PAPILEDEM KOT PRVI ZNAK NETIPIČNEGA POTEKA NEVROOFTALMOLOŠKE BOLEZNI



Prikaz dveh kliničnih primerov

Mateja Repar Kokalj, dr.med., Mihaela Pavlovič, dr.med.
Mentor: prof.dr. Marko Hawlina, dr.med.

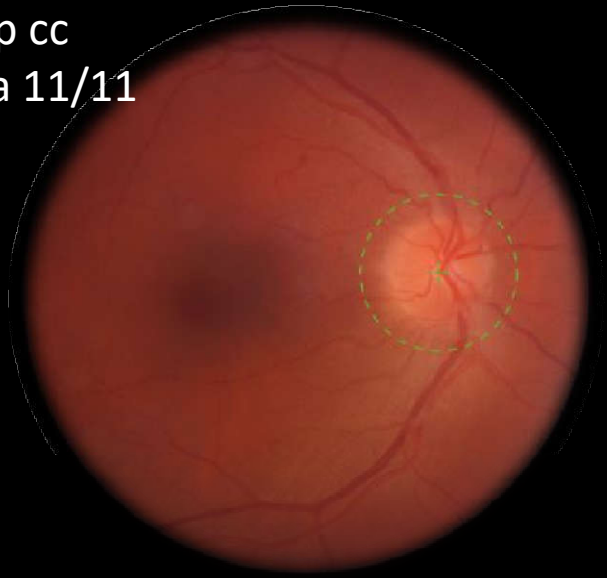
November 2018

1. primer: Ž, 63 let

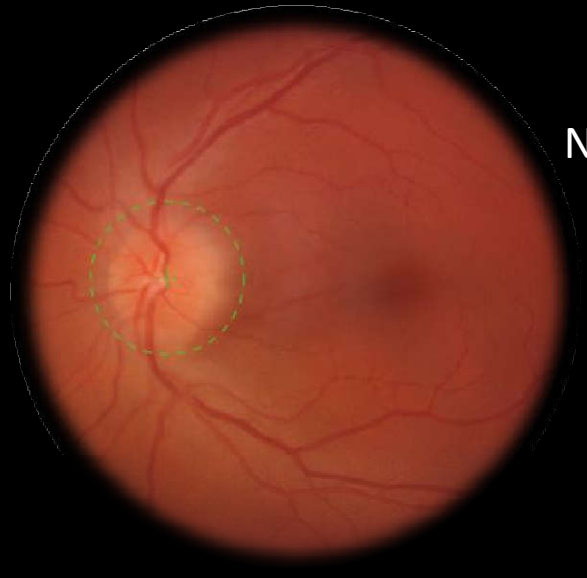
- 1 dan poslabšanje vida na LO, črna lisa v zunanjem delu vidnega polja
- Brez glavobola, dvojnih slik, bolečin v žvečnih mišicah
- Težave z zobmi, leta 2001: op. sinusov

- Vdo: +8.25:0.7p, Vlo:+7.50:0.4p
- Ishihara do 11/11, lo 8/11
- Levo nakazan RAPD
- BM DLo: leča blago kortikalno skaljena

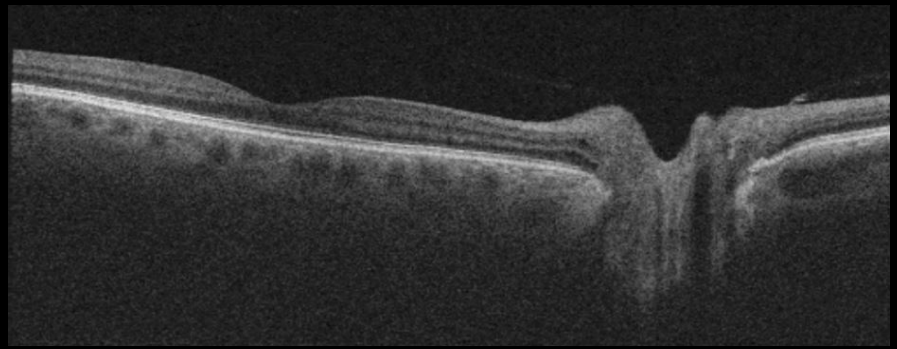
DESNO OKO
Vo: 0.7p cc
Ishihara 11/11

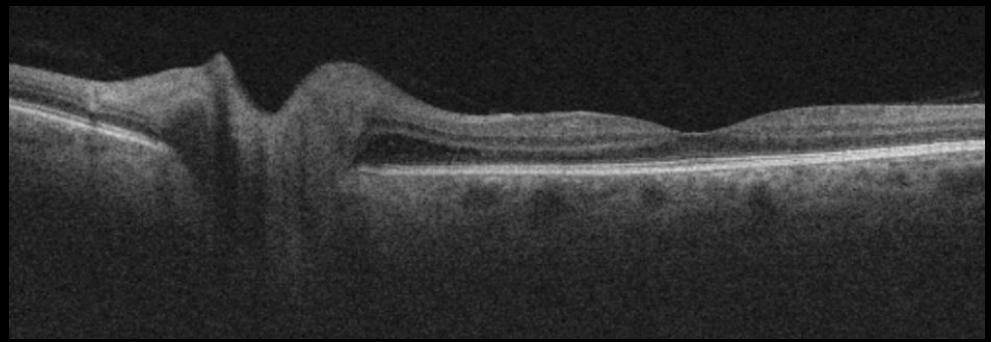


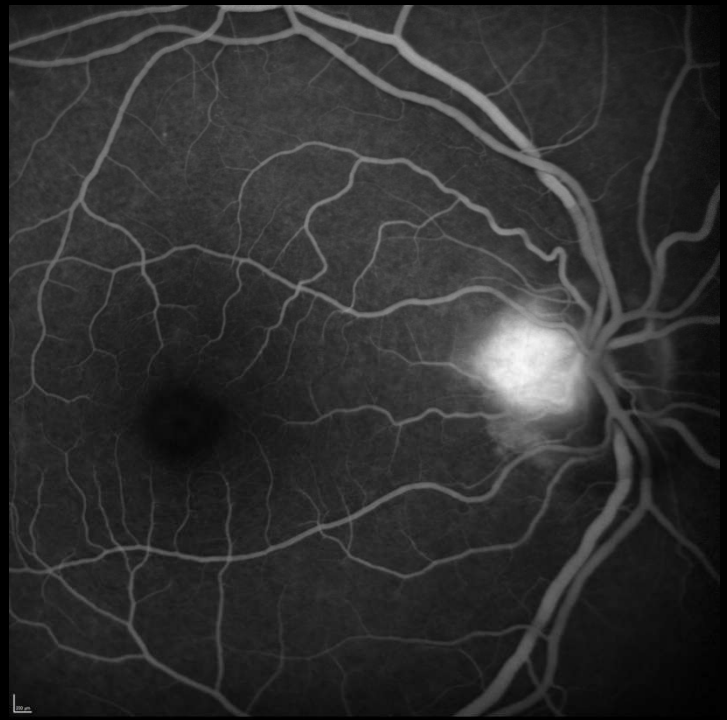
LEVO OKO
Vo: 0.4p cc
Ishihara 8/11
Nakazan RAPD



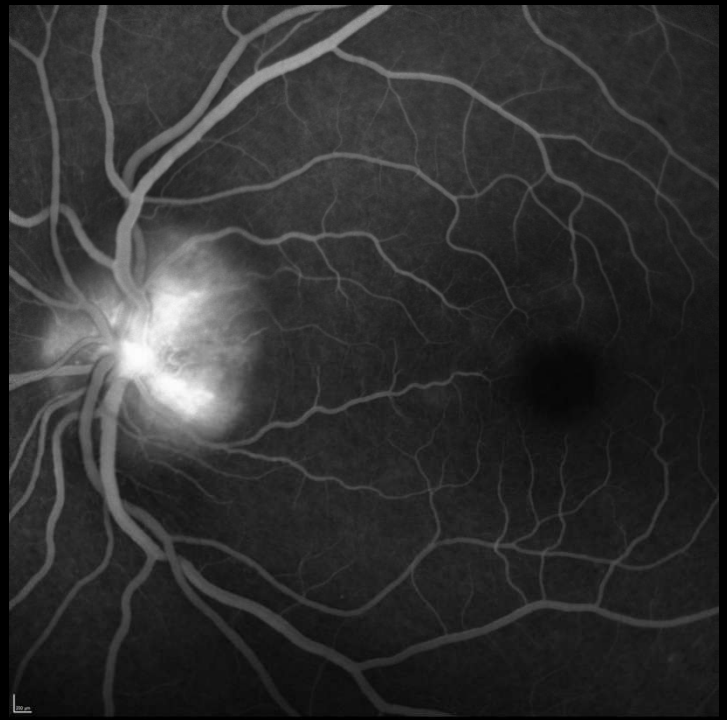
Sum na
AION levo
↓
uvedba
Aspirina







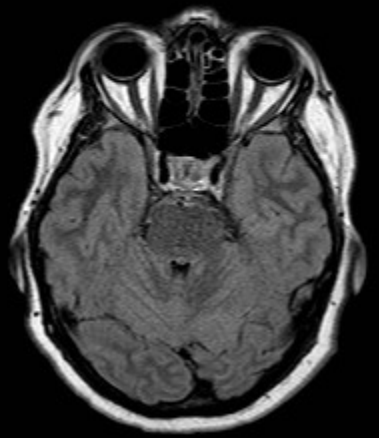
25.10.2018, OD
FA 7.14.36.30° ART(21) [HR]



