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Research for Nursing Practice

Living with Neurofibromatosis Type 2

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Living With Neurofibromatosis Type 2

This literature review will identify themes that stood out from current research that has taken place on Neurofibromatosis Type 2 (NF2) and provide a recommendation for future research in order to help people living with the condition, which currently has no cure. As the number of people around the world living with NF2 is limited, it is important to gain an understanding into the research that has already been carried out and also identify any 'gaps' within the research. Due to the rare nature of this condition, it is important to raise awareness to ensure that when nursing staff do come across Neurofibromatosis Type 2, they have sufficient information and knowledge to administer effective care and promote independence in patients suffering this condition,

Neurofibromatosis type 2 is a condition that causes benign tumours to grow on the central and peripheral nerves. According to (Neuro-foundation 2013) type 2 Neurofibromatosis generally affects 1 in 35,000 people. NF2 is caused by a genetic mutation, where the cells in the body have trouble copying the chromosomes; this leads to tumours growing on nerves around the body, (NHS 2013). In 50% of cases a misprinted gene is normally passed down from one of the parents, and in the other 50% the person is the first to create the faulty gene, this is known as a sporadic mutation. (NINDS 2013) The misprinted gene will be present in a person from birth but the signs and symptoms will not appear until early twenties. Patients with NF2 face many difficulties in life.

Research is essential to develop and refine knowledge so that nurses can improve their clinical care for the patients they treat and that it will be at a high standard. (Grove & Burns, 2011). According to NMC (2008) "you must keep your knowledge and skills up to date throughout your working life".

Literature Review

A literature review is the in depth analysis of the material about a specific topic. It is a useful nursing tool to help healthcare staff gain an overview of the knowledge available. (Aveyard 2010). It allows the nurses to keep their skills and knowledge up to date throughout the time they work as a healthcare professional.

For this particular search, relevant electronic databases were used to carry out the search: Google Scholar, Cumulative Index to Nursing, Allied Health Literature (CINAHL) and Proquest. The search used a variety of different words such as "Neurofibromatosis + Treatment", "Neurofibromatosis Type 2 in adults" , "Neurofibromatosis type 2" + "Deafness" + "Audiology", "NF2 & maintaining independence" and "NF2 + Disability + Symptoms" and it was limited to articles in only English based language. The searches brought up a total 1445 articles. The date that the research articles were written was then limited between the years 2008 to 2013. This then brought the total of articles down to 83. The relevant articles for this particular topic were identified and research articles were selected to be used within this literature review. Due to NF2 being very rare the research into this condition is very limited. The articles were analysed and the 3 themes for people living with Neurofibromatosis type 2 were identified. They were hearing loss, balance difficulties and psych-social impact.

Hearing Loss

One of the difficulties that NF2 patients are faced with is hearing loss due to a brain tumour known as an 'Acoustic Neuroma' (BANA 2013), 90% - 95% of patients will be affected. As the tumour grows the patient's hearing declines. NF2 patients will get an acoustic neuroma on both hearing and balance nerves (UCAL Health, 2013). If this tumour is left untreated it can press on the brain stem and ultimately can lead to death.

A quantitative study carried out by (Asthagiri et al, 2012), selected 100 NF2 patients to take part. All patients involved within this study underwent tests to find out the mechanisms of hearing loss. Firstly the participants had to enrol on National Institute of Neurological Disorders and Stroke (NINDS). Informed consent was also gained. This study was limited to adults only and English had to be their first language. The first test was a Clinical and Audiologic evaluation. Examinations were carried out to exclude any presence of middle ear disease. 44 patients were excluded from the hearing tests due to other medical reasons, a total of 56 patients totalling 89 treatment naïve ears were used. 62% of all evaluated ears had normal hearing. 38% had associated with mild hearing loss, 16% had moderate hearing loss, 13% had severe hearing loss and 4% had profound hearing loss. It was stated that hearing loss occurred gradually over months to years. In a small amount of patients (12%) sudden profound sensorineural hearing loss happened to them. All 56 patients underwent an MRI scan with and without contrast to measure protein levels within the inner ear. They also measured to see if there was an obstruction within the cochlear nerve on both ears. The results of study showed that 94.4% (84 of 89 ears) of participants showed evidence of Acoustic Neuroma. 88% had hearing difficulties associated with the tumour.

In a qualitative study carried out by (Neary, et al 2006a) investigated the effects that NF2 has on patients. The approach used was a semi-structured interview/questionnaire. It was sent out to 20 individuals of varying severity. The most common problem highlighted was hearing loss. 18 of the 20 patients said that they have severe hearing loss, 3 felt their hearing was moderately reduced, the final 3 felt their hearing was only mildly reduced all due to the acoustic neuroma. Seven patients stated they were able to use a telephone. Four reported they were unable to use a telephone due to the severity of their hearing and the remainder of the participants used other methods of communication. 17 out of the 20 patients mentioned that they use lip-reading to compensate for loss of hearing. More candidates could have been added to provide a more accurate outcome.

