Acquired motor speech disorders in childhood epilepsy

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Abstract

Aim

To evaluate a cohort of children with epilepsy and motor speech regression, with the aim of characterising their speech disorders, electrographic features and outcomes.

Method

Children referred to a tertiary Developmental Epilepsy Clinic with epilepsy and motor speech regression were retrospectively identified. Clinical history, speech and cognitive assessments and outcomes were recorded. Speech samples were scored for severity and speech features. Seizure frequency and epileptiform discharges in the interictal EEG were analysed.

Results

Eighteen children (ten female) were evaluated including seven with Landau-Kleffner syndrome (LKS) and six with Rasmussen syndrome (RS). Speech regression occurred at mean age 5 years (SD 2 years 6 months), concurrent with seizure onset or peak seizure burden in 47%. Speech features included dysarthria (74%), phonological errors (39%) and dyspraxia (33%). Electrographic abnormalities occurred most frequently in left centrotemporal and right frontal regions. Intelligibility of speech was affected in 83% of patients at baseline and 50% at follow-up. Expressive language standardised scores increased from mean 50.0 to 91.4 in LKS ($p=0.0498$) and decreased from 75.2 to 59.0 in RS ($p=0.002$) over follow-up.

Interpretation

Motor speech disorders in epilepsy were severe, multifarious, and often fluctuated with seizure burden. Symptoms typically improved, especially in LKS, but rarely fully resolved.

What this paper adds

- Acquired motor speech disorders in childhood epilepsy are complex and heterogeneous.
- Electrographic abnormalities are observed most frequently in brain regions involved in speech production.
- The typical course shows improvement over time but full resolution is rare.
- Interventions focus on bridging the communication gap between understanding and expression.

Short title: Acquired motor speech disorders in epilepsy

Language disorders occur commonly in children with early onset epilepsies, usually conceptualised as a consequence of the underlying aetiology (e.g. a focal epilepsy in the left hemisphere) causing language impairments which can emerge over time as the child reaches a certain developmental level. Another group of children with epilepsy syndromes such as Landau-Kleffner syndrome (LKS), a specific subtype of EE-SWAS (epileptic encephalopathy with spike-and-wave activation in sleep),[1] can present with language regression due to acquired disturbances in the symbolic processing of language (aphasia).[2] In contrast, motor speech disorders are characterised by impairments in the motor planning (dyspraxia) or execution (dysarthria) of speech. Motor speech disorders are less commonly seen in childhood epilepsy, except as part of the underlying aetiology that has led to the seizure disorder (e.g. cerebral palsy), or as transient ictal or post-ictal phenomena (e.g. in SeLECTS [self-limited epilepsy with centrotemporal spikes syndrome][13]). However children with epilepsy can also present with acquired, persistent and clinically significant motor speech difficulties. The presentation is challenging to assess and manage due to its rarity, difficulties in distinguishing motor speech characteristics from aphasia (which may be co-existent), complex interactions with normal speech development, underlying aetiology and fluctuating neurological symptoms, and a lack of published data to inform practice.

Here we describe a cohort of 18 children with epilepsy and acquired motor speech regression, with the aim of characterising their clinical histories, speech characteristics, electrographic features and outcomes.

Methods

Setting and institutional approval

This was a retrospective, single-centre study performed in the Developmental Epilepsy Clinic at Great Ormond Street Hospital. Patients were referred from secondary and tertiary paediatric epilepsy services (including the NHS England Children's Epilepsy Surgery Service) for diagnosis and management of developmental difficulties associated with epilepsy. Patients were assessed by a multi-disciplinary team including paediatricians, occupational therapists, speech and language therapists (SLT) and psychologists. All study data was collected as part of routine clinical care. The study was approved as a service evaluation (#2949) by Great Ormond Street Hospital for Children NHS Foundation Trust.

Participants

Patients were evaluated in the clinic between 2016 and 2019 and met the following inclusion criteria at first assessment: (1) age 3-16 years; (2) diagnosis of epilepsy; (3) history of regression in speech due to an acquired speech disorder (dysarthria, dyspraxia or phonological). Patients who had undergone epilepsy surgery prior to the first assessment were excluded.

