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ORIGINAL ARTICLE

Emergency or urgent splenectomy in children for non-traumatic reasons

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Abstract

Emergency splenectomy is rarely performed since a widespread consensus exists towards conservative management of splenic injury. However, in selected conditions, mainly hematological, there is a role for emergency or urgent splenectomy. This study aims to retrospectively review these cases and discuss outcome in relation to the pre-existing splenic pathologies. Between 2000 and 2015, 12 patients, five girls, and seven boys, with a median age of six years (3 months–13.11 years), underwent emergency or urgent splenectomy for non-traumatic conditions. All patients had major associated disorders; mainly hematological (11 cases) including hemolytic anemia with pancytopenia (1), sickle cell anemia (1), AML (1), ALL (2), CML (1), T cell lymphoma (1), Burkitt lymphoma (1), and ITP (3). One patient had a microvillous inclusion disease. Indications for splenectomy included diffuse resistant splenic abscesses (4), intracranial hemorrhage (4) or hypersplenism (3) with refractory thrombocytopenia, and spontaneous splenic rapture (1). Nine patients improved following surgery but three died, owing to massive intracranial hemorrhage (1) and severe respiratory failure (2) despite aggressive management.

Conclusions: Rarely, an emergency splenectomy is required in complex settings, mostly refractory hematological conditions, in a deteriorating patient when all other measurements have failed. A multidisciplinary team approach is mandatory in the treatment of these complex cases.

What is known
- *Conservative treatment is advised for splenic injury.*
- *Many hematological disorders are responsible of splenic pathology.*

What is new
- *Emergency splenectomy in children for reasons other than trauma is a treatment of last resort that should be performed in a multidisciplinary context.*
- *The outcome of emergency splenectomy in children for reasons other than trauma depends on the underlying medical condition.*

Keywords Emergency splenectomy · Non-traumatic · Children

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Abbreviations

ALL	Acute lymphoblastic leukemia
AML	Acute myeloid leukemia
CML	Chronic myeloid leukemia
CT scan	Computerized tomography scan
CVA	Cerebrovascular accident
ITP	Immune thrombocytopenic purpura

Introduction

In 1952 when King and Shumaker reported fatal sepsis in five infants who underwent splenectomy for hereditary spherocytosis, they changed the concept of the spleen as an expendable organ [6]. Patients without a spleen are known to have a fivefold increased risk for fatal sepsis [7, 9]. As trauma remains the most common cause of splenic rupture requiring splenectomy, pediatric surgeons were the first to apply the concept of non-operative treatment for children with blunt spleen injury and becoming gold standard during the 1980s [11]. The rare spontaneous non-traumatic splenic rupture is also life-threatening if unrecognized and untreated. This entity usually occurs with a pre-existent splenic pathology mostly hematological diseases as well as infection, malignancy, metabolic, and vascular disorders.

Rarely, these pre-existing splenic pathologies require low threshold for an aggressive approach and emergency or urgent splenectomy in attempt to prevent further clinical deterioration. We herein present our experience with 12 pediatric cases that underwent splenectomy for non-traumatic conditions and discuss clinical features, imaging studies, and outcome/effect of surgical approach in these highly complex patients. We hope our series will emphasize the need for multidisciplinary management of these difficult cases and help others in their management by raising awareness.

Material and methods

From January 2000 to October 2015, 117 children underwent splenectomy, 105 total and 12 partial. The medical records of the patients with non-traumatic indications for emergency or urgent splenectomy were reviewed retrospectively. They had no history of minor trauma before admission. They all received preoperative and 24 h postoperatively antibiotics. In addition, after splenectomy, all children received pneumococcal, haemophilus influenza, and meningococcal vaccines.

The splenectomy was considered emergency because if not performed, there would have been colossal risk to patient's life and urgent when further deterioration of the child's health would have been expected without surgery.

Patient data were evaluated for their characteristics, anamnesis, symptoms, method of diagnosis, etiology, surgical indication for emergency or urgent splenectomy, findings at surgery, pathology, and outcome.

Results

During a 15-year period, 117 splenectomies were performed, 93 electively, 24 as emergency or urgent procedures and amongst them 12 for non-traumatic reasons related to underlying systemic disorders in 11 and to spontaneous splenic rupture in 1.

Our series is composed of these 12 cases: five girls and seven boys, with a median age of six years (3 months–13.11 years). All patients had major associated disorders; mainly hematological (11 cases) including hemolytic anemia with pancytopenia (1), sickle cell anemia (1), AML (1), ALL (2), CML (1), T cell lymphoma (1), Burkitt lymphoma (1), and ITP (3). One patient had a microvillous inclusion disease.

