

Emotional recognition, experience and empathy in persons with Moebius syndrome and persons with congenital bilateral facial paralysis.

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Ethical review	Approved WMO
Status	Recruitment stopped
Health condition type	Other condition
Study type	Observational non invasive

Summary

ID

NL-OMON31744

Source

ToetsingOnline

Brief title

Moebius project

Condition

- Other condition
- Neurological disorders congenital

Synonym

facial paralysis, Moebius syndrome

Health condition

healthy subjects

Research involving

Human

Sponsors and support

Primary sponsor: Universitair Medisch Centrum Groningen

Source(s) of monetary or material Support: Ministerie van OC&W,NWO

Intervention

Keyword: Emotional recognition, Facial expression recognition, Functional magnetic resonance imaging, Moebius syndrome

Outcome measures

Primary outcome

The primary study parameters involved are listed below. Full details of the experiments can be found in the protocol.

1. PSYCHOMETRIC AND EMOTIONAL RECOGNITION

The core goal of our study is to investigate the recognition of emotional and facial expressions in the Möbius and congenital bilateral facial paralysis populations even though they cannot perform these movements themselves. To this end, we would therefore like to investigate:

- a. The extent to which Möbius and congenital bilateral facial paralysis research participants recognise facial expressions in others
- b. The threshold for that recognition
- c. The extent of the ability to generate visible facial expressions and emotions

Through the use of various tools we can test the following:

- Empathy (Davis Interpersonal Reactivity Questionnaire)
- Emotional functioning (Bermond-Vorst Alexithymia Questionnaire)
- General intelligence (Raven Progressive Matrix Questionnaire)
- Expression and emotional recognition (Montagne-Perrett Morphed Faces).

Including a group of healthy control volunteers throughout the investigation will provide information about the psychological, intellectual, emotional and empathic functioning of the Möbius research participants compared to the general population.

Montagne-Perrett Morphed Faces program quantifies the capacity of the participant to associate an emotional label to facial expressions. Using it, we will examine if the capacity to perform facial expressions (which is impaired in congenital bilateral facial paralysis) influences our sensitivity to recognise emotions.

2. NEUROLOGICAL TESTING

There has previously been some in depth electrophysiological testing carried out on Möbius patients showing a certain variability in how much motor capacity they have in their face. We will therefore need to quantify facial motility and somatosensory sensitivity in our Möbius and congenital bilateral facial paralysis participants, and in our control group to investigate if the processing of other individual*s faces is affected by the participant*s facial motor and somatosensory capacities.

For this purpose we will run a number of simple non-invasive tests:

- a. Semmes-Weinstein Monofilament Test
- b. Facial Motility (producing a smile and a frown)
- c. Ocular Motility (the Four Point Eye Movement Test)
- d. Visual Acuity (Warrington Face Memory Recognition Test)

3. fMRI STUDY

It has been shown by our research and that of others that neuroimaging using fMRI is a valuable tool to investigate the areas of the brain activated whilst observing the actions and emotions of others. Therefore, in the neuroimaging (fMRI) section of the design we would like to measure the premotor and somatosensory areas whilst viewing facial expressions that the research participants cannot produce (such as smiling) as well as movements, such as sucking through a straw, that they can. We intend to provide these facial expressions in the form of short 3s films of actors displaying the facial expressions of happiness, disgust, fear, sucking through a straw and neutral. A concomitant aim of this section of the design is also to determine the location of the premotor and somatosensory areas in the subjects when they are asked to execute emotional gestures and sucking movements.

The neuroimaging (fMRI) is composed of two substudies:

- a. Viewing sucking through a straw and facial expressions
- b. Facial Movement Execution

Our lab has extensive experience with this type of design, and has found a number of cortical areas to be involved in both viewing and executing facial expressions in subjects without bilateral facial paralysis so there is pilot data in support of this.

Secondary outcome

n.v.t.

Study description

Background summary

Social cognition, or the way in which people make sense of other people and themselves, is a crucial element of social interactions. We often effortlessly understand what goes on in other people despite the fact that their goals and feelings are hidden from outside sight, inside their own brains. The simple act of being able to smile or frown to indicate pleasure or annoyance is one that happens spontaneously in the majority of the population. If we are able to understand the emotions of others by sharing their facial expressions, the intriguing question arises as to what happens in individuals who are unable to make facial expressions. We have identified such a group of people: those with congenital bilateral facial paralysis and specifically Möbius Syndrome.

Möbius Syndrome is a condition present from birth affecting mainly the 6th and 7th cranial nerves. It is defined as congenital facial weakness combined with the impaired ability to move the eyes to the side, and often includes physical deformities such as clubfeet, webbed digits, drooling, and an abnormally small jaw. The most striking feature of patients with Möbius syndrome is the mask-like faces due to the affected seventh cranial nerve.

