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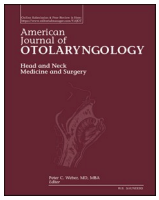



# Adenotonsillectomy in children with sickle cell disease: a systematic review

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ABSTRACT

*Objective:* Adenotonsillectomy (ADT) is a common indication for children with sickle cell disease (SCD) and comorbid obstructive sleep apnea (OSA) or recurrent infections (adenoidits and tonsillitis), despite the paucity of data in this population. This review aims to evaluate the surgical outcomes of ADT in pediatric patients with SCD.
*Methods:* We conducted a systematic review by searching the PubMed and Embase electronic databases. The PRISMA guidelines were followed, and the review protocol was registered in PROSPERO. A qualitative analysis was performed.
*Results:* Ten studies, comprising a total of 454 subjects, were included. The mean age ranged from 6 to 12 years, with a comparable sex distribution between groups. The combined cohort consisted of 303 cases of HbSS, 24 of HbSC, 16 of HbS/β-thalassemia, and 3 cases of HbSS-HPFH. Baseline hemoglobin (Hb) levels ranged from 6.7 to 9.3 g/dL, while mean Hb levels at the time of surgery varied between 8.9 and 11.4 g/dL. The proportion of patients receiving preoperative transfusions ranged from 72% to 100%. Follow-up duration spanned from 1 month to 1 year. The primary surgical indications were OSA and recurrent infections. Regarding surgical outcomes, the improvement in the AHI ranged from 50.56% to 82.90%, with mean hospital stays of 2 and 3.5 days. Postoperative acute chest syndrome and emergency department visits decreased significantly, whereas the reduction in vaso-occlusive crises was not significant. Finally, surgical complications were reported in 5.2% to 11.3% of cases.
*Conclusion:* ADT may provide clinical benefits for children with SCD. However, given the limited evidence, outcomes must be interpreted with caution, and a multidisciplinary approach is required to manage potential complications.

## 1. Introduction

Sickle cell disease (SCD) is a monogenic hemoglobinopathy caused by a point mutation that results in the substitution of valine for glutamic acid at the sixth position of the β-polypeptide chain, leading to formation of an abnormal type of Hb, defined as S (HbS) [1]. It accounts more than 300,000 new cases per year and it is clinically characterized by acute and chronic manifestations, where systemic vasculopathy leads to cumulative and multisystemic organ damage [2,3]. Children with SCD have an increased risk of obstructive sleep apneas (OSA) [4] and recurrent adenoiditis and/or tonsillitis [5,6] compared with the general pediatric population.

While the exact prevalence of obstructive sleep apnea (OSA) within SCD cohort remains to be definitively established, current literature suggests a broad range between 10% and 79% [6,7]. This markedly exceeds the estimated 1–5% prevalence observed in the general pediatric population [8,9]. The increased incidence of OSA in SCD seems to be due to adenotonsillar hypertrophy, a frequent clinical finding in these children that may arise as a compensatory mechanism following the loss of splenic function [10–12]. Untreated OSA can adversely affect disease outcome particularly in children with SCD. In fact, sleep apnea can lead to decreased nocturnal HbO2 saturation, a strong trigger of sickle hemoglobin polymerization and vaso-occlusive crisis, acute chest syndrome (ACS) and increased risk of stroke and other central nervous system events [13,14]. Adenotonsillectomy (ADT) is considered the gold standard for the treatment of pediatric OSA [15]. Although the procedure may not achieve complete resolution of sleep apnea in all cases, it consistently provides significant clinical improvement [16,17]. Acute tonsillitis and/or adenoiditis is common in children with SCD because they have not developed immunity to common organisms and have an increased susceptibility to bacterial infection. This susceptibility is due to an abnormal reticulo-endothelial system saturated with the by-

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products of haemolyzed red blood cells, abnormal phagocyte function, asplenia, and decreased opsonization against Streptococcus pneumonie [5]. The tonsils probably may act as chronic inflammatory foci, and surgical removal may decrease secondary systemic inflammatory mediators and endothelial damage in such patients [18].

