The child with acquired disorders of speech and language

EPNS Training Course

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Université Libre de Bruxelles (ULB)
Vrije Universiteit Brussel (VUB)
Universiteit Antwerpen (UA)

OVERVIEW OF ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- Dysarthria
- Apraxia of speech
- Aphemia
- Mutism
- Neurogenic stuttering
- Language of confusion
- Aphasia (and related language syndromes)

ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Dysarthria***

  A motor speech disorder caused by dysfunction of the muscles involved in speech production. Diagnosing dysarthria implies the presence of a neurologically based alteration of the nervous system that controls the functioning of the phono-articulatory apparatus: paresis, neuromuscular weakness, abnormal muscle tone, incoordination. Several subsystems are implicated in speaking: respiration, voice, resonance, articulation, speech rate, prosody.

SCHEMATIC REPRESENTATION OF THE COGNITIVE PROCESSES UNDERLYING NAMING

- Early visual processing ⇒ recognition of the picture;
- Accessing the semantic representation (or meaning);
- Accessing the phonological (or auditory) form of the word;
- Programming the articulatory movements (lips, tongue, jaw, palate, etc) and their sequence;
- Articulating the word by implementing the planned movements.

↓

Structural Representation

↓

Semantics

↓

Auditory Word Form

↓

Motor Planning

↓

Articulation

↓

« horse »

Hillis AE, Course N° 2DS.008, American Academy of Neurology, 53rd Annual Meeting, 2001.

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~~**Articulation**~~ ⇒ **Dysarthria**

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« horse »

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ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Apraxia of speech (also called "Anarthria" in French literature)***

  An articulatory disorder resulting from impairment, due to brain damage, of the capacity to plan and program the positioning of speech musculature for the volitional production of phonemes and the sequencing of muscle movements for the production of words, in the absence of significant damage to the motor or sensory pathways directly controlling the articulatory musculature.

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ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

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~~**Motor Planning**~~ ⇒ **Apraxia of speech**

↓

Articulation

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« horse »

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ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Aphemia***
  - Complete, neurologically based articulatory failure in the absence of damage to the neuromuscular system controlling the phono-articulatory apparatus.
  - Typically, auditory comprehension, writing, and reading are preserved, as well as voicing.
  - The responsible lesion is located in the language dominant insular and opercular region.

  Aphemia is to be considered the most severe form of apraxia of speech.

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ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Mutism***

  Failure to utter verbal and/or non-verbal vocal sounds following damage to the cerebrospinal axis. Auditory comprehension, writing, and reading are preserved.

  Some authors distinguish:
  - *Verbal mutism*: non-verbal vocal sound production still possible (e.g., grunting, yawning --cf. *aphemia*).
  - *Complete mutism*: complete absence of verbal and non-verbal sound production.

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ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Mutism (neurosurgical definitions)***

The total absence of speech and sound in an awake and conscious patient. [p. 894]

Van Calenbergh F et al. Transient cerebellar mutism after posterior fossa surgery in children. Neurosurgery 1995: 37: 894-898.

Mutism is a condition of complete absence of speech that is not associated with other aphasic symptomatology or alteration of consciousness. [p. 115]

Crutchfield JS et al. Postoperative mutism in neurosurgery: report of two cases. J Neurosurg 1994: 81: 115-121.

Mutism is represented by a lack of voice or speech output in the awake patient, with the comprehension of speech and language remaining intact. [p. 472]

Dailey AT et al. The pathophysiology of oral pharyngeal apraxia and mutism following posterior fossa tumor resection in children. J Neurosurg 1995: 83: 467-475.

ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Mutism***

Possible causes of organic mutism :

- Damage to Broca's area (transient mutism)
- Damage to SMA of dominant hemisphere
- Damage to reticular formation of mesencephalon (akinetic mutism)
- Bilateral hemispheric damage producing pseudobulbar palsy
- Peripheral lesions leading to bilateral pharyngeal or vocal cord paralysis

Benson DF. Aphasia, alexia, agraphia. New York: Churchill Livingstone, 1979.

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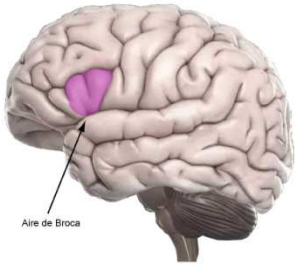

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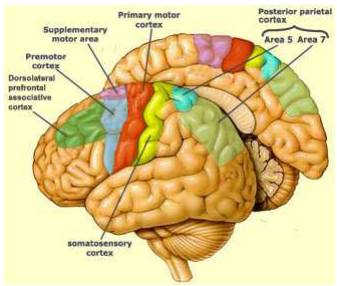

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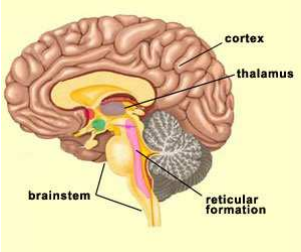

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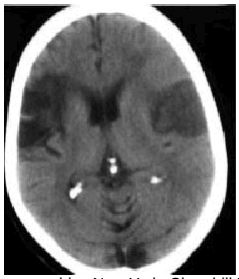

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ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

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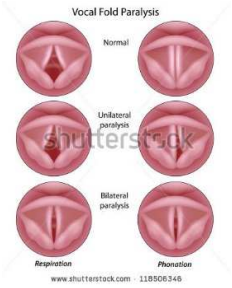
  Possible causes of organic mutism :

  - Peripheral lesions leading to bilateral pharyngeal or vocal cord paralysis



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ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Mutism***

  Possible surgical causes of neurologic mutism :

  - Thalamotomy for parkinsonism
  - Callosotomy
  - Surgery to SMA of dominant hemisphere
  - Posterior fossa surgery

Aguiar PH et al. Transient mutism following a posterior fossa approach to cerebellar tumors in children: a critical review of the literature. Child's Nerv Syst 1995: 11: 306-310.

