HFEA Licence Committee Meeting

26 August 2010

21 Bloomsbury Street London WC1B 3HF

Minutes – Item 4

Centre 0102 (Guys Hospital)–Application for Variation of licence to include embryo testing for Leigh Syndrome (Infantile Subacute Necrotising Encephalopathy) OMIM# 185620

Members of the Committee: Anna Carragher (lay) – (Chair) Jane Dibblin (lay) Sally Cheshire (lay) Sue Price (Professional) Debbie Barber (Professional) Mair Crouch (lay) Apologies: Rebekah Dundas (lay)	Committee Secretary: Joanne McAlpine Legal Advisers: Sarah Ellson – Field Fisher

Declarations of Interest: members of the Committee declared that they had no conflicts of interest in relation to this item, Debbie Barber noted that she works in a licensed centre.

The following papers were considered by the Committee:

- Executive summary
- Application for the variation of licence
- Lay summary of condition
- Redacted peer review
- Genetics Alliance UK opinion

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)
- HFEA (Licence Committees and Appeals) Regulations 1991 (SI 1991/1889)
- Decision trees for granting and renewing licences and considering requests to vary a licence (including the PGD decision tree)
- Guidance for members of Authority and Committees on the handling of conflicts of interest approved by the Authority on 21 January 2009.
- 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

1. The Legal Adviser drew the Committees attention to the application form in the papers, and informed them that the OMIM number on the application form varies to that on the genetic alliance page within the papers. The Legal Adviser advised the Committee that if they decide to approve this condition for Leigh's Syndrome, then they can only approve the condition that has been applied for within the application (OMIM 185620).

2. The Committee had regard to its Decision Tree. The Committee was satisfied that the Centre has been licensed to perform PGD for a number of years and has appropriately trained and experienced staff to deliver the service.

3. 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, 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'.

4. The Committee noted that this type of Leigh disease carries a dominant OMIM number but that it is inherited in a recessive pattern with a 25% risk where both parents carry a mutation.

5. The Committee noted that there is a significant risk that a person with the abnormality will develop a serious medical condition because it is a 100% penetrant, and onset is generally in infancy and early childhood, although it can be in teenage years.

6. The Committee considered that the condition is serious. Leigh syndrome as described by OMIM number 185620 is a severe progressive neurological disease characterised by deterioration of brain and spinal cord tissue. Affected individuals generally present normally at birth, but typically begin displaying symptoms within a few months to two years of age, although it can be later. Symptoms include seizures, fatigue, poor muscle tone, poor reflexes, poor motor function, poor coordination, and breathing, eating and swallowing difficulties. These symptoms progress until premature death. Treatment in the form of drugs and vitamin supplements is only partially effective in treating some symptoms, and not halting disease progression. There is no cure for Leigh syndrome and affected individuals usually die in early or late childhood.

7. Leigh syndrome is a rare inherited condition affecting the nervous system. It is caused by a number of mutations in a number of genes which build mitochondrial proteins.

The Committee's Decision

8. The Committee agreed that on the basis of the information presented, the Committee 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. Accordingly, it was appropriate to grant the application under 1ZA(1)(b) of Schedule 2 of the Act.

9. The Committee agreed that the licence should be varied to authorise the testing of embryos for the highly specific form of Leigh's syndrome that has been applied for in this application – OMIM# 185620, and that no conditions should be put on the licence in relation to this variation. The Committee confirmed that this specific form of Leigh's syndrome will be added to the published list of conditions for which PGD may be carried out.

Signed: Anna Carragher Date: 15.9.2010.

Anna Carragher (Chair)