

Medical

1. Our daughter's care home takes the temperature of all care workers and anybody entering the building. Is this a wise precaution?

APAS - Not everyone with COVID-19 will have a fever, so it is not a fail proof precaution. However, it is a quick and inexpensive way of detecting those who do have a fever. Provided non-contact thermometers are used, together with other measures, it could be somewhat helpful. Social distancing and hand hygiene remain the most effective measures to prevent spread of the disease. The quality of the thermometer could also be an issue, some are not very accurate.

2. If the PWRS gets symptoms how best to safely manage their care at home?

I guess this may vary as to a home with nursing or one without. With nursing we would expect they will have access to oxygen, SATs monitor, blood pressure monitors and the knowledge to provide care to a point, know when they person needs hospital admission.

If a home without nursing, they should have GP support but would expect to follow the health guidance <https://www.nhs.uk/conditions/coronavirus-covid-19/self-isolation-advice/> which also includes information about managing symptoms.

3. Can care homes where our Rett people are living get oxygen to keep in the home, just in case? Oxygen is a drug and hence needs to be prescribed by a qualified practitioner. Given wrongly it is highly dangerous and can be fatal. If someone needs oxygen, speak to the GP who can prescribe it on medical grounds.

Would it be possible to cover guidance for those living in a care home?

The home should have issued you with information about how they are managing the risk of infection, isolation, protocols they have put in place to help keep people safe. There is also this helpful document for the government. If you have concerns, do speak to the home manager.

<https://www.gov.uk/government/publications/covid-19-residential-care-supported-living-and-home-care-guidance>

4. One of our mums has been informed by GP that her daughter will get palliative care only, is this appropriate?

No this is not acceptable unless this has been previously discussed and agreed. We would advise families to do an [Advance Care Plan](#) and decide what you like to happen should the worst happen. There are also some good resources here for [Resus.org.uk](https://www.resus.org.uk) and guidance as to what you should have in a ReSpect Form.

5. Hi, I have been told by E's consultant that she isn't at increased risk because she doesn't have risk of chest infections or breathing problems - is that correct?

My 10-year-old daughter has Rett but i cannot remember the last time she was unwell with even a cough or cold. She has never had a chest infection or pneumonia and her seizures are under control. She has absences that is all. She takes regular medication so should I still register her as vulnerable on the government's website?

My child with Rett has very irregular breathing but no history of chest infections or immune issues to date - I am taking this as her being not in the very highest vulnerability category but should we be shielding her? We are isolating at the moment and have been for 3 weeks...apart from either my husband or myself going to the shops.

My daughter doesn't have any medical needs at the moment. Shall we still stay in self isolation for 12 weeks?

In response to the above four questions which all are very similar - Rett syndrome is a spectrum disorder; some people are more severely affected than others. They are all vulnerable to some extent, but some are certainly more frail and vulnerable than others. Everyone with Rett syndrome should be isolated as much as is reasonably possible, but those who would be deemed as being at even higher risk of having serious complications if they got the virus should be extra careful and shielded as much as possible.

To protect everyone and also to help the pressure on the NHS, everyone should be following the government guidance on - staying home unless it is an essential trip for food or medication, following strict social distancing rules (2 metres gap) if you do go for a walk (one form of exercise a day such as a walk or run) etc.

Coronavirus outbreak FAQs: what you can and can't do

<https://www.gov.uk/government/publications/coronavirus-outbreak-faqs-what-you-can-and-cant-do/coronavirus-outbreak-faqs-what-you-can-and-cant-do>

Also register your daughter or son on the government vulnerable persons register [here](#). Some people will have received letters but not everyone but you can still register [here](#) even if you have not received a letter, This should then give you priority for GP access and they will flag it up with supermarkets so you get a priority slot for home deliveries, as well as other local community organisations that can help. Rett is a rare disease on their list of people that fall into the extremely vulnerable groups.

6. What support shall we be seeing from our Paediatricians especially when our children are really at risk with specific lung issues? Also, shall these children be going out at all?

It does depend on the lung problem and the potential susceptibility of the child to lung infections. Social distancing and covering the mouth and nose and ensuring limited hand to face contact when going out should be sufficient. Parents should be able to make contact with their paediatrician via telephone consultation to take sensible advice.