Although ADT is commonly indicated for children with SCD presenting with adenotonsillar hypertrophy-induced OSA or recurrent adenotonsillar infections, limited data are available in this specific cohort. This is particularly relevant given their predisposition to perioperative complications and the potential for variable therapeutic outcomes. To the best of our knowledge, this is the first systematic review that aims to comprehensively evaluate current literature regarding ADT in pediatric SCD, focusing on surgical outcomes and complications.

## 2. Material and methods

### 2.1. Search strategy

This systematic review was conducted according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) 2020 guidelines [19]. The review protocol was registered in PROSPERO (registration number: CRD420261285530).

"We searched PubMed and Embase databases from inception, with the search last updated on January 31, 2026, for studies regarding ADT in children with sickle cell disease.

The following search terms were used in different combinations: "adenotonsillectomy" OR "adenoidectomy" OR "tonsillectomy" AND "tonsillitis" OR "recurrent tonsillitis" AND "OSAS" OR "sleep apnea syndrome" AND "sickle cell disease" OR "drepanocitosis" AND "children" OR "pediatric". The complete, reproducible search strings are provided in the Supplementary material (Table S1).

### 2.2. Study selection

We used the PICO framework to define eligibility criteria. Population (P): patients diagnosed with sickle cell disease who underwent adenotonsillectomy; Intervention/Exposure (I): adenoidectomy and/or tonsillectomy; Comparator (C): not applicable; Outcomes (O): surgical outcomes (evaluation of sleep study parameters, average admission length, number of pain crisis, acute chest syndrome and emergency department visits).

The subsequent criteria for exclusion from this review are as follows: 1) non-English articles, animals and laboratory studies, reviews and/or systematic review and/or meta-analysis, case report, editorials, conference abstract, or grey literature; 2) studies including mixed cohorts of both children and adult populations from which pediatric data could not be separately extracted or were incomplete; 3) studies lack sufficient data for extraction. Two authors (G.F. and G.W.) independently screened titles and abstracts against eligibility criteria. Full texts of potentially eligible studies were retrieved and independently assessed by the same reviewers. Data extraction and quality assessment were likewise performed independently and in duplicate by both authors. Any inter-reviewer disagreements at any stage were thoroughly discussed until a final consensus was reached.

### 2.3. Data extraction

A custom-designed Microsoft Excel spreadsheet was employed to systematically collect data. Extracted data encompassed key study attributes (authors, publication year, geographical location, type and duration study), population characteristics (number of cases, demographic and baseline data), follow-up duration and surgical outcome measures (sleep study parameters, average admission length and number of complications).

### 2.4. Quality assessment

The quality assessment of the included studies was evaluated using the Joanna Briggs Institute (JBI) critical appraisal checklist for case series assessment tools. As it is shown in Table S2, items were categorized as yes, no, or unclear. A 1-point score was assigned for a "yes" response, whereas "no" and "unclear" responses were assigned 0 points. For the case series analysis, "Good" quality was established when 7 or more of the 10 criteria were met; "Fair" quality encompassed 5 or 6 met criteria; and "Poor" quality indicated 4 or fewer criteria were satisfied. The mean score for each manuscript underwent independent appraisal by two authors (F.G. and G.W.), with any divergences resolved by consensus [20].

### 2.5. Data analysis

Due to the limited number of comparable studies and substantial clinical and methodological heterogeneity, a qualitative synthesis was performed, as the data did not permit a formal meta-analysis.

## 3. Results

The selection process is illustrated in the PRISMA flow chart in Fig. 1. A total of ten studies met the inclusion criteria, comprising included eight retrospectives and two prospective designs. Study duration varied from 4 [13] to 12 [21] years. The characteristics of the included studies are summarized in Table 1. Overall, 454 subjects were included. The mean age ranged from 6 [22] to 12 [5] years and the sex distribution was comparable between groups, showing an F:M ratio of 0.95. Seven studies provided detailed information regarding hemoglobin genotypes. The combined cohort included 303 cases of HbSS, 24 of HbSC, 16 of HbS/β-thalassemia, and 3 cases of HbSS-HPFH [13,21–26]. Baseline hemoglobin (Hb) levels were documented in four studies [13,24,26,27], with mean values ranging from 6.7 [27] to 9.3 [26] g/dL. Additionally, seven authors reported mean Hb levels at the time of ADT [13,22–27], which varied between 8.9 [13] and 11.4 [26] g/dL. Seven studies highlighted the implementation of preoperative transfusion protocols [5,21–23,25–27]. The proportion of patients receiving transfusions prior to surgery ranged significantly, from 72% [26] to 100% [22,23] across the reporting cohorts [22,23,26,27]. The follow-up duration ranged from 1 month [23,24] to 1 year [5,26].

