

ENERGETSKA IN BELJAKOVINSKA PODHRANJENOST GIBALNO OVIRANEGA OTROKA/MLADOSTNIKA S CEREBRALNO PARALIZO ALI PROGRESIVNO POTEKAJOČO ŽIVČNO-MIŠIČNO BOLEZNIJO: POSLEDICE TER TERAPEVTSKI PRISTOPI

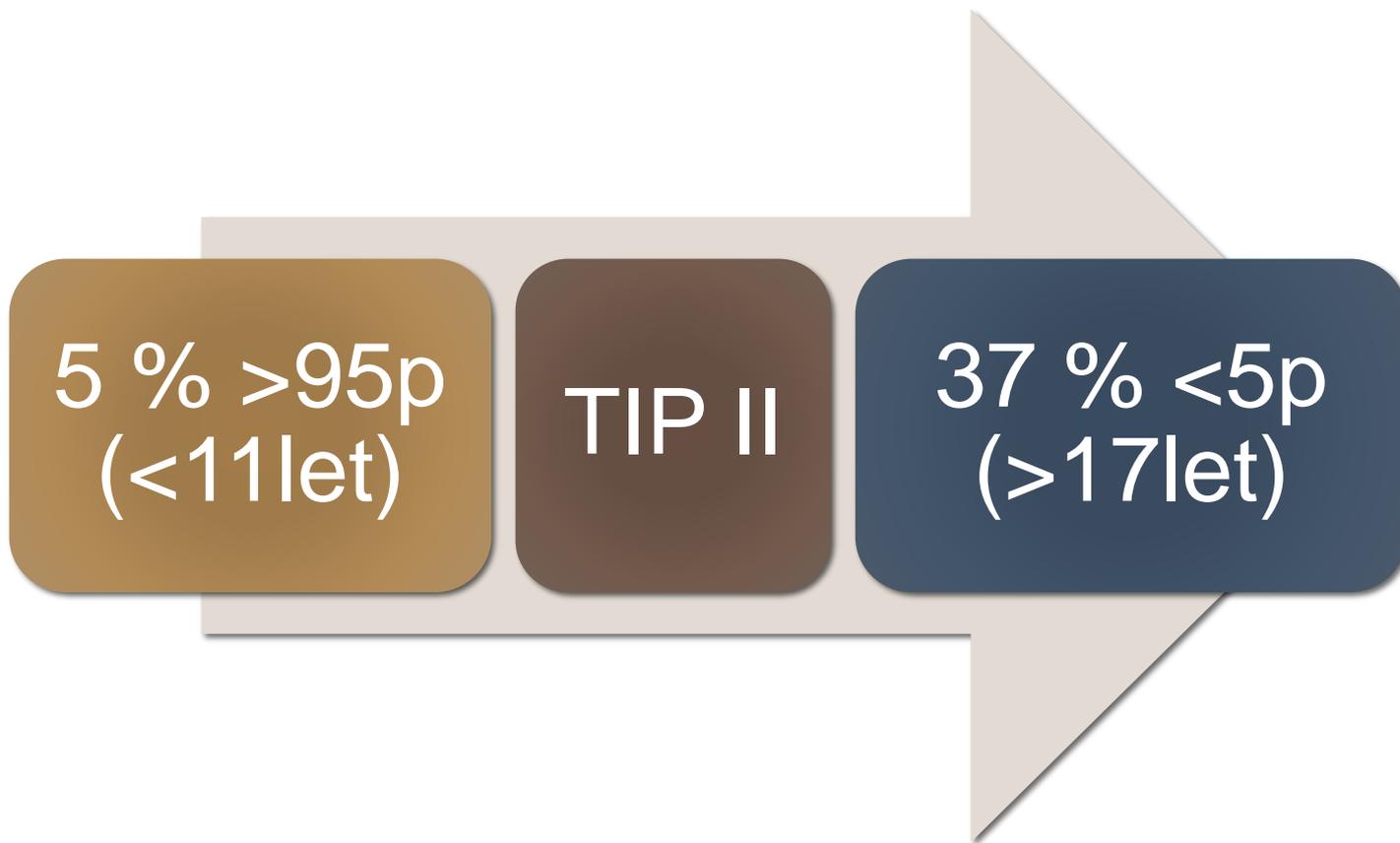
izr prof Matjaž Homan

ALI JE PODHRANJENOST OTROK S CP VELIK PROBLEM?

RAZISKAVA	GMCSF	NIZKA RAST	NIZKA TM	SESTAVA T
Samson-Fang 2002	3-5 (2-18LET)	68%	47%	47%
Stallings 1993	1-2 (2-18LET)	23%	23%	28%
Benfer 2017	1-5 (2-5LET)	11%	11%	13%