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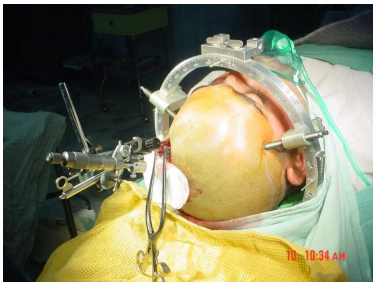

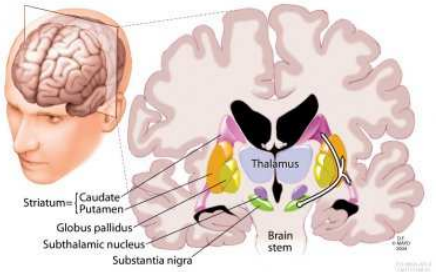

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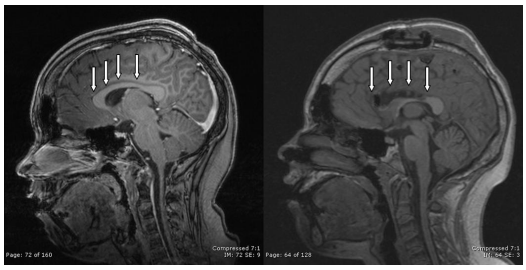
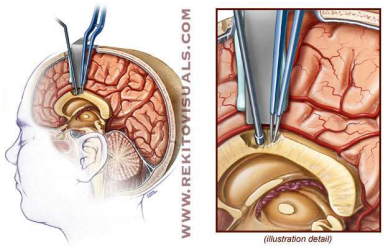
ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Mutism***

  Possible surgical causes of neurologic mutism :

  - Callosotomy





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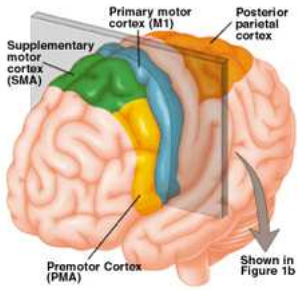

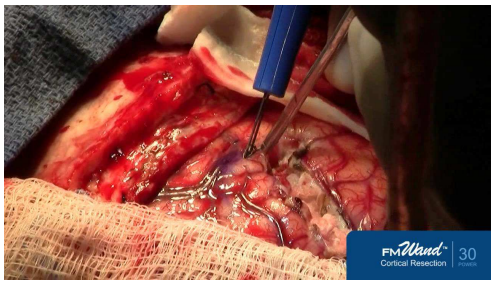

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ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Mutism***

  Possible surgical causes of neurologic mutism :

  - Posterior fossa surgery

  "A transient mutism occurring after a posterior fossa tumor approach in children with unimpaired consciousness, unimpaired symbolic functions, no detectable deficit of cranial nerves or peripheral organs of speech, and no lesions of long pathways in the course of the cranial nerves at the level of the brain stem." [p. 306]

Aguiar PH et al. Transient mutism following a posterior fossa approach to cerebellar tumors in children: a critical review of the literature. Child's Nerv Syst 1995: 11: 306-310.

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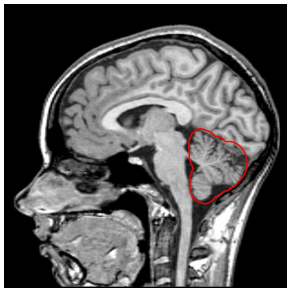

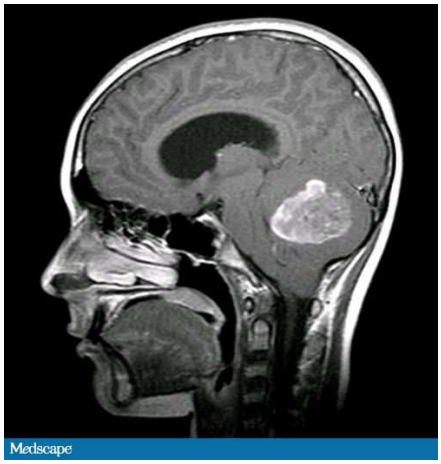

ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Neurogenic stuttering***

  "Deviation of speech attracting attention of speakers or listeners because of interruption of the normal rhythm of speech by involuntary interruption, prolongation or arrest of sounds. Acquired stuttering, mainly resulting from cerebral infarcts, has been described in association with unilateral left or right, or bilateral, cortical or subcortical lesions."

Abe K et al. Repetitive speech disorder resulting from infarcts in the paramedian thalami and midbrain. J Neurol Neurosurg Psychiatry 1993: 56: 1024-1026

ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Language of confusion***

  An inadequate language behavior resulting from brain damage, and characterized by a semantic incoherence the confused patient is usually unaware of (*anosognosia*).

  The inappropriateness and bizarreness of the content reflect the disorder of reasoning and the reduced clarity of thinking, which, in turn, are due to a profound alteration of attentional mechanisms.

