Inherited Metabolic Disease and Coronavirus (COVID-19)

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Coronavirus (COVID-19)

- You can submit questions through the online Q&A facility and we will try to provide answers to these questions after the webinar
- We will not be able to address questions relating to specific patients or conditions
- Please contact you patient organisation for practical, social care or financial support

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Expert Panel:

Dr Robin Lachmann
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Clinical Psychologist, Charles Dent Metabolic Unit, National Hospital for Neurology and Neurosurgery

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Shielding and the search for a vaccine

Dr Robin Lachmann

Charles Dent Metabolic Unit,
National Hospital for Neurology and Neurosurgery

London

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Public Health England

Guidance

Guidance on shielding and protecting people defined on medical grounds as extremely vulnerable from COVID-19

Published 21 March 2020

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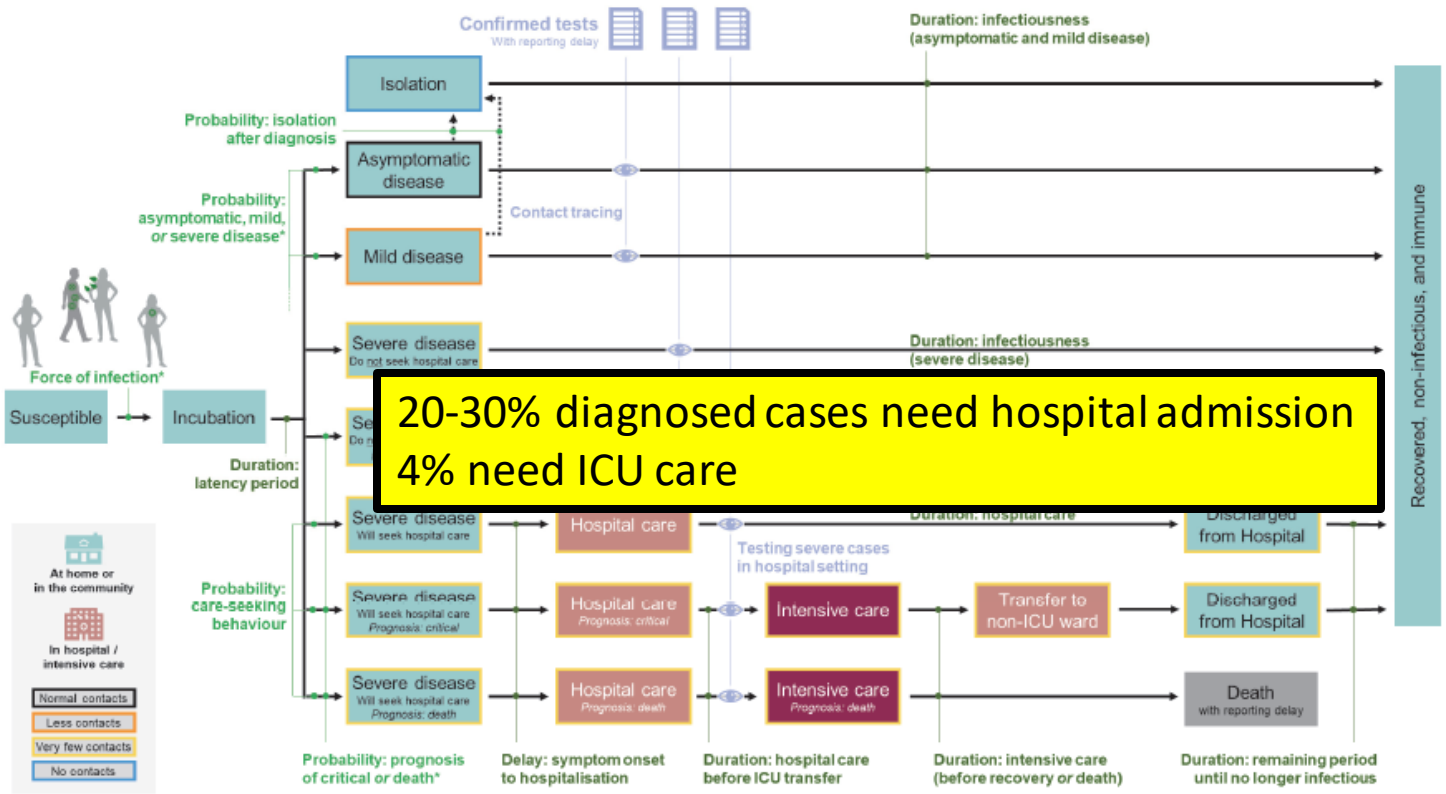
Shielding was initially introduced to protect ICU capacity during the first peak of COVID-19

Figure 1. Model compartment overview



20-30% diagnosed cases need hospital admission
4% need ICU care

https://www.ecdc.europa.eu/sites/default/files/documents/Projected-baselines-COVID-19-for-assessing-impact-measures.pdf

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Shielding

Can we identify those most likely to need ICU care and prevent them from becoming infected?

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Extremely Vulnerable

1. Solid organ transplant recipients.
2. People with specific cancers:
   - people with cancer who are undergoing active chemotherapy
   - people with lung cancer who are undergoing radical radiotherapy
   - people with cancers of the blood or bone marrow such as leukaemia, lymphoma or myeloma who are at any stage of treatment
   - people having immunotherapy or other continuing antibody treatments for cancer
   - people having other targeted cancer treatments which can affect the immune system, such as protein kinase inhibitors or PARP inhibitors
   - people who have had bone marrow or stem cell transplants in the last 6 months, or who are still taking immunosuppression drugs
3. People with severe respiratory conditions including all cystic fibrosis, severe asthma and severe chronic obstructive pulmonary (COPD).
4. People with rare diseases and inborn errors of metabolism that significantly increase the risk of infections (such as Severe combined immunodeficiency (SCID), homozygous sickle cell).
5. People on immunosuppression therapies sufficient to significantly increase risk of infection.
6. Women who are pregnant with significant heart disease, congenital or acquired.

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IMD patients for shielding

Those with significant respiratory compromise due to:

- Pompe disease
- Niemann-Pick disease type B
- MPS I (Hurler disease)
- MPS II (Hunter)
- MPS IV (Morquio)
- MPS VI (Maroteau Lamy)
- Glycogen Storage Disease type 3

Those at risk of metabolic decompensation due to:

- Ornithine transcarbamylase deficiency
- Citrullinaemia (type 1 and 3)
- Arginosuccinic aciduria
- Carbamoyl phosphate synthetase I (CPSI)deficiency
- N-acetylglutamate synthetase (NAGS) deficiency
- Arginase deficiency
- Very-long-chain acyl-CoA dehydrogenase deficiency (VLCAD)
- Long-chain L-3 hydroxyacyl-CoA dehydrogenase deficiency (LCHAD)
- Medium-chain acyl-CoA dehydrogenase (MCAD)
- Multiple acyl-CoA dehydrogenation deficiency (MADD)
- Maple Syrup Urine Disease
- Methymalonic aciduria
- Propionic aciduria
- Isovaleric aciduria
- Glutaric aciduria

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NHSE used ICD10 to identify patients for shielding

- Ornithine transcarbamylase deficiency
- Citrullinaemia (type 1 and 3)
- Arginosuccinic aciduria
- Carbamoyl phosphate synthetase I (CPSI)deficiency
- N-acetylglutamate synthetase (NAGS) deficiency
- Arginase deficiency
- Very-long-chain acyl-CoA dehydrogenase deficiency (VLCAD) E71.310
- Long-chain L-3 hydroxyacyl-CoA dehydrogenase deficiency (LCHAD) E71.310
- Medium-chain acyl-CoA dehydrogenase (MCAD) E71.311
- Multiple acyl-CoA dehydrogenation deficiency (MADD) E71.313
- Maple Syrup Urine Disease E71.0
- Methymalonic aciduria E71.120
- Propionic aciduria E71.121
- Isovaleric aciduria E71.110
- Glutaric aciduria

} **E71**

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What went wrong?

