Statutory Approvals Committee – minutes

Centre 0006 (The Lister Fertility Clinic)

Pre-implantation Genetic Diagnosis (PGD) application for

Bardet-Biedl Syndrome 10, OMIM #615987

Thursday, 29 June 2017

Church House Westminster, Dean's Yard, Westminster SW1P 3NZ

Committee members	Margaret Gilmore (Chair) Anne Lampe Ruth Wilde Bobbie Farsides	
Members of the Executive	Bernice Ash Paula Robinson	Secretary Head of Planning & Governance
External adviser	Dr Alan Fryer	
Legal Adviser	Ros Foster	Browne Jacobson LLP
Observers		

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
- Minutes of one previous Licence Committee meeting

1. Consideration of application

1.1. The committee welcomed the advice of its specialist adviser, Dr Alan Fryer, who confirmed that the condition was as described in the papers.

1.2. The committee noted that the description in the application for Bardet-Biedl Syndrome 10, OMIM #615987 is consistent with the Peer Review.

1.3. The committee noted that the Genetic Alliance opinion provided a patient perspective and supported the application.

1.4. 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.

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

1.6. 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'.

1.7. The committee noted that the condition is inherited in an autosomal recessive manner which means there is a 25% chance of having an affected child in each pregnancy, if each parent has a relevant mutation.

1.8. The panel noted the condition is a genetic disorder affecting the cilia structures in several different parts of the body. It is associated with early onset symptoms including visual loss preceded by night blindness, postaxial polydactyl (an additional finger or toe on the outside of the hands or feet), learning difficulties including speech delay, childhood obesity, cardiovascular abnormalities, renal failure and hypogonadism (reduced function of the testes or ovaries). Infertility and diabetes are also common in Bardet-Biedl Syndrome 10. Penetrance is considered to be close to 100%.

1.9. The committee noted there is currently no curative treatment for Bardet-Biedl Syndrome 10. Symptomatic treatment includes monitoring of blood pressure and renal function, haemodialysis and, ultimately, transplantation; surgical removal of extra digits; vision aids for visual impairment; hormone replacement therapy for hypogonadism; specialist dentistry; hearing aids; diabetes treatment; and assisted conception for infertility.

1.1. Affected individuals are usually eventually registered blind or partially sighted which can affect education, choice of occupation and independence. Intellectual disability is present in up to 80%, and is often mild to moderate. Cardiac defects including cardiomyopathy are relatively uncommon but can be fatal. Cystic kidney disease is an important complication that may be life-threatening or require dialysis or renal transplantation in due course.

1.2. The committee noted the inspectorate’s request to consider whether Bardet-Biedl Syndrome 10, OMIM #615987 should be approved for inclusion the PGD List. If approved, the condition would be named as Bardet-Biedl Syndrome 10, OMIM #615987 which is consistent with the information published by OMIM. The committee agreed to consider the application on this basis.

2. Decision

2.1. The committee considered that Bardet-Biedl Syndrome 10, OMIM #615987 is serious given the early onset and the impact on quality of life of this condition, including kidney failure, learning disabilities, obesity and associated complications, uncertainty over how the condition will progress, teenage onset of legal blindness in many cases and the lack of a cure. The committee also noted the peer reviewer's comment that this variant of Bardet-Biedl Syndrome 10 has a higher risk of having severe medical problems than other forms of the condition. The committee considered the impact on the family caring for affected individuals.

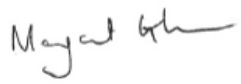
2.2. The committee had regard to its explanatory note and noted that on the basis of the information presented, given the condition's worst symptoms, it was satisfied that there is a significant risk that a person with the abnormality will have or develop a serious physical or mental disability, a serious illness or any other serious medical condition. The committee was therefore satisfied that the condition of Bardet-Biedl Syndrome 10, OMIM #615987 does meet the criteria for testing under paragraph 1ZA(1)(b) and (2) of Schedule 2 of the Act.

2.3. The committee agreed to authorise the testing for Bardet-Biedl Syndrome 10, OMIM #615987.

3. Chair's signature

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

Signature



Name

Margaret Gilmore

Date

25 July 2017