ACQUIRED NEUROLOGICAL SPEECH/LANGUAGE DISORDERS IN CHILDREN

- ***Aphasia***

  A childhood language disorder due to a cerebral lesion that is sustained after onset of language acquisition, and that disrupts already developed language skills.

  In contrast with congenital aphasia and developmental dysphasia (SLI), in acquired childhood aphasia (ACA) the pathological process which compromises the functioning of language is sustained after a period of language development.

ACQUIRED APHASIA IN CHILDREN: TERMINOLOGICAL AMBIGUITY

- ***Acquired aphasia*** or ***acquired lesion*** ?

  "This chapter addresses the language abilities only in children *with known brain insults*, that is, those *with acquired aphasia*."

  "Therefore, *acquired aphasia*, as used here, refers to language discrepancies or abnormalities accompanied by a known brain lesion, *irrespective of when during the course of language development that lesion occurred*."

  Aram DM. Acquired aphasia in children. In: MT Sarno (ed.), Acquired Aphasia. Orlando: Academic Press, 1991; 425-453.

  [Cf. definition of congenital aphasia !]

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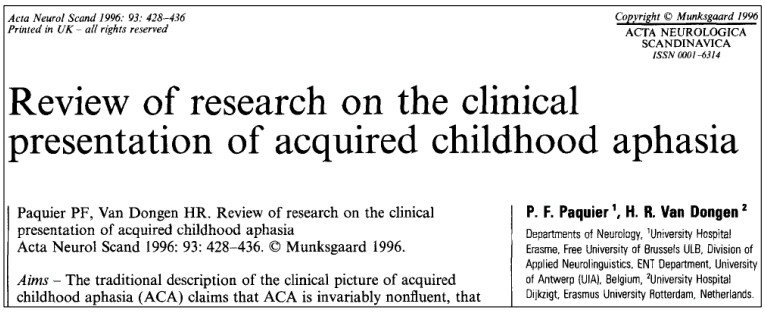
ACQUIRED APHASIA IN CHILDREN: TERMINOLOGICAL DELINEATION



Acta Neurol Scand 1996: 93: 428–436
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ACTA NEUROLOGICA SCANDINAVICA
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Review of research on the clinical presentation of acquired childhood aphasia

Paquier PF, Van Dongen HR. Review of research on the clinical presentation of acquired childhood aphasia
Acta Neurol Scand 1996: 93: 428–436. © Munksgaard 1996.

Aims – The traditional description of the clinical picture of acquired childhood aphasia (ACA) claims that ACA is invariably nonfluent, that

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Departments of Neurology, [1]University Hospital Erasme, Free University of Brussels ULB, Division of Applied Neurolinguistics, ENT Department, University of Antwerp (UIA), Belgium, [2]University Hospital Dijkzigt, Erasmus University Rotterdam, Netherlands.

Acquired childhood aphasia is the equivalent of *aphasia in children caused by a non-congenital brain lesion*.

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ACQUIRED APHASIA IN CHILDREN: CLINICAL PRESENTATION

- Traditionally considered a nonfluent aphasia irrespective of lesion location:

  "Nonfluent verbal output characterizes acquired aphasia in childhood regardless of the site of brain insult."

  Benson DF, Ardila A. Aphasia: A Clinical Perspective. New York: Oxford University Press, 1996.

- Currently acknowledged to be as heterogeneous as aphasia in adults:

  "All types of aphasia may affect children, showing an adult-like heterogeneity of aphasic syndromes."

  Chilosi AM et al. Brain Lang 2008: 106: 211-225.

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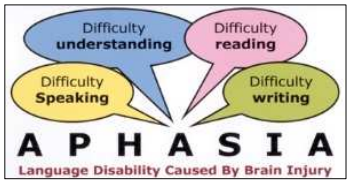
ACQUIRED APHASIA IN CHILDREN: CLINICAL PRESENTATION

Nonfluent Aphasia
Broca aphasia
Global aphasia
Transcortical motor aphasia
Mixed transcortical aphasia

Fluent Aphasia
Wernicke aphasia
Conduction aphasia
Transcortical sensory aphasia
Anomic aphasia

Related syndromes
Subcortical aphasia
Alexia with agraphia
Alexia without agraphia
Auditory agnosia



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ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

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ACQUIRED APHASIA IN CHILDREN: INCIDENCE

- Traditionally, acquired aphasia in children was considered a rare phenomenon (greater neuroplasticity of the young brain).
- Satz & Bullard-Bates' (1981) conclusion regarding the rareness of acquired childhood aphasia:
  - ACA is not rare if the lesion is unilateral and encroaches upon the "speech areas". However, there is a lower frequency of unilateral vascular disease in children as compared to adults.

Satz P, Bullard-Bates C. Acquired aphasia in children. In: MT Sarno, ed. Acquired Aphasia. San Diego: Academic Press, 1981; 399-426.

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ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

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ACQUIRED APHASIA IN CHILDREN: INCIDENCE

- Incidence of stroke and of aphasia in children:
  - Ischemic strokes : 7,91/100.000/yr
    (adults: 150/100.000/yr)
  - Hemorrhagic strokes : 5,11/100.000/yr
    (adults: 13,4/100.000/yr)
  - All strokes : 13,02/100.000/yr
    (adults: 130/100.000/yr)

Giroud M et al. J Clin Epidemiol 1995: 48: 1343-1348.

