COVID-19 - guidance on clinically extremely vulnerable children and young people

NOTE: this advice was published in November 2020.

This paper provides advice to members on which paediatric patient groups are considered to be clinically extremely vulnerable during the COVID-19 outbreak and at very high risk of severe illness from coming into contact with the virus.

This advice was developed in June 2020 (and updated in November 2020) in partnership with a wide range of paediatric specialty groups: British Association of Paediatric Nephrology, British Association of Perinatal Medicine, British Congenital Cardiac Association, British Inherited Metabolic Disease Group, British Paediatric Allergy, Immunity & Infection Group (working with the UK Primary Immunodeficiency Network), British Paediatric Neurology Association, British Paediatric Respiratory Society, British Society for Paediatric Endocrinology and Diabetes, British Society of Paediatric Gastroenterology, Hepatology and Nutrition, British Society for Rheumatology, Children’s Cancer and Leukaemia Group, Paediatric Special Interest Group of British Haematology Society. Many specialties also worked with parents and patient groups as they developed their advice.

Children who are clinically extremely vulnerable

The RCPCH has been reviewing the evidence base around the impact of SARS-CoV-2 infection on children and young people with comorbidities which has been developing over time. We are also working with paediatric specialties to review this evidence and advise on which children and young people are at the highest risk of severe disease due to SARS-CoV-2 infection because they are ‘clinically extremely vulnerable’ (CEV). This evidence includes the following:

- Research evidence summaries
- Service evaluation and audit on the care needs of children admitted to hospital (England)
- Systematic review of evidence about milder outcomes in children

We know that the vast majority of children with conditions including asthma, cystic fibrosis, diabetes, epilepsy and kidney disease are not CEV.

In principle:

- Children and young people who are cared for only by their GP will not be CEV.
- A small group of children are CEV due to their pre-existing condition or specialist treatment (Group A).
- A further group of children exists who due to their underlying condition and specialist treatment may possibly be CEV (Group B). This will be determined on individual basis, in discussions between the clinician, the child and their family. Of note, the majority of children in this group are not CEV.
Identifying children and young people who are CEV

Our advice identifies children and young people (under 18 years of age) who are clinically extremely vulnerable (CEV) due to the risk of severe disease caused by SARS-CoV-2 infection. The evidence gathered since the start of the pandemic indicates that the risk of severe disease caused by SARS-CoV-2 infection in children is extremely low and although no one group of conditions has been identified as being at particular risk, specialists have identified those conditions which may make the child or young person CEV.

These children are usually CEV under normal (non-pandemic) circumstances, and at risk of severe disease due to a variety of infections which would result in mild disease in the majority of the population. Before the pandemic many of the children and young people in Group A and some in Group B would have been advised from time to time to not attend school due to their clinical condition or the treatment required to manage it. Similarly, their families and households would be advised to take extra care around hygiene and infectious contacts.

Group A

Group A lists conditions that mean a child or young person is clinically extremely vulnerable (CEV).

**Immunodeficiency and immunosuppression**

- Children with risk of severe infection due to their primary immunodeficiency. Only a small number of children fall into this category. More advice for clinicians is available from [UK Primary Immunodeficiency Network](https://www.piduk.org.uk). Advice for parents is also available from [PIDUK](https://www.piduk.org.uk).
- Children at risk of severe infection due to immunodeficiency induced by their disease or their drugs as part of their therapy (for example, some post-transplant immunosuppression, severe vasculitis). This may include children who are clinically vulnerable during the period before and after transplants. The duration of immunosuppression may differ for solid organ transplant and stem cell transplant. Specific guidance for children and young people on immunosuppression will be specialty specific and depending on their disease as well as medications prescribed.

**Oncology**

Children with very specific immunosuppression as part of their cancer therapy. This means those who:

- are receiving induction chemotherapy for acute lymphoblastic leukaemia (ALL) and Non-Hodgkins Lymphoma
- are receiving chemotherapy for acute myeloid leukaemia (AML)
- are receiving intensive chemotherapy for relapsed and/or refractory leukaemia or lymphoma
- have received a donor stem cell transplant (allogeneic transplant) in the last 12 months
- have received their own stem cells back (autograft transplant) in the last 6 months
- are undergoing CAR-T therapy and for 6 months following CAR-T therapy.