7. I understand COVID-19 patients on ventilators are on dry circuits, would giving saline nebs be detrimental to a patient with COVID-19?

There is no evidence that nebulisers and humidifiers are helpful. Advice from PHE is that nebulisation is not a viral droplet generating procedure. The droplets are from the machine (liquid bronchodilator drug particles), not the patient. Nebulisation is not therefore considered a 'viral' aerosol generating procedure. Although, some experts disagree with this view as the patient will exhale aerosolized particles.

There are different nebuliser types and systems, some of which release no particles to the atmosphere and others that do. Standard jet nebulisers do produce a fine mist that would enter the atmosphere, but this had not been directly inhaled – it's "blown out" from the face mask from the nebuliser and almost certainly has very limited entry into the airways of the subject. Other devices work only on inhalation, so the patients gets the drug dose and none of the mist enters the atmosphere – just normal exhaled air from the subject. Correctly used these systems would seriously limit the potential for a virus loaded spray

Mucus thinning cough medicines do not work (in general) and cough suppressants (e.g. pholcodine) may be harmful as cough is an important protective reflex.

There are exercises that could help though. The lungs are full of tiny branching airways known as bronchi. Oxygen travels through these airways, ends up in tiny sacs called alveoli, and from there is absorbed into the bloodstream. These are the small airways. The inside walls of the bronchi are coated with sticky mucus, which protects against damage from particles moving down into the small airways. COVID-19 causes inflammation in the lungs which may close the small airways and/or alter the consistency and/or amount of mucus produced. There are breathing exercises (called active cycle of breathing) to help clear mucus and open small airways, although not every PWS will be able to do them. Some PWS may be able to do them by mirroring a carer. It is also important to avoid long hours lying on their back, this contributes to accumulation of mucus and closure of small airways.

There are several variations but the principle is the same, for a detailed demo see:

<https://www.youtube.com/watch?v=XvorhwGZGm8>

8. What about heart problems? The guidance is vague. I appreciate that the data may not be there, but is long QTC likely to be present a problem?

There is no specific information on how COVID-19 affects PWS. Those who have long QT could be at higher risk of arrhythmias if they develop hypoxemia (low blood oxygen), which happens in some people with COVID-19.

9. Also, after intensive care when somebody survives they may have significant problems afterwards, or need a prolonged recovery time?

Full recovery may take several weeks depending on severity. In the previous SARS epidemic (less infective but more deadly), a small percentage of patients had long-term effects from their illness, including depression or anxiety, cough, shortness of breath, chronic lung disease or kidney disease. However, most patients fully recovered.

10. I am worried should my daughter have to be taken to hospital, she would go alone. She could then die of something else because she would not get the care for a Rett syndrome person requires and could die of something else? My feeling is not to go to hospital in that situation.

There are parents scared of taking them to hospital because of being separated, some are deciding not to go in, they feel it's better to be with them than separated and having them potentially die alone. The young 13 year old in Kings was alone.

One parent or a carer can accompany a child to hospital and stay with them. Not sending the child would be putting them at risk and would potentially be a safe-guarding concern. Please always seek medical advice. Starting with NHS 111.

If it is an adult with Rett syndrome and they may have to go to ICU alone if they needed ventilation when they would be completely sedated.

We have been told that Brighton & Sussex University Hospital Trust has just agreed this policy in relation to adults with learning disabilities. 'Current hospital policy is that a carer remaining with a person with a learning disability is a reasonable adjustment. Their clinical presentation will be taken into account and if in consultation with yourself (carer) it is assessed as appropriate then they will be facilitated to have a carer present. Understandably things may change but this is the agreement with BSUH currently.'

Also, The University Hospitals Bristol & Weston NHS Trust has agreed that for a COVID+ patient in isolation can have a single visitor under exceptional circumstances provided a risk assessment has been undertaken and approval has been obtained from a senior clinician. For a person with Rett syndrome this should be regarded as an exceptional circumstance on the basis of communication limitations and specific carer-patient interactions.

