

Impaired balance and neurodevelopmental disabilities among children with congenital CMV infection

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CLINTEC



Background

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- PCR analysis and dried blood spots - new possibilities

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- Since then: all children with severe/profound HI are tested for cCMV
- Almost 50 children identified so far
- Longitudinal follow up

Background

- Children with cCMV and CI - other issues
- Late age of walking
- Strange head tossing
- Feeding problems



Aim

- To evaluate the potential comorbid conditions among children with HI resulting from cCMV infection
- To compare with children whom have HI caused by *GJB2* mutations

Methods

- Study group: 26 children with cCMV infection and CI, 1-16 years of age
- Controls: 13 children with *GJB2* mutations and CI, 1-12 years of age

Methods

- Assessed by a multidisciplinary team: pediatrician, neuropediatrician, SLP, ophtalmologist, orthoptist, physiotherapist and otolarygologist
- A full day at the hospital
- In addition: 11 older children with cCMV infection at the vestibular laboratory

Assessment methods

- **Balance:** Movement ABC-2
Vestibular tests: Caloric stimulation, video head impulse test, evoked myogenic potentials (VEMP)
- **Neurodevelopmental dysfunction:** journals, assessment of the child, interview of parents and parent questionnaires: five to fifteen (FTF)
- **Non-verbal cognitive and spoken language abilities:**
Ravens colored matrices, PPVT-3, Speech Intelligibility Rating scale (SIR-2)

Assessment methods

- **Ocular assessment:** Visual acuity, ocular motor score and fundus photography
- **cCMV infection and *GJB2* mutations (Cx26):**
DBS was tested/retested for CMV DNA
All children were tested for *GJB2* mutations

Results



CMV	Neo	Path	Age of	Movement	Vestibular					DBS			
Case	Sympt	head	walking	ABC-2	function	Feeding	Raven	PPVT-3	SIR-2	copy	Cx26	MRI	Visual
		move	(mo)	n = 18	n = 11	issues	n = 13	n = 20		number	mutations	n = 12	function
1	0	0	12	50	1	0	-	135	5	7.6·10 ⁶	0	-	1
2	1	3	14	1	2	1	75	69	4	3.0·10 ⁶	0	1	2
3	1	2	19	-	-	0	-	12	2	8.3·10 ⁶	1	1	1
4	1	5	Nw	-	-	4	-	-	0	NA	0	3	1
5	0	6	20	25	1	0	75	164	5	1.0·10 ⁴	0	-	1
6	0	2	15	2	-	0	25	75	5	6.5·10 ³	0	-	1
7	0	2	18 ^s	-	-	0	-	28	3	3.4·10 ³	0	1	1
8	0	5	36	0.1	-	3	-	-	1	>20·10 ⁶	0	-	1
9	1	6	19	1	1	1	50	172	4	4.5·10 ⁴	0	-	1
10	0	4	24	0.1	2	0	10	179	5	10·10 ⁶	0	2	1
11	1	3	25	1	-	0	90	128	5	3.2·10 ³	0	2	2
12	0	5	21	1	-	1	10	136	5	9.7·10 ⁶	0	-	1
13	0	3	22	0.5	-	3	-	180	5	1.7·10 ⁶	0	-	2
14	0	3	24	-	3	3	-	-	1	3.6·10 ⁴	0	-	1
15	0	3	18	9	-	0	5	98	3	7.5·10 ³	0	1	2
16	0	0	10	63	-	0	-	134	5	NA	0	-	1
17	1	5	23	0.5	1	4	75	138	5	>50·10 ⁶	0	-	1
18	0	3	18	-	-	0	-	-	1	>30·10 ⁶	0	1	1
19	0	3	12 ^s	-	0	3	-	12	2	2.1·10 ³	0	-	1
20	0	0	12 ^s	9	3 ²	0	25	37	3	3.8·10 ³	0	1	1
21	0	4	24	5	-	4	50	51	4	1.2·10 ⁶	0	1	1
22	0	3	18 ^s	0.5	-	1	75	123	5	3.1·10 ³	0	-	2
23	0	3	22	1	-	0	90	116	5	7.3·10 ⁶	0	1	1
24	0	3	Nw	-	2	0	-	-	2	1.3·10 ⁶	0	0	1
25	0	4	Nw	-	-	3	-	-	2	14.8·10 ⁶	0	0	1
26	0	4	18 ^s	9	2	0	-	123	5	2.6·10 ⁶	0	-	1
Mean			19***	10**			50	106	3.5				

