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1. Introduction

This clip, aired on the 'Today's Health' segment of the Today Show, tells the story Cameron Mott. At the age of three Cameron began to suffer from persistent symptomatic seizures. She was diagnosed with the rare autoimmune disease known as Rasmussen's Encephalitis (referred to as Rasmussen's Syndrome in the video) which causes the destruction of one hemisphere of the brain. As the disease progressed over the next three years her cognitive intelligence declined and she could barely talk. Cameron's condition resisted drug treatment and so at around the age of 6 she underwent a hemispherectomy – a removal of one hemisphere of the brain – in order to prevent seizures and further cognitive impairment. The result of the hemispherectomy was initial left hemiparesis. Cameron underwent 10 hours of intense physical therapy per week and in less than a month was able to walk. The post-surgery video shows her running, twirling and playing in a child's playground and she is documented to be performing well in school. She can also be heard talking fluently. Cameron is reported to have been seizure free since the hemispherectomy.

We chose this news excerpt because of its relevance to some of the broad areas of study in the Neuroscience Fundamentals course. These areas include seizures, neurotrauma and neural plasticity. The excerpt was also of interest because of the opportunity it afforded us to research an extremely rare autoimmune disease and to investigate current research and information on neural plasticity, a fundamental aspect of neuroscience which is of increasing importance to modern medical treatment.

Our research indicates that the information presented on Rasmussen's Encephalitis, surgical hemispherectomy and neural plasticity, while being simplified for the target audience, is quite accurate; and that her recovery is within normal expectations.

2. Neuroscientific Context

To explain the neuroscientific side of the clip, we chose to investigate Rasmussen's Syndrome, hemispherectomies and the concept of neural plasticity. Each of these was mentioned in the excerpt and is relevant to our studies in neuroscience. The young girl Cameron Mott underwent a hemispherectomy (where half of her brain was removed) to improve her quality of life and help reduce the seizures she was experiencing while suffering from Rasmussen’s Syndrome. Neural plasticity was important in this case as it allowed her to recover so quickly from such a dramatic surgery.
2.1 Rasmussen's Syndrome

Rasmussen's Syndrome is a rare brain disorder characterized by worsening seizures and gradual neurological deficit (Honavar, 1998). Observed onset is generally between 14 months and 14 years old and is primarily unilateral in its effects. The initial hypothesis of the origin of the syndrome was a viral infection, but has since been researched to be an auto immune disorder (Sheybani, Schaller & Seeck, 2011).

The progressive cortical degeneration present in Rasmussen's Syndrome patients is consistent with a continual and worsening process of neuronal damage governed by the immune system (Pardo, Vining, Guo, Skolasky, Carson & Freeman, 2004). Whereas it would normally destroy foreign particles and protect the body, the immune system can start destroying healthy cells in the body, as with other autoimmune disorders such as Type I Diabetes. This cortical pattern of damage can be classified into 4 stages, as outlined by Pardo et al. in their 2004 study involving hemispherectomies. Stage 1 is defined by the presence of lymphocytes in the layers of neurons in the brain in the absence of any neuronal damage. Stage 2 is characterized by increased infiltration of lymphocytes with evidence of neuronal damage in the cortex. Stage 3 involves a large decrease in the amount of neurons, either in clusters or spread evenly throughout the cortex. Stage 4 is widespread destruction of the cerebral cortex. These findings present important evidence for future study and treatment of Rasmussen Syndrome as an immune disease.

As seizures begin to worsen, therapy by drug treatment becomes less, if not at all effective, making the procedure of a hemispherectomy the best and most common option for treatment. There is little to no information on its mortality rate with only 100 cases reported between 1958-2000, making it rare amongst the cabinet of brain disorders and as such, research is limited and only at its genesis (Gilman, 2005).

2.2 Hemispherectomy

2.2.1 Definition and History

'Hemispherectomy' is the removal of one cerebral hemisphere of the brain (Stedman, 2006). It is one of the treatment options for children with seizures and epilepsy syndromes, such as Hemimegalencephaly, Infantile Hemiplegia, Sturge-Weber syndrome and Rasmussen's syndrome. These syndromes cannot be treated by medication (Holloway, Gadian, Vargha-khadem, Porter, Boyd & Connelly, 2000). Dr. Walter Dandy is the first person to have performed an anatomic hemispherectomy for treating a malignant brain tumour in 1928 (Cleveland Clinic, 2012). The first hemispherectomy to treat epilepsy on an adult patient was performed by Mackenzie in 1938, and he reported the great outcome of having curing epilepsy after the surgery (Fountas, Smith, Robinson, Tamburrini, Peitrini & Di Rocco, 2006).