TM pri otrocih s SMA^{MESSINA,2008.}

- ITM različen glede na tip SMA in starost otroka



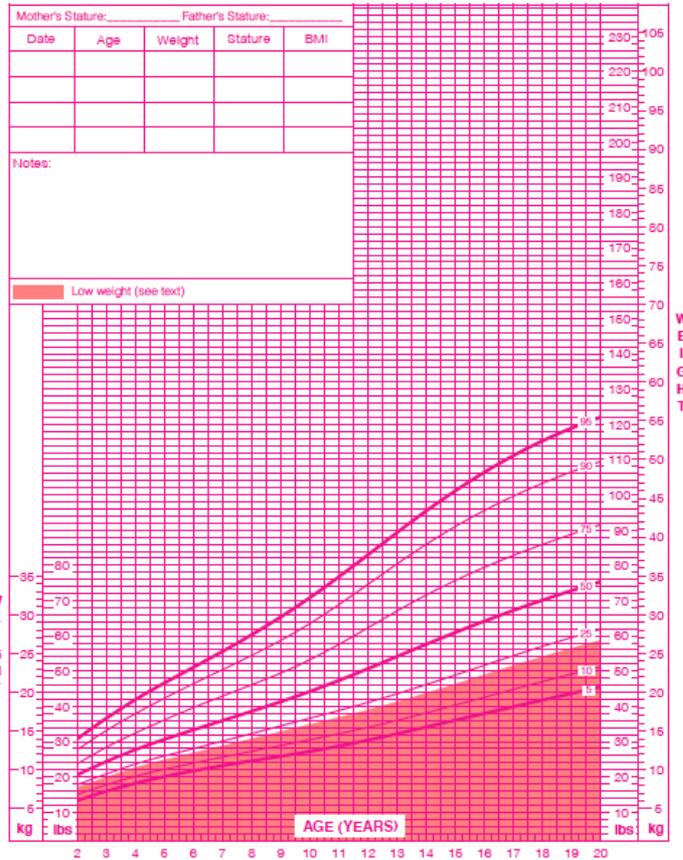
POMEN POSEBNIH RASTNIH KRIVULJ ZA OTROKE Z CP

- definirati maso in višino, sestavo telesa “zdravih” otrok z CP je težko
- napredovanje na TM in TV pri otrocih z CP zavisi predvsem od stopnje CP
- krivulje za TM, TV, ITM po starosti od 2 let naprej za otroke z CP glede na stopnjo CP (1-5)^{BROOKS, 2011.}
- rastne krivulje za otroke s CP iz Evrope^{ERGENOLF, 2019.}

<http://www.LifeExpectancy.org/Articles/NewGrowthCharts.shtml>

2 to 20 years: Girls
Cerebral palsy
GMFCS V, feeds orally
Weight-for-age percentiles

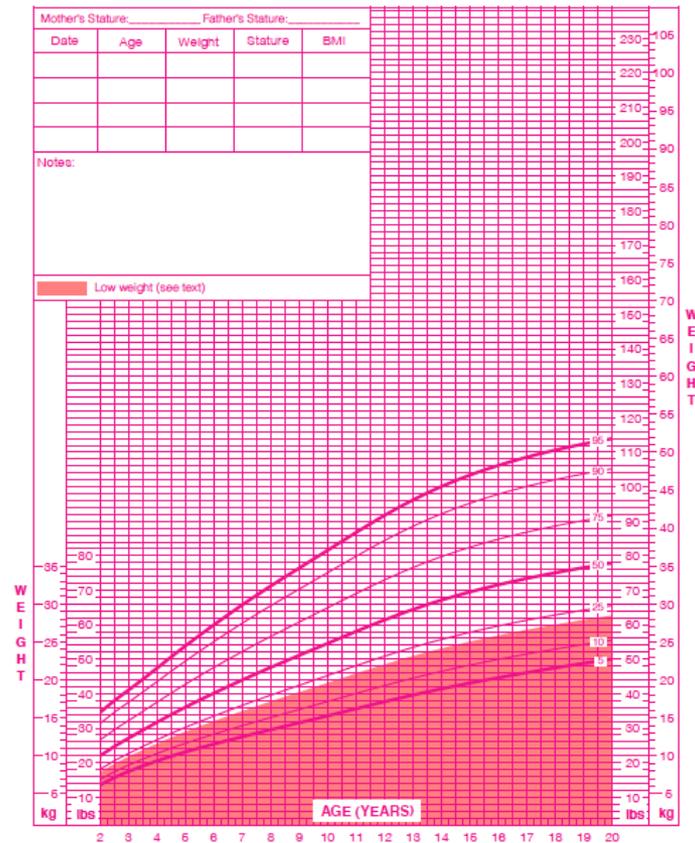
NAME _____
RECORD # _____



SOURCE: Life Expectancy Project (2011)
Based on data from the California Department of Developmental Services and California Bureau of Vital Statistics
<http://www.LifeExpectancy.org/Articles/NewGrowthCharts.shtml>

2 to 20 years: Girls
Cerebral palsy
GMFCS V, tube fed
Weight-for-age percentiles

NAME _____
RECORD # _____



SOURCE: Life Expectancy Project (2011)
Based on data from the California Department of Developmental Services and California Bureau of Vital Statistics
<http://www.LifeExpectancy.org/Articles/NewGrowthCharts.shtml>

