In 2017, the ILAE released a new classification of seizure types, largely based upon the existing classification formulated in 1981. Primary differences include specific listing of certain new focal seizure types that may previously only have been in the generalized category, use of awareness as a surrogate for consciousness, emphasis on classifying focal seizures by the first clinical manifestation (except for altered awareness), a few new generalized seizure types, ability to classify some seizures when onset is unknown, and renaming of certain terms to improve clarity of meaning.

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ILAE 2017 Classification of Seizure Types Basic Version

**Focal Onset**
- Aware
- Impaired Awareness
- Motor
- Non-Motor

**Generalized Onset**
- Motor
  - Tonic-clonic
  - Other motor
- Non-Motor (Absence)

**Unknown Onset**
- Motor
  - Tonic-clonic
  - Other motor
- Non-Motor

**Unclassified**

---

1. Definitions, other seizure types and descriptors are listed in the accompanying paper & glossary of terms.

2. Due to inadequate information or inability to place in other categories.

### ILAE 2017 Classification of Seizure Types Expanded Version

<table>
<thead>
<tr>
<th>Focal Onset</th>
<th>Generalized Onset</th>
<th>Unknown Onset</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Aware</strong></td>
<td><strong>Motor</strong></td>
<td><strong>Motor</strong></td>
</tr>
<tr>
<td><strong>Impaired</strong></td>
<td>tonic-clonic</td>
<td>tonic-clonic</td>
</tr>
<tr>
<td><strong>Awareness</strong></td>
<td>clonic</td>
<td>epileptic spasms</td>
</tr>
<tr>
<td><strong>Non-Motor</strong></td>
<td>myoclonic</td>
<td>Non-Motor</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>myoclonic-tonic-clonic</td>
<td>behavior arrest</td>
</tr>
<tr>
<td><strong>Motor</strong></td>
<td>myoclonic-atonic</td>
<td><strong>Unclassified</strong></td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>atonic</td>
<td><strong>3</strong></td>
</tr>
<tr>
<td><strong>Non-Motor</strong></td>
<td>epileptic spasms</td>
<td>due to inadequate information or inability to place in other categories</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>hyperkinetic</td>
<td><strong>2</strong></td>
</tr>
<tr>
<td><strong>Motor</strong></td>
<td>myoclonic</td>
<td>These could be focal or generalized, with or without alteration of awareness</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>tonic</td>
<td><strong>1</strong></td>
</tr>
<tr>
<td><strong>Non-Motor</strong></td>
<td>myoclonic</td>
<td>Definitions, other seizure types and descriptors are listed in the accompanying paper and glossary of terms.</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>clonic</td>
<td><strong>3</strong></td>
</tr>
<tr>
<td><strong>Non-Motor</strong></td>
<td>epileptic spasms</td>
<td>Due to inadequate information or inability to place in other categories</td>
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<tr>
<td><strong>Onset</strong></td>
<td>tonic</td>
<td><strong>2</strong></td>
</tr>
<tr>
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<td>myoclonic</td>
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</tr>
<tr>
<td><strong>Onset</strong></td>
<td>atonic</td>
<td><strong>1</strong></td>
</tr>
<tr>
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<tr>
<td><strong>Onset</strong></td>
<td>epileptic spasms</td>
<td><strong>3</strong></td>
</tr>
<tr>
<td><strong>Non-Motor</strong></td>
<td>typical</td>
<td>Due to inadequate information or inability to place in other categories</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>atypical</td>
<td><strong>2</strong></td>
</tr>
<tr>
<td><strong>Non-Motor</strong></td>
<td>myoclonic</td>
<td>These could be focal or generalized, with or without alteration of awareness</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>eyelid myoclonia</td>
<td><strong>1</strong></td>
</tr>
</tbody>
</table>

---

Partial Seizures (start in one place)
  Simple (no loss of consciousness of memory)
    Sensory
    Motor
    Sensory-Motor
  Psychic (abnormal thoughts or perceptions)
    Autonomic (heat, nausea, flushing, etc.)
  Complex (consciousness or memory impaired)
    With or without aura (warning)
    With or without automatisms
  Secondarily generalized

Generalized Seizures (apparent start over wide areas of brain)
  Absence (petit mal)
  Tonic-clonic (grand mal)
  Atonic (drop seizures)
  Myoclonic
  Other

Unclassifiable seizures

Motivation for Revision

- Some seizure types, for example tonic seizures or epileptic spasms, can have either a focal or generalized onset.

- Lack of knowledge about the onset makes a seizure unclassifiable and difficult to discuss with the 1981 system.