Fig. 2 illustrates the distribution of surgical indications within the study population, categorized into OSA and recurrent infections (defined as recurrent adenoiditis and tonsillitis).

Five studies encompassing a total of 182 children evaluated surgical outcomes post-ADT for OSA (Table 2). All studies reported an improvement in sleep study parameters; specifically, OSA severity decreased from mild-moderate to mild following ADT. The reported rate of Apnea-Hypopnea Index (AHI) improvement spanned from 50.56% [28] to 82.90% [26]. Average admission lengths were 2 and 3.5 days, as reported by Warrier et al. and Farrell et al., respectively [25,26].

Table 3 outlines the impact of ADT on SCD and associated surgical complications, integrating data from four studies. All included studies documented a reduction in postoperative pain crises and acute chest syndrome (ACS) [13,21,22,26]; however, this decrease was only statistically significant in the Liguoro et al. study [22]. A reduction in emergency department (ED) visits was observed by three authors, though only the findings from Farrell et al. reached statistical significance [26]. Postoperative complications were detailed by Farrell et al. and Liguoro et al. in 5.2% to 11.3% of cases, respectively [22,26].

## 4. Discussion

This systematic review, which includes ten studies and 454 subjects, provides a qualitative description of the analyzed literature regarding ADT in children with SCD. Specifically, individual studies suggest that

