

## INTRODUCTION

Beta-thalassemia is a group of genetic diseases that is common in the Mediterranean, Middle Eastern, Indian and Southeast Asian countries. Health resources are scarce in many of the countries where the disease is common (*Sheikha et al., 2007*). The total annual incidence of symptomatic individuals is estimated at 1 in 100,000 throughout the world and 1 in 10,000 people in the European Union. Three main forms have been described:  $\beta$ -thalassemia major,  $\beta$ -thalassemia intermedia and  $\beta$ -thalassemia minor (*Takeshita, 2010*).

Infections are frequent complications of  $\beta$ -thalassemia. They can be fatal. The types of these infections vary throughout the world depending on differences in the epidemiology of each infection and on the socio-economic level of each country and also vary depending on the preventive and therapeutic strategies adopted (*Di Carlo et al., 2008*).

Many patients with thalassemia major require splenectomy (*Ataga et al., 2007*). Patients who undergo splenectomy, are at an increased risk of contracting a life-threatening condition known as overwhelming postsplenectomy infection (OPSI) (*Howard-Jones et al., 2009*). The risk of invasive bacterial infection in splenectomized patients is well known (*Bisharat et al., 2001*).

Streptococcus pneumonia was responsible for the majority of the infections (66%), with a 55.3% mortality rate. It is followed in incidence by H. influenza type b, Escherichia coli, and Neisseria meningitidis (*Fuentes-*

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*Ferrer et al., 2008*). However, beta hemolytic streptococcal type A infection in thalassemic patients is not a common cause of OPSI (*LeBlanc, 1995*). Recommended strategies to prevent OPSI include education of the patients and their families, vaccination as well as preventive and emergency antibiotics (*Spelman et al., 2008*).

Splenectomised patients with better knowledge of the infection risks, those who received pneumococcal vaccine and those who had taken regular postsplenectomy antibiotics had a lower incidence of OPSI (*Jones et al., 2010*). As a general rule, asplenia is not a contraindication to routine vaccinations (both killed and living attenuated vaccines) and therefore these patients should receive all the vaccinations prescribed to normal individuals (*American Academy of Pediatrics, 2000*). Moreover, splenectomised patients should receive additional vaccinations, because of the high risk of life-threatening infections due to encapsulated bacteria observed in these patients (*Advisory Committee on Immunization Practices, 2009*).

Chemoprophylaxis with oral penicillin, 125 mg b.i.d. for children under two years and 250 mg b.i.d. for children two years and older, is recommended to reduce the risk of postsplenectomy sepsis (*Price et al., 2007*). The importance of compliance with prophylactic antibiotics should be stressed repeatedly to patients and parents. However, the limitations of antibiotic prophylaxis must also be emphasized (*Moffett, 2009*).