# Modified Atkins diet in GLUT1-DS

Stéphane Auvin CHU Robert-Debré & INSERM U1141 Université Paris-Diderot Paris







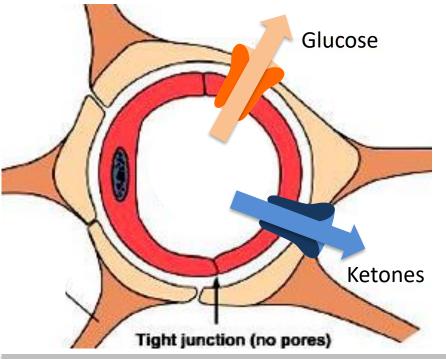


## Stéphane Auvin - Disclosures

Clinical trials: Advicenne Pharma, Eisai, UCB pharma, Ultragenyx, Zogenyx

Advice/Expertise: Advicenne Pharma, Biocodex, Eisai, GSK, Novartis, Nutricia, Shire, Servier, Ultragenyx, Zogenyx

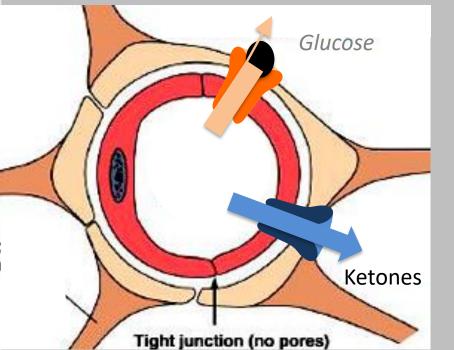
**Lectures:** Advicenne Pharma, Biocodex, Eisai, Novartis, Nutricia, Shire



#### Physiological condition

Regular food intake

Balanced energetic level in the brain

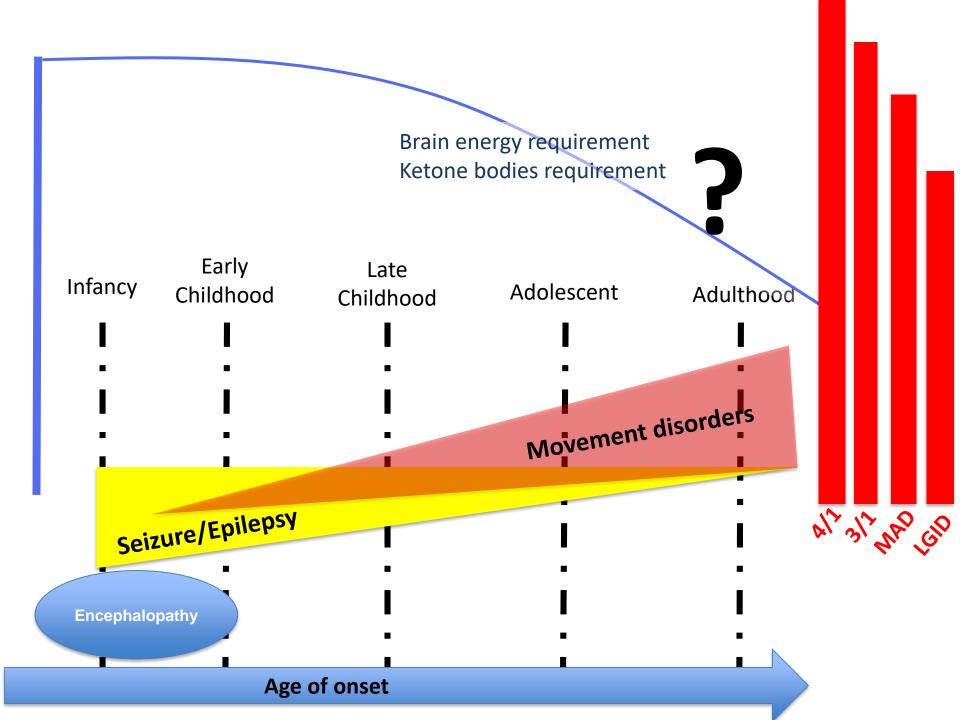


#### **GLUT1-DS**

Regular food intake: inadequate energetic brain level

Ketone bodies used as an alternative 'fuel'

Adapted Ketone bodies level?