- E71 Disorders of branched-chain amino-acid metabolism and fatty-acid metabolism
  - E71.0 Maple-syrup-urine disease
  - E71.1 Other disorders of branched-chain amino-acid metabolism
    - E71.11 Branched-chain organic acidurias
      - E71.110 Isovaleric acidemia
      - E71.111 3-methylglutaconic aciduria
      - E71.118 Other branched-chain organic acidurias
    - E71.12 Disorders of propionate metabolism
      - E71.120 Methylmalonic acidemia
      - E71.121 Propionic acidemia
      - E71.128 Other disorders of propionate metabolism
    - E71.19 Other disorders of branched-chain amino-acid metabolism
  - E71.2 Disorder of branched-chain amino-acid metabolism, unspecified
  - E71.3 Disorders of fatty-acid metabolism
    - E71.30 Disorder of fatty-acid metabolism, unspecified
    - E71.31 Disorders of fatty-acid oxidation
      - E71.310 Long chain/very long chain acyl CoA dehydrogenase deficiency
      - E71.311 Medium chain acyl CoA dehydrogenase deficiency
      - E71.312 Short chain acyl CoA dehydrogenase deficiency
      - E71.313 Glutaric aciduria type II
      - E71.314 Muscle carnitine palmitoyltransferase deficiency
      - E71.318 Other disorders of fatty-acid oxidation
    - E71.32 Disorders of ketone metabolism
    - E71.39 Other disorders of fatty-acid metabolism
  - E71.4 Disorders of carnitine metabolism
    - E71.40 Disorder of carnitine metabolism, unspecified
    - E71.41 Primary carnitine deficiency
    - E71.42 Carnitine deficiency due to inborn errors of metabolism
    - E71.43 Iatrogenic carnitine deficiency
    - E71.44 Other secondary carnitine deficiency
      - E71.440 Ruvalcaba-Myhre-Smith syndrome
      - E71.448 Other secondary carnitine deficiency
  - E71.5 Peroxisomal disorders
    - E71.50 Peroxisomal disorder, unspecified
    - E71.51 Disorders of peroxisome biogenesis
      - E71.510 Zellweger syndrome
      - E71.511 Neonatal adrenoleukodystrophy
      - E71.518 Other disorders of peroxisome biogenesis
    - E71.52 X-linked adrenoleukodystrophy
      - E71.520 Childhood cerebral X-linked adrenoleukodystrophy
      - E71.521 Adolescent X-linked adrenoleukodystrophy
      - E71.522 Adrenomyeloneuropathy
      - E71.528 Other X-linked adrenoleukodystrophy
      - E71.529 ..... unspecified type
    - E71.53 Other group 2 peroxisomal disorders
    - E71.54 Other peroxisomal disorders
      - E71.540 Rhizomelic chondrodysplasia punctata
      - E71.541 Zellweger-like syndrome
      - E71.542 Other group 3 peroxisomal disorders
      - E71.548 Other peroxisomal disorders

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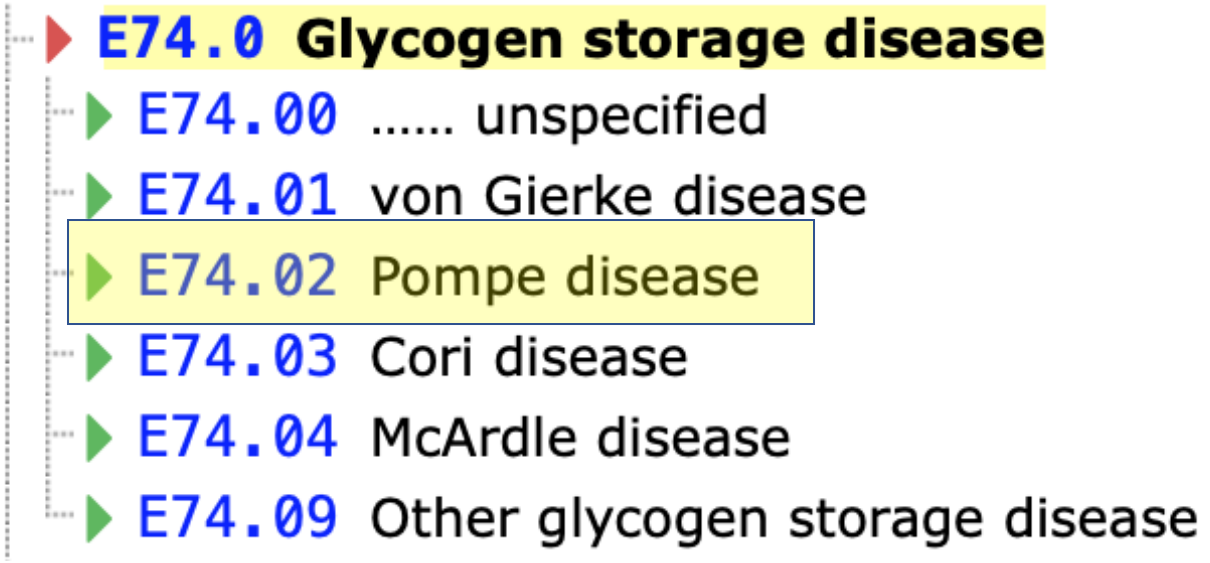
What went wrong?

- Other diseases were included – X-ALD
- Not everyone with the diseases included was extremely vulnerable - MCADD

What went wrong?



