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Recommended Citation

Arganbright, Jill M.; Tracy, Meghan; Narayanan, Srivats; and Ingram, David G. MD, "Tonsillectomy In Children With 22q11.2 Deletion Syndrome" (2022). *Research at Children's Mercy Month 2022*. 24.
https://scholarlyexchange.childrensmc.org/research_month2022/24

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Tonsillectomy in Children with 22q11.2 Deletion Syndrome



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Background

22q11.2 deletion syndrome (22qDS) is a common genetic disorder with an estimated risk of obstructive airway symptoms in 10-37% of affected children. While tonsillectomy is commonly pursued in an effort to ameliorate airway obstruction, there are currently no published studies evaluating tonsillectomy outcomes in children with 22qDS.

Objective

To investigate the indications, efficacy, and complications of tonsillectomy in a large cohort of patients with 22qDS.

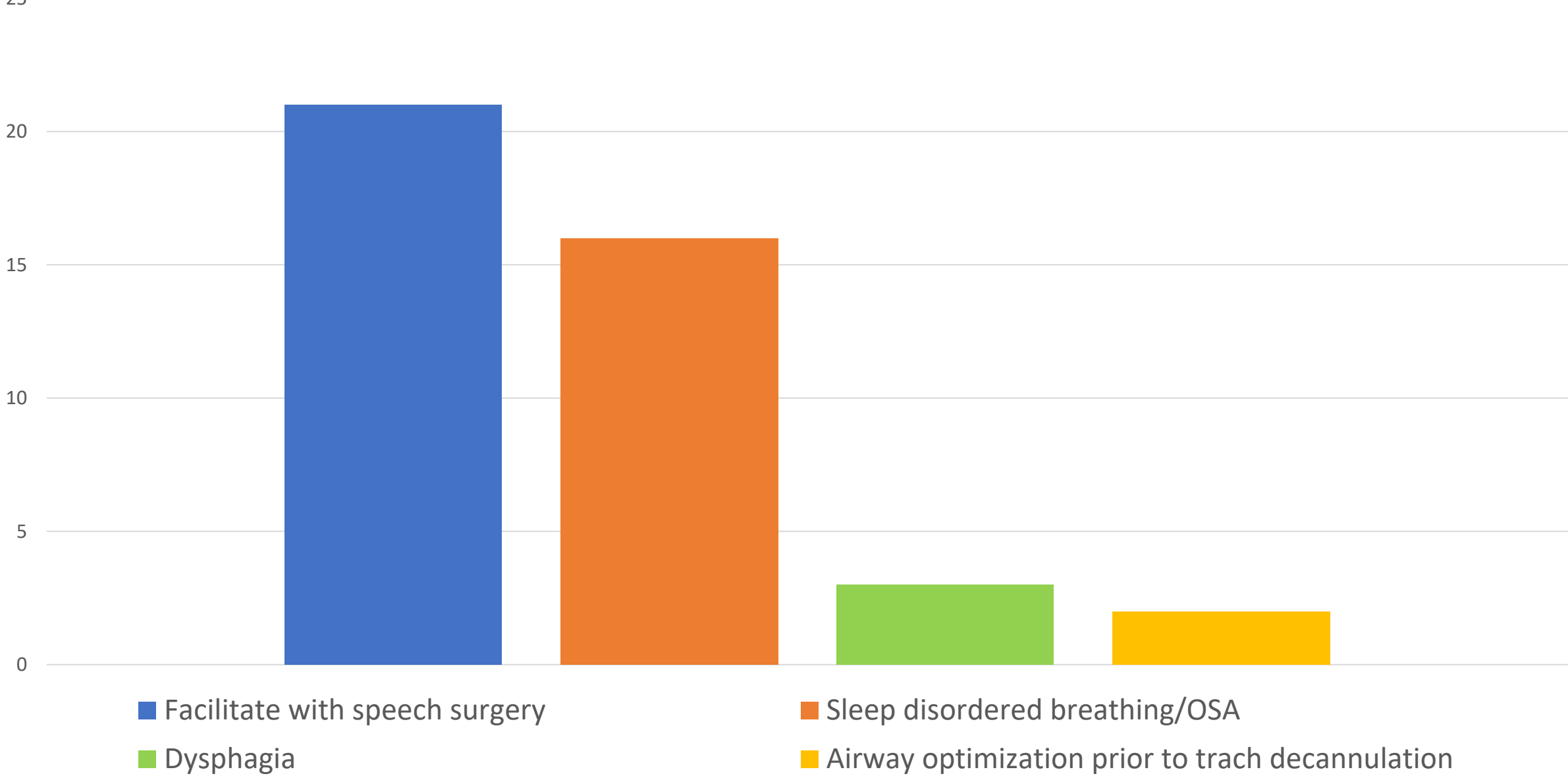
Methods and Materials

We performed a retrospective chart review of 168 consecutive patients with 22qDS from the institution's 22q repository from 1/1/2010-6/1/2021. Data was collected for tonsillectomy indications, length of hospital stay, complications, and pre- and postoperative sleep study results.

