HFEA Licence Committee Meeting

26 April 2012

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

Minutes – Item 4

Centre 0044 (Centre for Reproductive and Genetic Health) – PGD for Glutaric academia (aciduria) type 1 (OMIM #231670)

Members of the Committee:	Committee Secretary:
David Archard (lay) Chair Anna Carragher (lay) Rebekah Dundas (lay) (videoconference) Mair Crouch (lay) Sue Price (professional) Debbie Barber (professional) Jane Dibblin (lay)	Joanne McAlpine Legal Adviser: Stephen Hocking, Partner, Public Law Department For DAC Beachcroft LLP

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

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 Glutaric acidemia (aciduria) type 1 (OMIM #231670) is a disorder that is inherited in an autosomal recessive manner. Where both parents carry an affected copy of the relevant gene there is a 1 in 4 chance (25% risk) of the embryo exhibiting the syndrome.

4. The Committee noted that Glutaric academia (aciduria) type 1 has a varying degree of expression. This disorder presents during the first year of life and results in physical handicap and often mental impairment as well. The effects vary widely between affected individuals. There may be widespread muscle weakness due to the inability to break down the amino acids lysine, hydroxylysine and tryptophan, leading to a build-up in glutaric acid and brain damage. Dietary control may restrict neurological damage. Difficulty in moving and spasms, jerking, rigidity or decreased muscle tone and muscle weakness may be aided (but not cured) by special seating. The Committee noted the risk of an affected individual suffering acute metabolic crises/decompressions, potentially leading to profound physical and in some cases mental disability.

5. The Committee noted that treatment is available for Glutaric acidemia (aciduria) type 1 but is supportive only and the syndrome cannot be cured. Treatment may involve the use of oral supplements and/or strict dietary control which may help limit progression of the neurological damage.

6. The Committee considered that the condition amounts to a serious physical or mental disability because inheriting the mutation from both parents will result in at least some physical disability and maybe mental impairment. The Committee further considered that even if managed to the extent possible the effect on an individual's metabolism was a serious illness and/or a serious medical condition.

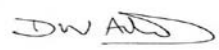
7. The Committee noted the application that both the Peer Reviewer and Genetic Alliance UK consider the condition to be serious.

8. The Committee had regard to its explanatory note, in particular paragraphs 5.4 *Where a condition has a range of penetrance (eg. 40-60%), the Licence Committee will base its decision on the highest penetrance figure* and 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'*. The Committee noted that on the basis of the information presented, given the highest degree of penetrance and 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 another serious medical condition. Accordingly, it was appropriate to grant the application under paragraph 1ZA(1)(b) of Schedule 2 to the Act.

9. 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).

10. The Committee agreed to authorise the testing of embryos for Glutaric acidemia (aciduria) type 1 (OMIM #231670). The Committee confirmed that this condition will be added to the published list of conditions for which PGD may be carried out.

Signed: Date: 10/05/2012



David Archard (Chair)