Statutory Approvals Committee – minutes

Item 1

Centre 0327 (Boston Place)

Prolidase Deficiency, OMIM #170100

Thursday, 27 June 2019

HFEA Medway Meeting Room, Level 2, 10 Spring Gardens, London, SW1A 2BU

Committee members	Margaret Gilmore (Chair) Bobbie Farsides (Deputy Chair) Anne Lampe Rachel Cutting Ruth Wilde	
Members of the Executive	Moya Berry Catherine Burwood	Committee Secretary Licensing Manager (Observer)
Specialist Adviser	Jenny Carmichael	
Legal Adviser	Sarah Ellson	Fieldfisher LLP
Observers	Dee Knoyle Vicky Brown	Committee Secretary Inspector

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:

- 9th 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 UK statement
- Email clarification from the Person Responsible on questions relating to the application

1. Consideration of application

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

1.2. The committee noted that the description in the PGD application for Prolidase Deficiency, OMIM #170100 was consistent with the peer review.

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

1.4. The committee noted that the Genetic Alliance UK statement provided a perspective on the impact of the condition on patients, their families and carers.

1.5. 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.

1.6. 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 Prolidase Deficiency is inherited in an autosomal recessive pattern, which means there is a 25% chance of an embryo being affected with the condition in each pregnancy if each parent has a relevant mutation.

1.8. The committee noted penetrance is high and close to 100%.

1.9. Prolidase Deficiency is a rare multi-system disorder which is usually detectable after birth or in early childhood. The condition is characterised by immune dysregulation leading to chronic infections, lung disease, skin ulcers which can lead to amputation, skin rash, enlargement of the liver and spleen and developmental delay. The condition can be life threatening, leading to death in childhood.

1.10. There is no cure for this condition and treatment is supportive.

1.11. The committee noted the inspectorate's request to consider Prolidase Deficiency, OMIM #170100 for inclusion on the list of conditions approved for PGD. The committee agreed to consider the application on this basis.

2. Decision

2.1. The committee considered that, in the worst-case scenario Prolidase Deficiency, OMIM #170100 is a severe multi-system condition that usually presents at birth or in early childhood. The condition can be life-threatening leading to death in early childhood. There is no cure for the condition which can have a significant effect on the quality of life for patients.

2.2. The committee had regard to its explanatory note and confirmed that, on the basis of the information presented, it was satisfied that there is a particular risk that an embryo may have the abnormality in question and that there is a significant risk, that a person with the abnormality will, given the conditions' worst symptoms, have or develop a serious physical disability, a serious illness or any other serious medical condition.

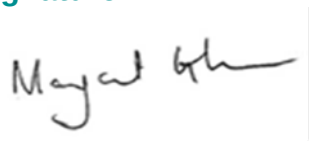
2.3. The committee was therefore satisfied that the following condition meets the criteria for testing under paragraph 1ZA(1)(b) and (2) of Schedule 2 of the Act. The committee agreed to authorise testing for:

- Prolidase Deficiency, OMIM #170100

3. Chairs signature

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

Signature



Name

Margaret Gilmore

Date

22 July 2019