#### **Use of modified Atkins Diet?**

Ketogenic diet considered as gold standard

Japan (questionnaire to Ped Neurol)

Fujii et al. 2016

- 39 patients: 31 KD > 1 month
- N=17 (55%) on MAD, N=11 (35%) on classic KD, 3 on MCT
- B-OHB: MAD 2.5 mM (0.75–4.1), KD 1.7 mM (0.23–3.5), MCT 2.6 mM (1.5–3.0)
- USA (families)

Kass et al. 2016

- 92 families (98% on dietary therapies).
- 59 classic KD, 29 MAD, 4 MCT, 2 LGI.
- Seizure-free younger on average (8.2 vs. 11.6 years, p = 0.01)
- Seizure-free slightly younger at GLUT1DS diagnosis (3.8 vs. 5.3 years, p = 0.05)
- KD/MCT Diets vs. MAD/LGIT (74% vs. 63%, p = 0.30)

# Methods

Retrospective observational study

 Pediatric Neurology at Robert Debré Children's Hospital, Paris, France.

 Completeness of data by 3 local databases (Pediatric neurology department, Neurophysiology Department, Dieticians)

 May 31, 2008 to June 1, 2015. Patients that discontinued MAD for at least 1 month were excluded.

	1	2	3	4	5	6	7	8	9	10
Sex	F	М	F	F	М	F	F	М	F	М
Age	14m	21m	4y 6m	6y 5m	7y 6m	9y 5m	14y	14y	16y	18y 5m
							7m	10m	4m	
First	2m	9m	6m	Зу	2y 7m	3у	9m	<b>4</b> y	6m	4m
symptoms										
Age at	4m	13m	8m	4y11	3y10m	4y 6m	12y	<b>11</b> y	16y	8y
diagnosis										
Sz type	-	Abs	-	Abs	-	Abs/FS	Abs	GTC	Abs/FS	Abs/GTC
Mvt D	AEM	-	AEM	-	CM	-	PMD	PMD	AEM	AEM
Gait	-	-	-	-	-	-	-	-	-	-
Ataxia	-	-	-	-	+	+	+	-	+	+
Spasticity	-	-	-	-	-	+	+	-	+	+
Hypotonia	+	-	+	-	-	-	-	+	+	+
Dystonia	+	-	-	-	+	-	+	-	+	+
Lang delay	-	-	-	-	-	-	-	-	-	-
Dyspraxia	S	- Mild	- N	- N	- N	- Mod.	- S	+ N	- S	- Mod.
ID	3	IVIIIU	IN	IN	IN	iviou.	3	IN	3	iviou.
AED	_	ESM	-	VPA/	-	VPA/LTG	VPA/	LEV/C	NA	PB/CBZ
(before diet)				ESM/LTG		/LEV/ESM	ESM/	LB/SUL		/VGB
(32333 333)						,,	LTG	,		,
CSF glucose	2.2	1.8	1.9	2.1	1.8	1.48	1.9	2.2	2.2	2.1
Ratio	0.47	0.38	0.39	0.45	0.4	0.3	0.34	0.45	0.41	0.43