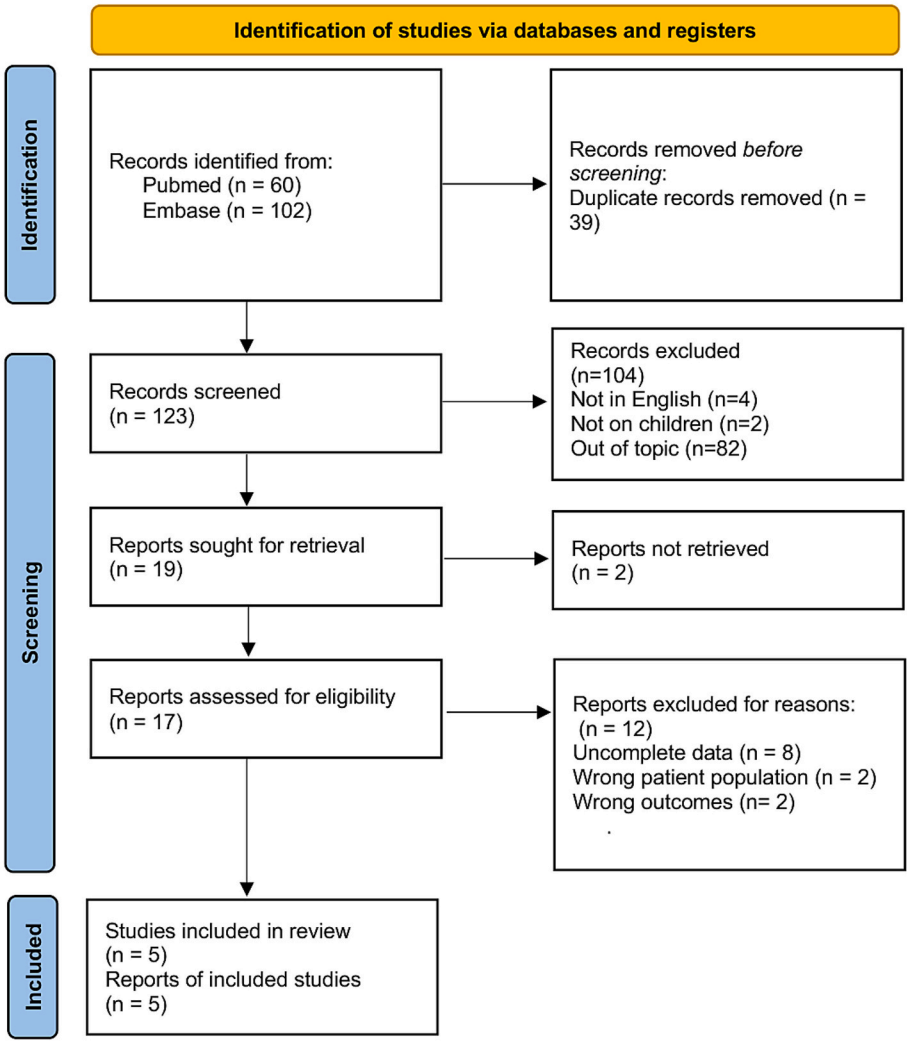


**Fig. 1.** PRISMA flow diagram of the study selection process.

these patients may benefit from ADT, as improving OSA and recurrent infections were reported to positively impact the reduction of sickling crises; second, evidence highlights that surgical complications may be prevented by adhering to perioperative multidisciplinary protocols. However, these observations should be interpreted with caution due to the limited quality of the available evidence. Most included studies were retrospective and heterogeneous; therefore, a quantitative meta-analysis was not feasible and no consolidated effect estimates can be inferred.

As it is shown in Table 1, children with SCD underwent ADT in middle-age period mainly for OSA rather than for recurrent adenoids and/or tonsils infections (Fig. 2). This is probably due, as it has been previously reported [10,11], to the major frequency of adenotonsillar hypertrophy in this population. Notably, a previous cross-sectional analysis of 85 pediatric patients with SCD reported a 55.3% prevalence of adenotonsillar hypertrophy [6]. The exact pathophysiology of this observation is not yet completely understood; however, three potential explanations are currently considered. These include: 1) the development of compensatory adenotonsillar hypertrophy following autosplenectomy; 2) a predisposition to frequent upper airway infections caused by deficient bacterial opsonization; and 3) the functional adaptation of lymphoid tissues (adenoids and palatine tonsils) into hematopoietic centers triggered by hemolytic anemia [11,28]. Recurrent adenoiditis and/or tonsillitis constitute surgical indications in approximately one-quarter of cases. Specifically, pediatric patients with SCD and obstructive adenotonsillar hypertrophy exhibit a significantly higher prevalence of recurrent upper respiratory tract infections (62.5% vs. 37.5%; $p = 0.010$) [6]. Furthermore, Salles et al. observed that exceeding five episodes of tonsillitis within a 12-month period strongly correlates with an increased frequency of vaso-occlusive crises, with a median of 12 episodes compared to 2 in less affected patients ($p = 0.017$) [6]. Moreover, a retrospective controlled study by Ajulo et al. demonstrated that tonsillectomy substantially reduces the incidence of complications secondary to tonsillar pneumococcal infections in children with SCD [5]. However, the studies included in this analysis did not provide sufficient data to further investigate the specific clinical outcomes of ADT for recurrent adenoiditis and tonsillitis. Nevertheless, this indication was included in our analysis for two main reasons: first, it represents a surgical indication that can significantly impact pediatric patients with SCD by resolving infectious episodes and their related complications; second, it frequently overlaps with OSA, further worsening the clinical picture. Therefore, future targeted studies are strictly required to clarify this crucial aspect.

Although ADT in pediatric population is a prevalent worldwide surgical intervention known for its safety and tolerability, it remains associated with inherent risks and potential complications [29]. In fact, postoperative bleeding, recognized as the most serious and feared complication, occurs in 2% to 5% of cases [30,31]. The hemorrhage-specific mortality incidence varying between 0.001% and 0.006% [32,33]. Furthermore, for children with complex chronic conditions (e.g., genetic disorders or neurological diseases), the mortality rate