25.10.2018, OS
FA 8.47.70.30° ART(13) [HR]

Preiskave

- Krvna slika bp, **CRP in SR nor**
- Serologija na borelijo, treponemo, mikoplazmo, bartonelo: neg.
- Quantiferon neg.
- Serologija na HSV, VZV, CMV: IgM neg., IgG poz.
- HEP-2 ANA, ENA, aCL, ANCA neg.
- UZ zrkel: zrkli krajši, leva papila minimalno dvignjena
- RTG pc: bp.
- CT glave: bp (orbiti in obnosne votline bp)
- MR glave: brez sprememb, ki pojasnile edem papile LO
- MRA možg. žilja: žilje prehodno, P1 segment ACP levo je aplastičen, desno hipoplastičen



Izključeni najpogostejši infektivni in avtoimuni vzroki ter kompresivne lezije CŽS.

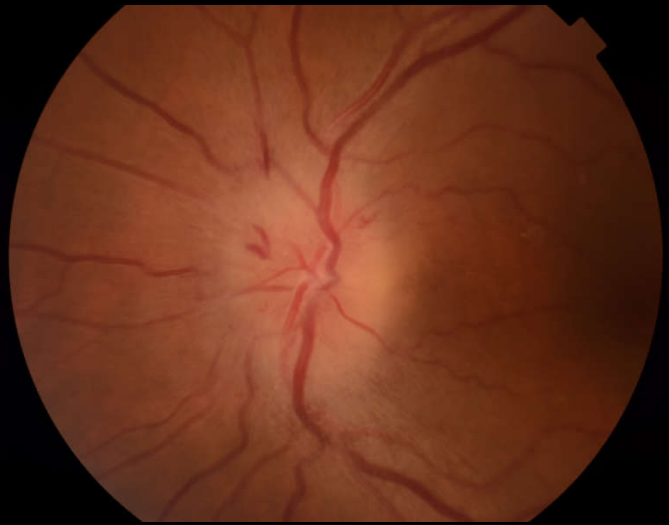
Poslabšanje klinične slike

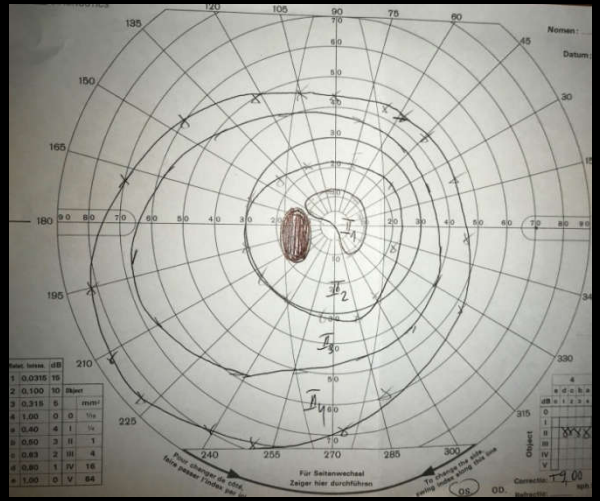
10. dan hospitalizacije

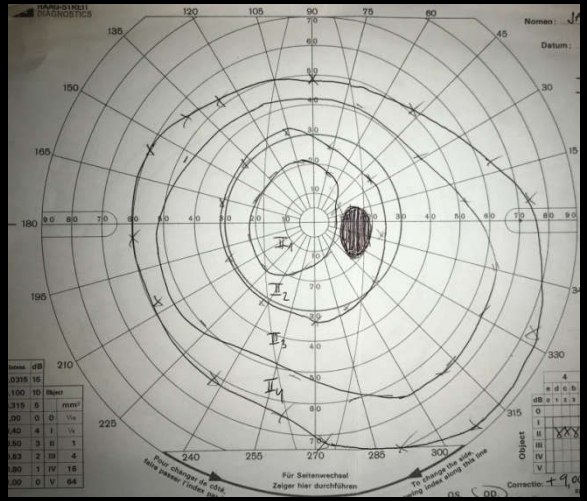
DESNO OKO
Vo: 0.6 cc
Ishihara 11/11

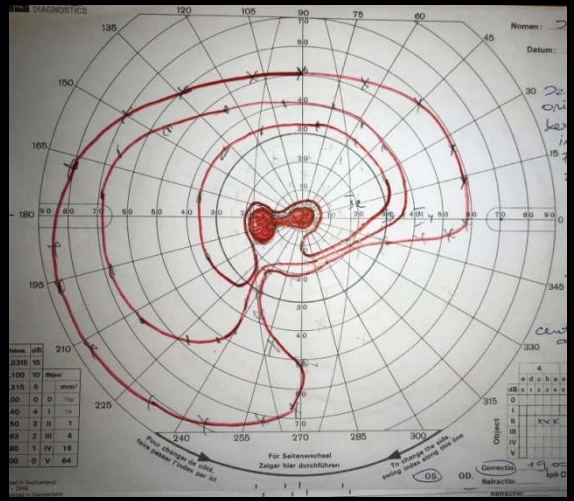


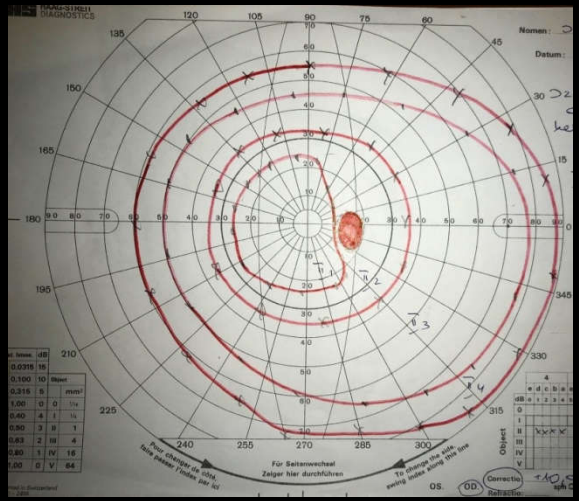
LEVO OKO
Vo: 0.2p cc
Ishihara 6/11



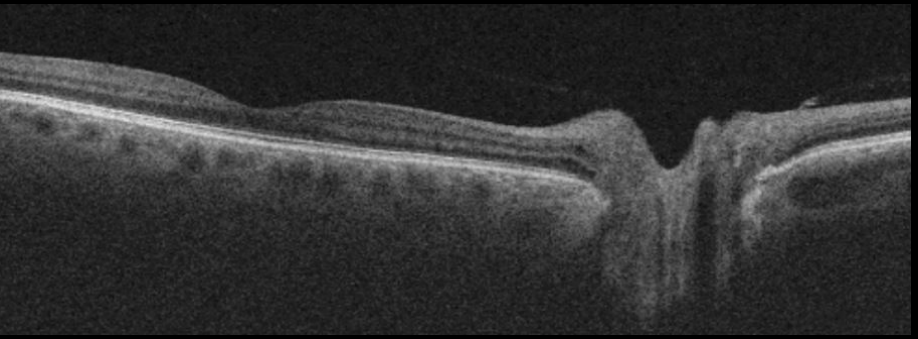

Nomen:
Datum:
Für Seitenwechsel Zeiger hier durchführen
To change the side swing index along this line
OS.
OD.