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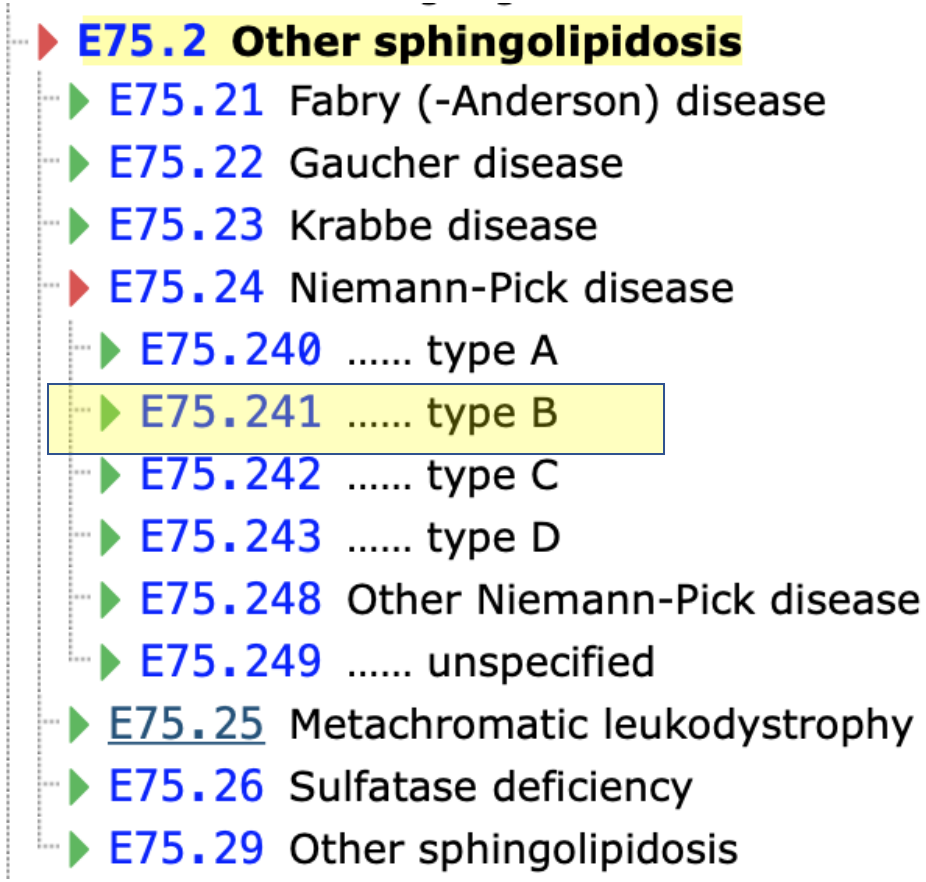
What went wrong?



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What else went wrong?

We are all different

Not everyone with one of these conditions is at extremely high risk

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What did shielding involve?

- **Those who are extremely vulnerable should:**
  - not leave their homes
  - minimise all non-essential contact with other members of their household

- **You should also follow these face-to-face distancing measures:**
  - Strictly avoid contact with someone who is displaying symptoms of coronavirus (new continuous cough, fever or loss of, or change in, sense of smell or taste (anosmia))
  - Don't leave your house
  - Don't attend any gatherings - this includes gatherings of friends and families in private spaces for example family homes, weddings and religious services
  - Don't go out for shopping, leisure or travel and, when arranging food or medication deliveries, these should be left at the door to minimise contact
  - Keep in touch using remote technology such as phone, internet, and social media
  - Use phone or online services to contact your GP or other essential services - if you require an ambulance, phone 999 and tell the call handler you're following shielding measures because of an underlying health condition

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What happened last week?

To: NHS Trust Medical Directors
NHS Trust Chief Nursing Officers
GPs
CCG AOs
ICS/ STP CEOs

Dear Colleagues,

NHS SUPPORT FOR PATIENTS WHO ARE SHIELDING

Since the start of the pandemic, you and your teams have devoted a great deal of time and effort identifying and advising patients who are clinically extremely vulnerable to COVID19, ensuring they are put on the Shielded Patient List (SPL), and most importantly of all, changing how the NHS provides care for this group of over 2 million people who were strongly advised to stay at home. We are hugely appreciative of your invaluable work in continuing to support these patients and helping to keep them safe and well.

The Government's updated advice

On Sunday evening 31 May, the Government published updated guidance on shielding on its website. This letter confirms the actions for the NHS.

The Government states that it has revised its advice by a small degree to reflect that COVID-19 disease levels are substantially lower now than when shielding was first introduced, with the most recently estimated prevalence in the community being, on average, 1 in 420 people.

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Who's at higher risk from coronavirus

Coronavirus (COVID-19) can make anyone seriously ill. But for some people, the risk is higher.

There are 2 levels of higher risk:

- high risk (clinically extremely vulnerable)
- moderate risk (clinically vulnerable)

Important

The lists below may not include everyone who's at higher risk from coronavirus and may change as we learn more about the virus.

People at high risk (clinically extremely vulnerable)

- have had an organ transplant
- are having chemotherapy or antibody treatment for cancer, including immunotherapy
- are having an intense course of radiotherapy (radical radiotherapy) for lung cancer
- are having targeted cancer treatments that can affect the immune system (such as protein kinase inhibitors or PARP inhibitors)
- have blood or bone marrow cancer (such as leukaemia, lymphoma or myeloma)
- have had a bone marrow or stem cell transplant in the past 6 months, or are still taking immunosuppressant medicine
- have been told by a doctor they have a severe lung condition (such as cystic fibrosis, severe asthma or severe COPD)
- have a condition that means they have a very high risk of getting infections (such as SCID or sickle cell)
- are taking medicine that makes them much more likely to get infections (such as high doses of steroids or immunosuppressant medicine)
- have a serious heart condition and are pregnant

If you're at high risk from coronavirus, you're advised to take extra steps to protect yourself.

This is called shielding.

https://www.nhs.uk/conditions/coronavirus-covid-19/people-at-higher-risk/whos-at-higher-risk-from-coronavirus/

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Shielding advice

Do

- only leave your home to spend time outdoors, for example to go for a walk
- stay at least 2 metres (3 steps) away from other people in your home as much as possible
- get food and medicine delivered and left outside your door – ask friends and family to help or register to get coronavirus support on GOV.UK if you need it
- prepare a hospital bag, including a list of the medicines you're taking, in case you need to go into hospital
- wash your hands with soap and water often – do this for at least 20 seconds
- make sure anyone who comes into your home washes their hands with soap and water for 20 seconds
- use hand sanitiser gel if soap and water are not available
- clean objects and surfaces you touch often (such as door handles, kettles and phones) using your regular cleaning products
- clean a shared bathroom each time you use it, for example by wiping the surfaces you have touched

Don't

- do not have visitors inside your home, including friends and family, unless they're providing essential care
- do not stop taking any prescription medicines without speaking to your doctor

Spending time outside your home

You can leave your home to spend time outdoors, for example to go for a walk.

If you decide to do this:

- always stay 2 metres (3 steps) away from other people
- try not to leave your home more than once a day
- go out on your own or with people you live with
- if you live alone, you can spend time outdoors with one other person - ideally the same person each time
- go outside when there are fewer people around, such as early in the morning
- do not go anywhere indoors such as into shops

https://www.nhs.uk/conditions/coronavirus-covid-19/people-at-higher-risk/advice-for-people-at-high-risk/

People at moderate risk (clinically vulnerable)

- are 70 or older
- are pregnant
- have a lung condition that's not severe (such as asthma, COPD, emphysema or bronchitis)
- have heart disease (such as heart failure)
- have diabetes
- have chronic kidney disease
- have liver disease (such as hepatitis)
- have a condition affecting the brain or nerves (such as Parkinson's disease, motor neurone disease, multiple sclerosis or cerebral palsy)
- have a condition that means they have a high risk of getting infections
- are taking medicine that can affect the immune system (such as low doses of steroids)
- are very obese (a BMI of 40 or above)

If you're at moderate risk from coronavirus, you can go out to work (if you cannot work from home) and for things like getting food or exercising. But you should try to stay at home as much as possible.

It's very important you follow the general advice on social distancing, including staying at least 2 metres (3 steps) away from anyone you do not live with.

https://www.nhs.uk/conditions/coronavirus-covid-19/people-at-higher-risk/whos-at-higher-risk-from-coronavirus/

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The Government is currently advising everyone who is considered clinically extremely vulnerable to continue to ‘shield’ at home, in this modified way, until the end of June 2020 to protect themselves from COVID-19.