More advice is available from the [Children’s Cancer and Leukaemia Group](https://www.ccleg.org.uk).
Group B
Group B lists conditions that require discussion between the clinician and the child and their family/carer to establish whether they are clinically extremely vulnerable (CEV) on a case by case basis. This decision will depend on the severity of the condition and knowledge that the secondary and tertiary care clinical teams have of the particular circumstances of the child. We recognise that most children with conditions listed in Group B will not be CEV. Although many diseases are treated with similar immunomodulatory drugs, advice may differ between conditions as an assessment of clinical vulnerability is based on a combination of the drug effect and the underlying disease.

Note: there may be other patients who do not fit these categories below or under other specialties, but secondary care clinicians feel, after discussions with families, that an individual child is CEV. We advise contacting their tertiary specialists for advice.

Cardiology
- Fontan, single ventricle physiology, especially with evidence of ‘failure’, and or end organ damage
- Persistent cyanosis
- Pulmonary Arterial Hypertension (PAH) especially those on pulmonary vasodilator therapy
- Severe and or symptomatic heart failure, particularly those on heart failure therapy
- Pregnant young mothers with some congenital cardiac abnormalities

More information is available from the BCCA.

Haematology
- For children with sickle cell disease, this means those:
  - with additional co-morbidities causing concern from their clinicians (for example, progressive critical neurovasculopathy, severe or symptomatic heart failure)
  - with a history, within the preceding 12 months, of either one or more chest crisis requiring intensive care treatment or two or more chest crises requiring treatment.
- For children with thalassaemia, this means those with severe iron overload (T2 *< 10 ms) and additional co-morbidity causing concern.
- For children with Diamond Blackfan Anaemia, this means those who have an associated immunodeficiency, severe iron overload (as per thalassaemia definition) or are on prednisolone (or equivalent) ≥0.5 mg/kg/day.
- For children with other rare inherited anaemias, for example. pyruvate kinase deficiency, congenital dyserythropoietic anaemia, if they are at particularly high risk due to iron overload as per thalassaemia guidelines above.

Note: Alone, asplenism due to surgery or functional asplenism is not a reason to shield, but could be considered if other co-morbidities.
Immunodeficiency

- **HIV:** Only children and young people who have a CD4 count less than 50 or who have had an opportunistic illness within the last six months (or who have one of the other CEV conditions listed) should be considered to be CEV. We recommend discussion with tertiary specialist if any doubt. Note that advice differs from that for primary immunodeficiency.

More advice for clinicians is available from Children’s HIV Association, as well as advice for parents.

- **Primary immunodeficiency:** Most immunodeficiencies, in particular those involving antibody deficiencies, do not make a child or young person clinically extremely vulnerable (CEV).

More advice for clinicians available from UK Primary Immunodeficiency Network. Advice for parents is available from PIDUK.

Neonatal

- Ex-premature infants with oxygen and/or intermittent non-invasive ventilation requirements

Neonatalogists may also consider the advice offered for cardiology and respiratory patients for information about specific cardio-respiratory risk factors that might also pertain to the neonatal group.

Nephrology (kidney medicine)

- Those with recent kidney transplants – first three months immediately after transplant
- Those on a high level of immunosuppressive medication for active disease undergoing induction treatment
- The kidney team determines with the family that the child is at high risk

More information available from the British Association for Paediatric Nephrology and the Renal Association.

Neurology

- Patients with significant difficulty with swallowing (eg myotonic dystrophy patients)
- Patients at significant risk of decompensation during infection (eg mitochondrial disease)
- Patients with symptomatic heart failure, particularly those on heart failure therapy (eg Duchenne muscular dystrophy)
- Patients with myasthenic syndromes

More advice is available from the British Paediatric Neurology Association.