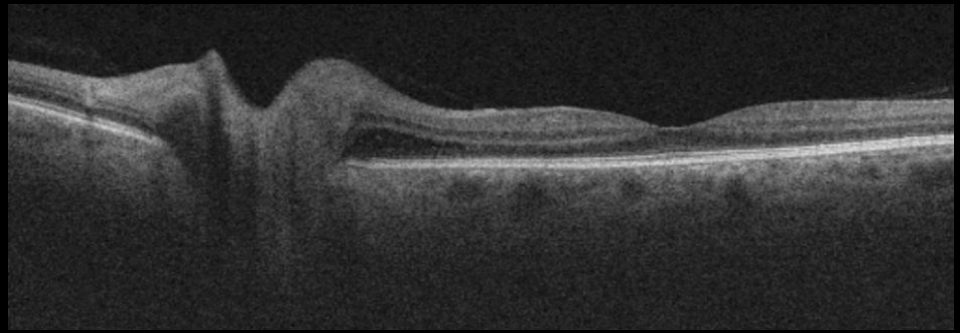

Nomen:
Datum:
Für Seitenwechsel Zeiger hier durchführen
To change the side swing index along this line
OS.
OD.


Nomen:
Datum:
Für Seitenwechsel Zeiger hier durchführen
To change the side swing index along this line
OS.
OD.

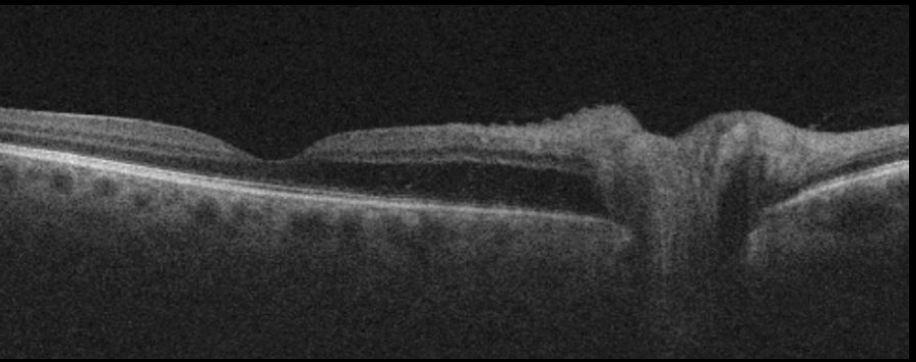

Nomen:
Datum:
Für Seitenwechsel Zeiger hier durchführen
To change the side swing index along this line
OS.
OD.

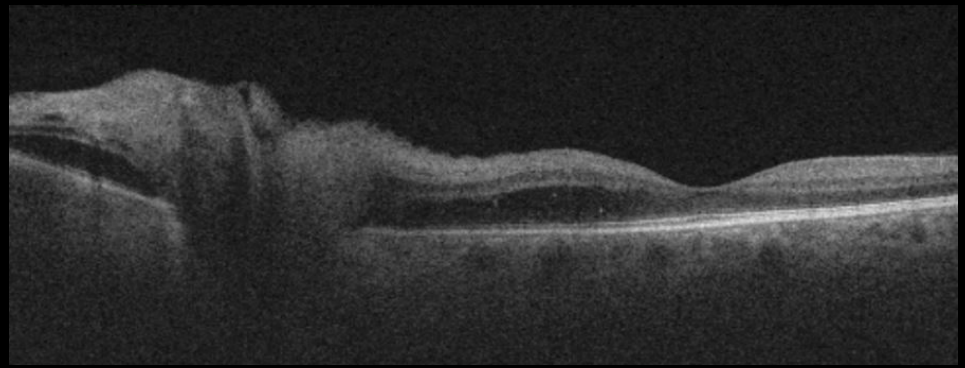
23.10.2018





5.11.2018



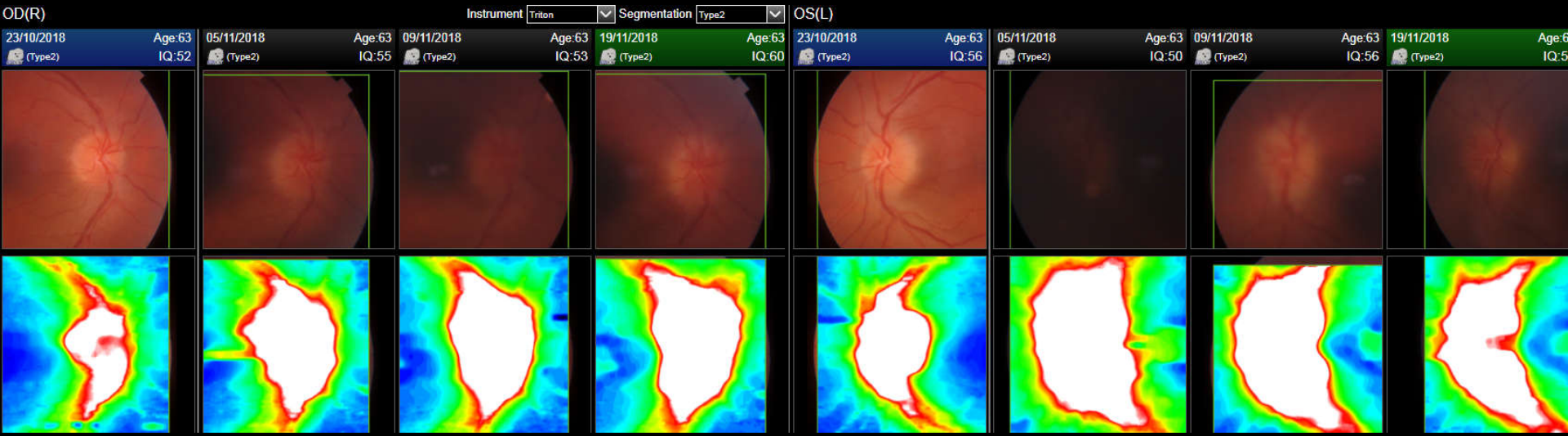


Lumbalna punkcija

1) 29.10.: nevrolog pritiska ni mogel izmeriti,
   likvor: mejno ↑proteini in levkociti

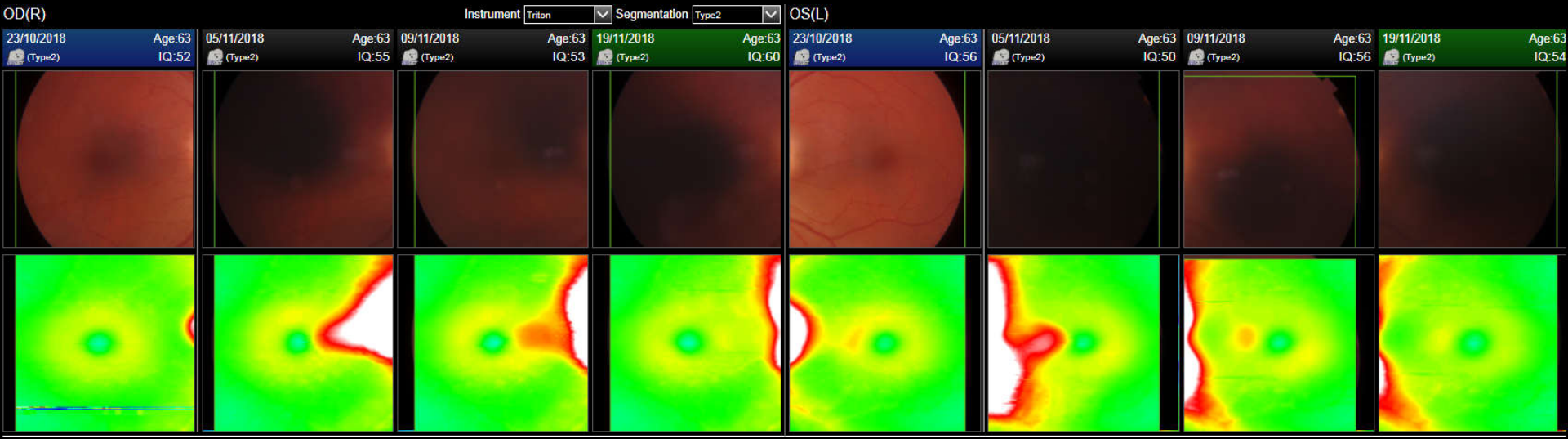
2) 5.11.: IKT = 18 cm$H_2$0
   Nevrolog: klinični potek ne govori v prid zvišanega intrakranialnega tlaka. Likvorski izvid bi lahko bil skladen z virusnim meningitisom.
   Infektolog