The Government has committed to reviewing the shielding guidance alongside each review of the wider social distancing measures to ensure that the latest epidemiology is directly considered with advice to clinically vulnerable groups.

They have confirmed the next review will take place the week commencing 15 June 2020. The government plans to write to all individuals on the SPL with information about next steps on shielding advice and the support that will be available to them after this review point.

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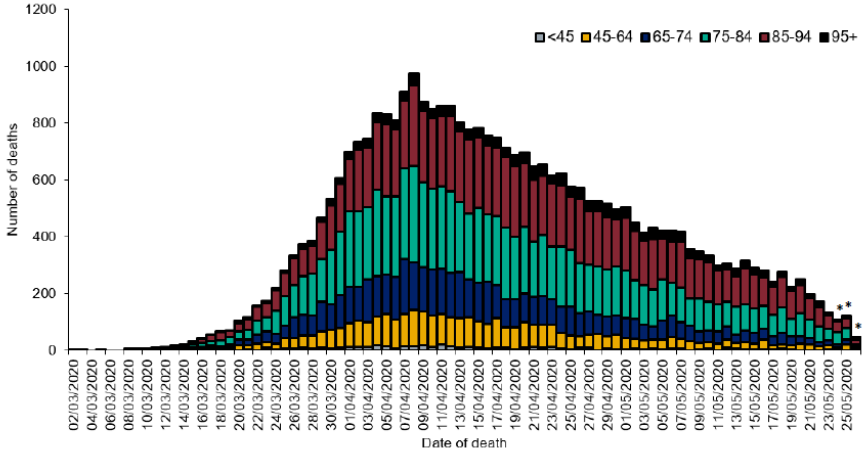
The first peak

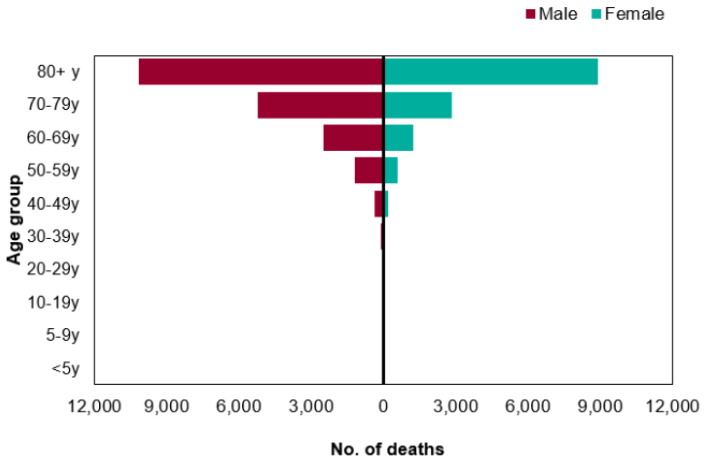
Figure 26: Cumulative number of deaths by date of death and age group, England (n=33,362)



* For the most recent dates, more deaths will be reported therefore the decrease seen in this graph should be interpreted with caution

Cumulative deaths

Figure 27: Age/sex pyramid of laboratory confirmed COVID-19 deaths (n=33,362)



https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/888254/COVID19_Epidemiological_Summary_w22_Final.pdf

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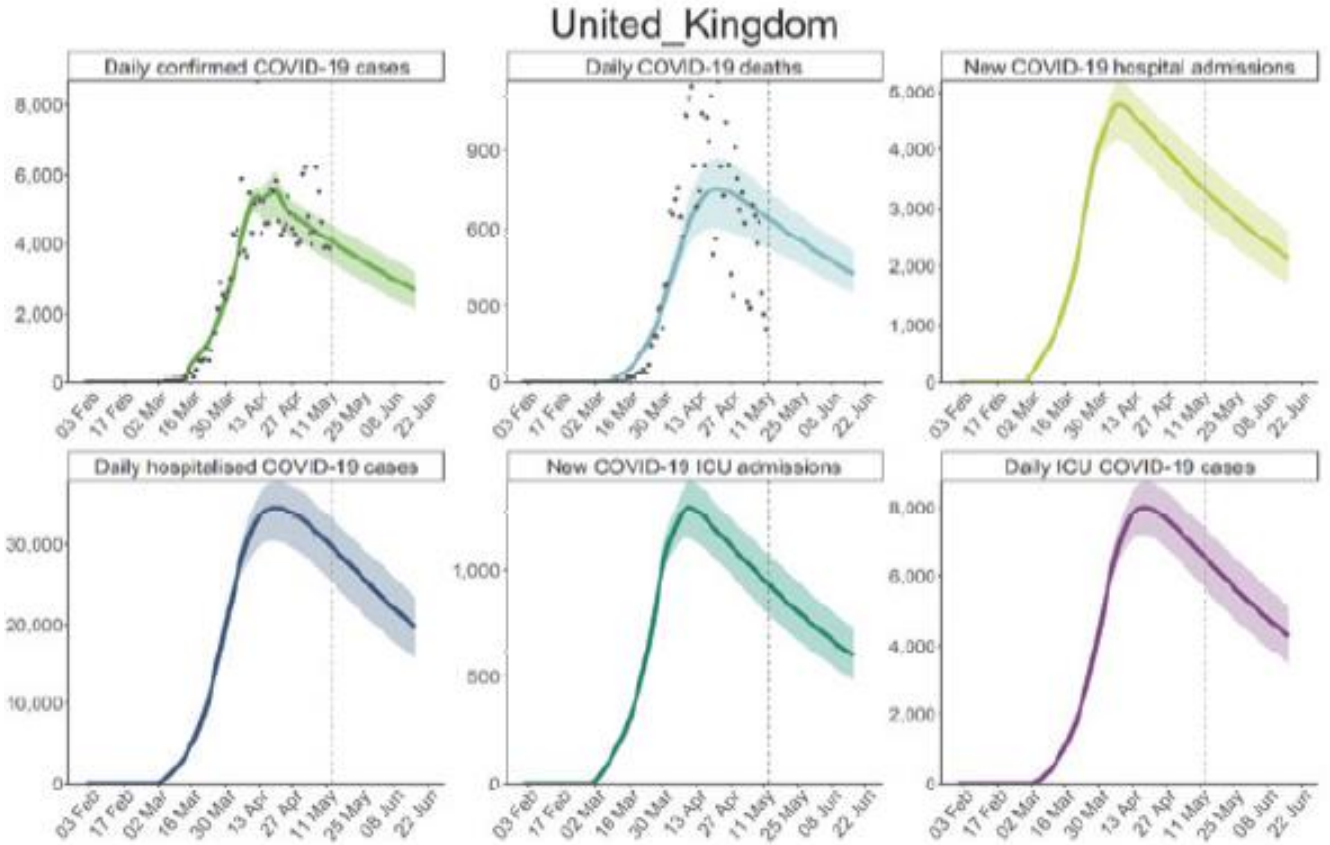

European Centre for Disease Prevention and Control. Projected baselines of COVID-19 in the EU/EEA and the UK for assessing the impact of de-escalation of measures – 26 May 2020. ECDC: Stockholm; 2020.

