webinar

HEALTH PROFESSIONALS

Thursdays Webinars

Indication for splenectomy

Maria Domenica Cappellini
Professor of Medicine and Hematology
Fondazione IRCCS Policlinico Hospital
University of Milan

22 May 2025

ERN | EuroBloodNet

Conflicts of interest

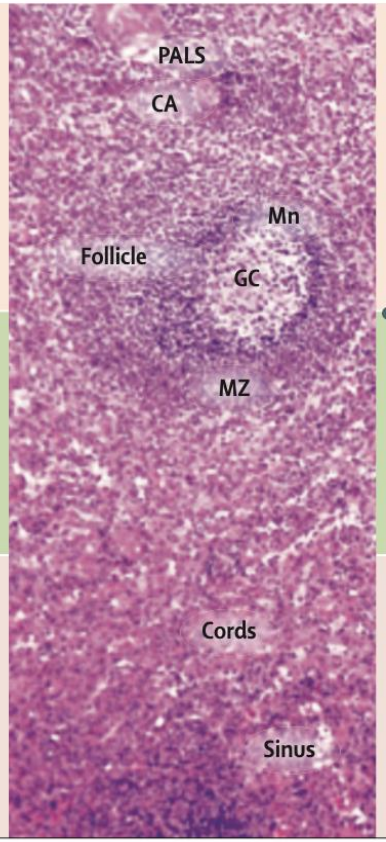
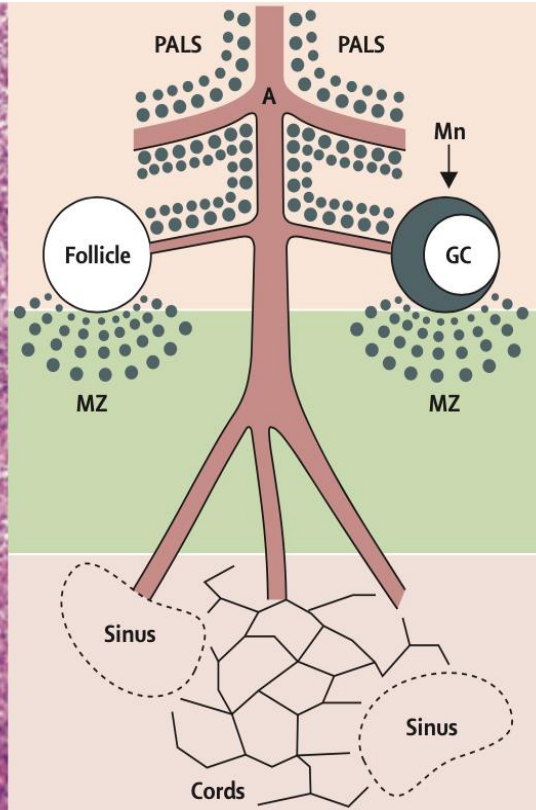
Advisory board member for:

- Sanofi Genzyme
- BMS/Celgene
- Vifor
- Vertex/CRISPR
- Novo Nordisk
- Pharmacosmos
- Novonordisk
- Pfizer

Learning objectives of the webinar

1. Splenomegaly and causes of Splenomegaly
2. Splenectomy indications
3. Consequences of splenectomy

Structure, function, and cell populations of the three functional compartments of the spleen

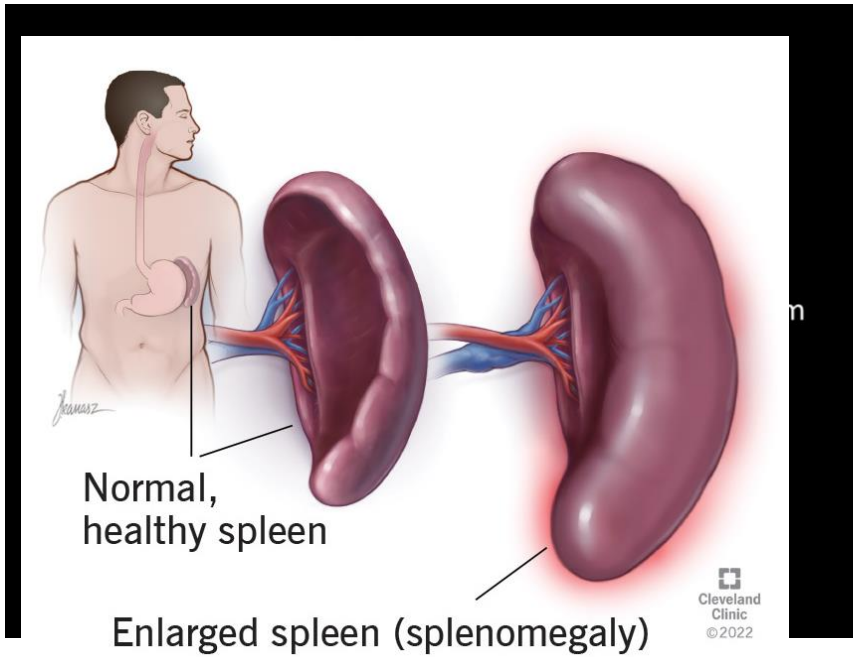
Compartment	Histology	Structure	Function	Cell
White pulp			Adaptive response (antigen specific) consequent to interaction between antigen-presenting cells (dendritic cells or marginal zone B lymphocytes) and B lymphocytes or T lymphocytes	**PALS (T-cell dependent)** Small $CD4^+$ T lymphocytes Dendritic cells B lymphocytes Macrophages Plasma cells **Follicle (B-cell dependent)** B lymphocytes or plasma cells Dendritic cells
Marginal zone			Innate response (first-line defence, non-antigen specific) characterised by IgM-memory B-lymphocyte production of natural antibodies	**Resident** B lymphocytes Macrophages **In transit** $CD4^+$ T lymphocytes $CD27^+$ memory B lymphocytes Dendritic cells
Red pulp			Innate response characterised by activation of macrophages in cords Adaptive response characterised by plasma-cell migration from the white pulp after antigen-specific differentiation in follicles Blood filter (pitting, culling)	**Cords of Billroth** $CD8^+$ T lymphocytes Fibroblasts Macrophages Natural killer cells **Sinusoids** $CD8^+$ endothelial cells

Di Sabatino A, Corazza G.Lancet 2011; 378: 86–97

Spleen

The spleen is a lymphoid organ that has a dual function, hemopoietic, especially during the in utero period, and immune.
Currently, hematologic disorders constitute the main indication for “elective” splenectomy.

Splenomegaly



- A normal spleen weighs 150 g and is approximately 11 cm in craniocaudal length
- The normal spleen is usually not palpable, although it can *sometimes be palpated in adolescents and individuals with a slender build*.
- Poulin et al defined **splenomegaly** as **moderate** if the largest dimension is **11-20 cm**, and **severe** if the largest dimension is **greater than 20 cm.**
- **Considerable variation in how massive splenomegaly is defined.**
- In recent publications describing techniques for laparascopic splenectomy, **massive** splenomegaly has been described as **≥17** cm craniocaudal length, **>20 cm**, or **splenic margin below the umbilicus or anteriorly extending over the midline**

Weinreb NJ & Rosenbloom BE 2013

Differential diagnosis of splenomegaly

The differential diagnosis of splenomegaly is extensive

Most often, the etiology is evident in light of historical and the concurrent presence of familiar, often pathognomonic, physical or laboratory findings (e.g. lymphadenopathy, stigmata of chronic cirrhosis or rheumatoid arthritis, abnormal blood morphology suggestive of hematological malignancies hemoglobinopathies or red cell cytoskeletal disorders)

.

Weinreb NJ & Rosenbloom BE 2013

Differential diagnosis of splenomegaly

Less commonly, there may be no relevant past or family history and accompanying findings may be non-specific (e.g. hematologic cytopenias without abnormal morphology), rare and unfamiliar (e.g. “gray” platelets), or simply not present. It is this scenario that so often leads to diagnostic error or delay in patients with splenomegaly that ultimately proves to be attributable to rare hereditary genetic diseases with which many physicians are unfamiliar.

Differential diagnosis of splenomegaly

Sustained diagnostic uncertainty is **particularly stressful** (for patient and physician) **when splenomegaly is "massive",** overtly symptomatic and sometimes accompanied by fear of an underlying malignancy.

