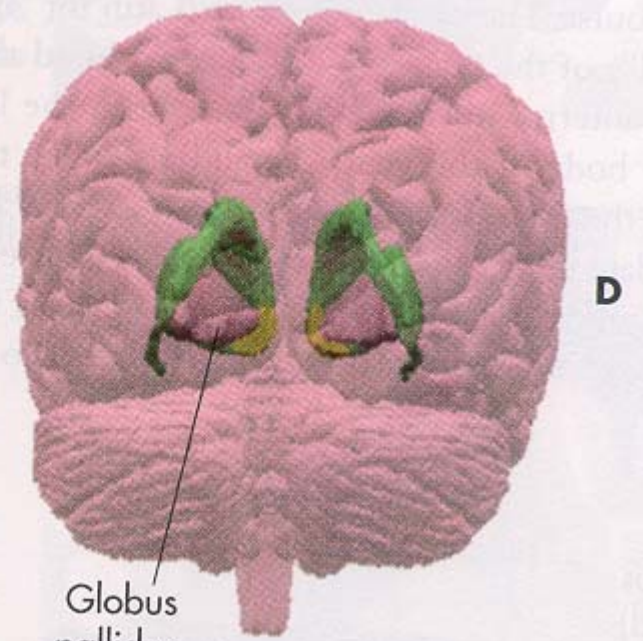
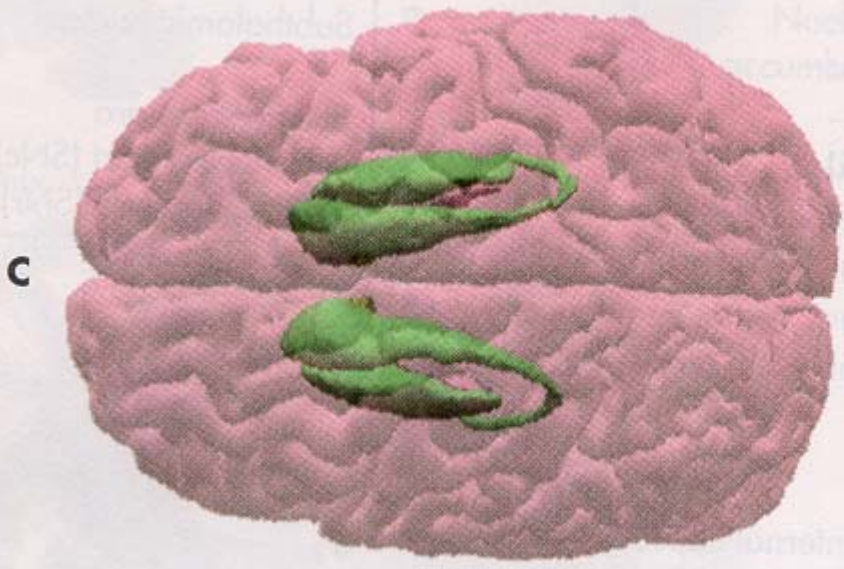
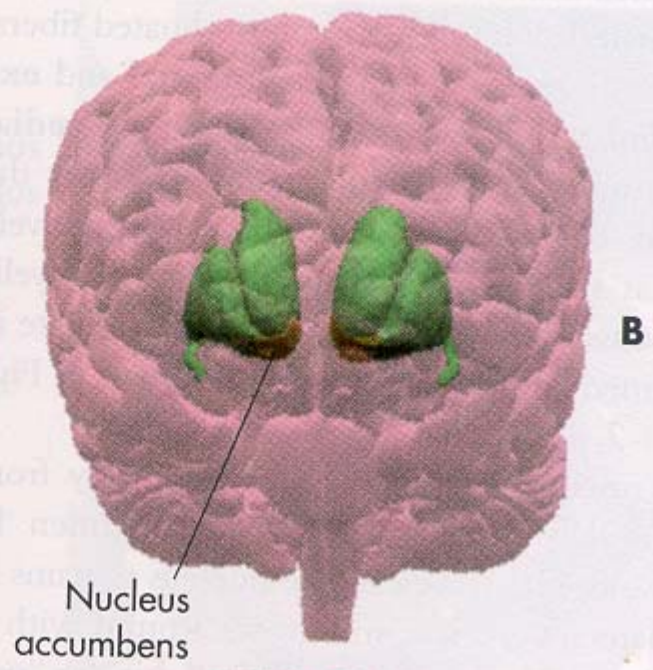
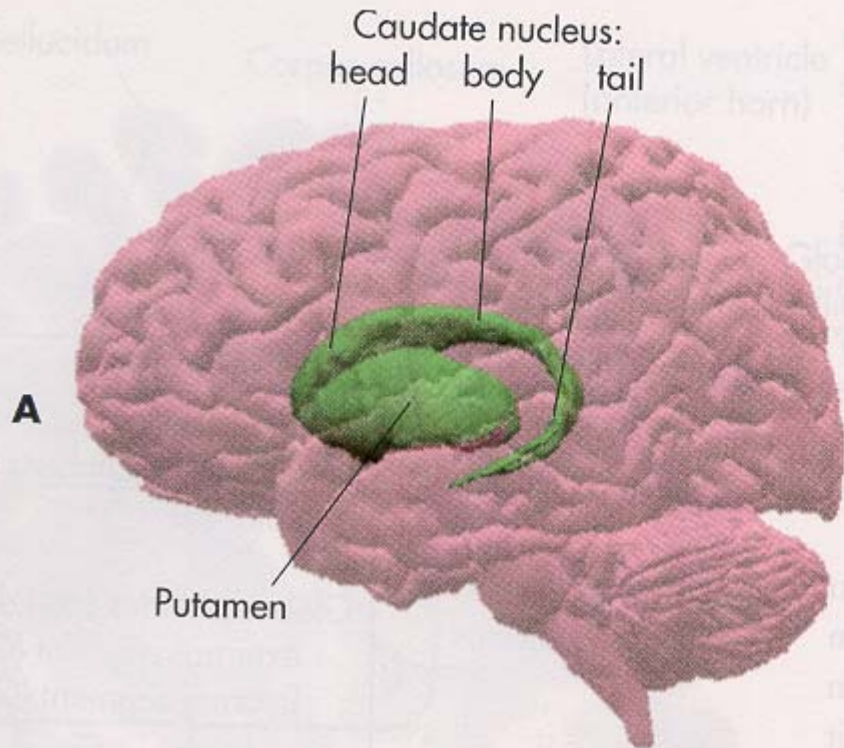
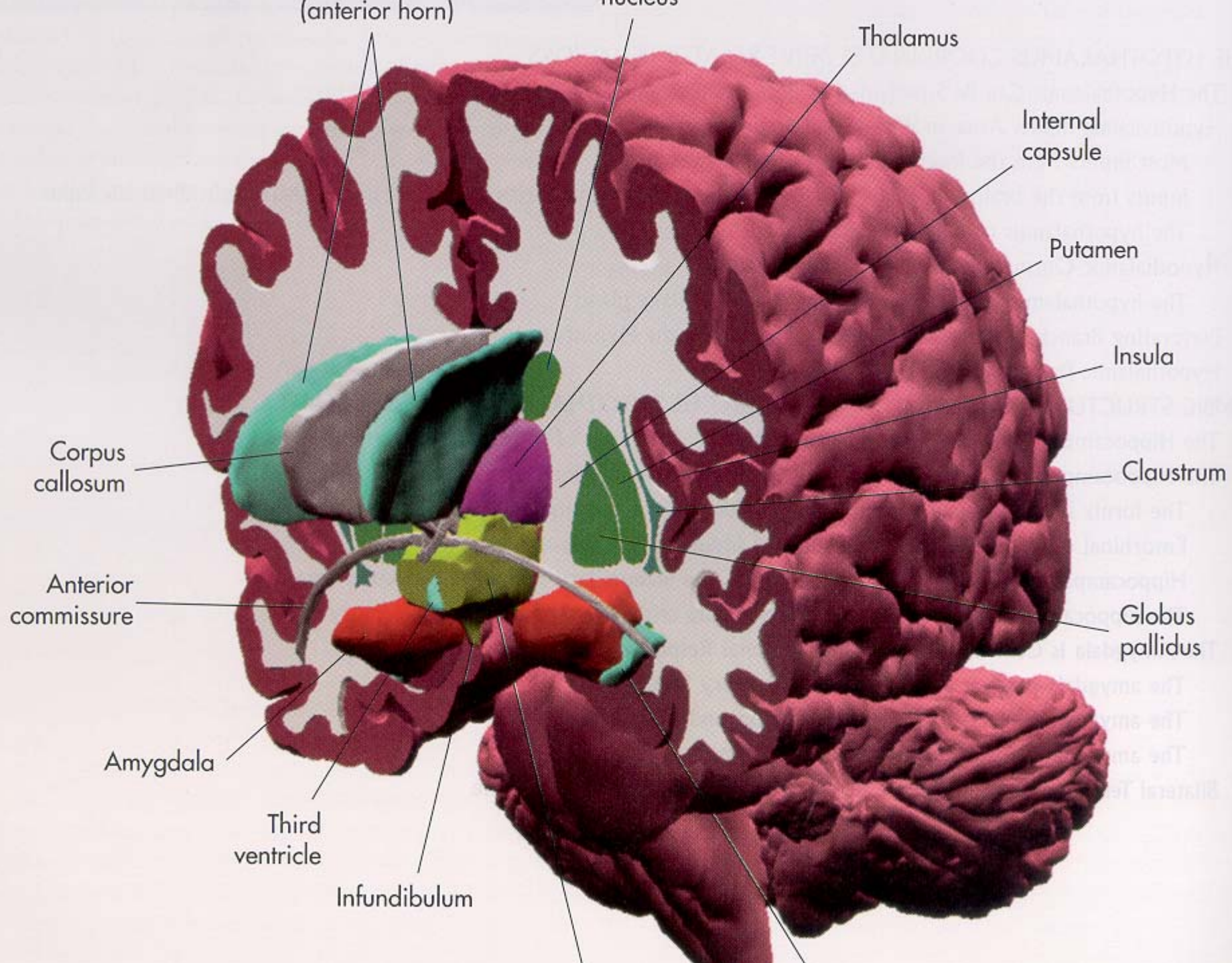