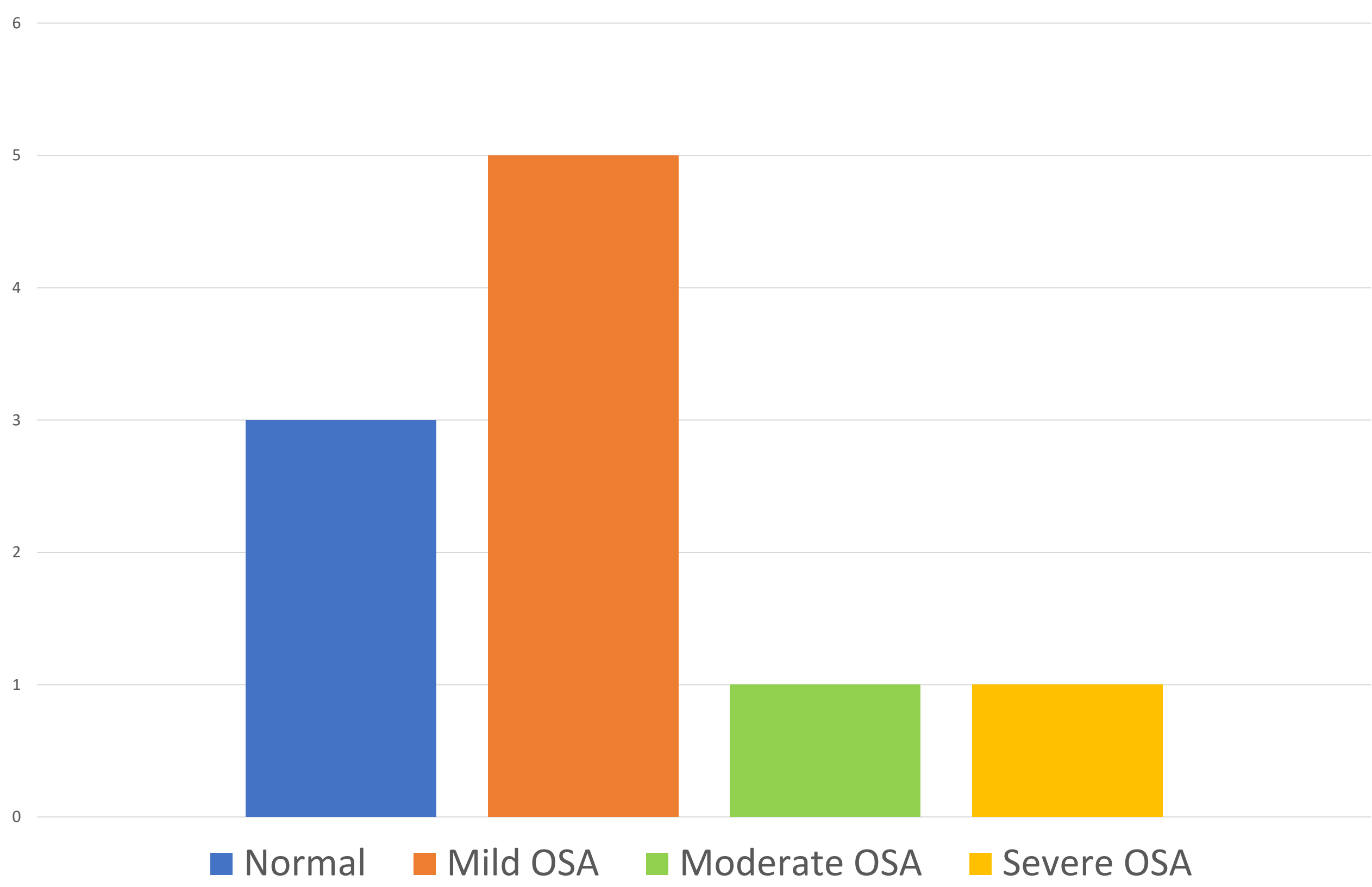
Results

Thirty-three of the 168 (20%) patients with 22qDS in our sample underwent tonsillectomy. Indications for tonsillectomy included: to facilitate with speech surgery (n=21), sleep disordered breathing/obstructive sleep apnea (OSA) (n=16), dysphagia (n=3), and airway management prior to trach decannulation (n=2). Average length of stay was 1.15 days (range 0-4 days). Six patients had a preoperative sleep study, 10 patients had a postoperative sleep study, and 3 patients had both pre- and post-operative sleep studies. Of those children who had postoperative sleep studies, 3 were normal, 5 had mild OSA, 1 had moderate OSA, and 1 had severe OSA. Three patients presented with postoperative tonsil bleeds, of whom 1 required surgical cauterization and 2 recovered with observation alone. Of the 8 patients who had postoperative calcium monitoring, 4 (50%) had hypocalcemia and received an endocrinology consultation; 1 patient was treated with oral supplementation, and 3 were observed with repeat labs. One patient experienced postoperative respiratory failure requiring overnight intubation.

Indications for Tonsillectomy in Patients with 22qDS



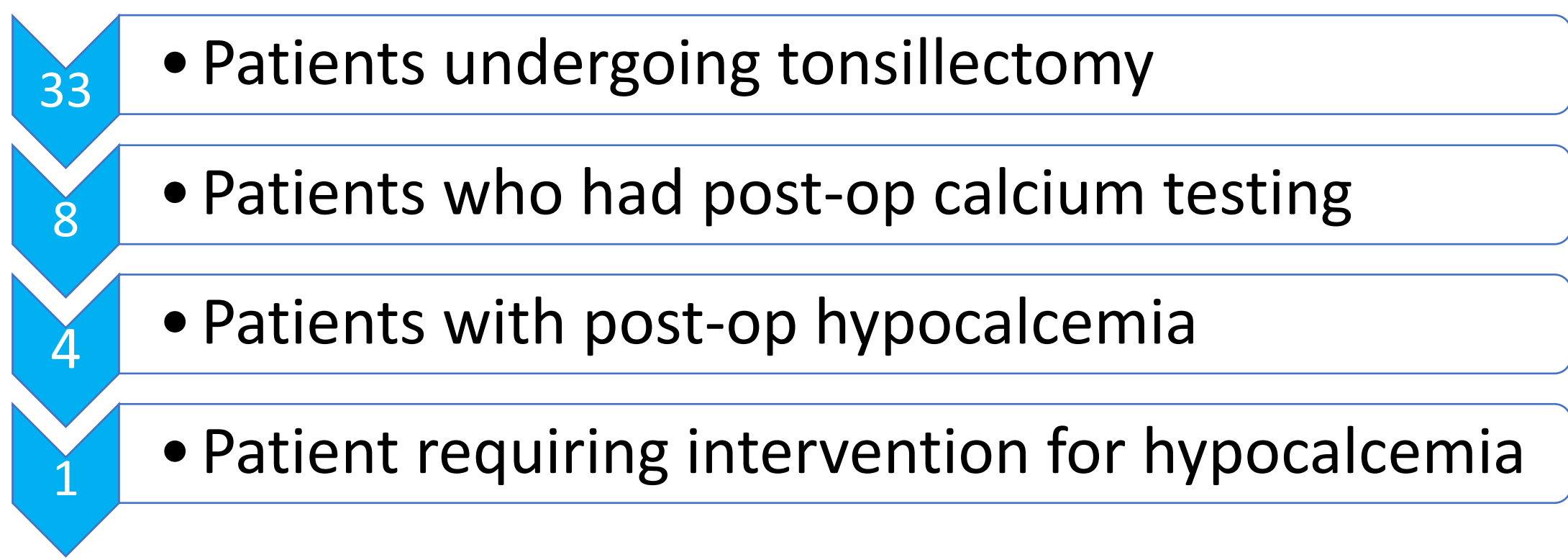
Post-Tonsillectomy Polysomnography Results



Discussion

Tonsillectomy was a commonly performed procedure in this large cohort of patients with 22qDS. Facilitation of speech surgery and sleep disordered breathing/OSA were the most common indications. Most patients experienced swift recovery with one night in the hospital prior to discharge, although these data highlight the potential need for close postoperative calcium and respiratory monitoring. Of note, for the eight patients that had postoperative calcium monitoring, 50% had hypocalcemia. Our data were limited with respect to polysomnographic outcomes. While a majority of this cohort had persistent OSA following tonsillectomy, we had only 3 patients with both pre- and post-operative sleep studies. This makes it difficult to comment on the success of tonsillectomy in treating OSA for this patient population.

Postoperative Calcium Monitoring



Conclusions

Tonsillectomy is common in patients with 22qDS. Providers should consider overnight observation and postoperative calcium monitoring following tonsillectomy. Future studies are needed to better characterize sleep disordered breathing/OSA outcomes following tonsillectomy for patients with 22qDS.

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