Clinical record review

Data were collected on demographics, developmental history, speech history, epilepsy syndromic and aetiological diagnosis and seizure history including peak seizure burden. Seizure types were classified according to ILAE criteria.[4]

Speech features and psychometric assessments

SLT reports were reviewed and descriptions of speech features extracted. Example features noted included developmental phonological processes, slow speech rate, consonant imprecision, vowel distortion and fluency changes. Note was made of any augmentative communication methods used. Expressive language standardised scores were obtained using the Clinical Evaluation of Language Fundamentals (CELF) or CELF Preschool (PS-CELF) if available, otherwise the Preschool Language Scale (PLS), Expressive Vocabulary Test (EVT) or expressive communication subscale of the Vineland Adaptive Behaviour Scales (VABS) (in descending order of preference).[5–9] Receptive language standardised scores were obtained using the CELF, PS-CELF, PLS, Peabody Picture Vocabulary Test (PPVT) or receptive communication subscale of the VABS.[10] Nonverbal cognitive standardised scores were obtained using the Wechsler Intelligence Scale for Children (WISC), Wechsler Preschool & Primary Scale of Intelligence (WPPSI), Wechsler Adult Intelligence Scale (WAIS) or Wechsler Nonverbal Scale of Ability (WNV).[11–14] A summary nonverbal cognitive score was calculated by taking the Nonverbal Index of the WISC/WPPSI/WAIS if available, otherwise the median of the available non-verbal indices (Visual Spatial Index, Fluid Reasoning Index, Processing Speed Index, Perceptual Reasoning Index/Perceptual Organisation Index) from the WISC/WPPSI/WAIS, or the WNV composite score. In rare cases where scores were only available as age equivalences, these were converted to a developmental quotient (cognitive age ÷ chronological age x 100).

Speech sample review

Audiovisual samples of speech were obtained in a subset of children as part of the routine clinic evaluation at first assessment. Written consent to obtain, store and retrospectively analyse recordings was routinely gathered in clinical practice. Speech samples were reviewed by an SLT independent of the original clinic assessment, including three different tasks (such as conversation, naming, repetition etc) with a minimum sample duration of 5 minutes. Speech samples were rated using audio-perceptual analysis with the Mayo Dysarthria Rating Scale.[15] Audio-perceptual changes in speech domains were identified and categorised. An overall severity rating was given using the Motor Speech Disorders Severity Rating (MSDSR) Scale (modified from Hillel et al.[16]), in which scores range from 1 (nonvocal)

to 10 (normal speech). A subset of samples were independently rated by another SLT (blinded to the first rater) to confirm consistency.

EEG review

Neurophysiologists' reports of EEGs acquired after onset of speech regression were reviewed for mention of epileptiform abnormalities, including continuous spike-wave during slow wave sleep (CSWS) or electrical status epilepticus in slow-wave sleep (ESES). EEG recordings acquired in the neurophysiology department of Great Ormond Street Hospital for Children after onset of speech regression and including a period of sleep were analysed where available. Where multiple recordings were available the one acquired closest to the first speech assessment was used. The standard EEG setup used 27 EEG electrodes placed according to the international 10-10 system. Recordings were analysed to quantify interictal epileptiform discharges (IEDs), reviewing the first three hours of awake EEG (sampling 60 seconds every 10 minutes) and the first one hour asleep (sampled in full). Each one-second interictal epoch was scored for the presence or absence of ≥1 IED using International Federation of Clinical Neurophysiology criteria.[17] The IED proportion was calculated as the number of epochs with ≥1 IED divided by the total number of epochs reviewed. Electrode locations showing maximal IED peak deflection on the common-average reference voltage maps (IED maxima) were also noted. The neurophysiologist's report was used to quantify seizure frequency and electrode locations implicated in seizure origin.