In such circumstances, clinicians, who are sometimes *unaware* of available biochemical or *genetic testing* possibilities, may feel pressed to seek a **quick answer** through invasive **procedures such as bone marrow and liver biopsy or even total splenectomy that they may regard as not only diagnostic but also therapeutic.**

Weinreb NJ & Rosenbloom BE 2013

1

Step-wise approach

Lab investigation

Histology

A **partial** differential diagnosis of splenomegaly

- Hematological
- Portal hypertension
- Storage diseases
- Systemic diseases
- Infections
- Tropical splenomegaly syndrome
- Cryptogenic syndrome

History, physical exam

Splenomegaly

Surgery

Imaging

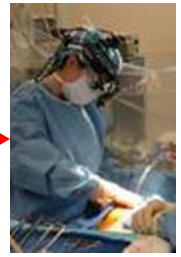
Treatment

Step-wise approach

2

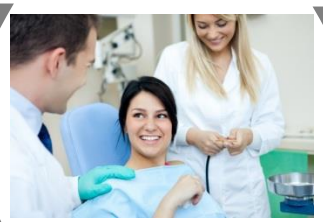
Imaging

Surgery

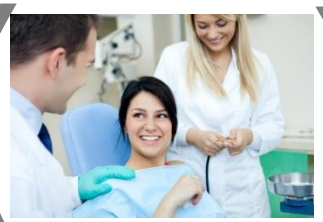


Histology

\+



\+



Benign and malign masses found incidently

- **Splenic cysts** (true*, false)
- **Benign tumors**

1. Hemangioma
2. Hamartoma
3. Litoral cell angioma
4. Lymphangioma
5. Inflammatory pseudotumor

- **Malign tumors**

1. Angiosarcoma
2. Metastases

Learning objectives of the webinar

1. Splenomegaly and causes of splenomegaly
2. Splenectomy indications
3. Consequences of splenectomy

Splenectomy

- With the progress in medical therapy, particularly with monoclonal antibodies, the indications and the results of splenectomy for hematologic disorders have changed enormously in recent years.
- However, certain benign and malignant diseases, including hematologic, red cell membrane, hemolytic, or platelet disorders, as well as lymphoproliferative or chronic myeloproliferative syndromes can still require splenectomy for management

Absolute indications for splenectomy

Table 1
Absolute indications for splenectomy.

Splenic trauma
Splenic rupture
•Spontaneous (tropical splenomegaly)
• Delayed rupture (subcapsular haematoma from trauma)
Splenic abscess (e.g. tuberculous infection)
Splenic cysts
Neoplasm
• As part of radical surgical oncological clearance of adjacent tumour. e.g. locally advanced gastric carcinoma, pancreatic carcinoma
• Angioma
• Primary (rare)
Aneurysm of splenic artery

E.P. Weledji / International Journal of Surgery 12 (2014) 113e119

Table 3
Relative indications and benefits of splenectomy.[11]

Condition	Degree of splenomegaly[a]	Indications for splenectomy[b]
1. Blood and reticuloendothelial disease		
a) Haemolytic		
i. Congenital haemolytic anaemia	+	+++
ii. Acquired haemolytic anaemia	++	++
iii. Thalassaemia	+	+
b) Haematological malignancy		
i. Acute leukaemia	+	0
ii. Chronic myeloid leukaemia	+++	+
iii. Chronic lymphatic leukaemia	++	+
iv. Lymphoma (Hodgkin's)	+	+
c) Myeloproliferative disorders		
i. Polycythaemia vera	++	0
ii. Myeloid metaplasia (myelofibrosis)	+++	+
d) Thrombocytopaenic disorders		
i. Acute ITP	+	+
ii. Chronic ITP	++	+++
2. Infective and inflammatory		
a) Parasites (hydatid)	++	+++
b) Protozoal (malaria)	+++	+
c) Inflammatory (Felty's syndrome)	++	+
3. Neoplastic		
a) Angioma	++	0
b) Cysts	++	+++
c) Metastases	+	0
4. Cryptogenic		
a) Tropical splenomegaly	+++	+
b) Non-tropical splenomegaly	+++	+++
5. Congestive		
a) Portal hypertension		
i. Intrahepatic	+	+
ii. Extrahepatic	++	+
6. Metabolic storage disorders		
a) Amyloidosis	++	0
b) Gaucher's disease	+++	+

[a] Degree of splenomegaly: +++ = marked (below umbilicus); ++ = moderate (4–8 cm below costal margin); + = slight (4 cm –just palpable).
[b] Indications for splenectomy: +++ = benefited by splenectomy; ++ = often benefited by splenectomy; + = splenectomy sometimes indicated.

E.P. Weledji / International Journal of Surgery 12 (2014) 113e119

Main hematological disorders potentially requiring splenectomy

Table 1 Main hematological disorders potentially requiring splenectomy.

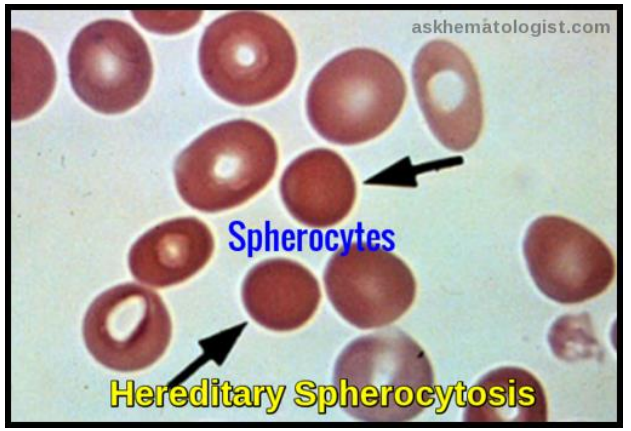
Hematological disorders		Proportion of indications for splenectomy[a]
Red cell membrane and hemolytic disorders	Constitutional hemolytic anemia Red cell membrane anomaly Hereditary Spherocytosis (HS) Familial Elliptocytosis (FE) Hereditary Pyropoikilocytosis (PH) Hemoglobinopathies Thalassemia Drepanocytosis Enzymatic deficits Pyruvate kinase Warm autoimmune hemolytic anemia (warm antibodies)	± 10% of indications for splenectomy
Platelet disorders	Immune Thrombocytopenic Purpura (ITP) Thrombotic Thrombocytopenic Purpura (TTP)	± 65% of indications for splenectomy
Lymphoproliferative syndromes	Splenectomy for diagnosis or for secondary complications of splenomegaly	1/4 of indications for splenectomy (23%)
Chronic myeloproliferative syndromes	For complications of splenomegaly (but high risk procedure in this context = thrombosis + +)	

[a] Data from the American College of Surgeons National Surgical Quality Improvement Program (over 7 years, from 2005 to 2011) [1]

Bonnet s. et al Journal of Visceral Surgery (2017) 154, 421—429

- Spherocytosis
- Thalassemia
- Sickle cell disease
- Platelets disorders

Spherocytosis



Indication of splenectomy in spherocytosis

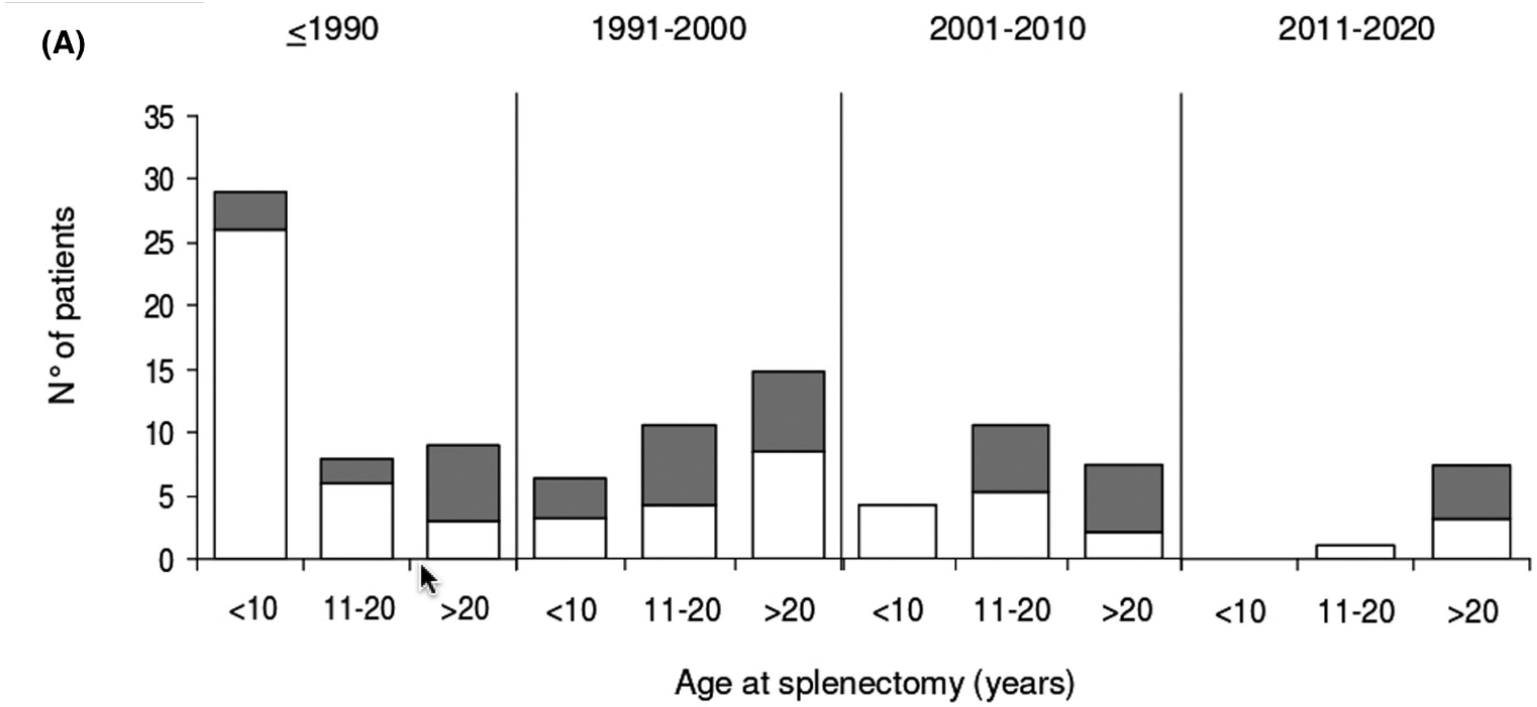
Table 2 Classification of spherocytosis and indications for splenectomy.