POMEN TM IN TV PRI OTROCIH Z CP

- **Uporabo posebnih krivulj ne priporoča CDC, niti ESPGHAN**

DMD- percentilne krivulje

- Manj puste mišične mase
- Več maščobnega tkiva

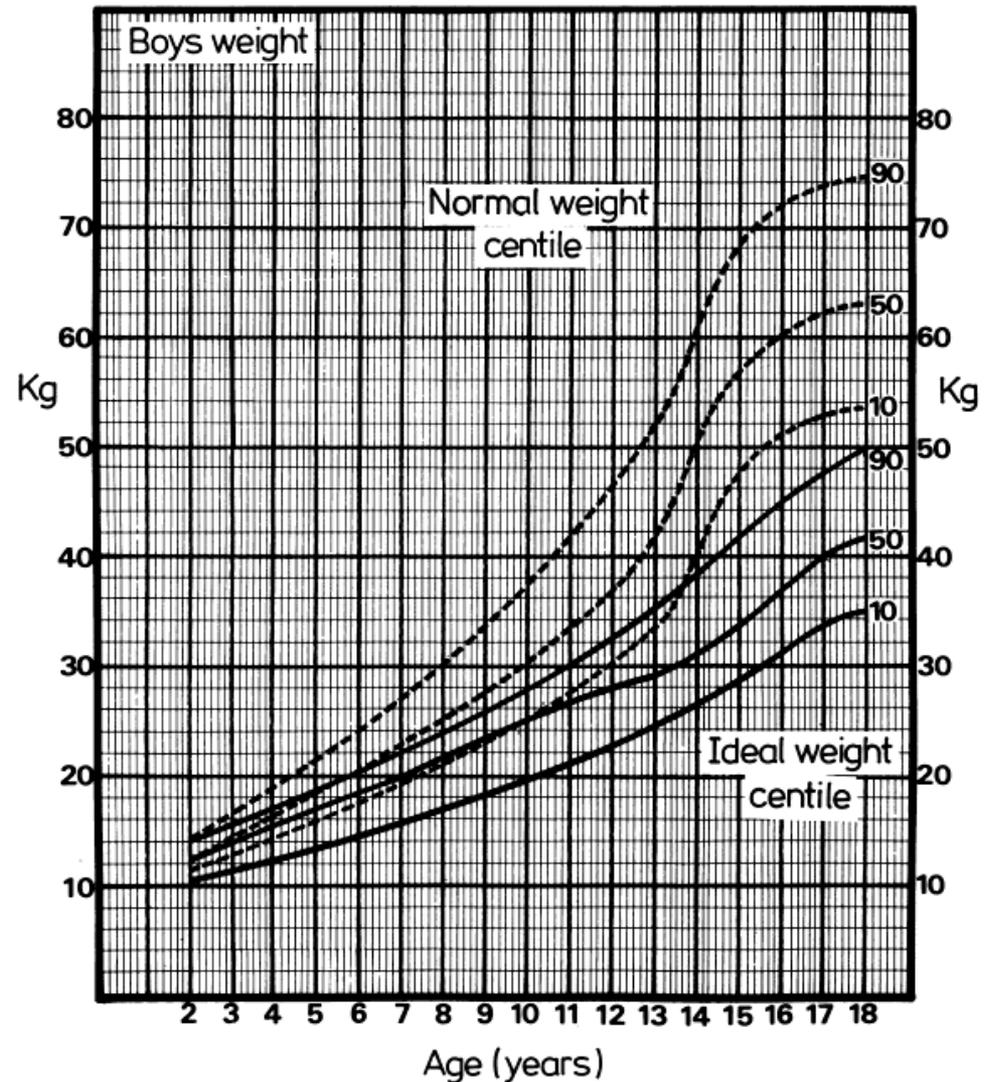


Figure. Ideal weight centile chart for boys with Duchenne muscular dystrophy. Based on data from Edwards et al,¹ Edmonds et al,⁴ and Tanner and Whitehouse.³ Assumes a 4% per year decline in muscle bulk.

KAKO POTEM PREPOZNAMO PODHRANJENOST?



VZROKI PODHRANJENOSTI

PREHRANSKI

premajhen vnos

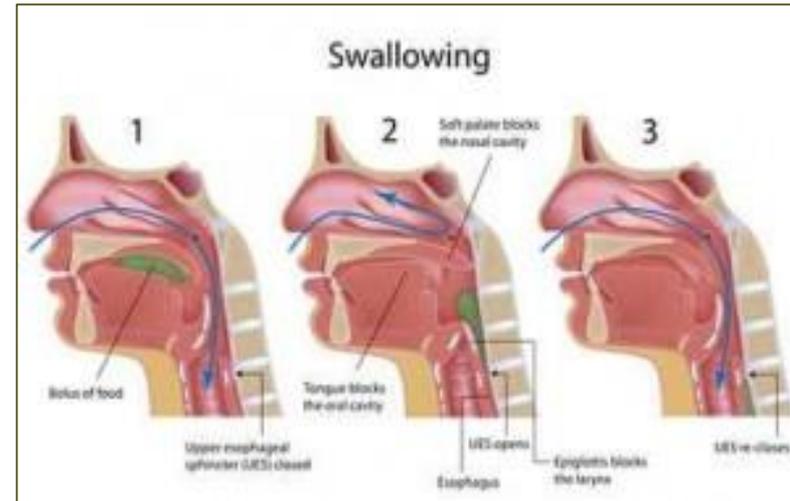
povečane izgube

povečane potrebe

OSTALI VZROKI

PREMAJHEN VNOS- motnje požiranja

- 1) **ORALNA PRIPRAVLJALNA FAZA** (hoteni nadzor): vnos hrane v usta in tvorba bolusa.
- 2) **ORALNA FAZA** (hoteni nadzor): transport tekočin ali bolusa hrane posteriorno skozi ustno votlino, začetek faringealnega požiranja.
- 3) **FARINGEALNA FAZA** (hoteni + nehoteni nadzor): začetek akta požiranja, inhibicija dihanja.
- 4) **EZOFAGEALNA FAZA** (nehoteni nadzor): transport bolusa skozi zg. požiralnikov sfinkter proti spodnjemu in nato v želodec.