# **W.A. Mozart: izvid nevrologa in kognitivnega nevrofiziologa**

Z. Pirtošek



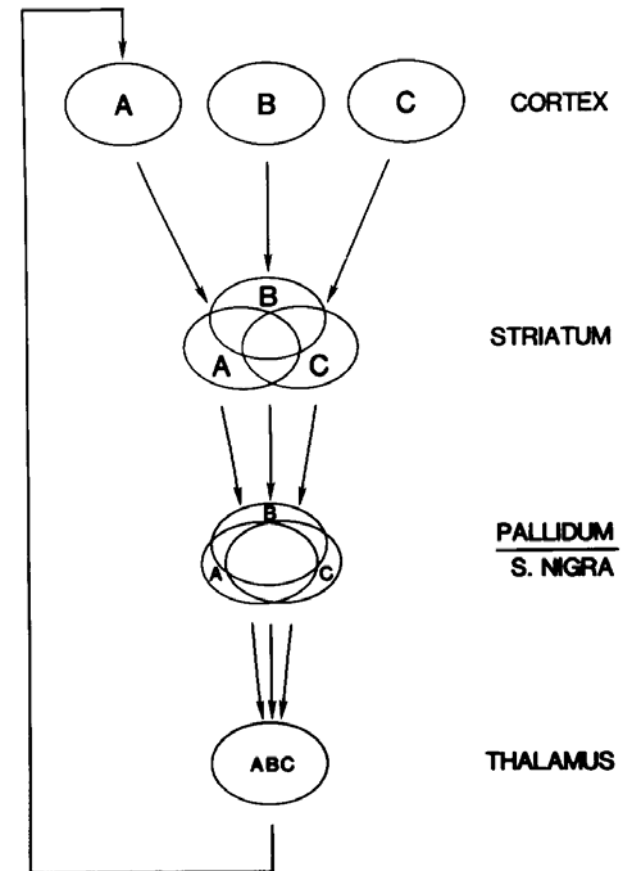
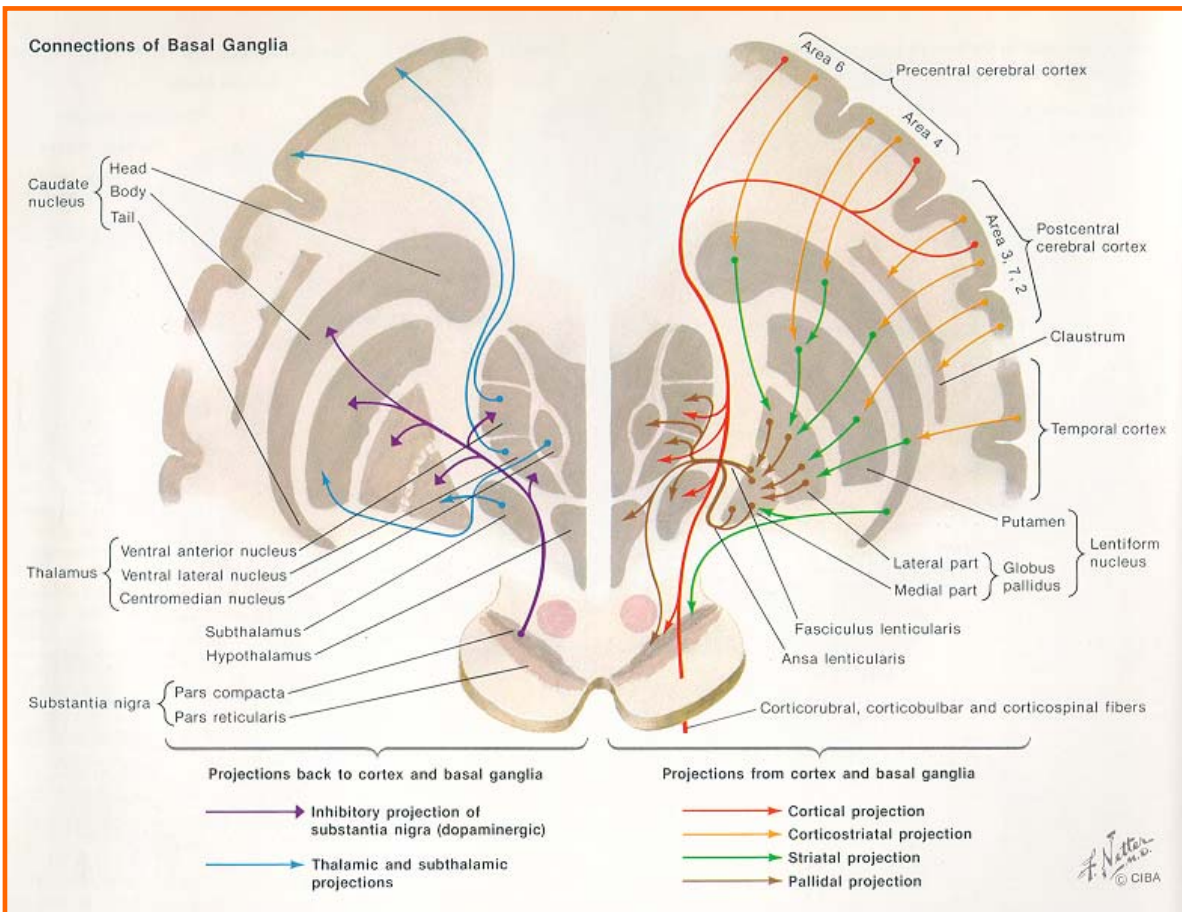




# Bazalni gangliji

Kaj so ?