Statistical analysis

Patients were organised into three groups (LKS, Rasmussen syndrome [RS] and others) to evaluate changes in standardised scores from first assessment to final follow-up using paired t-tests. The proportion of patients with impaired speech intelligibility was compared between initial assessment and final follow-up, and within final follow-up according to improvement in the interictal EEG, using Fisher's exact test. Two-tailed p-values <0.05 were regarded as significant. The topographical distributions of IED maxima and suspected seizure origin locations were visualised as contour maps, including only electrodes common across all recordings. Standardised scores are summarised as mean ± standard deviation. Analyses were carried out in Python 3.10 with *Scipy* v1.10.0.

Results

Participant characteristics

Eighteen children (ten female) met the inclusion criteria: seven with LKS (three with GRIN2A mutation), six with RS (four affecting the left hemisphere), three with other generalised epilepsies (including two with myoclonic atonic seizures), and two with other focal epilepsies (**Table 1**). There were seizure events

in 17/18 (94%) children including 13/18 (72%) with focal seizures and 9/18 (50%) with generalised seizures. Mean (SD) age at seizure onset was 5 years (2 years 6 months) and at speech regression was 5 years 11 months (2 years 6 months). Speech regression occurred within three months of either seizure onset or peak seizure burden in 8/17 (47%). Median seizure frequency at peak was 6/day (≥1/day in 11/17 [65%]). 16/18 (89%) children were on anti-seizure medications at the time of first assessment (median 2.5). Handedness prior to disease onset was recorded in 16/18, all right-handed.

Speech features and psychometric assessments at first assessment

There were pre-existing speech/language concerns in 10/18 children (56%). Speech features identified included dysarthria in 13/18 (74%), phonological errors in 7/18 (39%), dyspraxia in 6/18 (33%), stammer in 3/18 (17%) and dysphonia in 2/18 (11%) (**Table 2**). Among those with dysarthria, 7/13 (57%) had a history of drooling and 4/13 (36%) had a history of dysphagia. 9/18 (50%) had features that could be consistent with more than one type of childhood speech sound disorder, including 2/18 (11%) with features of both dysarthria and dyspraxia. Intelligibility of speech was affected in 15/18 (83%) (**Figure 1**). Mean (SD) standardised scores at first assessment were 64.9 (18.9) for expressive language, 70.6 (16.8) for receptive language and 77.0 (20.2) for nonverbal cognition (**Figure 2**).

Speech sample review

Audiovisual speech samples were available in 12 children (**Table 3**). All samples showed features of dysarthria across multiple domains, most prominently articulation (median 2/5 [40%] features per child) and prosody (median 3.5/10 [35%] features per child), while respiratory support of speech was least affected. 8/12 (67%) children scored at least one feature in every dysarthria domain that could be assessed except respiration. Median MSDSR score was 6.5 (obvious speech abnormalities/repeats messages on occasion). Scoring was generally consistent between the two independent raters, with complete concordance in 14/21 (67%) domain feature counts and a maximum discrepancy in MSDSR score of one (**Supplementary Table 1**).

EEG review

EEG after speech regression was reported to show generalised discharges or electrographic seizures in 11/18 children (61%) and focal abnormalities in 15/18 (83%), left-hemispheric in 12/18 (67%) and right-hemispheric in 9/18 (50%), localised predominantly to central and fronto-temporal regions. EEG recordings were analysed in 12 children (**Supplementary Table 2**). IEDs were identified in 11/12 recordings (92%); the median proportion of the recording containing IEDs in wakefulness was 14% (IQR 3.6-26%) and in sleep was 11% (IQR 4.2-49%). IED maxima were localised predominantly to centrotemporal and frontal regions (**Figure 3A**). Seizure events were identified in 9/12 recordings (75%),

with a median 6.5 events per recording (IQR 1.5-22). Regions frequently implicated in seizure origin included right frontal and left centrotemporal (**Figure 3B**).

