

# The brain network during cataplexy in children

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**BACKGROUND & OBJECTIVES:** Cataplexy is typically triggered by positive emotions. Animal data suggest that the amygdala and the anterior cingulate cortex are key regions in promoting emotion-induced cataplectic attacks. Here, we investigated the neural networks associated with laughter-induced cataplexy using the BOLD signal changes that occurred while viewing prolonged funny videos in a “naturalistic” fMRI protocol in drug-naïve children and adolescents with Narcolepsy type 1 (NT1).

**METHODS:** 21 drug-naïve patients with NT1 (13 males, mean age 11 years) (**Table 1**) were studied with functional MRI while viewing funny videos (**Figure 1**). SPM8 software was used for whole-brain fMRI data analysis. Whole brain hemodynamic correlates of (a) sign of fun and amusement (laughter) and of (b) cataplexy were analyzed and compared.

**RESULTS:** Emotion-induced laughter occurred in 16 patients, 10 showed cataplexy for a total of 77 events (mean duration = 4.4 sec). The contrast *laughter>baseline* shown an increased hemodynamic response in the bilateral motor/premotor cortex and anterior cingulate gyrus. The contrast *cataplexy>baseline* demonstrated suprapontine BOLD signal increase in the amygdala, frontal operculum – anterior insular cortex, ventromedial prefrontal cortex, and in the nucleus accumbens. For the contrast *cataplexy>laughter*, an higher hemodynamic demand over the bilateral (> right) opercular-insular cortex (global maxima), the right amygdala, the bilateral ventral striatum and the right nucleus accumbens was observed.

Table 1: Clinical, polysomnographic and laboratory data of investigated patients.

Pt. N°	Age (yrs) /Sex	Disease duration (yrs)	Time since cataplexy onset (yrs)	Cataplectic attack frequency*	MSLT (min)	MSLT-SOREMPs	CSF hcrf pg/mL	ESS*	HLA-DQB1*0602
1	13/F	2	1	1/week-1/day	3.30	5	10.7	13	p
2	11/M	4	2	1/yr-1/month	9.00	5	43.1	13	p
3	14/F	3	2	1/week-1/day	2.40	5	0	13	p
4	10/M	2	1	>1/day	1.90	4	0	21	p
5	10/F	4	2	>1/day	1.20	1	15.5	14	p
6	9/M	0	0	>1/day	1.90	5	0	16	p
7	15/M	5	2	1/week-1/day	3.30	4	0	17	p
8	9/F	0	0	>1/day	4.40	2	0	14	p
9	12/F	1	1	>1/day	1.70	3	33	10	p
10	11/F	1	1	1/week-1/day	1.00	5	0	14	p
11	11/M	3	2	>1/day	9.80	4	25	14	p
12	11/M	3	1	1/week-1/day	2.20	5	13.6	11	p
13	13/M	3	2	>1/day	13.00	4	55	19	p
14	11/F	1	1	>1/day	8.90	3	84.2	12	p
15	7/F	1	1	>1/day	9.60	1	0	20	p
16	16/M	4	2	1/week-1/day	2.40	5	0	13	p
17	14/M	2	1	>1/day	2.00	5	10.5	21	p
18	8/M	1	0	1/week-1/day	4.00	1	0	14	p
19	15/M	3	1	>1/day	7.00	3	0	12	p
20	9/M	0	1	1/week-1/day	3.50	4	20	9	p
21	8/M	2	2	>1/day	2.00	2	40	18	p