Kaj je njihova funkcija ?



# BOLEZNI BAZALNIH GANGLIJEV

- PREVEČ GIBANJA: Parkinsonizem
- PREVEČ GIBANJA: Tiki
  
- *MOTORIKA*
- *KOGNITIVNI PROCESI*
- *EMOCIJE, PSIHOZA*
- *OSEBNOST*

# PREVEČ GIBOV

Tik: ponavljajoča se, nehotena, nenadna, stereotipna motorična akcija

- Motorični tiki (enostavni ali kompleksni)
- Vokalni tiki (enostavni ali kompleksni)

Nehoten gib?

# Vzroki tikov

Neznani

genetski  
*GDT*

infekcije

*pediatrična avtoimuna nevropsihiatrična motnja povezana s streptokokno infekcijo (PANDAS)*

zdravila

*amfetamini, kokain, antipsihotiki*

toksini

razvojne motnje

nevrodegenerativne bolezni

drugo

*poškodba glave*

# GDT - diagnostični kriteriji

- multipli motorični in vsaj en vokalni tik
- tiki se pojavljajo večkrat dnevno vsaj eno leto
- tiki se skozi čas spreminjajo
- tiki se pojavijo pred 21 letom
- odsotnost druge bolezni ali stanja, ki bi pojasnila tike
- tike zabeleži zanesljiv preiskovalec ali se jih posname na videokamero



# GDT - vedenjske motnje

- obsesivno kompulzivno vedenje
- motnje pozornosti
- koprofalije
- eholalije

# GDT- zdravljenje

- nadzor tikov
- zdravljenje vedenjskih motenj
- medikamentozno zdravljenje
- psihoterapija in vedenjska terapija
- operativno zdravljenje

# GDT - zdravljenje tikov

- dopaminski antagonisti
  - tipični nevroleptiki (haloperidol, pimozyd)
  - atipični nevroleptiki (risperidon, ziprazidon, olanzapin)
- alfa-2-agonisti (klonidin, guanfacin)
- inhibitorji monoaminov (tetrabenazin)
- dopaminski agonisti (pergolid – nizki odmerki)
- drugo (klonazepam, botulin)

# GDT - zdravljenje motenj pozornosti

- alfa-2-agonisti (klonidin)
- stimulansi (metilfenidat, pemolin, amfetamin)

# GDT - zdravljenje obsesivno-kompulzivnega vedenja

- selektivni zaviralci privzema serotonina (SSRI)
  - fluoksetin
  - fluvoksamin
  - paroksetin
  - sertralin
  - venlafaksin
  - citalopram











# CURRICULUM VITAE

- 1756 Salzburg (oče Leopold, mati Anna Maria Pertl)
- 1756: Versuch einer gruendlichen Violinschule
- Wunderkind
- 1782 Constanze Weber
- 1791 'hitziges Frieselfieber' – revmatska vročica?
- 1809 Constanze poroka z von Nissen-om (biografija)



# NISSEN

“The ear was malformed, completely lacking the conch or outer convolution”

*....pri malformacijah srčnih zaklopk*

# SCHACHTNER

- “He was of a fiery disposition, no object held his attention by more than a thread. I think that if he had not had the advantageously good education which he enjoyed, he might have become the most wicked villain, so susceptible was he to every attraction, the goodness or badness he was not yet able to examine.”

# SUARD

“ His mind, limited to those ideas which concerned music, shed a bright light on all that interested his talent, but shewed little aptitude to occupy itself with other things. He was extremely irritable; his affection were lively, but superficial and of short duration.”

# Nissen (Aloysia Lang)

- “Even when he washed his hands in the early morning, he walked up and down the room at the same time, never standing still, at the same time tapped one heel against the other and was always thoughtful. At table he often took a corner of his napkin, crumpled it up tightly, rubbed it round below his nose, and appeared in his thoughtfulness to be unaware of what he was doing, and often he would grimace with his mouth at the same time...Also, he was always moving his hands and feet, always playing with something, e.g. with his hat, pockets, watch-ribbon, tables, chairs...”





# SESTRIČNI

“Well, I wish you good night, but first shit into your bed and make it burst. Sleep soundly, my love, into your mouth your arse you’ll shove...Oh, my arse is burning like a fire! What on earth does it mean? Perhaps some muck wants to come out? Why, yes, muck, see and smell you...and..what is that? Ye, gods, it is so - what a long melancholy note!”

# PISMO SESTRICNI

Ma tres chere cousine,  
were you never in Berlin?  
Your cousin of virtue rare  
In weather foul or fair  
W.A. Mozart,  
who shits without a fart.

# SESTRIČNI

*' I shit on your nose  
So it runs down your chin*

*Dreck! Dreck! O Dreck!*

*Blow into my rear  
Nothing could be finer  
Bon appetit'*

# POSVETILO *AVE VERUM*

Franz Susmayr, Muckshitter, Shitting house

# Karoline Pichler

(Mozart je mirno igral...)

“But then he suddenly tired of it, jumped up, and in the mad mood which so often came over him, he began to leap over tables and chairs, miaow like a cat, and turn somersaults...”

# BESEDILO ZA K231

Leck mich im Arsch

*Lick out my arsehole*

*Lick it till it's good and clean*

# W. Hildesheimer

“Mozart’s musical genius shines out in his words as well, when he manipulates them like musical associations... His compulsive delight and ease in association are capable not only of producing euphony and rhythm with disparate and seemingly arbitrary combinations of sound but also of keeping the connotations always in mind. In writing he yields himself up to the flow of words, going far beyond the comprehensible, reveling in, intoxicated by the sounds and the continually changing meanings they suggest. In this intoxication the inhibition of convention and bourgeois taste fall by the wayside.”