Follow-up

Fourteen children were followed up after median 3 years 1 month (**Table 2**). There were persisting motor speech difficulties in 13/14 (93%), although 9/14 (64%) had some degree of improvement; the proportion with impaired intelligibility of speech decreased from 15/18 (83%) at first assessment to 7/14 (50%) at final follow-up (p=0.06) (**Figure 1**). Four children with RS had epilepsy surgery (hemispherotomy) during the follow-up interval; excluding these, there was impaired intelligibility of speech in 1/5 of those with improvement of IED burden on follow-up EEG, compared to 3/3 of those without interictal EEG improvement (p=0.14). Mean (SD) standardised scores at follow-up were 76 (25) for expressive language, 77 (24) for receptive language and 75 (22) for nonverbal cognition. Mean expressive language scores in LKS increased by 41.4 points (95% CI 0.04 to 82.8, p=0.0498) from first assessment to follow-up (mean 91.4) and in RS decreased by 16.2 points (95% CI 9.0 to 23.4, mean at follow-up 59.0, p=0.002) (**Figure 2**). There were no significant changes in other standardised scores.

Discussion

This study is the first to our knowledge to characterise acquired motor speech disorders in a cohort of children with epilepsy. Dysarthria, phonological errors and dyspraxia were the most frequent speech disorders and a mixed picture was typical. Every speech sample assessed demonstrated multiple dysarthria features, most notably affecting speech articulation and prosody. These problems impacted significantly on functioning with 83% of children having impaired intelligibility of speech at initial assessment. Difficulties typically persisted with some improvement over time, and with potential for significant improvements in some children, especially those with LKS and/or those whose interictal EEG improved.

Speech differences were highly variable, tending to fluctuate from day to day, particularly in association with seizures. Epilepsy was typically severe, with most children receiving multiple medications and with a history of daily seizures. The initial motor speech regression was often temporally associated with either epilepsy onset or peak seizure burden. Interictal epileptiform activity was frequently localised to brain regions involved in speech production (**Figure 3A**), such as the left centrotemporal electrodes overlying the face and mouth regions of the motor cortex. The right prefrontal cortex was the most frequently implicated region in seizure origin (**Figure 3B**); this region has an important role in the production of speech prosody, impairments in which were universally observed in our speech samples. In adults,

acquired injuries to the right prefrontal cortex are associated with prosodic deficits, and seizures arising from the right hemisphere and frontal lobes are associated with ictal dysprosody.[18–21] Impaired prosody has also been reported in right frontal epilepsy of childhood onset.[22] While intriguing, these EEG localisations in our cohort are based on small numbers of patients, and focal EEG abnormalities were reported in many other brain regions across both hemispheres, consistent with an overall view of speech production as a function of bilaterally distributed large-scale network interactions[23] which may be disrupted in epilepsy by many different mechanisms.

Despite striking impairments at presentation, children with LKS had the most favourable motor speech and broader language/cognitive outcomes. In two children with LKS who did not improve (cases 6 and 10), one had ongoing seizures and the other ongoing epileptiform activity on EEG; good outcomes were generally associated with successful management of the epilepsy syndrome. Conversely, children with RS tended to progressively decline in language and cognitive skills with or without surgical management of the epilepsy and overall had less improvement in motor speech than other children, although 3/4 RS children managed with hemispherotomy did have some motor speech improvement, unlike 2/2 managed without.

Implications for assessment

The determination of acquired motor speech difficulties in children is complex; acquired characteristics can be hard to differentiate from other childhood speech-sound disorders, and many present with a mixed picture of dysarthria and dyspraxia, highlighting the limited applicability of conventional adult diagnostic speech categories in children. In contrast to children, dysarthria in adults is usually acquired. It is typically classified by the observed motor disorder (spastic, hypokinetic, hyperkinetic, ataxic, dyskinetic, dystonic, flaccid or mixed) and the presumed site or cause of the brain damage or dysfunction. The validity of applying this to children has been questioned.[24–26] Our data supports the notion that it is not possible to neatly categorise subtypes of dysarthria in children with acquired motor speech disorders. Children who acquire a motor speech difficulty in the context of epilepsy should receive an in-depth speech assessment using perceptual, structured and standardised assessment tools to document clearly what can be seen and heard. Detailed description of the baseline profile is essential for differential diagnosis and mapping changes over time; capturing an audiovisual sample of speech is highly recommended. We also recommend using a recently developed tool to assist differentiation of dysarthria and dyspraxia in children.[27]

Implications for intervention

At first assessment in our tertiary/quaternary service, 83% of children had impaired speech intelligibility to the level that would reduce participation in learning and social interactions and impact on self-esteem, however only 9/18 (50%) were already known to an SLT service, and often regarding language rather than speech needs. We referred a further 7/18 (39%) to SLT following our assessment. Thus, it seemed that SLT intervention is often not provided for children with acquired motor speech disorders and so we reviewed our own recommendations and potential barriers to accessing therapy for these children.