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Mvt D	AEM	-	AEM	-	CM	-	PMD	PMD	AEM	AEM
Gait	-	-	-	-	-	-	-	-	-	-
Ataxia	-	-	-	-	+	+	+	-	+	+
Spasticity	-	-	-	-	-	+	+	-	+	+
Hypotonia	+	-	+	-	-	-	-	+	+	+
Dystonia Dystonia	+	-	-	-	+	-	+	-	+	+
Lang delay	-	-	-	-	-	-	-	-	-	-
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Gait	-	-	-	-	-	-	-	-	-	-
Ataxia	-	-	-	-	+	+	+	-	+	+
Spasticity	-	-	-	-	-	+	+	-	+	+
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Dystonia	+	-	-	-	+	-	+	-	+	+
Lang delay	-	-	-	-	-	-	-	-	-	-
Dyspraxia	-	-	-	-	-	-	-	+	-	-
ID	S	Mild	N	N	N	Mod.	S	N	S	Mod.
AED	_	ESM	_	VPA/	_	VPA/LTG	VPA/	LEV/C	NA	PB/CBZ/
(before diet)		LJIVI		ESM/LTG		/LEV/ESM	ESM/	LB/SUL	INA	VGB
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-	Abs	-	Abs	-	Abs/FS	Abs	GTC	Abs/FS	Abs/GTC
AEM	-	AEM	-	CM	-	PMD	PMD	AEM	AEM
-	-	-	-	-	-	-	-	-	-
-	-	-	-	+	+	+	-	+	+
-	-	-	-	-	+	+	-	+	+
+	-	+	-	-	-	-	+	+	+
+	-	-	-	+	-		-		+
-	-	-	-	-	-		-	-	-
-	- N 4:1 -1	- NI	- NI		- N4l				- 0.4l
5	IVIIIa	IN	IN	IN	ivioa.	5	IN	5	Mod.
	FSM				VPA/ITG	<b>VPΔ</b> /	LFV/C	NΔ	PB/CBZ
	23141		-			_	-	147 (	/VGB
			LSIVITLIC		/ LL V/ L31VI	-	LD/JOL		/ VGB
									2.1
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	F 14m 2m 4m - AEM 2.2	F M 14m 21m  2m 9m  4m 13m  - Abs AEM	F       M       F         14m       21m       4y 6m         2m       9m       6m         4m       13m       8m         -       Abs       -         AEM       -       AEM         -       -       -         -       -       -         -       -       -         +       -       -         -       -       -         S       Mild       N         -       ESM       -         2.2       1.8       1.9	F         M         F         F           14m         21m         4y 6m         6y 5m           2m         9m         6m         3y           4m         13m         8m         4y11           -         Abs         -         Abs           AEM         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           +         -         -         -           -         -         -         -           -         -         -         -           S         Mild         N         N           -         ESM/LTG           2.2         1.8         1.9         2.1	F         M         F         F         M           14m         21m         4y 6m         6y 5m         7y 6m           2m         9m         6m         3y         2y 7m           4m         13m         8m         4y11         3y10m           -         Abs         -         Abs         -           -         AEM         -         CM           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           +         -         -         -           +         -         -         -           -         -         -         -           +         -         -         -           +         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         - <tr< td=""><td>F         M         F         F         M         F           14m         21m         4y 6m         6y 5m         7y 6m         9y 5m           2m         9m         6m         3y         2y 7m         3y           4m         13m         8m         4y11         3y10m         4y 6m           -         Abs         -         Abs/FS           AEM         -         CM         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -</td><td>F         M         F         F         M         F         F           14m         21m         4y 6m         6y 5m         7y 6m         9y 5m         14y 7m           2m         9m         6m         3y         2y 7m         3y         9m           4m         13m         8m         4y11         3y10m         4y 6m         12y           -         Abs         -         Abs 7FS         Abs           AEM         -         Abs 7FS         Abs           AEM         -         CM         -         PMD           -         -         -         -         -         -           -         -         -         -         -         -         -           -</td><td>F         M         F         F         M         F         F         M           14m         21m         4y 6m         6y 5m         7y 6m         9y 5m         14y         14y           2m         9m         6m         3y         2y 7m         3y         9m         4y           4m         13m         8m         4y11         3y10m         4y 6m         12y         11y           -         Abs         -         Abs/FS         Abs         GTC           AEM         -         CM         -         PMD         PMD           -</td><td>F         M         F         F         M         F         F         M         F           14m         21m         4y 6m         6y 5m         7y 6m         9y 5m         14y         14y         16y           2m         9m         6m         3y         2y 7m         3y         9m         4y         6m           4m         13m         8m         4y11         3y10m         4y 6m         12y         11y         16y           -         Abs         -         Abs/FS         Abs         GTC         Abs/FS           AEM         -         Abs         -         Abs/FS         Abs         GTC         Abs/FS           AEM         -         Abm         -         PMD         PMD         AEM           -         -         -         -         -         -         -         -           -         -         -         -         -         -         -         -         -         -           -         -         -         -         -         -         -         -         -         -         -         -         -         -         -         -         -</td></tr<>	F         M         F         F         M         F           14m         21m         4y 6m         6y 5m         7y 6m         9y 5m           2m         9m         6m         3y         2y 7m         3y           4m         13m         8m         4y11         3y10m         4y 6m           -         Abs         -         Abs/FS           AEM         -         CM         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -         -         -         -           -	F         M         F         F         M         F         F           14m         21m         4y 6m         6y 5m         7y 6m         9y 5m         14y 7m           2m         9m         6m         3y         2y 7m         3y         9m           4m         13m         8m         4y11         3y10m         4y 6m         12y           -         Abs         -         Abs 7FS         Abs           AEM         -         Abs 7FS         Abs           AEM         -         CM         -         PMD           -         -         -         -         -         -           -         -         -         -         -         -         -           -	F         M         F         F         M         F         F         M           14m         21m         4y 6m         6y 5m         7y 6m         9y 5m         14y         14y           2m         9m         6m         3y         2y 7m         3y         9m         4y           4m         13m         8m         4y11         3y10m         4y 6m         12y         11y           -         Abs         -         Abs/FS         Abs         GTC           AEM         -         CM         -         PMD         PMD           -	F         M         F         F         M         F         F         M         F           14m         21m         4y 6m         6y 5m         7y 6m         9y 5m         14y         14y         16y           2m         9m         6m         3y         2y 7m         3y         9m         4y         6m           4m         13m         8m         4y11         3y10m         4y 6m         12y         11y         16y           -         Abs         -         Abs/FS         Abs         GTC         Abs/FS           AEM         -         Abs         -         Abs/FS         Abs         GTC         Abs/FS           AEM         -         Abm         -         PMD         PMD         AEM           -         -         -         -         -         -         -         -           -         -         -         -         -         -         -         -         -         -           -         -         -         -         -         -         -         -         -         -         -         -         -         -         -         -         -