Home this site



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Wolfgang Amadeus Mozart

English Nederlands Français Português



# W. Hildesheimer

“He worked out an elaborate composition entirely in his head, and then, perhaps later, sat down to write it straight off; and the writing was so much a mechanical process that he could engage in a game of skittles, or a conversation, at the same time. Mozart’s genius went hand in hand with a near total absorption, which rendered him unfit for everyday life. He had an overmastering compulsion to compose”

# MATERI

Yesterday, though, we heard the king of farts  
It smelled as sweet as honey tarts  
While it wasn't in the strongest of voice  
It still came on as a powerful noise

# TIKI

- Grimase
- Ramena
- Tapkanje z nogami

# VOKALIZACIJE

- Mijavkanje
- Nesmiselne besede

# KOMPULZIJE

- Rituali pred spanjem
- Vrtenje
- Skok v naročje cesarice pri 8 letih
- Prevali

# OBSESIJE

- Patološki strah pred trobento
- Ure
- Bele mačke
- Varnost žene
  
- ? glasba v glavi

# FUGE

Ponovno odkritje (Haydnovi kvarteti)

Fuge: kaos, a obenem rigidna struktura

- 'Tourette pomeni stalno borbo med silo kompulzije in potrebo po kontroli le-te'

# MOZARTOV EFEKT

- Znižanje krvnega pritiska (A. Savanfound)
- Manj epileptičnih napadov (J. Jenkins)
- Podgane: bolj uspešno učenje labirinta
- Zvišan IQ



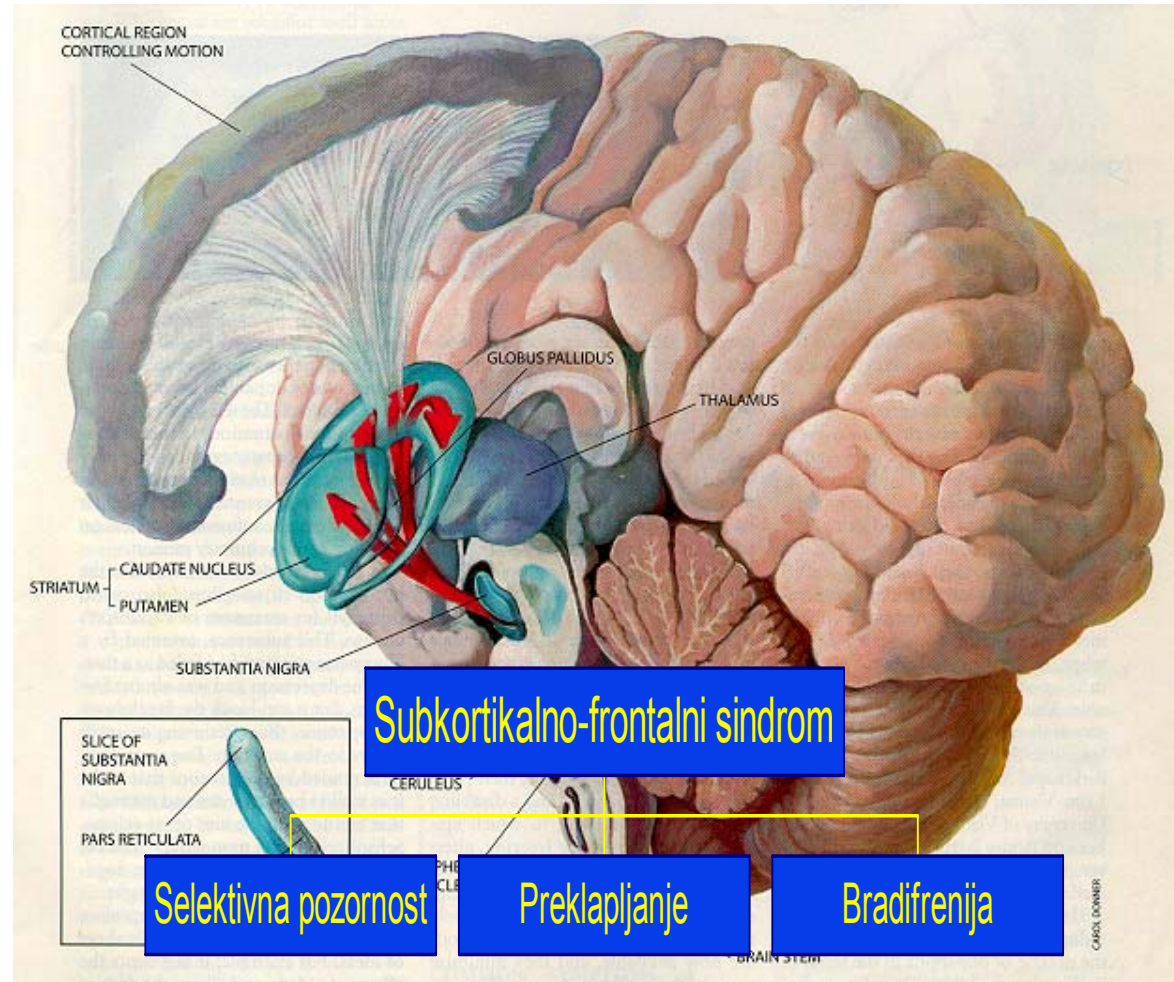
# DEPRESIJA

“I can't describe what I have been feeling - a kind of emptiness, which hurts me dreadfully - a kind of longing, which is never satisfied, which never ceases, and which persists, nay rather increases daily.”

# GDT: Izbrana metoda

Potenciali, izzvani s testnim vzorcem, ki odseva funkcije čelnega režnja

Reakcijski čas (RČ)



# GDT: OSNOVNA MOTNJA?

- ena od osnovnih motenj: selektivna pozornost??

# ELEKTROFIZIOLOGIJA SLUŠNEGA SIGNALA

## EVOCIRANI POTENCIALI

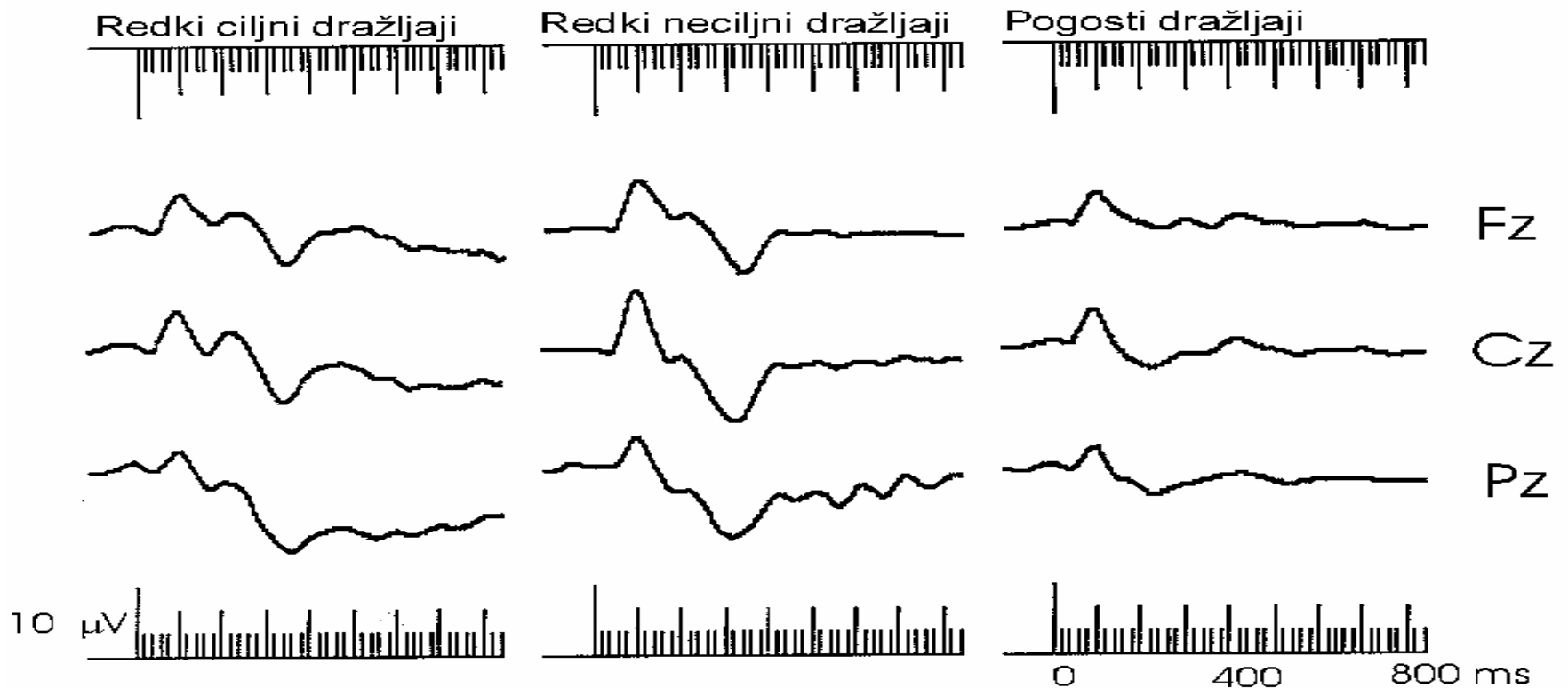
- kohlearni potenciali
- potenciali debla
- kortikalni potenciali (MMN, P1, N1, P2)
- endogeni potenciali (P3, pozornost, spomin...)