Criteria	Mild	Moderate	Severe
Hemoglobin (g/dL)	11 to 15	8 to 12	6 to 8
Reticulocytes (%)	3 to 6	> 6	> 10
Non-conjugated bilirubin (μmol/dL)	1.7 to 3.4	> 3.4	> 5.1
Splenectomy	Usually not necessary in infancy or in the adolescent	Necessary between 6 years and puberty	Necessary but after 6 years old if possible

Bonnet s. et al Journal of Visceral Surgery (2017) **154**, 421—429

Changing trends of splenectomy in hereditary spherocytosis: The experience of a reference Centre in the last 40 years

- Splenectomy:44% before 1990 to 7% in 2011– 2020)
- Age at splenectomy progressively increased (63%. in children before 1990 to 88% in patients aged ≥20 years in 2011– 2020



Vercellati et al. Br J Haematol. 2022;198:912–915.

Changing trends of splenectomy in hereditary spherocytosis: The experience of a reference Centre in the last 40 years

Conclusions:

- In any case, the effectiveness of splenectomy on anaemia in HS is clearly greater than that observed in other congenital haemolytic anaemias, such as PKD and dyserythropoietic forms.
- Therefore, a case- by- case evaluation is advisable in intermediate situations, considering lifestyle, patient's reported outcomes, quality of life, wish to become pregnant, concomitant cholecystectomy, and possible underlying thrombotic/infectious predisposition.

Vercellati et al. Br J Haematol. 2022;198:912–915.

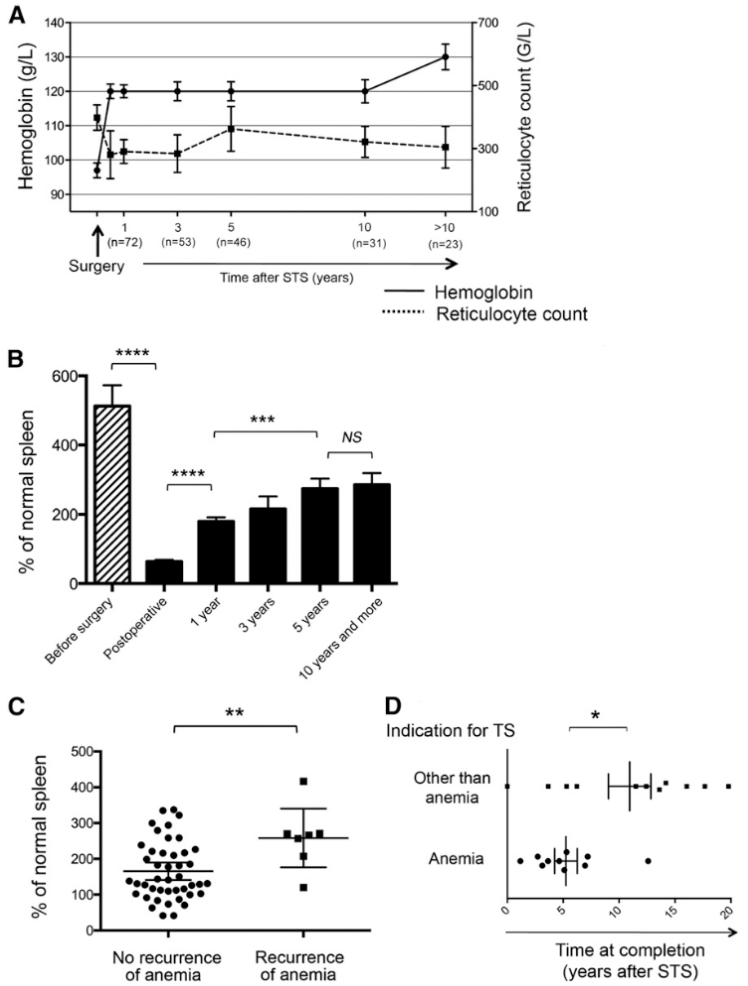
Table 3. Splenectomy guidelines in hereditary spherocytosis –2011 update[45] and the authors' recommendations.

N.	Guidelines 2011	Authors' recommendations
1	The laparoscopic approach to splenectomy is associated with less pain, shorter hospital stay and better cosmetic appearance; but is dependent on the availability of appropriately trained surgeons, and suitable equipment (grade 1 recommendation, grade B evidence).	No change.
2	In children undergoing splenectomy, the gall bladder should be removed concomitantly if there are symptomatic gallstones (grade C evidence, grade 2 recommendation).	No change.
3	In children who require cholecystectomy for symptoms of gallstones the use of concurrent splenectomy is controversial. It may be associated with a decreased risk of common bile duct stones in the future, but is also associated with a risk of post-splenectomy sepsis (grade 2 recommendation, grade C evidence).	In children > 6 years of age concomitant splenectomy is indicated according to severity of anemia (Table 1).
4	When splenectomy is indicated, ideally it should be done after the age of 6 years (grade 2 recommendation, grade C evidence).	No change.
5	Partial splenectomy is theoretically associated with a decreased risk of post-splenectomy sepsis, but it is possible that further surgery may be needed for either recurrence of hematologic problems or symptomatic cholelithiasis (grade 2 recommendation, grade C evidence).	No consensus among our experts.

* Reference 45 The Grading of Recommendation Assessment, Developing and Evaluation (GRADE) system was used to rate quality of evidence and strength of recommendations.

Iolascon a et al. Haematologica 2017;102,8

Subtotal Splenectomy in Sperocythosis



Pincez tBLOOD, 24 MARCH 2016 x VOLUME 127, NUMBER 12

Summary of splenectomy recommendatiopns for hemolytic disorders

Table 1. Summary of splenectomy recommendations for hemolytic disorders.

Disease	When splenectomy recommended? *
Hereditary spherocytosis	Patient is transfusion-dependent or suffers severe anemia. Patient has moderate disease: decision based on spleen size and quality of life parameters. No need to perform cholecystectomy.
Pyruvate kinase deficiency	Consider if patient is transfusion-dependent or severely anemic. Cholecystectomy should be performed at time of splenectomy.
Splenectomy in congenital non-spherocytic hemolytic anemia due to G6PD deficiency	Consider if patient is transfusion-dependent and/or has massive splenomegaly and/or has symptomatic splenomegaly.
Hereditary stomatocytosis	Contraindicated.
Congenital dyserythropoietic anemia type II	Consider if patient is transfusion-dependent and/or has symptomatic splenomegaly.
Sickle cell disease	Patient has had two acute splenic sequestration crises and/or has massive splenomegaly and/or suffers symptomatic hypersplenism.
Unstable hemoglobin	Consider only if patient has transfusion-dependent anemia and/or symptomatic splenomegaly.

*For all indications splenectomy should be performed after 6 years of age. G6PD: glucose-6-phosphate dehydrogenase.

