Language disorders

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Pagal LANGUAGE MODALITIES:

1) *verbal fluency* (**motor aphasia**)
2) *comprehension* (**sensory aphasia**)
3) *repetition* (**conduction aphasia**)
4) *naming* (**anomic aphasia**)
5) *reading* (**alexia**)
6) *writing* (**agraphia**)

Dar išskiriama LANGUAGE-RELATED DISORDERS:

7) **aphemia** – disturbance in *verbal output* (with preserved written language).
8) **aprosody** – kalbos *emocinio atspalvio* sutrikimas.
9) **auditory aphasia** (s. pure word deafness) – nesuvokia tik *girdimų* žodžių prasmės.

APHASIA

- *acquired inability* (of previously normal person) to **understand or/and express words as symbols for communication**;
primary sensory systems (vision, hearing), phonation motor mechanisms, mental state are intact.

ETIOLOGY & EPIDEMIOLOGY

1. Most common cause of focal aphasias – ***STROKE*** **in dominant hemisphere** (**a. cerebri media** supplies parasylvian zone exclusively!); 40% of stroke patients have aphasia!
2. Intracranial ***neoplasm*** (uncommon in sylvian region and are usually large before aphasia appears)
3. Traumatic brain ***injury***.
4. ***Neurodegenerative processes*** (e.g. Alzheimer's disease, vascular dementia).
5. ***Primary progressive aphasia*** (frontotemporal dementia such as Pick's disease) - insidious decline in language, either dysfluency or semantic anomia, that progresses to full dementia.

TYPES

Pagal ANATOMIC SUBSTRATE:

A. **Perisylvian**

I. Sensory aphasia (s. Wernicke aphasia, fluent aphasia)

II. Motor aphasia (s. Broca aphasia, nonfluent aphasia)
III. Conduction aphasia (s. associative aphasia)
IV. Global aphasia (s. mixed aphasia)

B. **Extrasylvian**
V. Transcortical aphasia

C. **Nonlocalizing**
VI. Nominal aphasia

D. **Subcortical**
VII. Thalamic, striatal, white matter aphasias

N.B. **NAMING**, **WRITING*** are impaired in ***all*** aphasias!
REPETITION impaired only in ***perisylvian*** aphasias

*writing errors typically parallel errors in spoken language

I. SENSORY aphasia (s. Wernicke aphasia, fluent aphasia, posterior aphasia)

- pažeistas **Wernicke center** (described in 1874, Karl Wernicke).

1. **Impairment in COMPREHENSION of language** – spoken (word deafness) & written (word blindness, s. alexia) & tactile.
   - comprehension ability may further decrease with testing (phenomenon called **fatigue** or **jamming**).
2. **Verbal OUTPUT is fluent** (articulated and effortless) – normal (or ↑) rate, sentence length, rhythm and melody (prosody); tačiau būdinga:
   1) PARAPHASIA (s. PARAPHRASIA, PARAGRAMMATISM) – patient uses wrong letters, words or combinations of words.
   2) LOGORRHEA – excessive language (pressured speech).
      paraphasia + logorrhea = "word salad"
   3) EMPTY SPEECH – kalba beprasmė.
   4) JARGON, NEOLOGISMS – naujų žodžių ir frazių sudarinėjimas (kuriuos suptanta (?) tik pats ligonis).
   5) impaired REPETITION and NAMING.
   - sunkiais atvejais kalba visai nesuprantama.
   - ACALCULIA commonly accompanies fluent aphasias.
3. Patients are ***unaware of their condition*** - are not depressed in acute stage (may exhibit elements of paranoia); making rehabilitation difficult.
4. Galima *right homonymous hemianopia* (or *superior quadrantanopia*) – dėl Meyer-Archambault loop of visual pathway;
   frequent ***absence of associated neurological deficits*** may lead to erroneous diagnosis of *psychosis* or *schizophrenia* (in younger patients) or *dementia* (in older patients).

