HFEA Licence Committee Meeting

27 September 2012

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

Minutes – Item 3

Centre 0044 (Centre for Reproductive and Genetic Health) – PGD for Muscular dystrophy, limb-girdle (LGMD) type 1B (OMIM# 159001)

Members of the Committee:	Committee Secretary:
David Archard (lay) Chair Sue Price (professional) Jane Dibblin (lay) Anna Carragher (lay) Mair Crouch (lay) Rebekah Dundas(lay)–videoconferencing	Lauren Crawford 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

- Cover sheet
- 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

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 Muscular dystrophy, limb-girdle (LGMD) type 1B (OMIM# 159001) 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 where one parent is affected.

4. The Committee noted that the disorder presents before 20 years of age. LGMD1B is quite a variable condition in terms of the severity of muscle weakness, but usually the progression is slow to moderate and people remain ambulant. Life expectancy depends upon the identification and treatment of the associated involvement of the heart and the breathing muscles. There are no specific treatments for LGMD1B.

5. The Committee noted that joint contractures (tightening) or foot drop can occur in LGMD1B and therefore regular physiotherapy is recommended. Splints are sometimes worn to enhance good positioning of the ankle joints or help with foot drop if there is weakness in the feet. Because of the risk of problems with the heart in LGMD1B (rhythm and conduction disturbance), regular heart checks are required and these should include echocardiograms. Some affected people are likely to need the insertion of a defibrillator. Regular monitoring of respiratory function is recommended. Sometimes overnight studies are indicated. Managing the symptoms of the condition improves a person's quality of life. Exercise should be undertaken, but with medical advice. ,

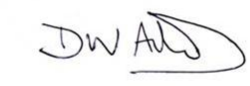
6. The Committee noted that the condition is serious because sudden cardiac death is very common in LGMD1B occurring in up to 50% of patients. Implantable cardioverter defibrilators (ICDs) can terminate many malignant ventricular arrhythmias, but have no impact on cardiomyopathy and progression to heart failure and death. Cardiac transplant prolongs survival in some. Currently there is no curative treatment so management is largely supportive, aiming to reduce the impact of weakness and contractures on disability through physiotherapy and exercise. Cardiac monitoring is essential in order to pick up and treat cardiac complications early. The use of medication may delay progression of cardiac complications but cannot prevent it. Similarly respiratory monitoring is essential as nocturnal ventilation may be necessary in a significant proportion of patients which can improve quality of life and prolong survival.

7. The Committee noted that the application is supported by the Peer Reviewer.

8. The Committee had regard to its explanatory note, in particular paragraph 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.

9. The Committee agreed to authorise the testing of embryos for Muscular dystrophy, limb-girdle (LGMD) type 1B (OMIM# 159001). The Committee confirmed that this condition will be added to the published list of conditions for which PGD may be carried out.

Signed: Date: 09/10/2012



David Archard (Chair)