# EVOCIRANI POTENCIALI

- latenca
- amplituda
- topografija
  
- Zgodnji (“obvezni”): 80 - 200 ms
- Pozni (“kognitivni”): 200 - 1000 ms

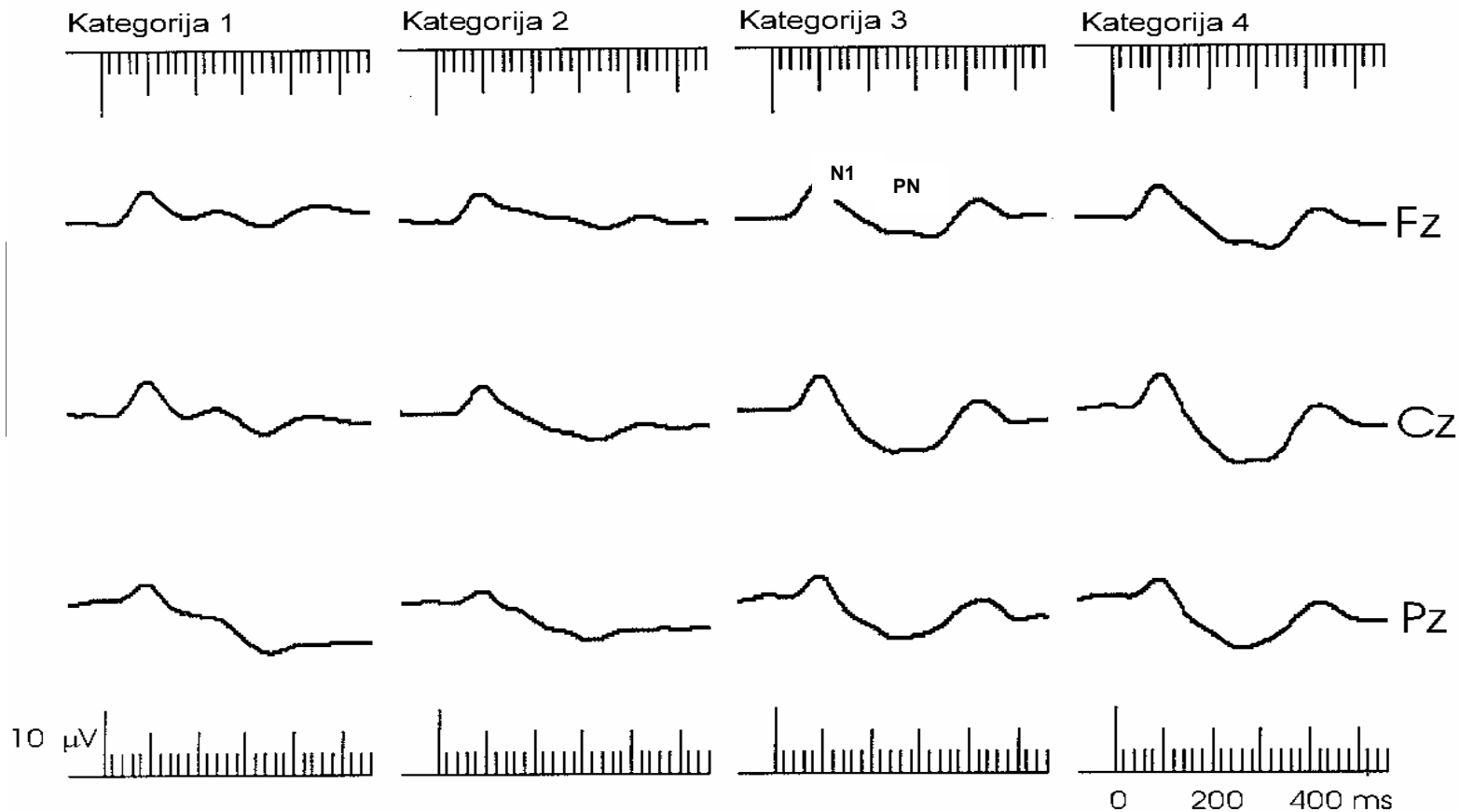
# VAL P300

Testni vzorec nenavadnosti (val P300)