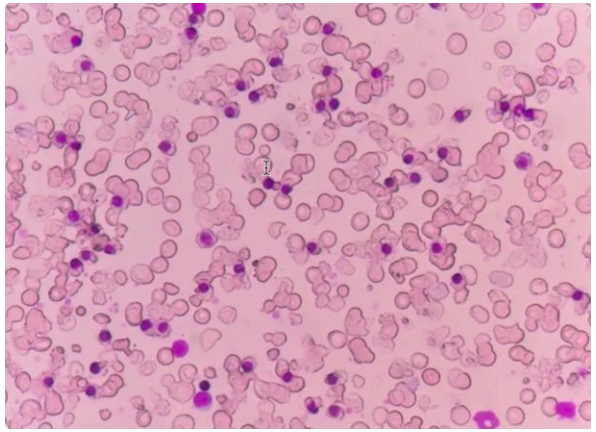
Iolascon a et al. Haematologica 2017;102,8

Table 4. Indications for splenectomy in pyruvate kinase deficiency based on severity of disease*.

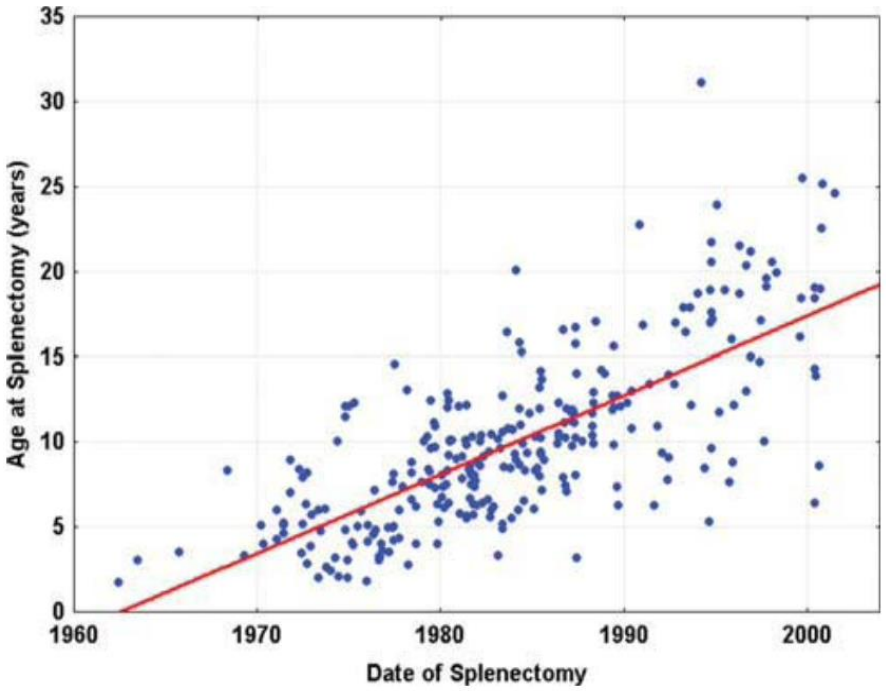
Disease severity	Age at diagnosis	Clinical manifestations	Median hemoglobin (g/dL)	Transfusion requirement	Indication for splenectomy
Severe	Birth/infancy	Most patients suffer severe neonatal jaundice requiring exchange transfusion, median age at diagnosis 4, almost all transfusion-dependent	6.8	Transfusion-dependent	Indicated after age of 6 years
Moderate	Variable, childhood to adult	Moderate anemia occasional exacerbations	9	Confined to exacerbations	Not indicated
Mild	Variable, childhood to adult	Lifelong history of mild anemia	11	Rare	Not indicated

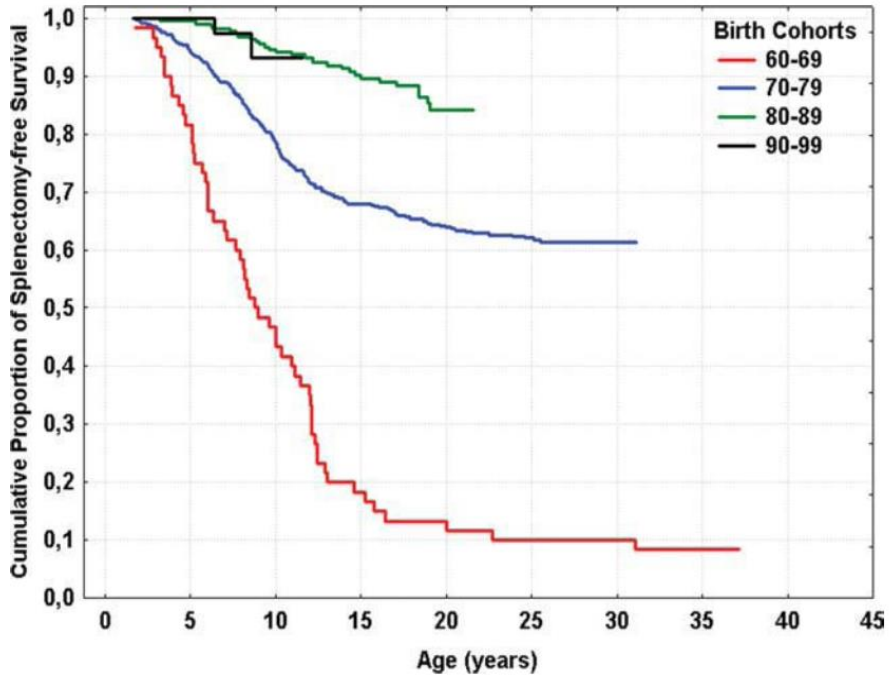
Iolascon a et al. Haematologica 2017;102,8

Thalassemia



Changing patterns of splenectomy in transfusion-dependent thalassemia patients

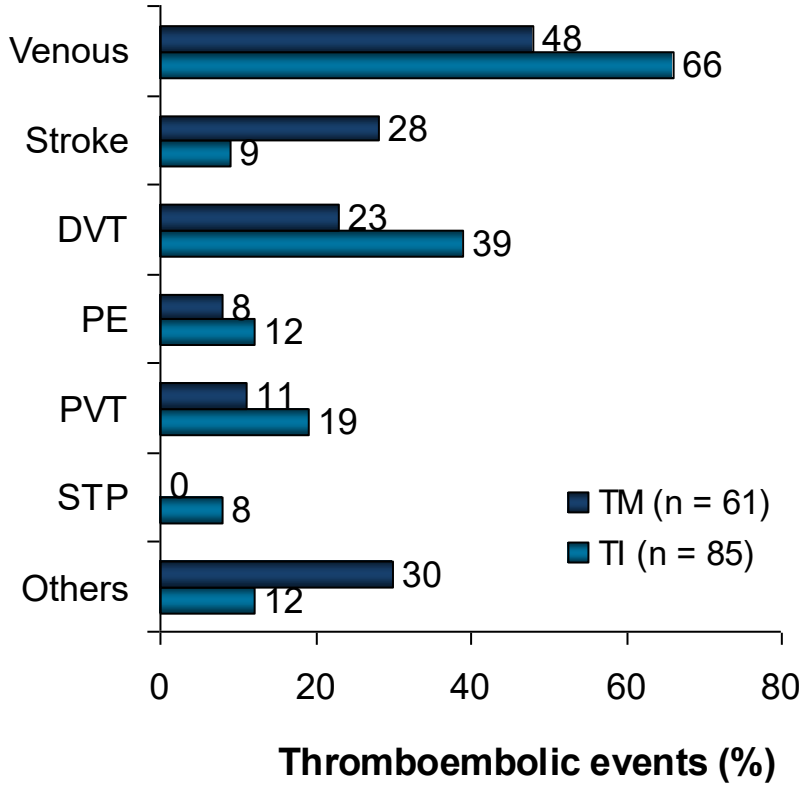




Piga A. et al Am J Hematol 2011

Thromboembolic events in a large cohort of β-TI patients

- Patients (N = 8,860)
  - 6,670 with β-TM
  - 2,190 with β-TI
- 146 (1.65%) thrombotic events
  - 61 (**0.9%**) with β-TM
  - 85 (**3.9%**) with β-TI
- Risk factors for developing thrombosis in β-TI were
  - age (> 20 years)
  - previous thromboembolic event
  - family history
  - **splenectomy**



DVT = deep vein thrombosis
PVT = portal vein thrombosis; STP = superficial thrombophlebitis

Taher A *et al. Thromb Haemost* 2006;96:488-491.

Patient stratification according to splenectomy and TEE status: OPTIMAL CARE

- Three groups of patients identified
  - **Group I**, splenectomized patients with a documented TEE (n = 73)
  - **Group II**, age- and sex-matched splenectomized patients without TEE (n = 73)
  - **Group III**, age- and sex-matched non-splenectomized patients without TEE (n = 73)

Type of thromboembolic event in splenectomized TI patients (Group I)	n (%)
DVT	46 (63.0)
PE*	13 (17.8)
STP	12 (16.4)
PVT	11 (15.1)
Stroke	4 (5.5)

*All patients who had PE had confirmed DVT.