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Mvt D	AEM	-	AEM	-	CM	-	PMD	PMD	AEM	AEM
Gait	-	-	-	-	-	-	-	-	-	-
Ataxia	-	-	-	-	+	+	+	-	+	+
Spasticity	-	-	-	-	-	+	+	-	+	+
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ID	S	Mild	N	N	N	Mod.	S	N	S	Mod.
AED	_	ESM		VPA/		VPA/LTG	VPA/	LEV/C	NA	PB/CBZ
(before diet)		LJIVI		ESM/LTG		/LEV/ESM	ESM/	LB/SUL	IVA	/VGB
(before diet)				LSIVITLIC		/ LL V/ L3IVI	LTG	LD/JOL		/ VGD
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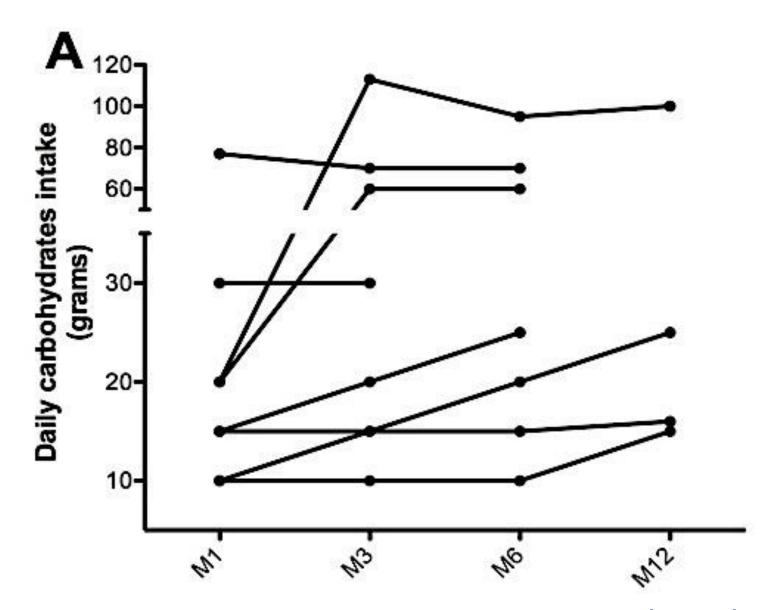
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Mvt D	AEM		AEM		CM		PMD	PMD	AEM	AEM
Gait										
Ataxia					+	+	+		+	+
Spasticity						+	+		+	+
Hypotonia	+		+					+	+	+
Dystonia	+				+		+		+	+
Lang delay								+		
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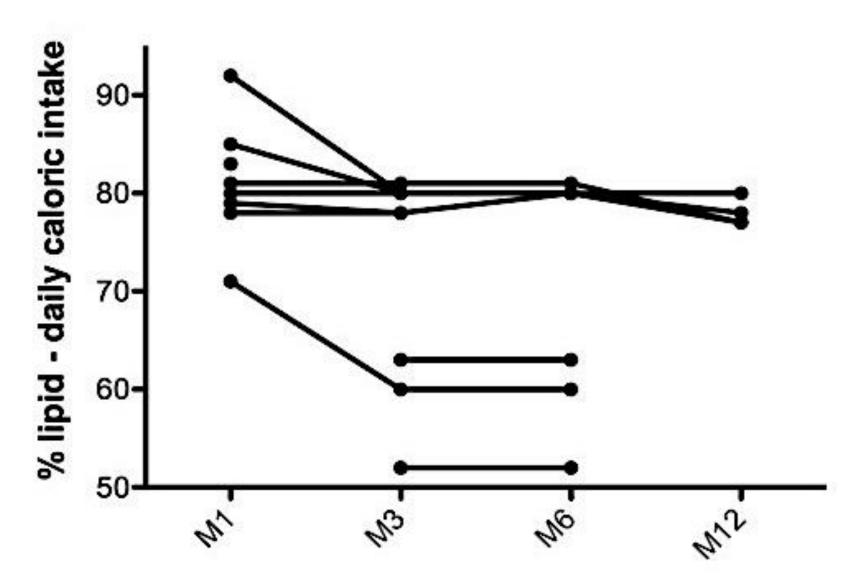
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Gait	-	-	-	-	-	-	-	-	-	-
Ataxia	-	-	-	-	+	+	+	-	+	+
Spasticity	-	-	-	-	-	+	+	-	+	+
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Lang delay	-	-	-	-	-	-	-	-	-	-
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#### Our 'Modified Atkins diet'

