



## COVID-19 Risk Assessment Grid for Patients with a Rare Genetic Disease

This grid is intended to be a helpful general guideline for clinicians and people with rare genetic disease, to help them decide on the degree of self-isolation. If you are under a specialist medical team and/or take prescribed medication, specific advice from them should be taken to confirm your level of risk.

Patients falling into the extremely vulnerable (very high risk) category should follow government advice on **shielding**:

<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>

Patients falling into the clinically vulnerable (high risk) category should follow government advice on **stringent social distancing**. See Section 8 of Stay Alert and Stay Safe: <https://www.gov.uk/government/publications/staying-alert-and-safe-social-distancing/staying-alert-and-safe-social-distancing#clinically-vulnerable-people>

Patients at population risk should follow the general **social distancing** advice in Stay Alert and Stay Safe:

<https://www.gov.uk/government/publications/staying-alert-and-safe-social-distancing/staying-alert-and-safe-social-distancing>

Extremely Vulnerable (Very High risk) – need to shield	Clinically Vulnerable (High risk) – should practice stringent social distancing	Population risk - should practice general social distancing (Stay Alert and Stay Safe)
<ol style="list-style-type: none"> <li>1. You have had an organ transplant</li> <li>2. You have chronic kidney disease that requires dialysis OR you have protein in your urine (nephrotic syndrome) OR you have a history of frequently relapsing nephrotic syndrome</li> </ol> <p>Further valuable advice is available from the <a href="#">Renal Association website</a>)</p> <ol style="list-style-type: none"> <li>3. You have severe liver disease such as:               <ol style="list-style-type: none"> <li>a. liver transplant requiring immunosuppression OR</li> <li>b. autoimmune hepatitis requiring immunosuppression OR</li> </ol> </li> </ol>	<ol style="list-style-type: none"> <li>1. You are over 70 years old</li> <li>2. You have learning difficulties</li> <li>3. You have chronic kidney disease but are NOT on dialysis</li> <li>4. You have compensated liver disease not requiring immunosuppression.</li> <li>5. You have a chronic lung condition but this is NOT severe. For example:               <ol style="list-style-type: none"> <li>a. Mild bronchiectasis</li> <li>b. Mildly reduced chest size</li> </ol> </li> </ol>	<p>Individuals with rare disease will not necessarily be at increased risk compared with the general population if they do not meet any of the criteria to the left.</p>

<p>c. decompensated liver disease.</p> <p>4. You have had your spleen removed (splenectomy).</p> <p><i>NB. Recent guidance suggests that <b>not all</b> splenectomy patients may need to shield. The decision as to which patients with splenectomy are extremely vulnerable is complex and depends upon the underlying condition and reason for splenectomy. For detailed specific guidance, see the <a href="#">British Society of Haematology</a>.</i></p> <p>5. You have a <b>severe</b> lung condition such as:</p> <ul style="list-style-type: none"> <li>a. Cystic fibrosis, primary ciliary dyskinesia or similar condition</li> <li>b. Pulmonary fibrosis, interstitial lung disease or pulmonary hypertension</li> <li>c. Severe asthma – you have needed steroids by mouth four or more times in the past 6 months AND you take long acting bronchodilators eg salmeterol with or without inhaled steroids or leukotriene</li> <li>d. Bronchiectasis AND you: <ul style="list-style-type: none"> <li>i. Are receiving prophylactic antibiotics OR</li> <li>ii. Have chronic pseudomonas infection OR</li> <li>iii. Have 3 or more chest infections a year OR</li> <li>iv. Have severe airflow obstruction or significant breathlessness (MRC breathlessness score of 3-5)</li> </ul> </li> <li>e. Have chronic obstructive pulmonary disease (COPD) or emphysema and are on triple inhaled therapy (long acting beta agonists, long acting muscarinic agonist and inhaled steroid) OR have been prescribed Roflumilast Since November 2019</li> <li>f. Have a significantly reduced chest size due to a skeletal condition such as a skeletal dysplasia or severe scoliosis</li> </ul>	<p>c. Mild scoliosis</p> <p>d. Obstructive sleep apnoea that does NOT require non-invasive ventilation</p> <p>6. You have a chronic neurological condition for example that causes hypotonia, significant developmental delay or increased risk of recurrent infections, but DOES NOT severely affect breathing or swallowing</p> <p>7. You have a heart condition that either <b>restricts daily life or causes symptoms</b> (such as heart failure, significant valvular disease, cardiomyopathy, or angina) but you are NOT pregnant and are otherwise healthy</p> <p>8. You have congenital heart disease in the following groups:</p> <ul style="list-style-type: none"> <li>a. Infants under 1 year with unrepaired congenital heart disease requiring surgery or catheter intervention e.g. VSD, AVSD or tetralogy of Fallot OR</li> <li>b. Patients with congenital heart disease on medication to improve heart function</li> </ul> <p>Further valuable advice is available from the <a href="#">British Congenital Cardiac Association website</a></p> <p>9. You have had a stroke or suffer from vascular dementia</p> <p>10. You have diabetes</p> <p>11. You have high blood pressure (hypertension)</p> <p>12. You are pregnant</p> <p>13. You have a BMI over 40</p> <p>14. You are taking a medication which <b>moderately</b> suppresses the immune system. <b>You should discuss your use of these medicines with the specialist who prescribed them.</b></p>	
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- g. Are on long term (non-invasive) ventilation, OR use a cough machine OR home oxygen regularly.

