HFEA Licence Committee Meeting

29 March 2012

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

Minutes – Item 4

Centre 0102 (Guys Hospital) – PGD for Long QT Syndromes Types 1, 2, 3, 5 and 6 (also known as Romano Ward Syndrome) (OMIM #613688)

Members of the Committee:	Committee Secretary:
David Archard (lay) Chair Anna Carragher (lay) Rebekah Dundas (lay) (videoconference) Mair Crouch (lay) (videoconference) Sue Price (professional) Debbie Barber (professional)	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
- 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 Long QT syndromes Types 1, 2, 3, 5 and 6 (OMIM #613688) is a disorder that is inherited in an autosomal dominant manner. There is a 1 in 2 chance of an embryo being affected by this condition.

4. The Committee noted that Long QT syndromes have several subtypes depending on the causative mutation. There is a varying degree of penetrance depending on the subtype. However the death rate is similar across all three phenotypes.

5. The Committee noted that condition is caused by an abnormal electrical signal in the heart that affects the way the heart beats. The part of the heart beat that is affected is called the QT interval and in this condition it is prolonged and can cause abnormal heart rhythm. As a result those affected can experience sudden loss of consciousness and fainting attacks. In a proportion of cases this can lead to cardiac arrest resulting in sudden, often unexplained death.

6. The Committee noted that the age of onset can vary greatly, from infancy to adulthood. Most cardiac events occur in the preteens to the 20s and become less common after 30-40 years of age.

7. The Committee noted that treatment starts in childhood in all gene carriers even if they have no symptoms as they may become symptomatic at a

later stage. Treatment includes beta blockers which will regulate the QT interval. Some of those affected may require pacemakers or implantable cardioverter defibrillators (ICD's). Although treatments are over 90% effective in preventing sudden death, families at risk of Long QT need long term screening and monitoring. The psychological impact of a well and healthy individual attending clinic and going away with a diagnosis of life long condition that needs monitoring cannot be underestimated.

8. The Committee noted for affected children living everyday life is a challenge. They are advised that competitive sports are a risk factor. Even though normal physical activities are suitable, parents remain constantly concerned for the health of their children which can have a major social or psychological impact on the child and his/ her family.

9. The Committee considered that the condition is serious because there is an ongoing risk of sudden death and noted the psychological impact of living with this risk. Any collapse or syncope may be feared as the first symptom to cause death. Treatment is life long and reviews of medication and symptoms are 3-4 monthly. This can be a challenge especially for teenagers and affected individuals will have a live in a limited fashion feeling unable to participate in many normal activities due to the fear of sudden death.

10. The Committee noted that the application is supported by the Peer Reviewer as well as by Genetic Alliance UK. The Genetic Alliance statement is supported by Sudden Adult Death Syndrome.

11. The Committee had regard to its explanatory note, in particular paragraphs 5.4 '*Where a condition has a range of penetrance (e.g. 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'*, and noted that on the basis of the information presented, given the conditions worst symptoms, it was also 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.

12. 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).

13. The Committee agreed to authorise the testing of embryos for Long QT syndrome 1, 2, 3, 5 and 6 (OMIM #613688). The Committee confirmed that this condition will be added to the published list of conditions for which PGD may be carried out.

Signed: Date: 12/04/2012

David Archard (Chair)