

Brain hemodynamics in patients with MSUD using Functional Near-Infrared Spectroscopy (fNIRS)

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Introduction

- **Maple Syrup Urine Disorder (MSUD)** is an inherited disease.
- Patients experience brain dysfunction.
- Brain biomarkers are helpful for diagnosis.

- **Functional Near-Infrared Spectroscopy (fNIRS)** is a noninvasive brain imaging method with low sensitivity to motion.
- Brain imaging is feasible in children using fNIRS.
- fNIRS can produce a map of the brain activity.

Goal

- Test if fNIRS can detect brain activity in patients with MSUD.
- Compare them with normal



What is fNIRS?



Non-invasive neuroimaging technique

Inexpensive

Portable

Not sensitive to motion

It measures changes in blood hemoglobin signals

Similar to fMRI

Why fNIRS?

- Performing measurements on patients with rare diseases requires planning and travel
- fNIRS can provide a unique opportunity to monitor rare diseases

The fNIRS Device

NIRSport2 from NIRx



Different cap sizes for all ages and head sizes

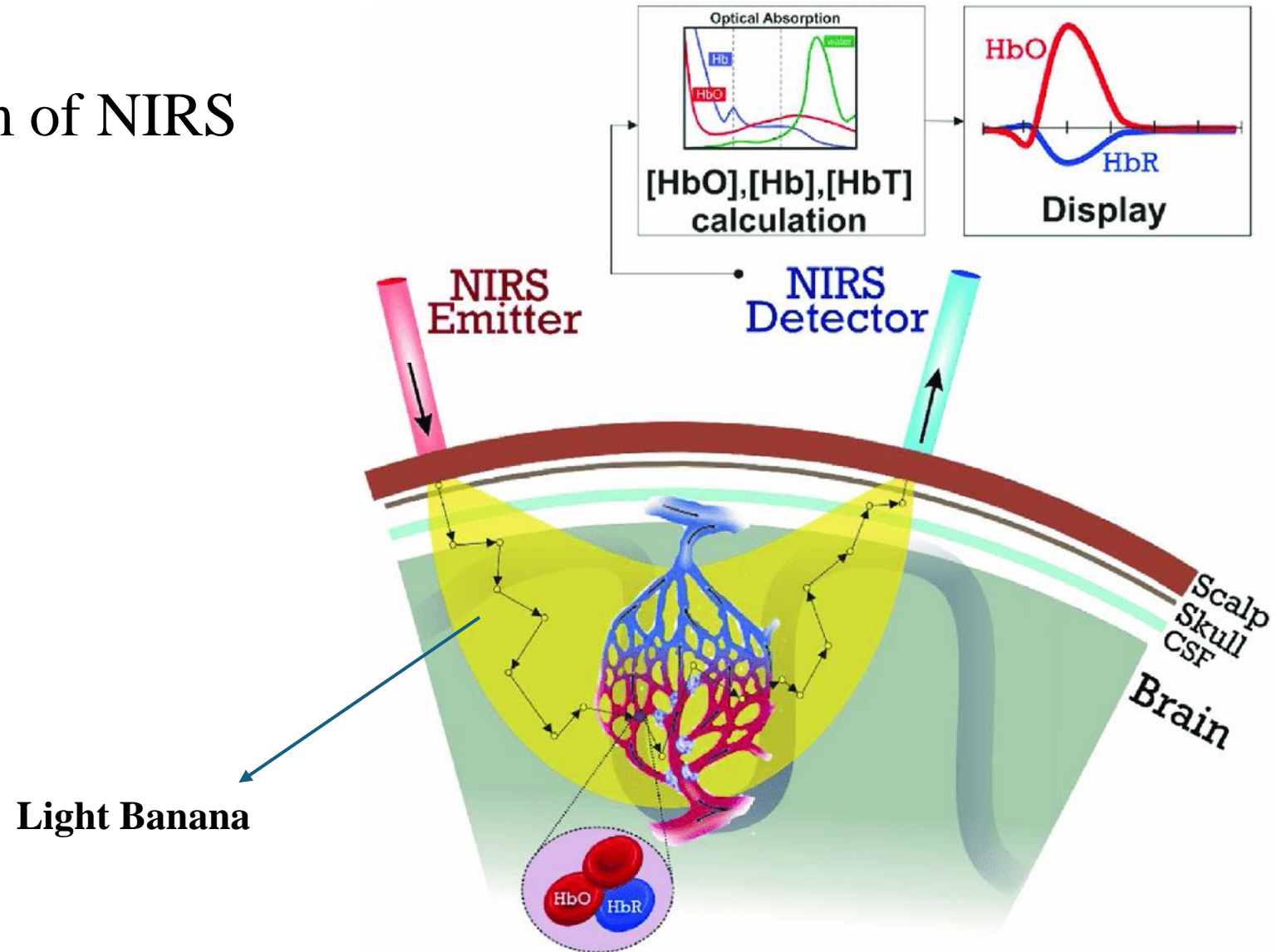
- Wireless (can be used wired)
- Uses Near-Infrared light:
Wavelengths 850 and 760 nm
- Eye safe
- 16 sources, 16 detectors, 44 channels
- Sampling frequency: 5.1 Hz