Children with acquired deficits are very different from those who have grown up with motor speech difficulties, as they have a background of successfully using speech as their main mode of expressing themselves and are often initially highly motivated communicators. Where speech is lost rapidly and completely (e.g. LKS) these children naturally use gesture to compensate and often do very well with sign language, as their motor issues do not preclude signing. In our cohort we recommended signing for 9/18 (50%) children, including 6/7 (86%) in the LKS group. In LKS, speech (and language) may recover with treatment and time, and only 1/5 (20%) LKS children still required signing long term. Signing is obviously more difficult where there is a motor deficit affecting hand function as in RS; we only recommended signing for one child with RS, who was at an early cognitive/language level, and we recommended Makaton to support his understanding and expression. An alternative way to bridge a communication gap between what a child understands and is able to express is to use a communication device and this was recommended for 4/18 (22%), including two children with RS. When speech deteriorates very gradually or is very fluctuant (e.g. varying with seizure burden) it can be challenging to engage children in alternative communication methods; often such children continue to have a strong preference to use speech as their main mode of communication even when it becomes unintelligible, and deterioration/fluctuation is over such a protracted time (with repeated experiences of communication failure) that they gradually become less persistent and motivated when expressing themselves. This group would probably benefit from early intervention to support their communication, particularly where the aetiology is known to be progressive (e.g. RS).

Although this cohort was defined by motor speech difficulties, we only recommended a trial of direct speech work (therapy focused on speech sound targets or speech quality features) for one child (a child with generalised epilepsy) and this was ineffective; and we know of another child with LKS who has had repeated courses of direct speech work with little impact. It could be that direct speech work is ineffective during periods of active seizures but helpful to build skills once seizures have settled, and this needs to be explored in future work. For all children we recommended using visuals to support communication, for both getting their message across and understanding. We made general recommendations around optimising intelligibility (e.g. attention to rate of speech, breath support etc.) and communication

competence (for both partners) and enhancing the communication environment. We emphasised to families that these children often have a fluctuant picture and are very vulnerable to fatigue, which needs to be taken into account in their daily lives and when planning any therapy. Impacts on social development and peer interactions should also be considered; it is concerning that three children in our cohort had been diagnosed with autism at final follow-up, with another currently awaiting assessment.

Limitations and implications for future research

Our study was limited by a small and heterogeneous cohort which restricted statistical power and precluded multivariable analysis. Assessments and follow-up were tailored to each child's needs rather than standardised. In future studies it would be advantageous to combine data from multiple centres; by providing detailed individualised data we hope to facilitate this. A key aim for future research will be to expand the evidence base for intervention, with study designs informed by extrapolation from evidence in other motor speech disorders. Patients should be evaluated longitudinally to capture fluctuations over time; the optimum timing for intervention will be an important consideration.

Conclusions

Acquired motor speech disorders in the context of childhood epilepsy are rare but important to recognise as they can impact significantly on quality of life and require thorough assessment. A mixed profile of speech characteristics is typical, including diverse manifestations of dysarthria alongside other speech features such as phonological errors and dyspraxia. Symptoms may fluctuate with seizure burden and interictal EEG activity. The typical course shows improvement over time but full resolution is rare, leading to long-term needs and highlighting the importance of compensatory therapy approaches to support communication.