Cx26	Neo	Path	Age of	Movement	Vestibular					DBS			
Case	Sympt	head	walking	ABC-2	function	Feeding	Raven	PPVT-3	SIR-2	copy	Cx26	MRI	Visual
		move	(mo)	n = 9	n = 0	issues	n = 7	n = 8		number	mutations	n =	function
1	0	0	12	-	-	0	-	12	3	-	3	-	1
2	0	0	14	-	-	0	-	15	2	-	3	-	1
3	0	0	13	5	-	0	-	-	2	-	3	-	1
4	0	0	17	-	-	0	-	-	2	-	3	-	1
5	0	0	12	50	-	0	75	130	5	-	3	-	1
6	0	0	12	-	-	0	-	-	4	-	3	-	1
7	0	0	12	25	-	0	50	73	5	-	3	-	1
8	0	0	11	98	-	0	90	146	5	-	2	-	1
9	0	0	12	63	-	0	-	-	5	-	3	-	1
10	0	1	10	50	-	0	95	137	5	-	3	-	1
11	0	0	12	37	-	0	50	-	4	-	3	-	1
12	0	0	11	16	-	0	90	45	5	-	2	-	1
13	0	1	14	63	-	0	95	100	5	-	3	-	1
Mean			12***	45**			78	82	4				

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Case	Sympt												
1	0	0	12	50	1	0	-	135	5	7.6·10 ⁶	0	-	1
2	1	3	14	1	2	1	75	69	4	3.0·10 ⁶	0	1	2
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Cx26	Neo	Path head move	Age of walking (mo)	Movement ABC-2 n = 9	Vestibular function n = 0	Feeding issues	Raven n = 7	PPVT-3 n = 8	SIR-2	DBS copy number	Cx26 mutations	MRI n =	Visual function
Case	Sympt												
1	0	0	12	-	-	0	-	12	3	-	3	-	1
2	0	0	14	-	-	0	-	15	2	-	3	-	1
3	0	0	13	5	-	0	-	-	2	-	3	-	1
4	0	0	17	-	-	0	-	-	2	-	3	-	1
5	0	0	12	50	-	0	75	130	5	-	3	-	1
6	0	0	12	-	-	0	-	-	4	-	3	-	1
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Mean			19 ^{***}	10 ^{**}			50	106	3.5				

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4	0	0	17	-	-	0	-	-	2	-	3	-	1
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4	0	0	17	-	-	0	-	-	2	-	3	-	1
5	0	0	12	50	-	0	75	130	5	-	3	-	1
6	0	0	12	-	-	0	-	-	4	-	3	-	1
7	0	0	12	25	-	0	50	73	5	-	3	-	1
8	0	0	11	98	-	0	90	146	5	-	2	-	1
9	0	0	12	63	-	0	-	-	5	-	3	-	1
10	0	1	10	50	-	0	95	137	5	-	3	-	1
11	0	0	12	37	-	0	50	-	4	-	3	-	1
12	0	0	11	16	-	0	90	45	5	-	2	-	1
13	0	1	14	63	-	0	95	100	5	-	3	-	1
Mean			12***	45**			78	82	4				