Balance Difficulties

In a qualitative study by (Evans, et al 2010) used a closed set questionnaire to measure the primary and secondary effects of Neurofibromatosis type 2. The aim of this research was to identify the greatest difficulties that NF2 patients are faced with. 89 adults under the care of Manchester NF2 team were sent out a pack inviting them to take part in the study; it also contained an information leaflet. 62 participants completed and returned the questionnaire. The participants were asked a variety of questions about the problems they face. The second highest problem reported were balance difficulties due to the tumour growing on hearing and balance nerves. 59 participants answered the question on balance problems. Only 6 out of these patients indicated that they had no problems with their balance. A further 18 indicated that they had slight problems. The final 35 indicated they had moderate or severe balance issues. Of the 62 patients that answered the question on

mobility, 31 indicated they had no such problems, 23 indicated they were slightly unsteady on their feet and a further 17 said that they were mobile; however they need to use a stick or walking frame to help them. Balance difficulties were documented in 80% of the participants. All the patients described difficulty in walking on a daily basis; they also explained they had trouble carrying objects with both hands. They felt they needed both hands free while walking in case they lost their footing while walking.

In a qualitative study by (Neary, et al, 2006) a semi structured interview was used to measure the psychosocial impact on NF2. It was administered to 20 individuals of varying severity. The second most common problem highlighted in the study was issues with their vestibular function. Out of the 20 participants 16 felt they did have issues. They were then asked balance difficulty in particular situations. Many patients felt they were limited to certain activities. All 20 patients said that they were able to leave the house, although three stated they needed to be accompanied. Three participants reported that due to their vestibular function they need to use a wheelchair. Finally, when asked about carrying objects, 12 patients responded saying that they need two hands free while moving in case they lost their balance, therefore carrying objects was not an option for these patients.

Psych- Social Impact

A qualitative study by (Patel, C et al 2011) wanted to find out about the impact of patients living with Neurofibromatosis type 2. Patients were taken from a clinic database at a large London teaching hospital. There were certain criteria to participant in the study, patients had to be over the age of 18 and English as their primary language. However, patients with severe communication difficulties were excluded from the study due to the process in which the study was being carried out. All participants were sent a letter explaining the study and consent was gained. The completed forms were analysed and 6 patients took part in the study. 5 interviews were conducted in a private room in the hospital and one was conducted in a patient's home. The interview provided a loose structure of open ended questions. The areas mainly discussed were the physical, emotional and social impact of NF2. All interviews were recorded and transcribed. It was narrowed down to 3 themes, "impact of the disease", "emotional response to the disease" and "awareness of NF2". The interview highlighted that the patients faced a range of emotional responses to their diagnosis, and uncertain about the nature of their condition and the future. Terms used by the patients were "shock", "cry", "feel alone", and "frustrating". Due to the facial weakness patients felt embarrassment and shame. Some spoke about the negative reactions that they had from other people. The patients also felt self-consciousness regarding their appearance, which then resulted in changes in activities and social withdrawal. Finally patients felt shocked upset with their diagnosis of NF2. They were particularly worried that the disease has no cure; they referred NF2 as to being an alien inside them.

As mentioned previously, in the first part of the study by (Neary, W et al 2006) 20 individuals took part. A semi structured interview/questionnaire was used. The specific psychosocial impact of NF2 on the affected individual and family has been reported in this study. Ethical approval was obtained. 19 patients were visited in their homes to complete the study, 1 patient completed the study by post and returned it. When the patients were asked about the diagnosis of NF2 and how it affected them, four patients described a positive experience as they were relieved when they finally got diagnosed and that a known label could be attached to their condition giving rise to the symptoms they were experiencing. 8 patients expressed negative emotions such as "fear", "anxiety" and

“disappointment”. 5 expressed the genetic implications and worry about the future, 2 patients blamed themselves for having the condition and the final 5 gave factual or neutral responses. The study looked at how it affected mobility on public transport, help required in everyday life, domestic life, social relationships, family relationships, work and employment. The patients that took part in the study gave both negative and positive responses. With regards to relationships, many felt that their family were very supportive, however with regards to their employment many felt they were given a hard time. All participants that took part in the study were within working age however only 8 were still able to work and the rest were unemployed due to their medical condition.

Recommendation for further research

Aim

The themes that stood out from the studies that have already been carried out show how living with Neurofibromatosis type 2 can affect the patients on a daily basis. There are many “gaps” within the current research that could be looked into in further detail. The chosen gap for further research of NF2 is the general management of NF2. The aim of this research is to highlight how patients feel with managing their own condition for example, hospital appointments for many different clinics such as ophthalmology, ear, nose and throat, neurosurgery, audiology clinics, physiotherapy, plastic surgery, pain management clinics and finally yearly MRI scans. It should also look into how this can affect patients emotionally. A qualitative study would be undertaken in this particular research.

Methodology

In nursing research two approaches can be used, either qualitative or quantitative study. Methodology strictly means “the study of the method” (Lindsay, B, 2007). A qualitative study focuses on an individual’s experience and their social phenomena. In the health care setting a qualitative research is mostly carried out as it will focus on a patients or carers own experience. The aim of this particular research is to find out about the general management of Neurofibromatosis type 2 and how it can affect patients emotionally, a qualitative research would be the most appropriate approach to use.