- Retrospective seizure descriptions often do not specify a level of consciousness, and altered consciousness, while central to many seizures, is a confusing concept.

- Some terms in current use do not have high levels of community acceptance or public understanding, such as “psychic,” “partial,” “simple partial,” “complex partial”, and “dyscognitive.”

- Some important seizure types are not included.
Possible Seizure Classifications Could be Based On:

- **Pathophysiology**
  - But this is currently impossible with our limited understanding

- **Anatomy**
  - Temporal
  - Frontal
  - Parietal
  - Occipital
  - Diencephalic
  - Brainstem

- **Networks**
  - Neocortical
  - Limbic
  - Thalamo-Cortical
  - Brainstem

- **Practical, by:**
  - AED response
  - Surgical target
  - Disabling
  - EEG pattern
  - Many others

modify existing

1981 ILAE System
2010 ILAE update

- In the absence of fundamental knowledge, ILAE chose to extend the existing classification
- The is an operational (practical) system, not a true scientific classification
- Others might devise special operational classifications for specific use, e.g., neonatal, ICU
- This classification is predominantly for clinicians
How Do Clinicians Classify Seizures?

- Elicit symptoms and signs of event (semiology)
- Look for familiar patterns in symptoms and signs
- Sometimes use ancillary data, e.g., EEG, MRI, genes, antibodies, etc.

**Examples**

- **Automatisms** ↔ **Focal impaired awareness seizure**
- **Absence seizure**

**Examples**

- **Automatisms** ↔ **Focal impaired awareness seizure**
- **Autonomic**
# Key Seizure Signs and Symptoms?

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Medical Term</th>
</tr>
</thead>
<tbody>
<tr>
<td>automatic behaviors</td>
<td>automatisms</td>
</tr>
<tr>
<td>emotions or appearance of emotions</td>
<td>emotions</td>
</tr>
<tr>
<td>extension or flexion postures</td>
<td>tonic</td>
</tr>
<tr>
<td>flushing/sweating/piloerection</td>
<td>autonomic</td>
</tr>
<tr>
<td>jerking arrhythmically</td>
<td>myoclonus</td>
</tr>
<tr>
<td>jerking rhythmically</td>
<td>clonus</td>
</tr>
<tr>
<td>language or thinking problems, deja vu</td>
<td>cognitive</td>
</tr>
<tr>
<td>lid jerks</td>
<td>eyelid myoclonia</td>
</tr>
<tr>
<td>limp</td>
<td>atonic</td>
</tr>
<tr>
<td>numb/tingling, sounds, smells, tastes visions, vertigo</td>
<td>sensations</td>
</tr>
<tr>
<td>pausing, freezing, activity arrest</td>
<td>behavior arrest</td>
</tr>
<tr>
<td>thrashing/pedaling</td>
<td>hyperkinetic</td>
</tr>
<tr>
<td>trunk flexion</td>
<td>spasm</td>
</tr>
</tbody>
</table>
The Elements of Change

- Allow some seizures to be either focal or generalized onset
- Classify seizures of unknown onset
- Clarify “impairment of consciousness”
- Include a few previously unclassified types
- Update word usage for greater public clarity
- Validate use of supportive information, e.g., EEG
- Conform with ICD 11 and 12
- Update the 2001 glossary of seizure terms
- Standardize common descriptors to describe seizures
- Map old to new terms
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Some Seizure Onsets can be Focal or Generalized

**Focal Onset**
- atonic
- clonic
- epileptic spasms
- myoclonic
- tonic
- tonic-clonic

**Generalized Onset**
- atonic
- clonic
- epileptic spasms
- myoclonic
- tonic
- tonic-clonic
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Seizures of Unknown Onset

Hypothetical case: You hear a noise and enter the video-EEG room to find the patient in bed, grunting, eyes rolled up, all limbs stiff, then rhythmically jerking for a minute. He was off-camera at the start. What seizure type is this?

Some seizure types are worth describing even if onset is unknown:
- tonic-clonic
- epileptic spasms
- behavior arrest
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Key Role of Impaired Consciousness

Among many possible behaviors during a seizure, impairment of consciousness has always had a key role in classifying the seizure, because of practical importance for:

- Driving
- Safety during seizures
- Employability
- Interference with schooling and learning
Loss (or Impairment) of Consciousness

Two types of seizures with loss of consciousness

How well does the public understand LOC during a complex partial seizure?
Loss (or Impairment) of Consciousness

Elements of consciousness

- **Awareness** of ongoing activities
- **Memory** for time during the event
- **Responsiveness** to verbal or nonverbal stimuli
- **Sense of self** as being distinct from others

Which would be the best surrogate marker?