Kompleksen nadzor živčnega sistema

(možganski živci V, VII, IX, X, možgansko deblo, suprabulbarni centri, možganski korteks) – dozoreva med razvojem.

PREMAJHEN VNOS- motnje požiranja

1) orofaringealna disfunkcija

motnja v nastanku bolusa hrane, sekrecija sline, inkoordinacija požiranja in dihanja, penetracija in aspiracija (80% tekočine, 40% čvrsta hrana)^{SULLIVAN,2008.}

2) disfunkcija požiralnika

spazm ZPS

retrogradna peristaltika

Sewell et al. BMJ 2014



PREMAJHEN VNOS- motnje požiranja

- **prevalenca (90-95%)**^{CALIS,2008.}
- 89% potrebuje pomoč pri hranjenju, 56% zaletavanje, 28% >3 ure trajajo obroki na dan, 22% bruhanje^{SULLIVAN,2000}
- Dejavniki tveganja: stopnja nevrološke okvare, pridružene strukturne anomalije nazofarinksa (sindromi), pridružen GERB.

Oropharyngeal Dysfunction

should be **considered in all NI patients** (especially with more significant gross motor problems) even in the absence of obvious clinical signs and symptoms

'Tiha' aspiracija^{ANDREW,2012.}

Table 1 Clinical symptoms and signs of aspiration

Clinical symptoms and signs of aspiration

Blinking, eye watering

Throat clearing, grimacing, head tilting

Difficulty handling oral secretions

Poor oral motor co-ordination

Limited endurance/increasing fatigue during feeding

Delayed swallow reflex

Multiple swallows to clear single bolus

Wet voice quality during or after feeding

Increased congestion during feeds

Noisy/wet upper airway sounds associated with feeding

Coughing or choking during/after feeds

Change in breathing rhythm or apnoea

Recurrent wheeze

Recurrent respiratory infection

DIAGNOSTIKA MOTENJ POŽIRANJA



- heteroanamneza in status (ORL področje...)
- opazovanje hranjenja
- **VFSS (videofluoroscopska ocena požiranja)**
- **NAZOLARINGOSKOPIJA**
- kontrastno slikanje prebavil
- EGDS z biopsijami
- Ph-impedanca



Source: Adapted from: Emergency Medicine Procedures, 2nd ed. Baltimore: www.lww.com/9780781768101
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MULTIDISCIPLINARNA OBRAVNAVA

