# BIMDG Bulletin

British Inherited Metabolic Diseases Group

## Spring 2020 – COVID-19 update

- Current BIMDG guidance & changes to 2020 calendar
- Launch of the BIMDG metabolic formulary

BIMDG
British Inherited Metabolic Disease Group

## Message from the chair — Elaine Murphy

I hardly know how to begin to write this. Within the space of a few weeks, our lives, working and personal, have changed beyond recognition. In the face of this pandemic, there has been an incredible pulling together of our IMD community, superbly assisted by the patient support groups – in particular Metabolic Support UK and the LSD Collaborative.

Zoom MDTs and WhatsApp calls are the new norms. Terms such as 'self-isolation' and 'shielding' describe the incredibly challenging advice we have given to some of our most vulnerable patients to reduce their exposure to COVID-19. Initiatives that we might have been trying to get off the ground for months or years – have suddenly become possible, with a melting away of red tape – if that initiative can help in any way with managing the pandemic.

We know that there are very hard times to come yet, and that we will be required to work as a group, and as a society, to minimize the impact on our patients and ourselves. Many of our members are doing this whilst living in a different country or region to their close family, parents or grandparents. This gives added importance to the 'team' at work – so let's all look out for each other over the next few months.

Our cover photo shows Manchester at night – restrictions in place, with empty streets. We know that we need the 'stay at home' message to be followed if we are to have any chance of reducing the impact on the NHS and preventing deaths. Apparently it was during quarantine from the Great Plaque in the 1660s that Issac Newton (aged 24 years!) described calculus and formulated the theory of universal gravitation…..so we can only hope that perhaps at the end of this very difficult period some pioneering ideas will also be formulated by another bright young scientist.

We have deliberately kept this bulletin brief – we know that everyone is extremely busy.

Stay safe all of you.

## Bulletin editorial

Given the ongoing COVID-19 epidemic and its impact on the NHS, the planned spring bulletin will be postponed in lieu of a concise bulletin aggregating all current BIMDG advice and notifications pertinent to healthcare professionals working in inherited metabolic disease.

Although events are changing daily, we intend to maintain open communication with our members and share advice that may be of benefit. To that end we would encourage our members to get in touch with their sub-specialty representative if there is information that they think should be shared. Specific advice issued by Public Health England and its devolved sister agencies is evolving constantly and as such should be looked for directly from their respective websites to ensure guidance is accurate and up to date.

## BIMDG events calendar for 2020 (as of April 2nd)

Due to the introduction of social distancing measures aimed at containing the spread of COVID-19, all BIMDG workshops due to take place over the next few months and the symposium in Manchester will be postponed until next year. Individuals who have already paid for the summer symposium will have their ticket automatically transferred to next years event or refunded on request. Events scheduled for later in the year (September and onwards) will hopefully be able to go ahead, pending further developments.

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| Cambridge Science Festival | Postponed to March 2021 |
| Gyrate atrophy workshop, London | Postponed until further notice. |
| BIMDG Symposium, Manchester | Rescheduled to 23-25th June 2021 |
| Inherited metabolic diseases for the paediatrician, Manchester | Rescheduled to 18th September 2020 |
| Inherited disorders of glucose homeostasis | (As scheduled) 14th-16th October 2020 |

## BIMDG Treasurers update — Rob Barski

Dear BIMDG Members

I appreciate that this may not be on the top of everyone's agenda at the current difficult time but can I remind everyone that as of this year all membership fees were **due by the 31st January**. In addition the membership fee has changed for some for 2020 onwards. The current membership rates are as follows:

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| £40 per annum | Clinical Consultant / AFC Band 8 and above |
| £25 per annum | Non-consultant grade / AFC Band 7 and below / Retired member |

My thanks goes to those people who have complete the membership questionnaire and amended their payments as appropriate. It is disappointing, however, that this only applies to approximately one third of the membership. I am sure there are many more members who are paying the correct rate but have not indicated to me which this is. Please can ask everyone who has not yet done so to take two minutes to inform me of their membership status and amend their payments as appropriate. **Please note if payment of membership fees continue to be unpaid or incomplete your membership status is at risk of being lost.**

Similarly, if you have not received such notifications by email it is likely that your contact details are not up to date.

To notify me of your membership status or update your details please email myself at robert.barski@nhs.net.

Thank you very much for your cooperation.

