Inhibitor Working Party

Membership

Dan Hart	Chairman
Kate Talks	Secretary
Trevor Baglin	
Liz Chalmers	
Peter Collins	
Charles Hay	
Ri Liesner	
Mike Makris	
Ben Palmer	
Savita Rangarajan	
Anne Riddel	
Mike Williams	

Olly Tunstall stepped down owing to conflicting commitments

Meetings

The group has met 3 times since the last AGM.

Activities

UKHCDO response to SIPPET study (Survey of Inhibitors in Plasma-Products Exposed Toddlers)

We have authored and disseminated our consensus position statement on the SIPPET study. A copy of our statement is available via the UKHCDO website.

Revised ITI recommendations – UKHCDO website and NHS England (NHSE) policy.

Our revised consensus guidance for inhibitor screening in PUPs and initiating ITI are also available on the UKHCDO website and have been adopted formally by NHSE. There are two fundamental changes of note:

To start ITI as soon as practicable after a reproducible inhibitor titre has been detected, i.e. to no longer wait for a reduction to below 10BU prior to commencing ITI.

A commissioning expectation of reporting prospective data to the NHD

We have commenced the prospective collection of data, which will be key to underpin our continued advocacy for adequate funding for ITI. Since November 2015, 14 PUP/MTP patients from 10 centres have been reported to have an inhibitor and on the ITI pathway. The ITI protocol document, available on the UKHCDO website, is explicit about both ITI dosing initiation, monitoring frequency and response interpretation, concluding with clear advice about de-escalation of dosing or necessity for second line/cessation considerations.

Acquired Haemophilia A prospective outcome data reporting

In parallel to the above ITI data collection, a similar rationale underpins the implementation of prospective data reporting of Acquired Haemophilia A outcomes. Since implementation in March 2016, 66 patients have been reported from 27 centres and now on the prospective data collection track. As another high cost area of haemostasis care, we recognized the need to report outcomes at a national level to continue to advocate for our patient group. This is timely both for the existing bypassing agent availability as well as understanding how recombinant porcine FVIII might fit into a future treatment algorithm. On behalf of the UKHCDO and CRG, I will input the clinical contribution to the NHSE evaluation of Obizur, which is just commencing.

Laboratory contact

As our Biomedical scientist representative on the group, Anne Riddel has been liaising with NEQAS colleagues to coordinate both a national written survey of laboratory practice for inhibitor surveillance as well as a NEQAS exercise for detection of anti-porcine-FVIII antibodies. Both these exercises should now be taking place together in Oct/Nov 2016. Please highlight these to your laboratory Chief Biomedical Scientist/team. Anne is also procuring a laboratory contact cascade for comprehensive care centres' laboratories to enable a closer relationship between UKHCDO working parties, particularly the inhibitor and paeds groups, and laboratory colleagues. This will facilitate communication to clarify existing assay practice and offer support/guidance about the laboratory implications of new products.

NHD Inhibitor reporting portal revision

Discussions have taken place with MDSAS to explore revising the NHD Inhibitor reporting portal. Although not quite ready to launch, these revisions aim to align the required data to the clinical background of severe haemophilia A, severe haemophilia B or non-severe haemophilia A.

THUNDER project (Treatment of Haemophilia, Unmet Need, and Disease Epidemiology in the Real world)

This is a recently initiated collaboration between the working party, NHD and Roche aiming to describe the prevalence of inhibitors in patients with haemophilia A of all severities and consequent national burden of care for inhibitor occurrences at all stages of life. The project aims to report in early 2017.

European Medicines Agency meta-analysis collaboration

We have been collaborating with the independent investigators from the Paul-Ehrlich Institute, Germany, led by Dr Brigitte Keller-Stanislawski. Ben Palmer and the NHD team worked hard to share our agreed data sets to enable the UKHCDO, FranceCoag and PedNet data to be analysed together. Dr Keller-Stanislawski presented her preliminary analysis at the World Federation of Hemophilia congress in July 2016 and publication of a peer-reviewed manuscript is imminent.

Dr Dan Hart,
Chairman, Inhibitor Working Party
October 2016