CMV	Neo	Path	Age of	Movement	Vestibular	Feeding	Raven	PPVT-3		DBS		MRI	Visual
Case	Sympt	head	walking	ABC-2	function	issues	n = 13	n = 20	SIR-2	copy	Cx26	n = 12	function
		move	(mo)	n = 18	n = 11					number	mutations		
1	0	0	12	50	1	0	-	135	5	7.6·10 ⁶	0	-	1
2	1	3	14	1	2	1	75	69	4	3.0·10 ⁶	0	1	2
3	1	2	19	-	-	0	-	12	2	8.3·10 ⁶	1	1	1
4	1	5	Nw	-	-	4	-	-	0	NA	0	3	1
5	0	6	20	25	1	0	75	164	5	1.0·10 ⁴	0	-	1
6	0	2	15	2	-	0	25	75	5	6.5·10 ³	0	-	1
7	0	2	18 ^s	-	-	0	-	28	3	3.4·10 ³	0	1	1
8	0	5	36	0.1	-	3	-	-	1	>20·10 ⁶	0	-	1
9	1	6	19	1	1	1	50	172	4	4.5·10 ⁴	0	-	1
10	0	4	24	0.1	2	0	10	179	5	10·10 ⁶	0	2	1
11	1	3	25	1	-	0	90	128	5	3.2·10 ³	0	2	2
12	0	5	21	1	-	1	10	136	5	9.7·10 ⁶	0	-	1
13	0	3	22	0.5	-	3	-	180	5	1.7·10 ⁶	0	-	2
14	0	3	24	-	3	3	-	-	1	3.6·10 ⁴	0	-	1
15	0	3	18	9	-	0	5	98	3	7.5·10 ³	0	1	2
16	0	0	10	63	-	0	-	134	5	NA	0	-	1
17	1	5	23	0.5	1	4	75	138	5	>50·10 ⁶	0	-	1
18	0	3	18	-	-	0	-	-	1	>30·10 ⁶	0	1	1
19	0	3	12 ^s	-	0	3	-	12	2	2.1·10 ³	0	-	1
20	0	0	12 ^s	9	3 ²	0	25	37	3	3.8·10 ³	0	1	1
21	0	4	24	5	-	4	50	51	4	1.2·10 ⁶	0	1	1
22	0	3	18 ^s	0.5	-	1	75	123	5	3.1·10 ³	0	-	2
23	0	3	22	1	-	0	90	116	5	7.3·10 ⁶	0	1	1
24	0	3	Nw	-	2	0	-	-	2	1.3·10 ⁶	0	0	1
25	0	4	Nw	-	-	3	-	-	2	14.8·10 ⁶	0	0	1
26	0	4	18 ^s	9	2	0	-	123	5	2.6·10 ⁶	0	-	1
Mean			19***	10**			50	106	3.5				

Cx26	Neo	Path	Age of	Movement	Vestibular	Feeding	Raven	PPVT-3		DBS		MRI	Visual
Case	Sympt	head	walking	ABC-2	function	issues	n = 7	n = 8	SIR-2	copy	Cx26	n =	function
		move	(mo)	n = 9	n = 0					number	mutations		
1	0	0	12	-	-	0	-	12	3	-	3	-	1
2	0	0	14	-	-	0	-	15	2	-	3	-	1
3	0	0	13	5	-	0	-	-	2	-	3	-	1
4	0	0	17	-	-	0	-	-	2	-	3	-	1
5	0	0	12	50	-	0	75	130	5	-	3	-	1
6	0	0	12	-	-	0	-	-	4	-	3	-	1
7	0	0	12	25	-	0	50	73	5	-	3	-	1
8	0	0	11	98	-	0	90	146	5	-	2	-	1
9	0	0	12	63	-	0	-	-	5	-	3	-	1
10	0	1	10	50	-	0	95	137	5	-	3	-	1
11	0	0	12	37	-	0	50	-	4	-	3	-	1
12	0	0	11	16	-	0	90	45	5	-	2	-	1
13	0	1	14	63	-	0	95	100	5	-	3	-	1
Mean			12***	45**			78	82	4				