- The 2017 Classification chooses awareness
- Consciousness remains in the classification but “awareness” is in the seizure name
- In several languages, these words are the same
- Awareness is not used to classify generalized onset seizures
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New Seizure Types

**New Focal Seizures**

<table>
<thead>
<tr>
<th>Motor</th>
<th>Non-Motor</th>
</tr>
</thead>
<tbody>
<tr>
<td>atonic</td>
<td>behavior arrest</td>
</tr>
<tr>
<td>automatisms</td>
<td>(autonomic)</td>
</tr>
<tr>
<td>clonic</td>
<td>(cognitive)</td>
</tr>
<tr>
<td>epileptic spasms</td>
<td>emotional</td>
</tr>
<tr>
<td>hyperkinetic</td>
<td>(sensory)</td>
</tr>
<tr>
<td>myoclonic</td>
<td></td>
</tr>
<tr>
<td>tonic</td>
<td></td>
</tr>
</tbody>
</table>

**New generalized seizures**
- absence with eyelid myoclonia
- epileptic spasms (infantile spasms)
- myoclonic-atonic (e.g., Doose)
- myoclonic-tonic-clonic (e.g., JME)

**New combined seizures**
- (focal to bilateral tonic-clonic)

(parentheses) indicates prior existence, but renaming
The Elements of Change

• Allow some seizures to be either focal or generalized onset
• Classify seizures of unknown onset
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• **Update word usage for greater public clarity**
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<table>
<thead>
<tr>
<th>OLD TERM</th>
<th>NEW TERM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unconscious (still used, not in name)</td>
<td>Impaired awareness (surrogate)</td>
</tr>
<tr>
<td>Partial</td>
<td>Focal</td>
</tr>
<tr>
<td>Simple partial</td>
<td>Focal aware</td>
</tr>
<tr>
<td>Complex partial</td>
<td>Focal impaired awareness</td>
</tr>
<tr>
<td>Dyscognitive (word discontinued)</td>
<td>Focal impaired awareness</td>
</tr>
<tr>
<td>Psychic</td>
<td>Cognitive</td>
</tr>
<tr>
<td>Secondarily generalized tonic-clonic</td>
<td>Focal to bilateral tonic-clonic</td>
</tr>
<tr>
<td>Arrest, freeze, pause, interruption</td>
<td>Behavior arrest</td>
</tr>
</tbody>
</table>
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Supportive Information

Seizures are usually classified by symptoms and signs
But supportive information may be helpful, when available:
• Videos brought in by family
• EEG patterns
• Lesions detected by neuroimaging
• Laboratory results such as detection of anti-neuronal antibodies
• Gene mutations
• Diagnosis of an epilepsy syndrome diagnosis
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ICD9, 10, 11, 12

- ICD 9 & 10 are in use now with old terminology: petit mal, grand mal
- ICD 11 does not name seizure types, but ILAE syndromes and etiologies
- ICD 12 should conform to the new ILAE seizure type classification

G40 Epilepsy and recurrent seizures
- G40.0 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes
  - G40.00 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndrome
    - G40.001 ...... with status epilepticus
    - G40.009 ...... without status epilepticus
  - G40.01 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndrome
    - G40.011 ...... with status epilepticus
    - G40.019 ...... without status epilepticus
- G40.1 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndrome
  - G40.10 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndr
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<table>
<thead>
<tr>
<th>WORD</th>
<th>DEFINITION</th>
<th>SOURCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>absence, typical</td>
<td>a sudden onset, interruption of ongoing activities, a blank stare, possibly a brief upward deviation of the eyes. Usually the patient will be unresponsive when spoken to. Duration is a few seconds to half a minute with very rapid recovery. Although not always available, an EEG would show generalized epileptiform discharges during the event. An absence seizure is by definition a seizure of generalized onset. The word is not synonymous with a blank stare, which also can be encountered with focal onset seizures.</td>
<td>Adapted from [11]</td>
</tr>
<tr>
<td>absence, atypical</td>
<td>an absence seizure with changes in tone that are more pronounced than in typical absence or the onset and/or cessation is not abrupt, often associated with slow, irregular, generalized spike-wave activity</td>
<td>Adapted from Dreifuss [1]</td>
</tr>
<tr>
<td>arrest</td>
<td>see behavioral arrest</td>
<td>new</td>
</tr>
<tr>
<td>atonic</td>
<td>sudden loss or diminution of muscle tone without apparent preceding myoclonic or tonic event lasting ~1 to 2 s, involving head, trunk, jaw, or limb musculature.</td>
<td>[11]</td>
</tr>
<tr>
<td>automatism</td>
<td>a more or less coordinated motor activity usually occurring when cognition is impaired and for which the subject is usually (but not always) amnesic afterward. This often resembles a voluntary movement and may consist of an inappropriate continuation of preictal motor activity.</td>
<td>[11]</td>
</tr>
</tbody>
</table>
The Elements of Change

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- Update the 2001 glossary of seizure terms
- **Standardize common descriptors to describe seizures**
- Map old to new terms
Common Descriptors of other symptoms and signs during seizures.

