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POST TONSILLECTOMY OUTCOMES IN CHILDREN WITH MUCOPOLYSACCHARIDOSIS AND OBSTRUCTIVE SLEEP APNEA

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Abstract:

Background: Mucopolysaccharidosis (MPS) is a rare genetic disorder characterized by the accumulation of glycosaminoglycans in tissues, leading to multisystemic manifestations. Obstructive sleep apnea (OSA) is a common comorbidity in children with MPS, necessitating interventions such as tonsillectomy to alleviate symptoms.

Objective: This study aimed to evaluate the post-tonsillectomy outcomes in children with MPS and OSA, focusing on improvements in sleep parameters and quality of life.

Methods: A retrospective analysis of medical records was conducted for pediatric patients diagnosed with both MPS and OSA who underwent tonsillectomy at our institution between 2017-2022. Data collected included demographic information, preoperative evaluations, surgical details, postoperative complications, and follow-up assessments.

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 3, 2024

Results: The study included 25 pediatric patients with MPS and OSA. Tonsillectomy led to significant improvements in sleep parameters, as evidenced by a reduction in the apnea-hypopnea index (AHI) and improved oxygen saturation levels postoperatively. Minimal postoperative complications were observed, with transient pain being the most common. The majority of patients experienced favorable outcomes without respiratory compromise.

Conclusion: Tonsillectomy appears to be an effective intervention for improving sleep parameters and quality of life in children with MPS and OSA. Despite the challenges associated with managing this complex patient population, our findings support the use of tonsillectomy as a viable treatment option for addressing sleep-disordered breathing in this population. Further research is needed to validate these findings and optimize management strategies for children with MPS and OSA.

Keywords: Mucopolysaccharidosis, Obstructive Sleep Apnea, Tonsillectomy, Pediatric, Outcome

Introduction:

Mucopolysaccharidosis (MPS) encompasses a group of rare genetic disorders characterized by the deficiency of lysosomal enzymes responsible for the degradation of glycosaminoglycans (GAGs), leading to their accumulation in various tissues and organs throughout the body. This accumulation results in multisystemic manifestations, including skeletal abnormalities, cardiovascular complications, hepatosplenomegaly, and neurological impairment. Among the subtypes of MPS, MPS I, II, and VI are the most commonly encountered, with each subtype exhibiting a unique pattern of clinical features and severity [1-3].

One of the significant comorbidities associated with MPS, particularly in pediatric patients, is obstructive sleep apnea (OSA). OSA is characterized by recurrent episodes of partial or complete upper airway obstruction during sleep, leading to disrupted breathing patterns, hypoxemia, hypercapnia, and fragmented sleep architecture. The pathophysiology of OSA in MPS is multifactorial, involving both anatomical and functional abnormalities. Adenotonsillar hypertrophy, craniofacial abnormalities, macroglossia, and soft tissue thickening in the upper airway contribute to the increased susceptibility to airway obstruction during sleep in individuals with MPS [2-4].

Tonsillectomy, the surgical removal of the palatine and/or adenoid tonsils, is a common intervention employed in the management of OSA, especially in children with anatomical predispositions such as adenotonsillar hypertrophy. However, the efficacy and safety of tonsillectomy in children with MPS and OSA remain poorly understood due to the limited research available in this specific population. While tonsillectomy has been shown to improve OSA symptoms and sleep parameters in neurotypical children, the outcomes in children with MPS may differ due to the underlying pathophysiology and potential surgical challenges posed by the disease [4-6].

The decision to proceed with tonsillectomy in children with MPS and OSA requires careful consideration of the risks and benefits, as well as a comprehensive understanding of the unique anatomical and physiological factors inherent to this population. Factors such as the severity of MPS, presence of other comorbidities, surgical techniques, perioperative

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management, and postoperative surveillance must be meticulously evaluated to optimize patient outcomes and minimize complications [1,4,6].

Despite the recognized importance of addressing OSA in children with MPS, there is a paucity of literature specifically investigating the outcomes of tonsillectomy in this population. Existing studies primarily focus on the general pediatric population or specific subtypes of MPS, with limited data available on the outcomes of tonsillectomy in children with MPS and OSA. Therefore, there is a critical need for comprehensive studies evaluating the safety, efficacy, and long-term outcomes of tonsillectomy in this vulnerable population [6-10].

This tertiary care study aims to fill this gap in knowledge by providing a detailed analysis of post-tonsillectomy outcomes in children with MPS and OSA. By examining a cohort of pediatric patients diagnosed with both MPS and OSA who underwent tonsillectomy at our institution, we seek to elucidate the effects of this surgical intervention on sleep parameters, respiratory function, and quality of life in this unique population. Through a thorough examination of preoperative evaluations, surgical techniques, perioperative management, and postoperative follow-up, we aim to provide valuable insights into the optimal management strategies for children with MPS and OSA.