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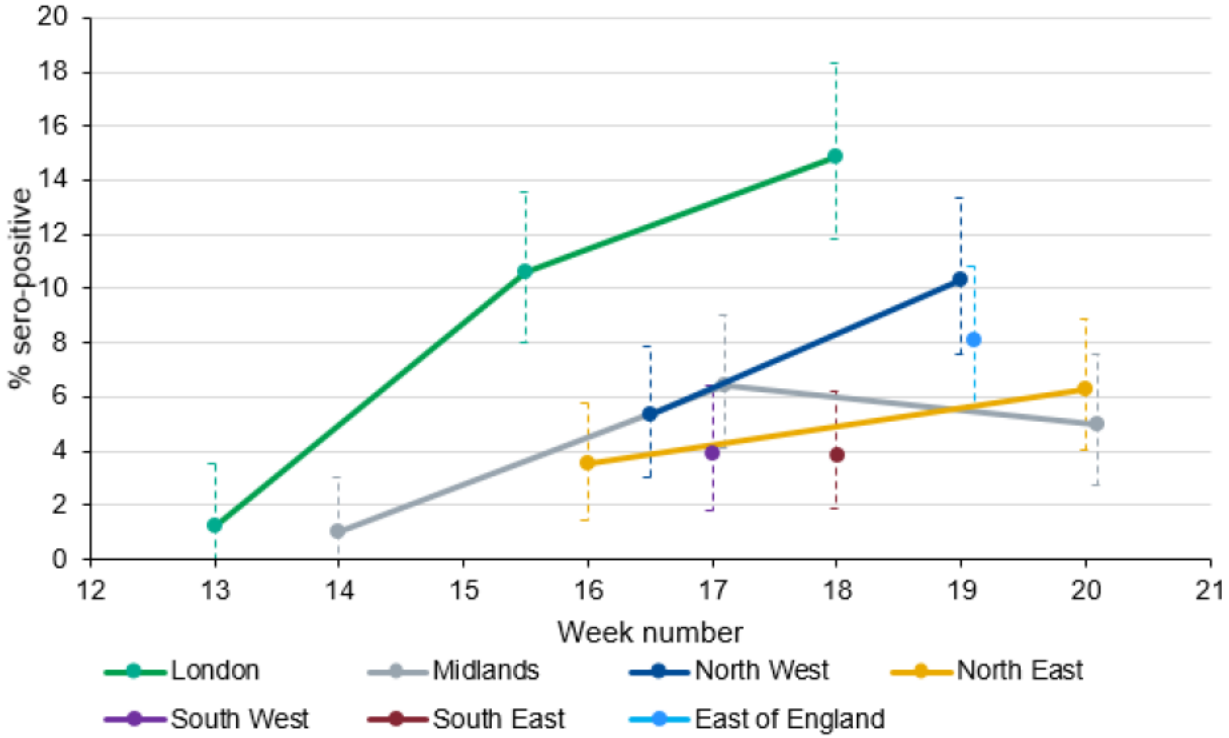
Antibodies to SARS CoV-2

Figure 29: Overall SARS-CoV-2 antibody Seroprevalence (%) in blood donors by PHE centres, using Euroimmun test adjusted for sensitivity (79%) and specificity (99%) and 95% confidence intervals (dashed lines)



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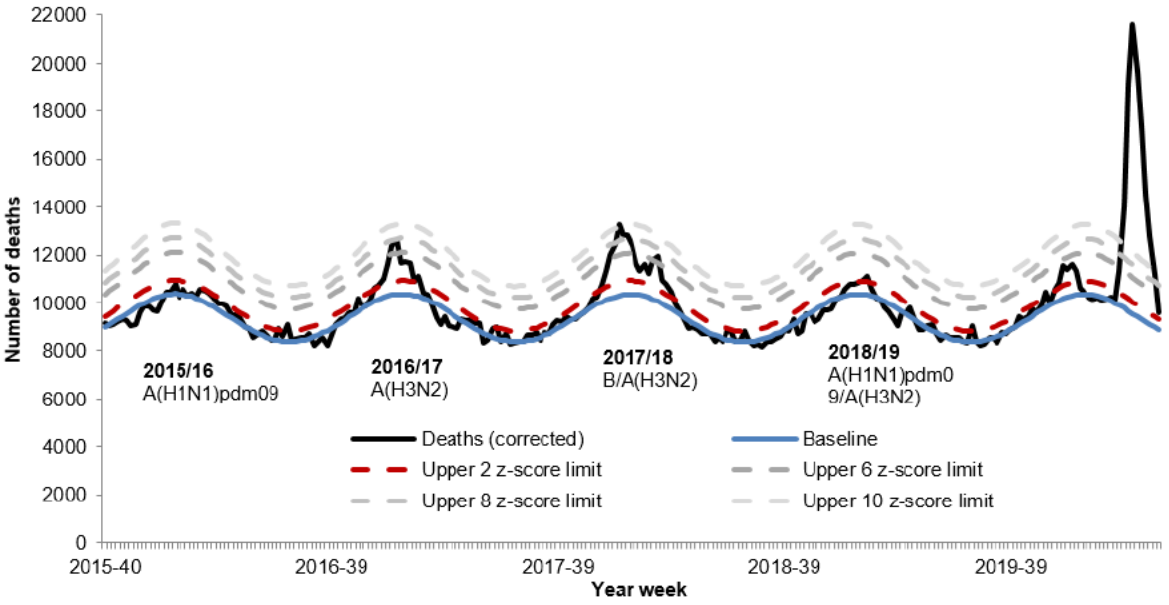
COVID-19 compared to annual influenza deaths

Figure 30: Weekly observed and expected number of all-cause deaths in all ages, with the dominant circulating influenza type(s), England, 2015 to week 21 2020



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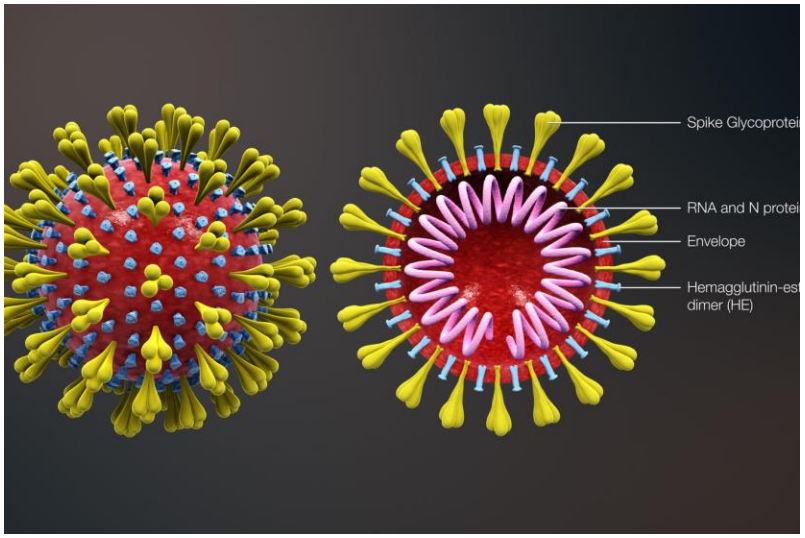
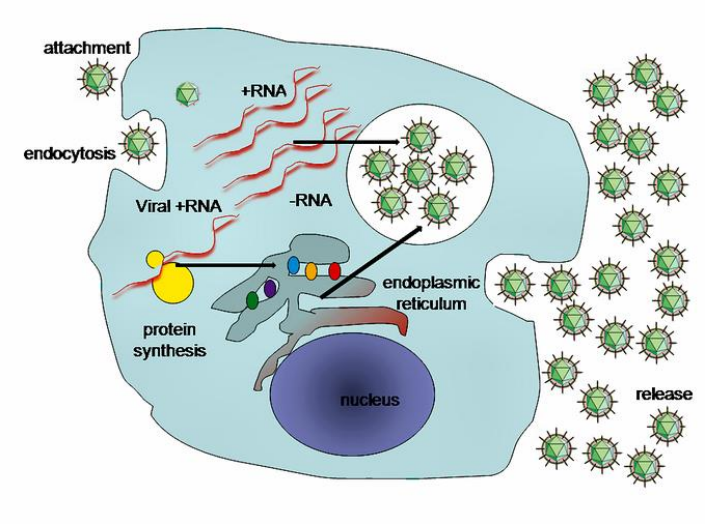
SARS-CoV-2: Severe Acute Respiratory Syndrome Coronavirus 2





