Acta Scientific Gastrointestinal Disorders (ISSN: 2582-1091)

Volume 4 Issue 10 October 2021

Research Article

Surgery and Sickle Cell Disease

Alexander Forero-Torres*, Francisco Reinoso Lozano, Fuad López Fernández and Santiago Valderrabano González

General and Digestive Surgery Department, La Paz University Hospital, Madrid, Spain

***Corresponding Author:** Alexander Forero-Torres, General and Digestive Surgery Department, La Paz University Hospital, Madrid, Spain.

Received: August 26, 2021

Published: September 23, 2021

Abstract

Sickle cell anemia is considered the most common inherited red blood cell disorder. Patients with SCD present a very particular physiological response to surgical and anesthetic stress, and due to the chronic clinical manifestations, they are a perioperative challenge. Since neonatal screening campaigns and prevention and treatment of this disease crises have been carried out, these patients have increased their survival, reaching adulthood and manifesting chronic organic dysfunctions that may require surgical treatment [1].

Surgery in patients with SCD raises an unique challenges due to the chronic manifestations of their disease and especially their physiological response to surgical stress and anesthesia. The perioperative management of the patient with SCD is of vital importance to minimize the appearance of complications associated with surgical interventions, and it is essential to create multidisciplinary teams that offer adequate perioperative management of these patients.

Keywords: Surgery; Sickle Cell Disease (SCD); Sickle Cell Anemia

Core Tip

Since the creation of campaigns for neonatal screening and for the prevention and treatment of sickle cell anemia, these patients have increased their survival, reaching adulthood and manifesting chronic organic dysfunctions that may require surgical treatment. Surgery in patients with SCD poses a unique challenge due to the chronic manifestations of their disease and especially their physiological response to surgical stress and anesthesia, and the perioperative management of these patients is of vital importance to minimize the appearance of complications associated with surgical interventions and the creation of multidisciplinary teams that offer adequate perioperative management of these patients is essential. It is important to know the most frequent surgeries and their complications to offer the best treatment to these patients.

Introduction

Sickle cell anemia is considered the most common inherited red blood cell disorder. In 1910 James B. Herrick described and published the first case of Sickle Cell Disease (SCD). Since neonatal screening campaigns and prevention and treatment of this disease crises have been carried out, these patients have increased their survival, reaching adulthood and manifesting chronic organic dysfunctions that may require surgical treatment [1].

Materials and Methods

Patients with SCD present a very particular physiological response to surgical and anesthetic stress, and due to the chronic clinical manifestations they constitute a perioperative challenge. It is very important that the medical health personnel who care for these patients know the epidemiology, pathophysiology, clinical characteristics and treatment to offer the best conditions for local trauma, anemia and anesthetic and surgical stress that these patients will suffer when being operated on.

Around 25% to 30% of patients may present postoperative complications that are associated with a longer hospital stay and a higher economic cost [2]. This particularity leads them to be in fifth place in the ranking of Medicaid "super-users" with an approximate expenditure to the United States health system of 1.6 billion dollars [3].

This disease produces a distortion of the erythrocyte that generates greater adherence to the vascular endothelium, leading to a continuous increase in shear stress and oxidation, and ultimately causing long-term vascular inflammation. All these processes produce alterations in vascular tone, coagulation, fibrinolysis, inflammation, lipid transport, permeability and alter nitric oxide signals [4], which are ultimately responsible for clinical manifestations and increase the probabilities in these patients of requiring some surgical interventions with respect to the general population [2].

Results and Discussion

Among the most frequent surgeries in patients with SCD are adenotonsillectomy, cholecystectomy and splenectomy [2].

Adenotonsillectomy is a surgery frequently performed in children with SCD. It is probably due to adenoid hypertrophy related to early functional hyposplenism. Occasionally, obstructive sleep apnea secondary to adenoid growth is also identified, increasing perioperative complications due to apnea. Furthermore, a higher rate of complications has been described in children under 4 years of age [2].

The most common abdominal surgeries in patients with SCD are cholecystectomy and splenectomy. Splenectomy is usually performed in children and is secondary to episodes of splenic sequestration [2]. There are variations in inflammatory markers such as C-reactive protein (CRP) and IL-1 in patients with cholecystitis and SCD when compared to the general population; These markers present higher levels during the preoperative and postoperative periods, probably secondary to the permanent inflammatory state of this disease [5], so it is very important to take these variations into account during the postoperative clinical course in these patients. The surgical technique of choice is laparoscopic cholecystectomy, although the frequency of complications and clinical evolution is similar to open cholecystectomy [5].

Total hip arthroplasty is another of the surgeries that is frequently performed in SCD patients. A higher risk of complications has been described in total hip and knee arthroplasty compared to the general population, generating an increase in the mean stay and hospital costs [3].

Bariatric surgery is another surgical intervention of great interest in patients with SCD. Obese patients frequently present with clinical manifestations of obstructive sleep apnea-hypopnea syndrome (OSAS), which leads to a state of hypoxemia during sleep that causes the appearance of vaso-occlusive crises more frequently. Obesity surgery reduces the frequency of these vaso- occlusive crises in addition to weight loss. Currently there are no specific preoperative indications other than those of obese patients in general who would undergo this surgery [6].

Another possible manifestation of SCD is organ damage. This damage can sometimes lead to a liver or kidney transplant. One of the pathologies that generates more morbidity and mortality in these patients is nephropathy secondary to SCD. Postoperative complications are around 25%, with postoperative infection being one of the major complications in these patients, produced not only by postoperative immunosuppressive treatment, but also by changes in the immune system of patients with SCD that affect and predispose them to a higher rate of infections [7]. There is little experience in patients with SCD and kidney transplantation, accompanied by an unfavorable prognosis in patients requiring dialysis [8].

Approximately 25 - 30% of SCD patients develop chronic liver disease [9]. It can be due to the ischemic lesions typical of the disease, and complications derived from transfusions such as iron accumulation or the probability of suffering from viral hepatitis (especially in transfusions received before 2000) or having suffered acute intrahepatic sickle cell cholestasis [10]. Survival of SCD

liver transplant patients is similar to a cohort of African American transplant patients with other etiologies of chronic liver disease and a propensity score group of 2: 1. The most common causes of graft loss were seizures. sickle cell disease, hemochromatosis and portal and hepatic vein thrombosis [9].

Degenerative valve disease and pulmonary hypertension represent two diseases that can affect the life expectancy of the SCD patient. The most frequent complications in this type of surgery are: sickle cell crisis, acute chest syndrome and acute kidney damage. The prevention of these injuries through methods such as electrophoresis, exchange transfusion (to obtain a Hemoglobin S < 30%), blood cardioplegia, temperature control, avoiding hypoxemia and medication control during the preoperative period, reduces the probability of complications in the perioperative period [11].

Conclusion

Surgery in patients with SCD raises an unique challenges due to the chronic manifestations of their disease and especially their physiological response to surgical stress and anesthesia. These patients are more likely to undergo certain surgical interventions and present a greater number and severity of complications than the general population, which also conditions a longer hospital stay and higher healthcare costs [3].

The perioperative management of the patient with SCD is of vital importance to minimize the appearance of complications associated with surgical interventions. Due to the improvement in the treatments, and to an increase in the survival of these patients, the probability of undergoing any surgical intervention is increasing. Therefore, it is essential to create multidisciplinary teams that offer adequate perioperative management of these patients, based on communication and the coordinated work of surgeons, anesthesiologists and hematologists.

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