

## REVIEW

# ROLE OF SPLENECTOMY IN CONGENITAL HEMOLYTIC ANEMIA AND THERE OF POSTOPERATIVE COMPLICATIONS

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### SUMMARY

In the hematological disorders, splenectomy has multiple indications: diagnostics, palliative or curative. In recent years an increasing number of indications of splenectomy in hematological disorders either benign or malignant. It is a consequence of expanding the list of indications for blood disorders and liberalization splenectomy for many diseases. Splenectomy increases the life span of red blood cells, decreased transfusion requirements, and is the first-line treatment in some hematologic diseases. In thalassemia, splenectomy is done after a proper assessment of the moment when, according to patient's age, who must overcome childhood to complete immune system in order to reduce infectious risk. However, splenectomized patients, regardless of age, are at risk of severe infection after splenectomy, which lessened enthusiasm for routine use of splenectomy in some hematologic diseases. Fulminant infection after splenectomy syndrome is caused mainly by *Streptococcus pneumoniae*, *Haemophilus influenzae* and *Neisseria meningitidis*, which can be prevented by vaccination and antibiotic prophylaxis preoperatively.

**Key words:** hematological diseases, splenectomy, infection after splenectomy, thalassemia

### RÉSUMÉ

*Le rôle de la splénectomie dans les anémies congénitales et dans les complications post-opératoires de celles-ci*

Dans le cadre des affections hématologiques, la splénectomie a des indications multiples: diagnostiques, palliatives ou curatives. Dernièrement on a enregistré une augmentation du nombre d'indications de splénectomie dans les maladies hématologiques, qu'elles soient bénignes ou malignes. C'est une conséquence de l'élargissement de la liste de troubles hématologiques et de la libéralisation des indications de splénectomie pour un grand nombre de maladies. La splénectomie conduit à la croissance de la durée de vie des érythrocytes et à la diminution du nécessaire transfusionnel, en représentant le traitement de première ligne dans certaines maladies hématologiques. Dans les thalassémies, on effectue la splénectomie après l'évaluation juste du moment où il convient de la faire, compte tenu de l'âge du patient, qui doit avoir dépassé l'âge de l'enfance pour que son système immunitaire soit complètement développé et afin de réduire ainsi le risque d'infection. Quand même, les patients ayant subi une splénectomie, sont, à tout âge, confrontés au risque d'infections sévères post-splénectomie, ce qui a fait diminuer l'enthousiasme pour l'utilisation de routine de la splénectomie dans certaines maladies hématologiques. Le syndrome d'infection fulminante post-splénectomie est provoqué, principalement, par *Streptococcus pneumoniae*, *Neisseria meningitidis* et *Haemophilus influenzae*, qu'on peut éviter par la prophylaxie antibiotique et par la vaccination pré-opératoire.

**Mots-clés:** maladies hématologiques, splénectomie, infection post-splénectomie, thalassémies

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## INTRODUCTION

**H**aematological diseases include a wide range of entities, sometimes diametrically opposed, such as anemia and polycythemia, hypercoagulant statuses or bleeding diatheses, haemopathies malignant syndrome of intravascular hemolysis may affect the cardiovascular system in various ways.

### *Short history*

First splenectomy for haematologic disease was successfully achieved in 1887 by Sir Spencer at the fourth attempt in England. Wells operated a patient aged 24 years for uterine fibroids but found a "stray spleen" that he removed. Later it was found that the patient was suffering from hereditary spherocytosis and Wells, unintentionally, made the first surgical treatment of autoimmune hemolytic anemia.

In 1910, Sutherland and Burghard published the first report referring to two cases of splenectomy in patients with hereditary hemolytic anemia. Both parents were cured. Their statements were: "We believe that the spleen is directly involved in the destruction of blood cells." Kaznelson, a medical student in Prague, proposed Schloffer professor of surgery, splenectomy for a patient aged 36 years, known with idiopathic thrombocytopenic purpura. The increase platelet counts to normal was reported four weeks later after surgical procedure, in 1916.

Splenectomy was performed frequently in the past to extend staged disease in patients with Hodgkin's disease, to improve disease in patients with hairy cell leukemia or proliferative diseases. Also, splenectomy is an effective treatment for some types of chronic lymphoproliferative, namely B-cell leukemia or hairy cell leukemia, but also lymphocytic leukemia pro, especially the splenic marginal zone lymphoma. In these cases, splenectomy is associated with a significant reduction of tumor load. In most cases, therapeutic excision of the spleen can correct cytopenia, particularly anemia and thrombocytopenia.[1, 2, 3]

### *Blood diseases*

#### *Hereditary spherocytosis*

Hereditary spherocytosis is the most common type of chronic hemolytic anemia through membrane defect common in Caucasians, with an incidence of 1: 2500. Jaundice, anemia, reticulocytosis, changing erythrocyte indices (MCHC increase, decrease VEM), identification spherocytes on the blood smear, hyperbilirubinemia and increased levels of LDH are part of the clinical picture – biological disease. Many complications can arise in evolution of hereditary spherocytosis, such as acute hemolytic crisis, crisis megaloblastic, aplastic crisis, cholelithiasis, post – splenectomy infections.[4]

The treatment of choice is splenectomy for hereditary spherocytosis, spherocytes massage in splenic cords, where lowering the pH and glucose levels, as local hypoxia, spherocytosis enhances and erythrocyte uptake in spleen.

