50%)
Kappa index for version	0.84	0.67	0.725
Patients with version contralateral to focus	9/9 (100%)	8/8 (100%)	13/13 (100%)

GTC, generalized tonic–clonic; ATLP, asymmetric tonic limb posturing.

[a] Six patients had temporal lobe epilepsy, 16 had extratemporal epilepsy, one patient's seizures could be lateralized but not localized, and three had symptomatic generalized epilepsy.

In group I patients, the overall interobserver agreement as measured by the kappa index was 0.71; 0.68 in TLE patients and 0.71 in XTLE ($p < 0.01$). This denotes good agreement between observers for the presence or absence of the sign as well as the side on which it was seen. The extended arm was contralateral to the seizure focus in 10 of 12 temporal lobe cases and seven of eight extratemporal cases. Version, on the other hand occurred in nine (56.3%) of 16 TLE patients and eight (53.3%) of 15 XTLE patients. Its kappa index was high, 0.76 ($p < 0.01$) overall; 0.84 for TLE and 0.67 for XTLE. Version was always contralateral to the side of ictal onset.

In group II patients, ATLP occurred in 18 (69.2%) of 26 patients and version in 13 (50%) of 26 patients. ATLP was once again found to have good interobserver agreement with a kappa score of 0.711 ($p < 0.001$) overall. Because of the smaller patient numbers, the kappa index could not be calculated separately for TLE and XTLE patients. ATLP was contralateral to the side of ictal EEG onset in 17 (94.4%) of 18 patients. The kappa index for version was 0.725 ($p < 0.001$), and it was always contralateral to the side of ictal onset. The appearance of ATLP occurred at a median of 23.5 s from clinical onset (range, 4–108 s); in TLE, onset of ATLP occurred at a median onset time of 28.5 s (range, 11–108 s) versus 22.5 s (range, 4–52 s) in XTLE. This difference was not statistically significant ($p = 0.46$). The median duration of ATLP was 10 s (range, 2–20 s) in the extratemporal patients versus 4.5 s (range, 2–20 s) in the temporal patients ($p = 0.14$). In group II, 11 of 18 patients with ATLP underwent focal resection (five temporal, six extratemporal), and six patients became seizure free. In the remaining five patients, seizures continued to arise from regions adjacent to the resection site (three patients) or from foci in the opposite hemisphere (two patients), which were identified before surgery. In the latter two patients, ATLP was contralateral to the epileptogenic region resected and was not observed in any of the seizures arising from the other side. Please see Table 1 for more details regarding group I and group II patients.

Other characteristics of ATLP were

1. ATLP followed M2e posturing. M2e posturing was noted in eight seizures from three (11.5%) of 26 patients in group II. Ajmone-Marsan coined this term to describe

   > abduction and external rotation of the shoulder with flexion of the elbow, with or without head turning; the head may look at the postured hand or be turned to the opposite side. The leg ipsilateral to the postured arm may extend while on the opposite side of the body, the arm assumes slight elbow flexion and that leg is flexed at the hip and knee (6).

   M2e was seen to evolve into ATLP, in which that same limb is extended while the ipsilateral arm remains flexed for several seconds before it too is extended (i.e., the typical tonic posturing of both upper extremities), which is then followed by the clonic movements of all extremities during a GTC seizure. M2e was contralateral to the focus in all three patients.
2. ATLP follows head and eye version. At the time of ATLP, the head position was midline or returning to midline from version in all instances (Fig. 3A). ATLP was on the same side as version in all but one patient in group II. This sole exception was a patient with symptomatic generalized epilepsy in whom left head version was followed by right head version, right ATLP, and then generalized clonic movements. EEG showed a generalized ictal pattern that was higher over the left hemisphere.
3. The extended arm is usually in front of the trunk, and occasionally it is raised in the air. The ipsilat-

FIG. 3. Right head version **(A)** is followed by tonic extension of the right arm with flexion of the left arm at the elbow **(B).**

eral upper extremity is also generally in the same plane. The flexed arm may be positioned either in front or behind the extended arm.