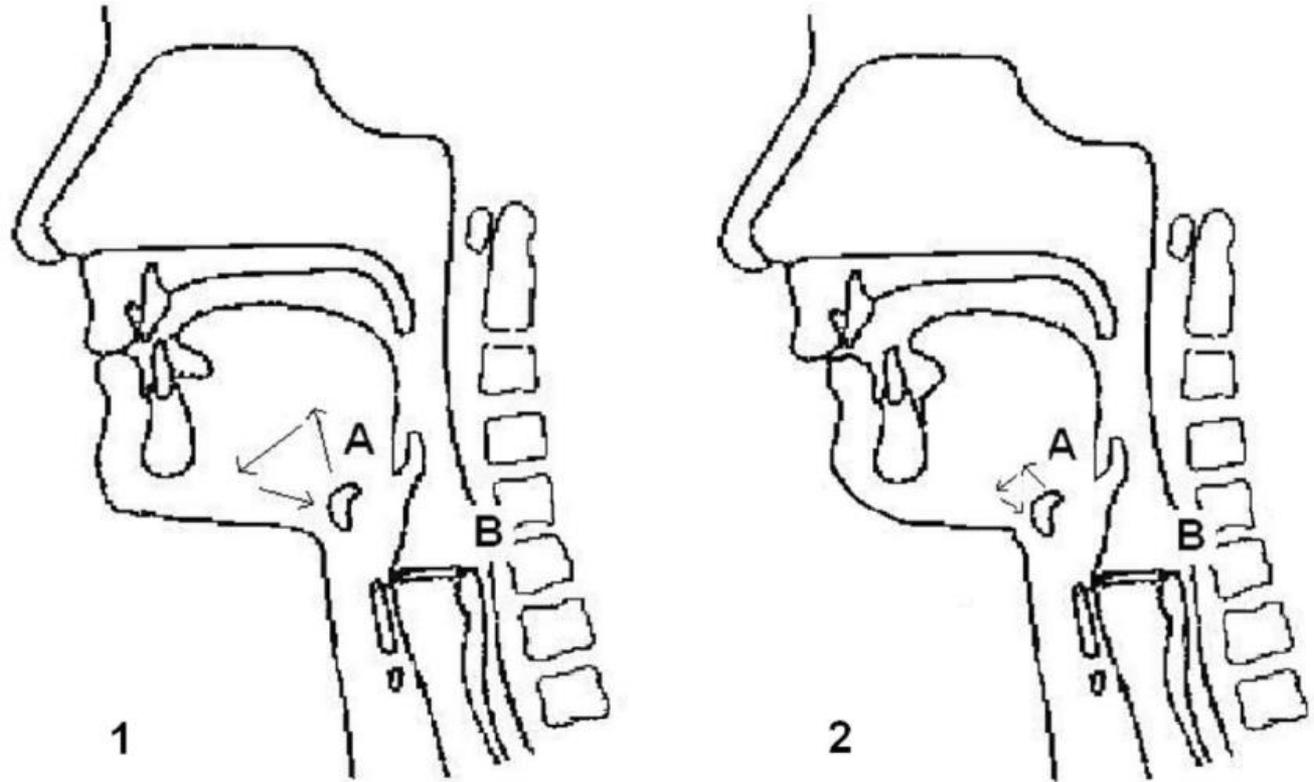
- TIM ZA MOTNJE POŽIRANJA IN PREHRANJEVANJA

ORL, URI-SOČA, klinični dietetik, psiholog, nevrolog, pulmolog, osebni in razvojni pediater,

- konzistenca hrane, čas hranjenja, hiperkalorična prehrana- DIETETIK
- položaj pri hranjenju, “učenje” hranjenja- LOGOPED



TIMSKA OBRAVNAVA otrok z ŽMB



- Pomen položaja glave
- **Pogosto spiranje hrane med obroki**

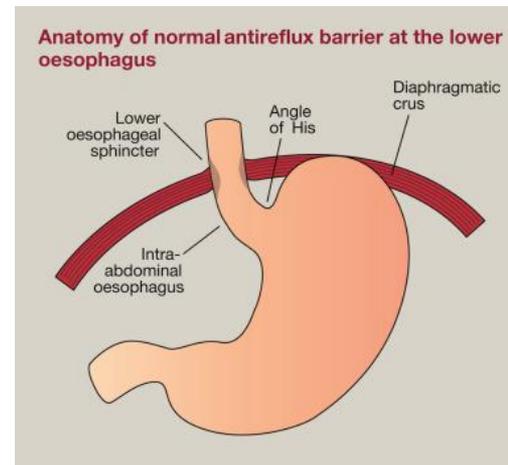
- Ledvena lordoza s kompenzatornim položajem vratu in čeljusti
- Omejena gibljivost čeljusti in jezika
- Pozno odpiranje ZPS

PREMAJHEN VNOS- drugi vzroki

- omejen nabor živil (okus, tekstura- senzorni vzroki)
- averzija do hrane
- nezmožnost izražanja lakote/sitosti
- zobni karies, nepravilen ugriz
- **GERB**
- **ZAPRTJE**

Otroci z NO in GERB

- 70 % NO otrok ima GERB Bayram, BrainDev 2016.



PREPOZNAVANJE IN SPREMLJANJE GERB-a PRI NO

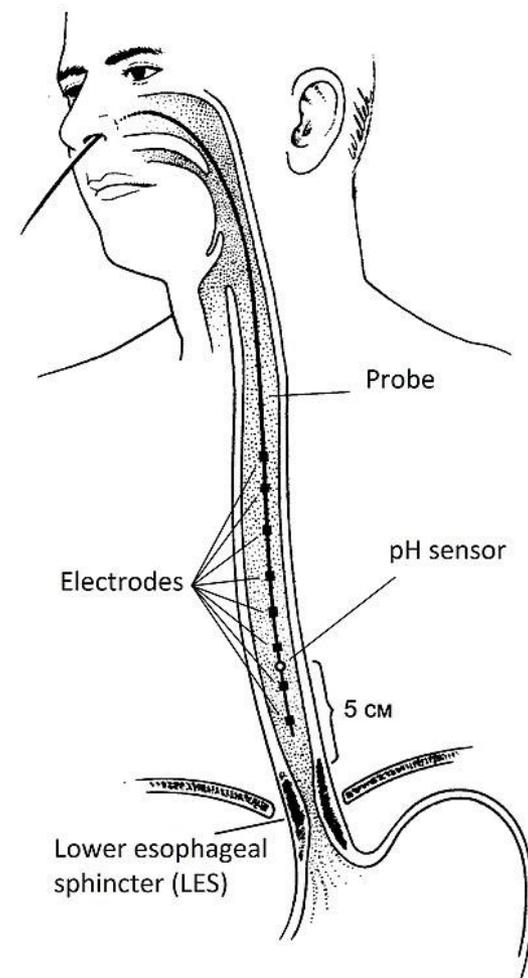
- anamneza GERB-a ni mogoča
- prekrivanje simptomatike in znakov GERB-a, cerebralnih paroksizmov in motenih faz požiranja

- Ph-impedanca
- EGDS z biopsijami
- Kontrastno slikanje zgornjih prebavil

PH-IMPEDANČNA PREISKAVA

PAZI PRI otrocih z NO !