The characteristic cellular defect of the disease is not corrected by splenectomy, but significantly reduces hemolysis. The risk associated with splenectomy is, however, increased incidence of severe infection after splenectomy, particularly those with *Streptococcus pneumoniae*. In children, surgery is postponed until 4-6 years if not given the worsening anemia and hemolysis. Recent data have shown that these infections are rare (<1% in 1657 splenectomy). Pneumococcal polysaccharide vaccine after splenectomy and anti-biotic prophylaxis is recommended for lowering the risk of developing severe pneumococcal infections. [5]

#### *Thalassemias*

Thalassemias are hereditary diseases characterized by mutations in the genes encoding the globin chains synthesis by decreasing or abolishing the synthesis of certain types of chains. Depending on the affected chain of  $\alpha$ -thalassemia discuss with the decrease / absence of chain  $\alpha$ ,  $\beta$ -thalassemia with the decrease/absence chain and  $\beta$ -thalassemia  $\delta\beta$  with the decrease or absence of both chains  $\beta$ , and  $\delta$ . In the past, splenectomy was performed frequently in patients with thalassemia due to hypersplenism. More recently, achieving more transfusions, it has diminished the need for splenectomy. It is necessary to evaluate the risks and benefits of splenectomy on postoperative infections on these patients because they are more severe than on other patients with haematologic disorders. [6, 7]

#### *Other congenital anemias*

Another blood disease that can perform a splenectomy is sickle cell anemia, characterized by structural abnormality in hemoglobin by replacing glutamic acid to valine in position 6 of the chain  $\beta$  hemoglobin giving birth to hemoglobin mutant HbS ( $\alpha_2\beta_2s$ ) that alters the deformability of red blood cell with the advent of eliptocyte. Splenectomy is performed for recurrent crises vasoocclusive in case of hypersplenism, splenic abscess and massive splenic infarction. Surgical removal of the spleen is an obvious cause of hypersplenism. Treatment of sickle cell disease is based on a program of repeated transfusions that can prevent infarction of the spleen (a process called autosplenectomy). Thus, the presence of a palpable spleen in a patient with sickle cell disease after age of 5 years, suggests the coexistence of hemoglobinopathies, for example, thalassemia or hemoglobin C. [8]

Hemolytic anemia in patients with high phosphatidylcholine, should not be practiced because splenectomy surgery, hemolysis anemia worsens and remains unchanged. Also, splenectomy is not indicated in patients with Gaucher's disease (caused by deficiency of an enzyme,  $\beta$ -glucocerebrosidase), but might be worth in those with pyruvate kinase deficiency where splenectomy is recommended if blood needs exceed 200 – 250 ml/kg/year to maintain the Hb 10 g/dl or if there are phenomena of hypersplenism.

Autoimmune hemolytic anemia in patients with splenectomy is indicated as second-line treatment failure occurs when the autoimmune haemolysis corticosteroid therapy or relapse after treatment with corticosteroids. [9, 10]

### Complications after splenectomy

Splenectomy may be followed by severe systemic infections, because such surgery eliminates splenic macrophages that filter and remove bacteria and other pathogens that arise in the blood. Post-splenectomy infection, fatal complication in this case as it is called, is not common, but causes high mortality. Most infections after splenectomy are caused, usually by bacteria *Streptococcus pneumoniae*, *Haemophilus influenzae* and *Neisseria meningitidis*, and over half of those contaminated die. Such infections can be caused by other pathogens, such as *Escherichia coli* and *Pseudomonas aeruginosa*, *Capnocytophaga canimorsus*, group B streptococci, *Enterococcus*, *Ehrlichia* and protozoa such as those of the species *Plasmodium* that cause malaria. Besides the increased susceptibility to bacterial infection, splenectomized patients are more susceptible to parasitic disease called babesiosis. Splenectomized patients should avoid regions parasite *Babesia* endemic.[11]

The first description of fatal infection after splenectomy was published in 1952 by King and Schumaker. The disease may begin as a minor influenza disease that evolves rapidly into a fulminant infection. It occurs most frequently in the first two years after splenectomy, but may occur after several decades. The true incidence is not known, but the estimated annual rate among patients who have had splenectomy, is 0.18 to 0.42% lethal risk is 5%. Mainly due to lack of systematic studies, there are only very few data on morbidity and mortality. Patients need to know both the nature of infection after splenectomy and the fact that it is compulsory to consult their doctor if they are sick and fever. [12]

Postoperative infections after splenectomy can have a fulminant course, often preceded by nonspecific symptomatic picture: fever, headaches, nausea, diarrhea, malaise. Most of the patients developed disseminated intravascular coagulation. Acidosis and kidney failure are often major complications and mortality is between 50% and 60%.

The infections after splenectomy can be attributed to the loss of two important functions of spleen: epitopes specific polysaccharide antibody production of the batteries

encapsulated; phagocytosis of opsonized particles in splenic red pulp cords.

After surgery, the patient is indicated to receive: antibiotics (amoxicillin administration/three or four generation cephalosporin, levofloxacin); analgesics. [13,14]

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