

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

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Splenectomy is a widely accepted therapeutic strategy in the management of a variety of hematological disorders^[1]. 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^[2]. 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^[3,4]. 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^[2,5].

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^[6,7], sickle cell disease^[8,9], and HS^[10] 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 pneumoniae*^[11-13], 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^[13-15].

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

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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.
3. Styrt B. Infection associated with asplenia: risks, mechanisms, and prevention. *Am J Med* 1990;88(5N):33N-42N.
4. 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.
5. 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.
6. Katkhouda N, Hurwitz MB, Rivera RT, et al. Laparoscopic splenectomy: outcome and efficacy in 103 consecutive patients. *Ann Surg* 1998;228(4):568-78.
7. 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.
8. 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.
12. Ejstrud P, Hansen JB, Andreassen 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.