CLINICAL RULE: sudden onset of fluent aphasia without hemiparesis suggests **embolus** (to posterior branch of middle cerebral artery).

Differentiate from:

1) ***word salad*** of **SCHIZOPHRENIA** and ***confused speech*** of **DELIRIUM** (both have normal auditory comprehension).
2) **PURE WORD DEAFNESS** (disorder of *auditory* input with preserved *written* input).
3) **DYSARTHRIA**
   Dysarthric speech - *stereotyped* speech errors (repeating the same errors when trying to produce the same sounds).
   Paraphasic speech - substituted letters occur in *variable* pattern.

II. MOTOR aphasia (s. Broca aphasia, nonfluent aphasia, anterior aphasia, speech apraxia)

- pažeistas **Broca center** (described in 1861 by Paul Broca).

1. **Impairment in language OUTPUT** – central feature; spontaneous speech:
   - **decreased, slow, halting** (words are hard to come by); sunkiais atvejais – almost complete mutism.
     N.B. *complete muteness* is seen in some psychiatric syndromes (e.g. catatonia, elective mutism) but not in aphasia!
   - **dysarthric**
     Patient may be able to hum *melody* normally (however, if patient is musician and views music as language, deficits in "producing" music will be experienced).
     Curses or other ejaculatory speech may be well articulated.
   - **simple grammar ("telegraphic" speech)**:
     – naudoja tiktai **key words** (vardininko linksnyje, praleidžia artikelius, įvardžius, būdvardžius) - ***agrammatic*** speech;
     – 2-3 words express the whole range of meaning and emotion (markedly ***dysprosodic*** speech);
     – sometimes the words retained are those which were being spoken at accident time.
   - REPETITION, NAMING impaired.
   - AGRAPHIA (agraphia esti visų afazijų atvejais, kuomet sutrikęs verbal output!) – nebelikę būdų kaip save išreikšti → tremendous frustration! (may contribute to mutism, depression)
2. **Normal COMPREHENSION** – supranta kas parašyta, bet negali perskaityti garsiai;
   - some difficulty with relational words (such as up and down, inside and outside).
   - patient is ***aware of impairment***.
3. Dažna (80%) *right hemiparesis* (esp. brachiofacial).
4. *Conjugate ocular deviation to left* is often present initially (due to frontal eye field lesion).

Differentiate from **APHEMIA (s. little Broca aphasia, cortical anarthria, speech apraxia)** - disturbance in *verbal output* with preserved written language (žr. žemiau).

III. CONDUCTION aphasia (s. associative aphasia)

- it was thought to be due to **arcuate fasciculus** lesions (most often deep to supramarginal gyrus); now it appears that it is due to lesions **in and around auditory cortex (areas 40, 41, 42)**.

- most common cause - occlusion of *angular branch of middle cerebral artery*.

1. Wernicke center intact - **normal COMPREHENSION**.
2. Broca center intact - **verbal OUTPUT is fluent** (but paraphasic).
3. Severely **impaired REPETITION and NAMING**.
   – PARAPHASIAS are common (esp. substitutions of phonemes), and naming is often limited by these paraphasic intrusions.
   – ***reading aloud is disturbed*** (severely paraphasic output), but ***reading for comprehension is normal***.

IV. GLOBAL aphasia (s. mixed aphasia)

- pažeista **visa left parasylvian area**.

- **sutrikę VISI kalbos, kaip bendravimo priemonės, aspektai**.
- kartu būna right hemiplegia-hemianesthesia-homonymous hemianopia.

- patients who do not make rapid recovery soon after onset have poor prognosis.

