

fNIRs studies in Glut-1

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Functional Near-Infrared Spectroscopy or fNIRs is a noninvasive neuroimaging technique. The advantages of fNIRs is that it is inexpensive and portable and can provide real time evaluation of brain function. fNIRs works by optical imaging in the near infrared spectrum of light which is 650-950 nm. A subject is presented a motor or cognitive task, and the area of the brain extracts oxygen which is carried on our blood cells. This converts the oxygenated hemoglobin to deoxygenated hemoglobin in the brain which is an indication of brain activity. fNIRs is similar to fMRI in that regard, except there are some differences. There is low sensitivity to motion that makes it attractive for those who are not able to lie still in the scanner such as children. It has good temporal resolution but because the light has to penetrate the hair and scalp, so it cannot measure deep activity. A NIRs circuit is composed of a light emitter and a detector as shown in the figure. These are placed onto a cap that the subject wears while performing tasks on a computer. The tasks may be motor or language. When an area of the brain is involved in the task, oxygen is abstracted from our red blood cells in the blood that flows there and causes a change in the wavelength of light that causes a signal to be emitted through the detector. We can map out the pathways in the brain that are activated with tasks and compare Glut-1 to controls or another metabolic disorder to look at what is specific to that condition. We have done this with another condition called ornithine transcarbamylase deficiency and shown that the damaged areas of the brain don't activate, but new areas do, like the brain is sending in reinforcements. We have done a preliminary study in 6 patients with Glut-1 prior to the conference and showed that when the subjects view a 5 minute video of Pippa pig, which has real language, and Timmy Time, that has sounds but no words, there are differences in brain activation. We were fortunate to collect another 11 subjects at the conference so we can analyze what this means and if we can use this device to look at responses to therapies such as the ketogenic diet. Thanks to those who participated, and I

think they will agree that it was not hard to wear the device on the head.
The figure is showing the activation of the total hemoglobin signal which is more in Glut-1. The brain is working ha