TEE = thromboembolic events

Taher AT *et al. J Thromb Haemost* 2010;8:2152-2158.

OPTIMAL CARE study: multivariate analysis on risk factors for thrombosis in splenectomised patients

Parameter	Group	OR	95% CI	p value
NRBC count ≥ 300 x 10^6/L	Group III	1.00	Referent	
	Group II	5.35	2.31–12.35	< 0.001
	Group I	11.11	3.85–32.26	
Platelet count ≥ 500				< 0.001
PHT	Group III	1.00	Referent	
	Group II	4.00	0.99–16.13	0.020
	Group I	7.30	1.60–33.33	
Transfusion naivety	Group III	1.00	Referent	
	Group II	1.67	0.82–3.38	0.001
	Group I	3.64	1.82–7.30	

Group I patients had significantly higher NRBC, platelets, and PHT occurrence, and were mostly non-transfused

NRBC = nucleated red blood cell; PHT = pulmonary hypertension; OR = adjusted odds ratio; CI = confidence interval.

Taher AT *et al. J Thromb Haemost* 2010;8:2152-2158.

Silent cerebral infarction in splenectomized patients

Study	Number of patients	Age (years)	MRI technique	Definition of infarction	Prevalence of SCI (95% CI)
Manfre et al. 1999	16	Mean: 29 Range: 9-48	• 0.5 Tesla • T2- spin-echo weighted imaging	• Abnormally high signal intensity on long TR-weighted images • Two blinded neuroradiologists	**37.5% (18.4-61.7)**
Taher et al. 2010	30	Mean: 32.1 Range: 18-54	• 3.0 Tesla • T1-, T2-gradient-echo-, FLAIR-, diffusion-weighted imaging	• Abnormally high signal intensity on the T2- and FLAIR-weighted images[a] • Two blinded neuroradiologists	**60.0% (42.2-75.5)**
Karimi et al. 2010	30	Mean: 24.3 Range: 18-34	• 1.5 Tesla • T1-, T2-, FLAIR-weighted imaging	• Abnormally high signal intensity on the T2- and FLAIR- weighted images • One neuroradiologist	**26.7% (14.2-44.6)**
Teli et. al 2012	24	Mean: 12 Range: 4.5-20	N/A	N/A	**0%**

[a]Other diagnoses were ruled out based on the radiological appearance of lesions (e.g. viral encephalopathy or multiple sclerosis) or absence of associated risk factors and clinical symptoms (e.g. vasculitis or Binswanger's disease).
FLAIR, fluid attenuation inversion recovery; N/A, data not available; CI, confidence interval.

Musallam KM *et al. Thromb Res* 2012;130:695-702.

Patients susceptible to PHT may be prospectively identified using a variety of risk factors

Parameters	β-TI patients (N = 64)	
	PHT+*	PHT−
Splenectomized		
%	84.4	46.9
AOR (95% CI)	4.9 (1.9–8.5)	
History of thromboembolic events		
%	40.6	7.8
AOR (95% CI)	3.69 (2.38–7.05)	
Nucleated RBC count		
Mean (SD; x 10^6/L)	354.2 (199.2)	214.7 (94.5)
AOR (95% CI)	2.59 (1.69–6.05)	
Transfusion naivety		
%	56.2	78.1
AOR (95% CI)	3.5 (2.1–6.25)	
Iron chelation naivety		
%	37.5	62.5
AOR (95% CI)	2.3 (1.2–4.25)	
Hydroxyurea naivety		
%	17.2	34.4
AOR (95% CI)	2.6 (1.1–5.25)	

* $p < 0.001$ for all parameters for patients with PHT versus those without PHT
AOR = adjusted odds ratio

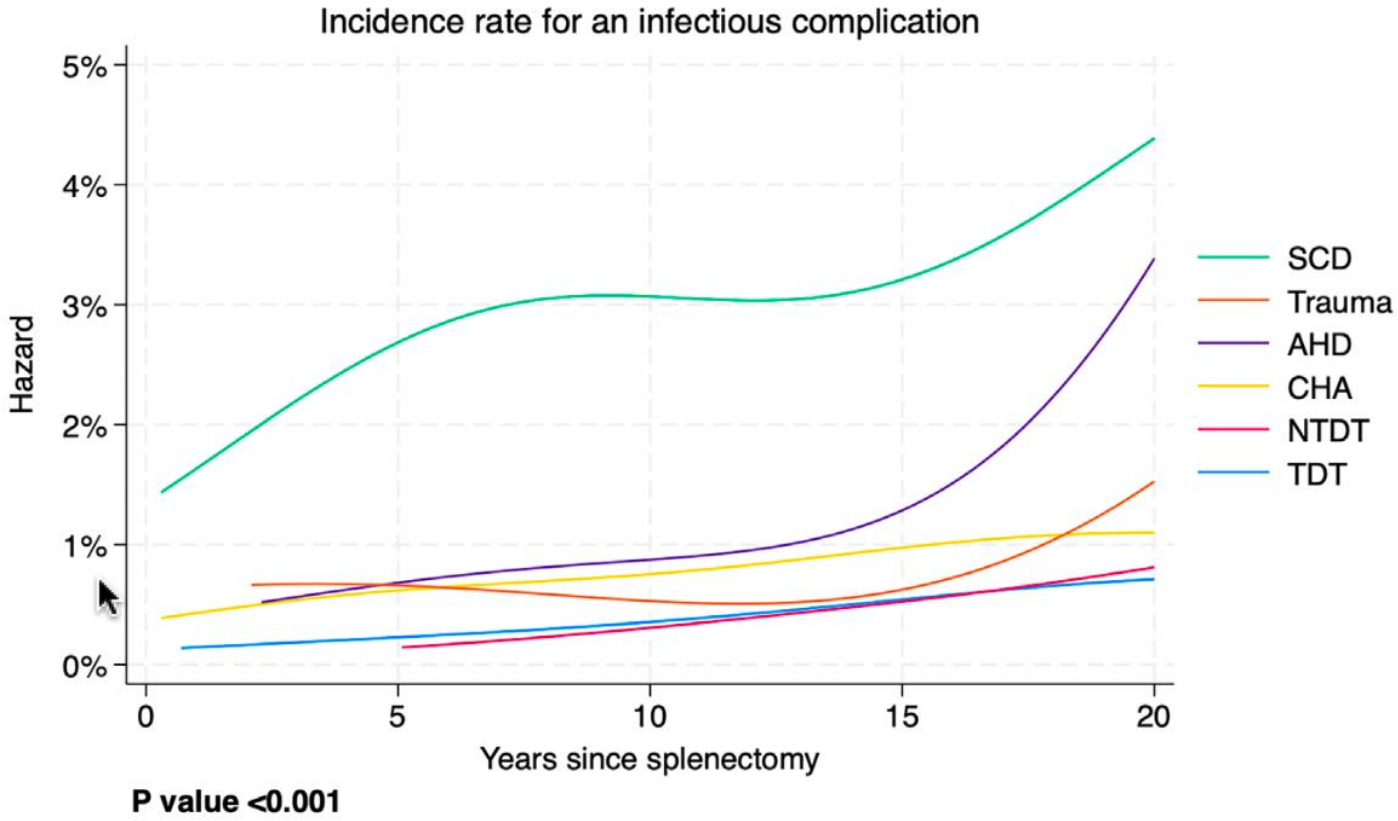
Karimi M *et al. Eur J Intern Med* 2011;22:607-610.

The Pros and Cons of Splenectomy in Transfusion Dependent Thalassemia Patient

- Fifty TDT patients were included from the data base
- Splenectomy had benefit in some TDT patients
- Factors that predict a higher response to splenectomy were thalassemia type (HbH and β/E), higher neutrophil percentage and older age at the time at splenectomy
- Hb and platelet was significantly increased after splenectomy.
- In term of complication, splenectomy group had significant higher TRV and tended to have more prevalence of PHT and thrombosis
- The longer time post splenectomy was the only significant factor for developing PHT.