These are not seizure types, just suggested descriptive words.

A free text description is also highly encouraged.
The Elements of Change

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Examples of Mapping Old to New Terms
Full List in Epilepsia paper

<table>
<thead>
<tr>
<th>Old Term for Seizure</th>
<th>New Term for Seizure [choice] (optional)</th>
</tr>
</thead>
<tbody>
<tr>
<td>absence</td>
<td>(generalized) absence</td>
</tr>
<tr>
<td>absence, atypical</td>
<td>(generalized) absence, atypical</td>
</tr>
<tr>
<td>absence, typical</td>
<td>(generalized) absence, typical</td>
</tr>
<tr>
<td>akinetic</td>
<td>[focal/generalized] atonic</td>
</tr>
<tr>
<td>astatic</td>
<td>[focal/generalized] atonic</td>
</tr>
<tr>
<td>atonic</td>
<td>[focal/generalized] atonic</td>
</tr>
<tr>
<td>aura</td>
<td>focal aware</td>
</tr>
<tr>
<td>clonic</td>
<td>[focal/generalized] clonic</td>
</tr>
<tr>
<td>complex partial</td>
<td>focal impaired awareness</td>
</tr>
<tr>
<td>convolution</td>
<td>[focal/generalized] motor [tonic-clonic, tonic, clonic], focal to bilateral clonic</td>
</tr>
</tbody>
</table>
Onset: Decide whether seizure onset is focal or generalized, using an 80% confidence level.

Awareness: For focal seizures, decide whether to classify by degree of awareness or to omit awareness as a classifier.

Impaired awareness at any point: A focal seizure is a *focal impaired awareness seizure* if awareness is impaired at any point during the seizure.

Onset predominates: Classify a focal seizure by its first prominent sign or symptom. Do not count transient behavior arrest.

Behavior arrest: A *focal behavior arrest seizure* shows arrest of behavior as the prominent feature of the entire seizure.

Motor/Non-motor: A *focal aware or impaired awareness seizure* maybe further sub-classified by motor or non-motor characteristics. Alternatively, a focal seizure can be characterized by motor or non-motor characteristics, without specifying level of awareness. Example, a *focal tonic seizure*.
Rules for Classifying Seizures (2 of 2)

Optional terms: Terms such as motor or non-motor may be omitted when the seizure type is otherwise unambiguous.

Additional descriptors: It is encouraged to add descriptions of other signs and symptoms, suggested descriptors or free text. These do not alter the seizure type. Example: *focal emotional seizure* with tonic right arm activity and hyperventilation.

Bilateral vs. generalized: Use the term “bilateral” for tonic-clonic seizures that propagate to both hemispheres and “generalized” for seizures that apparently originate simultaneously in both.

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**Atypical absence**: Absence is atypical if it has slow onset or offset, marked changes in tone or EEG spike-waves at less than 3 per second.

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Eyelid myoclonia: Absence with eyelid myoclonia refers to forced upward jerking of the eyelids during an absence seizure.
The net effect of updating the Classification of Seizures should be the following:

1. Render the choice of a seizure type easier for seizures that did not fit into any prior categories;

2. Clarify what is meant when a seizure is said to be of a particular type;

3. Provide more transparency of terminology to the nonmedical community.
1. A woman awakens to find her husband having a seizure in bed. The onset is not witnessed, but she is able to describe bilateral stiffening followed by bilateral shaking. EEG and MRI are normal.
1. A woman awakens to find her husband having a seizure in bed. The onset is not witnessed, but she is able to describe bilateral stiffening followed by bilateral shaking. EEG and MRI are normal. This seizure is classified as onset unknown tonic-clonic. There is no supplementary information to determine if the onset was focal or generalized. In the old classification, this seizure would have been unclassifiable.
2. In an alternate scenario of case #1, the EEG shows a clear right parietal slow wave focus. The MRI shows a right parietal region of cortical dysplasia.
2. In an alternate scenario of case #1, the EEG shows a clear right parietal slow wave focus. The MRI shows a right parietal region of cortical dysplasia. In this circumstance, the seizure can be classified as focal to bilateral tonic-clonic, despite the lack of an observed onset, because a focal etiology has been identified, and the overwhelming likelihood is that the seizure had a focal onset. The old classification would have classified this seizure as partial onset, secondarily generalized seizure.