Indications for surgery are listed in Table 1.

All four patients with splenic abscesses (diffuse resistant splenic mycosis after chemotherapy in two, splenic abscess during acute attack of sickle cell anemia in one, splenic abscess in child with AML in remission after chemotherapy in one) improved after the urgent surgery and adapted antimycotic and/or antibiotic treatment. The patients treated after 2006 received fluconazole or itraconazole.

All four patients who displayed intracerebral hemorrhage had an underlying hematological disease, three with resistant ITP and one with CML. No thrombopoietin receptor agonist treatment was offered because it was not yet available in our country for the corresponding patients.

1. A 3-year-old boy suffering from juvenile CML (diagnosed eight months prior) underwent chemotherapy and twice bone marrow transplantation and developed refractory thrombocytopenia. After presenting a hemiparesis, an intracerebral hemorrhage was seen on CT scan and he underwent a splenectomy 12 hours after appearance of the neurological symptoms. His platelet count went up progressively to 164000 but he died one month later from severe respiratory failure.
2. A 10-year-old boy suffering from newly diagnosed ITP, resistant to high-dose steroids and to intravenous immunoglobulins, presented with deteriorating consciousness. On the CT scan, an active left frontal hemorrhage was found. He then underwent an urgent craniotomy and splenectomy. In the postoperative period, his platelet count went up but because of an elevation in intra-cranial pressure, he underwent a second craniotomy with an anoxic brain death and died.
3. A 3-year-old boy with a history of ALL following chemotherapy, in remission, developed a resistant ITP with

Table 1 Indications for splenectomy

Indication for surgery	No	Underlying pathology	Outcome
Infectious–splenic abscess (diffuse splenic mycosis)	4	Burkitt lymphoma	Improvement
		Sickle cell anemia	Improvement
		AML – down syndrome	Improvement
		ALL	Improvement
Intracerebral hemorrhage	4	ITP 3 (2 with ALL)	1 death, 2 improved
		CML 1	1 death
Hypersplenism–resistant thrombocytopenia	3		
• With massive pulmonary bleeding	1	Fragile X syndrome	Improvement
• Increased abdominal pressure precluding ventilation	1	T cell lymphoma	Death
• Immunodeficiency	1	Hemolytic anemia	Improvement
Spontaneous splenic rupture	1	Microvillous inclusion disease	Improvement

an intracranial hemorrhage. He received rituximab treatment but still required a splenectomy without neurosurgical intervention. He fully recovered neurologically and hematologically.

4. A 3-year-old boy with also an underlying ALL presented with an exacerbation of his ITP, resistant to steroids, associated with a hypogammaglobulinemia. During that exacerbation, a CVA with a left parietal hemorrhage was diagnosed and an emergency splenectomy performed. He recovered from his thrombocytopenia and pursued his oncological treatment.

Three patients with hypersplenism and severe thrombocytopenia underwent emergency splenectomies.

1. A 21-month-old boy with an underlying T cell lymphoma presented a resistant thrombocytopenia and required very high ventilator pressures because of his enlarged spleen and thus underwent splenectomy. There was no improvement in the platelet count or on the ventilator pressures. On the 5th postoperative day, he underwent a salvage bone marrow transplantation that failed and the boy died.
2. A 3-month-old girl was born preterm at 32 weeks with a fragile X syndrome. At three weeks of age, she presented a septicemia with a cholestatic liver disease, a hepatosplenomegaly with pancytopenia. Because of a massive pulmonary bleeding due to her thrombocytopenia, she underwent splenectomy followed by a recovery (396000 platelet count) and discharge at 11 days postoperatively.
3. A 7-year-old girl with an underlying hemolytic anemia presented with hepatosplenomegaly, thrombocytopenia requiring repeated blood and platelets transfusions, and an undefined immunodeficiency. She underwent splenectomy and her platelet count went up to 107000. She was discharged on the 11th postoperative day.

One patient presented a spontaneous splenic rupture, a 4 and a half year-old girl with microvillous inclusion disease. She underwent an emergency splenectomy with an uneventful postoperative course and was discharged from hospital.

Discussion

Most elective splenectomies in children are done for hematological and immunological disorders [9]. Emergency splenectomy was commonly performed in patients with traumatic splenic injuries until the 80s when a conservative non-operative management for spleen preservation was introduced in children [2]. This approach emerged from the fatal consequences of overwhelming postsplenectomy infections in the asplenic patient that resulted with a high mortality rate. The majority of infections occur within two years and are fatal in almost 50% [7, 9].