- No fasting period in the in-patient department
- No caloric restriction
- Proteins: 30-35% of caloric intake
- Intially 10-15g of carbohydrates daily initially
- After clinical improvement, allowed to increase carbohydrates by 5 g/day at intervals of at least 1 month, up to a maximum of 10% of caloric intake per day.
- In most patients, we allowed a maximum of 40g of carbohydrates if tolerated.



Amalou et al. DMCN 2016



Amalou et al. DMCN 2016

		-				Patie					
		1	2	3	4	5	6	7	8	9	10
Age a	at diet start	3y 10m	13m	8m	4y 11m	8y	4y6m / 8y3m	12y	<b>11</b> y	16y	8y / 1
Тур	pe of diet	MAD	MAD	MAD	MAD	MAD	KD/MAD	MAD	MAD	MAD	KD/M
Duration of	f MAD treatment	11m	11m	4y	1y10m	3y9m	15m	2y6m	3у	6m	6у
	Seizure type	0	0	0	Absences	0	0/FS	0	↓GTC	0	0
MAD M1	MD	Free of MD	0	Free of MD	0	Free of MD	0	Free of MD	↓ PMD	Free of MD	Free o
	Antiepileptic drug	0	ESM	0	VPA/ESM/LT G	0	VPA/LTG	VPA	VPA/CLB	0	0
	CGI	Much	Very much	Very much	Much	Very	Much	Much	Much	Mini	Mud
			improved	improved	improved	much improved	improved	improved	improved		impro
	Seizure type	0	0	0	Absences		0/↓FS	↓Absence s	0	0	0
MAD M3	MD	Free of MD	0	Free of MD	0		0	Free of MD	Free of MD	Free of MD	Free o
	Antiepileptic drug	0	ESM	0	VPA/LTG		LTG/ESM	↑ VPA	VPA	0	0
	CGI	Much	Very much	Very much	Much		Much	Mini	Much	Mini	Mud
		improved	improved	improved	improved		improved	mproved	improved	improved	impro
MAD	Seizure type	0	0	0	Absences		0/↓FS	个Absence s	0		0
M6	MD	Free of MD	0	Free of MD	0		0	Free of MD	→PMD		Free o
	Antiepileptic drug	0	0	0	VPA/↑LTG		LTG/ESM	ESM	SUL		0
	CGI	Much	Very much	Very much	Min		Much	Mini	Mini		Mud
		improved	improved	improved	improved		improved				impro
MAD	Seizure type			0	Absences		0/↓FS	→Absence s	0		0
M12	Movement disorder			Free of MD	0		0	Free of MD	→PMD		Free of
	Antiepileptic drug			0	↑VPA/LTG		LTG/ESM	ESM/LTG	SUL		0
	CGI			Very much	No change		Much improved	Mini	Mini		Mud