# SELEKTIVNA POZORNOST

Slušna selektivna pozornost



Dražljajev 167  
Poprečenj 156  
Pritiskov 162

Kategorija

Reak. čas 482

Fz-L+R

15,0  $\mu$ V/raz

Cz-L+R

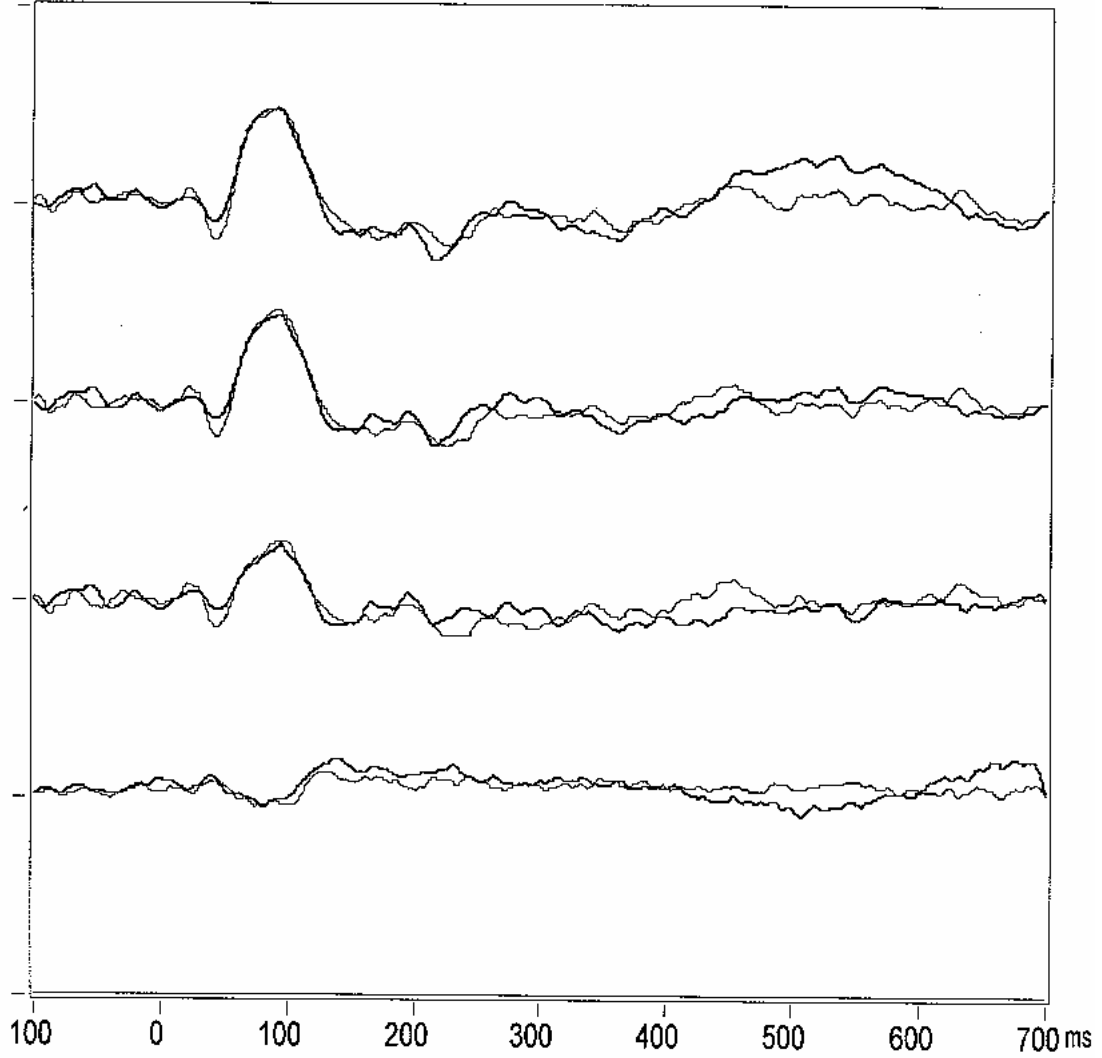
15,0  $\mu$ V/raz

Pz-L+R

15,0  $\mu$ V/raz

Pg2-Fp2

15,0  $\mu$ V/raz





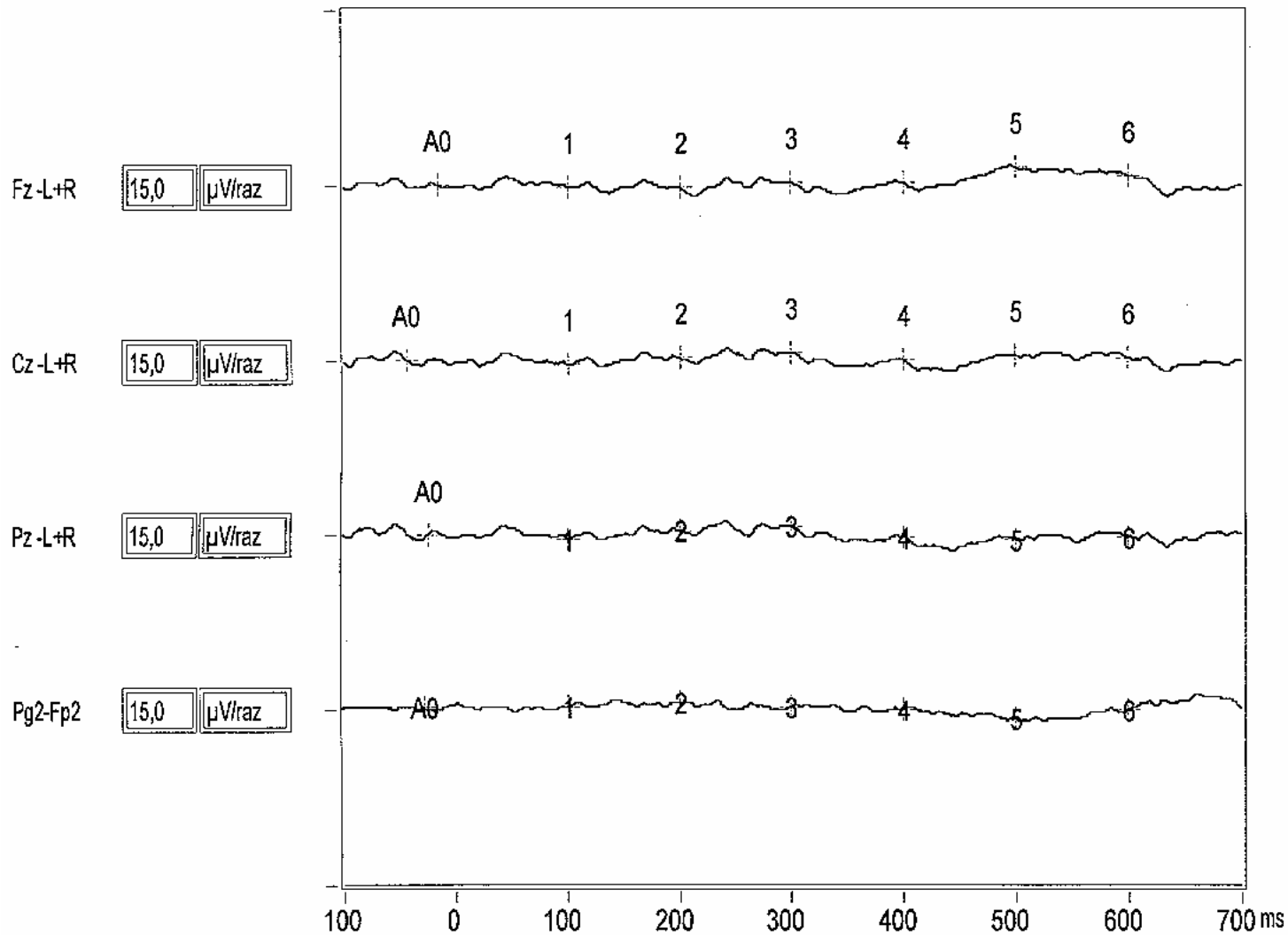
Dražljajev 331

Kategorija

Poprečenj 309

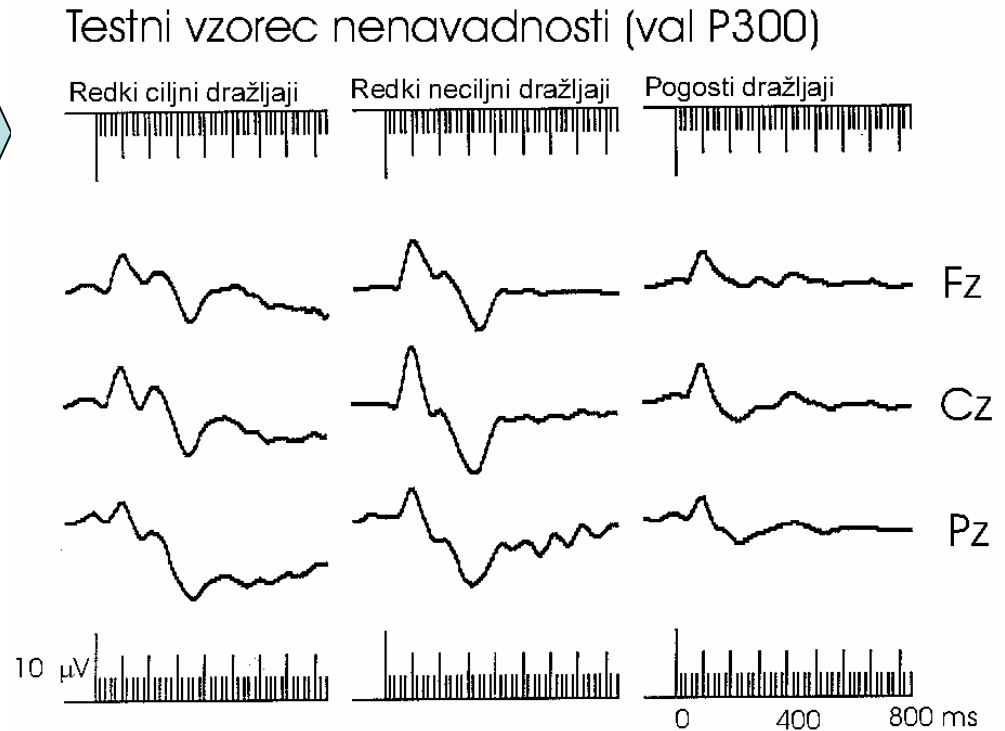
Pritiskov 171

Reak. čas 477



# Izbrana metoda

- Testni vzorec nenavadnosti (“oddball”, val P300)
- Slušna selektivna pozornost
- Vidna selektivna pozornost
- Reakcijski čas