4. The lower extremity on the same side as the extended arm also was extended, except in two seizures from two patients in which the leg was flexed.
5. In nine (39.1%) 23 of patients in group II with partial-onset seizures evolving to GTC seizures, ATLP occurred without prior version and in fact was the only lateralizing sign observed.
6. In two patients from group II, ATLP occurred first on the side contralateral to the ictal onset (Fig. 3B), followed by extension of both upper extremities (Fig. 3C), which was followed by bilateral clonic movements more prominent on the side of initial ATLP, while the opposite arm remained extended (Fig. 3D). Thus ATLP may change sides during the evolution of the seizure. In such instances, we found the initial ATLP to be contralateral to the side of ictal onset. Without exception, ATLP occurred on the same side in all of the patient's seizures that showed this phenomenon.

We present this anecdotal information from two patients who were not included in either group I or II. The first patient had independent seizure onsets from the left and right temporal lobes. ATLP was seen contralateral to the side of ictal onset in both types of seizures. The second patient had both juvenile myoclonic epilepsy (JME) and temporal lobe epilepsy. ATLP occurred only in his four temporal lobe complex partial seizures with secondary generalization but not the two primary GTC seizures.

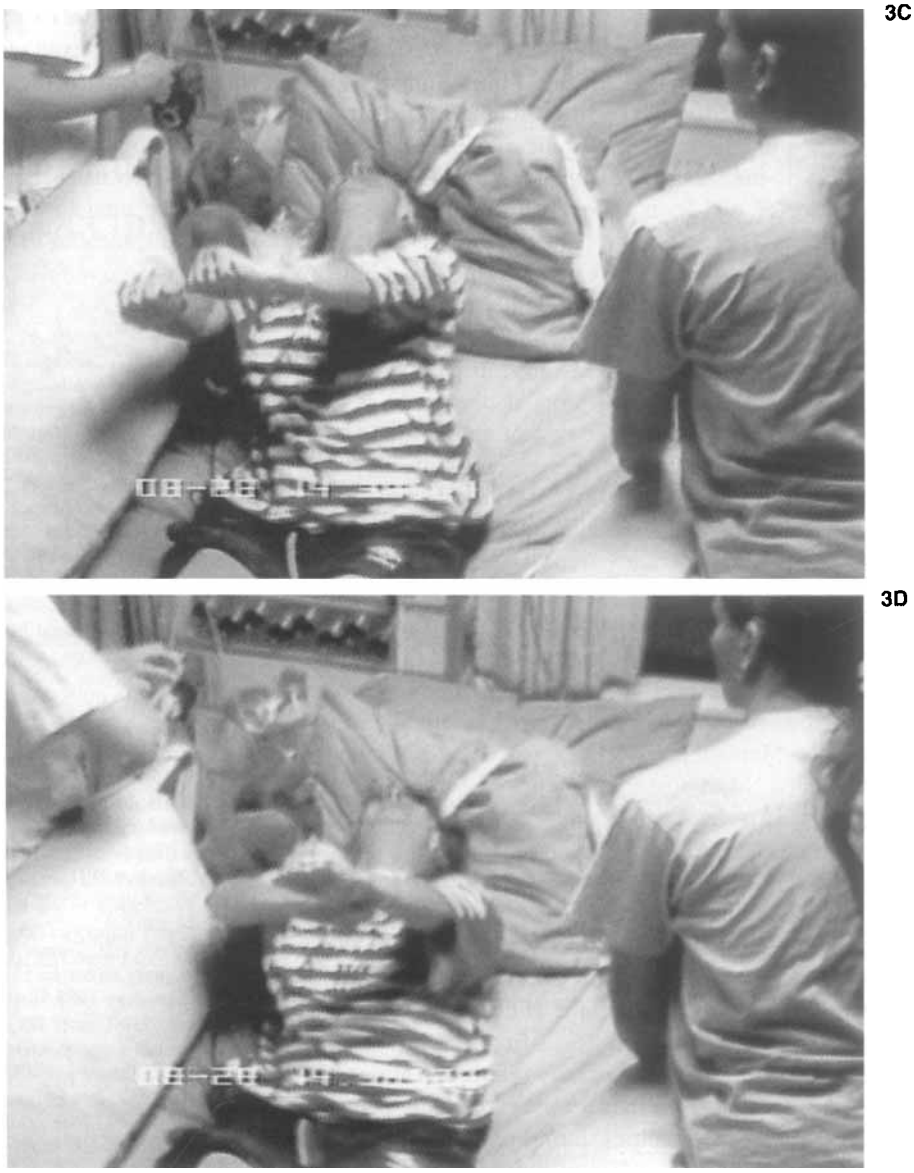

Fig. 3. *Continued.* Thereafter both upper limbs are rigidly extended at the elbow **(C)** followed by asynchronous clonic jerking of all extremities and the facial muscles. This is much more pronounced on the right side; at this stage, the left arm is still extended, and the ATLP has changed sides **(D).**

DISCUSSION

The clinical characteristics and EEG findings of GTC seizures have been described in detail by Gastaut and Broughton (7) and Theodore et al. (8). In his book on metrazol-induced seizures, Ajmone-Marsan and Ralston (6) described asymmetric posturing of the arms with one arm flexed and the other arm extended at the elbow, which they termed the "asymmetrical generalized seizure." This was seen in 9% of metrazol-induced seizures but thought not to be of lateralizing significance. In our prospectively collected cases with secondarily generalized seizures (group II), we observed ATLP in nearly 70% of patients. In both group I and II patients, there was good agreement beyond chance between observers for the presence or absence of ATLP as shown by the kappa scores, which were comparable to those for version.

The lateralizing value of this sign appears to be quite good, and it agrees with version. The sole exception was one patient with symptomatic generalized epilepsy whose EEG showed a generalized ictal pattern that was higher over the left hemisphere. In this case, version occurred first to the left and then to the right followed by right ATLP and generalized clonic movements. In retrospect, had the head movement immediately preceding generalization been interpreted as version, it would have agreed with the side of ATLP. In nearly 40% of patients with secondarily generalized seizures, it was the only clinical sign available to lateralize the side of ictal onset.