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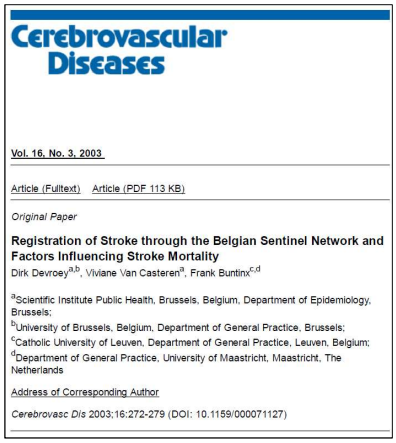
ACQUIRED APHASIA IN CHILDREN: INCIDENCE

- Stroke incidence in Belgium (1998-1999): 185/100.000/year
  ⇒ 18.500 pts/year
- ± 30% aphasia
  ⇒ 5.550 aphasic pts/year



Cerebrovascular Diseases

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Article (Fulltext) Article (PDF 113 KB)

Original Paper

Registration of Stroke through the Belgian Sentinel Network and Factors Influencing Stroke Mortality

Dirk Devroey[a,b], Viviane Van Casteren[a], Frank Buntinx[c,d]

[a]Scientific Institute Public Health, Brussels, Belgium, Department of Epidemiology, Brussels; [b]University of Brussels, Belgium, Department of General Practice, Brussels; [c]Catholic University of Leuven, Department of General Practice, Leuven, Belgium; [d]Department of General Practice, University of Maastricht, Maastricht, The Netherlands

Address of Corresponding Author

Cerebrovasc Dis 2003;16:272-279 (DOI: 10.1159/000071127)

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  - If the left hemisphere has been damaged, the risk of acquired aphasia is approximately the same in right-handed children as it is in right-handed adults.

Satz P, Bullard-Bates C. Acquired aphasia in children. In: MT Sarno, ed. Acquired Aphasia. San Diego: Academic Press, 1981; 399-426.

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ACQUIRED APHASIA IN CHILDREN: FREQUENCY

- Frequency of stroke and of aphasia in children:
  - 54 children with arterial stroke (30M, 24F, mean age 10,25 yrs):
    - 31 cases of ischemic stroke (57%)
    - 23 cases of hemorrhagic stroke (43%)
  - Aphasia present in:
    - 14/31 (45%) of ischemic cases
    - 9/23 (39%) of hemorrhagic cases
  - Total: 23/54 (43%) children aphasic

Giroud M et al. J Clin Epidemiol 1995: 48: 1343-1348.

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ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

ACQUIRED APHASIA IN CHILDREN: FREQUENCY

- Frequency of aphasia in adults:

  European population-based studies report the frequency of aphasia among stroke patients to be between 23 and 38 %.

Stegmayr B et al. Stroke 1994: 25: 1738-1745.
Pedersen PM et al. Ann Neurol 1995: 38: 659-666.

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ACQUIRED APHASIA IN CHILDREN: OUTCOME

- Traditionally considered to have a favorable outcome with rapid and complete recovery.
- If the aphasiogenic lesion occurs after the onset of language development and is confined to a single hemisphere, language will invariably return if the child is less than 9 years old at the time of lesion occurrence.
- “Apparently aphasia runs a different course before the end of the first decade than after it” [p. 146].
- Aphasia acquired at the time of puberty commonly leaves some traces behind.

Lenneberg E. Biological Foundations of Language. New York: John Wiley, 1967.

ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

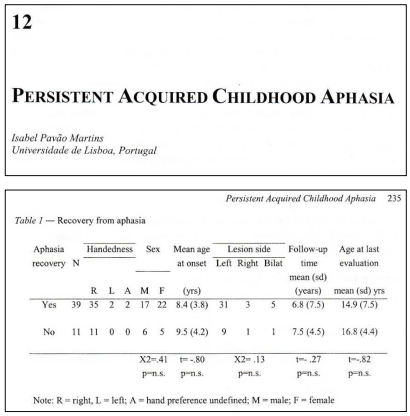
ACQUIRED APHASIA IN CHILDREN: OUTCOME

- Current insights acknowledge that recovery of acquired childhood aphasia depends upon a number of interrelated factors, among them etiology, site and size of the lesion, diaschisis, and concomitant neurological disturbances.
- Recovery from acquired childhood aphasia may be slow and incomplete, leading to disruption of scholastic achievement and persisting linguistic disabilities.

Paquier P & Van Dongen HR. Acta Neurol Scand 1996: 93: 428-436.

ACQUIRED APHASIA IN CHILDREN: OUTCOME

- 11/50 patients, mean age 17.5 yrs (sd 5.2) chronically aphasic for ≥ 2 yrs following a non-progressive brain lesion sustained in childhood.
- Factors associated with persistent aphasia:
  - traumatic or infectious etiology
  - poor verbal comprehension at onset
  - epilepsy
  - lesions involving classical L hemisphere language areas



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PERSISTENT ACQUIRED CHILDHOOD APHASIA

Isabel Pavão Martins
Universidade de Lisboa, Portugal

Persistent Acquired Childhood Aphasia 235

Table 1 — Recovery from aphasia

Aphasia recovery	N	Handedness R	L	A	Sex M	F	Mean age at onset (yrs)	Lesion side Left	Right	Bilat	Follow-up time mean (sd) (years)	Age at last evaluation mean (sd) yrs
Yes	39	35	2	2	17	22	8.4 (3.8)	31	3	5	6.8 (7.5)	14.9 (7.5)
No	11	11	0	0	6	5	9.5 (4.2)	9	1	1	7.5 (4.5)	16.8 (4.4)
					X2=.41 p=n.s.		t= -.80 p=n.s.		X2= .13 p=n.s.		t=- .27 p=n.s.	t=-.82 p=n.s.

