HFEA Statutory Approvals Committee

27 February 2014

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

Minutes – Item 3

Centre 0035 (Oxford Fertility Unit) – PGD application for Autosomal Dominant Acute Necrotizing Encephalopathy OMIM #608033

Members of the Committee: David Archard (lay) Chair Rebekah Dundas (lay) Hossam Abdalla (professional) Jane Dibblin (lay) Advisor: Dr Mary Porteous	Committee Secretary: Lauren Crawford Legal Adviser: Stephen Hocking, DAC Beachcroft Also in attendance: Sam Hartley, Head of Governance and Licensing, HFEA

Declarations of Interest: The Members declared no conflicts in relation to this item

The following papers were considered by the Committee

- Executive summary
- Application form
- Redacted peer review

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) ("the Act")
- 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 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 proposed purpose of testing the embryos was as set out in paragraph 1ZA(1)(b) of schedule 2 of the Act, i.e. '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 that Autosomal Dominant Acute Necrotizing Encephalopathy (OMIM #608033) is inherited in an autosomal dominant pattern and there is a 1 in 2 chance of an embryo being affected with the condition where one parent is affected.

4. The Committee noted that Autosomal Dominant Acute Necrotizing Encephalopathy is a rapidly progressive encephalopathy that can occur in otherwise healthy children after common viral infections such as influenza and parainfluenza. Affected children become comatose after onset of a febrile illness. Outcomes include full recovery, permanent neurologic impairment, and death. Recurrences produce more severe impairments.

5. The Committee noted that the mutation in gene RANBP2 (2q12.1) provides a genetic predisposition to Acute Necrotizing Encephalopathy and it is considered as a likely causative for the disease. Disease penetrance was estimated at 40%, and recurrent episodes occurred in half of affected individuals. Mortality rate can be as high as 25% and 25% of survivors develop severe neurological sequels.

6. The Committee noted that the only symptomatic treatment is available with steroid therapy and this treatment is not always successful.

7. The Committee noted that the application is supported by the Peer Reviewer.

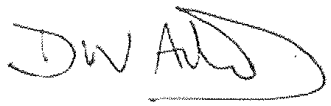
8. The Committee welcomed the advice of its Advisor, Mary Porteous, who confirmed that the condition was as described in the papers. She also clarified for the Committee that this disorder is also caused by mutations on the CPT2 gene and that this application captures those patients as well.

9. The Committee considered that the condition is serious because although it can resolve itself it has a substantial mortality rate with a very substantial chance of moderate to severe residual neurological impairments in those who survive.

10. The Committee had regard to its explanatory note and noted that on the basis of the information presented, given the condition's worst symptoms, it was 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. The Committee was therefore satisfied that the condition meets the criteria for testing under paragraph 1ZA(1)(b) and (2) of Schedule 2 to the Act.

11. The Committee agreed to authorise the testing of embryos for Autosomal Dominant Acute Necrotizing Encephalopathy (OMIM #608033).

Signed: Date: 11/03/2014



David Archard (Chair)