CMV	Neo	Path	Age of	Movement	Vestibular					DBS			
Case	Sympt	head	walking	ABC-2	function	Feeding	Raven	PPVT-3	SIR-2	copy	Cx26	MRI	Visual
		move	(mo)	n = 18	n = 11	issues	n = 13	n = 20		number	mutations	n = 12	function
1	0	0	12	50	1	0	-	135	5	7.6·10 ⁶	0	-	1
2	1	3	14	1	2	1	75	69	4	3.0·10 ⁶	0	1	2
3	1	2	19	-	-	0	-	12	2	8.3·10 ⁶	1	1	1
4	1	5	Nw	-	-	4	-	-	0	NA	0	3	1
5	0	6	20	25	1	0	75	164	5	1.0·10 ⁴	0	-	1
6	0	2	15	2	-	0	25	75	5	6.5·10 ³	0	-	1
7	0	2	18 ^s	-	-	0	-	28	3	3.4·10 ³	0	1	1
8	0	5	36	0.1	-	3	-	-	1	>20·10 ⁶	0	-	1
9	1	6	19	1	1	1	50	172	4	4.5·10 ⁴	0	-	1
10	0	4	24	0.1	2	0	10	179	5	10·10 ⁶	0	2	1
11	1	3	25	1	-	0	90	128	5	3.2·10 ³	0	2	2
12	0	5	21	1	-	1	10	136	5	9.7·10 ⁶	0	-	1
13	0	3	22	0.5	-	3	-	180	5	1.7·10 ⁶	0	-	2
14	0	3	24	-	3	3	-	-	1	3.6·10 ⁴	0	-	1
15	0	3	18	9	-	0	5	98	3	7.5·10 ³	0	1	2
16	0	0	10	63	-	0	-	134	5	NA	0	-	1
17	1	5	23	0.5	1	4	75	138	5	>50·10 ⁶	0	-	1
18	0	3	18	-	-	0	-	-	1	>30·10 ⁶	0	1	1
19	0	3	12 ^s	-	0	3	-	12	2	2.1·10 ³	0	-	1
20	0	0	12 ^s	9	3 ²	0	25	37	3	3.8·10 ³	0	1	1
21	0	4	24	5	-	4	50	51	4	1.2·10 ⁶	0	1	1
22	0	3	18 ^s	0.5	-	1	75	123	5	3.1·10 ³	0	-	2
23	0	3	22	1	-	0	90	116	5	7.3·10 ⁶	0	1	1
24	0	3	Nw	-	2	0	-	-	2	1.3·10 ⁶	0	0	1
25	0	4	Nw	-	-	3	-	-	2	14.8·10 ⁶	0	0	1
26	0	4	18 ^s	9	2	0	-	123	5	2.6·10 ⁶	0	-	1
Mean			19***	10**			50	106	3.5				