V. TRANSCORTICAL aphasia

- **left parasylvian area** is intact but **isolated** from rest of hemispheric cortex.
  - dažniausia priežastis – ***infarction in watershed area*** (junction of vascular territories) – from prolonged hypotension or hypoxia in patients with severe carotid stenosis, cardiac arrest.
- **preserved REPETITION**!!!
- skirstoma:
  1) TRANSCORTICAL MOTOR APHASIA (damage in left **supplementary motor area** or between that area and Broca center) – kaip Broca aphasia (but repetition normal!).
  2) TRANSCORTICAL SENSORY APHASIA (damage in left **posterior parietal region**, e.g. in Alzheimer's disease) – kaip Wernicke aphasia (but normal repetition, with apparent unawareness of what is said).
  3) MIXED (unusually frequent in Creutzfeldt-Jakob disease) – *ECHOLALIA*, o kitos kalbos funkcijos iškritę (t.y. kaip global aphasia, but repetition normal!).

SUPPLEMENTARY MOTOR AREA APHASIA (variant of transcortical motor aphasia) - damage to left **medial frontal structures** (cingulate cortex, supplementary motor area); differences from transcortical motor aphasia:
- occlusion of **anterior cerebral artery**.
- slow hypophonic output that improves considerably with repetition.
- weakness of right lower extremity and shoulder + normal strength in arm & face.

VI. NOMINAL aphasia (s. anomic aphasia, amnestic aphasia)

- lesion **anywhere in cerebrum**.*

*N.B. anomia is ***not reliable localizing abnormality*** - may result from ***toxic / metabolic encephalopathies*** or ***space-occupying lesions*** (far from speech area that exert pressure effects) – always search for reversible, metabolic causes!

Frequent cause is Alzheimer disease!

- vienintelis sutrikimas – **difficulty in finding names for seen words or pictures** (visual information is not processed and transmitted to Wernicke area), i.e. defect of confrontational naming → results in:
  - **empty speech** (lack of substantive words, with substitution of many nonspecific words that fail to communicate idea satisfactorily);
  - excessive **word-finding pauses**!
  - *patient acts as though the name has been forgotten, and may give functional descriptions instead*; if description demands substantive word that cannot be produced, another description is tried - this rapidly produces circuitous output (**circumlocution**).
- **no difficulty with speech or understanding of auditory information**! reading and writing may be entirely normal!

N.B. ***naming is disturbed in all aphasias***! (anomia often remains complaint of many well-recovered aphasics).

Lesion of **left temporal pole** (**area 38**) - inability to retrieve **names of places & persons** but preserved ability to retrieve common nouns (i.e. **names of nonunique objects**), verbs and adjectives.

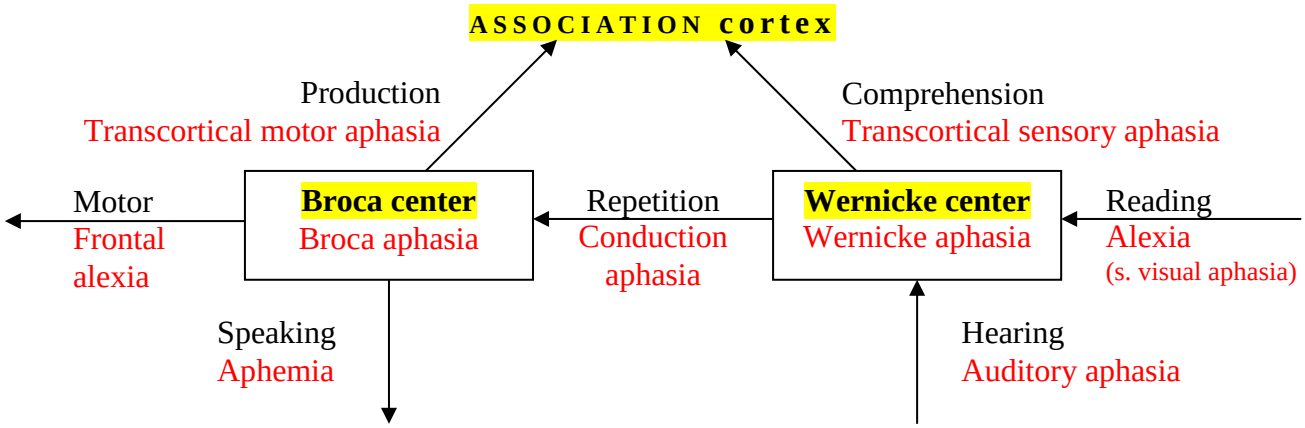
VII. SUBCORTICAL aphasia – pažeidus **subkortikalines struktūras**:

1) **thalamic (anterolateral nuclei of thalamus, thalamic peduncles)** - Wernicke-like aphasia with good comprehension and repetition (≈ transcortical sensory aphasia).
2) **striatal (basal motor nuclei)** ≈ transcortical motor aphasia + paraphasias*.
3) **internal capsule** - usually manifest dysarthria.

