British Society of Haematology
Guidance on shielding for Children and Adults with splenectomy or splenic dysfunction during the COVID-19 pandemic

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6th May 2020

This document aims to provide a clear and consistent summary of the recommendations as to which patients with splenectomy or functional asplenia should be classed as being at high risk of developing Covid-19-related complications and, as a consequence, be placed on the Shielded Patient List (SPL).

All other asplenic patients may be considered to be lower risk ('clinically vulnerable' instead of 'clinically extremely vulnerable') but should follow national guidance concerning social distancing measures.

Background

Individuals with an absent or dysfunctional spleen are at increased risk of overwhelming bacterial sepsis with encapsulated pathogens. The commonest pathogen is *Streptococcus pneumoniae*, but other organisms also present significant risks, including Haemoph*ilus influenzae type b (Hib)* and *Neisseria meningitidis*.

Asplenia or hyposplenism may result from congenital asplenia, surgical removal of the spleen (for any reason), from therapeutic splenic embolization , splenic irradiation and as a complication of certain medical conditions or their treatment.

Guidelines recommend that all asplenic patients , irrespective of underlying cause, should have been immunised with certain specific vaccines – pneumococcal polysaccharide and conjugate, Hib conjugate and meningococcal – and be on long-term prophylactic anti-microbial therapy as appropriate (Davies et al 2011, Rubin and Schaffner 2014).

Asplenic/hyposplenic patients are also advised to receive influenza immunisation due to the increased risk of secondary bacterial infection after contracting influenza. Patients normally invited for annual flu immunisation are currently defined as "vulnerable", being at moderate risk of Covid-19 complications, and advised to maintain strict social distancing. However, the need for influenza immunisation is not, in itself, an indication of a higher risk of Covid-19 and therefore, not an indication for shielding in the asplenic/ hyposplenic patient.

Based on knowledge of the immunological functions of the spleen, there is no evidence that the lack of a spleen or part of a spleen or a non -functioning spleen on its own renders patients at higher risk of Covid-19. Recommendations for shielding will therefore depend in the underlying cause for splenectomy or asplenia and any associated comorbidities and treatments. Further guidance is available from the British Society for Haematology in relation to the management of different underlying conditions which may be associated with splenectomy or functional asplenia (see below).

However, since fever could indicate bacterial as well as viral infection, all patients should be instructed to seek medical advice by contacting NHS 111 and/or their clinical team if they develop a new fever, stating that have an absent or non -functioning spleen. Consideration should be given to the presence of bacterial infection, particularly with capsulated pathogens.

Recommendations applying to all patients

- Patients should ensure they are up to date with their vaccinations
- Patients taking regular prophylactic antibiotics should be encouraged to continue
- Those who are not taking antibiotics should have a supply at home to take if unwell and instructed to do so by a clinician
- All patients reporting a new fever should be evaluated for bacterial as well as viral infection

Patients who should be placed on the Shielded Patient List

Children or adults who are already on the SPL due to their underlying diagnosis and treatment; some will have had splenectomy or functional hyposplenism. See also Shielded Patients List at https://digital.nhs.uk/coronavirus/shielded-patient-list.

Haemato-oncology

- Those on anticancer treatments (chemotherapy, immunotherapy including CAR-T, cell therapy, antibody treatments or targeted treatments)
- Those with blood cancer (leukaemia, lymphoma) at any stage of treatment
- Those within 12 months of having an autologous stem cell transplant
- Those within 24 months of a donor stem cell transplant

General Haematology

Immune Thrombocytopenic Purpura (ITP), Autoimmune Haemolytic Anaemia (AIHA) and other autoimmune disorders post splenectomy with ongoing immunosuppressive treatment (see BSH ITP and Covid-19 guidance):

The British Society of Rheumatology (BSR) have stratified patients on immunosuppression into risk categories to identify those at highest risk requiring shielding (https://www.rheumatology.org.uk/News-Policy/Details/Covid19-Coronavirus-update-members). Those taking the following immunosuppression are advised to shield irrespective of splenectomy status:

- Corticosteroids ≥20mg (0,5mg/kg) prednisolone, or equivalent, per day for > 4 weeks
- Corticosteroid dose of ≥5mg prednisolone (or equivalent) per day for >4 weeks plus at least one other immunosuppressive medication (e.g. azathioprine, mycophenolate, ciclosporin) or rituximab within the last 12 months.
- A combination of 2 immunosuppressive medications including rituximab within the last 12 months plus an additional co-morbidity (age >70, Diabetes Mellitus, any pre-existing lung disease, renal impairment, any history of Ischaemic Heart Disease or hypertension).

Haemoglobinopathy/inherited red cell disorders

Sickle Cell Disease (SCD), of all genotypes, is associated with hyposplenism. Guidance from the Haemoglobinopathy Coordinating Centres (HCC) recommends which patients with SCD and Thalassaemia are considered clinically extremely vulnerable and need to shield. The initial advice was that all patients with SCD and high risk patients with Thalassaemia and other inherited red cell anaemias should shield. This latter group included patients with Thalassaemia or other inherited red cell disorders with splenectomy plus significant iron overload and/or comorbidities e.g. diabetes, cardiac disease This advice is being updated regularly based on accumulated data of outcomes in SCD and Thalassaemia with Covid-19.
(https://b-s-h.org.uk/media/18244/hbp-hccs-response-to-covid-v9-200420.pdf).
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Based on this data and following the initial period of shielding, due to end in June 2020, it is proposed that some of the patients will be classified as 'clinically vulnerable instead of 'clinically extremely vulnerable' and will no longer be recommended to continue shielding. after this time.

This list is not exhaustive, and some patients will need consideration by their treating clinician in light of their individual circumstances.

Patients who do not require shielding (providing no other indication for shielding)

All other patients. This will include:

- Splenectomy for trauma
- Thalassaemia or other inherited red cell disorders with splenectomy but without significant iron overload or comorbidities
- Splenectomy for autoimmune disorders but not currently taking immunosuppressive treatment and not on SPL due to underlying disease.

References

Further advice on the SPL is given here:
https://digital.nhs.uk/coronavirus/shielded-patient-list.

Further advice on how to shield is given here:
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

Further advice on social distancing is given here:
https://www.gov.uk/government/publications/full-guidance-on-staying-at-home-and-away-from-others

John M. Davies, Michael P. N. Lewis, Jennie Wimperis, Imran Rafi, Shamez Ladhani and Paula H. B. Bolton-Maggs; Review of guidelines for the prevention and treatment of infection in patients with an absent or dysfunctional spleen: Prepared on behalf of the British Committee for Standards in Haematology by a Working Party of the Haemato-Oncology Task Force *British Journal of Haematology, 155, 308–317*

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https://b-s-h.org.uk/about-us/news/covid-19-updates/

Haemoglobinopathy HCCs: Advice on COVID-19 in patients with Sickle Cell Disease and Thalassaemia Haemoglobinopathy Co-ordinating Centres V9 20 April 2020
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Rubin LG, Schaffner W. Care of the asplenic patient. New Engl J Med 2014;371:349-56