

## Research Work

**Acquired epileptic aphasia in children (landau-kleffner syndrome): case report from a pediatric neurology clinic of a tertiary health facility in nigeria.****Igwe Wilson C\*<sup>1</sup>, Umeadi Esther N<sup>2</sup>, Ezeudu Chijioke E<sup>3</sup>, Chukwuka John O<sup>4</sup>,**Senior Lecturer, Department of Pediatrics Faculty of Medicine, College of Health Sciences, Nnamdi Azikiwe University Nnewi Campus Anambra State, Nigeria<sup>1</sup>Lecturer, Department of Pediatrics Faculty of Medicine, College of Health Sciences Nnamdi Azikiwe University Teaching, Nnewi Campus Anambra State, Nigeria<sup>2</sup>Senior Lecturer, Department of Pediatrics Faculty of Medicine, College of Health Anambra State, Nigeria<sup>3</sup>Prof of Pediatrics, Dept of Paediatrics Faculty of Medicine, College of Health Sciences Nnamdi Azikiwe University Nnewi Campus, Anambra State, Nigeria<sup>4</sup>Email id: [wc.igwe@unizik.edu.ng](mailto:wc.igwe@unizik.edu.ng)<sup>1</sup>, [zigor2k3@yahoo.com](mailto:zigor2k3@yahoo.com)<sup>2</sup>, [ezeuducj@yahoo.com](mailto:ezeuducj@yahoo.com)<sup>3</sup>, [onuorachukwuka@yahoo.com](mailto:onuorachukwuka@yahoo.com)<sup>4</sup>**Corresponding author:** Igwe Wilson C Department of Pediatrics, Faculty of Medicine, CHS Nnamdi Azikiwe University, Nnewi Campus. Anambra State, Email: [wc.igwe@unizik.edu.ng](mailto:wc.igwe@unizik.edu.ng)

Telephone: +2347036531408

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**Abstract:**

Acquired epileptic aphasia is a rare childhood epileptic encephalopathy characterized by acquired aphasia, paroxysmal electroencephalographic pattern, behavioral and psychomotor disturbances. Seizures may not occur in all cases, and when present are infrequent and often remits spontaneously with age. It is often the aphasia that brings most patients to the physicians' attention. Children who have not developed language before onset of the seizures/paroxysmal EEG pattern should not be considered for this syndrome. It is important that acquired epileptic aphasia be excluded in patients suspected to have autism, attention deficit disorder, aggressive, oppositional or psychotic behaviors. A multidisciplinary approach comprising of pediatric neurologists, psychologists, audiologists and speech therapists are necessary for early identification and management in order to restore reasonable language function in these children. This report describes 3 cases of this rare syndrome seen in an outpatient pediatric neurology clinic of a tertiary health facility.

**Keywords:** Acquired epileptic aphasia, Landau-Kleffner syndrome, case reports**Introduction:**

Acquired epileptic aphasia, also known as Landau Kleffner syndrome (LKS) is a rare age-dependent epileptic encephalopathy characterized by acquired aphasia, abnormal EEG pattern, behavioral and psychomotor disturbances.<sup>1</sup>This childhood encephalopathy was first described by Landau and Kleffner in 1957 as acquired

Childhood aphasia with convulsive disorder. It presents typically as a sudden or gradual loss of language skills in a previously normal child in association with paroxysmal electroencephalographic (EEG) changes.<sup>1</sup>

Behavioral disorders such as attention deficit hyperactivity, impulsivity, aggression, and other bizarre behaviors commonly accompany the language impairment. The prevalence and incidence of LKS is unknown but has been noted to be commoner in males than females.<sup>2</sup> The literature on this rare encephalopathy is scarce in Africa and to the best of our knowledge only one case has been reported in Nigeria.<sup>3</sup>

Acquired aphasia in previously normal children which is the defining feature of this disorder develops between the ages of 3-9 years with about

70% presenting by 6 years. Some of these patients may recover the language skills fully, partially or remain aphasic for variable periods.<sup>4</sup>

Behavioral and psychomotor disturbances are seen in about 75% of the cases following the aphasia.<sup>4</sup> the most common behavioral abnormalities are hyperactivity, impulsivity and outbursts of rage. Other symptoms noted in these patients include anxiety disorder, avoidance of interpersonal contact and other bizarre behaviors.

About 70-85% of patients with LKS will have seizures which may precede, follow or occur concurrently with onset of aphasia.<sup>5</sup>The seizure when present is infrequent, easy to control and often remits spontaneously with age. About 30% of patients may even have only an episode of seizures in this syndrome. In some cases a few months or even years may pass between seizures and the onset of aphasia.<sup>6,7</sup> The seizures may be nocturnal simple partial seizures, myoclonic seizures, generalized tonic-clonic seizures or atypical absences.<sup>8,9</sup> The paroxysmal EEG finding is the diagnostic indicator in LKS.<sup>4,10</sup> This abnormal EEG pattern is most evident in sleep recording.

Children with LKS can easily be misdiagnosed, since the outstanding features may resemble autism, emotional or behavioral disorder, or other types of acquired aphasia.