*paraphasic errors are not due to lesion of cerebral surface, as was claimed traditionally

- very good prognosis (transient nature can be accepted as diagnostic characteristic!).

Wernicke-Geschwind model of language and language disorders:



DIAGNOSIS

- differentiate from DISORDERS OF MECHANICAL PROCESS OF SPEECH (dysarthria, dysphonia) - grammar and word choice are correct.
- various formal tests for diagnosing aphasia (e.g. Boston Diagnostic Aphasia Examination) are available. However, bedside interaction usually suffices! see D1 p.
- all other *COGNITIVE FUNCTIONS* are intact (except – ***verbal memory***).

Ligonio tyrimo metodika – žr. EXAM TECHNIQUE.

APHASIA	Verbal output	Compre-hension	Repetition	Naming	Reading aloud / comprehension	Associated signs
Broca	**nonfluent**	normal	impaired	marginally impaired	poor / good	RHP (esp. lower face)
Wernicke	fluent (paraphasic)	**impaired**	impaired	impaired	poor / poor	± RHH
Conduction	fluent (paraphasic)	normal	**impaired**	impaired (paraphasic)	poor / good	± RHS
Global	**nonfluent**	**impaired**	**impaired**	**impaired**	poor / poor	RHP, RHS, RHH
Anomic	fluent	normal	normal	**impaired**	variable	-
Transcortical: MOTOR SENSORY MIXED	 nonfluent fluent nonfluent	 normal impaired impaired	 **normal** **normal** **normal**	 impaired impaired impaired	 poor / good poor / poor poor / poor	 RHP ± RHH RHP, RHS

RHP – right hemiparesis
RHS – right hemisensory deficit

RHH – right homonymous hemianopia

N.B. **NAMING**, **WRITING** are impaired in all aphasias!

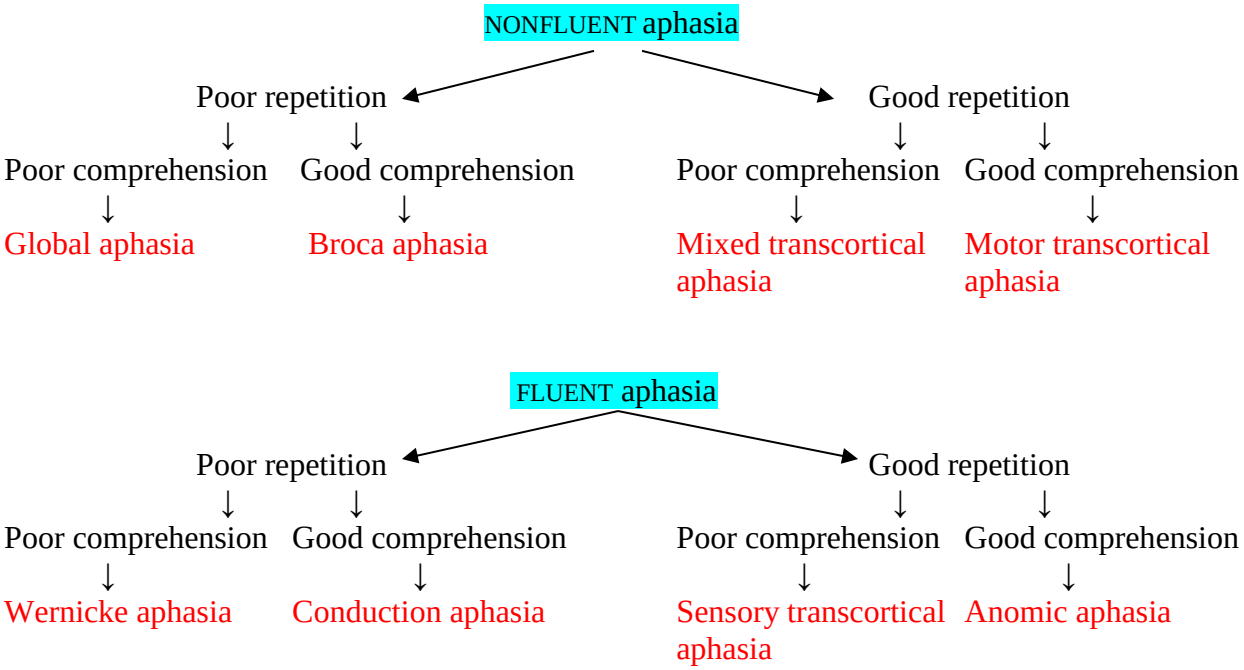
Characteristic responses when shown ***picture of chair***:
Broca aphasia: "Tssair"
Wernicke aphasia: "Stool" or "Choss" (neologism)
Conduction aphasia: "Flair….no, swair…tair"
Anomic aphasia: "I know what it is… I have a lot of them"