It would seem that different trusts have different policies. Speak to the learning disability nurse at the hospital or PALS if you are not sure what the policy is where you are. Rett UK is lobbying along with other related charities to put pressure on the health service to facilitate this if it was at all possible.

Our advice at the moment is to do everything you reasonably can to reduce the risk of them being exposed to the virus, manage mild symptoms at home following the NHS advice about managing symptoms but do call 111 for further advice if temperature cannot be controlled and/or breathing becomes difficult.

For both children and adults it would be very good to make sure you have their medical/health and communication preferences up to date in accessible document such as the [Rett UK Health Passport](#) and have an Advance Care Plan following the ReSpecT principles covered in Q4.

11. So, it does seem that oxygen could be of significant help?

Up to a point. As the small airways close, delivering more oxygen to the ones that are still opened will be helpful. If the lung surface area for oxygen absorption is excessively reduced and/or the patient is not breathing well on their own, assisted ventilation will be or even extra-corporeal circulation may be required. There is no evidence that oxygen alleviates the infection itself it only alleviates a symptom.

12. What to do about the hand mouthing in Rett syndrome? I wash E hands as much as I can, and she isn't going out other than to the garden, but her hands are always in her mouth.

Ensuring surfaces are regularly cleaned and thoroughly washing hands, including the backs, as much as possible, is probably as much as you can do. Splinting may cause further distress.

13. Are there any statistics for the survival rate if you require ICU?

World Health Organisation 50/50 from going into ICU in the general population but this is lower in elderly people. Only 20% of those affected need to go to hospital and of those, 20% will need ICU.

14. What is the expectation of this virus to impact the brain of Rett girls?

Some specialists are suggesting that the virus could penetrate the brain, particularly the brainstem, and negatively affect brain activity (see <https://dx.doi.org/10.1002/jmv.25728>). There is no data in PWRs and no evidence of long-term damage. It is expected that a severe infection (by any pathogen) and/or hypoxemia (low blood oxygen) would negatively affect brain function even in neurotypical people. This could be more pronounced in the elderly and those who already have some level of cognitive impairment. However, the main tropism of the virus is for the lungs and digestive system.

15. My daughter is due to come onto the neuro ward at the Evelina for a procedure under anaesthetic which has been cancelled once due to the current situation, she urgently needs her gastro tube changing so how and when is it safe to let this happen?

All planned procedures under general anaesthetic are being reviewed, and where ever possible are being postponed. There is very limited theatre capacity running at the moment, and so when procedures can't be cancelled, they can be performed.

How long things are postponed will probably depend upon how long the current pandemic conditions last. It would be advisable to contact the team who had been planned to perform the PEG change and to discuss when it might be performed.

16. Could I please ask, is there anything that we can put onto hands specifically to help them? Due to the amount of hand sanitizer that we have been using along with washing them constantly seems to be causing really sore and dry hands which is difficult as the hands are already sore before this.

A thick heavy-duty moisturiser to stop cracks appearing and posing an infection risk would be good.

Hygiene/Infection Control

17. Should we be washing loose fruit and veg in something more than plain water and if so, what?

Yes, preferably buy packaged fruit and vegetables, then remove and dispose safely of the package. Wash fruit and vegetables even if not eating the peel, as cutting into the unwashed peel can contaminate the flesh.

Wash everything in soapy water but rinse copiously. Be aware that this may reduce the shelf life of some items, especially citrus fruit. Pre-packed salads are generally safe as they are washed in chlorine. Chlorine tablets for disinfecting fruits and vegetables can also be bought online (always follow label instructions).

Avoid pre-packed sandwiches or any food that has potentially been prepped/handled by several other people. Food that can be heated/re-heated at home is safe as the virus is destroyed by heat (>63° C).

18. Are wearing face masks effective? Can you please talk through in what situations they would be appropriate to wear in a care home please?

When using a mask, should you throw it away after each use/contact with the poorly person?

Some reliable information here from the World Health Organisation

Yes, it would be appropriate to wear masks in a care home and they should have been issued with PPE (personal protective equipment)

19. How do you clean a supermarket trolley, will wipes, gels do?

Anti-bacterial wipes will work if they contain alcohol or soap (check label for surfactants). Antibacterial chemicals like chlorhexidine or benzalkonium chloride are not as effective at killing the virus. In any case, avoid touching your face while outside your home; upon returning wash your hands thoroughly before touching surfaces inside your home.