Note: R = right, L = left; A = hand preference undefined; M = male; F = female

Fabbro F (ed). Neurogenic Language Disorders in Children. Amsterdam: Elsevier, 2004; 231-251.

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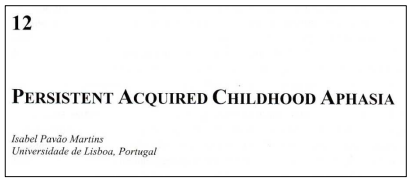
ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

ACQUIRED APHASIA IN CHILDREN: OUTCOME

- 3/11 children never returned to school.
- 5/8 attended special education classes.
- 7/8 repeated grades.
- At adulthood:
  - lower socio-economic status as compared to their parents;
  - female patients economically dependent on husband or parents;
  - male patients living with their parents and economically dependent;
  - no new and stable emotional relationships.



12

PERSISTENT ACQUIRED CHILDHOOD APHASIA

Isabel Pavão Martins
Universidade de Lisboa, Portugal

Fabbro F (ed). Neurogenic Language Disorders in Children. Amsterdam: Elsevier, 2004; 231-251.

Language recovery is less complete than that reported after congenital lesions … persistent aphasia has a negative impact on the patients' educational, social, and professional life.

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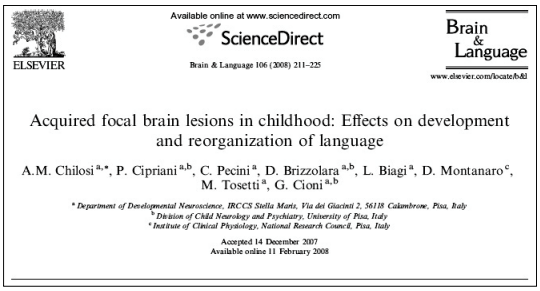
ACQUIRED APHASIA IN CHILDREN: OUTCOME

- Outcome of ACA:



Available online at www.sciencedirect.com
ScienceDirect
Brain & Language 106 (2008) 211–225
Brain & Language
www.elsevier.com/locate/b&l
ELSEVIER

Acquired focal brain lesions in childhood: Effects on development and reorganization of language

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Illusory recovery is the presence of subtle language sequelae and frequent learning disabilities, in spite of apparent good clinical recovery, many years after onset of aphasia.

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ACQUIRED APHASIA IN CHILDREN: OUTCOME

Illusory recovery results from a combination of factors:

- Aphasic children need to recover the components of language acquired before onset of aphasia.
- Aphasic children must acquire new language skills and processes.
- However, the cognitive prerequisite for acquisition of new (written) language skills might be impaired because of disrupted cortico-subcortical pathways –even deep lesions can interfere with cognition.

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ACQUIRED APHASIA IN CHILDREN: OUTCOME

Impact of brain lesions on cognition:

- Early brain damage may result in a cumulative effect on ongoing development.
- Increasing deficits may emerge through childhood as more functions are expected to mature and will need to be subsumed within undamaged tissues.
- New long-term problems may result from changes during further development which lead to disruption of pathways that fail to develop properly as a consequence of the lesion.

⇒ phenomenon of ***growing into deficits***

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ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

ACQUIRED APHASIA IN CHILDREN: OUTCOME

Because of the phenomenon of *growing into deficits*, children with initially good clinical recovery may develop cognitive, social, and behavioral deficits years after onset of aphasia.

⇒ *illusory recovery* is linked to *growing into deficits*

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ACQUIRED APHASIA IN CHILDREN: CONCLUSION

- When L hemisphere lesions are sustained after onset of language acquisition, childhood aphasia frequently occurs.
- Substantial language recovery after acquired brain injury has been documented in children, supporting the greater potential for recovery in the immature and less functionally organized brain.
- However, the statement “the earlier the better” is not always confirmed –cf. Landau-Kleffner Syndrome and duration of CSWS.
- Age at onset of aphasia has not always emerged as a predictor of linguistic recovery (as would be expected from data on children with congenital lesions).

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ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

ACQUIRED APHASIA IN CHILDREN: CONCLUSION

- When left hemisphere lesions are sustained after onset of language acquisition, childhood aphasia frequently occurs.
- Substantial language recovery after acquired brain injury has been documented in children, supporting the greater potential for recovery in the immature and less functionally organized brain.
- However, the young child’s brain, though plastic, does lack the consolidated representations of complex language knowledge, which is typical of adults.
- Age at onset of aphasia has not always emerged as a predictor of linguistic recovery (as would be expected from data on children with congenital lesions).

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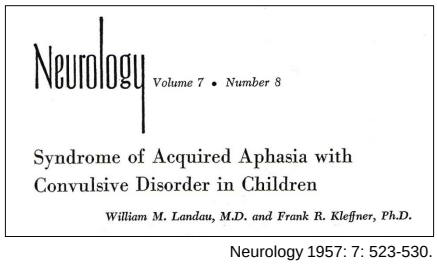
ACQUIRED APHASIA IN CHILDREN: CONCLUSION

- Incomplete –illusory– recovery from aphasia despite early acquired lesions urges us to reconsider the critical and favorable role of age at onset of aphasia.
- The statement “the earlier the better” is not always confirmed –cf. Landau-Kleffner Syndrome and duration of CSWS.
- Recovery of language disorders depends upon a number of interrelated variables such as etiology, lesion site and size, aphasia type, and concomitant neurological disturbances. Recovery from ACA may be slow and incomplete, leading to disruption of scholastic achievement and persisting linguistic disabilities.

