Minutes of the CJDSN's FSM/AGM
16th November 2019
Burlington Hotel
Birmingham

Present: Graham Blades, Annette Beal, Gill Chanter, Kate Dahill, Marie Dahill & Laura, Michaela Dunn, Dr Kieran Jayaprakash, Martin Kelly, Stephen Kelly, Michael Kelly, James Kelly, Prof Richard Knight, Terence Lee, Margaret Leitch, Mary Lyon, Beth Marsh, Meg Marsh, Prof Simon Mead, Dr Tzehow Mok, Barbara Moore, Jamie Newton, Dr Akin Nihat, Aisha Oakley, Mark Ratcliffe, Susan Ratcliffe, Jeff Robinson, Anna Savva, Helena Savva, Greg Savva, Samantha Shed, Julia Simmons, Blaire Smith-Bathgate, Andrew Takoushis, Anita Tipping, Florence Tittensor & Mary, Andy Tomaso, Sam Kraven, Gill Turner (40)

There were 35 members who attended the dinner-reception held last night.

Apologies: No names given.

1.0 Welcome
Prof Knight welcome everyone for giving their time to attend the annual Family Support Meeting. Our main goal is for families to meet other families to share and talk about their experience, meet and talk to the experts. Members were re-assured that everyone in the room had been through the same experience who had cared for and still caring for their love ones with CJD.

Prof Knight also informed the members to be vigilant and keep their belongings with them at all times and informed the members of the health and safety guidelines to follow in case of an emergency.

The committee members briefly introduced themselves to the members prior to the proceedings.

2.0 Ice-breaker
Prof Mead led the activity by grouping the members according to the criteria given. It was a wonderful way of breaking the ice, get to know and talk to the members in different groups and putting everyone at ease before the start of the day's proceedings.

3.0 Introduction to Prion Disease/CJD – Dr Kiran Jayaprakash
Dr Jayaprakash opened his talk with normal biology. The body is made up of organs which are made of cells. A cell is a collection of proteins and a nucleus which contain the DNA. Our DNA holds 46 chromosomes. Protein is a chain of amino acids. The protein can misfold into a wrong shape which can cause diseases. Plaques are dead proteins in the brain.

3.1 Where does CJD fit into this?
Dr Jayaprakash spoke about scrapie from sheeps, cannibalism and kuru which shows spongyform encephalopathy on the brain similar to that of a CJD patient. It could be the misfolded protein that is behaving like an infection as stated by Prof Prusner, as referenced by Dr Jayaprakash.

We all produce prion protein. Prion protein is found in the brain and the spinal cord which helps and keeps our neurons healthy. If misfolded it is very resistant to breakdown causing damage to the neurons.

Misfolding of the protein is the root cause of CJD.

3.2 Types of CJD
Genetic- this is due to faulty genes that lead to faulty proteins which are more likely to misfold.

Acquired - the misfolded protein is sourced from the outside which starts a chain reaction. The misfolded protein enters the body by injection of infected human growth hormone (iatrogenic CJD), historical fertility treatment, through blood transfusion, surgery or transplantation and ingestion of infected meat in the food chain (vCJD).

Chance – the most common is sporadic CJD where spontaneous misfolding of the protein takes place and there are no known clear risk factors. There are around 120 cases in the UK per year. Life expectancy is around 4-6 months.

3.3 How is CJD diagnosed?
With the exclusion of diseases presenting similar symptoms that of a CJD patient, assessment of the patient's memory, language, vision, behavior (physical,

emotional and psychological), swallowing, spatial awareness, balance and cognitive capacity are closely monitored.

3.4 Tests to support scientific diagnosis
MRI is a more detailed investigation than a CT scan.
EEG is an electrical recording of the brainwaves/activity of a patient. CJD patients show slower brain activity.
Lumbar puncture is the collection of spinal fluid checking for white blood cells, bacteria, viruses and evidence of inflammation.
RT Quick is the synthesizing of a normal protein if misfolded protein is found.

Postmortem is a process to confirm misfolded proteins are present in the brain.

3.4 What next?
There is still a lot the experts do not know about the condition. They need to know how the misfolding of the protein occur in order to move on. Developing new tools will help early diagnosis.

Questions and answers followed after the presentation.

4.0 Bereavement – Ms Bethan Marsh
Ms Marsh presented her talk by defining "loss" and how it affects all of us.

Loss is defined as someone leaving you or something taken away from you. Loss is an inevitable part of life and death is the event most often thought of as a loss and grief is a natural part of the healing process.

Loss affects our life in more ways than one. It impacts on us psychologically, socially, emotionally, physically, intellectually and spiritually.

Bereavement refers to the death of a significant person and grieving is the psychological part of bereavement.

Ms Marsh went on to explain the stages of grief and loss - denial, anger, bargaining, depression and acceptance by (Kubler Ross, E (1969) On Death and Dying).

Not everyone goes through all the stages but we'll find ourselves in any one of the stages after experiencing the loss of a loved one.

One grows through the support of others while his/her grief remains the same (Tonkin, L (1996) Growing around Grief).

Help and professional support are available nationally, regionally and locally.

Questions and answers followed after the presentation.

5.0 Research Grants Presentation

5.1 Prediction of Care Milestones in CJD – Dr Akin Nihat
Dr Nihat is a neurologist working at the National Prion Clinic.

Dr Nihat's research work is the analysis of data and to use the system of rating of clinical aspects to predict certain criteria when a client is going to need a round the clock care.

After a confident diagnosis of CJD, the aim of his research work revolves around sCJD, to see if change have occurred and put them in sequence to tract their progress, pick up key changes that are relevant to their mobility, continence, speech and cognitive function. Marked change in condition is between 4-6 months. The research also aims to predict on how fast or slow the rate of change is occurring and when extra support is likely to be required when changes start to impact on the client and his/her family.