						Patie	nts				
		1	2	3	4	5	6	7	8	9	10
Age a	at diet start	3y 10m	13m	8m	4y 11m	8y	4y6m / 8y3m	12y	11y	16y	8y / 1
Тур	pe of diet	MAD	MAD	MAD	MAD	MAD	KD/MAD	MAD	MAD	MAD	KD/M
Duration of	f MAD treatment	11m	11m	4y	1y10m	3y9m	15m	2y6m	3у	6m	6у
	Seizure type	0	0	0	Absences	0	0/FS	0	↓GTC	0	0
MAD M1	MD	Free of MD	0	Free of MD	0	Free of MD	0	Free of MD	↓ PMD	Free of MD	Free of
	Antiepileptic drug	0	ESM	0	VPA/ESM/LT G	0	VPA/LTG	VPA	VPA/CLB	0	0
	CGI	Much	Very much	Very much	Much	Very	Much	Much	Much	Mini	Muc
			improved	improved	improved	much improved	improved	improved	improved		impro
	Seizure type	0	0	0	Absences		0/↓FS	↓Absence s	0	0	0
MAD M3	MD	Free of MD	0	Free of MD	0		0	Free of MD	Free of MD	Free of MD	Free o
	Antiepileptic drug	0	ESM	0	VPA/LTG		LTG/ESM	↑ VPA	VPA	0	0
	CGI	Much	Very much	Very much	Much		Much	Mini	Much	Mini	Mud
		improved	improved	improved	improved		improved	mproved	improved	improved	impro
MAD	Seizure type	0	0	0	Absences		0/↓FS	↑Absence s	0		0
M6	MD	Free of MD	0	Free of MD	0		0	Free of MD	→PMD		Free of
	Antiepileptic drug	0	0	0	VPA/↑LTG		LTG/ESM	ESM	SUL		0
	CGI	Much	Very much	Very much	Min		Much	Mini	Mini		Mud
		improved	improved	improved	improved		improved				impro
MAD	Seizure type			0	Absences		0/↓FS	→Absence s	0		0
MAD M12	Movement disorder			Free of MD	0		0	Free of MD	→PMD		Free of
	Antiepileptic drug			0	个VPA/LTG		LTG/ESM	ESM/LTG	SUL		0
	CGI			Very much	No change		Much improved	Mini	Mini		Muc

						Patie	nts				
		1	2	3	4	5	6	7	8	9	10
Age a	at diet start	3y 10m	13m	8m	4y 11m	8y	4y6m / 8y3m	12y	11y	16y	8y / 1
Тур	pe of diet	MAD	MAD	MAD	MAD	MAD	KD/MAD	MAD	MAD	MAD	KD/M
Duration of	f MAD treatment	11m	11m	4y	1y10m	3y9m	15m	2y6m	3у	6m	6у
	Seizure type	0	0	0	Absences	0	0/FS	0	↓GTC	0	0
MAD M1	MD	Free of MD	0	Free of MD	0	Free of MD	0	Free of MD	↓ PMD	Free of MD	Free o
	Antiepileptic drug	0	ESM	0	VPA/ESM/LT G	0	VPA/LTG	VPA	VPA/CLB	0	0
	CGI	Much	Very much	Very much	Much	Very	Much	Much	Much	Mini	Mud
			improved	improved	improved	much improved	improved	improved	improved		impro
	Seizure type	0	0	0	Absences		0/↓FS	↓Absence s	0	0	0
MAD M3	MD	Free of MD	0	Free of MD	0		0	Free of MD	Free of MD	Free of MD	Free o
	Antiepileptic drug	0	ESM	0	VPA/LTG		LTG/ESM	↑ VPA	VPA	0	0
	CGI	Much	Very much	Very much	Much		Much	Mini	Much	Mini	Mud
		improved	improved	improved	improved		improved	mproved	improved	improved	impro
MAD	Seizure type	0	0	0	Absences		0/↓FS	↑Absence s	0		0
M6	MD	Free of MD	0	Free of MD	0		0	Free of MD	→PMD		Free o
	Antiepileptic drug	0	0	0	VPA/↑LTG		LTG/ESM	ESM	SUL		0
	CGI	Much	Very much	Very much	Min		Much	Mini	Mini		Mud
		improved	improved	improved	improved		improved				impro
MAD	Seizure type			0	Absences		0	→Absence s	0		0
M12	Movement disorder			Free of MD	0		0	Free of MD	→PMD		Free o
	Antiepileptic drug			0	个VPA/LTG		LTG/ESM	ESM/LTG	SUL		0
	CGI			Very much	No change		Much improved	Mini	Mini		Mud