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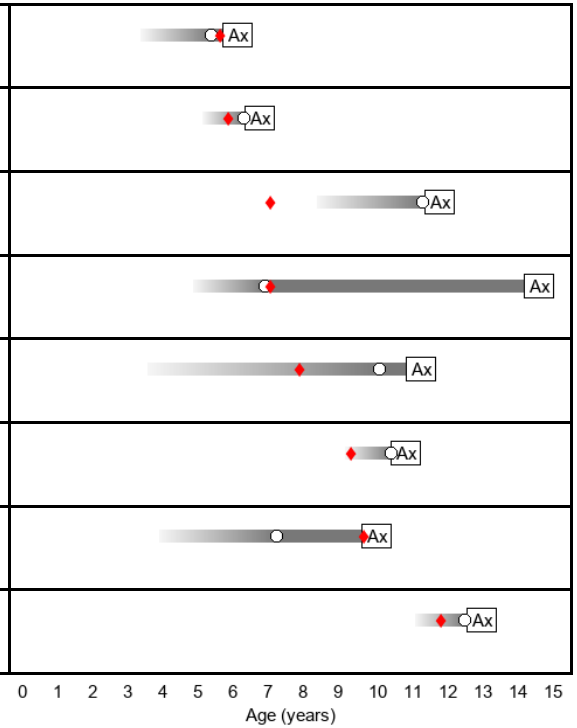
Tables

Table 1: Epilepsy clinical features and chronology

Case no.	Sex	Epilepsy syndromic diagnosis	Epilepsy aetiological diagnosis	Affected hemi-sphere	Age at seizure onset (years: months)	Age at seizure nadir (years: months)	Seizure burden at nadir	Generalised seizure types	Focal seizure types	Age at motor speech regression (years: months)	Age at first assessment (years: months)	Chronology
1	M	Structural/lesional	Parieto-occipital infarction	L	2:6	12:0	30/day	Atonic, GTCS, Tonic	Motor, Non-motor	2:6	12:1	
2	M	EMAtS	Unknown	-	3:0	3:1	100/day	Atonic, Myoclonic, Tonic	-	3:0	3:10	
3	M	Structural/lesional	RS	L	2:7	3:6	10/day	Spasm	Motor	3:0	3:6	
4	M	LKS	GRIN2A mutation	-	-	-	-	-	-	3:6	10:6	
5	M	LKS	Unknown	-	3:7	4:0	8/day	Tonic	Motor	3:9	4:0	
6	F	LKS	GRIN2A mutation	-	5:2	5:2	1 isolated seizure	-	Motor	4:7	5:6	
7	F	LKS	Unknown	-	3:1	5:1	1/month	GTCS, Myoclonic	-	4:8	6:2	
8	M	EMAtS	Unknown[a]	-	4:0	5:9	16/day	Atonic, Myoclonic	Motor	5:4	6:4	
9	F	LKS	Unknown	-	5:2	5:4	30/day	Absence, Atonic, GTCS, Gelastic, Spasm	Motor	5:4	6:3	
10	M	LKS	Unknown	-	5:7	5:7	<1/month	-	Motor	5:7	6:3	

11	F	Structural/lesional	RS	R	3:6	5:6	5/day	-	Motor	5:7	5:9	
12	F	Uncharacterised generalised epilepsy	Unknown	-	5:3	6:4	100/day	Absence, Atonic, GTCS, Tonic	-	5:10	6:6	
13	M	Structural/lesional	RS	R	8:6	11:6	4/week	-	Motor, Non-motor	7:0	11:7	
14	F	Structural/lesional	RS	L	5:0	7:1	1/week	-	Motor	7:1	14:6	
15	F	Lesion-negative focal epilepsy	Unknown	R[b]	3:8	10:3	5/day	-	Motor	7:10	11:1	
16	F	Structural/lesional	RS	L	9:4	10:7	50/day	-	Motor, Non-motor	9:4	10:7	
17	F	LKS	GRIN2A mutation	-	4:0	7:4	Unknown	GTCS	-	9:8	9:9	
18	F	Structural/lesional	RS	L	11:3	12:9	6/day	-	Motor, Non-motor	11:10	12:9	



Cases are ordered by ascending age at motor speech regression. In the chronology plots red diamonds show age at speech regression, Ax shows age at first assessment in the Developmental Epilepsy Clinic (DEC), white circles show age at seizure nadir, and horizontal bars show the time period from seizure onset to first assessment. Abbreviations: EMAtS, epilepsy with myoclonic atonic seizures; GTCS, generalised tonic-clonic seizure; LKS, Landau-Kleffer syndrome; RS, Rasmussen syndrome.