**Table 1**
Summary of the baseline characteristics of the included studies.

| Author, y | Country | Study design | Study duration, y | No. cases | Mean age, y | Sex F: M | Hb type | Mean baseline Hb | Mean Hb at time ADT | Pre-operative transfusion (%) | Follow up |
|---|---|---|---|---|---|---|---|---|---|---|---|
| Ajulo et al., 1998 [5] | UK | Retrospective with control group | nr | 45 | 12 | nr | nr | nr | 'normal' | Yes | 1y |
| Waldron et al., 1999 [23] | USA | Prospective randomized | 5 | 107 | 8.2 | 63:44 | HbSS 107 | nr | 10 | 100 | 1 m |
| Wali et al., 2003 [24] | Oman | Prospective | 6 | 39 | 9.8 | 20:19 | HbSS 29 HbSB-th 10 | 7 | 10 | nr | 1 m |
| Duke et al., 2006 [27] | USA | Retrospective | 7 | 41 | 8.6 | nr | nr | 6.7–12.1 | >9 | 91 | nr |
| Warrier et al., 2010 [25] | USA | Retrospective | nr | 8 | 10.3 | 3:5 | HbSS 6 HbSC 2 | nr | 10 | Yes | 6 m |
| Rogers et al., 2010 [39] | USA | Retrospective | nr | 10 | 10.6 | 6:24 | nr | nr | nr | nr | nr |
| Finch et al., 2013 [13] | USA | Retrospective | 4 | 13 | 7.0 | 6:7 | HbSS 8 HbSC 4 HbSB-th 1 | 8.8 | 8.9 | nr | nr |
| Farrell et al., 2018 [26] | USA | Retrospective | 6 | 132 | 7.6 | 68:64 | HbSS 108 HbSC 16 HbSS HPHF 3 HbSB-th 5 | 9.3 | 11.4 | 72.7 | 1y |
| Liguoro et al., 2020 [22] | UK | Retrospective | 6 | 19 | 6 | 9:10 | Hb SS 17 Hb SC 2 | nr | 9.0 | 100 | nr |
| Marzouki et al., 2022 [21] | Saudi Arabia | Retrospective | 12 | 40 | 10 | 14:25 | Hb SS 28 HbSB 9 | nr | nr | yes | nr |

nr: not reported; ADT: adenotonsillectomy; Hb: hemoglobin; HbSS: homozygous Hb S; HbSC: double heterozygosity for Hb S and C; HbSS HPHF: Homozygous Hb S with Hereditary Persistence of Fetal Hb; HbSB-th: sickle β-thalassemia; y: year.

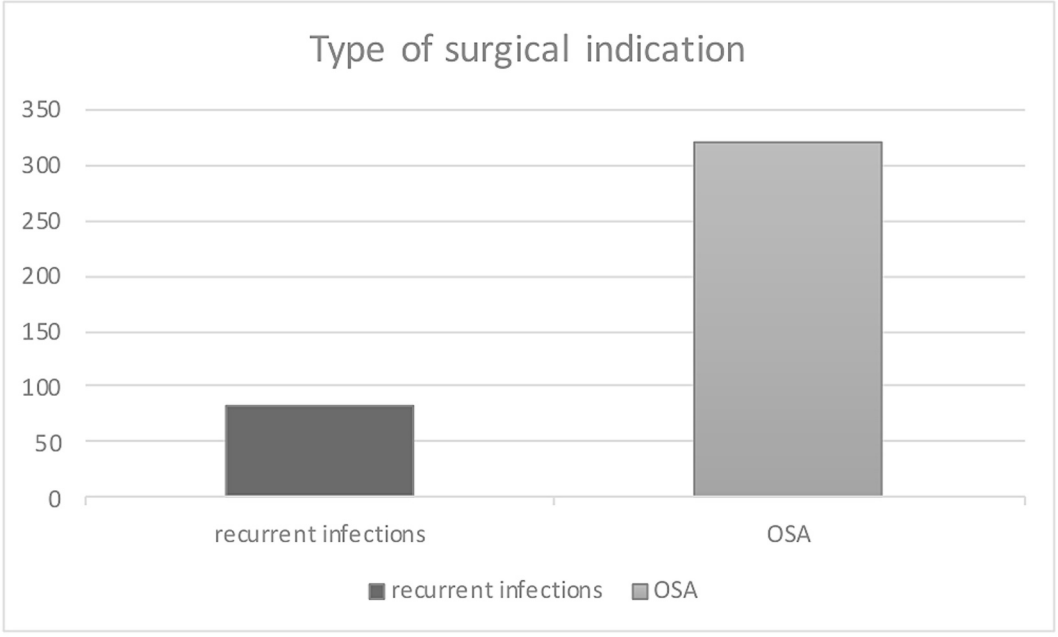


**Fig. 2.** Surgical indications for adenotonsillectomy in children with sickle cell disease.

increases significantly to 117.2 per 100,000 operations [34,35]. However, to date, these data are unavailable for ADT in children with SDC.