A fNIRS channel

Schematic diagram of NIRS

Oxyhemoglobin
Deoxyhemoglobin
Total Hemoglobin



MSUD/fNIRS protocol

Children's National Hospital in collaboration with MSUD family support group

Title: Brain function examination in patients with maple syrup urine disease (MSUD) using functional near infrared spectroscopy (fNIRS)

Subjects: 20 MSUD patients and 20 age and sex matched controls

Tasks:

- 8 years old and younger: watch cartoons
- Older than 8 years old: N-back task



- **Subjects**

- So far → Patients: 2 Controls: 4
- Symposium → Patients: 8 Controls: 2

- Full siblings with a genetically confirmed diagnosis of MSUD
-
- P1 is a 5-year-old male
- P2 is a 4-year-old female

Methods

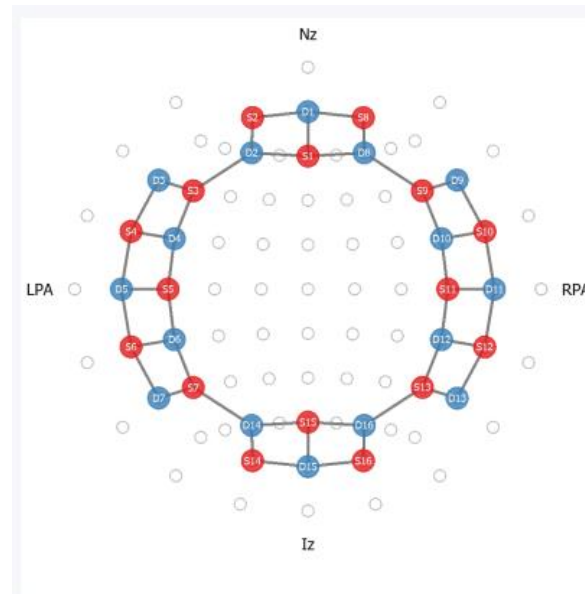
Our designed configuration:

16 sources

16 detectors

44 channels

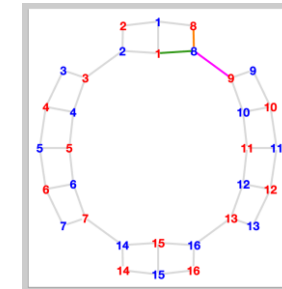
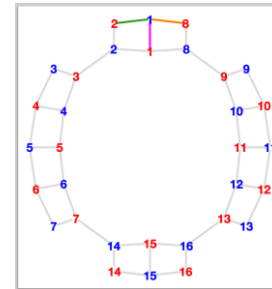
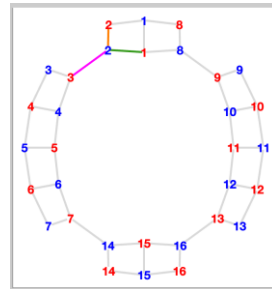
Cap Configuration



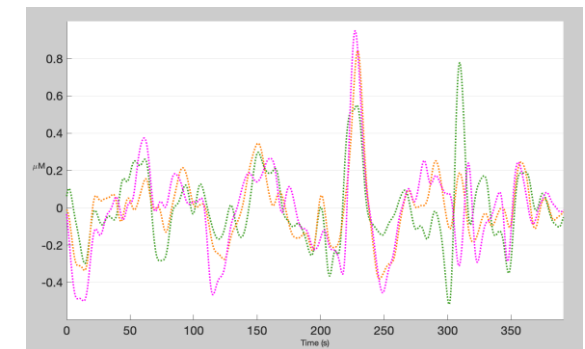
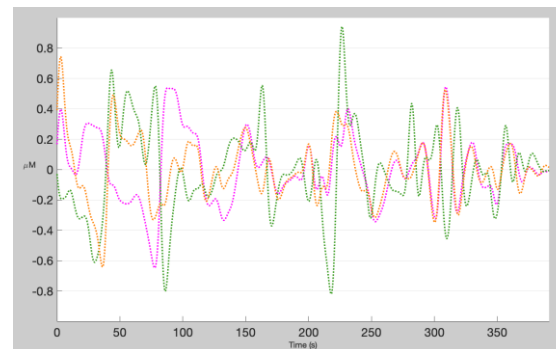
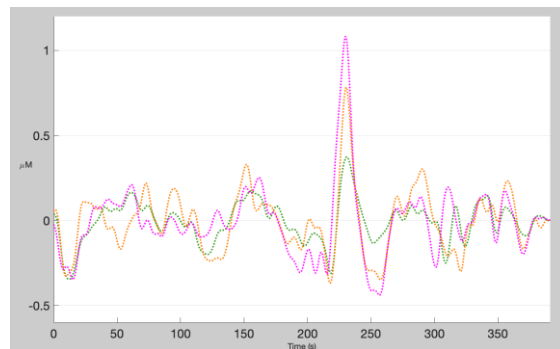
Data Analysis

