Statutory Approvals Committee - minutes

Centre 0102 (Guys Hospital) – PGD application for reducing body myopathy OMIM #300717

Friday, 5 August 2016

HFEA, Level 2, 10 Spring Gardens, London, SW1A 2BU

Committee members	Rebekah Dundas (Chair) Margaret Gilmore Anthony Rutherford Anne Lampe	
Members of the Executive	Ian Brown Trent Fisher	Head of Corporate Governance Secretary
External adviser	Peter Turnpenny	
Legal Adviser	Sarah Ellson	Fieldfisher LLP
Observers	None	

Declarations of interest:

- members of the committee declared that they had no conflicts of interest in relation to this item.

The committee had before it:

- 8th edition of the HFEA Code of Practice
- standard licensing and approvals pack for committee members

The following papers were considered by the committee:

- executive summary
- PGD application form
- redacted peer review
- Genetic Alliance opinion

1. Consideration of application

1.1. The committee welcomed the advice of its specialist advisor, Professor Peter Turnpenny, who confirmed that the condition is as described in the papers, a serious muscular disorder with complications that may lead to loss of mobility.

1.2. The committee noted that the application is consistent with the Peer Review and Genetic Alliance opinion.

1.3. 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.

1.4. The committee noted that the condition being applied for is not on the approved PGD condition list.

1.5. The committee noted that the proposed purpose of testing the embryos was as set out in paragraph 1ZA(1)(b) of schedule 2 of the Act, i.e. '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'.

1.6. The committee noted that the condition is inherited in an x-linked dominant pattern and there is a 1 in 2 chance of an embryo being affected with the condition where one parent is affected. Although this is an x-linked condition, females who are carriers of the disease can manifest symptoms and may be as severely affected as males.

1.7. The committee noted that the condition is a X-linked subtype of Myofibrillar Myopathy (MFM) and is caused by a mutation in the FHL 1 gene on the X chromosome. The condition causes progressive muscle weakness, especially in the muscles that run parallel to the spine and in the limbs.

1.8. The committee noted other symptoms that may present include weakness of the respiratory muscles and heart muscles that can lead to premature death. Scoliosis of the spine can be a consequence of the disorder.

1.9. The committee noted that during development children will struggle to be able to sit up and stand unaided. For those who develop the ability to walk, they will deteriorate over time and will need the use of a wheelchair.

1.10. The committee noted that the condition demonstrates complete penetrance in males and penetrance is high in females. Age of onset for males is infancy to early childhood and for females' teenage years to early adulthood.

1.11. The committee noted that there is no curative treatment for the condition.

2. Decision

2.1. The committee considered that the condition is serious due to the profound effect the condition has on the affected individual and risk of premature death.

2.2. 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) and (2) of Schedule 2 to the Act.

2.3. The committee agreed to authorise the testing of embryos for reducing body myopathy OMIM #300717.

3. Chair's signature

3.1. I confirm this is a true and accurate record of the meeting.

Signature

Name

Rebekah Dundas

Date

19 August 2016