Paediatric gastroenterology, hepatology and nutrition

Paediatric inflammatory bowel disease (IBD) patients who meet one or more of the following criteria:

- Commencement of biologic therapy plus immunomodulatory or systemic steroids within previous six weeks
- Moderate to severely active disease not controlled by moderate risk treatments who may require an increase in treatment
Intestinal failure patients requiring Home Parenteral Nutrition (HPN) who meet one or more of the following criteria:

- Primary immunodeficiency or immunodeficiency induced by drugs as part of their therapy.
- Other significant conditions or other organ involvement (renal, haematology, cardiac, GI, respiratory, diabetes mellitus)

Liver disease who meet one of more of the following criteria:

- Decompensated liver disease
- Receiving post-transplant immunosuppression or on Liver/small bowel/multivisceral transplant waiting list
- Liver disease and other significant conditions or other organ involvement (renal, haematology, cardiac, GI, respiratory, diabetes mellitus)
- Active or frequently relapsing autoimmune liver disease where they are likely to need increase in treatment

More information is available from the [British Society for Paediatric Gastroenterology, Hepatology and Nutrition](http://www.rcpch.ac.uk).

**Respiratory**

Most of the children in the respiratory groups listed below are not CEV but special consideration should be given to those with a recent PICU or HDU admission.

- Children with significant impairment in ability to cough and to clear airway secretions due to disease severity. This will include those children with severe neurological diseases including severe cerebral palsy, neuromuscular disabilities, severe motor impairment and those with severe metabolic disease
- Children who otherwise require a cough assist device to help with clearance of airway secretions
- Children who are life-dependent on long term ventilation, both invasive (via tracheostomy) and non-invasive (CPAP and BiPAP)
- Children with severe lung disease requiring continuous or overnight supplementary home oxygen and/or intermittent non-invasive ventilation
- Children with:
  - Cystic fibrosis and Primary ciliary dyskinesia
  - Severe bronchiectasis
  - Severe restrictive lung disease such as interstitial lung disease or obliterative bronchiolitis
  - Severe asthma: children treated with biological agents or maintenance oral steroids. **Note** the large majority of children with the most severe asthma including those treated with biological agents and daily prednisolone will not be CEV
  - Repaired congenital thoracic abnormalities such as congenital diaphragmatic hernia / trachea-oesophageal fistula only if significant airway or lung problem.
Notes on other conditions

Diabetes
There is no evidence that children with diabetes are more likely to be infected with COVID-19 compared to children without diabetes. More information is available from the Association of Children’s Diabetes Clinicians.

Down Syndrome
There is evidence that some adults with Down Syndrome may be at risk of complications from COVID-19, this primarily appears to be age related. There is no evidence that children with Down Syndrome and without co-morbidities need to take more care than is currently advised for all. Some children with Down Syndrome will have co-morbidities from either Group A or Group B, and they and their families will need to have conversations with their clinicians to determine if they are clinically extremely vulnerable. More information is available from the Down’s Syndrome Association.

Endocrinology
Children and young people who have hormone problems and in particular who are taking steroids (hydrocortisone, prednisolone, dexamethasone) because their adrenal glands do not work properly (steroid replacement therapy) are at no more risk of catching COVID-19 than other children. More information is available from the British Society of Paediatric Endocrinology and Diabetes.

Inherited metabolic diseases (IMD)
Children with an IMD who as a consequence fulfil one of the criteria in Group A will be CEV. Children with an IMD who fulfil one of the criteria in Group B may be considered to be CEV depending on discussion with the multidisciplinary team and parental assessment of the individual circumstances. Children with an IMD who do not fulfil Group A or B criteria should follow the advice given to the general population.

Rheumatology / paediatric ophthalmology
There is no evidence that children and young people with rheumatological or inflammatory ophthalmic conditions are more likely to be infected with COVID-19 than those without. If children and young people with rheumatological or inflammatory ophthalmic conditions do become infected with COVID-19 there is no evidence that they will become more unwell compared with children and young people without these conditions. This advice includes those on immunosuppressive medications. Our advice is paediatric rheumatology and paediatric ophthalmology patients should attend school in accordance with government advice.

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1 Clift AK, Coupland CAC, Keogh RH et al. Living risk prediction algorithm (QCOVID) for risk of hospital admission and mortality from coronavirus 19 in adults: national derivation and validation cohort study. BMJ Oct 2020 doi.org/10.1136/bmj.m3731