[a]Variant of uncertain significance in SLC6A1 gene.

[b]Semiology and ictal EEG findings demonstrating weak lateralising features to the right hemisphere.

Table 2: Speech features and psychometric assessments

				Speech features at initial assessment						Standardised scores at initial assessment			Motor speech outcome at final follow-up				Standardised scores at final follow-up		
Case no.	Diagnostic group (affected hemisphere)	Pre-existing speech/ language concerns	Motor speech presentation	Dysarthria	Dyspraxia	Phonological error	Dysphonia	Stammer	Impaired intelligibility	Expressive language	Receptive language	Nonverbal cognition	Follow-up interval (years:months)	Persisting difficulties	Persisting difficulties without improvement	Impaired intelligibility	Expressive language	Receptive language	Nonverbal cognition
4	LKS	•	Sudden and complete loss of speech		•	•			•	59[b]	34[h]	76[i]							
5	LKS		Slurred speech, postictal then gradually progressive	•		•	•		•	35[c]	98[f]	99[j]	8:9	•			110[d]	107[d]	106[k]
6	LKS	•	Gradual onset of drooling and loss of speech	•					•	65[c]	45[c]	50[j]	5:0	•	•	•	71[d]	65[d]	62.5[k]
7	LKS	•	Gradual slowing of speech	•					•	52[d]	85[d]	111[k]	2:8	•			122[d]	127[d]	117[k]
9	LKS		Gradual loss of prosody then complete loss of speech		•				•	39[b]	63[f]	77.5[j]							
10	LKS	•	Sudden loss of speech at epilepsy onset		•	•			•	50[e]	50[e]	111[i]	4:2	•	•	•	55[d]	70[d]	103[k]
17	LKS	•	Gradual loss of motor-speech skills		•			•		61[d]	54[d]	57[k]	6:0				99[d]	103[d]	57[k]
2	Other generalised epilepsy	•	Sudden loss of speech at epilepsy onset			•			•	91[f]	77[f]	72[j]	1:0	•		•	102[f]	100[f]	97[j]
8	Other generalised epilepsy		Slurred speech, postictal then gradually progressive	•		•			•	100[g]	93[f]	97[j]	1:8	•			94[d]	67[d]	86[k]

12	Other generalised epilepsy	•	Gradual slowing of speech and loss of articulation	•		•			•	50[b]	82[h]	60[j]	5:10	•		•	50[d]	48[d]	53[k]
3	RS (L)	•	Gradual slowing of speech and loss of articulation	•	•	•	•		•	65[e]	64[e]	69[j]	1:7[m]	•		•	50[e]	50[e]	60[j]
11	RS (R)		Gradual loss of speech clarity	•					•	91[f]	77[f]	77.5[j]	3:6[m]	•			69[d]	59[d]	57[k]
13	RS (R)	•	Progressive slurring and loss of speech clarity	•					•	69[d]	76[d]	100[k]	1:10[m]	•	•		57[d]	62[d]	55[k]
14	RS (L)	•	Gradual deterioration in speech clarity	•					•	63[d]	60[d]	63[k]	3:0	•	•	•	50[d]	57[d]	59.5[n]
16	RS (L)		Gradual slowing and slurring of speech	•					•	98[d]	90[d]	92[k]	3:1	•	•		73[d]	88[d]	75[k]
18	RS (L)		Gradual deterioration in speech	•	•			•	•	65[b]	81[h]	54.5[k]	2:10[m]	•		•	55[b]	80[h]	68[k]
1	Other focal epilepsy (L)		Sudden deterioration in speech after prolonged seizure	•				•		44[c]	69[c]	43[l]							
15	Other focal epilepsy (R[a])		Gradual increased drooling and speech deterioration	•						71[d]	73[d]	76[k]							