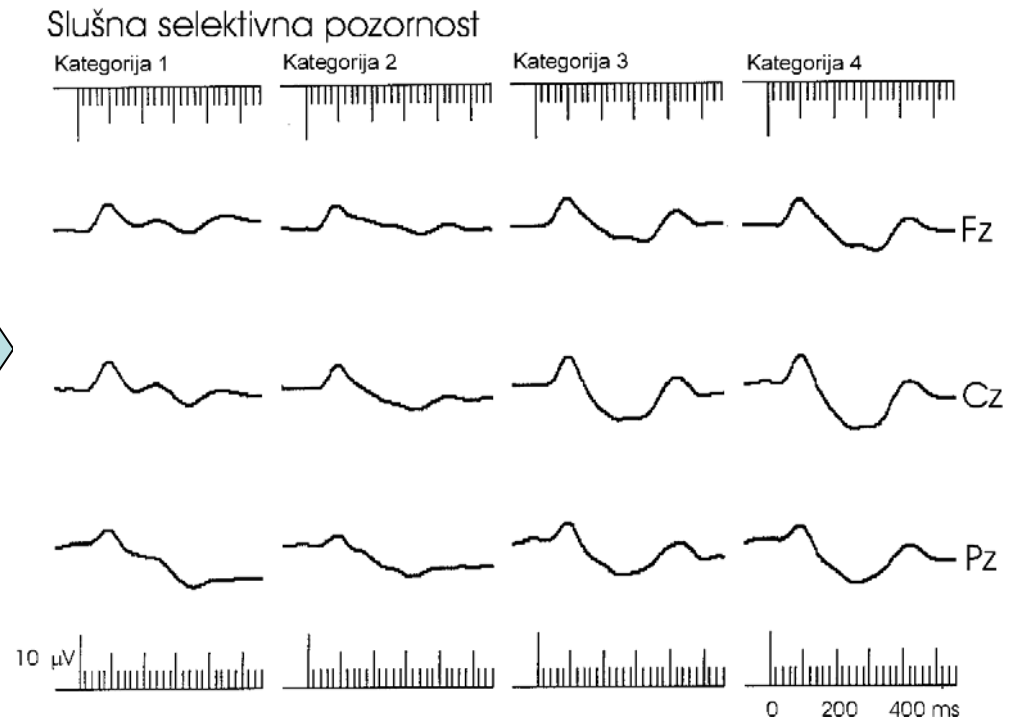
# Izbrana metoda

- Testni vzorec nenavadnosti (“oddball”, val P300)

• Slušna selektivna pozornost

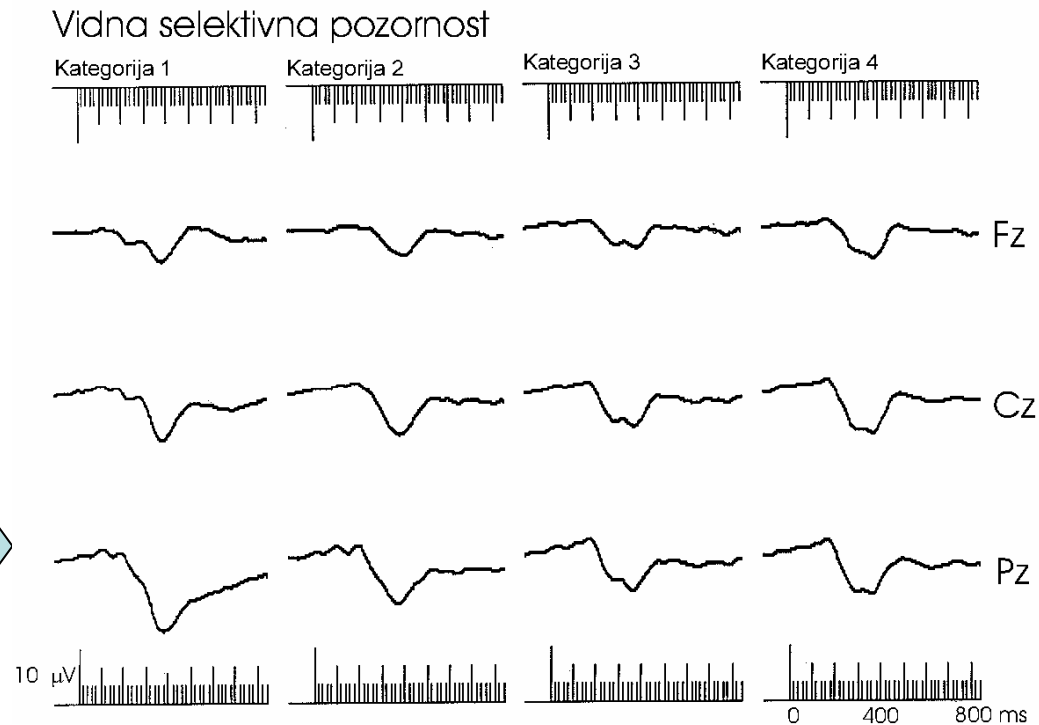
- Vidna selektivna pozornost

- Reakcijski čas



# Izbrana metoda

- Testni vzorec nenavadnosti (“oddball”, val P300)
- Slušna selektivna pozornost
- Vidna selektivna pozornost
- Reakcijski čas



# Izbrana metoda

- Testni vzorec nenavadnosti (“oddball”, val P300)
- Slušna selektivna pozornost
- Vidna selektivna pozornost
- Reakcijski čas