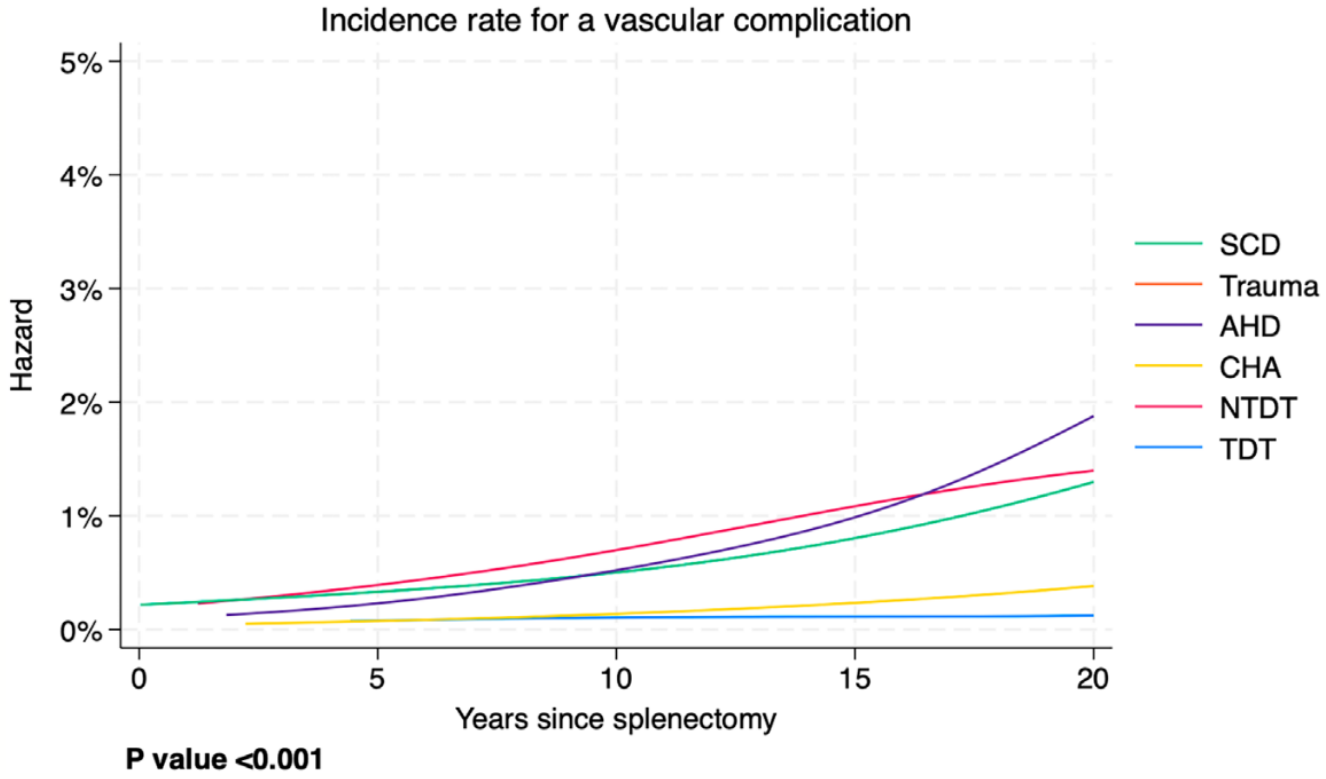
Tantiworawit A. et al. **132, Issue Supplement 1** November 29 2018

Underlying disease is the main risk factor in post- splenectomy complication risk: Data from a national database



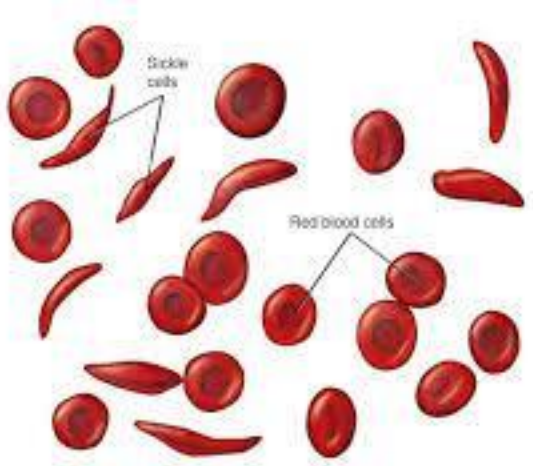
Casale M et al. Br J Haematol. 2025;00:1–11.

Underlying disease is the main risk factor in post- splenectomy complication risk: Data from a national database

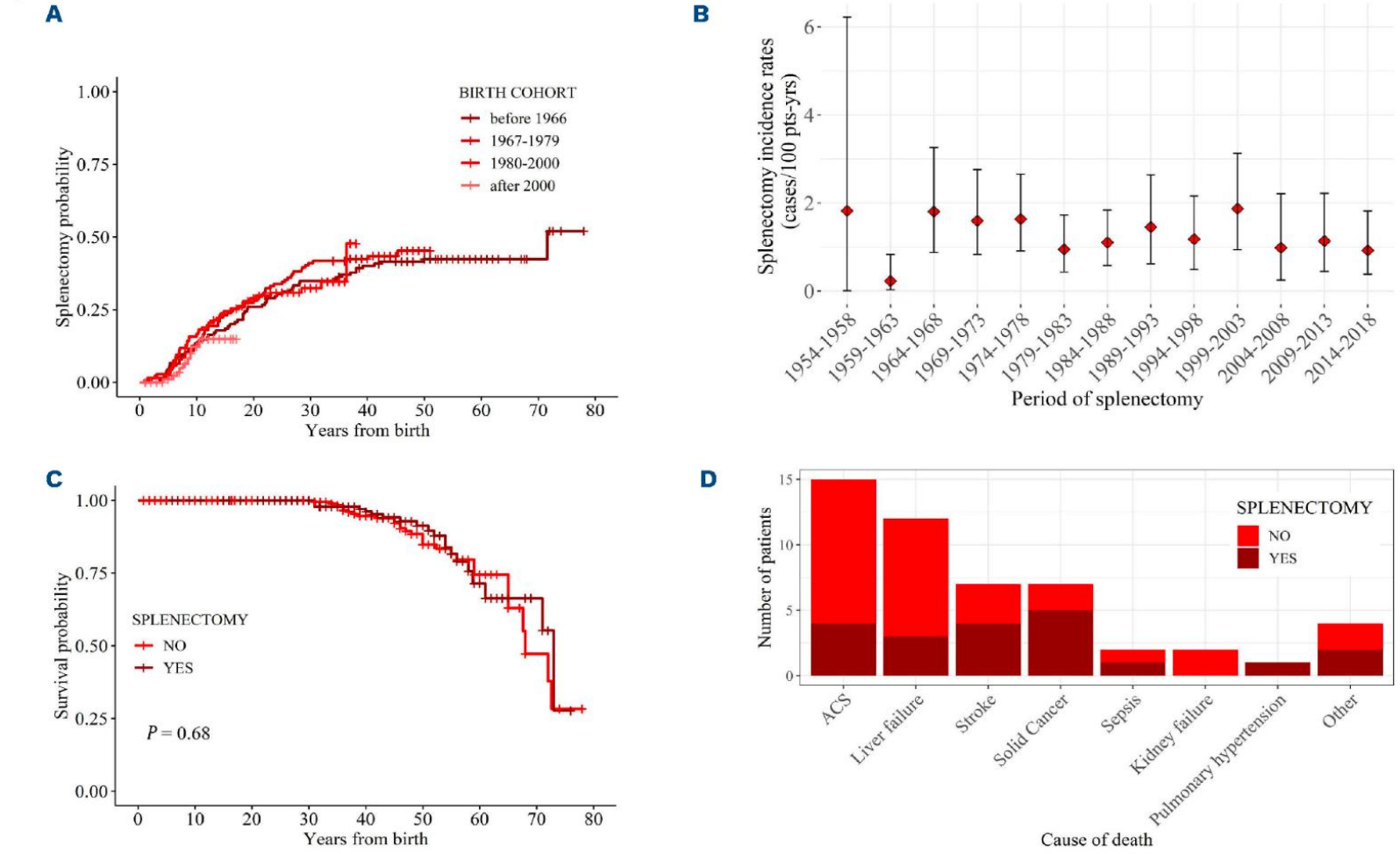


Casale M. et al. Br J Haematol. 2025;00:1–11.