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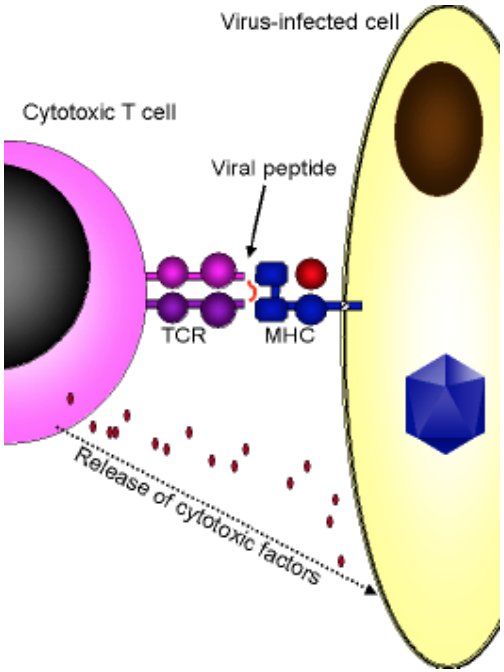
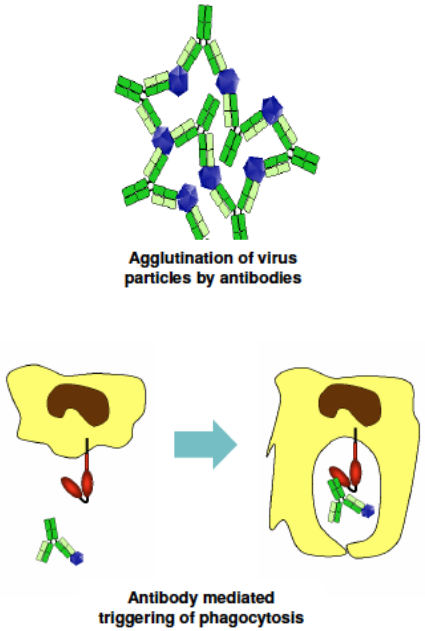
Immunity to viruses

Cell-mediated Immunity



Antibody-mediated Immunity



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How do vaccines work?

Vaccination is the safest way to protect your child against an infectious disease. Once your child has been vaccinated, they should have **immunity** to the disease.

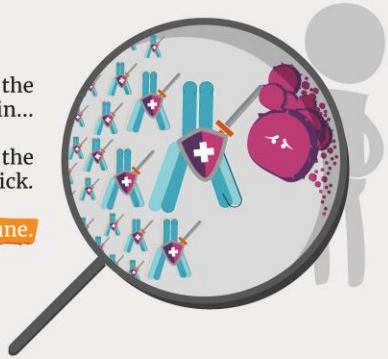
You are given a small amount of a harmless form of a disease...

...Then your body makes antibodies to fight it off

Then if you encounter the disease again...

...your body already has the antibodies, so you don't get sick.

You are immune.



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Celebrate Vaccines

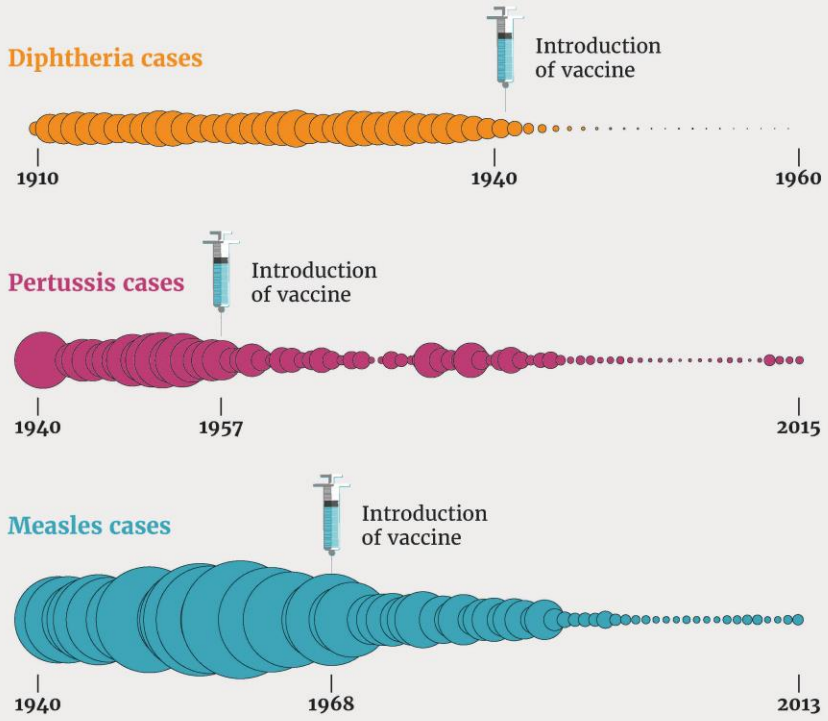
with the British Society for Immunology

https://www.immunology.org/celebrate-vaccines/public-engagement/infographics

How effective is vaccination?

Vaccines are considered one of our greatest global health achievements and are estimated to save **2-3 million lives a year**.

Thanks to vaccines, life-threatening diseases that used to be common in young children in the UK are now relatively rare.



British Society for immunology

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Celebrate Vaccines with the British Society for Immunology

https://www.immunology.org/celebrate-vaccines/public-engagement/infographics

Questions

- Why has the guidance changed for those shielding when the impact of the other relaxing measures is not yet known and we are still at stage 4?
- Is it really safe for shielding patients/families to go outside now? (Many state that when they have tried to go out for a walk they do not feel safe due to the number of people around)
- Will those shielding have to remain largely so until a vaccine is found or will there be further relaxation of the guidance?
- When will it be safe to resume carers coming into the home (where the service has been stopped)?

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The impact of COVID on IMD and plans going forward

Dr Elaine Murphy

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National Hospital for Neurology and Neurosurgery

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How many IMD patients tested positive with COVID-19 and what where the outcomes?

Paediatric Hospitalisations

(confirmed COVID positive)

Hospitalisations = 5 patients

Non-invasive ventilation required = 0

Intubation required = 0

Deaths = 0

Adult Hospitalisations

(confirmed COVID positive)

Hospitalisations = 8 patients

Non-invasive ventilation required = 0

Intubation required = 1

Deaths = 0 (all recovered)

As far as we are aware no patients with IMD / LSD in the UK have died due to a COVID infection

No one IMD in particular was more associated with risk of hospitalisation due to COVID

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How many IMD patients were admitted with a metabolic decompensation?