# Premalo (IPB) in preveč (GDT) gibov

N Starost Trajanje Začetek Zdravilo

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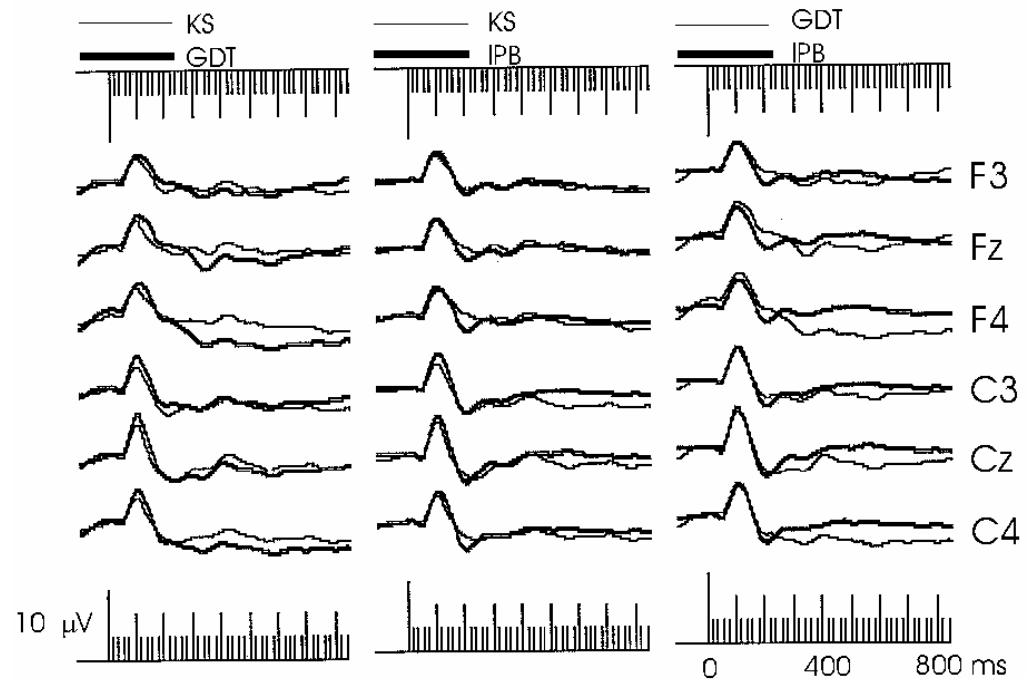
Zdravi	6	34			
GDT	6	31	184	6	Sulpirid
IPB	6	44	29	41	L-dopa, bromokriptin, deprenil

---

# DVA NASPROTNA POLA: PD - GDT

*GDT*

- *daljši RČ*
- *“P3”- pogosti dražljaji*
- *krajši P1v*
- *nenormalna SP*



# DVA NASPROTNA POLA: PB - GDT

- *RČ:*

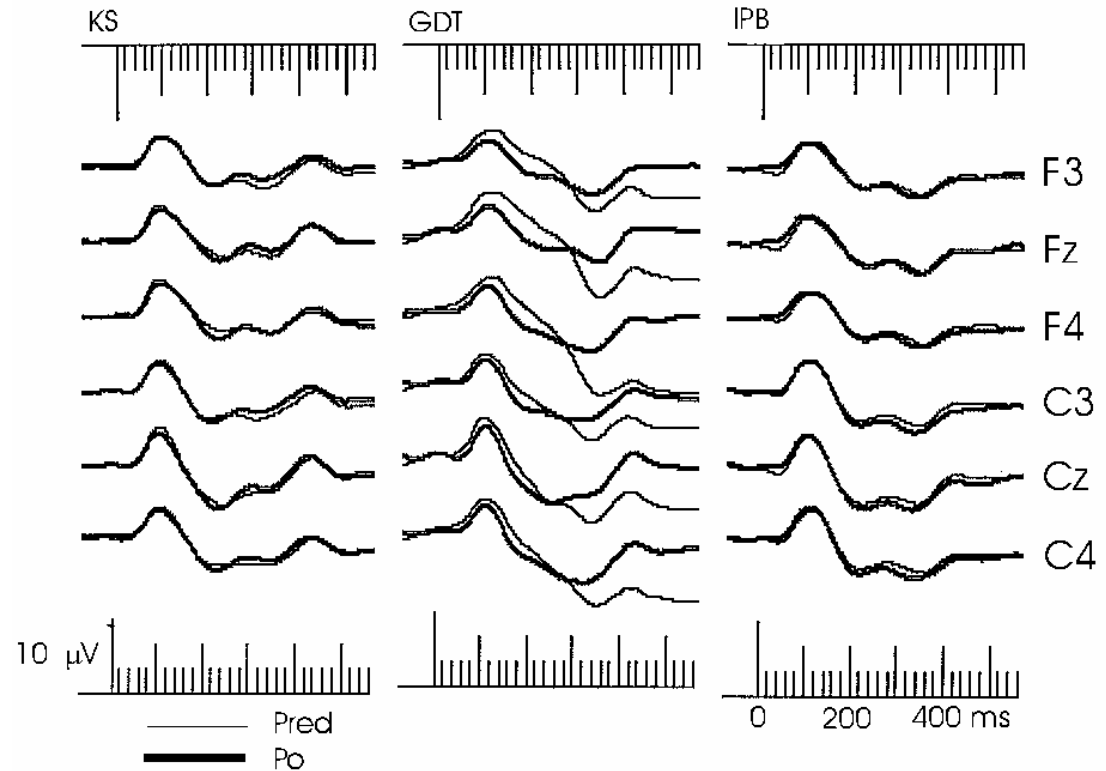
GDT daljši

IPB krajši

- *EP:*

GDT bolj "normalni"

IPB nespremenjeni





# DVA NASPROTNA POLA: PD - GDT

## Podobnosti

- IPB in GDT: moteni mehanizmi selektivne pozornosti

## Razlike

- GDT - večja pozornost nepomembnemu
- IPB - manjša pozornost pomembnemu
- GDT - perseverativna in obsesivna pozornost



# IZVID NEVROLOGA 1

WA Mozart, 34 let, desničen, glasbenik. Že od otroštva ima multiple tike, vokalizacije, koprofalijo, vrsto obsesij in kompulzij. Glasba za hipno in kar nehote 'preplavi'. Precej impulziven, v zadnjem času depresiven.

Koprofalijo ima tudi mati. V mladosti imel revmatsko vročico. V zadnjem času pogosto otečeni sklepi.

Priporočam še pregled CT, EEG, val P300, elektrofiziološko opredelitev selektivne pozornosti pri nevrofiziologu in pregled za antistreptokokna antitelesa.

Izpolnjuje klinične kriterije za sindrom GDT, ima tudi OCD in ADHD.



# IZVID KOGNITIVNEGA ELEKTROFIZIOLOGA

WA Mozart, 34 let, desničnega glasbenika pošilja nevrolog zaradi suma na sindrom GDT.

EEG je v mejah normalnega. Slušni evocirani potenciali (kohleja, deblo) v mejah normalnega.

Val P100 (skorja temporalnega režnja) normalne latence in amplitude.

Val P300 je normalno prisoten v kategoriji redkih ciljnih, se javlja tudi v kategoriji pogostih dražljajev.

Pri testiranju slušne pozornosti je 'Val Pozornost' dobro izražen, vendar med prvo ('pozornostno') in četrto ('nepozornostno') kategorijo ni razlik.

Elektrofiziološki izvid potrjuje napotno diagnozo.



# IZVID NEVROLOGA 2

Gospod Mozart izpolnjuje klinične kriterije za sindrom GDT. V zadnjem času bolj depresiven, piše Requiem.

EEG, CT v mejah normalnega. Elektrofiziološki izvid je skladen z napotno diagnozo, titer antistreptokoknih antiteles je zvišan.

V terapiji priporočam Prozac, kasneje po potrebi se antipsihotik, n.pr. risperdal in klonidin.

Kontrola čez 6 mesecev.





# IZVID NEVROLOGA 3

Gospod Mozart izpolnjuje klinične kriterije za sindrom GDT.

Po terapiji je izboljššan, tikov manj, impulzivnosti tudi, glasba ga ne 'preplavlja' več. Pomaga ženi v pri kuhinjskih opravilih, sprejel dobro plačano delo na dvoru in poplačal precej dolgov. Jemlje Prozac, antipsihotik in klonidin, čez dan še Lexaurin.

Glasba ga ne preplavlja več, Requiema ne piše več, pač pa 2x tedensko igra pri narodnozabavnem ansamblu. Prav nič pa se ni izboljšala koprofalija, gre celo za poslabšanje. Terapevtski učinek bi morda še izboljšali z injekcijo botulina v glasilke.

Kontrola čez 6 mesecev.