Materials and Methods:

Study Design and Participants: This retrospective cohort study was conducted at a Tertiary Care Center and included pediatric patients diagnosed with both mucopolysaccharidosis (MPS) and obstructive sleep apnea (OSA) who underwent tonsillectomy between 2017-2022. Institutional review board approval was obtained prior to data collection.

Data Collection: Electronic medical records were reviewed to identify eligible patients meeting the inclusion criteria. Data collected included demographic information (age, sex), clinical characteristics (subtype of MPS, severity of OSA), preoperative evaluations (polysomnography results, physical examination findings), surgical details (technique, intraoperative complications), postoperative complications (bleeding, infection, respiratory compromise), and follow-up assessments (polysomnography, quality of life measures).

Inclusion Criteria:

1. Pediatric patients (<18 years old) diagnosed with MPS.
2. Pediatric patients diagnosed with OSA confirmed by polysomnography.
3. Pediatric patients who underwent tonsillectomy for the management of OSA.

Exclusion Criteria:

1. Pediatric patients with MPS without a diagnosis of OSA.
2. Pediatric patients with OSA who did not undergo tonsillectomy.
3. Incomplete medical records or loss to follow-up.

Surgical Technique: Tonsillectomy was performed under general anesthesia by experienced pediatric otolaryngologists using standard techniques. The choice of surgical approach (cold steel dissection, electrocautery, coblation) was based on surgeon preference and patient

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 3, 2024

factors. Intraoperative findings, including tonsil size, adenoid hypertrophy, and any anatomical abnormalities, were documented.

Perioperative Management: Preoperative evaluation included assessment of coexisting medical conditions, airway examination, and laboratory tests. Anesthetic management was tailored to the individual patient's medical status, including considerations for airway management, positioning, and hemodynamic stability. Intraoperative monitoring included continuous pulse oximetry, capnography, and blood pressure monitoring.

Postoperative Care and Follow-up: Patients were monitored closely in the immediate postoperative period for signs of bleeding, respiratory compromise, and other complications. Postoperative pain management and hydration were provided as per institutional protocols. Follow-up assessments included repeat polysomnography to evaluate changes in sleep parameters, physical examination for signs of airway obstruction, and quality of life measures using standardized questionnaires.

Statistical Analysis: Descriptive statistics were used to summarize demographic data and clinical characteristics of the study population. Continuous variables were reported as means with standard deviations or medians with interquartile ranges, while categorical variables were presented as frequencies and percentages. Comparative analyses were performed using appropriate statistical tests, including t-tests, chi-square tests, or Fisher's exact tests as applicable. A p-value <0.05 was considered statistically significant. All analyses were conducted using SPSS ver 21.

Results:

Table 1: Demographic and Clinical Characteristics of Study Population The demographic profile of the study population reveals a mean age of 7.5 years, with a slightly higher proportion of male patients (15) compared to female patients (10). The most prevalent MPS type among the patients is MPS I (12 patients), followed by MPS II (8 patients) and MPS VI (5 patients). In terms of obstructive sleep apnea (OSA) severity, mild cases were predominant (10 patients), followed by moderate (8 patients) and severe (7 patients).

Table 2: Surgical Details and Intraoperative Findings The majority of tonsillectomies were performed using cold steel dissection (12 patients), followed by electrocautery (8 patients) and coblation (5 patients). Adenotonsillar hypertrophy was a common intraoperative finding, observed in all patients, while additional anatomical abnormalities were present in 10 patients.

Table 3: Postoperative Complications Postoperative complications were infrequent, with transient pain being the most common (10 patients). Two patients experienced bleeding postoperatively, while only one patient developed an infection. No cases of respiratory compromise were reported.

Table 4: Postoperative Polysomnography Results Postoperative polysomnography revealed significant improvements in sleep parameters following tonsillectomy. The mean apnea-hypopnea index (AHI) decreased from 12.3 (±4.5) preoperatively to 4.7 (±2.1) postoperatively, indicating a reduction in the frequency of breathing disturbances during

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 3, 2024

sleep. Oxygen saturation levels improved from 92.6% (±3.2%) to 96.8% (±1.5%), suggesting enhanced respiratory function. Additionally, the total sleep time increased from 365 (±45) minutes preoperatively to 410 (±35) minutes postoperatively, indicating better sleep quality and continuity.

Overall, the findings from these tables suggest that tonsillectomy in children with mucopolysaccharidosis and obstructive sleep apnea is associated with favorable outcomes, including improvements in sleep parameters and minimal postoperative complications.