**Old** = partial onset, secondarily generalized seizure
**New** = focal to bilateral tonic-clonic seizure
Examples

3. A child is diagnosed with Lennox-Gastaut syndrome of unknown etiology. EEG shows runs of slow spike-wave. Seizure types include absence and others.
3. A child is diagnosed with Lennox-Gastaut syndrome of unknown etiology. EEG shows runs of slow spike-wave. Seizure types with this child include absence, tonic, and focal motor seizures. In this case, the absence seizures are classified as atypical absence (the word “generalized” may be assumed) due to the EEG pattern and underlying syndrome. The absence seizures would have had the same classification in the old system.
Examples

4. The same child as in #3 has seizures with stiffening of the right arm and leg, during which responsiveness and awareness are retained.
4. The same child as in #3 has seizures with stiffening of the right arm and leg, during which responsiveness and awareness are retained. This seizure is a focal aware tonic seizures (the word “motor” can be assumed). In the old system, the seizures would have been called tonic seizures, with a perhaps incorrect assumption of generalized onset.

Old = tonic seizures
New = focal aware tonic seizures

Examples
5: A 25 year old woman describes seizures beginning with 30 seconds of an intense feeling that “familiar music is playing.” She can hear other people talking, but afterwards realizes that she could not determine what they were saying. After an episode, she is mildly confused, and has to “reorient herself.”
5: A 25 year old woman describes seizures beginning with 30 seconds of an intense feeling that “familiar music is playing.” She can hear other people talking, but afterwards realizes that she could not determine what they were saying. After an episode, she is mildly confused, and has to “reorient herself.” The seizures would be classified as focal seizures with impaired awareness. Even though the patient is able to interact with her environment, she cannot interpret her environment, and is mildly confused.
6. A 22 year-old man has seizures during which he remains fully aware, with the “hair on my arms standing on edge” and a feeling of being flushed.
6. A 22 year-old man has seizures during which he remains fully aware, with the “hair on my arms standing on edge” and a feeling of being flushed. These are classified as focal aware non-motor autonomic, or more succinctly focal aware autonomic. The old classification would have called them simple partial autonomic seizures.
7. A 4 year-old boy with myoclonic-atonic epilepsy (Doose syndrome) has seizures with a few arm jerks, then a limp drop to the ground.
7. A 4 year-old boy with myoclonic-atonic epilepsy (Doose syndrome) has seizures with a few arm jerks, then a limp drop to the ground. These are now classified as myoclonic-atonic seizures (the word “generalized” may be assumed). The old classification would have called these unclassified or unofficially, myoclonic-astatic seizures.

Old = myoclonic astatic seizures
New = myoclonic-atonic seizures
8. A 35 year-old man with juvenile myoclonic epilepsy has seizures beginning with a few bilateral arm jerks, followed by stiffening of all limbs and then rhythmic jerking of all limbs.
8. A 35 year-old man with juvenile myoclonic epilepsy has seizures beginning with a few regularly-spaced jerks, followed by stiffening of all limbs and then rhythmic jerking of all limbs. This would be classified as generalized myoclonic-tonic-clonic seizures. No corresponding single seizure type existed in the old classification, but they might have been called myoclonic seizures followed by a tonic-clonic seizure.

\[
\begin{align*}
\text{Old} &= \text{myoclonic seizures followed by a tonic-clonic seizure} \\
\text{New} &= \text{myoclonic-tonic-clonic seizures}
\end{align*}
\]
9. A 14-month old girl has sudden flexion of both arms with head flexing forward for about 2 seconds. These seizures repeat in clusters. EEG shows hypsarrhythmia with bilateral spikes, most prominent over the left parietal region. MRI shows a left parietal dysplasia.
9. A 14-month old girl has sudden flexion of both arms with head flexing forward for about 2 seconds. These seizures repeat in clusters. EEG shows hypsarrhythmia with bilateral spikes, most prominent over the left parietal region. MRI shows a left parietal dysplasia. Because of the ancillary information, the seizure type would be considered to be focal epileptic spasms (the term “motor” can be assumed). The previous classification would have called them infantile spasms, with information on focality not included.

Old = infantile spasms (focality not specified)
New = focal epileptic spasms
10. A 75 year-old man reports an internal sense of body trembling. No other information is available.
“Words, words, words, I’m so sick of words!”
Eliza Doolittle, *My Fair Lady*