The etiology of LKS is unknown but possible proposed etiologies include sub-acute bitemporal encephalitis, neurocysticercosis and cerebral vasculitis.<sup>4</sup>

Currently there are no standard treatments for acquired epileptic aphasia. Many treatment modalities such as anticonvulsant drugs, corticosteroids (adrenocorticotropin hormone [ACTH], prednisolone), ketogenic diet, and surgical interventions have been used. Valproic acid, ethosuximide and benzodiazepines alone or in combination have been effective in controlling the seizures and reversing the EEG abnormalities in LKS. Anticonvulsants such as phenobarbitone, carbamazepine and pheytoin have not been found to be effective and may even worsen the EEG abnormalities. Steroids have been linked to improvements or at least stabilization of language abilities. Prednisolone at 2mg/kg/day for 1 month, followed by reduction to 1mg/kg/day for another 1 month is recommended.<sup>1</sup> with improvement this is further reduced to 0.5mg/kg/day for 6-12 months. Steroid reduction may be associated with

recurrence of symptoms and thus treatment may be prolonged.

All patients with acquired epileptic aphasia should be referred for audiologic evaluation and speech therapy and special education may be necessary.

Psychotherapy and psychiatric consultation are indicated in patients with behavioral problems who may likely need behavioral therapy and drug management.

### **Case 1**

CK was a 6 ½ year old male who presented with loss of language ability of 4weeks duration following 2 episodes of generalized seizures. Other aspects of the history including delivery were essentially normal. Except for the aphasia, other aspects of the physical examination were normal. The EEG could not be done due to financial constraints. He was placed on tab carbamazepine. He remained seizure free and recovered his language skills a month later but was lost to follow up.

### **Case 2**

AC was a 4yr old male that presented with a 2 month history of a sudden loss of language ability, impulsivity and hyperactivity following an episode of prolonged generalized tonic-clonic seizure. He was previously having recurrent episodes of myoclonic seizures of a year's duration. The birth history was unremarkable. Other aspects of the history and neurological examination were essentially normal. The sleep electroencephalogram (EEG) showed generalized high amplitude spikes mainly on the left hemisphere. The cranial computed scan (CT scan) was normal. He has remained seizures-free during the 8 months follow- up on valproic acid and prednisolone. He is still aphasic and hyperactive. There are currently no facilities in our centre to address the speech impairment however the impulsivity and hyperactivity are being managed in conjunction with the child psychologists.

### **Case 3**

AZ was a 6yr old male who presented with 8 months history of aphasia and abnormal behavior after an episode of generalized tonic-clonic seizures. The abnormal behaviors consisted of being withdrawn and eating anything including his clothing. He had a past medical history of recurrent generalized seizures of 5 years' duration. The birth history was essentially normal. On examination he had macrocephaly and exhibited

some inappropriate behaviors. The receptive and expressive language skills were absent. The sleep EEG showed bilateral high amplitude slow waves with multifocal spikes mainly at the left temporo-parietal area. The cranial CT scan was essentially normal. He was commenced on Sodium valproate and prednisolone. He has been seizure-free during the 10-months follow-up period but the language skills and the abnormal behavior have persisted. The abnormal behaviors are being managed by the psychiatrists.

### **Discussion:**

The defining feature of LKS is the onset of acute or progressive loss of language ability in a previously normal child coincident with the appearance of paroxysmal EEG patterns that are compatible with epilepsy.<sup>5</sup> All the patients in this report had seizures prior to the onset of the aphasia. The aphasia also developed within the age group characteristic of LKS. The two patients who did EEG all had paroxysmal EEG patterns that were compatible with the diagnosis of epilepsy.

The most obvious symptom of LKS is often the acute and sometimes gradual language regression in a previously normal child. The patients in this case report presented due to the aphasia and the behavioral abnormalities.

The neurologic examination and neuroimaging studies which were essentially normal in the cases presented in this report helped further in confirming LKS and exclude other causes of aphasia in these patients.

Two of the patients presented with behavioral abnormalities such as hyperactivity, impulsivity and other bizarre behaviors which were in keeping the findings in LKS.

The prognosis with regards to seizures was good as the seizures in all the 3 cases in this report are under control. Caraballo et al<sup>5</sup> in a case series involving 29 patients that were followed up for 12yrs also noted that all were seizure free during the follow-up period.

While aphasia persisting beyond 1year of onset is said to be predictive of poor language recovery, there is yet no agreement on the influence of age of onset of aphasia and the prognosis for language recovery.<sup>4,8,10</sup> The prognosis for language recovery was variable in this report as only one of the 3 cases regained the language ability. This occurred in the oldest of the patients who was

aged 6yrs 6months. Robinson et al<sup>8</sup> and Duran et al<sup>10</sup> in separate follow up studies did not observe any association between age of onset of aphasia and outcome in language recovery. Caraballo et al<sup>5</sup> also observed that normalization or a significant improvement of the EEG was a necessary condition for complete language recovery, but however not all their patients with a normalized EEG recovered language.

In a follow up study of 7 patients by Duran et al<sup>10</sup>, 1 patient recovered language completely, 3 recovered language partially while the rest did not recover language during the study period.

In conclusion, LKS is rare and can easily be misdiagnosed since the outstanding features may suggest other diagnoses. There is therefore a need for increased awareness and early intervention in this rare childhood epilepsy syndrome.

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