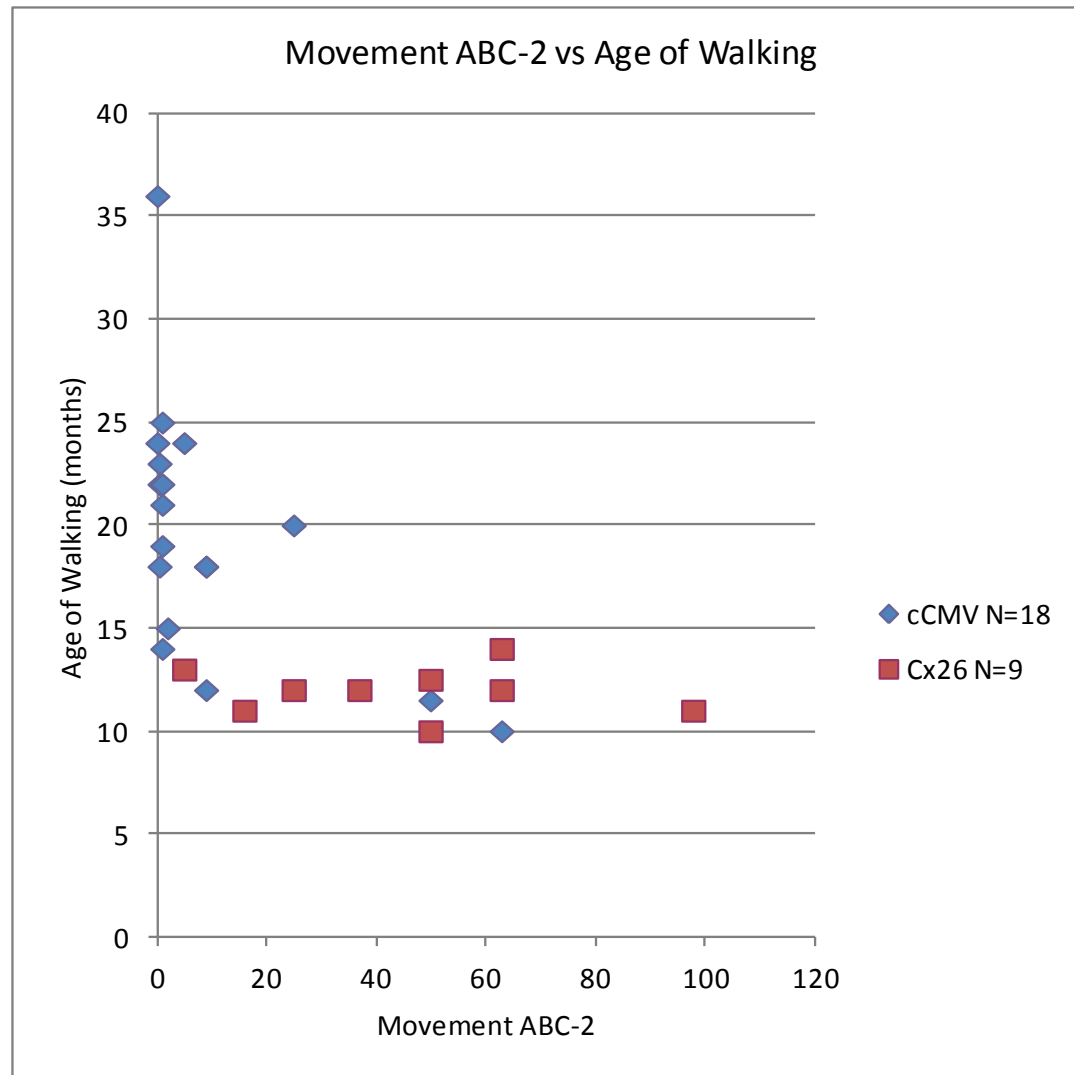
Cx26	Neo	Path	Age of	Movement	Vestibular					DBS			
Case	Sympt	head	walking	ABC-2	function	Feeding	Raven	PPVT-3	SIR-2	copy	Cx26	MRI	Visual
		move	(mo)	n = 9	n = 0	issues	n = 7	n = 8		number	mutations	n =	function
1	0	0	12	-	-	0	-	12	3	-	3	-	1
2	0	0	14	-	-	0	-	15	2	-	3	-	1
3	0	0	13	5	-	0	-	-	2	-	3	-	1
4	0	0	17	-	-	0	-	-	2	-	3	-	1
5	0	0	12	50	-	0	75	130	5	-	3	-	1
6	0	0	12	-	-	0	-	-	4	-	3	-	1
7	0	0	12	25	-	0	50	73	5	-	3	-	1
8	0	0	11	98	-	0	90	146	5	-	2	-	1
9	0	0	12	63	-	0	-	-	5	-	3	-	1
10	0	1	10	50	-	0	95	137	5	-	3	-	1
11	0	0	12	37	-	0	50	-	4	-	3	-	1
12	0	0	11	16	-	0	90	45	5	-	2	-	1
13	0	1	14	63	-	0	95	100	5	-	3	-	1
Mean			12***	45**			78	82	4				