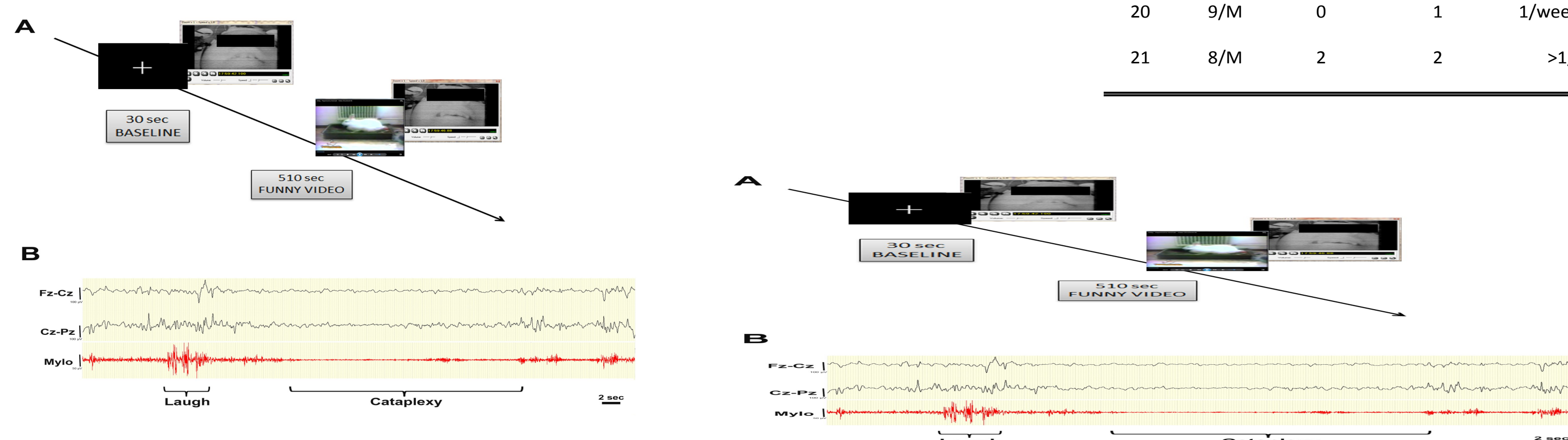
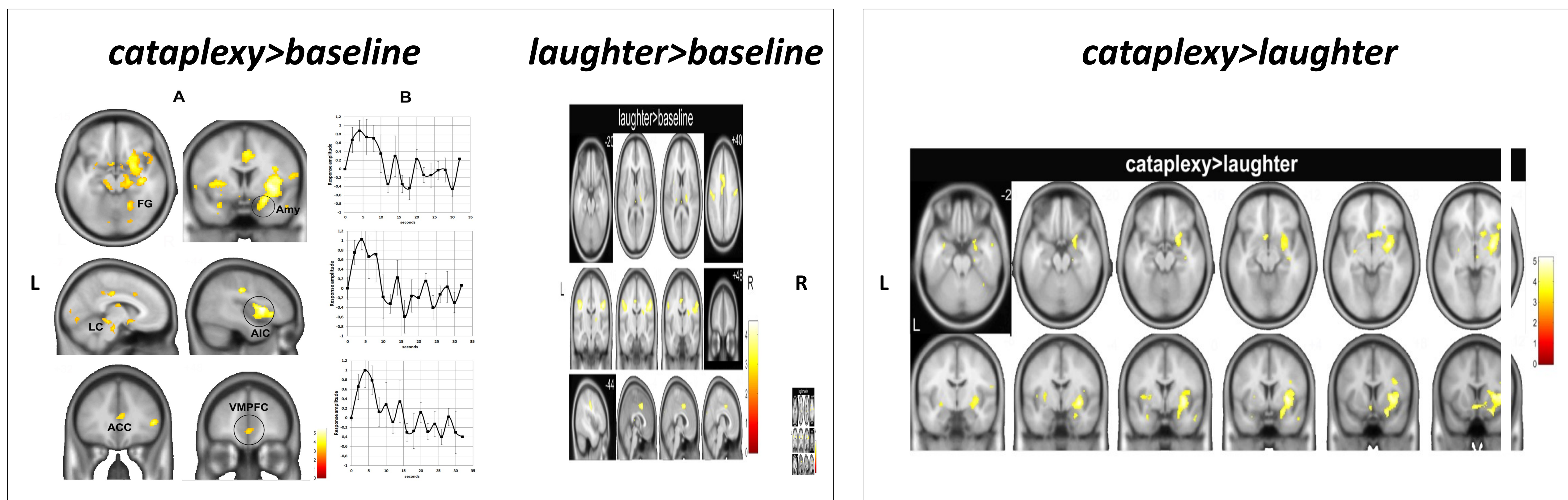


Figure 1: EEG-fMRI protocol



**CONCLUSIONS:** Cataplexy in NT1 occurs with an increased neural activity of the anterior insular and cingulate cortices, and in particular of the amygdala, that in turn could inhibit pontine structures mediating muscle tone.