## Splenectomy for Hematological Disorders in Iranian Pediatric Patients: A Single Center Study

Bibi Shahin Shamsian<sup>\*1</sup>, MD; Mohammad Thaghi Arzanian<sup>1</sup>, MD; Raheleh Kaviani<sup>1</sup>, MD; Samin Alavi<sup>1</sup>, MD; Mona Hedayat<sup>2,3</sup>, MD; Nima Rezaei<sup>2,3</sup>, MD, PhD

- 1. Department of Pediatric Hematology-Oncology, Mofid Children's Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran
- 2. Research Center for Immunodeficiencies, Pediatrics Center of Excellence, Children's Medical Center, Tehran, Iran
- 3. Molecular Immunology Research Center and Department of Immunology, Tehran University of Medical Sciences, Tehran, Iran

Received: Jan 26, 2011; Accepted: Nov 15, 2011

*Key words:* Splenectomy; Pneumococcal Vaccines; Sepsis

Splenectomy is a widely accepted therapeutic strategy in the management of a variety of hematological disorders<sup>[1]</sup>. Regardless of its therapeutic advantages, it may be followed by overwhelming post-splenectomy infection (OPSI), the most devastating complication in asplenic individuals, which is a fulminant and rapidly fatal condition mainly due to encapsulated organisms<sup>[2]</sup>. The true incidence of OPSI is not well known; however, age at time of splenectomy, the underlying diagnosis, and the time elapsed from splenectomy constitute the main determinants of fatal outcome <sup>[3,4]</sup>. Vaccinations against Streptococcus pneumoniae, Haemophilus influenzae type b, and Neisseria meningitidis, and long-term antibiotic prophylaxis, in conjunction with patient education, have been strongly advocated in splenectomized patients <sup>[2,5]</sup>.

The present study was conducted to address issues related to the enforcement of and compliance with current prevention guidelines among splenectomized patients in a tertiary care setting. In a retrospective cross-sectional study, we evaluated 53 patients aged 1.5 to 23 years (32 males and 21 females) undergoing splenectomy within a 5-year period at Mofid Children's Hospital, Shahid Beheshti University of Medical Sciences and Health Services, Tehran, Iran. The most common indications for splenectomy were thalassemia major (43.3%), hereditary spherocytosis (HS) (15.1%), and idiopathic thrombocytopenic purpura (ITP) (11.3%). Findings from other studies indicated that ITP<sup>[6,7]</sup>, sickle cell disease<sup>[8,9]</sup>, and HS<sup>[10]</sup> were the most common single hematologic disorders requiring elective splenectomy in the course of the disease. The noted discrepancy might be partially due to the fact that beta-thalassemia is particularly prevalent among the Mediterranean countries, justifying its high prevalence among Iranian patients undergoing splenectomy.

Studies conducting retrospective audits of vaccination status in splenectomized individuals reported 62-80.6% vaccination rate against Streptococcus pneumonia <sup>[11-13]</sup>, whereas in the present study, all 53 patients (100%) received pneumococcal vaccination perioperatively, with 41.5% and 35.8% of them being respectively vaccinated against Neisseria meningitidis and Haemophilus influenzae type b as well. On the whole, 58.5% received all three vaccines.

Of all studied patients, 38 (71.7%) were provided with follow-up care, of whom, 4 developed OPSI and died at the time of study. A review of available follow-up data revealed that 33 (out of 34) patients were receiving daily prophylactic oral penicillin, compared to 63-93 % of patients being prescribed long-term antibiotic prophylaxis in other studies <sup>[13-15]</sup>.

Besides appropriate and timely immunization with pneumococcal vaccine and a high compliance rate to daily antibiotic prophylaxis, adherence to the remaining key guidelines concerning proper immunization against Haemophilus influenzae and Neisseria meningitidis and, more important, patient education, i.e. providing patients with clear

<sup>\*</sup> **Corresponding Author; Address:** Department of Pediatric Hematology-Oncology, Mofid Children's Hospital, Shariati Avenue, Tehran, Iran

discharge instructions emphasizing on the importance of timely follow-up visits and recognizing the need for urgent referral for any suspected infection, is not fully satisfying. Many of these could be met affordably by developing a registry of splenectomized patients, which will be aiding in providing patients with continued follow-up cares.

## **References**

- 1. Torelli P, Qasaimeh GR, Bani-Hani KE, et al. Laparoscopic splenectomy for hematological diseases. *Surg Endosc* 2002;16(6):965-71.
- 2. Clinical Haematology Task Force. Working Party of the British Committee for Standards in Haematology. Guidelines for the prevention and treatment of infection in patients with an absent or dysfunctional spleen. *BMJ* 1996;312(7028): 430-4.
- Styrt B. Infection associated with asplenia: risks, mechanisms, and prevention. *Am J Med* 1990; 88(5N):33N-42N.
- Kyaw MH, Holmes EM, Toolis F, et al. Evaluation of severe infection and survival after splenectomy. *Am J Med* 2006;119(3):276 e1-7.
- Davies, JM, Barnes R, Milligan D. Update of guidelines for the prevention and treatment of infection in patients with an absent or dysfunctional spleen. *Clin Med* 2002;2(5):440-3.
- Katkhouda N, Hurwitz MB, Rivera RT, et al. Laparoscopic splenectomy: outcome and efficacy in 103 consecutive patients. *Ann Surg* 1998; 228(4):568-78.

- Delaitre B, Champault G, Barrat C, et al. Laparoscopic splenectomy for hematologic diseases. Study of 275 cases. French Society of Laparoscopic Surgery. *Ann Chir* 2000;125(6): 522-9.
- Meshikhes AW, Mubarek MA, Abu-Alrahi AI, et al. The pattern of indications and complications of splenectomy in Eastern Saudi Arabia. *Saudi Med J* 2004;25(12):1892-5.
- 9. Al-Salem AH, Naserullah Z, Qaisaruddin S, et al. Splenectomy for hematological diseases: The Qatif Central Hospital experience. *Ann Saudi Med* 1999;19(4):325-30.
- 10. de Lagausie P, Rorlich P, Benkerrou M, et al. Laparoscopic splenectomy in children: experience and results. *Arch Pediatr* 2001;8(6): 584-7.
- 11. Brigden ML, Pattullo A, Brown G. Pneumococcal vaccine administration associated with splenectomy: the need for improved education, documentation, and the use of a practical checklist. *Am J Hematol* 2000;65(1):25-9.
- Ejstrud P, Hansen JB, Andreasen DA. Prophylaxis against pneumococcal infection after splenectomy: a challenge for hospitals and primary care. *Eur J Surg* 1997;163(10):733-8.
- 13. Pickering J, Campbell H. An audit of the vaccination and antibiotic prophylaxis practices amongst patients splenectomised in Lothian. *Health Bull (Edinb)* 2000;58(5):390-5.
- 14. Ramachandra J, Bond A, Ranaboldo C, et al. An audit of post-splenectomy prophylaxis -- are we following the guidelines? *Ann R Coll Surg Engl* 2003;85(4):252-5.
- 15. Glass JM, Gilbert JM. Splenectomy in a general hospital. *J R Soc Med* 1996;89(4):199-201.