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What are the Current Indications and Contraindications for a Splenectomy?

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ABSTRACT

Splenectomy can treat conditions characterized by hemolysis or thrombocytopenia caused by autoantibodies or splenic reticuloendothelial function, as well as disorders characterized by massive splenomegaly and hypersplenism with cytopenias, such as hereditary spherocytosis, transfusion-dependent thalassemia, immune thrombocytopenia, autoimmune hemolytic anemia, splenic marginal zone lymph Splenectomy is no longer used for Hodgkin disease staging and is typically avoided in autoimmune lymphoproliferative syndrome, hereditary stomatocytosis or xerocytosis, cold agglutinin disease, paroxysmal cold hemoglobinuria, Gaucher disease, and cirrhosis-related hypersplenism.

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INTRODUCTION

Certain disorders, such as inherited hemolytic anemias, autoimmune cytopenias, or symptomatic splenomegaly, may need therapeutic splenectomy. Splenectomy is performed less frequently to reach a diagnosis when there is unexplained splenomegaly or a splenic mass. To provide the greatest results and the least morbidity, various concerns must be addressed before, during, and after the surgery. [1]

INDICATIONS

When less invasive medical interventions have failed to minimize hemolysis and improve hemoglobin levels, splenectomy is an acknowledged therapy for numerous hereditary hemolytic anemias. When there is severe splenomegaly that interferes with normal functioning, it may also give symptomatic relief. [2]

Hereditary hemolytic anemias caused by variations in genes that encode components of the red blood cell (RBC) membrane and cytoskeleton include spherocytosis, elliptocytosis, pyropoikilocytosis, and Southeast Asian ovalocytosis. Splenectomy may be utilized in chosen persons (typically children) who are still transfusion reliant or extremely symptomatic beyond one year of age, while it is better to postpone the treatment until after the age of six years owing to the risk of sepsis. [3]

Thalassemias are hereditary hemolytic anemias of various severity caused by a reduction in alpha or beta globin production. Some people with severe disease (transfusion-dependent thalassemia, growth retardation, hypersplenism, splenic infarction) may require splenectomy, but severe complications are becoming less common when people are treated with regular transfusions, chelation therapy, and activin receptor ligand trap therapy. Gene therapy will also most likely affect the natural course of this illness, potentially reducing the necessity for splenectomy. [4]

Sickle cell disease is characterized by early splenic infarction and is seldom accompanied with splenomegaly or hypersplenism. However, certain individuals may develop life-threatening splenic sequestration as a result of significant red blood cell pooling in the spleen, leading in quickly increasing splenomegaly, anemia, and hypovolemia. This is a dangerous complication with a high death rate. There have been no randomized controlled trials in individuals with a history of splenic sequestration crisis comparing the benefits of splenectomy with continuous transfusion treatment versus surveillance to avoid recurrent episodes. After one or two occurrences of splenic sequestration, some specialists urge splenectomy. [5]

Pyruvate kinase deficiency is an uncommon congenital nonspherocytic hemolytic anemia characterized by decreased ATP generation in RBCs. Individuals with severe or transfusion-dependent anemia frequently undergo splenectomy. Surgery is postponed if feasible until beyond the age of 5 years to reduce the risk of sepsis. A registry

research in people with PK deficiency found that splenectomy increased hemoglobin by 1.6 g/dL, which is frequently less than what is reported in other hereditary hemolytic anemias treated with splenectomy.[6]

The spleen may be the major location of cell death owing to reticuloendothelial macrophages in autoimmune cytopenias, or it may be the source of the clone of cells expressing the autoantibodies responsible for targeting the cells for destruction. [7]

Splenectomy is often done to lower the risk of major bleeding in persons (usually adults) with immune thrombocytopenia (ITP) when early medications are ineffective in raising the platelet count to a safe level (eg, >30,000/microL). Benefits and dangers are discussed individually in comparison to other therapy choices.[8]

Splenectomy is occasionally employed in people with autoimmune hemolytic anemia (AIHA) when other treatments have failed to improve hemoglobin levels and reduce hemolysis. Benefits and dangers are discussed individually in comparison to other therapy choices. [9]

Splenectomy is only utilized in rare cases of autoimmune neutropenia. Felty syndrome (splenomegaly and autoimmune neutropenia in people with rheumatoid arthritis), for which splenectomy may be undertaken if there are recurring or serious infections and other treatments have failed. [10]

Splenic marginal zone lymphoma (SMZL) is a B-cell non-Hodgkin lymphoma that commonly manifests as splenomegaly and lymphocytosis in adults. Splenectomy may be done to treat symptoms caused by localized splenic involvement in certain people.[11]

An abscess in the spleen is an uncommon consequence of other infections that are often seeded hematogenously (eg, from bacterial endocarditis). Antibiotics and splenectomy are commonly used in treatment, while expertise with image-guided percutaneous drainage is growing.[12]

Bleeding gastric varices can aggravate splenic or portal vein thrombosis. If a person experiences frequent bleeding episodes, a splenectomy may be performed. [13]

Splenectomy (deliberate or unintentional) may be included in surgical excision or tumor debulking, particularly for the following cancer types:[14]

- Ovarian cancer
- Colon cancer involving the splenic flexure
- Gastric cancer with direct splenic extension
- Pancreatic cancer that requires distal (or total) pancreatectomy

CONTRAINDICATIONS

While there are no definite contraindications to splenectomy, it has been shown in some cases that procedure is either unsuccessful or causes considerable toxicity. Splenectomy is typically not recommended in the following cases:

Autoimmune lymphoproliferative syndrome (ALPS) is a hereditary immune dysregulation condition (lymphoproliferation, autoimmune cytopenias) that may not manifest until maturity. Splenectomy causes unacceptably high rates of sepsis and mortality owing to encapsulated organisms in these patients.[15]

Cold agglutinin illness is a kind of autoimmune hemolytic anemia in which autoantibodies bind to complement at cold temperatures. Splenectomy is unlikely to be beneficial since the liver, not the spleen, is the principal site of RBC phagocytosis. IgG autoantibodies bind at cold temperatures and fix complement, resulting in intravascular hemolysis in paroxysmal cold hemoglobinuria. Splenectomy is ineffective because intravascular hemolysis occurs regardless of splenic function.[16]

The most prevalent hereditary lysosomal storage disorder is Gaucher disease. Splenectomy is no longer essential in the majority of patients due to the development of enzyme replacement or substrate reduction therapy. Splenectomy hastens Gaucher cell accumulation at other places and can aggravate bone disease, liver illness, pulmonary hypertension, and bone marrow involvement.[17]

Hereditary stomatocytosis and hereditary xerocytosis are inherited RBC illnesses characterized by abnormal RBC membrane ion channels and hemolytic anemia caused by genetic variations. Splenectomy does not reduce hemolysis appreciably and is linked with a considerably increased risk of vascular consequences such as thromboembolic events, vaso-occlusive episodes, and/or pulmonary hypertension.[18]

Cirrhosis can produce hypersplenism and thrombocytopenia, however there are other treatment options, and splenectomy is associated with a high complication risk.[19]

CONCLUSION

The table lists the situations for which splenectomy may be acceptable therapy, those for which it is seldom used, and those for which it is often contraindicated. Close collaboration is required between the primary care physician, the consultant hematologist or oncologist, and the surgeon to decide if splenectomy is the best therapeutic choice for the individual patient and what preoperative, intraoperative, and postoperative factors apply.

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