Cases are ordered by diagnostic group. Abbreviations: LKS, Landau-Kleffer syndrome; RS, Rasmussen syndrome.
[a]Semiology and ictal EEG findings demonstrating weak lateralising features to the right hemisphere
[b]Expressive Vocabulary Test (EVT)
[c]Vineland Adaptive Behaviour Scales (VABS)
[d]Clinical Evaluation of Language Fundamentals (CELF)
[e]Preschool Language Scale (PLS)
[f]CELF Preschool (PS-CELF)
[g]Expressive Vocabulary Subtest of PS-CELF
[h]Peabody Picture Vocabulary Test (PPVT)
[i]Wechsler Nonverbal Scale of Ability (WNV)
[j]Wechsler Preschool & Primary Scale of Intelligence (WPPSI)
[k]Wechsler Intelligence Scale for Children (WISC)
[l]WPPSI developmental quotient
[m]Epilepsy surgery (hemispherotomy) during follow-up interval
[n]Wechsler Adult Intelligence Scale (WAIS)

Table 3: Speech sample audio-perceptual analysis and severity rating

		Mayo Dysarthria Rating Scale: domain (total no. features)							Motor Speech Disorders Severity Severity Rating Scale	
Case no.	**Diagnostic group (affected hemisphere)**	**Pitch (6)**	**Loudness (5)**	**Voice quality (7)**	**Resonance (5)**	**Respiration (4)**	**Prosody (10)**	**Articulation (5)**	**Score**	**Descriptor**
4	LKS	•	•	•	•		••	•••	4	Speech plus AAC
5	LKS	O	O	•••	O	O	O	O	2	Vocalises for emotion
6	LKS		•	•	•		••	••	7	Obvious speech abnormalities
7	LKS	•	•	•	•	•	•••••••	•••	7	Obvious speech abnormalities
10	LKS	•	•	••	••		•••	••	5	Frequent repetition needed
17	LKS	•	•				••	•	8	Perceived speech changes
8	Other generalised epilepsy	•	••	•••	•••		•••••	••••	6	Repeats messages on occasion
12	Other generalised epilepsy		•	•	•••		••••••••	••••	7	Obvious speech abnormalities
3	RS (L)	O	O	O	O	•	O	O	2	Vocalises for emotion
16	RS (L)	•	•	•	•		••••••	•	7	Obvious speech abnormalities
18	RS (L)	O	•	••	O	O	••••	O	5	Frequent repetition needed
15	Other focal epilepsy (L)		•	•			••	•	9	Nominal speech abnormality

Cases are ordered by diagnostic group. • indicates a feature present within a domain (multiples = multiple features); O indicates inability to rate domain due to insufficient speech produced. The Motor Speech Disorders Severity Rating Scale score ranges from 1 (nonvocal) to 10 (normal speech). Abbreviations: AAC, augmentative and alternative communication; LKS, Landau-Kleffer syndrome; RS, Rasmussen syndrome.

Figure legends

Figure 1: Motor speech disorders at first assessment and follow-up

Data are shown for the whole cohort in the top panel and organised by diagnostic group in the bottom panels. Stacked bars show the proportions of children with no motor speech disorder (green), motor speech disorder with preserved intelligibility of speech (orange) or motor speech disorder with impaired intelligibility of speech (red) at first assessment and at follow-up (FU). Grey bars indicate the proportion of children with some degree of motor speech improvement from first assessment to follow-up.

Figure 2: Language and nonverbal cognitive scores at first assessment and follow-up

The top row shows the mean standardised score for each diagnostic group at first assessment and follow-up. The bottom row shows the individual scores for each child (including only those assessed at both timepoints). Change in score from first assessment to follow-up was tested within each diagnostic group using paired t-tests; results significant at $p<0.05$ are displayed. FU, follow-up; LKS, Landau-Kleffner syndrome; RS, Rasmussen syndrome.

Figure 3: Interictal epileptiform discharges and suspected seizure origin locations

Contour maps generated from EEG electrode locations of interictal epileptiform discharge (IED) maxima (A) and electrodes implicated in seizure origin (B) in 12 children.