Due to their underlying disease, SCD children face a higher risk of complications in case of bleeding. Their low baseline blood volume (mean baseline Hb varied from 6.7 to 9.3 g/dL) leaves minimal reserve, and clinical deterioration can be rapid. Consequently, most authors advocate the implementation of preoperative transfusion protocols [5,21–23,25–27]. In these included studies, 72–100% of the patients received transfusions prior to surgery, achieving target Hb levels at the time of surgery between 8.9 [13] and 11.4 [26] g/dL.

Post-ADT bleeding in the general pediatric population is associated with substantial morbidity. Clinical consequences include readmission, extended hospital stays, the need for blood transfusions, emergency return to the operating theatre, and increased healthcare costs [36].

**Table 2**
Adenotonsillectomy outcomes for obstructive sleep apnea.

| Author, y | No. cases | Sleep study parameters preop | | Sleep study parameters postop | | *p* value | Improving %[a] | Average admission length, d |
|---|---|---|---|---|---|---|---|---|
| | | AHI | ODI | AHI | ODI | | | |
| Warrier et al., 2010 [25] | 8 | 5 | nr | 2 | nr | nr | 60 | 2 |
| Rogers et al., 2010 [39] | 10 | 8.9 | nr | 4.4 | nr | nr | 50.56 | nr |
| Finch et al., 2013 [13] | 13 | 6.3 | nr | 2.0 | nr | 0.003 | 68.25 | nr |
| Farrell et al., 2018 [26] | 132 | 7.6 | nr | 1.3 | nr | 0.0001 | 82.90 | 3.5 |
| Liguoro et al., 2020 [22] | 19 | nr | 5.7 | nr | 1.8 | 0.003 | 68.39 | nr |

AHI: Apnea-Hypopnea Index; ODI: Oxygen Desaturation Index; nr: not reported; y: years; d: days.
[a] The percentage of improvement represents the relative reduction from the preoperative baseline, calculated using the standard formula: [(Preoperative Value − Postoperative Value) / Preoperative Value] × 100.

**Table 3**
Impact of adenotonsillectomy on sickle cell disease.

| Author, y | Preop pain crisis (mean events/y) | Postop pain crisis (mean events/y) | *p* value | Preop ACS (mean events/y) | Postop ACS (mean events/y) | p value | Preop ED visits (mean events/y) | Postop ED visits (mean events/y) | p value | Complications |
|---|---|---|---|---|---|---|---|---|---|---|
| Finch et al., 2013 [13] | 0.3 | 0.2 | 0.75 | 0.2 | 0.2 | 1 | nr | nr | nr | nr |
| Farrell et al., 2018 [26] | 1.3 ± 2.3 | 1.0 ± 1.8 | 0.274 | 0.27 ± 0.6 | 0.2 ± 0.4 | 0.213 | 2.6 ± 2.8 | 1.8 ± 2.2 | 0.0002 | 15 |
| Liguoro et al., 2020 [22] | 1.1 | 1.0 | 0.729 | 0.6 ± 1.22 | 0.1 ± 0.2 | 0.03 | 1.7 | 1.0 | 0.148 | 1 |
| Marzouki et al., 2022 [21] | 1.6 | 1 | 0.337 | 9 | nr | nr | 2.4 | 1.9 | 0.386 | nr |

ACS: acute chest syndrome; ED: emergency department; nr: not reported; y: year.

Despite children with SCD being a high-risk population (Table 2), two studies have reported a low incidence of surgical complications, such as bleeding, ACS, pain crisis, reintubation, and hypotension, occurring in 5.2% to 11.3% of cases [22,26]. However, the mean length of stay was 2 and 3.5 days, respectively [25,26], which is slightly longer than that of the general pediatric population [37].

Therefore, despite data limitations hindering a broader comparison, our findings suggest that ADT in children with SCD is associated with a favorable postoperative course, largely due to pre-operative transfusion protocols that have significantly reduced surgical complications. Achieving an adequate preoperative target Hb level is absolutely critical given the potential for severe intra- and postoperative hemorrhage, an event that could acutely worsen the clinical status of a pediatric patient with SCD. Nevertheless, these results warrant further validation through larger prospective studies. In this regard, while the administration of prophylactic tranexamic acid to minimize postoperative bleeding has been proposed [38], no current evidence supports its routine use in the pediatric SCD population. Indeed, given the baseline hypercoagulable state inherent to SCD, the use of an antifibrinolytic agent could theoretically exacerbate the already elevated risk of thrombotic events and vaso-occlusive crises in these fragile patients.