- ezofagitis in dizmotiliteta znižujeta bazalno upornost, zato je število RE manjkrat prepoznano pri otrocih z NO
- povezanost ekstraesofagealne simptomatike z RE (+ CMCRF)



The Los Angeles Classification System for Esophagitis

Los Angeles Grade A



One or more mucosal breaks no longer than 5 mm, not bridging the tops of mucosal folds

Los Angeles Grade B



One or more mucosal breaks longer than 5 mm, not bridging the tops of mucosal folds

Los Angeles Grade C



One or more mucosal breaks bridging the tops of mucosal folds involving <75% of the circumference

Los Angeles Grade D



One or more mucosal breaks bridging the tops of mucosal folds involving >75% of the circumference

Lundell LR, et al. *Gut*. 1999;45:172-180.
Armstrong D, et al. *Gastroenterology*. 1996;111:85-92.

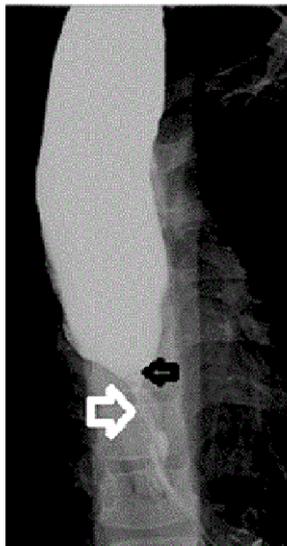
EGDS Z BIOPSIJAMI

- stopnja GERB-a (LA- A,B,C,D)
- Barrettovim požiralnik^{Raicevic, WJP} 2018.
- lokacija katetra
- stenoza
- groba ocena gastropareze
- histopatološka analiza- DD
- TE fistula
- ahalazija



KONTRASTNO SLIKANJE ZGORNJIH PREBAVIL

- **sindrom zgornje mezenterične arterije** Neuman, Case RepMed2014.
 - skolioza
 - distrofija
- **zožitev požiralnika**
- **malrotacijo, ahalazija**



Zdravljenje- konzervativni ukrepi

- gostitev hrane in tekočin z zgoščevalci^{Aggett, JPGN 2002.}
- več manjših obrokov
- manj mastna hrana
- položajno zdravljenje

Zdravljenje z ZPČ

Given their high prevalence of GORD, a trial of PPIs with careful clinical follow-up is acceptable management in this clinically fragile group of children.

JPGN 2017.

- ponavadi 1x na dan zjutraj pol ure pred obrokom
- dolgotrajna terapija- občasna prekinitev zdravljenja in ponovna ocena potrebe po ZPČ s preiskavami
- ~~probiotiki~~

Proton Pump Inhibitors (PPIs)

Omeprazole	1-4 mg/kg/day	40 mg
Lansoprazole	2 mg/kg/day for infants	30 mg
Esomeprazole	10 mg/day (weight <20kg) or 20 mg/day (weight >20kg)	40 mg
Pantoprazole	1-2 mg/kg/day	40 mg

Rosen, JPGN 2018.

Fundoplikacija

Fundoplication caution!

Complications of Gastroesophageal Antireflux Surgery in Neurologically Impaired Versus Neurologically Normal Children

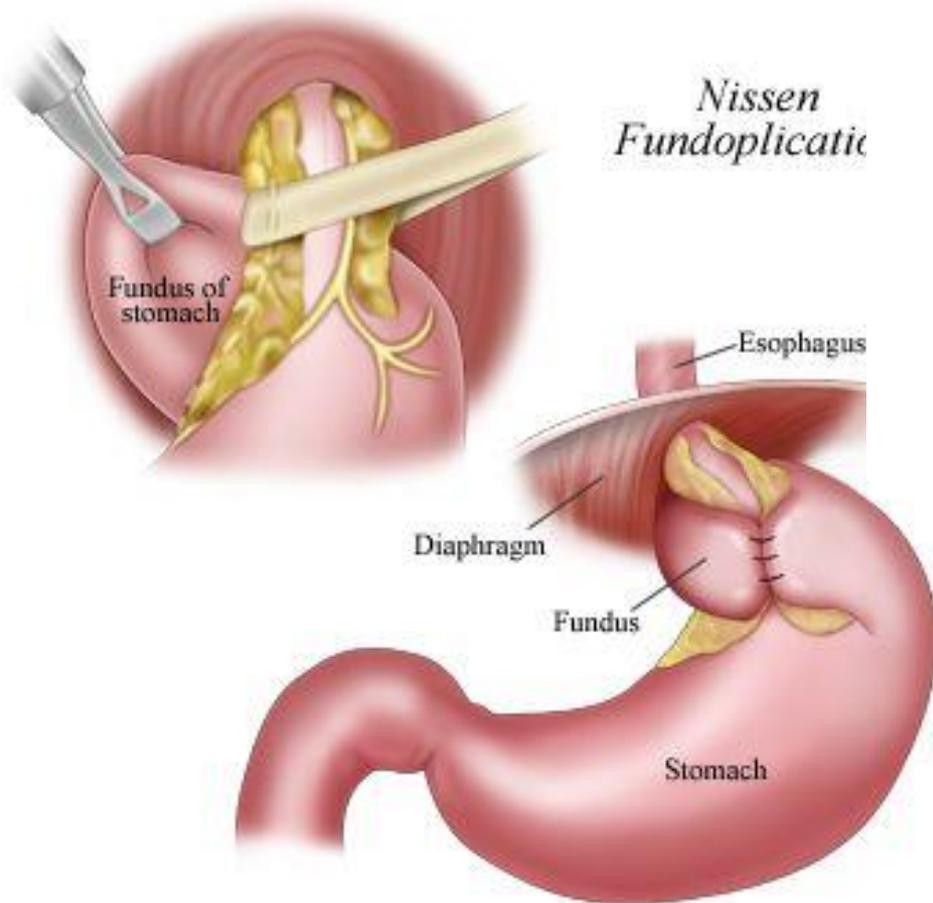
By R.H. Pearl, D.K. Robie, S.H. Ein, B. Shandling, D.E. Wesson, R. Superina, K. McTaggart, V.F. Garcia, J.A. O'Connor, and R.M. Filler
Washington, DC and Toronto, Ontario