It is only recently that a more definite diagnosis of Möbius syndrome has been agreed upon as *congenital facial palsy with impairment of ocular abduction*. Until the advent of the genetic studies (Verzijl et al 2003) it was believed that a group containing what is now known as *congenital facial palsy* (in some cases hereditary) also had Möbius syndrome. The genetic background now appears to be different: Möbius syndrome arises from a development disorder of the lower brainstem; hereditary congenital facial palsy does not. The latter is a general nuclear development disorder. We have also come to understand that since Möbius is such a rare disorder its diagnosis may often take years before it is recognised. The afflicted persons are labelled as having congenital

bilateral facial paralysis. For our purposes, we are inherently interested in the fact that the person cannot perform facial expressions.

There has been some research carried out on Möbius patients, but the vast majority of this has been based on medical analysis i.e. which nerves are affected, possible genetic factors and the resulting physical deformities. There has been very little emotional or psychological research done in this area.

Understanding others by automatically *putting yourself into someone else*s shoes* is an essential component of social life, as it gives an intuitive nature to the relationships between humans. This is basically, in lay terms, what we mean by empathy. Looking at the normal development of empathy more specifically, it is not considered to be innate in life. However, this function seems to develop gradually while persons participate in daily social life: it involves acting yourself (and by doing this, experiencing emotions) and observing actions of other people. Coupling other peoples* emotions with your own experience - which is the basis of empathy - takes place by means of communication (e.g. facial expressions, (body) language). People with Möbius Syndrome and congenital bilateral facial paralysis are unable to perform facial expressions themselves, so does this mean that they are deprived of the capacity to share the facial expressions of other people? In other words, are the internal neural circuits still activated, despite the fact that the responding facial expression cannot be generated?

The investigation of social cognition and its dysfunctions has long been dominated by the idea that a specific module is responsible for our capacity to attribute mental states to others (Theory of Mind module). In the last decade however, our understanding of how we understand the actions, emotions and sensations of others has significantly changed. In this newly emerging model, understanding others can be achieved through two normally complementary routes: one automatic and based on an internal simulation of the actions, emotions and sensations of the other (*shared circuits*), the other, more deliberate and cognitive, is based on the entertainment of explicit thoughts about the beliefs and emotions of others .

We have shown that neurons in the premotor cortex of the monkey respond both to the monkey*s own execution of hand actions and to the sight of someone else performing similar actions. This is true, even when the actions of others can only be heard or guessed. These neurons appear to translate the sight of someone else*s actions into a language well known to the observer: his or her own actions. This translation creates a strong and intuitive link between the observing and the observed individual. Recently, we have shown that in analogy to these mirror neurons, the observation of someone else*s disgust triggers activity in the anterior insula in voxels also activated while the subject himself is being disgusted in the scanner. Lesions in the anterior insula have been shown to decrease both ones ability to feel disgusted, and to recognise

disgust in the facial expression of others showing the importance of this system in social cognition. We have also shown that observing someone else being touched activates the observer's secondary somatosensory cortex as if he/she had been touched. Together this body of evidence suggests that understanding others may rely not only on the existence of a domain unspecific theory-of-mind module, but also on a series of systems (e.g. premotor cortex, insula and SII) normally involved in our own actions, emotions and sensations, that we will call 'shared-circuits'.

Study objective

The core approach of our proposed project is to investigate three aspects of emotional recognition in the Möbius and congenital bilateral facial paralysis populations through observed facial expressions, and the possible link between them, as follows:

- a. By quantifying the degree of facial paralysis (deafferentiation) and somatosensory sensitivity
- b. By quantifying the capacity to recognise emotions from observed facial expressions
- c. By quantifying the degree to which the motor somatosensory areas are activated whilst watching emotional facial expressions during neuroimaging (fMRI) studies.

The overall goal of this proposed project is to explore the link between the capacity to generate facial expressions and the capacity to recognise emotions displayed through facial expressions in others. Through this research we hope to provide better understanding of the cognitive impact of congenital bilateral facial paralysis to help patients deal with the challenges of their condition that may not yet have been recognised.

Study design

The proposed study is separated into two distinct participatory sections: Part I and Part II. Part I is to be carried out in the volunteer's own home; Part II takes places at the Neuroimaging Centre in Groningen. Research participants can elect to take part only in Part I or in Part I AND Part II.