Regarding surgical outcomes, 182 children with SCD underwent ADT for OSA (Table 2). These patients exhibited variable improvements in sleep parameters, ranging from a numerical reduction of 50.56% [39] to a statistically significant improvement of 82.90% [26]. However, although the overall severity of OSA decreased, mild OSA persisted. Consistent with previous literature [16,17], complete resolution of OSA following ADT is frequently not achieved. Consequently, clinical and polysomnographic postoperative follow-up is mandatory to manage residual OSA in children with SCD and to prevent potential complications, such as cerebrovascular events and cognitive impairment [15,40]. Nevertheless, no consensus currently exists regarding the optimal timing for polysomnography execution, despite the inherently high-risk nature of these pediatric patients.

In line with previous meta-analyses, our results suggest that ADT may contribute to an increase in mean nocturnal oxygen saturation. This stabilization of oxygen levels is critical in preventing hemoglobin S polymerization, thereby reducing the risk of microvascular occlusion and tissue hypoxia. However, the clinical significance of these changes has yet to be fully elucidated [41].

When evaluating the impact of ADT on SCD (Table 3), statistically significant benefits were selectively demonstrated by Farrell et al. [26], who reported a highly significant decrease in ED visits ($p = 0.0002$), and Liguoro et al. [22], who documented a significant reduction ACS ($p = 0.03$). Conversely, the remaining data across the cohorts showed mostly non-significant trends, with no statistical reductions observed for postoperative painful crises. Taken together, these findings provide crucial clinical evidence that ADT may mitigate the overall disease burden and healthcare utilization in children with SCD, reinforcing the importance of timely surgical evaluation in this fragile population. However, given the limited and overall inconsistent reporting, these findings must be interpreted with caution.

Several methodological limitations and inherent risks must be acknowledged when interpreting the synthesized evidence. Regarding the critical appraisal, although no study was classified as 'Poor' quality according to the JBI checklist, specific risks of bias warrant caution (Table S2). First, the lack of consecutive participant inclusion in the majority of the series introduces potential selection bias, indicating that the reported clinical profiles might not fully represent the broader patient population. Second, because statistical analyses were consistently descriptive or inappropriate across almost all series, the current evidence lacks inferential power, meaning conclusions are grounded in high-level descriptive data rather than robust statistical correlations.

Beyond the quality of the primary literature, this review itself faces structural constraints. The inclusion of retrospective and prospective non-randomized designs introduced heterogenous sample sizes; given the rarity of the disease, larger cohorts remain essential to validate these findings.

Furthermore, substantial clinical heterogeneity was observed across the cohorts, particularly regarding the specific surgical indications for adenotonsillectomy, perioperative management strategies, and preoperative transfusion protocols, alongside variations in outcome definitions.

This overall heterogeneity precluded a formal meta-analysis,

restricting our output to a qualitative synthesis. Finally, the variable and generally short follow-up durations across the included studies prevent the drawing of definitive, long-term clinical conclusions.

## 5. Conclusions

Based on the available literature, ADT represents a therapeutic option for OSA and recurrent adenoiditis and tonsillitis in children with SCD. However, its precise safety and efficacy profile cannot be definitively established at this level of evidence. Achieving optimal outcomes remains highly dependent on a comprehensive, multidisciplinary perioperative strategy. Additional large-scale, prospective research is required to confirm these findings and establish a consensus-based protocol for this high-risk population.

## Code availability (software application or custom code)

Not applicable.

## CRediT authorship contribution statement

**Francesca Galluzzi:** Writing – review & editing, Writing – original draft, Software, Methodology, Formal analysis, Data curation, Conceptualization. **Werner Garavello:** Writing – review & editing, Writing – original draft, Validation, Supervision, Methodology, Formal analysis, Conceptualization.

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## Declaration of competing interest

The authors declare that they have no conflicts of interest/Competing interests.

## Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amjoto.2026.104905.

## Data availability

Articles were identified through a search of the PubMed and Embase databases.

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