ACQUIRED APHASIA IN CHILDREN: LANDAU-KLEFFNER SYNDROME

- 6 children with normal intelligence
- Language regression
- Cause ??
- Children seemed to be deaf, yet normal hearing and premorbid language development
- Waking EEG: epileptic anomalies in all children
- Diagnosis: acquired aphasia with epilepsy


Neurology Volume 7 • Number 8

Syndrome of Acquired Aphasia with Convulsive Disorder in Children

William M. Landau, M.D. and Frank R. Kleffner, Ph.D.

Neurology 1957: 7: 523-530.

“Persistent convulsive discharge in brain tissue largely concerned with linguistic communication results in the functional ablation of these areas for normal linguistic behavior”. [p. 529]

ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

Hôpital Erasme | ULB

ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

Hôpital Erasme | ULB

ACQUIRED APHASIA IN CHILDREN: LANDAU-KLEFFNER SYNDROME

- Occurs in children aged 1-8, but mostly between 3 and 8 years of age.
- Boys > girls.
- Initially children develop language in a (near-) normal way.
- Epileptic insults are often not the first manifestation.
- At onset, children seem to become progressively deaf: they understand language less and less, and no longer respond when spoken to –yet normal audiogram and BAEP !
- If normal hearing: psychogenic deafness? Hypothesis nullified after referral to the child psychiatrist.
- Loss of recognition and meaning of non-verbal sounds (e.g., phone, doorbell, klaxon, animal sounds, etc.) ⇒ auditory agnosia.
- Progressive regression of expressive language: word-finding difficulties, jargon or telegraphic speech, mutism, gestures…

⇒ Aphasia !

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ACQUIRED APHASIA IN CHILDREN: LANDAU-KLEFFNER SYNDROME

- Aphasia develops over a period ranging from a few days to a few months.
- Fluctuating course of the aphasia, characterized by remissions and exarcerbations.
- Versatile and unpredictable mode of presentation :

  "Le propre du syndrome de Landau et Kleffner est d'échapper à toute systématisation" (Dugas M et al. Rev Neurol 1982: 138: 755-780).

- Normal clinical neurological exam except for the aphasia.
- Clinical insults not always present.
- CT-scan, MRI: without particularities.
- Routine EEG: often bilateral temporal spike-wave activity that increases during sleep –when ≥ 85% of non-REM sleep:

Continuous Spikes and Waves during slow Sleep (CSWS)
Electrical Status Epilepticus during slow Sleep (ESES)

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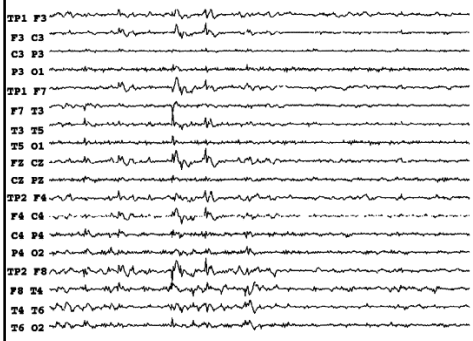
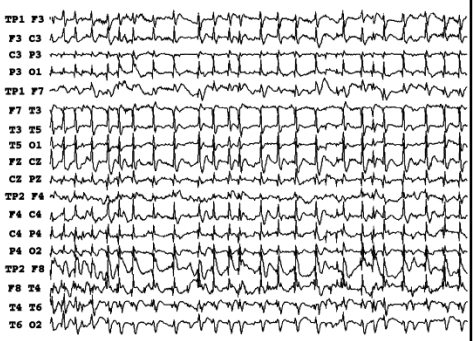
ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

ACQUIRED APHASIA IN CHILDREN: LANDAU-KLEFFNER SYNDROME

Continuous Spikes and Waves during slow Sleep (CSWS)



Waking EEG showing paroxysmal spike-wave discharges occurring against normal background activity.



Sleep EEG showing continuous spike-wave discharges during slow sleep (CSWS).

Metz-Lutz MN et al. J Neurolinguistics 1999: 12: 167-179.

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ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

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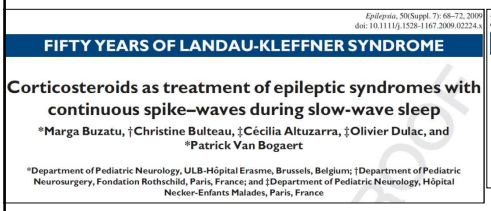
ACQUIRED APHASIA IN CHILDREN: LANDAU-KLEFFNER SYNDROME

- Classified among the epileptic encephalopathies of unknown etiology.
- Considered part of the spectrum of idiopathic focal epilepsies with CSWS.
- No constant relationship between severity and duration of the aphasia, and the paroxysmal abnormalities on a waking EEG → sleep recording !
- Epileptic insults well-controlled with AEDs, but limited effect on the aphasia.
- CSWS and epilepsy disappear by adolescence, yet clinical recovery from aphasia in only ≈ 30% children → unfavorable long-term outcome.
- Therapeutic strategies: (a) corticosteroids, (b) immunoglobulins.