Sickle cell diseases

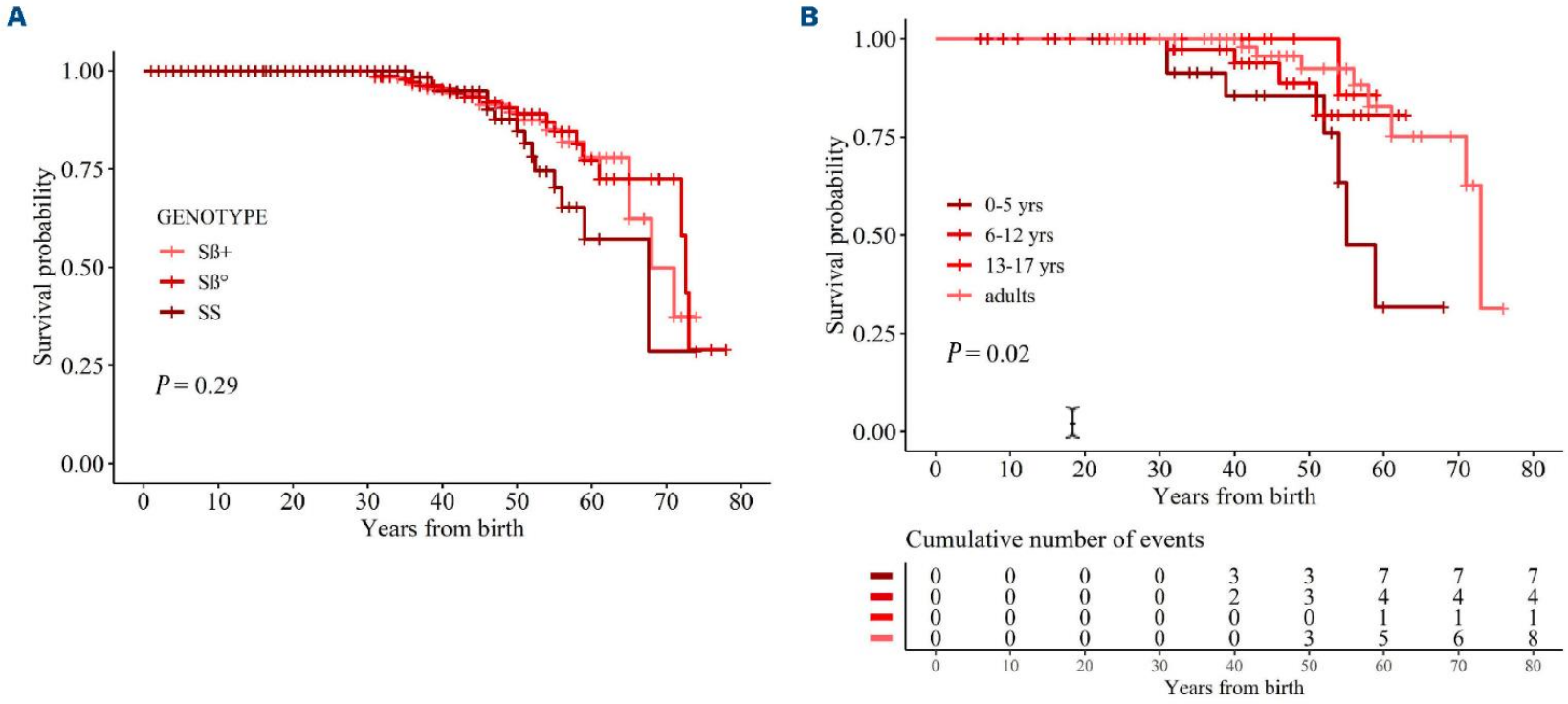


Splenectomy does not affect survival rate and the incidence of fatal infectious events in patients with sickle cell disease



Pinto V. et al. Haematologica 108: 4, 2023

Probability of survival of patients with sickle cell disease according to genotype and splenectomy

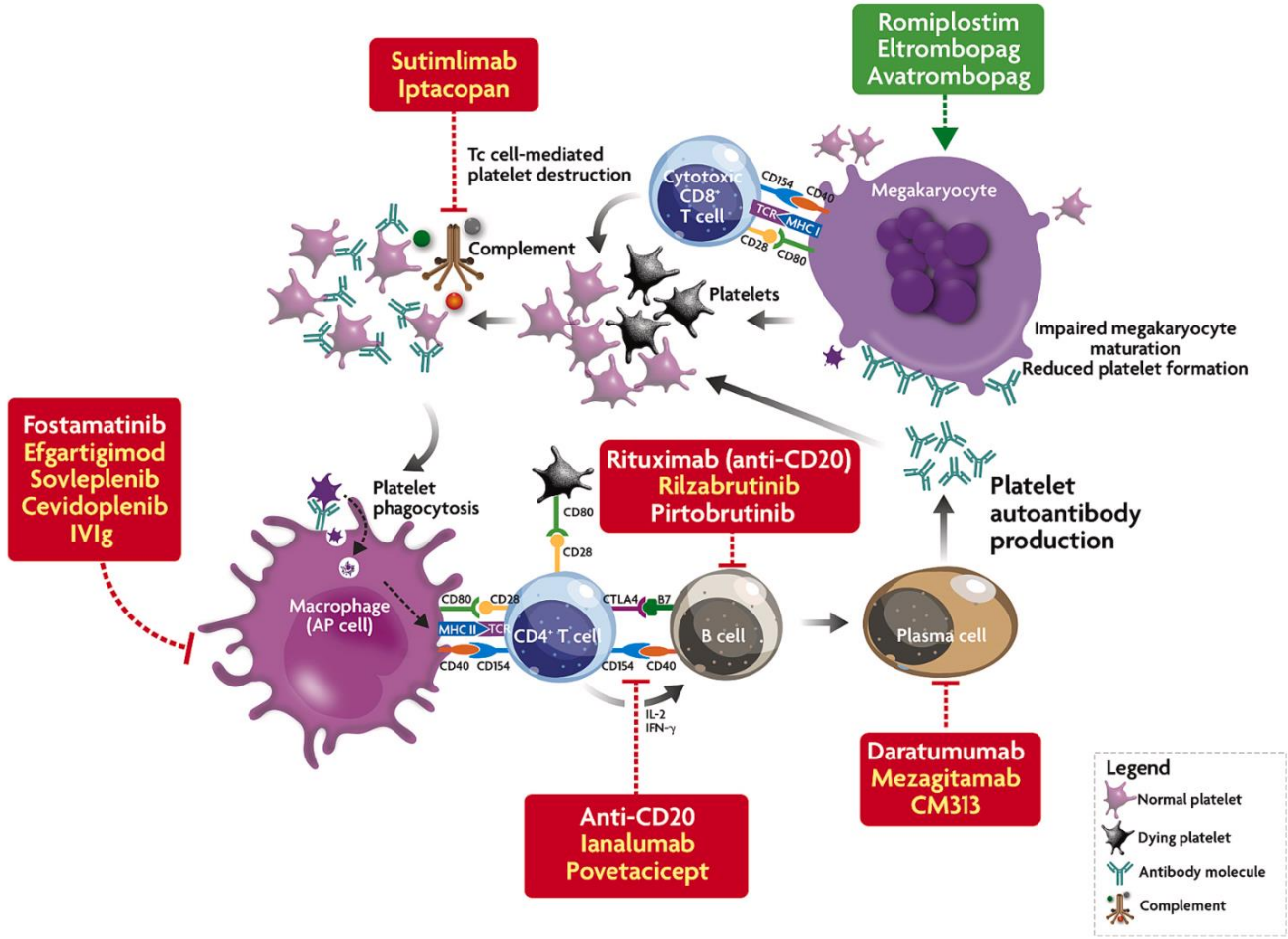


Pinto V. et al. Haematologica 108: 4, 2023

Platelets disorders

Immune Thrombocytopenic Purpura (ITP)

Thrombotic thrombocytopenic purpura (TTP)



Tomás José González-López, Drew Provan, Blood Reviews, https://doi.org/10.1016/j.blre.2025.101300

Contents lists available at ScienceDirect

Blood Reviews

journal homepage: www.elsevier.com/locate/issn/0268960X

Review

The new era of primary immune thrombocytopenia management in adults: A narrative review of current and emerging treatments

Tomás José González-López [a,*], Drew Provan [b]

[a] Hematology Department, Hospital Universitario de Burgos, Burgos, Spain
[b] Department of Haematology, Barts and The London School of Medicine, Queen Mary University of London, London, UK

Since the introduction of the TPO-RAs and other approved therapies for ITP splenectomy is carried out at a much lower rate than previously and is now a late treatment option.

Tomás José González-López, Drew Provan, Blood Reviews, https://doi.org/10.1016/j.blre.2025.101300

Splenectomy for ITP: down but not out

- The decision to proceed with splenectomy in a specific patient depends on the characteristics of their ITP as well as their age, general health, lifestyle, and goals
- Because there are few data comparing long-term outcomes of splenectomy with other second-line approaches, it is difficult to provide evidence-based recommendations, and patient preferences weigh heavily in treatment decisions.
- If possible, we defer splenectomy for 12 months after ITP onset to allow for a possible spontaneous or therapy-induced remission to occur
- Before splenectomy or even second-line medical therapies, it is important to rule out underlying conditions that may contribute to persistent or recurrent thrombocytopenia.