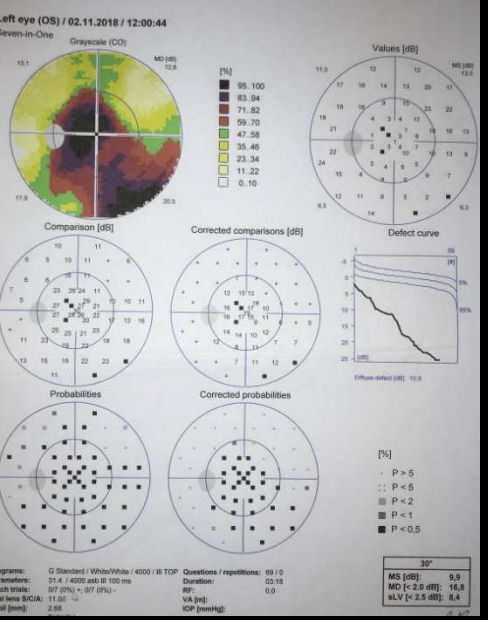
Nadaljnji potek → Edemox 250mg 2x2 tbl

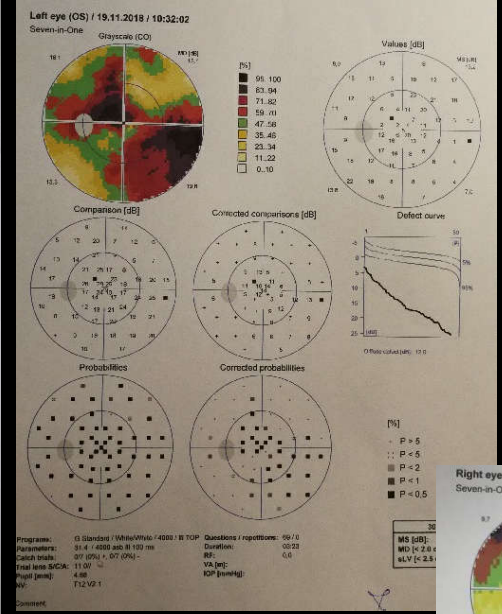


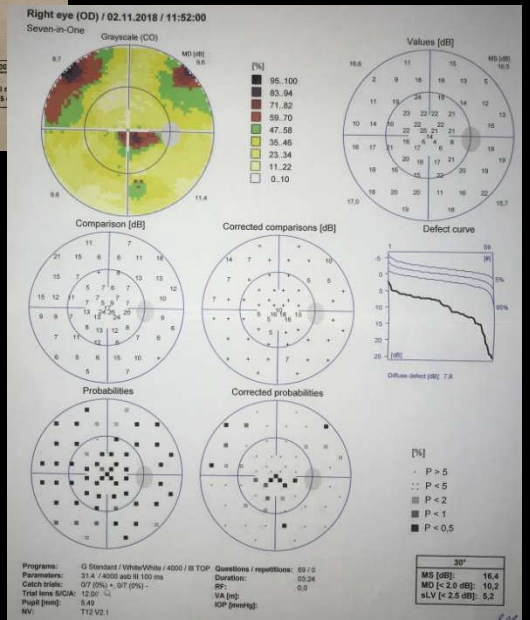
DO: Vo: 0.6p cc, Ishihara 15/15

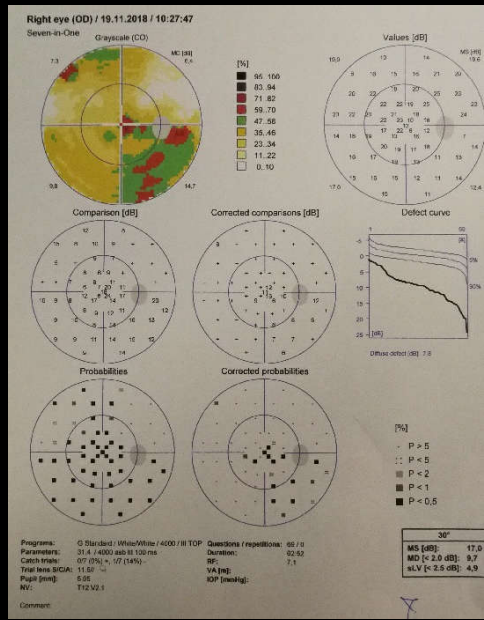
LO: Vo: 0.1 cc, Ishihara 4/15




Left eye (OS) / 02.11.2018 / 12:00:44
Seven-in-One
Grayscale (CO)
Values [dB]
Comparison [dB]
Corrected comparisons [dB]
Defect curve
Probabilities
Corrected probabilities
[%]
P > 5
P < 5
P < 2
P < 1
P < 0,5
30°
MS [dB]: 9,9
MD [< 2,0 dB]: 16,8
sLV [< 2,5 dB]: 8,4


Left eye (OS) / 19.11.2018 / 10:32:02
Seven-in-One
Grayscale (CO)
Values [dB]
Comparison [dB]
Corrected comparisons [dB]
Defect curve
Probabilities
Corrected probabilities
[%]
P > 5
P < 5
P < 2
P < 1
P < 0,5


Right eye (OD) / 02.11.2018 / 11:52:00
Seven-in-One
Grayscale (CO)
Values [dB]
Comparison [dB]
Corrected comparisons [dB]
Defect curve
Probabilities
Corrected probabilities
[%]
P > 5
P < 5
P < 2
P < 1
P < 0,5
30°
MS [dB]: 16,4
MD [< 2,0 dB]: 10,2
sLV [< 2,5 dB]: 5,2


Right eye (OD) / 19.11.2018 / 10:27:47
Seven-in-One
Grayscale (CO)
Values [dB]
Comparison [dB]
Corrected comparisons [dB]
Defect curve
Probabilities
Corrected probabilities
[%]
P > 5
P < 5
P < 2
P < 1
P < 0,5
30°
MS [dB]: 17,0
MD [< 2,0 dB]: 9,7
sLV [< 2,5 dB]: 4,9

2. primer: M, 40 let

- Pol leta poslabšanje vida na DO, meglen vid, vidna ostrina niha
- Občasno občutek pekočih oči
- Brez glavobola in bolečin v žvekalnih mišicah
- Kadilec, pogosto je imel klopa
- Mama ima glavkom

- Vdo: 1.0 sc, Vlo: 1.0 sc
- Ishihara do: 8/11, lo 10/11 (od otroštva delna okvara barvnega vida)
- RAPD –
- Spr. deli bp

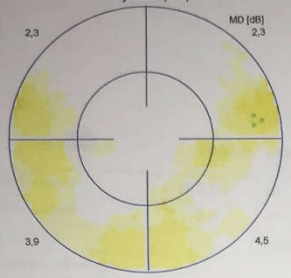




Left eye (OS) / 15.10.2018 / 14:16:35

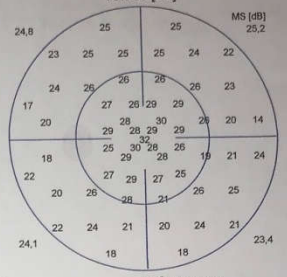
Seven-in-One

Grayscale (CO)

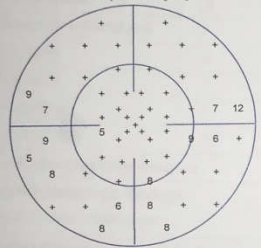


[%]
95..100
83..94
71..82
59..70
47..58
35..46
23..34
11..22
0..10

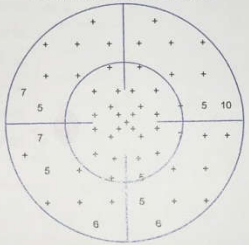
Values [dB]



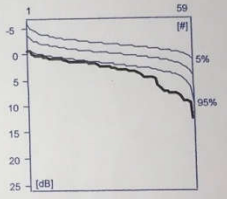
Comparison [dB]



Corrected comparisons [dB]