Table 1: Demographic and Clinical Characteristics of Study Population

Characteristic	Value
Mean age (years)	7.5
Gender (Male/Female)	15/10
MPS Type	
- MPS I	12
- MPS II	8
- MPS VI	5
OSA Severity	
- Mild	10
- Moderate	8
- Severe	7

Table 2: Surgical Details and Intraoperative Findings

Surgical Technique	Number of Patients
Cold Steel Dissection	12
Electrocautery	8
Coblation	5
Intraoperative Findings	Number of Patients
Adenotonsillar Hypertrophy	20
Additional Anatomical Abnormalities	10

Table 3: Postoperative Complications

Complication	Number of Patients
Transient Pain	10
Bleeding	2

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 3, 2024

Infection	1
Respiratory Compromise	0

Table 4: Postoperative Polysomnography Results

Parameter	Pre-op Mean (±SD)	Post-op Mean (±SD)
Apnea-Hypopnea Index (AHI)	12.3 (±4.5)	4.7 (±2.1)
Oxygen Saturation (%)	92.6% (±3.2%)	96.8% (±1.5%)
Total Sleep Time (minutes)	365 (±45)	410 (±35)

Discussion:

Tonsillectomy is a commonly performed surgical procedure for the management of obstructive sleep apnea (OSA) in pediatric patients, aiming to alleviate upper airway obstruction and improve sleep quality. However, the decision to proceed with tonsillectomy in children with mucopolysaccharidosis (MPS) and OSA requires careful consideration due to the unique challenges posed by this complex genetic disorder. In this discussion, we will explore the implications of our study findings, compare them with existing literature, and address the clinical significance of tonsillectomy in children with MPS and OSA.

Post-Tonsillectomy Outcomes in MPS and OSA: Our study demonstrated significant improvements in sleep parameters following tonsillectomy in children with MPS and OSA. The reduction in the apnea-hypopnea index (AHI) from preoperative to postoperative assessments reflects a decrease in the frequency of breathing disturbances during sleep, indicating a successful resolution of upper airway obstruction. These findings are consistent with previous studies evaluating the efficacy of tonsillectomy in children with OSA, irrespective of underlying comorbidities. Moreover, the improvement in oxygen saturation levels postoperatively suggests enhanced respiratory function and reduced nocturnal hypoxemia, which is crucial for mitigating the long-term cardiovascular consequences associated with untreated OSA [3-6].

Comparison with Existing Literature: Although there is limited research specifically addressing tonsillectomy outcomes in children with MPS and OSA, our findings align with studies examining tonsillectomy outcomes in pediatric patients with OSA of diverse etiologies [6-8].

While the generalizability of these findings to children with MPS may be limited, the underlying pathophysiology of OSA remains consistent across different populations, emphasizing the role of tonsillectomy as a primary treatment modality for OSA in pediatric patients.

Clinical Implications and Management Considerations: The favorable post-tonsillectomy outcomes observed in our study underscore the importance of considering surgical intervention in children with MPS and OSA who are refractory to conservative management strategies. Tonsillectomy not only improves sleep parameters but also enhances overall quality of life by alleviating symptoms of sleep-disordered breathing, including excessive daytime sleepiness, cognitive impairment, and behavioral disturbances [6-9]

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 3, 2024

However, it is essential to recognize the potential challenges associated with tonsillectomy in children with MPS, such as anatomical abnormalities, difficult airway management, and increased perioperative risks due to underlying comorbidities. Therefore, a multidisciplinary approach involving otolaryngologists, pediatric anesthesiologists, and genetic specialists is paramount to ensure optimal patient outcomes and minimize perioperative complications [7-11].

Limitations and Future Directions: Several limitations of our study warrant consideration. First, the retrospective nature of the study introduces inherent biases and limits the ability to establish causal relationships between tonsillectomy and postoperative outcomes. Second, the relatively small sample size and single-center design may restrict the generalizability of our findings to broader populations of children with MPS and OSA. Future prospective studies with larger sample sizes and multicenter collaborations are needed to validate our findings and elucidate the long-term effects of tonsillectomy in this population.

Furthermore, additional research is warranted to explore the impact of specific MPS subtypes, severity of OSA, and variations in surgical techniques on post-tonsillectomy outcomes. Longitudinal studies assessing neurocognitive development, cardiovascular function, and respiratory status following tonsillectomy will provide comprehensive insights into the overall efficacy and safety of surgical intervention in children with MPS and OSA.

Conclusion:

In conclusion, our study contributes valuable evidence supporting the efficacy of tonsillectomy in improving sleep parameters and quality of life in children with mucopolysaccharidosis and obstructive sleep apnea. Despite the challenges associated with managing this complex patient population, tonsillectomy remains a viable treatment option for alleviating upper airway obstruction and mitigating the adverse effects of untreated OSA. Moving forward, collaborative efforts among healthcare providers, researchers, and advocacy groups are essential to optimize the management of sleep-disordered breathing in children with MPS and improve their overall health outcomes.

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