HFEA Licence Committee Meeting

29 September 2011

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

Minutes – Item 3

Centre 0102 (Guy's Hospital) – PGD for Peroxisome Biogenesis Disorders PBD (Zellweger Syndrome Spectrum ZSS)

Members of the Committee: David Archard (lay) – Chair Debbie Barber (professional) Anna Carragher (lay) Sally Cheshire (lay) (videoconference) Mair Crouch (lay) (videoconference) Rebekah Dundas (lay) (videoconference) Sue Price (professional)	Committee Secretary: Terence Dourado Legal Adviser: Graham Miles, Morgan Cole

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:

- Executive Summary
- PGD application form
- Redacted peer review form
- 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

1. The Committee had regard to its Decision Tree. The Committee was satisfied that the Centre has considerable 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 that Peroxisome Biogenesis Disorders
PBD (Zellweger Syndrome Spectrum ZSS) is a disorder that is inherited in an autosomal recessive manner. If an embryo inherits a copy of the faulty gene from both parents it will develop the disease, ie there is a 1 in 4 chance of the embryo having the abnormality.

4. The Committee considered that there is a significant risk that an individual with the condition will be affected because it is fully penetrant

5. The Committee considered that the condition was serious because the three phenotypes are all serious and have similar overlapping features. There is also some variation of symptoms within the three phenotypes. There are twelve genes known to be involved in these disorders and it is not possible to consistently assign these to the different phenotypes. PBD (ZSS) symptoms include floppiness in the newborn, poor feeding, fits, liver cysts with liver dysfunction, hearing loss, vision impairment and developmental delay. The Committee noted that most individuals will be diagnosed in the newborn period or in childhood. The condition is often slowly progressive.

6. The Committee noted that treatment for all forms of PBD (ZSS) focus on symptomatic treatment and palliative care. This may include gastrostomy, antiepileptic drugs, hearing aids, cataract removal and glasses. Quality of

life is affected in the most serious form, and almost all children die in the first six to twelve months of life.

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

8. The Committee agreed to authorise the testing of embryos for Peroxisome Biogenesis Disorders PBD (Zellweger Syndrome Spectrum ZSS). The Committee confirmed that this condition will be added to the published list of conditions for which PGD may be carried out.

Signed: Date: 11/10/2011

David Archard (Chair)