HFEA Licence Committee Meeting

31 March 2011

21 Bloomsbury Street London WC1B 3HF

Minutes – Item 6

Centre 0044 (Centre for Reproductive and Genetic Health (CRGH)) –PGD for Mucopolysaccharidosis type VI OMIM# 253200

Members of the Committee:	Committee Secretary:
David Archard (lay) – Chair Debbie Barber (professional) Anna Carragher (lay) Sally Cheshire (lay) Mair Crouch (lay)	Terence Dourado Legal Adviser: Tom Rider, Field Fisher

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

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

Tabled documents

- Genetic Alliance opinion

1. The Committee noted that it was in receipt of a Genetic Alliance Opinion regarding the condition. The legal adviser reminded the Committee of the provisions of Regulation 9.2 of the Human Fertility and Embryology (Procedure for Revocation, Variation or Refusal of Licences) Regulations 2009 (as amended), and the Committee accepted the document in to its consideration of the item.

2. 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 form had been received by the HFEA.

3. The Committee was satisfied 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'.

4. The Committee noted that Mucopolysaccharidosis type VI OMIM #253200 is inherited in an autosomal recessive pattern. If an embryo inherits a copy of the faulty gene from both parents it will develop the disease,i.e.there is a 1 in 4 chance of the embryo having the abnormality.

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 fully penetrant with variable expressivity.

6. The Committee considered that the condition is serious because in its severest form it has an early onset with symptoms appearing between the first few months of life and certainly by two years of age, progressing to death before the second decade. The Committee noted that there is a less severe and slower progressive form of the condition where symptoms do not appear until the end of the first decade or in to the second, with long term survival reported in to the fifth decade. The Committee noted that there is significant intrafamilial variability. The symptoms of the disease

include progressive skeletal dysplasia with short stature, joint contractures. Additionally, there are problems with recurrent otitis media (inflammation of the middle ear), sleep apnoea (cessation of breathing during sleep, of longer than 10 seconds) hepatosplenomegaly (enlargement of both the liver and spleen), cardiac valve disease, cranial and peripheral nerve impingement and instability of the cervical spine. Cognitive function is usually normal and remains so throughout the course of the disease. Symptomatic treatment is available and in addition more recently treatment by haematopoietic stem cell transplantation (HSCT) has been carried out. This has lead to some improvement in symptoms but in many cases the skeletal features have continued to progress, and there is a significant morbidity and mortality associated with the treatment.

7. The Committee had regard to its explanatory note and noted that on the basis of the information presented given the conditions 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. Accordingly, it was appropriate to grant the application under paragraph 1ZA(1)(b) of Schedule 2 to the Act.

8. The Committee reminded centres that given the rarity of the condition and its visual identifiers all centres must be mindful of preserving the confidentiality of its patients.

9. The Committee noted paragraph 10.5 of the Code of Practice (8th edition)/ HFEA guidance for Centres: ‘The use of PGD should be considered only where there is a significant risk of a serious genetic condition being present in the embryo. When deciding if it is appropriate to provide PGD in particular cases, the seriousness of the condition in that case should be discussed between the people seeking treatment and the clinical team. The perception of the level of risk for those seeking treatment will also be an important factor for the centre to consider.’

10. The Committee agreed that the licence should be varied to authorise the testing of embryos for Mucopolysaccharidosis type VI OMIM #253200, and that no conditions should be put on the licence in relation to the variation. The Committee confirmed that this condition will be added to the published list of conditions for which PGD may be carried out.

Signed..... Date..14.4.2011.

David Archard (Chair)