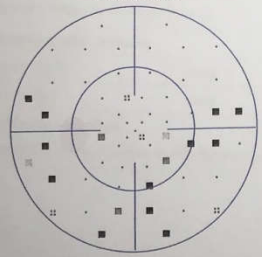


Defect curve

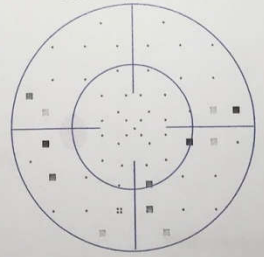


Diffuse defect [dB]: 2,3

Probabilities



Corrected probabilities



[%]
P > 5
P < 5
P < 2
P < 1
P < 0,5

30°	
MS [dB]:	24,5
MD [< 2.0 dB]:	3,2
sLV [< 2.5 dB]:	3,0

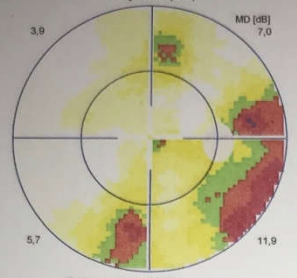
Programs:	G Standard / White/White / 4000 / III TOP	Questions / repetitions:	70 / 0
Parameters:	31.4 / 4000 asb III 100 ms	Duration:	02:12
Catch trials:	0/7 (0%) +, 0/7 (0%) -	RF:	0,0
Trial lens S/C/A:	1.5//	VA [m]:	
Pupil [mm]:	3.71	IOP [mmHg]:	
NV:	T12 V2.1		

Comment:

Right eye (OD) / 15.10.2018 / 14:13:36

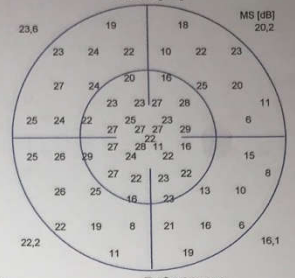
Seven-in-One

Grayscale (CO)

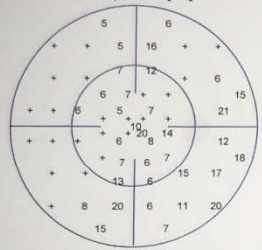


[%]
95..100
83..94
71..82
59..70
47..58
35..46
23..34
11..22
0..10

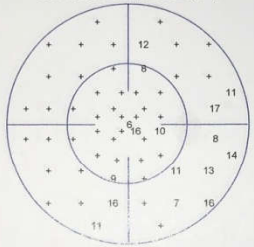
Values [dB]



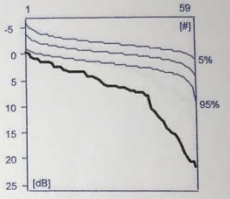
Comparison [dB]



Corrected comparisons [dB]

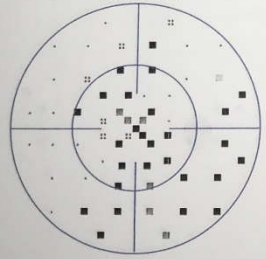


Defect curve

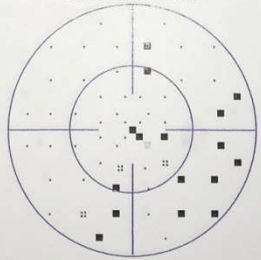


Diffuse defect [dB]: 4,0

Probabilities



Corrected probabilities



[%]
P > 5
P < 5
P < 2
P < 1
P < 0,5

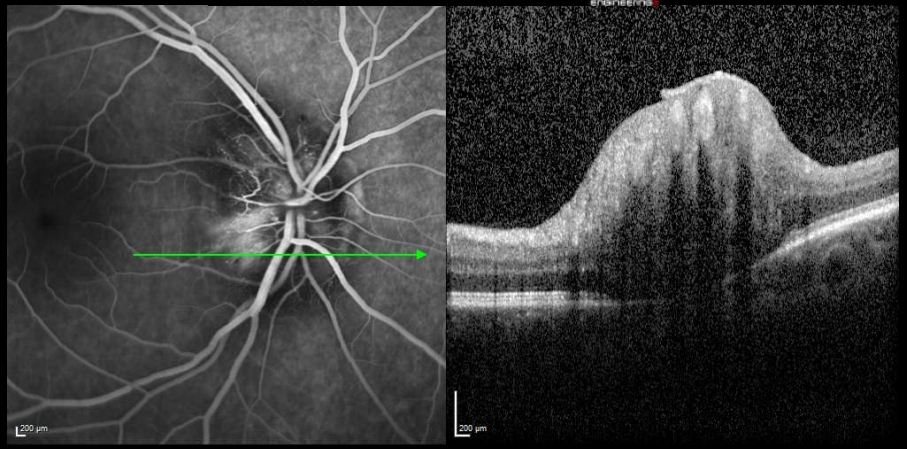
30°	
MS [dB]:	20,6
MD [< 2.0 dB]:	7,1
sLV [< 2.5 dB]:	5,8

Programs:	G Standard / White/White / 4000 / III TOP	Questions / repetitions:	69 / 0
Parameters:	31.4 / 4000 asb III 100 ms	Duration:	02:09
Catch trials:	0/7 (0%) +, 0/7 (0%) -	RF:	0,0
Trial lens S/C/A:	1.5//	VA [m]:	
Pupil [mm]:	4.22	IOP [mmHg]:	
NV:	T12 V2.1		

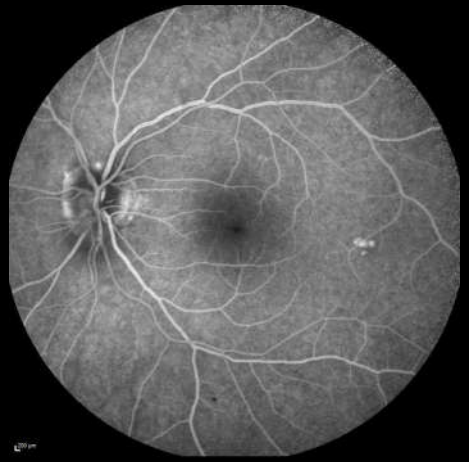
Comment:



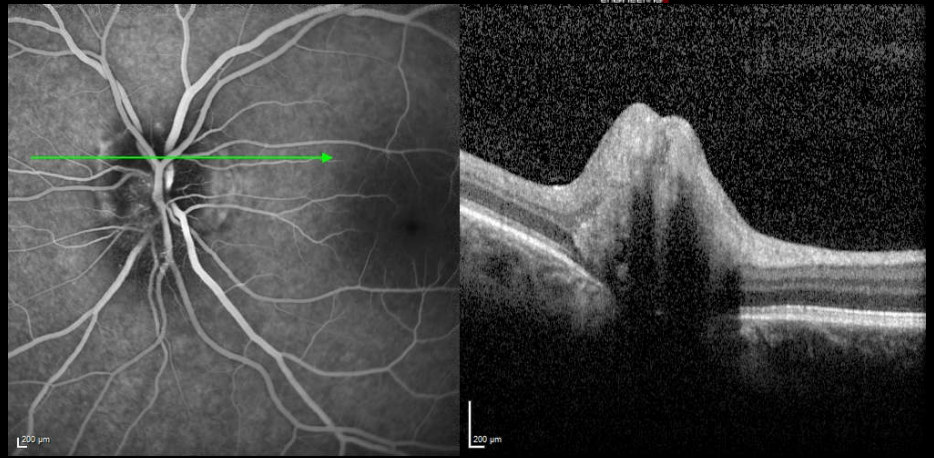
16.10.2018, OD
FA 9:04.23 55° ART(28)



16.10.2018, OD
FA&OCT 3:59.14 30° ART [HS] ART(7) Q: 22



16.10.2018, OS
FA 9:13.83 55° ART(29)



16.10.2018, OS
FA&OCT 4:52.08 30° ART [HS] ART(9) Q: 20

Preiskave



- Serologija na borelijo in treponemo: neg.
- CRP, SR nor.
- HEP-2 ANA, ENA, aCL, ANCA neg.
- Anti-MOG, aquaporin-4 protitelesa: neg.
- UZ zrkel: druze papil