2.2.2 Types of Hemispherectomy

A. Anatomical hemispherectomy
B. Functional hemispherectomy.

Hemispherectomy can be divided into Anatomical Hemispherectomy and Functional Hemispherectomy. Anatomical Hemispherectomy involves the removal of the spot of the seizure's origin on the frontal, parietal, temporal and occipital lobes. It does not involve structures such as the basal ganglion, thalamus and brain stem. Anatomical hemispherectomy has a higher risk of blood loss, delayed hydrocephalus and complications compared to Functional Hemispherectomy. It is usually performed for patients with Hemimegalencephaly. Functional Hemispherectomy involves partial removal and disconnection of the brain. Functional hemispherectomy involves less risk of blood loss and complaints (Cleveland Clinic, 2012).

2.2.3 Cameron Mott's case

The hemispherectomy which was performed on Cameron Mott was an anatomical hemispherectomy. This hemispherectomy is performed to treat intractable epilepsy caused by extensive cerebral unilateral hemispheric injuries. These injuries are secondary to congenital malformations, such as Plurilobar Cortical Dysplasia, HME, and Sturge–Weber syndrome or, as in this case, Rasmussen’s Syndrome (Fountas et al., 2006).

Cameron underwent surgery at the John Hopkins Medical Center to remove the entire right side of her brain, a dramatic surgical procedure that lasted seven hours. The neurosurgeon, Dr. George Jallo was confident that Cameron would make a full recovery after the hemispherectomy. This surgery can be performed successfully on children because of the ability of the remaining hemisphere to compensate for the removed, diseased hemisphere, as will be explored below in Neural Plasticity.

2.3 Neural Plasticity

Studies in recent decades have revealed that the cerebral cortex, rather than being fixed in structure and function, is highly dynamic (Nudo, Plautz & Frost, 2001). The dynamic nature of the brain is also referred to as neural plasticity. Neural plasticity can be defined as the ability of the brain to form or eliminate synaptic junctions between specific neurons. This ability to reorganise synaptic connections plays a crucial role in early brain development, life-long
memory and learning, as well as recovery from neurotrauma.

Children have a greater capacity for neural plasticity than adults do. This is made evident in their faster acquisition of new languages and skills, such as instrument playing in musical adults, as compared to average adults (Johnston, 2004). The specific type of plasticity in children that enhances acquired skills and allows them to recover from brain injuries is known as adaptive plasticity. (Nudo et al., 2001) Children who have undergone hemispherectomies are often able to regain the ability to talk and walk, although fine motor control of the contra-lateral side remains impaired. (Johnston, Ishida, Matsushida, Nishimura & Tsuji, 2009).

When a hemisphere of the brain is removed the contra-lateral side of the body becomes paralysed. This is because the somatosensory and motor cortical maps of the paralysed side of the body have been removed. (Nudo et al., 2001) Functional magnetic resonance imaging (fMRI) has been used to demonstrate that cortical activation associated with both motor and sensory function are transferred to motor and sensory association cortices in the remaining hemisphere. (Johnston et al., 2009) Research indicates that new ipsilateral cortical maps are made in similar, anterior or overlying positions to the previously existing contra-lateral maps. (Holloway et al., 2000) Thus, to some extent, the remaining hemisphere is able to compensate for the function of the lost hemisphere.

An important aspect of neural plasticity is the formation of new, long-lasting synapses. In order for these synapses to form pre-synaptic neurons must fire onto a post-synaptic neuron receiving simultaneous input from other neurons. If the post-synaptic neuron is not strongly activated in this way then a strong neural connection will not be established. Glutamate is one of the primary neurotransmitters involved in long-term synaptic potentiation. Its receptors are categorised as either AMPA receptors or NMDA receptors. The NMDA receptor is different from the AMPA receptor in that it alone appears on the cell membrane of a newly formed post-synaptic neuron. It also requires cellular depolarisation of the post-synaptic membrane as well as the presence of glutamate before the channel will open to allow the influx of Ca2+ ions. The difficulty in reaching depolarisation because NMDA receptors are normally blocked by Mg2+ ions means that the cell can only be activated when there is a great enough summation of input received simultaneously from multiple pre-synaptic neurons to reach action potential threshold. When this occurs the channels open and the influx of Ca2+ ions produces a response in the cell which results in the insertion of AMPA receptors into the cell membrane. Thus long-term potentiation is achieved by the correlated activity of multiple pre-synaptic neurons onto a post-synaptic neuron. (Bear, Connors
3. Analysis of Relevance

3.1 Target Audience and Intention

As a segment of the "Today Show" the media item's primary intent is not to inform viewers of Rasmussen's Syndrome, hemispherectomies or brain plasticity, but rather to highlight the positive outcome of a "medical miracle". Its time slot as a daytime news segment indicates that its target audience is the general population, particularly those at home during the day which would include stay-at-home mothers and members of the retired population. Its focus on a young child makes it relevant particularly to mothers of young children.

Broadly speaking, this target audience would have little knowledge of neuroscience and as such would not be watching the show primarily for its neuroscientific content. The appeal of the item lies in its presentation of something unique, dramatic and emotional. Being a live show, the inclusion of an interview with the patient and family makes the item feel more personal which would enhance its emotional appeal to its target audience and allows the interviewed family to emphasize the allegedly miraculous nature of their daughter's recovery.