Approach

Within a qualitative research there are three different types of approach; phenomenological research, grounded theory research, and ethnographic research. (Burns & Grove 2011) Phenomenology looks at the lived experience of an individual and tries to gain a better understanding of how a patient feels with regards to a certain subject (Lindsay, B, 2007). The best way to carry out this approach is to do one to one interviews on the individual or send out a questionnaire. Grounded theory means to collect data and analyse the data in order to generate a pattern of human behaviour and social contexts (Engward, H 2013) Ethnography is the study of a culture or a community using a range of field works such as observation or randomised control trials. (Lindsay, B, 2007). For this topic a phenomenological approach is the most appropriate as it will look into the experience of individuals with Neurofibromatosis type 2, and how managing the condition can affect the patient emotionally.

Sample

There are two different types of sampling when carrying out research. If your sample needs to represent an overall population then it is most likely to be representative sampling. Whereas if you want to go out and target specific individuals for example people suffering from Neurofibromatosis type 2 then it will be purposive sampling as the researcher is purposively going out to speak to the individuals. (Lindsay, B, 2007). A phenomenological study requires a purposive sample will be carried out. To be included in this study the participants must be suffering from Neurofibromatosis type 2, they must be over the age of 18 as Neurofibromatosis normally affects older patients. The sample size for this specific research is going to consist of large amount of patients that suffer from NF2. Information leaflets would be placed in local support groups for NF2 encouraging them to participate in the study. The specialist clinic based in Manchester (Manchester Royal, 2013) would also be contacted to see if anyone is willing to take part in the study. Finally it is important to reach the saturation. If this does not happen then further participants must be encouraged to take part in the research until there is no new information or data can be collected.

Data Collection

The recommendation for further research is a phenomenological approach. The most appropriate and effective way to carry out this research would be sending out a questionnaire. According to (Gerrish & Lacey 2010) questionnaires provide a standardised set of results which represent the views of the sample studied. The main benefit of a questionnaire is it is relatively easy to complete and does not take up a great amount of time. A questionnaire would be made up with the appropriate questions in relation to the specific topic. The questionnaire would consist of open ended questions which allow the participants to provide detailed responses about how the management of NF2 affects them. It allows the researcher to see exactly what the participants are experiencing. The questionnaire would be sent in a pre-paid envelope. A covering letter will also be added along with the questionnaire explaining the research. The purpose of this is to establish credibility and what stands out in the study to convince the participant that it is worth taking part (Czaja & Blaire 2005 cited in Gerrish & Lancy 2010). The covering letter must also explain that everything will remain confidential and anonymous and the participants have the right to withdraw at any point. A contact number will also be added in the letter in case any of the participants have any questions. Doing all this should hopefully provide a good response rate.

Data Analysis

Thematic analysis will be used. Thematic analysis is seen to be one of the most useful and most widely used methods in qualitative data analysis (Grbich, 2007 cited in Lindsay, 2007). After all the participants have completed and returned their questionnaire all the data is looked at and divided into themes which have emerged. Certain words or phrases can be coded which is easier for the themes to be identified. A team of researchers will then look at the data to make sure it has been analysed correctly before it can be published. The participants and the specialist clinic will then receive the analysed data to make sure that everything is correct.

Ethics

To go ahead with any research, approval must be gained by the National Health Service (NHS). Approval must also be gained by the specialist clinic which is based in Manchester; approval must be gained by the local support groups to place posters. You must also gain approval from the Research Ethics Committee (Gerrish & Lacey, 2010).

Consent must be gained from the participants that are willing to take part. According to Gerrish & Lacey 2010 a signature is required to indicate that informed consent has been given. As touched on previously the covering letter must explain everything in detail to the participant and the benefits and risks of the study (Burns & Grove 2007).

Data collection should always be carried out in ways to ensure confidentiality and anonymity remains. (Lindsay, 2007). The participants must be aware that their names will not be given to anyone and that the information they provide will remain confidential. They must also be aware that all the data will be stored in a password protected computer that only the researchers have access too. Finally, once all the research has been analysed and completed and published all the findings prior to carrying out the research will be destroyed.

Conclusion

When reading the literature review relating to the specific topic it became clear that patients suffering from NF2 face many problems on a daily basis. Although not having much hearing does not seem like a big issue compared to many other things in life, people should be aware that it can affect an individual's confidence a lot. Due to NF2 being less common it is important that everyone is aware of the emotional impact of having a lifelong condition that has no cure. It is also important to gain an understanding of how tiring and emotional it can be for an individual to attend many hospital appointments and having to deal with new tumours appearing anywhere on their body unexpectedly. Although someone suffering from Neurofibromatosis type 2 looks ok from the outside, they have to deal with constant neuropathic pain, and emotional pain on the inside.

Finally, it is important that the results of the study are published so that nursing and medical staff have more knowledge and awareness on this particular condition as it is not well known. This means that the patient would feel a little bit at ease and not have to explain the condition to every new doctor or nurse that they came across at new appointments.

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