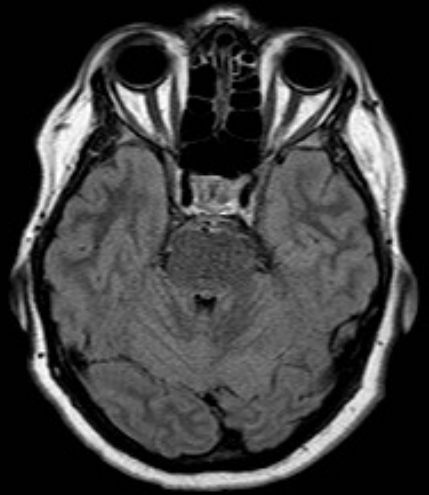
⇩

Izključeni najpogostejši infektivni in avtoimuni vzroki

- MR glave s kontrastom: likvorski prostor primerne širine, optična kjazma prosta, papili optičnih diskov nežno prominirata v zrklo

⇩

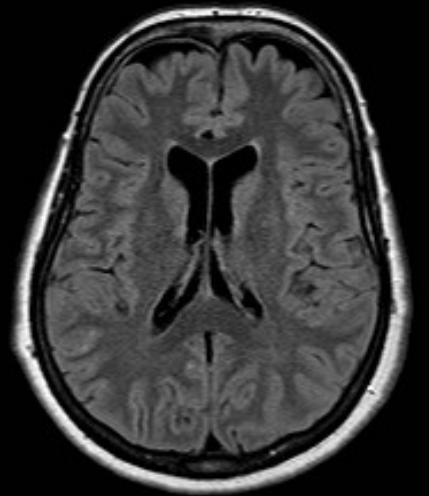
Izključene kompresivne lezije CŽS



- Nevrolog: Lumbalna punkcija: IKT = 24 cmH_20

⇩

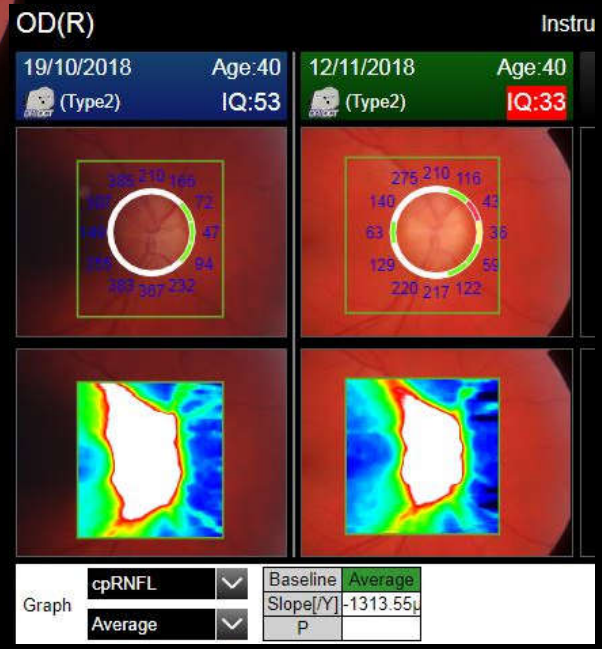
Sum na idiopatsko intrakranialno hipertenzijo



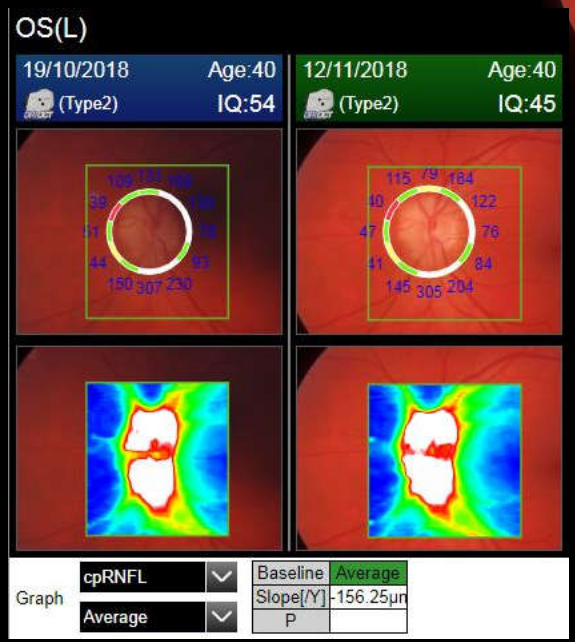
Potek bolezni po terapiji

Edemox 250mg 2x1tbl

Vdo: 1,0 sc



Vlo: 1,0 sc



Idiopatska intrakranialna hipertenzija brez glavobola

IDIOPATSKA INTRAKRANIALNA HIPERTENZIJA (IIH)

- Klinični sindrom zvišanega IKT v odsotnosti strukturnih lezij in odsotnosti hidrocefalusa
- Brez znanega vzroka
- *Benigna intrakranialna hipertenzija, pseudotumor cerebri*
- Najpogostejši vzrok papiledema
- Prevalenca: 0.9/100.000 v splošni populaciji, 19/100.000 mladih žensk s prekomerno telesno težo

Kanski's Clinical Ophthalmology: A Systematic Approach, 8th Edition, 2016

Tipični fenotip:

mlade ženske (15-44 let) s prekomerno telesno težo, zlasti nedavno pridobljeno

BMI > 30 kg/m^2



GLAVOBOL

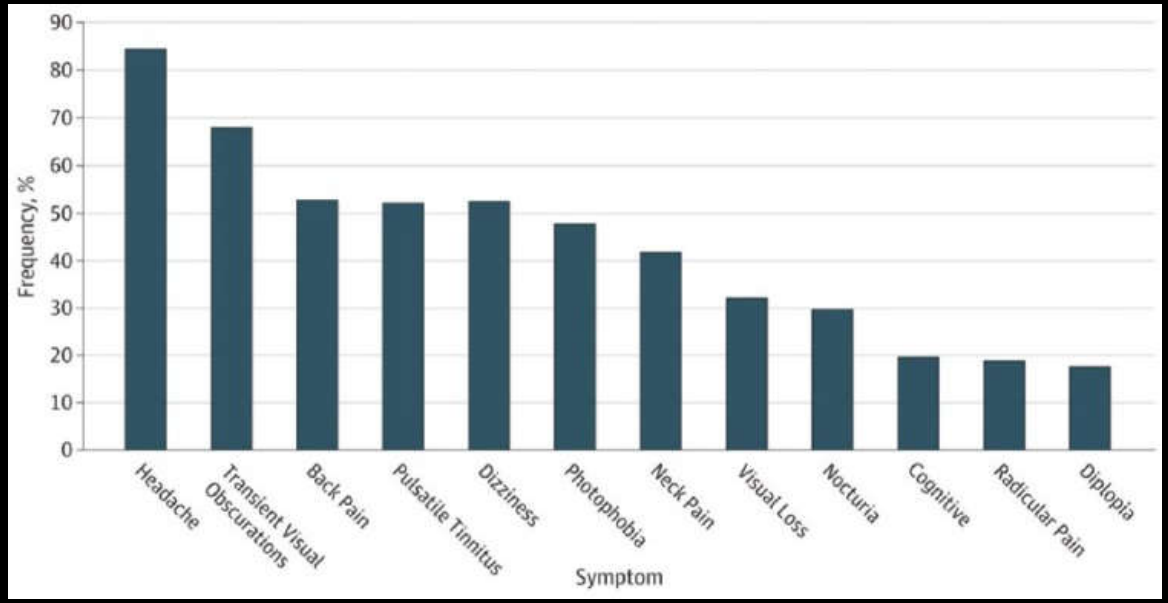


MOTNJE VIDA



Klinična slika

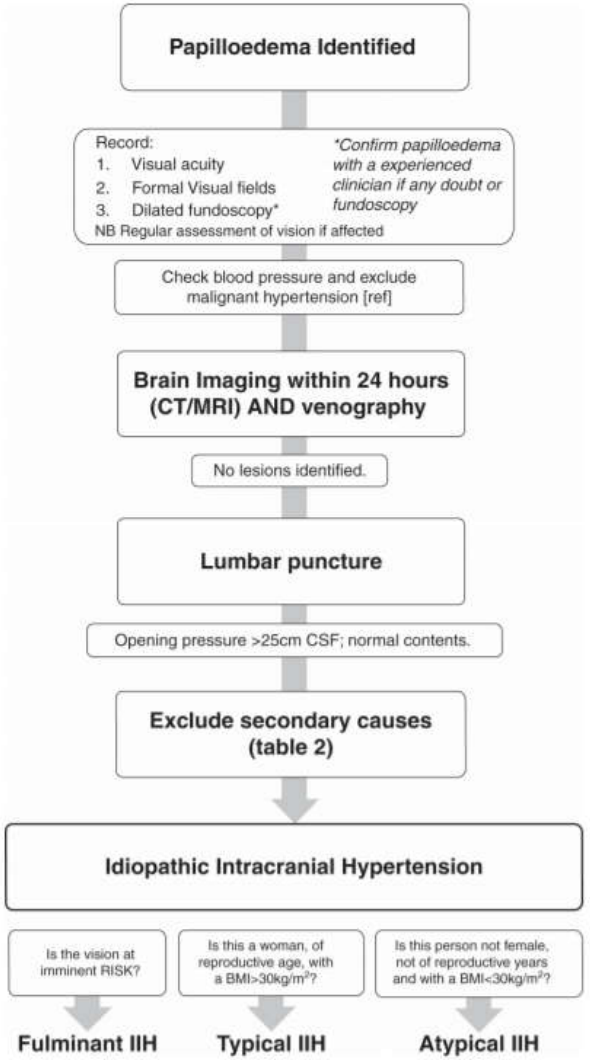
- Glavobol 76-94% (bilateralen frontalno, retrobulbarno)
- Prehodne motnje vida 68-72% (sive, temne sence)
- Pulzatilni tinitus 52-61%
- Bolečina v hrbtu 53%
- Omotica 52%
- Bolečina v vratu 42%
- Meglen vid 32%
- Kognitivne motnje 20%
- Diplopija 18 %
- ...



Percentage of patients and their presenting symptoms in Idiopathic Intracranial Hypertension, modified from Wall and colleagues [Wall *et al.* 2014a].

Diagnostični postopek

- Izključitvena diagnoza



Mollan SP, et al. Idiopathic intracranial hypertension: consensus guidelines on management, J Neurol Neurosurg Psychiatry 2018;89:1088–1100.

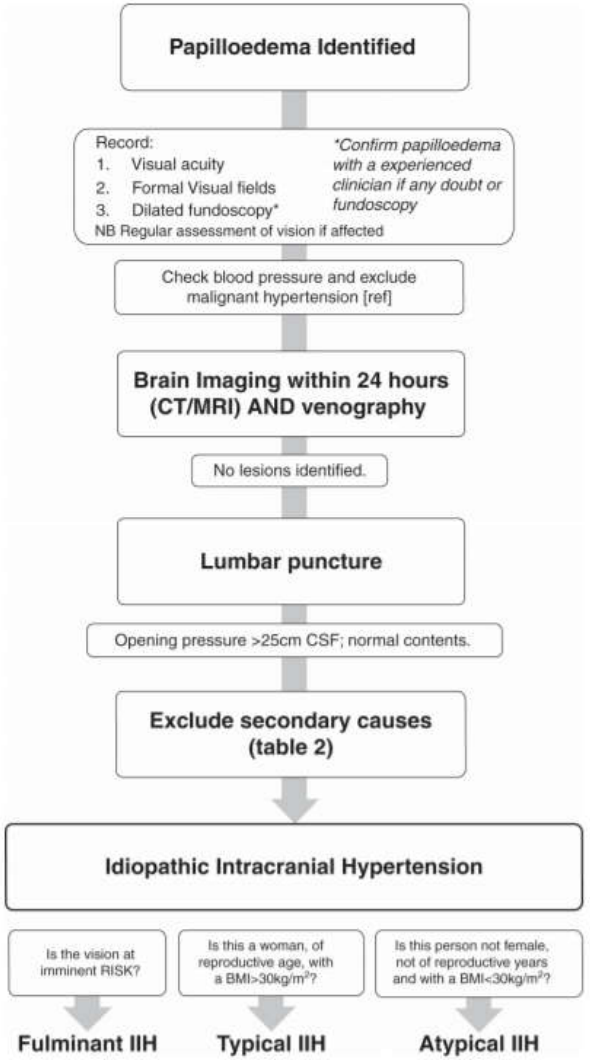
Diagnostični postopek

Papilloedema Identified

Record:
1. Visual acuity
2. Formal Visual fields
3. Dilated fundoscopy*

Confirm papilloedema with a experienced clinician if any doubt or fundoscopy

NB Regular assessment of vision if affected



Check blood pressure and exclude malignant hypertension [ref]



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Brain Imaging within 24 hours (CT/MRI) AND venography

No lesions identified.

Lumbar puncture

Opening pressure >25cm CSF; normal contents.

Exclude secondary causes (table 2)

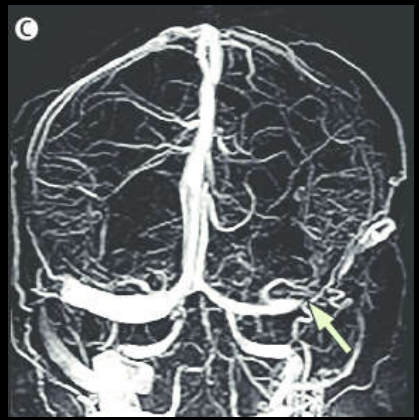
Idiopathic Intracranial Hypertension

Is the vision at imminent RISK?	Is this a woman, of reproductive age, with a BMI>30kg/m^2?	Is this person not female, not of reproductive years and with a BMI<30kg/m^2?
Fulminant IIH	**Typical IIH**	**Atypical IIH**

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No lesions identified.





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NAJDBE NA MR PRI IIH:

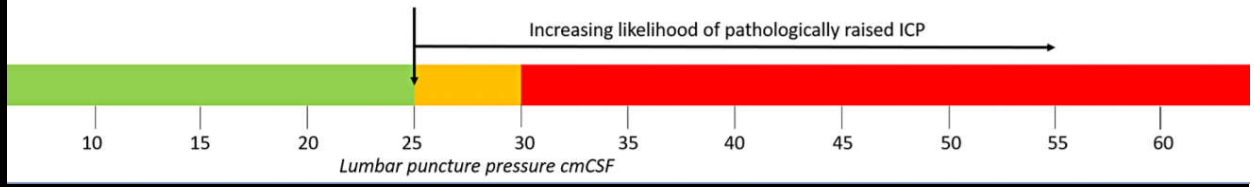
- Tortuozni potek vidnega živca in distenzija ovojnice vidnega živca (45%)
- Izravnanje zadnjega dela zrkla (80%)
- Protruzija papile vidnega živca (30%)
- Delna empty sella (70%)
- Stenoza transverzalnih venskih sinusov



Markey KA, et al. Understanding idiopathic intracranial hypertension: mechanisms, management, and future directions, Lancet Neurol 2016; 15: 78–91.



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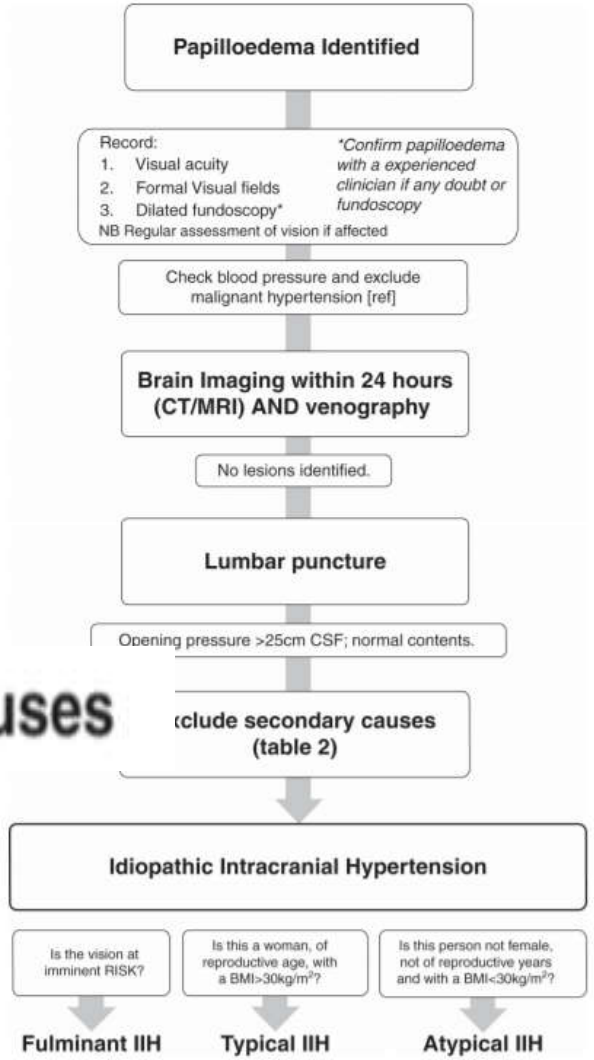
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Fulminant IIH	**Typical IIH**	**Atypical IIH**