PART I

To be carried out in the volunteer research participant's home involves:

Davis Interpersonal Reactivity Index Questionnaire

De Bermond-Vorst Alexithymia Questionnaire

Raven Progressive Matrix Questionnaire

Neurological testing: Semmes-Weinstein Monofilament Test

Ocular Motility: Four Point Eye Movement Test

Visual Acuity: Warrington Face Memory Recognition Test

Emotional expression recognition: Montagne-Perrett Morphed Faces Program (Montagne et al. 2007)

Emotional/facial expression generation: filming of participant generated facial expression corresponding to happiness, disgust, fear, neutral, and the motor function of chewing

PART II

To be carried out at the Neuroimaging Centre, University Medical Centre Groningen involves:

Neuroimaging (fMRI) studies whilst participant views short film clips of emotions

Neuroimaging (fMRI) studies whilst participant executes emotional gestures and chewing movements

Rating of the film clips (as seen during the scan) after completion of the fMRI scan using the basic six emotions

Autism Diagnostic Observation Schedule (ADOS) Questionnaire/Interview

Study burden and risks

BENEFIT/GROUP RELATEDNESS

Through this research we hope to provide better understanding of the cognitive impact of Möbius syndrome and congenital bilateral facial paralysis to help patients deal with the challenges of their condition that may not yet have been recognised. We also want to elucidate further the areas involved in the brain in empathic functioning.

BURDEN

As discussed in E9 and E9a there is minimal burden to participants in this study.

RISKS

The extent of the minimal risks are due to the MRI scanning section of the proposed project, and so far no side effects have been described in the literature. We will responsibly advise participants of these (as in E7), and they also have access to our independent medical expert in the event of any worries or queries.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age

Adults (18-64 years)
Elderly (65 years and older)

Inclusion criteria

For the Moebius Participants and congenital bilateral facial paralysis the following inclusion criteria are required for Part I of the study:

- * Males and females between the ages of 18 and 65 years
- * Normal visual acuity or corrected to normal vision
- * The participants must be willing and able to participate in the measurements
- * The participants must have given written informed consent to be included in the study
- * Clinical diagnosis of Möbius Syndrome or congenital bilateral facial paralysis from a registered neurologist; For the Volunteer Control Participants for Part I, the following inclusion criteria are required:

- * Males and females between the ages of 18 and 65 years
- * Normal visual acuity or corrected to normal vision
- * The participants must be willing and able to participate in the measurements
- * The participants must have given written informed consent to be included in the study; We shall begin by scanning for Part II the first SIX fMRI suitable volunteers who apply. Depending on the results (see section 5.3) from this, this will determine how we will proceed further with Part II.

Exclusion criteria

For the Moebius Participants and congenital bilateral facial paralysis in Part I, the following exclusion criteria apply:

- * Those suffering from schizophrenia
 - * Those suffering from a neurological disorder not related to Möbius e.g. epilepsy, Parkinson*s disease, and Huntington*s disease.;
- For the Moebius Participants and congenital bilateral facial paralysis in Part II of the study the following criteria apply:

- * Visual disorder that cannot be corrected through the use of corrective lenses to a level of 20-40 in both eyes (we have MRI safe glasses for a very broad range of prescriptions)
 - * Another significant CNS disorder (e.g. brain damage, epilepsy)
 - * Pregnancy or suspected pregnancy
 - * Those suffering from claustrophobia and/or a panic disorder
 - * Subjects who do not fulfil the criteria for participating in an fMRI assessment (e.g. people who have metal implants (pacemaker, heart valves, vascular clips, eye-implants or piercing))
 - * Wishes not to be informed of brain abnormalities that may be noticed in the scans
 - * If they are unable to suck through a straw;
- For the Control Participants in Part I the following exclusion criteria apply:

- * Those with signs of facial paralysis (as assessed from the rated video-taped facial expressions)
 - * Those suffering from a neurological disorder
 - * Those with a psychiatric history (as assessed by the Suitability Questionnaire in Appendix VA of the protocol);
- For the Control Participants in Part II, the following exclusion criteria apply:
- * Visual disorder that cannot be corrected through the use of corrective lenses to a level of 20-40 in both eyes (we have MRI safe glasses for a very broad range of prescriptions)
 - * Another significant CNS disorder (e.g. brain damage, epilepsy)
 - * Pregnancy or suspected pregnancy
 - * Those suffering from claustrophobia and/or a panic disorder
 - * Participants who do not fulfil the criteria for participating in an fMRI assessment (e.g. people who have metal implants (pacemaker, heart valves, vascular clips, eye-implants or piercing))
 - * Wishes not to be informed of brain abnormalities that may be noticed in the scans

Study design

Design

Study type:	Observational non invasive
Intervention model:	Other
Allocation:	Non-randomized controlled trial
Masking:	Open (masking not used)

Control:	Active
Primary purpose:	Other

Recruitment

NL	
Recruitment status:	Recruitment stopped
Start date (anticipated):	07-08-2008
Enrollment:	50
Type:	Actual

Ethics review

Approved WMO	
Application type:	First submission
Review commission:	METC Universitair Medisch Centrum Groningen (Groningen)

Study registrations

Followed up by the following (possibly more current) registration

No registrations found.

Other (possibly less up-to-date) registrations in this register

No registrations found.

In other registers

Register	ID
CCMO	NL20869.042.07