CMV	Neo	Path	Age of	Movement	Vestibular					DBS			
Case	Sympt	head	walking	ABC-2	function	Feeding	Raven	PPVT-3	SIR-2	copy	Cx26	MRI	Visual
		move	(mo)	n = 18	n = 11	issues	n = 13	n = 20		number	mutations	n = 12	function
1	0	0	12	50	1	0	-	135	5	7.6·10 ⁶	0	-	1
2	1	3	14	1	2	1	75	69	4	3.0·10 ⁶	0	1	2
3	1	2	19	-	-	0	-	12	2	8.3·10 ⁶	1	1	1
4	1	5	Nw	-	-	4	-	-	0	NA	0	3	1
5	0	6	20	25	1	0	75	164	5	1.0·10 ⁴	0	-	1
6	0	2	15	2	-	0	25	75	5	6.5·10 ³	0	-	1
7	0	2	18 ^s	-	-	0	-	28	3	3.4·10 ³	0	1	1
8	0	5	36	0.1	-	3	-	-	1	>20·10 ⁶	0	-	1
9	1	6	19	1	1	1	50	172	4	4.5·10 ⁴	0	-	1
10	0	4	24	0.1	2	0	10	179	5	10·10 ⁶	0	2	1
11	1	3	25	1	-	0	90	128	5	3.2·10 ³	0	2	2
12	0	5	21	1	-	1	10	136	5	9.7·10 ⁶	0	-	1
13	0	3	22	0.5	-	3	-	180	5	1.7·10 ⁶	0	-	2
14	0	3	24	-	3	3	-	-	1	3.6·10 ⁴	0	-	1
15	0	3	18	9	-	0	5	98	3	7.5·10 ³	0	1	2
16	0	0	10	63	-	0	-	134	5	NA	0	-	1
17	1	5	23	0.5	1	4	75	138	5	>50·10 ⁶	0	-	1
18	0	3	18	-	-	0	-	-	1	>30·10 ⁶	0	1	1
19	0	3	12 ^s	-	0	3	-	12	2	2.1·10 ³	0	-	1
20	0	0	12 ^s	9	3 ²	0	25	37	3	3.8·10 ³	0	1	1
21	0	4	24	5	-	4	50	51	4	1.2·10 ⁶	0	1	1
22	0	3	18 ^s	0.5	-	1	75	123	5	3.1·10 ³	0	-	2
23	0	3	22	1	-	0	90	116	5	7.3·10 ⁶	0	1	1
24	0	3	Nw	-	2	0	-	-	2	1.3·10 ⁶	0	0	1
25	0	4	Nw	-	-	3	-	-	2	14.8·10 ⁶	0	0	1
26	0	4	18 ^s	9	2	0	-	123	5	2.6·10 ⁶	0	-	1
Mean			19***	10**			50	106	3.5				

Cx26	Neo	Path	Age of	Movement	Vestibular					DBS			
Case	Sympt	head	walking	ABC-2	function	Feeding	Raven	PPVT-3	SIR-2	copy	Cx26	MRI	Visual
		move	(mo)	n = 9	n = 0	issues	n = 7	n = 8		number	mutations	n =	function
1	0	0	12	-	-	0	-	12	3	-	3	-	1
2	0	0	14	-	-	0	-	15	2	-	3	-	1
3	0	0	13	5	-	0	-	-	2	-	3	-	1
4	0	0	17	-	-	0	-	-	2	-	3	-	1
5	0	0	12	50	-	0	75	130	5	-	3	-	1
6	0	0	12	-	-	0	-	-	4	-	3	-	1
7	0	0	12	25	-	0	50	73	5	-	3	-	1
8	0	0	11	98	-	0	90	146	5	-	2	-	1
9	0	0	12	63	-	0	-	-	5	-	3	-	1
10	0	1	10	50	-	0	95	137	5	-	3	-	1
11	0	0	12	37	-	0	50	-	4	-	3	-	1
12	0	0	11	16	-	0	90	45	5	-	2	-	1
13	0	1	14	63	-	0	95	100	5	-	3	-	1
Mean			12***	45**			78	82	4				