3.2 Neuroscientific Relevance

Initially the Today Show reported that Cameron's left hemisphere was removed and that she was paralysed on the left side of her body as a result. However, further into the segment Dr Nancy Snyderman, chief medical editor of NBC News, reported that it was Cameron's right hemisphere that was removed which gave rise to her left hemiparesis. We have reconciled this conflicting information using research articles which support contra-lateral paralysis, indicating that it would have been Cameron's right hemisphere that was removed.
The media item provides overall good quality neuroscientific information which is appropriately simplified for its target audience without losing much of its scientific veracity. The definitions provided for Rasmussen's Syndrome, hemispherectomy and neural plasticity are mostly accurate. Rasmussen's Syndrome is correctly defined near the beginning of the segment as an auto-immune disease and not as a viral infection as was once thought, or as a type of epilepsy, although its symptoms include seizures. The definition provided of neural plasticity as the “ability of the other side of the brain to take over and control the function of the diseased half being removed” is more or less correct. It could be reworded to emphasise the fact that the ability of one hemisphere to take over the functions of the other is a result of neural plasticity; instead the segment defines neural plasticity in terms of its outcomes. It is a simplistic definition with no mention of the mechanisms of neural plasticity but it is appropriate for the target audience and the level of information required. The level and quality of information on the hemispherectomy is also accurate and appropriately pitched to its target audience.

Within the live interview section of the interview, Dr Nancy Snyderman says this about the cause of Rasmussen's syndrome: "It's an electrical storm that sends the wrong kind of electricity." Rather than providing information on what causes the disease Dr Snyderman is presumably focusing on what causes the seizure symptoms. Her answer is so generalised and non-technical that it contains almost no useful neuroscientific information. Her response also includes no mention of Rasmussen's Syndrome being an auto-immune disease or of the causes behind other symptoms, including speech problems. While Dr Snyderman is obviously attempting to simplify information surrounding the neurological disease she does so at the expense of accurate and relevant neuroscientific content.

3.3 Conclusion

Taking into consideration the time limit given to the segment, the audience and its neuroscientific relevance, the media article does a sufficient job at reporting the basic principles of the neurological topic. Facts are narrated in simple, understandable sentences that encompass both the information and the audience's ability to comprehend. Its stage-like presentation of information is reflective of how textbooks would introduce disorders with following steps and treatments and outcomes. Its intention was not to inform the viewing public with great detail but rather to report, which it was achieved with sufficient underlying, factual neuroscience.

4. Appendix

We began with looking for articles or youtube videos on either biologically interesting topics or psychological issues to do with the brain. We chose this news excerpt because of its relevance to some of the broad areas of study in the Neuroscience Fundamentals course. These areas include seizures, neurotrauma and neural plasticity. The excerpt was also of interest because of
the opportunity it afforded us to research an extremely rare autoimmune disease and to investigate current research and information on neural plasticity, a fundamental aspect of neuroscience which is of increasing importance to modern medical treatment. As a group we didn't know much, if anything, about Rasmussen's Syndrome, hemispherectomies and we were limited in our knowledge of brain plasticity, so we took it as a chance to be able to learn outside and around of what we were being taught in lectures.

Following this, research was undertaken in what the group perceived as important aspects of the video: the syndrome itself (cause), hemispherectomy (procedure) and brain plasticity (what made the recovery possible). Subsequently, more detailed information for the project was mainly found amongst research articles, peer reviewed journals and textbooks. As the topics became more detailed, additional articles were found to support our statements and explanations. We searched through the UNSW library catalogue, online search engines by means of Sirius, and other referenced material from the articles we had found to try and find relevant and reliable information from the internet to better inform us about our topics.

Reviewer's comments advised us of our many grammatical errors, lack of structure, pictures, referencing style and (thankfully) pointed out that parts required in the marking criteria were missing. We sought to edit and restructure, as well as try to make the page look more vibrant. In addition we worked on creating uniformity in referencing and on adding and rewriting some paragraphs to better meet the marking criteria.

5. References


http://my.clevelandclinic.org/epilepsy_center/services/epilepsy_surgery/hemispherectomy.aspx


6. Group Work: Who We Are (original proposal and some notes)

Topic:
Rasmussen's syndrome & hemispherectomy

Elise McPherson z3330765
Bianca Abela z3289362
Matilde Nielsen z3433230
Sarah Moore z3329503
Josh Lee z3329177

Jobs:
Sarah: Rasmussen's syndrome
Bianca: Rasmussen's syndrome
Josh: Surgery (hemispherectomy) – procedure
Matilde: Brain plasticity - research
Elise: Brain plasticity - research

Schedule: (ongoing posting on Group Facebook page)
August 24 - meet up after Lab (clearer instructions on division of sections, articles and more detailed information)
- Active posting on Group Facebook page
September 3 - Group Meeting and start posting onto wiki
September 21 - Final Meeting (discussion of final points to fix, edits and completion of project)