						Patie	nts				
		1	2	3	4	5	6	7	8	9	10
Age a	at diet start	3y 10m	13m	8m	4y 11m	8y	4y6m / 8y3m	12y	11y	16y	8y / 1
Тур	pe of diet	MAD	MAD	MAD	MAD	MAD	KD/MAD	MAD	MAD	MAD	KD/M
Duration of	f MAD treatment	11m	11m	4y	1y10m	3y9m	15m	2y6m	3у	6m	6у
	Seizure type	0	0	0	Absences	0	0/FS	0	↓GTC	0	0
MAD M1	MD	Free of MD	0	Free of MD	0	Free of MD	0	Free of MD	↓ PMD	Free of MD	Free of
	Antiepileptic drug	0	ESM	0	VPA/ESM/LT G	0	VPA/LTG	VPA	VPA/CLB	0	0
	CGI	Much	Very much	Very much	Much	Very	Much	Much	Much	Mini	Muc
			improved	improved	improved	much improved	improved	improved	improved		impro
	Seizure type	0	0	0	Absences		0/↓FS	↓Absence s	0	0	0
MAD M3	MD	Free of MD	0	Free of MD	0		0	Free of MD	Free of MD	Free of MD	Free of
	Antiepileptic drug	0	ESM	0	VPA/LTG		LTG/ESM	↑ VPA	VPA	0	0
	CGI	Much	Very much	Very much	Much		Much	Mini	Much	Mini	Muc
		improved	improved	improved	improved		improved	mproved	improved	improved	impro
144D	Seizure type	0	0	0	Absences		0/↓FS	↑Absence s	0		0
MAD	- NAD	5 f NAD	0	For a fAAD	0			Free of MD	\ D\ 4D		F 4
M6	MD 	Free of MD	0	Free of MD	0		0	Free of MD	→PMD		Free of
	Antiepileptic drug	0	0	0	VPA/↑LTG		LTG/ESM	ESM	SUL		0
	CGI	Much	Very much	Very much	Min		Much	Mini	Mini		Muc
		improved	improved	improved	improved		improved	\ \ \ \ \ \ \ \ \ \ \ \ \ \ \ \ \ \ \	0		impro
MAD	Seizure type			0	Absences		0	→Absence s	0		0
M12	Movement disorder			Free of MD	0		0	Free of MD	→PMD		Free of
	Antiepileptic drug			0	个VPA/LTG		LTG/ESM	ESM/LTG	SUL		0
	CGI			Very much	No change		Much improved	Mini	Mini		Muc

		Patients									
		1	2	3	4	5	6	7	8	9	10
Age at diet start		3y 10m	13m	8m	4y 11m	8y	4y6m / 8y3m	12y	11y	16y	8y / 12
Type of diet		MAD	MAD	MAD	MAD	MAD	KD/MAD	MAD	MAD	MAD	KD/MA
Duration of	f MAD treatment	11m	11m	4y	1y10m	3y9m	15m	2y6m	3у	6m	6у
	Seizure type	0	0	0	Absences	0	0/FS	0	↓GTC	0	0
MAD M1	MD	Free of MD	0	Free of MD	0	Free of MD	0	Free of MD	↓ PMD	Free of MD	Free of
	Antiepileptic drug	0	ESM	0	VPA/ESM/LT G	0	VPA/LTG	VPA	VPA/CLB	0	0
	CGI	Much	Very much	Very much	Much	Very	Much	Much	Much	Mini	Mucl
			improved	improved	improved	much improved	improved	improved	improved		improv
	Seizure type	0	0	0	Absences		0/↓FS	↓Absence s	0	0	0
MAD M3	MD	Free of MD	0	Free of MD	0		0	Free of MD	Free of MD	Free of MD	Free of
	Antiepileptic drug	0	ESM	0	VPA/LTG		LTG/ESM	↑ VPA	VPA	0	0
	CGI	Much	Very much	Very much	Much		Much	Mini	Much	Mini	Mud
		improved	improved	improved	improved		improved	mproved	improved	improved	impro
MAD	Seizure type	0	0	0	Absences		0/↓FS	↑Absence s	0		0
M6	MD	Free of MD	0	Free of MD	0		0	Free of MD	→PMD		Free of
	Antiepileptic drug	0	0	0	VPA/个LTG		LTG/ESM	ESM	SUL		0
	CGI	Much	Very much	Very much	Min		Much	Mini	Mini		Mud
	CG1	improved	improved	improved	improved		improved	1711111	1411111		impro
	Seizure type	IIIIpro.c.	IIIIpi 0.02	0	Absences		0/↓FS	→Absence s	0		0
MAD M12	Movement disorder			Free of MD	0		0	Free of MD	→PMD		Free of
	Antiepileptic drug			0	个VPA/LTG		LTG/ESM	ESM/LTG	SUL		0
	CGI			Very much	No change		Much improved	Mini	Mini		Mud