The anecdotal observations from a single patient who had both TLE and JME were remarkable in that ATLP occurred only in partial-onset seizures with secondary generalization but not in the primarily GTC seizures. This observation needs to be validated in a larger series

of patients having GTC seizures who have idiopathic or symptomatic generalized epilepsy.

After the onset of ATLP, both upper limbs become tonically extended, followed by bilateral clonic movements; in some patients, the clonic movements may appear first on the limb that extended first, thus giving the appearance of ATLP on the opposite side. Such an evolution occurred in one of our patients (Fig. 3). The asymmetric tonic posturing usually involves the lower limb on the same side as well, which is rigidly extended.

Although not statistically significant, the median duration of ATLP tended to be longer in XTLE than in TLE. Asymmetric tonic posturing of the limbs is typically observed in supplementary motor area (SMA) seizures (9) and can also be reproduced with electrical stimulation of the SMA (10). Ictal SPECT studies have shown focal cortical and basal ganglia hyperperfusion in frontal lobe seizures arising from the primary or supplementary motor areas (11). This suggests that ATLP may be due to asymmetric activation of the primary supplementary motor cortices and/or basal ganglia. The occurrence of ATLP in GTC seizures might suggest a focal origin; however, this hypothesis must be confirmed.

Ajmone-Marsan and Ralston (6) described another phenomenon, M2e posturing occurring before the tonic phase of GTC seizures induced by injection of metrazol. They found that M2e posturing was contralateral to the side of seizure onset in 96% of his cases. It was seen in 45% of metrazol-induced seizures. In our group of patients undergoing EEG-video monitoring, many of whom were discontinued from antiepileptic medication, M2e posturing occurred in only three (11.5%) of 26 patients and was contralateral to the focus in all cases.

In conclusion, ATLP (popularly referred to at our center as the figure 4 sign) indicates that the tonic phenomena in the upper extremities during secondarily generalized seizures occurs at different times on the two sides. First, one upper extremity is extended at the elbow while the other is flexed at the elbow, the extended arm being contralateral to the hemisphere of seizure onset. This sign has good agreement between observers, and it is of lateralizing value, particularly when version does not occur. Given its relatively brief duration, careful attention must be paid to the evolution of the asymmetric tonic limb phenomena during secondarily GTC seizures.

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