There were fewer admissions than normal with metabolic decompensation

- Protective measures against COVID-19 also work against other viruses / infections (social distancing, hand-washing etc)

- Very good compliance with taking medications and supplements; focus on good dietary routine

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How has the pandemic affected provision of treatment?

Enzyme replacement therapy (by intravenous infusion) was paused / put on hold for some vulnerable patients

- to facilitate shielding and protect patients from exposure to COVID
- patient choice

Fabry disease	87 of 560 patients treatment on hold / switched to oral therapy (82 adults)
Pompe disease	34 of 134 patients treatment on hold (33 adults)
Gaucher disease	25 of 215 patients treatment on hold / switched to oral therapy (25 adults)
MPS I	11 of 64 patients treatment on hold (11 adults)
MPS II	7 of 47 patients treatment on hold (4 adults)
MPS IVa	13 of 66 patients treatment on hold (8 adults)
MPS VI	6 of 38 patients treatment on hold (5 adults)

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How has the pandemic affected provision of treatment?

Surveillance or monitoring for complications has been reduced dramatically

Echocardiogram; Pulmonary function testing; Brain MRI; Bone density scans

Planned surgery has been delayed

So as the number of cases of COVID decreases we now need to decide on how best to continue to protect the most vulnerable patients, and to adjust services to manage chronic conditions.

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Virtual meetings - Paediatric

Paediatric – representative(s) from each centre invited (including NI, Wales, Scotland)

- Led by Drs James Davison and Hugh Lemonde
  - Friday May 1st
  - Agreement to support RCPCH document (on their website)
  - Two groups with regard to ongoing shielding
    - A – ongoing shielding will be recommended
    - B – ongoing shielding may be recommended depending on MDT discussion

Virtual meetings - Adult

Adult – representative(s) from each centre (including NI, Wales, Scotland)

- Friday May 29th
- Agreement to align with RCPCH (paediatric) document and guidance from other specialities

BIMDG Principles of Recovery (1)

We recognize that…

- There may be different geographical prevalence of COVID-19 in UK
- Staffing and resources (including staff illness and redeployment) may differ between centres
- The case mix of extremely vulnerable / not so vulnerable patients may differ between centres
- Trust requirements may differ with regard to re-opening outpatient clinics, arranging surveillance investigations, day-ward admissions etc
- There will need to be a flexible approach to any recommendations as these may need to change rapidly depending on any local or national resurgence of COVID-19

BIMDG Principles of Recovery (2)

We agreed in general…

- Patients should be advised to attend hospital for urgent / emergency care as usual
- Teams should ensure that all patients who require an emergency regimen have access to this
- As local services allow, and as deemed clinically appropriate, we will move towards seeing more newly referred patients face to face from July 2020

BIMDG Principles of Recovery (3, Shielding)

We agreed in general…

- Each unit will review patients currently on their shielding list in June.
- Decisions will be made locally by each team as to which patients remain on this list from July 1.
- Patients will be written to inform them whether or not they need to remain on the shielding list.
- A reason for remaining on the list, or removal from the list should be given to the patient.
- These letters should be copied to the patient's GP.
- From July 1, patients for whom enzyme replacement therapy (ERT) is currently on hold will be advised that the risk-benefit is likely to be in favour of restarting ERT. Patients will still have a choice whether to restart or not.
- Patients will be encouraged to become independent / semi-independent with ERT (if possible).
- Patients who are shielded will be contacted by their metabolic team every 8-10 weeks (staff permitting).

Re-opening hospital services….

Routine face to face outpatient appointments have generally been suspended and replaced with either telephone or video consultations

Only those patients deemed 'urgent' have been seen face to face (F2F)

Most trusts are now slowly moving towards re-opening more face to face services

This needs to be done in the context of

- social distancing in hospitals (corridors, waiting rooms)
- infection control measures (e.g. cleaning of rooms between patients; cleaning of equipment (x-ray scanners) between patients; adequate airflow and ventilation in clinic rooms)
- Staffing numbers (may be reduced to due illness / self-isolation / redeployment)

Fewer patients can be seen per day in outpatients; fewer investigations can be done per day; fewer people can be admitted electively to the hospital ie. waiting times for non-urgent issues will increase

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Re-opening hospital services….

- Those patients who are booked for elective procedures may be asked to isolate at home for 14 days prior to hospital admission
- Those patients who are booked for elective procedures will be tested for COVID-19 prior to hospital admission
- Hospitals will have specific designated COVID-positive (and COVID-free) areas
- There may be specific entrances and routes through the hospital (e.g. one-way, COVID-free)
- Staff will be wearing masks – and patients may also be asked to wear masks if possible
- The number of parents / carers allowed in the hospital with a patient will be limited
- Time slots for clinic appointments will be given (and may need to be adhered to more strictly)

This is to avoid the reintroduction of COVID into hospitals and transfer of COVID to vulnerable patients

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Rapidly increased access to technology….

As an example, in our trust, since the start of the pandemic……

We have been given
- dual computer screens
- good quality headsets and microphones
- webcams

And access to new software
- **Attend anywhere** – allows video consultations – patients' home, GP surgeries, outreach clinics etc
- **Health Information Exchange** – allows access to patients' records (GP and other hospitals)
- **Microsoft Teams** – allows us to run virtual meetings with the whole clinical team

Video / telephone consultation - first-line option for many patients at least until the end of this year. These options will continue to be offered if clinically appropriate even when the pandemic is over.

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Rapidly increased access to technology….

Video consultation

- May be possible to arrange at GP surgeries
- Already being used for outreach clinics
- Could be used to liaise with local hospital inpatient teams
- Trainee doctors

‘Patient-initiated appointments’ are being considered as a possibility

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Issues with telephone / video consultations

Less personal, harder to pick up on non-verbal cues

Not all patients have access to technology (we may be able to work with GPs to support patients) or WiFi

May be more challenging

- visual / hearing impairment
- Non-native English speakers

Not possible to examine patients or arrange tests on the same day

Getting blood tests is challenging

- Some specialist bloods – need processing quickly, may need to be stored frozen.
- We are investigating using more 'bloodspot' technology

Weight and heights needed (particularly for children to adjust diets and medications)

- Please consider buying a home scales and tape measure

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Restarting enzyme replacement therapy (ERT) at home

Centres have agreed that the risk-benefit assessment now favours restarting ERT for most patients who have discontinued this during the last few months

- More patients are contacting the metabolic centres with recurrence of symptoms (some patients have restarted ERT already)
- The R value is lower in the community
- Hospitals have capacity to treat patients if needed
- The homecare companies have comprehensive plans in place (including access to appropriate PPE) to protect patients (and themselves) – 'infection control procedures'
- More patients have expressed an interest in becoming (semi)independent with cannulation and infusions, and we will support this

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Over the next few weeks…

If you have previously been told to shield (by your metabolic centre) then you should expect a letter or contact informing you as to whether you should continue to shield, or follow advice as per the general population

If you are not contacted, or are unsure (or feel that you have / have not been sent a letter in error), then please contact your metabolic team directly