The spleen is commonly involved in a wide spectrum of hematologic, immunologic, neoplastic, infectious, and vascular systemic disorders [13]. Elective splenectomy remains a key treatment in some hematological and immunological conditions such as sickle cell disease, resistant ITP, hereditary spherocytosis. In these cases, splenectomy is almost always performed electively, which enables adequate preparation. Excluding trauma-related cases (including those who did not respond to conservative treatment), splenectomy is rarely performed as an emergency or urgent procedure in children. Our series shows that there are cases when a decision to perform a splenectomy is taken emergently in a deteriorating patient.

In our series, splenectomy was an emergency procedure in cases of intracranial hemorrhage in the presence of severe refractory thrombocytopenia with platelet counts improving after surgery and in one case of spontaneous splenic rupture. Nowadays, a thrombopoietin receptor agonist treatment is first given, the indication for splenectomy taken only in case of persistent hemorrhage with persistent thrombocytopenia. It

was an urgent procedure performed in deteriorating complex patients with a multidisciplinary team decision-making (hematologist, intensive care, infectious disease pediatrician, surgeon). The indications were hypersplenism with resistant thrombocytopenia and high ventilatory pressures partially due to the enlarged spleen causing increased abdominal pressure, hypersplenism with resistant pancytopenia accompanied by massive pulmonary bleeding, and diffuse candidiasis contaminating the spleen.

All the patients with splenic abscesses improved after splenectomy because surgery removes the infective load and with the combination of systemic antibiotics and effective antifungals, it allowed an improvement and eventually recovery from the sepsis. Aggressive antifungals need to be given first and only in presence of insufficient or no recovery the need for splenectomy arises. Another two patients who improved following splenectomy had severe thrombocytopenia due to ITP and hypersplenism, which dramatically improved in the postoperative period.

Another described cause for emergency splenectomy consists of non-traumatic splenic rupture that occurs in less than 1% of patients with no trauma and no medical disease presenting signs of spleen rupture [8]. Despite its rarity, it is important to highlight awareness of this pathology that requires a high index of suspicion for diagnosis. The actual reason for the rupture is not fully understood but it rarely occurs in histologically proven normal spleen, the size of the spleen plays a role. In 2008, Gedik described three mechanisms implicated: (i) increased intrasplenic tension caused by cellular hyperplasia and engorgement, (ii) compression of the spleen by abdominal muscles during activities like coughing or defecation, (iii) vascular occlusion [4].

The spleen can be the site of severe infections and may undergo spontaneous bleeding or rupture without any trauma [10, 12, 13]. Splenic infarctions and infective endocarditis, splenic vascular diseases, acute splenic infections and abscesses, splenic hemorrhage, and rupture (due to pregnancy, peliosis, amyloidosis, coagulopathy such as ITP, hemodialysis, anticoagulant or thrombolytic therapy, colonoscopy, 7% idiopathic rupture in normal spleen) were found responsible.

Patients presenting with acute non-traumatic abdominal symptoms and/or hypotension - clues hinting to a spontaneous splenic injury are limited even for experienced surgeons. A CT scan gives quick identification of acute splenic abnormalities [13]. The overall mortality rate for spontaneous non-traumatic rupture is 10–15% [10]. In the current series, the patient survived.

Aggressive management with early surgical intervention and appropriate hemoderivative support is important. Non-operative management is risky because a diseased (like in infectious mononucleosis) spleen may not heal as readily as a normal, traumatized spleen [5]. Splenectomy remains the treatment of choice for patients with underlying malignancies. In hematological diseases, in cases of severe thrombocytopenia and splenic abscesses, the general condition of the patients is improved by splenectomy [3]. Splenectomy in situations of hypersplenism is always a risk with mortality up to 20% [3].

Some patients are specifically investigated for suspected splenic pathology but most of the time, splenic emergencies are detected in patients being investigated for non-specific abdominal pain. There is a need to be vigilant in identifying these non-traumatic splenic emergencies as they can have life-threatening consequences if not recognized and managed promptly [1]. The survival of patients following splenectomy is correlated with the course of the underlying disease [4].

We do not presume to determine indications for surgery in these cases considering our small cohort. However, in our experience, it is crucial to implement an on-going close multidisciplinary evaluation and follow-up to decide upon the need for emergency or urgent splenectomy and the appropriate timing for these patients. More recently, the arrival of thrombopoietin receptor agonists has reduced the need for splenectomy in patients with severe chronic ITP. Since these are rare, difficult, and complex cases, surgery is performed as the last resort hence the poor outcome and high mortality rate.

Authors' contributions Study conception and design: Dr. I. Samuk, Dr. A. Baazov, Pr. E. Freud
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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This article does not contain any study with human participants or animals performed by any of the authors.

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