HFEA Licence Committee Meeting

29 March 2012

Finsbury Tower, 103-105 Bunhill Row, London, EC1Y 8HF

Minutes – Item 3

Centre 0102 (Guys Hospital) – PGD for Frontotemporal dementia with Parkinsonism (also known as Picks disease) (OMIM #600274)

Members of the Committee:	Committee Secretary:
David Archard (lay) Chair Anna Carragher (lay) Rebekah Dundas (lay) (videoconference) Mair Crouch (lay) (videoconference) Sue Price (professional) Debbie Barber (professional)	Lauren Crawford Legal Adviser: Juliet Oliver, Field Fisher Waterhouse

Declarations of Interest: members of the Committee declared that they had no conflicts of interest in relation to this item.

The following papers were considered by the Committee

- Cover sheet
- Executive Summary
- PGD Application form
- Redacted Peer Review
- Genetic Alliance opinion
- Redacted Public Comment

The Committee also had before it

- HFEA Protocol for the Conduct of Licence Committee Meetings and Hearings
- 8th edition of the HFEA Code of Practice
- Human Fertilisation and Embryology Act 1990 (as amended)
- Decision trees for granting and renewing licences and considering requests to vary a licence (including the PGD decision tree); and
- Guidance for members of Authority and Committees on the handling of conflicts of interest approved by the Authority on 21 January 2009.
- Guidance on periods for which new or renewed licences should be granted
- Standing Orders and Instrument of Delegation

- Indicative Sanctions Guidance
- HFEA Directions 0000 – 0012
- Guide to Licensing
- Compliance and Enforcement Policy
- Policy on Publication of Authority and Committee Papers
- HFEA Pre-Implantation Diagnostic Testing ("PGD") Explanatory Note For Licence Committee

Discussion

1. The Committee had regard to its Decision Tree. The Committee noted that the Centre is licensed to carry out PGD. The Committee was also satisfied that the Centre has much experience of carrying out PGD and that generic patient information about its PGD programme and associated consent forms had previously been received by the HFEA.

2. The Committee noted that the Centre's proposed purpose of testing the embryos was as set out in paragraph 1ZA(1)(b) of schedule 2 of the Act, ie. 'where there is a particular risk that the embryo may have any gene, chromosome or mitochondrion abnormality, establishing whether it has that abnormality or any other gene, chromosome or mitochondrion abnormality'.

3. The Committee noted Frontotemporal dementia (OMIM #600724) is a disorder that is inherited in an autosomal dominant manner. There is a 1 in 2 chance of an embryo being affected by this condition.

4. The Committee noted that the condition is fully penetrant in most cases, although some mutations show reduced penetrance. Frontotemporal dementia is caused by mutations in the gene called MAPT on chromosome 17. The main problems fall into two groups, psychiatric and neurological. The disease initially presents as presenile dementia affecting the frontal and temporal cortex. This leads to progressive behavioural changes including: personality change, loss of interest in surroundings, personal hygiene neglect, disinhibition, loss of initiative, obsessive compulsive disorders, hallucinations or delusions, verbal and physical aggressiveness and restlessness.

5. The Committee noted that the age of onset can vary from between 40-60 years of age, however some develop symptoms earlier in adulthood. Physical presentations include: Parkinsonism (rigidity and bradykinesia), decreased facial expression, postural instability, supranuclear palsy (difficulty chewing and swallowing) loss of saccadic eye movement

~~disorders~~. Epileptic seizures are related to specific mutations within the MAPT gene.

6. The Committee noted that there is no curative treatment for Frontotemporal dementia, but medication is available to alleviate some of the symptoms such as sedative and antipsychotic drugs to help aggression, agitation and restlessness. Levadopa is used to help reduce the involuntary movement problems associated with the Parkinsonism.

7. The Committee noted in the Public Comment “affected individuals undergo significant personality change at an early stage of illness. Family members report a painful process of grieving for loss of the person they once knew, now not recognisable, although that person is still alive in body. Families that have been through this process with multiple loved ones have often been stretched to break-point and family relationships and care has broken down. At risk family members report high levels of anxiety, whilst waiting onset of symptoms”

8. The Committee considered that the condition is serious because it is a rapidly progressive neurodegenerative disorder that requires long term physical and mental health care. Those affected become fully care dependent and the impact of this on their families is enormous.

9. The Committee noted that the application is supported by the Peer Reviewer as well as by Genetic Alliance UK. The Genetic Alliance statement is supported by Frontotemporal Dementia support group.

10. The Committee had regard to its explanatory note, in particular paragraph 5.5 ‘*Where a condition has variable symptoms, the Licence Committee will base its determination of how serious the disability, illness or condition is, on the worst possible symptoms’*, and noted that on the basis of the information presented, given the conditions worst symptoms, it was also satisfied that there is a significant risk that a person with the abnormality will have or develop a serious physical or mental disability, a serious illness or any other serious medical condition. Accordingly, it was appropriate to grant the application under paragraph 1ZA(1)(b) of Schedule 2 to the Act.

11. The Committee noted that while they may grant approval, the centre providing PGD for the condition must also ensure that each individual case treated fulfills the severity criteria of Schedule 2 paragraph 1ZA of the HFE Act 1990 (as amended).

12. The Committee agreed to authorise the testing of embryos for Frontotemporal dementia (OMIM #600274). The Committee confirmed that this condition will be added to the published list of conditions for which PGD may be carried out.

Signed: Date: 12/04/2012

David Archard (Chair)