If your ERT is currently on hold your metabolic team will discuss with you what happens next

Ask your metabolic team; Metabolic Support UK; the LSD Collaborative or another reliable patient support organisation

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Paediatrics and COVID-19

Dr Roshni Vara

Evelina Children's Hospital
St Thomas' Hospital
London

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Social distancing and Self Isolation

- Children with IMD and families to adhere to current Government advice
- Social distancing where possible and isolate if patient / family or contact with symptoms
- Can only 'self isolate' as much as practically possible
- If a family member (not a main carer) is symptomatic, try to isolate as much as possible from child with IMD, particularly if high risk
- Consider asking neighbours and friends to help
- Higher risk; those at risk of metabolic decompensation (e.g. UCD, OA, GA1, FAOD, MSUD, Cbl disorders, mitochondrial) and those with pre-existing lung or heart disease (NPA/B, MPS with severe lung or airway disease, severe neurological disease)

Remember: the evidence is that COVID-19 is a mild infection for the majority of children – so we hope they will be able to cope as with any other infection

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Shielding

‘People have also been classed as clinically extremely vulnerable, based on clinical judgement and an assessment of their needs’

1.Patients should continue to take precautions but can now leave their home if they wish and continue to practice social distancing.
2.If you choose to spend time outdoors, this can be with members of your own household.
3.You should stay alert when leaving home: washing your hands regularly, maintaining social distance and avoiding gatherings of any size.
4.You should not attend any gatherings, including gatherings of friends and families in private spaces
5.You should strictly avoid contact with anyone who is displaying symptoms of COVID-19 (a new continuous cough, a high temperature, or a loss of, or change in, your sense of taste or smell).

The Government is currently advising people to shield until 30 June 2020 and is regularly monitoring this position.

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Acute care during crisis: a child with IMD

- If your child with IMD is unwell at home – please commence your oral ER as advised / usual
- Contact your metabolic team as per normal guidance
- If the oral emergency is not tolerated at home and your child has no respiratory symptoms – then follow your usual open access process
- If your child has respiratory symptoms and needs medical assessment / not tolerating ER at home – the metabolic team will liaise with your local hospital and advice accordingly (i.e. where and when to attend)
- End of life care issues – please contact your local PPC or metabolic team

Remember – you can still call 999 in an emergency.

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Clarification on the Kawasaki type symptoms in children

1. A child presenting with persistent fever, inflammation (neutrophilia, elevated CRP and lymphopenia) and evidence of single or multi-organ dysfunction (shock, cardiac, respiratory, renal, gastrointestinal or neurological disorder)

2. This may include children fulfilling full or partial criteria for Kawasaki disease.

3. Exclusion of any other microbial cause, including bacterial sepsis, staphylococcal or streptococcal shock syndromes, infections associated with myocarditis such as enterovirus (waiting for results of these investigations should not delay seeking expert advice).

4. SARS-CoV-2 PCR testing may be positive or negative

All stable children should be discussed as soon as possible with specialist services to ensure prompt treatment (paediatric infectious disease / cardiology / rheumatology*). There should be a low threshold for referral to Paediatric Intensive Care using normal pathways.

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Should parents/carers be requesting testing in case they are asymptomatic?

- Currently all inpatient paediatric patients are tested with swab testing
- For elective admissions (green pathway) – shielding is required for 14 days prior to admission with testing 48 hours prior to admission
- No travel on public transport

- Testing for asymptomatic individuals remains uncertain

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Returning to school

1.Clinically extremely vulnerable children to remain shielded and not to return to school, even if their year group has.
2.Clinically vulnerable children who are only under the care of primary care are overwhelmingly likely to benefit from returning to school when their year group does.
3.Clinically vulnerable children, who are under secondary or specialist care for an underlying health condition are, on the balance of probabilities, more likely to benefit from returning to school when their year group does so. These families may need a conversation with their treating teams to balance the potential risks and any familial anxiety.
4.All other children should attend school when their year group returns.

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Returning to school

- Should children with IMDs and/or their siblings (including those not shielding but social distancing) return to school if it is open for their year group?
  Yes

- If unaffected siblings can go, what safeguarding measures can be put in place for patients in household?
  As per standard guidelines for hand and respiratory hygiene

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Research and clinical trials

Dr Simon Jones

Willink Unit, Saint Mary's Hospital
Manchester

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Research and clinical trials

- Most clinical trials suspended or paused at the start of the COVID-19 crisis
- Research staff and hospital support systems realigned and redeployed to deal with expected COVID-19 patients
- Only trials continuing are those essential for delivery of treatment not otherwise available (a number of metabolic studies)
- Research staff also asked to become part of nationally and regionally prioritised COVID-19 related trials
  - Testing trials
  - Treatment trials

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Research and clinical trials

- As hospitals consider a return to routine care, other research may also restart
- The timescales for this vary widely and depend on research staff able to work on non-COVID-19 trials and hospitals being able to deliver the study
- COVID-19 related trials will continue for many months more using up research staff and infrastructure
- Gradual reopening of ongoing and new trials where the ability to deliver these exists and on priority
- Significant variability across regions and country

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Advice regarding coping and mental wellbeing

Dr Lynne Aitkenhead

Charles Dent Metabolic Unit,
London

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Coping and mental wellbeing

“We are all in the same storm, but we are not all in the same boats”

We are all facing a situation that is very uncertain, and that continues to cause difficult feelings including anxiety, sadness, guilt, and loneliness.

However, everyone is facing different circumstances, challenges, and decisions. We also each have different strengths and resources to help us.

This makes it hard to give general tips or advice.

There are a couple of psychology ‘tools’ that might be helpful, and extra support is available if you need it. Talk to your metabolic team, GP, or look back at the suggested resources in the last webinar.

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Coping and mental wellbeing

What should I do to move forwards now?

It might help to set set some positive goals for the next few months

Think about them in relation to your values

- Deep down inside, what is important to me?
- What sort of person do I want to be?
- What sort of relationships do I want to build?
- What do I want to channel my time and energy into doing?

Values provide the direction you want to travel in life, and can help motivate you

Make goals realistic and achievable, as well as specific and clearly defined

Break your goals down into small, manageable steps, and build up gradually from easy to difficult ones

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Coping and mental wellbeing

What can help me to make decisions that are right for me and my family?

Taking a structured approach can help you to think through problems clearly, especially if they are associated with strong emotions.

1. **Identify the problem.** What emotions are associated with it?
2. **List all possible solutions.** Be creative!
3. **Eliminate any less desirable or reasonable solutions and list the remaining ones in order of preference**
4. **Write down the advantages and disadvantages of the top few solutions**
5. **Decide on a plan, and break it down into steps**
6. **Put it into action, and check how well it worked.** What works well and what not so well? Do you need to make any changes?

You can download example problem-solving worksheets from www.getselfhelp.co.uk

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Questions

If you have questions relating to your / your child's clinical care, please contact your metabolic team direct.

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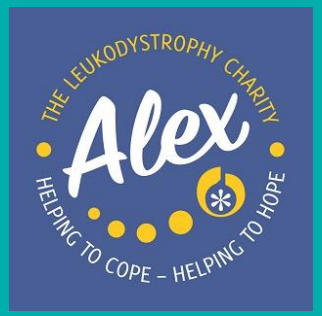
AGSD UK
Association for Glycogen Storage Disease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