HFEA Licence Committee Meeting

15 January 2012

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

Minutes – Item 5

Centre 0044 (The Centre for Reproductive and Genetic Health) Wolcott-Rallison Syndrome (OMIM # 226980)

Members of the Committee: David Archard (lay) Chair Sue Price (professional) Debbie Barber (professional) Jane Dibblin (lay) Rebekah Dundas (lay)	Committee Secretary: Lauren Crawford Legal Adviser: Juliet Oliver, Field Fisher Waterhouse

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 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

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 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 Wolcott-Rallison Syndrome (WRS) (OMIM #226980), is inherited in an autosomal recessive manner which means that carrier parents have a 1 in 4 chance of having an affected child in each pregnancy.

4. The Committee noted that Wolcott-Rallison Syndrome (WRS) is a rare autosomal recessive disease, characterised by neonatal/early-onset non-autoimmune insulin-requiring diabetes associated with skeletal dysplasia and growth retardation. Typically, diabetes occurs before six months of age, and skeletal dysplasia is diagnosed within the first year or two of life. Other manifestations vary between patients in their nature and severity and include frequent episodes of acute liver failure, renal dysfunction, exocrine pancreas insufficiency, intellectual deficit, hypothyroidism, neutropenia and recurrent infections. Bone fractures may be frequent.

5. The Committee noted that early diagnosis is beneficial, in order to ensure rapid intervention for episodes of hepatic failure, which is the most life threatening complication. Close therapeutic monitoring of diabetes and treatment with an insulin pump are recommended because of the risk of acute episodes of hypoglycaemia and ketoacidosis. Interventions under general anaesthesia increase the risk of acute aggravation, because of the toxicity of anaesthetics, and should be avoided. Prognosis is poor and most patients die at a young age.

6. 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) of Schedule 2 to the Act.

7. The Commiitee noted that the application is supported by a Peer Reviewer and the Genetic Alliance UK.

8. The Committee agreed to authorise the testing of embryos for Wolcott-Rallison Syndrome (WRS) (OMIM #226980). The Committee confirmed that this condition will be added to the published list of conditions for which PGD may be carried out.

Signed: Date: 08/02/2013

David Archard (Chair)