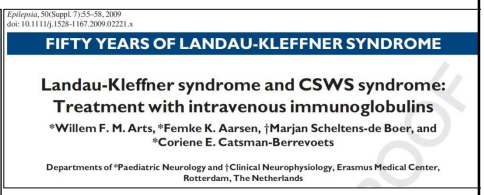

Epilepsia, 50(Suppl. 7): 68–72, 2009
doi: 10.1111/j.1528-1167.2009.02224.x

FIFTY YEARS OF LANDAU-KLEFFNER SYNDROME

Corticosteroids as treatment of epileptic syndromes with continuous spike–waves during slow-wave sleep

*Marga Buzatu, †Christine Bulteau, ‡Cécilia Altuzarra, ‡Olivier Dulac, and *Patrick Van Bogaert

*Department of Pediatric Neurology, ULB-Hôpital Erasme, Brussels, Belgium; †Department of Pediatric Neurosurgery, Fondation Rothschild, Paris, France; and ‡Department of Pediatric Neurology, Hôpital Necker-Enfants Malades, Paris, France


Epilepsia, 50(Suppl. 7):55–58, 2009
doi: 10.1111/j.1528-1167.2009.02221.x

FIFTY YEARS OF LANDAU-KLEFFNER SYNDROME

Landau-Kleffner syndrome and CSWS syndrome: Treatment with intravenous immunoglobulins

*Willem F. M. Arts, *Femke K. Aarsen, †Marjan Scheltens-de Boer, and *Coriene E. Catsman-Berrevoets

Departments of *Paediatric Neurology and †Clinical Neurophysiology, Erasmus Medical Center, Rotterdam, The Netherlands

ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

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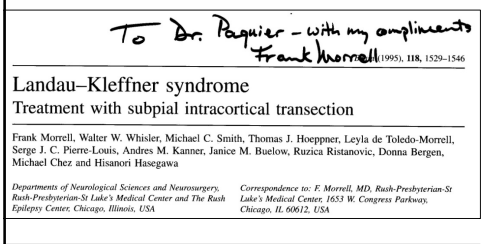
ACQUIRED APHASIA IN CHILDREN: LANDAU-KLEFFNER SYNDROME

- Therapeutic strategies: (c) surgery, (d) ketogenic diet, (e) vagus nerve stimulation.


To Dr. Paquier – with my compliments
Frank Morrell

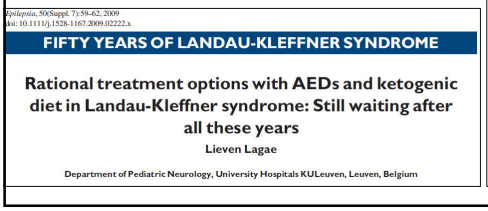
(1995), 118, 1529–1546

Landau–Kleffner syndrome
Treatment with subpial intracortical transection

Frank Morrell, Walter W. Whisler, Michael C. Smith, Thomas J. Hoeppner, Leyla de Toledo-Morrell, Serge J. C. Pierre-Louis, Andres M. Kanner, Janice M. Buelow, Ruzica Ristanovic, Donna Bergen, Michael Chez and Hisanori Hasegawa

Departments of Neurological Sciences and Neurosurgery, Rush-Presbyterian-St Luke's Medical Center and The Rush Epilepsy Center, Chicago, Illinois, USA

Correspondence to: F. Morrell, MD, Rush-Presbyterian-St Luke's Medical Center, 1653 W. Congress Parkway, Chicago, IL 60612, USA

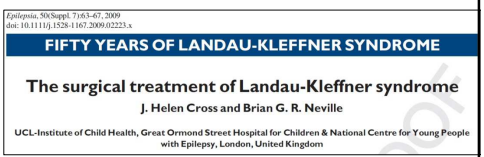

Epilepsia, 50(Suppl. 7):59–62, 2009
doi: 10.1111/j.1528-1167.2009.02222.x

FIFTY YEARS OF LANDAU-KLEFFNER SYNDROME

Rational treatment options with AEDs and ketogenic diet in Landau-Kleffner syndrome: Still waiting after all these years

Lieven Lagae

Department of Pediatric Neurology, University Hospitals KULeuven, Leuven, Belgium

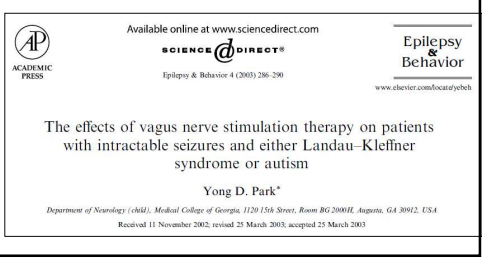

Epilepsia, 50(Suppl. 7):63–67, 2009
doi: 10.1111/j.1528-1167.2009.02223.x

FIFTY YEARS OF LANDAU-KLEFFNER SYNDROME

The surgical treatment of Landau-Kleffner syndrome

J. Helen Cross and Brian G. R. Neville

UCL-Institute of Child Health, Great Ormond Street Hospital for Children & National Centre for Young People with Epilepsy, London, United Kingdom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Epilepsy & Behavior 4 (2003) 286–290

Epilepsy & Behavior
www.elsevier.com/locate/yebeh

The effects of vagus nerve stimulation therapy on patients with intractable seizures and either Landau–Kleffner syndrome or autism

Yong D. Park*

Department of Neurology (child), Medical College of Georgia, 1120 15th Street, Room BG 2000JL, Augusta, GA 30912, USA

Received 11 November 2002; revised 25 March 2003; accepted 25 March 2003

Hôpital Erasme | ULB

ACQUIRED APHASIA IN CHILDREN: LANDAU-KLEFFNER SYNDROME

- Epileptic encephalopathy with CSWS: the abundant epileptic activity enhanced by sleep plays a major role in the cognitive and behavioral deficits presented by these children. This view is supported by:
  - A close temporal relationship between cognitive deterioration and enrichment and diffusion of the sleep discharges on EEG;
  - Clinical remission is usually associated with EEG improvement;
  - There is an association between the importance of cognitive sequelae and the long duration of CSWS ⇒ age at onset impacts on the long-term outcome.
- However, the presence of CSWS is not a prerequisite for a diagnosis of LKS !