N.B. aphasia localizes lesion in left cerebral cortex!

Quick differential:

Aphasia Subtype	Fluency	Comprehension	Repetition
Nominal	Normal	Normal	Normal
Broca	Impaired	Normal	Impaired
Wernicke	Normal	Impaired	Impaired
Conduction	Normal	Normal	Impaired
Transcortical Motor	Impaired	Normal	Normal
Transcortical Sensory	Normal	Impaired	Normal
Transcortical Mixed	Impaired	Impaired	Normal
Global	Impaired	Impaired	Impaired

Differentiation algorithm:



TREATMENT

Treatment is controversial.

- patients treated **soon after onset** do best; but *treatment delays* are not detrimental (are warranted until patient is neurologically stable).
- patients systematically treated by ***qualified speech therapists*** improve more.
- special **language therapy techniques (speech therapy)** are available (e.g. *melodic intonation therapy*).
- training in use of **visual imagery** as internal cue helps to overcome word blocking of Broca aphasia.

- **picture board** may circumvent expressive language deficit.
- *drugs* have little success.

N.B. **depression** is associated with left hemisphere injury (esp. deep frontal regions - Broca aphasia, global aphasia, subcortical aphasia with anterior extension).

PROGNOSIS

- **children < 8 yr** often regain language after severe damage to either hemisphere.
- **> 8 yr old** - most recovery occurs within first 3 months (but improvement continues to variable degree up to 1 year).
- ***comprehension*** improves more than ***language*** (i.e. ***fluent aphasics*** respond better to rehabilitation than do ***non-fluent aphasics***).
- ***if right hemisphere is dominant*** (15%) for hand and speech - injuries to either hemisphere can cause aphasia, but nearly all recover rapidly!!!
  In most left-handers hemispheric dominance for language is incomplete!
  Left-handers are more likely to become aphasic because of bilateral representation, but their aphasia tends to be milder and more brief!
- prognozės blogėjimo tvarka:

  subcortical, anomic, conduction, transcortical > Broca > Wernicke > global

- **traumatic** cases do better than **stroke**.
- patients are often assumed to be incompetent (because of reduced communication ability), but patients have intact nonverbal communication, thinking, expressing opinions!

ALEXIA (s. visual alexia, sensory alexia, visual aphasia, word blindness, alexia without agraphia)

- pažeista left **occipital medial surface (visual cortex) + splenium corporis callosi** – pacientas gali naudotis tik right visual cortex, bet informacija į kitą pusę nepatenka - ***visual information has lost access to language area*** (DISCONNECTION theory).

