Rev Paul Pediatr. 2015;**33(2)**:150-153

REVISTA PAULISTA DE PEDIATRIA

www.rpped.com.br

ORIGINAL ARTICLE

Infectious complications after surgical splenectomy in children with sickle cell disease

Cypriano Petrus Monaco Junior, Patricia Belintani Blum Fonseca*, Josefina Aparecida Pellegrini Braga

Universidade Federal de São Paulo (Unifesp), São Paulo, SP, Brazil

Received 3 June 2014; accepted 26 September 2014

KEYWORDS
Sickle cell disease;
Acute splenic sequestration;
Splenectomy;
Infection

Abstract
Objective: To evaluate the frequency of infectious complications in children with sickle cell disease (SCD) after surgical splenectomy for acute splenic sequestration crisis.
Methods: Retrospective cohort of children with SCD who were born after 2002 and were regularly monitored until July 2013. Patients were divided into two groups: cases (children with SCD who underwent surgical splenectomy after an episode of splenic sequestration) and controls (children with SCD who did not have splenic sequestration and surgical procedures), in order to compare the frequency of invasive infections (sepsis, meningitis, bacteremia with positive blood cultures, acute chest syndrome and/or pneumonia) by data collected from medical records. Data were analyzed by descriptive statistical analysis.
Results: 44 patients were included in the case group. The mean age at the time of splenectomy was 2.6 years (1-6.9 years) and the mean postoperative length of follow-up was 6.1 years (3.8-9.9 years). The control group consisted of 69 patients with a mean age at the initial follow-up visit of 5.6 months (1-49 months) and a mean length of follow-up of 7.2 years (4-10.3 years).All children received pneumococcal conjugate vaccine. No significant difference was observed between groups in relation to infections during the follow-up.
Conclusions: Surgical splenectomy in children with sickle cell disease that had splenic sequestration did not affect the frequency of infectious complications during 6 years of clinical follow-up.

DOI of original article: http://dx.doi.org/10.1016/j.rpped.2014.09.006
*Corresponding author.
E-mail: patricia_blum@uol.com.br (P.B.B. Fonseca).

PALAVRAS-CHAVE
Doença falciforme;
Sequestro esplênico;
Esplenectomia;
Infecção

Complicações infecciosas em crianças com doença falciforme após esplenectomia cirúrgica

Resumo

Objetivo: Avaliar a frequência de complicações infecciosas em pacientes portadores de doença falciforme (DF) submetidos à esplenectomia cirúrgica, após episódio de sequestro esplênico (SE).

Métodos: Coorte retrospectiva de crianças com DF que nasceram após 2002 e que estavam em acompanhamento regular até julho de 2013. Os pacientes foram divididos em dois grupos,casos (constituído pelas crianças com DF que fizeram esplenectomia cirúrgica após sequestro esplênico) e controles (crianças com DF que não tiveram SE e não foram submetidas ao procedimento), a fim de comparar a frequência de infecções invasivas (sepse, meningite, bacteremia com hemocultura positiva, síndrome torácica aguda e/ou pneumonia) por meio de informações obtidas do prontuário. A análise estatística foi descritiva.

Resultados: Foram avaliados 44 pacientes com idade média no momento da esplenectomia de 2,6 anos (1-6,9 anos) e com tempo médio de seguimento após esplenectomia de 6,1 anos (3,8-9,9 anos). O grupo controle foi formado por 69 pacientes com idade média do início do seguimento de 5,6 meses (1-49 meses) e tempo de acompanhamento médio de 7,2 anos (4-10,3 anos). Todos receberam a vacina pneumocócica conjugada. Não foi observada diferença significativa entre os grupos em relação aos processos infecciosos durante o período de seguimento.

Conclusões: A esplenectomia cirúrgica nas crianças com doença falciforme e que sofreram sequestro esplênico não se associou ao aumento na frequência de complicações infecciosas após seis anos de acompanhamento clínico.

Introduction

Sickle-cell disease (SCD) is a type of hemolytic anemia caused by hemoglobinopathy that, due to the substitution of valine for glutamic acid at the sixth position of the beta globin chain, leads to the formation of an abnormal hemoglobin, called "S" hemoglobin (HbS).[1] SCD comprises sickle-cell anemia (SCA) and the associations that occur when the hemoglobin S gene is associated with the gene of other hemoglobinopathies, such as SC hemoglobinopathy (HbSC) and HbS-beta thalassemia (HbSβ).[1]

Among the complications of SCD, splenic sequestration (SS) affects 7.5% to 30% of patients, usually between 3 months and 5 years of age, being the second leading cause of death in the first decade of life.[1,2] The mortality rate for SS crisis can reach 12% and may occur in more than 50% of patients.[1,3,4] Treatment should be immediate, aiming to restore blood volume through transfusion of red blood cells.[1] The prevention of SS recurrence can be performed through periodic transfusions, chronically, or via splenectomy.[1,5-8]

The effectiveness of chronic transfusion in preventing recurrence is not well established. A study showed that the SS crisis occurs in spite of the reduction of hemoglobin S (Hb) to less than 30%, with the risk of recurrence being similar in patients receiving chronic transfusions compared to the ones who remained under clinical observation.[6] However, randomized studies are needed to confirm these data.[2] Brousse et al. found that the risk of recurrence was higher when the first SS episode occurred before two years of age and concluded that a more aggressive preventive treatment should be carried out at this age range.[9]

The performance of a splenectomy early in life is always debatable due to the increased risk of infection by encapsulated bacteria.[10] However, it should be considered that children with sickle-cell disease show splenic hypofunction since the first months of life,[11] and that advances in the prevention of these infections through prophylaxis with penicillin and conjugate vaccines have decreased the risk.[12,13]

Therefore, the aim of this study was to evaluate the frequency of infectious complications after surgical splenectomy in children with SCD.

Method

This is a retrospective cohort study carried out with data obtained from medical records of patients followed in a Pediatric Hematology service. The study population (PG) consisted of children with SCD born from 2002 to 2007, submitted to surgical splenectomy after the first episode of splenic sequestration between January 2003 and February 2009 and who had regular follow-up until July 2013. After the first episode of splenic sequestration, these patients were submitted to a regimen of chronic red blood cell trans-