Journal of Pediatric Surgery, Vol 25, No 11 (November), 1990: pp 1169-1173

Nissen Fundoplication 234 children
- 153 neurologically impaired (NI)
- 81 neurologically normal (NN)

Postoperative complications 26% (NI) vs 12% (NN)
Re-operation 19% (NI) vs 5% (NN) (Mortality 9:1)

Combined failure rate 28% (NI) vs 6% (NN)



Otroci z NO in ZAPRTJE

- 56 % NO otrok ima težave z zaprtjem^{Veugelers, 2010.}

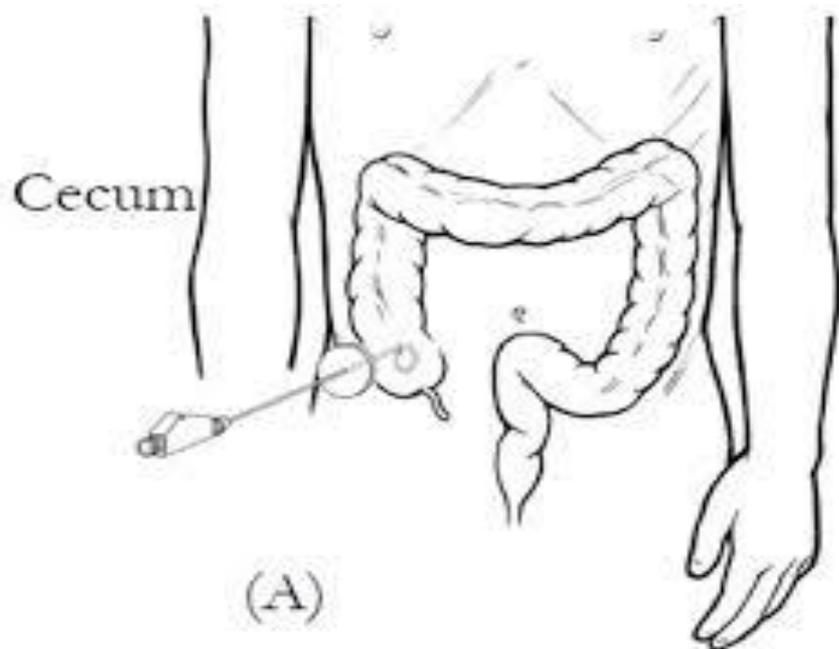


ZDRAVLJENJE ZAPRTJA^{TABBERS,2014.}

- PEG se priporoča kot zdravilo prvega reda
- 1-1.5g/kg/dan ali klizma za dezimpaktacijo
- začetni odmerek vzdrževalnega zdravljenja je 0.4 g/kg/dan
- laktuloza 1-2g/kg/dan za dojenčke < 6 mesecev starosti
- glicerinska, lecikarbon svečka, bisakodil svečke, preparati senne
- mikrokлизme
- prehrana- več poudarka na balastnih snoveh, tekočini



CEKOSTOMIJA KING,2005.



“POVEČANE” POTREBE po energiji^{WALKER,2012.}

ENERGY REQUIREMENTS IN CEREBRAL PALSY

TABLE 3

Anthropometric, energy requirement, and body-composition results in children with cerebral palsy according to motor type and in typically developing children¹

	Spasticity (<i>n</i> = 26)	Dyskinesia (<i>n</i> = 5)	Hypotonia (<i>n</i> = 1)	Typically developing children (<i>n</i> = 16)
Age (y)	3.7 ± 0.5	3.7 ± 0.7	4.0	3.7 ± 0.5
Weight <i>z</i> score	-0.4 ± 1.3 ²	-1.9 ± 2.1	0.3	0.6 ± 0.6
Height <i>z</i> score	-0.5 ± 1.3 ²	-1.1 ± 1.4	-1.5	0.5 ± 0.7
FFMI (kg/m ²)	11.7 ± 1.4 ²	10.7 ± 1.7	11.1	12.7 ± 1.0
Body fat (%)	25.2 ± 7.4	20.9 ± 8.6	36.0	22.9 ± 3.6
ER (kJ/d)	5722 ± 1378 ²	4924 ± 875	4707	6411 ± 755

¹ All values are means ± SDs, except for the child with hypotonia. ER, energy requirement; FFMI, fat-free mass index.