DEVELOPMENTAL MEDICINE & CHILD NEUROLOGY CASE REPORT

Acquired auditory agnosia in childhood and normal sleep electroencephalography subsequently diagnosed as Landau–Kleffner syndrome: a report of three cases

PATRICK VAN BOGAERT[1] | MARY D KING[2] | PHILIPPE PAQUIER[3] | CATHERINE WETZBURGER[1] | CATHERINE LABASSE[4] | JEAN-MARIE DUBRU[5] | THIERRY DEONNA[6]

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DMCN 2013: 55: 575-579.

ACQUIRED PEDIATRIC SPEECH/LANGUAGE DISORDERS

Hôpital Erasme | ULB

ACQUIRED APHASIA IN CHILDREN: LANDAU-KLEFFNER SYNDROME

- 3 children with normal sleep EEG several months after onset of language regression.
- LKS diagnosed when a subsequent sleep EEG showed IEDs.
- A normal sleep EEG does not rule out LKS [*cave* day-to-day fluctuations in frequency of IEDs].

DMCN 2013: 55: 575-579.

Table I: Clinical and EEG data of patients

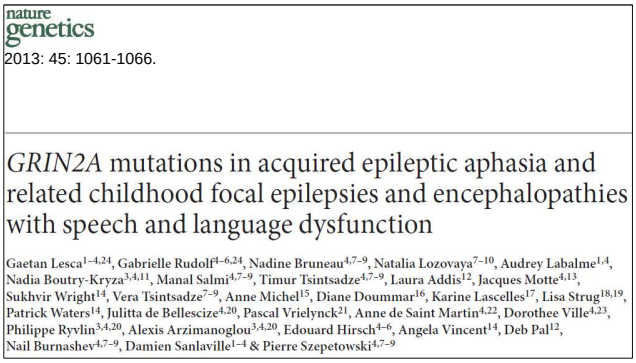
	Patient (sex)		
	1 (Male)	2 (Female)	3 (Female)
Age at onset of language regression	3y 6mo	2y 6mo	2y 3mo
Age at first sleep EEG (duration: results)	4y (overnight: normal)	3y 9mo (nap: normal)	4y 3mo (nap: normal)
Non-verbal IQ (Leiter International Performance Scale), age, result	4y 8mo, 93	3y 9mo, 110	5y 5mo, 111
Age at first sleep EEG showing IEDs (results)	4y 9mo (R and L rolandic IEDs awake, CSWS)	6y (R temporal IEDs awake, CSWS)	4y 5mo (L temporal and R occipital, no CSWS)
Drug trials	Hydrocortisone (21m cure)	Dexamethasone (1m cure), and various conventional AED	Corticosteroids (6wks cure) and various conventional AED
EEG evolution under treatment	Complete and sustained normalization after 1m treatment	No significant changes	Initial normalization, then reappearance of IEDs and CSWS pattern (5y 2mo)
Epileptic seizures	None	A few GTCS between ages 6 and 7y	Three focal seizures between ages 7 and 9y
Long-term outcome of auditory agnosia	Partial recovery	No recovery	No recovery

EEG, electroencephalography; IEDs, interictal epileptiform discharges; R, right; L, left; CSWS, continuous spike–waves during slow-wave sleep; AED, antiepileptic drug; GTCS, generalized tonic–clonic seizures.

Hôpital Erasme | ULB

ACQUIRED APHASIA IN CHILDREN: LANDAU-KLEFFNER SYNDROME

- LKS represents an epileptic encephalopathy of unknown etiology.
- However, about 20% of cases of LKS, syndrome of CSWS, and electroclinically atypical rolandic epilepsy often associated with speech/language impairment can have a genetic origin sustained by *de novo* or inherited mutations in the *GRIN2A* gene (Lesca et al., 2013).



nature genetics
2013: 45: 1061-1066.

GRIN2A mutations in acquired epileptic aphasia and related childhood focal epilepsies and encephalopathies with speech and language dysfunction

Gaetan Lesca[1–4,24], Gabrielle Rudolf[4–6,24], Nadine Bruneau[4,7–9], Natalia Lozovaya[7–10], Audrey Labalme[1,4], Nadia Boutry-Kryza[3,4,11], Manal Salmi[4,7–9], Timur Tsintsadze[4,7–9], Laura Addis[12], Jacques Motte[4,13], Sukhvir Wright[14], Vera Tsintsadze[7–9], Anne Michel[15], Diane Doummar[16], Karine Lascelles[17], Lisa Strug[18,19], Patrick Waters[14], Julitta de Bellescize[4,20], Pascal Vrielynck[21], Anne de Saint Martin[4,22], Dorothee Ville[4,23], Philippe Ryvlin[3,4,20], Alexis Arzimanoglou[3,4,20], Edouard Hirsch[4–6], Angela Vincent[14], Deb Pal[12], Nail Burnashev[4,7–9], Damien Sanlaville[1–4] & Pierre Szepetowski[4,7–9]