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Original Article

Ear, Nose and Throat Section

Association of Adenotonsillectomy with Vaso-occlusive Crisis Frequency in Sickle Cell Patients of Eastern Saudi Arabia: A Retrospective Study

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ABSTRACT

Introduction: The Red Blood Cells (RBCs) in Sickle Cell Disease (SCD) are distorted into a crescent shape in conditions of low oxygen because of a mutant type of haemoglobin. Vaso-occlusive Crisis (VOC), a defining symptom of SCD, is acute painful assaults caused by vaso-occlusion in the bone marrow. SCD is substantially more prevalent in some regions of Saudi Arabia than in others, with the Eastern Province having the highest incidence rates, followed by the Southern and Western provinces. A 90% of SCD patients were hospitalised due to a painful crisis, often brought on by several reasons, including infection, dehydration, acidosis, hypoxia and many others. Many childhood infections are linked to tonsil and adenoid infections, often requiring tonsillectomy or adenoidectomy.

Aim: To evaluate the relationship between adenotonsillectomy and VOC frequency in SCD patients who underwent adenotonsillectomy.

Materials and Methods: A retrospective cohort study was conducted in the Otorhinolaryngology Department in Qatif Central Hospital, Qatif, Saudi Arabia, in June 2023. Data were collected retrospectively from January 2019 and January 2022. Patients diagnosed with SCD and who underwent Adenoidectomy and/or Tonsillectomy was included in the study. Clinical and demographic data, along with co-morbidities and laboratory results, were retrieved from electronic medical records. Intragroup comparison of presurgical and postsurgical intervention variables was done using a paired t-test. A p-value of 0.05 or less was considered statistically significant.

Results: A total of 32 patients were included in the study. Among the participants, females accounted for 13 (40.6%) and males accounted for 19 (59.4%) of cases. The mean age at surgery of the participants was 10.16 years. The mean of preadenotonsillectomy admissions was less than that of postadenotonsillectomy admissions in all periods; however, this difference was not statistically significant, with p-values of 0.156 for one year, 0.376 for two years and 0.162 for three years. The mean preadenotonsillectomy VOC was higher than the postadenotonsillectomy VOC for the one-year, two year and three-year periods, with p-values of 0.400, 0.167 and 0.088, respectively. Regarding Blood Transfusions (BT), the mean preadenotonsillectomy value was higher than the postadenotonsillectomy mean; however, this difference was not statistically significant, with a p-value of 0.395

Conclusion: Over three years, Adenotonsillectomy did not affect the frequency of VOCs, BTs and hospitalisations, indicating that these procedures may not be suitable as preventive measures for the majority of patients. Future research with larger sample sizes, preferably randomised controlled trials, is required to confirm the findings and assess whether performing the procedure prophylactically in SCD patients with recurrent tonsillitis lowers the frequency of VOCs, potentially reducing the number of ED visits for pain management and control.

Keywords: Adenoidectomy, Blood transfusion, Tissue hypoxia, Tonsillectomy

INTRODUCTION

In SCD, RBCs become distorted, taking on a crescent form when oxygen levels are low. This deformation is caused by an abnormal variant of haemoglobin [1]. Deoxygenation causes Haemoglobin S (HbS) polymers to form sickle RBCs, enhance their adhesion to the endothelium, causing vaso-occlusion and tissue hypoxia [2]. Vaso-occlusive Crisis (VOCs), a defining symptom of SCD, are acute painful assaults caused by vaso-occlusion in the bone marrow [3]. Clinical manifestations of VOCs include priapism, acute chest syndrome and stroke [1].

The SCD is the most prevalent haemoglobinopathy, affects 312,000 newborns annually worldwide [4]. SCD is substantially more prevalent in some regions of Saudi Arabia than others, with the eastern province having the highest incidence rates, followed by the Southern and western provinces [5]. It is reported that between 2% to 27% of the Saudi population have sickle cell trait and up to 2.6% of the population has SCD [5]. A 90% of SCD patients are hospitalised due to a painful crisis [6], often brought on by several reasons including an infection, dehydration, acidosis, hypoxia and many others [7].

Many childhood infections are caused by infections of the tonsils and adenoids, necessitating tonsillectomy or adenoidectomy, which are two of the most common paediatric procedures [8]. A study of 260 SCD patients in Ghana found that 93.1% of them had tonsillar hypertrophy in varying degrees [9]. Patients with SCD may experience frequent pain episodes and lengthy hospitalisation time as a result of VOC due to either adenotonsillar hypertrophy or infection (adenotonsillitis) [10,11]. Recurrent infections in the adenoids and tonsils may serve as long-lasting sites of inflammation that trigger secondary inflammatory mediators, which in turn will damage the endothelium and increase RBC sickling [12].

Additionally, one of the primary causes of paediatric obstructive sleep apnoea syndrome is adenotonsillar hypertrophy [13]. The increased frequency of adenotonsillar hypertrophy, which is most likely an immune system response to functional hyposplenism, increases the incidence of Obstructive Sleep Apnoea (OSA) in children with SCD [14]. Obstructive sleep apnoea can cause hypoxemia, which can cause ischaemic stroke and transient ischaemic episodes [15].