Chaturvedi S et al.,Blood. 2018;131(11):1172-1182)

Splenectomy for ITP: down but not out

There are several situations in which we would strongly consider splenectomy:

- Occasionally, patients present with severe ITP characterized by profound thrombocytopenia, bleeding, a poor or transient response to corticosteroids and IVIg, and a suboptimal response to TPO-RA. In such cases splenectomy provides an expeditious and generally effective approach. Even if splenectomy does not result in a complete remission, the response to subsequent interventions with previously used agents may improve
- Young patients with an active lifestyle, including those who participate in contact sports or high-risk activities, may prefer splenectomy. For these patients, the long- term risk of infection after splenectomy may be minimized through vaccination, patient education, and rapid institution of antibiotic therapy at the earliest indication of infection. They should also be counseled on the risk of thrombosis.
- Other patients who may benefit from splenectomy include those who are noncompliant with medications or for whom medical alternatives are inaccessible

Chaturvedi S et al.,Blood. 2018;131(11):1172-1182)

Splenectomy for ITP: down but not out

Table 2. Splenectomy vs TPO-RA and rituximab in refractory/relapsed ITP

Therapy	Response rate and durability	Time to response	Adverse effects	Contraindications	Preferred in	Approximate cost
Splenectomy	Overall response rate >80%, 50%-75% at 5 y	Days	Surgical mortality (<0.2% with laparoscopic splenectomy), surgery-related complications (9.6%; bleeding, infection, thrombosis) Lifetime risk of overwhelming sepsis Possible vascular complications: VTE, ATE	Multiple comorbidities, poor surgical candidate Relative: advanced age (higher rate of complications, lower response rate at age >60-70) *Helicobacter pylori*, hepatitis C (treat underlying cause first)	Fulminant ITP refractory to corticosteroids/IVIg with poor response to TPO-RA, desire to avoid drug therapy or close medical monitoring, uncertain compliance with medical therapy, prohibitive cost of medical therapy	20 000 USD
TPO-RA (eltrombopag and romiplostim)	80% overall response rate, high rates of durable response on continued therapy	10-14 d	Headache, rebound thrombocytopenia, elevated liver enzymes (eltrombopag), bone marrow reticulin fibrosis, possible small increased risk of venous thrombosis	Pregnancy (category C) and lactation, MDS Caution in patients with liver disease and a history of thrombosis	Patient preference, patients not interested in or unable to undergo splenectomy	Annually ~108 000 USD*
Rituximab	60% overall response rate; 21%-26% of responders at 1 y have responses at 5 y	1-8 wk	Infusion-related adverse events (fever, chills, dyspnea, hypotension), neutropenia, hypogammaglobulinemia, reactivation of viral infections (hepatitis B), progressive multifocal leucoencephalopathy (rare)	Active hepatitis B infection, pregnancy (category C) and lactation	Patient preference, patients not interested in or unable to undergo splenectomy, patient seeks medical long-term remission	10 000-40 000 USD per 4-infusion course

MDS, myelodysplastic syndrome; USD, United States dollars.

*Cost is estimated based on average wholesale cost for the following doses: eltrombopag 50 mg daily and romiplostim 3 μg/kg per week for a 70-kg individual.

Chaturvedi S et al.,Blood. 2018;131(11):1172-1182)

Systematic Review and Meta-Analysis

Medicine®

OPEN

Comparison of clinical efficacy of laparoscopic splenectomy versus open splenectomy for idiopathic thrombocytopenic purpura

A meta-analysis

Quan-Li Zhu, MD*, Wei Wu, MD

- The therapeutic effect of LS was the same as that of OS in Overall response
- Complication: accessory spleen
- The operative time was longer,
- The Estimated blood loss was less,
- The postoperative length of stay was shorter.

Zhu and Wu Medicine (2021) 100:4

Splenectomy in TTP

- First-line treatment consists of therapeutic plasma exchange plus corticosteroids whose dose can be intensified in case of failure, which occurs in up to 40% of patients
- Second-line therapy utilizes rituximab with good response rates
- Splenectomy is proposed exceptionally when TTP is refractory to previous treatments, and usually returns platelet counts to normal
- A 70% 10-year recurrence-free survival rate has been reported after splenectomy for refractory TTP and a recent study comparing splenectomy to cyclophosphamide confirmed that outcomes were similar with 80% remission rates in this indication [1].

Bonnet s. et al Journal of Visceral Surgery (2017) **154**, 421—429

Table 3 Lymphoid proliferative diseases that can be revealed by ''splenic lymphoma''.

Lymphomas in which splenic involvement is usual if not typical
Splenic marginal zone lymphoma
Splenic diffuse red pulp small B-cell lymphoma
Hairy cell leukemia
T-cell prolymphocytic leukemia
T-cell large granular lymphocytic leukemia
Hepatosplenic T-cell lymphoma
Lymphomas for which splenic involvement can be predominant
Mantle cell lymphoma
Follicular lymphoma
Diffuse large B-cell lymphoma

Bonnet s. et al Journal of Visceral Surgery (2017) **154**, 421—429

Principal complications of splenectomy

Complications	Approximate percentage
Intraoperative	
Bleeding	5%
Pancreatic tail injury	2%
Early postoperative	
Bleeding	1%
Subphrenic abscess	1%
Pneumonia, atelectasis	2%
Portal vein thrombosis	2%
Pancreatic fistula/pancreatitis	0.5%
Surgical wound complications	1%
Pulmonary embolism/thrombophlebitis	1—2%
Late postoperative	
Infective complications	3—5%
Thromboembolic risk	Variable according to indication
Risk of cancer	Suggested

Bonnet s. et al Journal of Visceral Surgery (2017) **154**, 421—429

Table 5
Summary of British haematology guidelines on timing and type of vaccinations in elective and emergency splenectomy.[29]

Class	OPSI prophylaxis	Timing
• <2yrs	• None (immature immune system)	
• >2yrs • Functional hyposplenism (e.g. sickle cell, ITP, coeliac dis)	• Immunization	• At least 2 weeks before splenectomy for optimal antibody response
• Emergency splenectomy	• Immunization	•Following emergency splenectomy • Immunization effect not as good • Still better than not being given
• Asplenic patients	• Immunization • Add influenza vaccine (prophylaxis against secondary bacterial infection)[9]	
• Splenectomy in underlying immunosuppressive disease (e.g. lymphoproliferative) or sickle-cell disorder[37]	• Immunization • Life-long prophylactic antibiotics	• Monitor response to pneumococcal vaccination • Timing of revaccination determine by levels of protective antibody
• Splenectomy in patients on immune suppressing therapies (chemotherapy and/or radiotherapy)	•Immunization • Life-long prophylactic antibiotics	• Immunization delayed at least 6 months following chemo/radiotherapy
• High risk patients (<18 yrs, immunosuppressed)	• Life-long oral prophylactic antibiotics against pneumococcal infection (penicillins/macrolides) • Prompt systemic antibiotic treatment for infection	•Regularly reviewed in light of local pneumococcal resistance patterns
• Low risk patients	• Counselled on risks and benefits of life-long antibiotics and may choose to discontinue • Carry a supply of appropriate antibiotics for emergency	
• Malaria belt	• Antimalarial prophylaxis • Treat malaria infection early and aggressively	

Key messages

- Splenectomy should be proposed routinely in patients with severe hereditary spherocytosis, and remains indicated in patients with moderate severity especially when gallstones are associated.
- Splenectomy can be indicated in sickle-cell anemia after a major crisis of acute splenic sequestration, or when hypersplenism is symptomatic.
- Splenectomy is indicated in warm-type autoimmune hemolytic anemia after failure of corticosteroid therapy (cortico-resistence or cortico-dependency), but is tending to be supplanted by rituximab.
- Splenectomy is indicated in ITP in patients who are young, sports-minded or whose work puts them at risk for trauma, or who are unwilling to continue long-term medical treatment. It should be delayed until after one year of evolution of disease (taking into account the possibility of spontaneous remission during the first year).
- Splenectomy is exceptionally indicated for lymphoproliferative disorders or chronic myeloproliferative syndromes.
- Multi-trocar laparoscopic splenectomy is considered the gold standard for surgical treatment of normal size or moderately enlarged spleens (diameter equal or less than 15 cm).

THANK YOU

webinar

www.eurobloodnet.eu

@ERNEuroBloodNet

eurobloodnet-european-reference-network-on-rare-hematological-diseases

Eurobloodnet - European Reference Network on Rare Hematological Diseases

ERN-EuroBloodNet's EDUcational Youtube channel

Funded by the European Union. Views and opinions expressed are however those of the author(s) only and do not necessarily reflect those of the European Union or European Health and Digital Executive Agency (HaDEA). Neither the European Union nor the granting authority can be held responsible for them.

Co-funded by the European Union