						Patie					
		1	2	3	4	5	6	7	8	9	10
Age at diet start		3y 10m	13m	8m	4y 11m	8y	4y6m / 8y3m	<b>12</b> y	<b>11</b> y	16y	8y / 1
Type of diet		MAD	MAD	MAD	MAD	MAD	KD/MAD	MAD	MAD	MAD	KD/M
Duration of MAD treatment		11m	11m	4y	1y10m	3y9m	15m	2y6m	3у	6m	6у
	Seizure type	0	0	0	Absences	0	0/FS	0	↓GTC	0	0
MAD M1	MD	Free of MD	0	Free of MD	0	Free of MD	0	Free of MD	↓ PMD	Free of MD	Free o
	Antiepileptic drug	0	ESM	0	VPA/ESM/LT G	0	VPA/LTG	VPA	VPA/CLB	0	0
	CGI	Much	Very much	Very much	Much	Very	Much	Much	Much	Mini	Mud
			improved	improved	improved	much improved	improved	improved	improved		impro
	Seizure type	0	0	0	Absences		0/↓FS	↓Absence s	0	0	0
MAD M3	MD	Free of MD	0	Free of MD	0		0	Free of MD	Free of MD	Free of MD	Free o
	Antiepileptic drug	0	ESM	0	VPA/LTG		LTG/ESM	↑ VPA	VPA	0	0
	CGI	Much	Very much	Very much	Much		Much	Mini	Much	Mini	Mud
		improved	improved	improved	improved		improved	mproved	improved	improved	impro
MAD	Seizure type	0	0	0	Absences		0/↓FS	个Absence s	0		0
M6	MD	Free of MD	0	Free of MD	0		0	Free of MD	→PMD		Free o
	Antiepileptic drug	0	0	0	VPA/↑LTG		LTG/ESM	ESM	SUL		0
	CGI	Much	Very much	Very much	Min		Much	Mini	Mini		Mud
		improved	improved	improved	improved		improved				impro
MAD	Seizure type			0	Absences		0/↓FS	→Absence s	0		0
M12	Movement disorder			Free of MD	0		0	Free of MD	→PMD		Free o
	Antiepileptic drug			0	个VPA/LTG		LTG/ESM	ESM/LTG	SUL		0
	CGI			Very much	No change		Much improved	Mini	Mini		Mud

#### Discussion

- Already 20 patients reported in the literature
- Mostly case reports
  - Including 6 patients Mov Disorders

- Mostly seizure free on MAD (62-69% with KD)
- Also effective on MD (41% with KD)

Leen et al. 2010 Pong et al. 2012

#### **Discussion**

- 2 patients were switch from KD to MAD
  - No change in neurological symptoms

- Growth
- Physical activity
- Fatigue

### Discussion

Various level of ketosis by KD, MAD, MCT

How to conduct the diet?

What is the ketosis requirement in each patient?

 Always have in mind the possibility to switch to KD (symptoms / development)

# Conclusion

- MAD seems to have similar efficacy than KD on Sz and MAD
- KD should be offered in non-responder to MAD
- Most important point remains diagnosis delay
- Interest to have a diet than can be keep for the long-term
- Need to be further studied
- Same cognitive outcome?????

## How to address the cognitive issue?

- The inclusion criteria will be:
  - GLUT1-DS diagnosis (clinical history + CSF)
  - Diagnosis delay <1y (first symptom to diagnosis)
  - MAD duration for at least 2 years (possible to include patient initially on KD but duration MAD>duration KD)
  - Age > or = 6-year-old and Age<18-year-old</p>

#### **Evaluation of cognition by WISC**

#### Merci pour votre attention

DEVELOPMENTAL MEDICINE & CHILD NEUROLOGY

ORIGINAL ARTICLE

# Use of modified Atkins diet in glucose transporter type 1 deficiency syndrome

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