The impact of conducting an adenotonsillectomy on the frequency of VOC has been studied in several studies. Results varied between populations and were often inconsistent. One of the studies found a substantial decline in the vaso-occlusive crisis [16]. According to another study, Tonsillectomy and Adenoidectomy (T&A) significantly reduced the number of acute chest syndrome incidents that occurred for each patient [17]. One study showed no discernible difference in the incidence of pain crisis or acute chest syndrome but showed a decrease in the rate of ischaemic stroke [18], while some studies did not observe any notable changes [19,20].

Therefore, to address this gap, the present study was conducted to evaluate the association between adenotonsillectomy and the frequency of VOC in SCD patients who underwent adenotonsillectomy.

Hence the primary objective of the study was to assess the association of adenotonsillectomy with the frequency of VOC in SCD patients. The secondary objective of the study were to assess the association of adenotonsillectomy with the frequency of emergency room visits and hospital admissions in SCD patients and to assess the association of adenotonsillectomy with the frequency of blood transfusions in SCD patients.

MATERIALS AND METHODS

A retrospective cohort study was conducted in the Otorhinolaryngology Department in Qatif Central Hospital, Qatif, Saudi Arabia, in June 2023. The data was collected retrospectively between January 2019 and January 2022, with a follow-up for a minimum of one year. Ethical approval for this retrospective cohort study was obtained from the Local Committee for Scientific Research Ethics in the Qatif Health Network, registered under QCH-SRECO 40/2023. Due to the nature of the study, informed consent was waived by the Institutional Review Board (IRB). All data were anonymised to ensure patient confidentiality. The study was conducted in compliance with the ethical standards of the responsible committee.

Inclusion criteria: Patients diagnosed with SCD who were older than two years of age were included. The diagnosis of SCD was based on the result of haemoglobin electrophoresis, who underwent adenoidectomy and/or tonsillectomy.

Exclusion criteria: Patients diagnosed with haematological disorders other than SCD, patients who had a family history or clinical picture of SCD but were not confirmed by haemoglobin electrophoresis test and patients who were lost to follow-ups were excluded from the study.

Study Procedure

Indications for surgical intervention were recurrent tonsillitis, symptoms of obstructive sleep apnoea, or both. Any co-morbidities, whether renal, hepatobiliary, or neurological, were diagnosed and managed by a physician in Qatif Central Hospital, Qatif, Saudi Arabia. The medical records of all SCD patients who underwent the surgery were collected. Each patient's Intensive Care Unit (ICU) and Emergency Room (ER) visits before and after the surgery and any supplemental data with progress notes which included details on any complications that may have occurred at other facilities, spanning the 1st, 2nd and 3rd years both pre- and post-surgery, were noted. All data collected from the medical records and used for the analysis was saved and kept in a separate hard disk protected with a password.

STATISTICAL ANALYSIS

Intragroup comparison of presurgical and postsurgical intervention variables was done using a paired t-test. A p-value of 0.05 or less was considered statistically significant.

RESULTS

A total of 32 patients were included in the study. Among the participants, females comprised 13 individuals (40.6%), while males accounted for 19 individuals (59.4%). The mean age of the participants at the time of surgery was 10.16 years [Table/Fig-1].

Variables	Number	Percentage
Age at surgery (years)	10.16 (Mean)	N/A
Gender		
Female	13	40.6
Male	19	59.4
Surgical procedure		
Adenotonsillectomy	23	71.9
Tonsillectomy	9	28.1
Surgical indication		
Recurrent tonsillitis	14	43.8
Obstructive sleep apnoea	8	25.0
Both	10	31.3

[Table/Fig-1]: The socio-demographic and clinical characteristics of the participants.

The mean number of preadenotonsillectomy admissions was less than that of postadenotonsillectomy admissions in all periods; however, this difference was not statistically or clinically significant, with a p-value of 0.156, 0.376 and 0.162 at one year, two year and three year, respectively [Table/Fig-2].

Period	Preadenotonsillectomy admissions (mean±SD)	Postadenotonsillectomy admissions (mean±SD)	p-value
1st year	0.42±0.672	0.55±0.925	0.156
2nd year	0.32±0.832	0.39±0.761	0.376
3rd year	0.23±0.560	0.39±0.761	0.162

[Table/Fig-2]: The mean number of preadenotonsillectomy admissions versus postadenotonsillectomy.

The mean of preadenotonsillectomy VOC was higher than that of postadenotonsillectomy Vaso-occlusive crisis in the 1st year, 2nd year and 3rd year periods, with p-values of 0.400, 0.167 and 0.088, respectively [Table/Fig-3].