Robert Barski - BIMDG Treasurer

## Launch of the BIMDG formulary

This month saw the launch on the website of the BIMDG formulary. This extensive piece of work was led by Will Batten, with the support of the BIMDG Pharmacists Group and others with particular expertise in specific areas of IMD.

The formulary has been developed to support prescribers across the NHS in the prescription and monitoring of treatments for rare inherited metabolic disorders. In conjunction with this formulary, a **Shared Care Guidance Plan** has been produced to support prescribers in primary and secondary care.

The aims of the formulary are:
• To support prescribers in primary and secondary care – with a single resource for information on treatment for these conditions.
• To reduce the postal costs to tertiary care in sending out medications / supplements to patients who may live far from a specialist centre, by supporting their local secondary or primary care in the safe prescription of treatment.
• To reduce the inconvenience, loss of working time, and potential gaps in treatment caused by patients being asked to travel long distances to collect medications / supplements from a specialist centre.
• To clarify the responsibilities of tertiary, secondary and primary care in the prescribing and monitoring of these treatments.
• To provide a national information resource to pharmacists.

We are grateful to Metabolic Support UK (www.metabolicsupportuk.org) who provided financial support for the completion of this project.

Feedback on this formulary is very welcome. Please email feedback on the **BIMDG Formulary Feedback Form** found on our website.

The dietitians are continuing work on a similarly styled formulary for dietetic products and supplements and we hope to include that on the website shortly.

## BIMDG COVID-19 guidance

BIMDG (together with Metabolic Support UK and the LSD Collaborative) has released several statements now on COVID-19. The latest statement can be found at http://www.bimdg.org.uk/site/news.asp

Members of the BIMDG also contributed to a webinar for patients. A recording of this webinar can be found at https://www.youtube.com/watch?v=I9jyO-bOQU4&feature=emb_title

The PHE guidance on shielding and protection for extremely vulnerable persons has now been released:

https://www.gov.uk/government/publications/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19

The definition of people falling into the extremely vulnerable group includes:

PEOPLE WITH RARE DISEASES AND INBORN ERRORS OF METABOLISM THAT SIGNIFICANTLY INCREASE THE RISK OF INFECTIONS (such as SCID, homozygous sickle cell).

These patients are being advised to start shielding measures:

'Shielding is a measure to protect extremely vulnerable people by minimising interaction between those who are extremely vulnerable and others. This means that those who are extremely vulnerable should not leave their homes, and within their homes should minimise all non-essential contact with other members of their household. This is to protect those who are at very high risk of severe illness from coronavirus (COVID-19) from coming into contact with the virus.'

This is going to be a major undertaking for individuals and families involved, so it is important to define which of our IEM patients are at 'significantly increased' risk if they become infected. This judgement is going to be down to each individual metabolic team, but I thought it might be useful for you to have the information the CRG submitted to NHSE which led to IEM being included in these measures.

NHSE specifically asked us for information on patients from our cohort who would be at high risk of severe COVID-19 infection. Rather than providing a list of individual conditions, we responded as below:

"A number of our patients have had either solid organ or HSC transplants as a treatment for their underlying metabolic disease. We believe these patients will have been flagged up by transplant services. Some metabolic diseases have neutropenia as a feature, putting patients at increased risk of bacterial infection, but not necessarily a virus such as that responsible for COVID19. Examples would be Glycogen Storage Disorder 1b, alfa mannosidosis, cobalamin deficiencies. Other patients have had splenectomies (eg some patients with Gaucher disease) which might also be a risk factor.

Our main concern is that a number of the conditions we look after either cause pulmonary disease or put patients at risk of metabolic decompensation with any intercurrent infection. Examples of conditions causing respiratory compromise would be Pompe disease, Niemann-Pick disease type B, mucopolysaccharidoses, Glycogen Storage Disease type 3. Many neurodegenerative conditions due to metabolic disorders progress to a point where patients are wheelchair bound and PEG-fed: for most of these patients an intercurrent pulmonary infection can prove fatal.

In addition, we have a large cohort of patients who may suffer a metabolic decompensation if they contract a viral illness, regardless of whether or not the lungs are affected. These patients, particularly the younger ones, will then need hospital admission and intensive, expert management, which can include ICU admission. Examples of such disorders would be urea cycle defects, fatty acid oxidation disorders, maple syrup urine disease, organic acidaemias (methylmalonic aciduria, propionic aciduria, isovaleric aciduria, glutaric aciduria)."

Hopefully you will find this useful in identifying the subgroup of your patients who should be following these stringent shielding measures.

Dr Robin Lachmann

CRG Chair