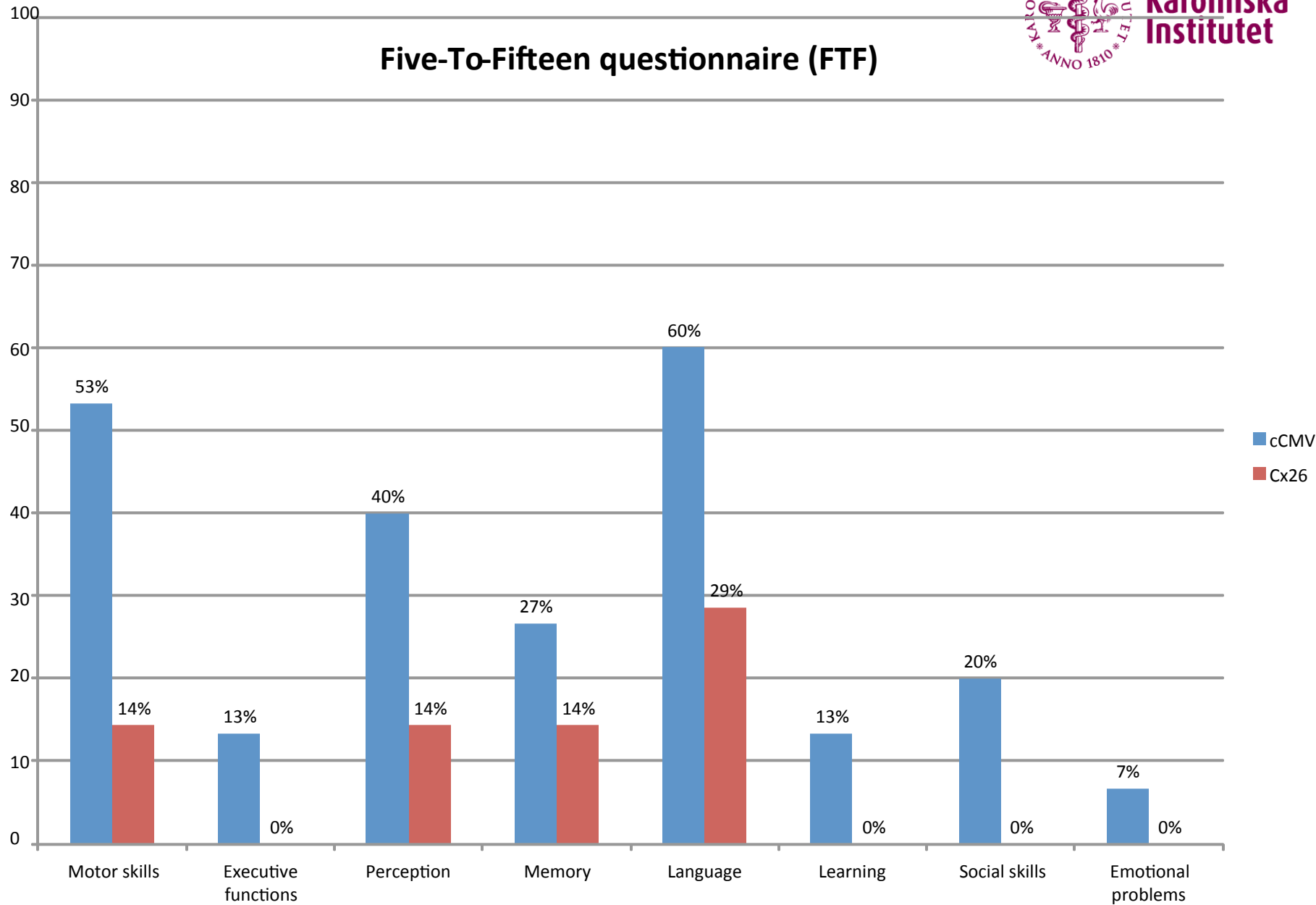
CMV	Neo	Path	Age of	Movement	Vestibular					DBS			
Case	Sympt	head	walking	ABC-2	function	Feeding	Raven	PPVT-3	SIR-2	copy	Cx26	MRI	Visual
		move	(mo)	n = 18	n = 11	issues	n = 13	n = 20		number	mutations	n = 12	function
1	0	0	12	50	1	0	-	135	5	7.6·10 ⁶	0	-	1
2	1	3	14	1	2	1	75	69	4	3.0·10 ⁶	0	1	2
3	1	2	19	-	-	0	-	12	2	8.3·10 ⁶	1	1	1
4	1	5	Nw	-	-	4	-	-	0	NA	0	3	1
5	0	6	20	25	1	0	75	164	5	1.0·10 ⁴	0	-	1
6	0	2	15	2	-	0	25	75	5	6.5·10 ³	0	-	1
7	0	2	18 ^s	-	-	0	-	28	3	3.4·10 ³	0	1	1
8	0	5	36	0.1	-	3	-	-	1	>20·10 ⁶	0	-	1
9	1	6	19	1	1	1	50	172	4	4.5·10 ⁴	0	-	1
10	0	4	24	0.1	2	0	10	179	5	10·10 ⁶	0	2	1
11	1	3	25	1	-	0	90	128	5	3.2·10 ³	0	2	2
12	0	5	21	1	-	1	10	136	5	9.7·10 ⁶	0	-	1
13	0	3	22	0.5	-	3	-	180	5	1.7·10 ⁶	0	-	2
14	0	3	24	-	3	3	-	-	1	3.6·10 ⁴	0	-	1
15	0	3	18	9	-	0	5	98	3	7.5·10 ³	0	1	2
16	0	0	10	63	-	0	-	134	5	NA	0	-	1
17	1	5	23	0.5	1	4	75	138	5	>50·10 ⁶	0	-	1
18	0	3	18	-	-	0	-	-	1	>30·10 ⁶	0	1	1
19	0	3	12 ^s	-	0	3	-	12	2	2.1·10 ³	0	-	1
20	0	0	12 ^s	9	3 ²	0	25	37	3	3.8·10 ³	0	1	1
21	0	4	24	5	-	4	50	51	4	1.2·10 ⁶	0	1	1
22	0	3	18 ^s	0.5	-	1	75	123	5	3.1·10 ³	0	-	2
23	0	3	22	1	-	0	90	116	5	7.3·10 ⁶	0	1	1
24	0	3	Nw	-	2	0	-	-	2	1.3·10 ⁶	0	0	1
25	0	4	Nw	-	-	3	-	-	2	14.8·10 ⁶	0	0	1
26	0	4	18 ^s	9	2	0	-	123	5	2.6·10 ⁶	0	-	1
Mean			19***	10**			50	106	3.5				