² Significantly different from the typically developing children, *P* < 0.05 (independent *t* tests). Statistical comparisons between other motor types could not be made because of small sample sizes.

- intenzivna fizioterapija
- kronične okužbe dihal
- pogosti cerebralni paroksizmi
- kronična bolečina

VITAMINI, MINERALI

- pomanjkanje mikronutrientov^{HILLESUND,2007.}

ŽELEZO

KALCIJ, VITAMIN D

VITAMIN A

ZINK

B₁₂, FOLNA KISLINA

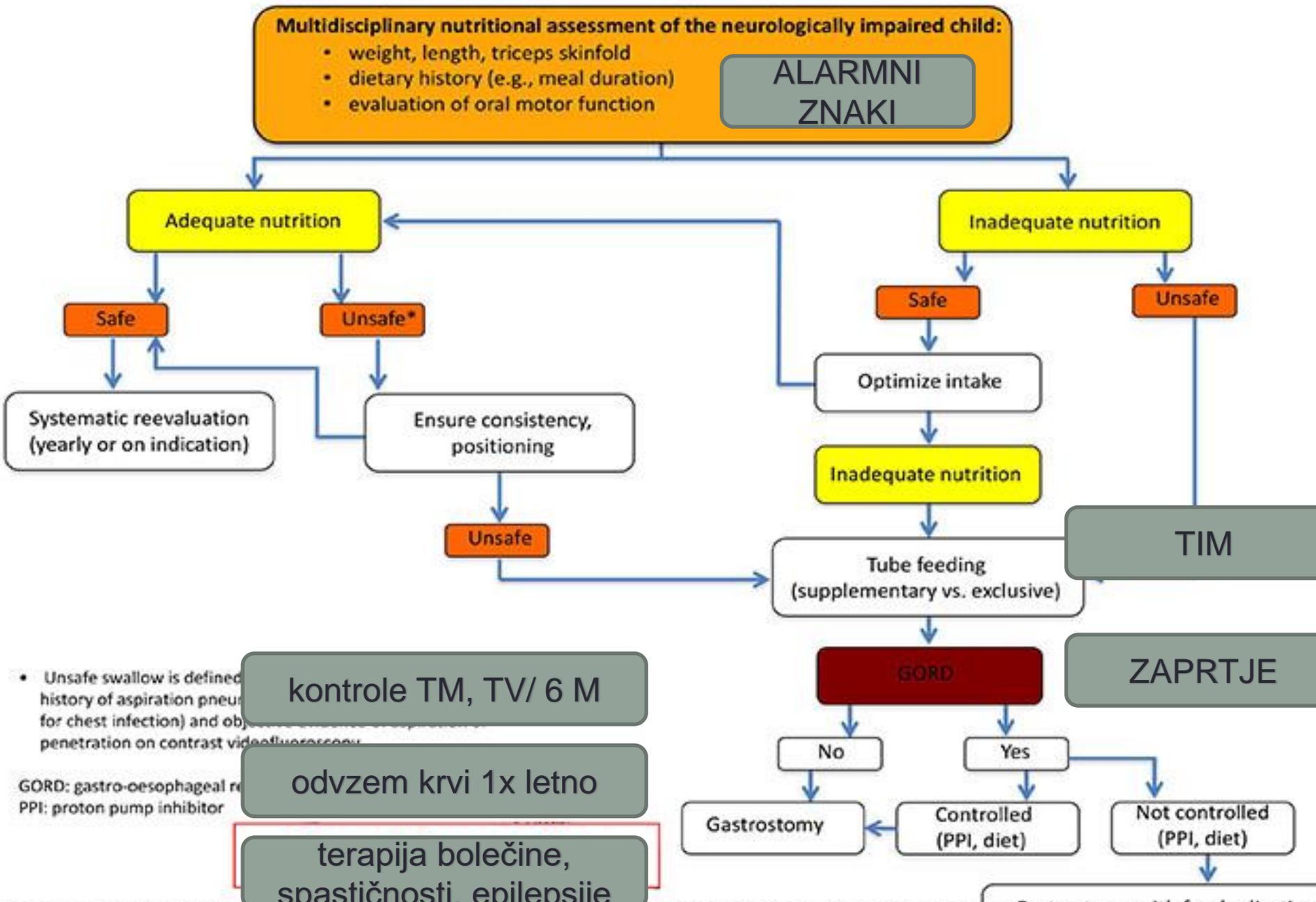
KONTROLE 1X LETNO

+hemogram, ledvični retenti,
hepatogram

POSLEDICE PODHRANJENOSTI

- vpliv na imunski sistem^{CUNNINGHAM,2005.}
- upočasnjeno celjenje operativnih ran
- znižana kostna gostota
- upočasnjen PMR^{SACHDEV,2005. BLACK, 2007.}
- življenska doba^{BROOKS, 2011.}
- slabša mišična moč

TAKE HOME MESSAGE





TIMSKI PRISTOP

HVALA ZA POZORNOST