- A 5-minute of Timmy Time cartoon was considered as a baseline for data analysis
- Deoxyhemoglobin signals of forehead were analyzed

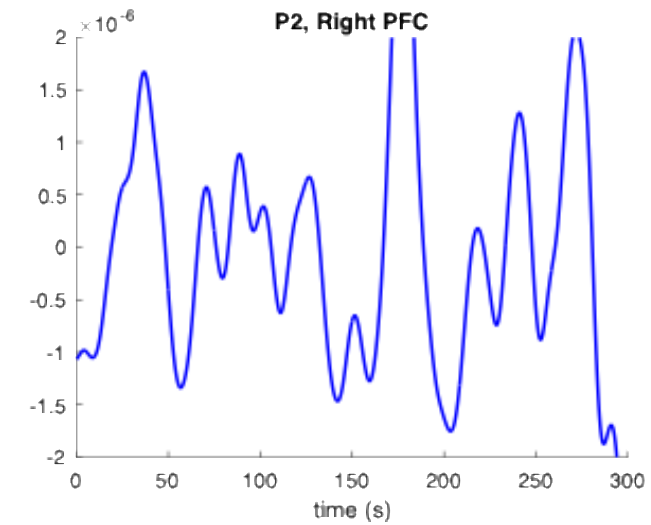
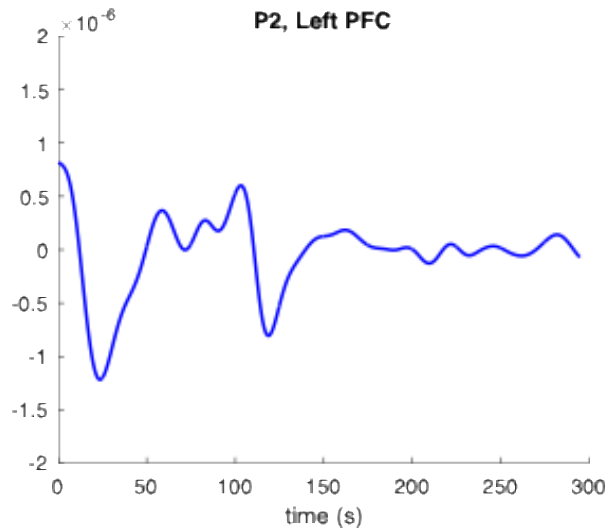
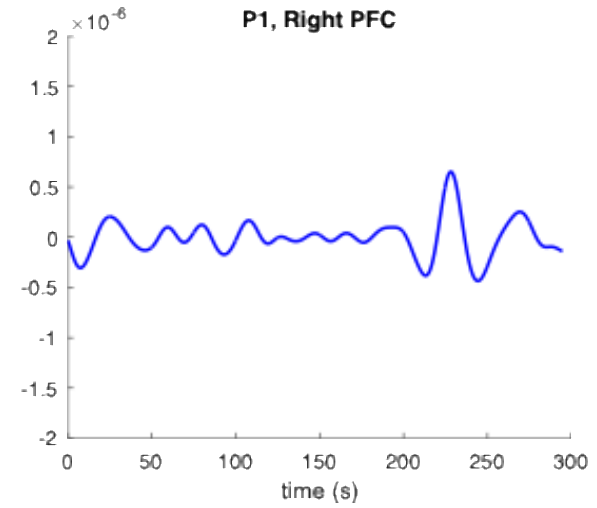
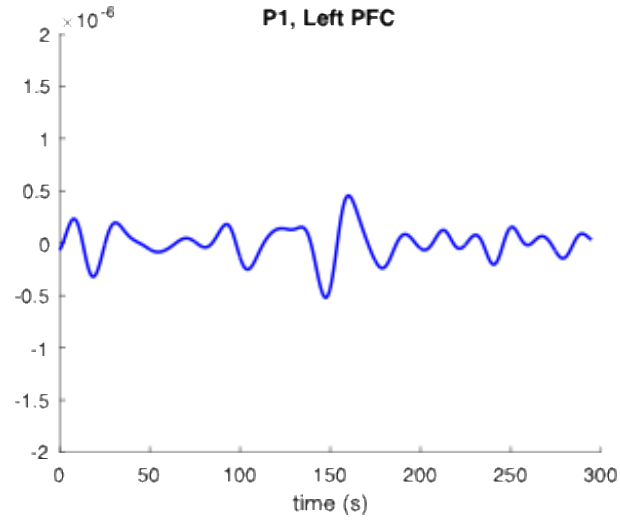
Channels on the PFC



HOMER3 output:
Deoxyhemoglobin



Results



Conclusion

fNIRS can detect
brain activity in
MSUD patients.

We will have more
disease biomarkers.

It will help
physicians to
diagnose and treat
MSUD disease.

Future Direction

An on-going project. We will work on the data.

We are planning to recruit 40 subjects. 20 MSUD patients and 20 normal subjects.

We will be collecting data at the MSUD symposium.

All channels will be considered.

Compare MSUD with normal subjects.



Children's National.

Our team

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Thank you!

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