

Statutory Approvals Committee - minutes

Centre 0035 (Oxford Fertility) – PGD application for Gerstmann- Straussler-Scheinker syndrome OMIM #137440

Thursday, 25 February 2015

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

Committee members	David Archard (Chair) Margaret Gilmore Anthony Rutherford Anne Lampe	
Members of the Executive	Trent Fisher	Secretary
External adviser	Professor John Walter	
Legal Adviser	Dawn Brathwaite	Mills & Reeve
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 John Walter, who confirmed that the condition is as described in the papers.
- 1.2. The committee noted that the application is consistent with the Peer Review.
- 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 autosomal dominant pattern and there is a 1 in 2 chance of an embryo being affected with the condition where one parent is affected with the condition.
- 1.7. The committee noted that the condition is a type of prion disease which is part of a group of degenerative conditions that affect the nervous system. In the late stages of the condition affected individuals can be bedridden due to severe ataxia, unable to communicate due to profound dysarthria and unable to eat due to lack of coordination in swallowing. The condition ultimately causes severe disability and death.
- 1.8. The committee noted that symptoms can include progressive unsteadiness and difficulty walking, cognitive dysfunction leading to slowness of thought processing, slurred speech and rigid muscle tone, visual disturbances leading to blindness, deafness and swallowing problems.
- 1.9. The committee noted that the condition has a high penetrance. Onset of symptoms is between 35 and 50 years of age.
- 1.10. The committee noted that there is no curative treatment for the condition, only treatment for symptoms that present.

2. Decision

- 2.1. The committee considered that the condition is serious as it is an unrelenting degenerative condition.
- 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 Gerstmann-Straussler-Scheinker syndrome OMIM #137440.

3. Chair's signature

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

Signature

A handwritten signature in black ink, appearing to read 'DWA' followed by a stylized flourish.

Name

David Archard

Date

10/03/2016