VPI following Adenotonsillectomy in Prader-Willi Syndrome

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ABSTRACT

BACKGROUND & PURPOSE: Sleep disordered breathing (SDB) is common among children with Prader-Willi Syndrome (PWS). SDB is multifactorial and is associated with the life threatening risk of obstructive sleep apnea (OSA), often requiring management with adenotonsillectomy. VPI is a rare complication of adenotonsillectomy, or adenoidectomy, surgery. It is characterized by persistent hypernasality with or without clinical hypernasal speech, which is a significant adverse outcome for patients and their families. The objective of this study was to assess the prevalence of VPI following adenotonsillectomy surgery among patients with PWS.

METHODS: Surveillance review of 8 PWS patients who underwent adenotonsillectomy, tonsillectomy, or both. Ten patients were diagnosed with OSA and 2 with sleep apnea disorder (SBD). Tonsil size was measured by subjective evaluation of the operating surgeon (0 = not enlarged, 1 = mildly enlarged, 2 = moderately enlarged, 3 = severe enlargement). Patient records were reviewed for patient demographics (age and sex), surgeries performed, surgical indications, and occurrence of VPI.

RESULTS: 4 patients did receive preoperative speech therapy for preexisting hypernasality. Five patients received postoperative speech therapy, 4 of whom received it because of documented perceptual measures used for conformational diagnosis of VPI. The average follow-up period after surgery was 26 months, with a range from 3 to 93 months. Of these 8 PWS patients, 4 patients (50%) developed hypernasality and underwent postoperative speech therapy for VPI. All 4 PWS-patients with VPI received phoneme “backing” (see Tables 2 and 3).

CONCLUSIONS: Children with Prader-Willi syndrome are at an increased risk of developing velopharyngeal insufficiency following adenotonsillectomy. As adenotonsillectomy is often necessary for management of OSA and SDB in the PWS population, more emphasis should be placed on preoperative counseling and postoperative monitoring concerning the high risk of VPI in this population.

REFERENCES


DISCUSSION

Given their genetic predisposition for hypotonia and obesity, along with any underlying condition such as obstructive sleep apnea, children with PWS are at an increased risk of developing sleep disordered breathing (SDB) and hypernasality. Furthermore, the increasing rate of adenotonsillectomy (OSA) in PWS patients has been found to improve the management of sleep disordered breathing (SDB). Moreover, PWS patients have a higher occurrence of VPI following adenotonsillectomy. Adenotonsillectomy, or tonsillectomy, has been shown to improve the symptoms of hypernasality, hypernasal speech, and hypernasal breathing in PWS patients. This improvement is likely due to the removal of the tonsils and adenoids, which are known to contribute to the obstruction of the nasal passage. Furthermore, the removal of these structures may reduce the risk of sleep apnea, which is a common complication in PWS patients. According to the literature, the incidence of VPI following adenotonsillectomy in PWS patients has been reported to be between 20-50%, with the majority of cases occurring within the first year after surgery.

CONCLUSIONS

Children with Prader-Willi syndrome are at an increased risk of developing velopharyngeal insufficiency following adenotonsillectomy. As adenotonsillectomy is often necessary for management of OSA and SDB in the PWS population, more emphasis should be placed on preoperative counseling and postoperative monitoring concerning the high risk of VPI in this population.