Further valuable advice is available from the [British Thoracic Society website](#).

- 6. You have a severe neurological or neuromuscular condition:
  - a. Such as a muscular dystrophy which currently affects your breathing or lung function due to muscular weakness of the chest or diaphragm OR
  - b. That currently affects your ability to swallow OR
  - c. That currently affects the function of your heart e.g. includes cardiomyopathy OR
  - d. Involving a mitochondrial condition.

Further valuable advice is available from the [Association of British Neurologists website](#) and the [British Paediatric Neurology Association website](#).

- 7. You have heart disease of the following types:
  - a. Fontan, single ventricle physiology with evidence of 'failure' and / or end organ damage OR
  - b. Those with chronic cyanosis (oxygen saturations <85% persistently) OR
  - c. Pulmonary Arterial Hypertension (PAH) especially those on pulmonary vasodilator therapy OR
  - d. Patients with severe cardiomyopathies requiring medication OR
  - e. You have congenital heart disease and other significant illnesses e.g. chronic kidney disease or chronic lung disease

<p>f. You have congenital or acquired heart disease AND you are pregnant</p> <p>Further valuable advice is available from the <a href="#">British Congenital Cardiac Association</a> and the <a href="#">British Heart Foundation</a>.</p> <p>8. You have a rare genetic disease which:</p> <p style="padding-left: 20px;">Gives you a much higher risk of getting infections, for example, immunodeficiency syndromes or sickle cell anaemia OR</p> <p style="padding-left: 20px;">Has led to 2 or more infections requiring hospital admissions in the last 12 months OR</p> <p style="padding-left: 20px;">Has resulted in you receiving prophylactic antibiotics to prevent recurrent infections</p> <p>9. If you currently have cancer or have previously been diagnosed with cancer, you <b>may be</b> extremely vulnerable. You should check with your doctor as your level of risk depends on the type of cancer, when it was diagnosed and the treatment you have received.</p>		
<p>If you are taking a medication which <b>significantly</b> suppresses the immune system (immunosuppressants), have received a bone marrow transplant (BMT) or have required immunosuppression over many years e.g. repeated courses of cyclophosphamide/biologics /or repeated high dose corticosteroids you <b>may be</b> at very high risk. <b>You should discuss your use of these medicines with the specialist who prescribed them for further advice.</b></p>	<p>If you meet <b>more than 1 of the High risk criteria</b> in this column this <b>may</b> increase your risk to Very High. You should discuss this with your doctor as a decision regarding your risk will require a clinical judgement based on your individual circumstances</p>	

**NOTES:**

1. We use the terms ‘very high risk’/ ‘extremely vulnerable’ to indicate a person who should shield, and ‘high risk’/ ‘vulnerable’ to indicate those at an increased risk compared with the general population but are currently advised to self-isolate in the same way as everyone at present, as defined by NHS England and Public Health England. However please note that some groups are using different terminology eg ‘high risk’ to determine the criteria for those who should shield

2. Rare diseases lead to many systemic manifestations. In addition to the original criteria defined by NHS England, this document summarises guidance from the [British Society of Gastroenterology](#), the [British Paediatric Neurology Association](#), the [Association of British Neurologists](#), the [British Thoracic Society](#), the [British Congenital Cardiac Association](#), the [Renal Association](#), the [British Society for Haematology](#) and the [British Society of Rheumatology](#) and we therefore wish to acknowledge their contribution to this work.
3. Further practical information and advice for individuals with rare genetic disease can be found in our Covid19 Information sheets. See <https://www.geneticdisordersuk.org/information-and-advice-on-coronavirus-covid-19/>