Period	Preadenotonsillectomy VOC (mean±SD)	Postadenotonsillectomy VOC (mean±SD)	p-value
1st year	2.48±5.847	1.68±2.072	0.400
2nd year	3.16±6.404	1.48±2.580	0.167
3rd year	2.07±2.374	1.10±2.093	0.088

[Table/Fig-3]: The mean number of preadenotonsillectomy Vaso-occlusive Crisis (VOC) versus postadenotonsillectomy Vaso-Occlusive Crisis (VOC).

Regarding blood transfusion, the preadenotonsillectomy mean was higher than the postadenotonsillectomy; however, this difference was not statistically significant, with a p-value of 0.395 [Table/Fig-4].

Variables	BT preadenotonsillectomy	BT postadenotonsillectomy	p-value
Number	0.45±1.06	0.26±0.514	0.395
Mean±SD			

[Table/Fig-4]: The mean number of preadenotonsillectomy blood transfusions versus postadenotonsillectomy blood transfusions.

DISCUSSION

The SCD is a prevalent condition with a significant impact. Despite the widespread use of several disease-controlling strategies, the morbidity and mortality rates related to sickle cell crisis are still quite high. The present study observed that adenotonsillectomy in SCD patients had no impact on the frequency of VOCs, BTs and hospitalisations.

The SCD is widespread in Saudi Arabia; however, the frequency varies among the country's many areas [5]. The national rate of consanguineous marriages is 57.7% and exceeds 80% in some regions, which complicates the management of SCD [21].

The VOC is a frequent and troubling side effect of SCD that affects people of all ages. Additionally, it is the main factor in these patients' frequent ED visits [22]. Although it is well established that infection is

one of the causes of VOC, less emphasis has been paid to the fact that the grounds for conducting an adenotonsillectomy in patients with SCD are the same as those for the general population [10,23].

There are several methods for decreasing VOC in SCD; one of them entails lowering the frequency of infections that trigger the VOC [24]. Penicillin prophylaxis and SCD-specific vaccination programs are two strategies other than adenotonsillectomy, that have been used globally to reduce the risk of infections in people with SCD [24]. However, none of these actions have not, however, produced a cure, which can only be achieved by bone marrow transplantation [7].

Consistent with the findings of the current study, hospital-based research conducted in Saudi Arabia involving 40 SCD patients concluded that adenotonsillectomy does not appear to reduce the frequency of VOCs in patients with SCD over ten years. The study found no significant differences in the rates of blood transfusions, Emergency Department admissions, or Intensive Care Unit admissions before and after the adenotonsillectomy [19]. Similarly, a retrospective analysis of 27 SCD patients who had tonsillectomies revealed a lower annual incidence of VOC compared to 307 individuals who had not had the procedure; however, the difference was not statistically significant [12].

The impact of adenotonsillectomy on several kinds of VOCs was evaluated in a cohort research involving 768 patients. That research included 256 individuals who had adenotonsillectomy and the rates of visits for cerebrovascular ischaemia and obstructive sleep apnoea were much lower. On the other hand, there were no changes in the frequency of visits for acute chest syndrome/pneumonia or VOCs [18].

In another study, a total of 108 children with sickle cell disease {Haemoglobin SS (HbSS)} underwent T&A. Their average age was 7.6 years and their average haemoglobin before surgery was 9.3 g/dL. A total of 70% received blood transfusions, raising the average haemoglobin to 11.4 g/dL at the time of surgery. They stayed in the hospital for an average of 3.5 days, with complications occurring in 11.4% of cases. Before T&A, 104 children had polysomnography and 45 had it after surgery. The Apnoea-Hypopnea Index significantly decreased from 7.6 to 1.3 and the lowest oxygen levels improved from 81.2% to 89.3%. Emergency room visits also significantly dropped from 2.6 per year to 1.8 [20].

In the present research, when the authors stratified the subjects based on the surgical indication (infection vs. hypertrophy), they found no changes in the frequency of VOC episodes pre- and postoperatively over any period (1 year, 2 years and 3 years).

Although the present study observed a decrease in the number of VOC and BT, when compared to presurgical intervention, this change was not statistically significant. However, it still shows the possibility of a link between them. Further larger studies with larger samples are needed to clarify this potential link.

Limitation(s)

A notable limitation of the present research is the restricted sample size, which inhibited the ability to perform multivariate analyses necessary for controlling confounding variables. In the present study, individuals were treated with varying haemoglobin electrophoresis results as equivalent and importantly, patients were included across a diverse age range. Additionally, authors were unable to categorise patients based on the type of surgical procedure- namely, tonsillectomy, adenoidectomy, or adenotonsillectomy- due to the constraints imposed by the limited sample size.

CONCLUSION(S)

The present study found no significant link between adenotonsillectomy and reduced incidence of VOCs in SCD patients over a three-year observation period. The procedure also had no impact on the frequency of VOCs, blood transfusions, or hospitalisations, suggesting it may not be an effective preventive measure for most patients. To confirm these findings, further research with larger sample sizes and randomised controlled trials is necessary. Additionally, such studies could investigate whether adenotonsillectomy reduces VOC frequency in SCD patients with recurrent tonsillitis, potentially decreasing Emergency Department visits for pain management.

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