

14th November 2020

Since the announcement of the second national U.K. lockdown, many families have contacted Rett UK and Reverse Rett regarding whether their child is considered clinically extremely vulnerable and should stay home as per government [guidance on shielding and protecting people who are clinically extremely vulnerable from Covid 19](#).

There is no clear evidence-based guidance on whether all patients with Rett Syndrome should be considered clinically extremely vulnerable or not. These are the recommendations from the Centre for Personalised Medicine in Rett Syndrome (CPMRS), based on the limited information available, and consensus from multiple clinicians, researchers, and patient groups' perspectives.

A. Patients with Rett Syndrome should be considered extremely clinically vulnerable if:

1. The patient is on assisted long term ventilation, either via tracheostomy or via non-invasive ventilation
2. The patient has significant impairment in the ability to cough and clear away secretions.
3. The patient requires a cough assist device to help with clearance of airway secretions.
4. The patient has an unsafe swallow with poor saliva control and experiences frequent choking / aspirating on their own secretions.
5. The patient has another condition, unrelated to Rett Syndrome that in and of itself qualifies them for CEV status.

The above guidance re CEV status is aligned with current guidance from the Royal College of Paediatrics and Child Health (RCPCH) and government guidance on Shielding and Protecting People who are Clinically Extremely Vulnerable from COVID-19 dated 4 November 2020

<https://www.rcpch.ac.uk/resources/covid-19-guidance-clinically-extremely-vulnerable-children-young-people#children-who-are-clinically-extremely-vulnerable>

<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#cev>

B. All other patients with Rett Syndrome remain at higher risk for complications from Covid-19 than the general population but are not necessarily considered 'extremely clinically vulnerable'.

C. As would be expected, one should contact health professionals for advice if you think that the child/adult with Rett syndrome's health has deteriorated.

D. What can be done to identify Covid-19 and to reduce risk in Rett Syndrome?

1. Patients with Rett are at increased risk from complications than healthy, neurotypical children and adults.
2. It is more difficult to identify Covid-19 in patients with Rett of any age at an early stage, than in healthy populations due to the inability to communicate pain or discomfort, lack of taste and smell and a low baseline body temperature which is often present.
3. Apart from the typical symptoms of fever and persistent cough, the key is to look for rapid breathing with declining oxygen saturation levels.
4. **Preliminary research suggests that Vitamin D supplementation** may reduce the risk of complications (Ali N, 2020). This is of particular relevance to patients with Rett Syndrome who often have low Vitamin D levels (Motil KJ et al, 2011).

References:

- Ali N. Role of vitamin D in preventing of COVID-19 infection, progression and severity. *J Infect Public Health*. 2020 Oct;13(10):1373-1380. doi: 10.1016/j.jiph.2020.06.021. Epub 2020 Jun 20. PMID: 32605780; PMCID: PMC7305922.
- Motil KJ, Barrish JO, Lane J, Geerts SP, Annese F, McNair L, Percy AK, Skinner SA, Neul JL, Glaze DG. Vitamin D deficiency is prevalent in girls and women with Rett syndrome. *J Pediatr Gastroenterol Nutr*. 2011 Nov;53(5):569-74. doi: 10.1097/MPG.0b013e3182267a66. PMID: 21637127; PMCID: PMC3638258.