Cx26	Neo	Path	Age of	Movement	Vestibular					DBS			
Case	Sympt	head	walking	ABC-2	function	Feeding	Raven	PPVT-3	SIR-2	copy	Cx26	MRI	Visual
		move	(mo)	n = 9	n = 0	issues	n = 7	n = 8		number	mutations	n =	function
1	0	0	12	-	-	0	-	12	3	-	3	-	1
2	0	0	14	-	-	0	-	15	2	-	3	-	1
3	0	0	13	5	-	0	-	-	2	-	3	-	1
4	0	0	17	-	-	0	-	-	2	-	3	-	1
5	0	0	12	50	-	0	75	130	5	-	3	-	1
6	0	0	12	-	-	0	-	-	4	-	3	-	1
7	0	0	12	25	-	0	50	73	5	-	3	-	1
8	0	0	11	98	-	0	90	146	5	-	2	-	1
9	0	0	12	63	-	0	-	-	5	-	3	-	1
10	0	1	10	50	-	0	95	137	5	-	3	-	1
11	0	0	12	37	-	0	50	-	4	-	3	-	1
12	0	0	11	16	-	0	90	45	5	-	2	-	1
13	0	1	14	63	-	0	95	100	5	-	3	-	1
Mean			12***	45**			78	82	4				



Five-To-Fifteen questionnaire (FTF)

Percentages of associated difficulties



Conclusion

- Balance impairment (86 %)
 - Late walking age, head tossing, low results on Movement ABC-2
- Neurodevelopmental disabilities (15% ASD)
- Visual impairment (20%)
- Feeding problems (46%)