20. Does bleach work as well as soap and water for surfaces?

Yes, chlorine solutions of 1000 ppm killed the virus in < 10 seconds in laboratory tests.

Isolation related

21. We have had COVID-19 diagnosed in our household when would it be safe for us to see our daughter who is in supported living?

Please refer to the government advice

<https://www.nhs.uk/conditions/coronavirus-covid-19/self-isolation-advice/>

22. Is my daughter safe to go into a supermarket if I cover her over with a rain cover?

This would provide some additional protection but you need to make sure it is cleaned thoroughly and put away before taking your daughter out of the buggy or wheelchair. Ideally best if she did not go to the supermarket with you. Can you arrange to have shopping delivered using one of the local support groups or online shopping?

23. Hello, my daughter has mild Rett with no underlying health issues, I am a key worker and she will have to go to childcare next week, is this OK or should I isolate her for the 12 weeks? I am front line NHS with a daughter with Rett who lives with me. Thank you for the advice.

Generally, as long as the childcare setting are taking all the necessary precautions that everyone else is taking in terms of social distancing, hand

washing etc. and that carers wear masks etc. then it is probably OK. If any carers have symptoms they should not be in at all. When home, change of clothes etc. as response to Q24 would seem reasonable for both parent and child.

24. If my partner is a paramedic is it still safe for him to live in the home?

Ideally, they should stay somewhere else. If not possible, upon arriving home, remove shoes, change clothes and have a shower before having contact with the PWS. Keep outdoor items (e.g. keys, money, credit cards, handbags) in a safe 'dirty' area and avoid touching them while at home. Clean items you will be touching whilst inside the house (e.g. mobile phone). If feasible, wearing a surgical face mask at home when in the same room as the PWS would be ideal. Keep windows opened as much as possible and let the sun in. Always observe strict hygiene of hands and surfaces. Ideally, high risk frontline worker should avoid preparing food and feeding the PWS. If not possible, wear face mask while handling food.

25. Is it safe to send my daughter to school? She does not have chest issues but has epilepsy. We're both NHS workers but one of us can work from home.

As much as possible the government wants children to stay home so if one parent can work from home that would be the recommended option.

26. Is it ok for a child with Rett to travel between parents if they don't live together?

If both parents had been sensible about isolating, social distancing, hygiene measures in the homes etc. and the mode of transport was parents car to and from each other's homes then yes it should be safe, although the government is discouraging non-essential travel. If the arrangement is giving the other parent a much needed break then we would think this was justified.

27. Is it safe to take her out for a walk?

Yes, if you follow the social distancing rules and follow the hygiene guidance upon returning home - shoes off at door, washing hands thoroughly straight away, etc. Those who are particularly vulnerable and are living in 'virus hot spots' should perhaps limit this to the garden if they have one.

28. My daughter's dad is still going to work, is she at higher risk of catching COVID-19, and how can we keep her safe?

That may depend on his type/place of work. It would certainly be advisable for him to have clean clothes to change into when he comes in, leave shoes at the door, put worn clothes straight in the washing machine, wash hands thoroughly and have a shower/wash hair before doing anything else. Wipe

down germ hot spots like door handles, keys, money etc – e.g. don't empty coins on table.

Additional info Extremely Vulnerable Person Register

Should everyone with Rett syndrome [register](#) on the government extremely vulnerable person list? Advice is yes as long as they meet the criteria.

Many people have and did get a text message yesterday saying the NHS will review your info and get back to you. Some people have had letters. There does seem to be regional variations. Other people have been flagged up either by their learning disability team within adult social care or through Continuing Health Care nurse at their CCG if they receive CHC funding.

Any further information and advice about this system please and it's benefits?

If you think you have been missed of the register and should be flagged as being extremely vulnerable contact your GP in the first instance for advice.

Once you have been flagged as extremely vulnerable it should give you priority access to your GP, priority for online shopping home delivery and other local support.

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