- Tromboza venskih sinusov, obstrukcija venske drenaže
- Anemija
- Sindrom obstrukcijske apneje v spanju (OSA)
- Ledvična odpoved
- Endokrinološki vzroki: debelost, pridobitev telesne teže, PCOS, hipoparatitroidizem, Cushingova bolezen, Addisonova bolezen
- SLE
- Hipervitaminoza A
- ZDRAVILA (tetraciklini, vitamin A in retinoidi, rastni hormon, tiroksin, ukinitev kroničnega jemanja kortikosteroidov,…)

Exclude secondary causes



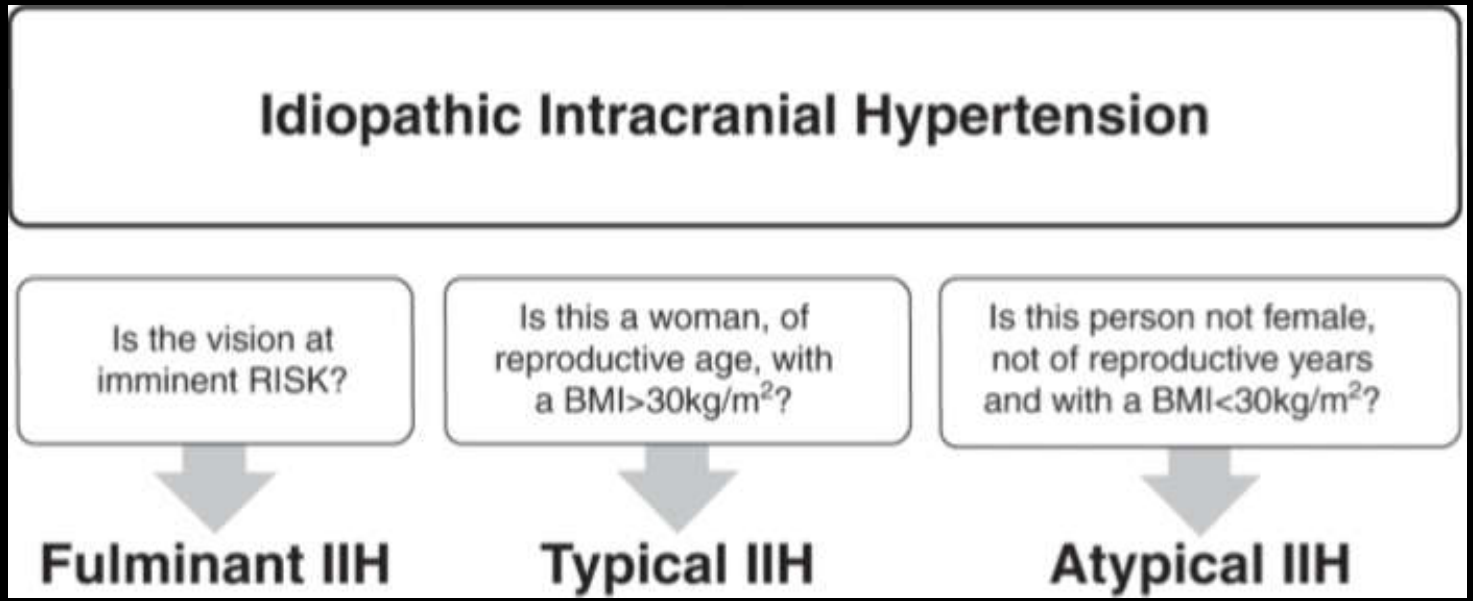
Consensus in diagnosing IIH

IIH diagnostic criteria, adapted from Friedman et al.

IIH Diagnostic criteria

A. Papilloedema
B. Normal neurological examination (except sixth cranial nerve palsy)
C. Neuroimaging: normal brain parenchyma (no hydrocephalus, mass, structural lesion or meningeal enhancement). Venous thrombosis excluded in all.
D. Normal CSF constituents
E. Elevated lumbar puncture pressure ≥ 25cmCSF

Susan P Mollan et al. J Neurol Neurosurg Psychiatry 2018;89:1088-1100


Idiopathic Intracranial Hypertension
Is the vision at imminent RISK?
Is this a woman, of reproductive age, with a BMI>30kg/m2?
Is this person not female, not of reproductive years and with a BMI<30kg/m2?
Fulminant IIH
Typical IIH
Atypical IIH

Potek

- Večina ima dobro prognozo (opazovanje, zdravila).
- Kronični glavoboli ⇨ slabša kvaliteta življenja.
- IZGUBA VIDA!
- Do 30% bo imelo ireverzibilno okvaro vida.
- Pomembno redno sledenje.
- Lahko pride do relapsa (nepredvidljiv, pridobitev na TT).

Zdravljenje

- Hujšanje
- Acetazolamid: **2x 250/500 mg** (max. odmerek 2g/dan)
- Analgetiki - Topiramat
- Kirurški posegi: fenestracija ovojnice optičnega živca, ventrikulo-peritonealni obvod, stentiranje stenoze transverzalnega sinusa

Mollan SP, et al. Idiopathic intracranial hypertension: consensus guidelines on management, J Neurol Neurosurg Psychiatry 2018;89:1088–1100.

IIH brez glavobola

- Glavobol je najpogostejši simptom pri IIH, pojavlja se v 90% primerov.
- V 10% (9-38%) primerov gre lahko za IIH brez glavobola.

- PRI MOŠKIH
- PRI OTROCIH
- VEČ NEVROLOŠKIH SIMPTOMOV
- HUJŠA IZGUBA VIDA IN IZPADI V VIDNEM POLJU OB PRVEM PREGLEDU
- SLABŠA PROGNOZA GLEDE VIDA

⇨ pogosto spremljanje in bolj agresivna terapija.

Aylward SC, Aronowitz C, Reem R, Rogers D, Roach ES. Intracranial hypertension without headache in children. J Child Neurol 2015;30(6):703–6.

Razlike med MOŠKIMI in ŽENSKAMI z IIH

	MOŠKI	ŽENSKE
GLAVOBOL	55%	75%
VIDNI SIMPTOMI	35%	20%
TINITUS	26%	38%
VIDNA FUNKCIJA	slabša	boljša

- Glavobol kot simptom IIH je manj pogost pri moških (bolečinski prag, hormonski, etiološki vzroki).
- Pri moških so pogosteje prisotni hujši vidni simptomi.
- Tveganje za trajno izgubo vida je večje.

Bruce, et al. Idiopathic intracranial hypertension in men. Neurology 2009;72(4):304–9. (721 bolnikov, 66 moških (9%), 655 žensk (91%))

IIH PRI OTROCIH

- Enaki diagnostični kriteriji, le tlak > 28 cm H20
- Pogosto vidni simptomi in netipični znaki zvišanega IKT
- Pogosto brez glavobola

OTROCI	Z GLAVOBOLOM	BREZ GLAVOBOLA
POVPREČNA STAROST V LETIH	9,5	7,3
NEVROLOŠKI ZNAKI	10%	33%
OKVARA VIDA	4%	33%
IZPADI V VIDNEM POLJU	5%	50%
TRAJNA OKVARA VIDA	3%	33%

Lim M, Kurian M, Penn A, Calver D, Lin JP. Visual failure without headache in idiopathic intracranial hypertension. Arch Dis Child 2005;90(2):206–10. (41 otrok z IIH, 29 z glavobolom (71%), 12 brez glavobola (29%))

Zaključek

- Glavobol in papiledem sta prisotna v večini primerov IIH, poznamo pa primere IIH brez papiledema in IIH brez glavobola.
- IIH se lahko pojavi pri moških, osebah z nizkim BMI in pri ženskah izven rodne dobe (atipična oblika).
- IIH brez glavobola je redka, pogostejša pri moških, otrocih, mlajših in osebah z nizkim BMI.
- Pri osebah z vidnimi simptomi brez glavobola je pomembno agresivno zdravljenje in spremljanje, saj pogosteje pride do trajnih okvar vida.
- Vloga oftalmologa pri IIH: diagnostika, zdravljenje, monitoriranje vidne funkcije (ostrina, barvni vid, vidno polje), izgled vidnega živca.