Dr. Nihat stated that we (experts) are not very good at predicting when outside care is going to be needed however collection of information and data as the disease progresses is a tool to aid the prediction. The "monitoring cohort data" has been going on for 10 years and 500+ patients with sCJD are included. The data and information will assess the increasing care needs of the client. Information from this predictive data is very sensitive and holding data and samples are controlled by regulations. The research work aims to get better at predicting mortality.

Questions and answers followed after the presentation.

5.2 When do prion infection and brain damage begin in individuals at risk of prion disease? – Dr Tzehow Mok (Neurologist)

Dr Mok's research work is to gather a big number of spinal fluid samples and to find out how many clients have positive RT Quick results and to optimize the amplification of the amino acid in order to build a scientific connection to prion disease.

Aims
To build a library of spinal fluid samples.
Develop a test to detect very small amount of prion.
Find proteins leaked by damaged brain cells.
Follow the changes overtime.

Who are at risk?
Individuals with mutation carriers and their untested relatives.
Recipients of human growth hormone sourced from cadavers before 1985.
Recipients of blood transfusion contaminated by VCJD.

Why does it matter?
To refine diagnosis.
Implement timing of treatment- early treatment stands a better chance of a good outcome.
Future drug trials for a rare disease if there is a biomarker.

Questions and answers followed after the presentation.

6.0 Chairman's Report – Prof Knight
Prof Knight gave a brief overview of the Network's achievements, activities and performance this year. The Network is currently funding two CJD research projects. Prof Knight also thank the fundraisers and friends of the Network who donated and fundraised on behalf of the CJD Support Network.

6.1 Approval of the minutes of the 2018 FSM/AGM

Copies of the minutes of the 2018 FSM/AGM were given to the members to peruse and review. The minutes were reviewed and there were no changes or

corrections made. The minutes were proposed true by Grahame Blades and seconded by Florence Tittensor.

7.0 Financial Report – Mr Andy Tomaso
Copies of the financial report were distributed to the members for their perusal.

Profit and Loss
CJD Support Network
01 April 2019 to 08 November 2019

Total Income	£29186.31
Total Operating Expenses	£45955.79
Net Profit	-£15296.48
Current Account	£3867.44
Reserve Account	£149924.21

Most of the money come from fundraising events and others. The research grants awarded to Drs Mok and Nihat are paid for from the Network's excess funds.

The Network has a small surplus this year 3K compared to last year's which is 10K. We aim to have enough money to run the charity for the 3 years and if we have more, fund other research projects in the future.

8.0 Election of officers
The incumbent committee members were duly re-elected en-block and Mr Marston had been proposed and elected to join the committee. The election of officers was accepted and agreed by the members, proposed by Terence Lee and seconded by Andrew Takoushis.

8.1 Dinner-Reception
Following a good attendance of the dinner-reception for the past two years, Prof Knight asked the members if they are happy for the Network to hold another dinner-reception next year and the response was a positive one.

Sam asked if we can make allowance for late commers attending the dinner-reception.

8.2 New venue
Change of a venue was again brought up. Majority of the members are open to moving to a different venue, however there were members who raised the issue that the current venue is easily accessible from the point of its location being close to the train station.

The cost of getting to the FSM/AGM was also raised. A good number of attendees come with their families. Cost of the train tickets can be beyond some families could afford and it is a factor that stops them from attending the Family Support Meeting. Prof Knight has informed the members that the Network has helped families with their train ticket expenses in the past and the Network would again help the families in need of support if they need it.

8.3 Other ideas from the Members
Someone from the audience suggested, could the day's proceedings be recorded and posted on the Network's website to encourage newly bereaved families to attend the FSM/AGM.

Facilitate a meeting in regional centre for sharing common experience.

8.4 Raffle tickets sales
The Network raised £1900.00 with the sale of the raffle tickets. Prof Knight thanked the members and friends of the Network who sold the tickets on behalf of the Network.

9.0 Where are we with Treatment? – Prof Mead
PRN 100 is the humanized antibody version of ICMS 18 produced by genetic engineering. The concept of protein antibody treatment was developed 20 years ago. The Prion Unit developed and manufactured the PRN 100.

The antibody sticks to the normal prion and stops it binding to the prion seed and prevents it from joining the disease process.

In 2013 the humanized antibody, PRN 100, was produced. There was a great interest from pharmaceutical companies but they thought it as commercially risky to manufacture it.

It was through the legal route that the administration of PRN 100 was made possible.

9.1 Clinical approach for the administration of PRN 100
The treatment plan was approved by the Hospital Oversight Committee.

IV administration of 1mg, 10mg, 80mg/kg increment at 48-72hour interval.

Admit the patient to hospital and give a test dose while being closely monitored and followed by subsequent increment in dose up to 120mg which is the maximum dose.

Patients can go home and will have a follow up care from the National Prion Clinic team.

9.2 First Drug Trial
The first patient to receive the treatment was in 2018. It is too early to be clear whether any clinical response is favourable or otherwise, given the highly variable clinical course of CJD.

The results are currently being analysed and the data collected will soon be published.

The potential outcome and overall conclusion is that PRN 100 is potentially a feasible treatment for CJD patients.

Questions and answers followed post presentation.

10.0 Group round table/informal discussion
Members randomly chose their choice of group for the informal discussion. There are experts, professionals and committee members in each group. Members are given the time to talk and ask experts about the day's proceedings that they felt need further explanation.

11.0 Summary/Close- Prof Knight
Prof Knight summed up the days meeting and thanked everyone for their continuing support and wished everyone a safe journey home.

The FSM/AGM finished at 1600hrs.