(12y) with PHS, Panhypopituitarism and HH. She developed an DRE with multiple seizure types, cognitive disability, severe aggressive and disinhibited behaviour and interictal psychosis, resulting in DEE. Presurgical epilepsy diagnostic showed large bilateral confluent HH, highly active epilepsy and high risk of surgery complications.

Objectives: Clarifying therapeutic options in patients with DRE/DEE due to HH in terms of seizure reduction, cognitive and psychiatric outcome.

Methods: A bibliographic search on PubMed was performed (last checked on 31 October 2024).

Results: Risk factors for cognitive impairment are earlier onset of epilepsy, increased seizure burden, increased number of antiseizure medications, and, for some, larger lesion burden. Epilepsy surgery is the gold-standard treatment for DRE. Open microsurgical resection provides the best outcome in seizure reduction (90 % Engel I+II) and halts neurocognitive/-psychiatric symptoms (88 % of patients). Due to higher complication rate, minimal-invasive techniques are common and achieve high level of seizure control: Endoscopic resection bipolar, radiofrequency, or laser coagulation (77 % Engel I+II), Stereotactic radiofrequency thermocoagulation (88 % Engel I) with improved IQ (69 % of patients) and resolution of behavioral problems, Laser interstitial thermal therapy (93 % Engel I), Gamma Knife radiosurgery (57 % Engel I + II).

In cases of large hamartomas with extensive attachment to the hypothalamus epilepsy surgery is not feasible. Therefore, deep brain stimulation (DBS) has been utilized by targeting HH, anterior thalamic nucleus (ANT) or mammillothalamic tract. Two patients were successfully treated and simultaneous stimulation of the ANT and HH has also been reported with resolution of gelastic seizures and reduction of focal seizures.

Stereo electroencephalography demonstrated an intrahypothalamic epileptogenic zone for gelastic seizures, whereas other seizure types were related to discharges affecting cortical regions, triggered by HH, forming complex epileptogenic networks. Encephalopathy may be mediated by disruption of hypothalamic projections and distributed networks.

Conclusions: Hamartoma removal is linked to improved cognitive and behavioral outcomes and early surgical treatment can minimize the risk of DEE. We propose to widen the indication for early epilepsy surgery in cases of HH with severe behavioral dysfunction, even in patients with good seizure control, and considering risk factors for developing DEE. Early HH-diagnosis and further studies on risk stratification for DEE are required. DBS can be an effective therapy option in inoperable patients as our patient might be. Further data concerning therapy of our patient will be submitted at the time of conference.

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Investigating expectation effects on face detection: a Bayesian model-based fMRI analysis

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Background: Visual illusions arise from a delicate balance of bottom-up sensory input and top-down expectations. While there is extensive literature on many visual illusions, the picture is less clear for face-related illusions. While the stimulus-bound bottom-up perspective has been explored extensively for face-like stimuli, the expectancy-driven top-down aspect has received much less attention. Here, we leverage the predictive processing framework (PP) to formalize cognitive expectancy effects during face-related visual illusions. The PP framework divides cognitive processes conceptually into three phases: forming predictions about future stimuli, calculating the prediction error, and updating beliefs in light of new observations.

Objectives: As an instance of face-related visual illusions, this work aims at delineating expectancy effects driving (illusory) face detection (IFD or face-pareidolia) using a Bayesian model-based fMRI analysis approach.

Question: Specifically, we aimed to identify functional specialization across a population of traditionally face-sensitive neural regions of interest (ROIs), i.e., the 'core-' and 'extended' face networks, through latent-process signals extracted from a PP-consistent cognitive-computational model. Finally, our goal was to identify a single trial-bytrial model parameter per PP phase that best explains IFD-associated BOLD activity across ROIs.

Methods: A pure-noise illusory face detection task required participants to indicate whether they perceived a face in a pure-noise stimulus array. First, fMRI data of face-sensitive regions was collected during the IFD task and converted to an ROI-averaged trial-by-trial beta-series. Second, the subject behavior of the IFD task was modeled with a hierarchical cognitive-computational model called the hierarchical Gaussian filter (HGF). The HGF allows extracting trial-by-trial model parameters (i.e., estimates of latent cognitive processes) from all three phases. Finally, we used Bayesian hierarchical linear models to fit the extracted model parameters to the beta-series.

Results: Consistent with the expectancy-driven nature of IFD, we identified multiple trial-by-trial model parameters corresponding to two of the three PP phases through a model selection procedure. A parameter from the prediction-forming phase, i.e., the probabilistic expectation to perceive an (illusory) face in the upcoming trial, positively predicted BOLD-activity in the right inferior occipital gyrus (IOG, includes the occipital face area). Notably, a parameter of uncertainty in this probabilistic expectation was negatively associated with BOLD activity in the left IOG and positively associated with the right inferior frontal gyrus (IFG). A parameter from the prediction-error phase, i.e., a measure of prediction error, was associated negatively with activity in the left IOG.

Conclusions: In the face domain, lateralized expectancy-driven functional specializations appear within face-selective cortical regions. The right IOG appears to represent probabilistic expectations of future face stimuli. In contrast, the left IOG seems sensitive to prediction uncertainty and expectation violations, with BOLD activity inversely related to both measures. In contrast, activity in the right IFG positively scales with a measure of prediction uncertainty, likely representing activity associated with statistical learning and prefrontal modulation. This work provides a new perspective into the functional specialization of face-selective cortical regions.

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