- first described by Dejerine in 1892.
- cause is nearly always **stroke** in ***a. cerebri post. sin***.
- nesuvokia tik **PARAŠYTŲ žodžių prasmės** - word blindness (***verbal alexia***);
  - retained ability to read letters - no letter blindness (***literal alexia***)!
  - rega nesutrikusi.
  - palpuodamas (taktiliškai) raštą supranta; supranta raides nupieštas ant delno.
  - rašyba nesutrikusi (patient can write but cannot read his / her own written output!).
  - many have disturbed *color naming* as well as mild *anomia*.
- diferencijuojama nuo **anarthria (s. motor alexia)** – loss of power to *read aloud* but *reading for comprehension* is normal.

ALEXIA with AGRAPHIA

- pažeista left **gyrus angularis**.

Gerstmann syndrome (finger agnosia, agraphia, acalculia, right-left confusion) - pažeista **parietal-temporal-occipital association cortex**.

Angular gyrus syndrome (alexia with agraphia + Gerstmann + anomia) – when Gerstmann localization includes gyrus angularis.

- **speaking & understanding spoken language** are normal.
- ACQUIRED ILLITERACY - previously educated patient is rendered unable to read and write.
- inability to read both letters and words (***verbal & literal alexia***); **cues** are of little help:
  - tracing letter with finger does not aid in identification;
  - cannot decipher word when it is spelled aloud.
- pažeidus tik **corpus callosum**:
  a) ***užpakalinę dalį*** – gali skaityti tik kai tekstas in right visual field (**HEMIALEXIA**).
  b) ***priekinę dalį*** – negali rašyti kaire ranka.

Writing is abnormal in **all aphasias**! (writing is consistently affected even in subtle aphasia; writing errors parallel speaking errors) - **tests of writing ability** can be used as screening device to detect aphasia!

N.B. agraphia may be without aphasia!

APHEMIA (s. little Broca aphasia, cortical anarthria, speech apraxia)

- disturbance in ***verbal output*** (BUCCOFACIAL APRAXIA) with preserved ***written language*** (i.e. nonfluent aphasia without agraphia) - mute patient able to communicate using written language (normal grammatical)!

N.B. speech problem rather than impairment of language!

- manoma, kad pažeidimas ***paviršinis Broca center*** arba ***šiek tiek žemiau Broca center*** (klasikinės Broca afazijos atveju pažeidimas nusitęsia gan giliai į požievį).
- laryngeal pathology should be ruled out!
- ligoniai greitai pasveiksta.

AUDITORY APHASIA (s. word deafness, auditory verbal agnosia)

- two known loci of pathology:
  a) single lesion **deep in left superior temporal region** (deep to Wernicke; affects primary auditory cortex or pathways to it from medial geniculate nucleus).
  b) bilateral lesion involving **mid-portion of superior temporal gyrus**.

N.B. Wernicke area is not involved!

It is **sensory transmission problem** rather than language disturbance - resembles deafness more than aphasia (so also been called AUDITORY VERBAL AGNOSIA).

- nesuvokia tik **GIRDIMŲ žodžių prasmės**; cannot repeat; klausa nesutrikusi.
- **reading is intact** – patients often carry with them ***writing tablet*** for others to use.

APROSODY

- lesions of **parasylvian area** in *nondominant (representational) hemisphere*.

Sutrikę kalbos **affective components** - emocinė gestikuliacija, intonacija:

a) pažeidus **mirroring area of Broca center** – nesuteikia affective component ***savo kalbai*** ("negyva" kalba);
   - AMELODIA (MOTOR APROSODIA) - loss of melody, prosody, emotional intonation in verbal output.
   - EXPRESSIVE AMUSIA - inability to produce melody when singing;

– decreased facial expression, sparse use of gestures.
N.B. easily misinterpreted as depression!

b) pažeidus **mirroring area of Wernicke center** – nesuvokia ***svetimos kalbos*** affective components (SENSORY APROSODIA, RECEPTIVE AMUSIA, etc).

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