

Obstructive sleep apnea and the role of tongue reduction surgery in children with Beckwith-Wiedemann syndrome (2018)

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Background

Patients with Beckwith-Wiedemann syndrome (BWS) can be affected by a large tongue (macroglossia). Similar to other features of BWS, macroglossia can vary in severity between patients. Studies suggest that children with macroglossia are at an increased risk for obstructive sleep apnea (OSA), a condition that is also highly variable, ranging from mild sleep obstruction to severe respiratory distress. No recommendations regarding OSA management in patients with BWS and macroglossia exist.

Purpose

This article reviews all available evidence regarding children with Beckwith-Wiedemann Syndrome (BWS) and macroglossia. The prevalence of obstructive sleep apnea (OSA) and management strategies in this population are discussed.

Findings

Evaluations

Children suspected of having BWS and macroglossia should receive the following evaluations. No clear guidelines exist for at what age children should be evaluated.

- **Clinical Genetics:** Any child with a feature suggestive of BWS should be referred to a clinical geneticist, who can evaluate the patient and determine whether the patient meets criteria for a clinical diagnosis of BWS.
- **Plastic Surgery:** Patients with macroglossia should be referred to a plastic surgeon, who can evaluate the size of the tongue to determine whether a tongue reduction surgery is necessary.
- **Pulmonology:** A pulmonologist can evaluate the degree to which the large tongue affects breathing, as an increased tongue size can narrow the airway and cause upper airway obstruction.
 - Polysomnography (sleep study) is used for evaluation of OSA in children and has been used in certain studies of BWS children to detect the following: moderate- severe OSA, upper airway obstruction, apnea, upper airway resistance, severe desaturation, sleep-disordered breathing, and snoring.

Conservative management: Positioning and CPAP

Conservative management of macroglossia for children with BWS includes therapies that avoid surgery. Positioning the baby during sleep can help prevent the tongue from blocking the airway. Positions include supine (flat on back) or prone (flat on chest). Continuous positive airway pressure (CPAP) works to keep the airway constantly open. Neither treatment has been studied well enough to fully understand their effectiveness in children with BWS.

Surgical Management: Tongue Reduction

The most common form of treatment for macroglossia in children with BWS is a tongue reduction surgery with the goal of improving sleep apnea at night as well as to potentially improve speech, feeding, and jaw or dental malformations. A number of small studies have reported improved OSA after a tongue reduction surgery and other studies have shown variable results regarding the effect of

a tongue reduction on speech and feeding. Various surgical techniques have also been reported. Neither a standard surgical technique nor an agreed upon ideal time for a tongue reduction have been established.

Other surgical techniques for patients with severe OSA can include a tracheostomy, tonsillectomy, adenoidectomy, uvulectomy, tongue-lip adhesion, and/or lingual frenectomy.

Conclusion

A relationship between tongue size and OSA severity has not been established. Success has been reported in treating OSA with tongue reduction, but larger studies are needed to understand which patients benefit the most from this procedure.

Key Points

- Children with BWS and macroglossia are at risk for OSA.
- There are a variety of treatment options for OSA including conservative and surgical management.
- More research is needed to determine the best treatment.

Reference

Cielo CM et al. Obstructive sleep apnoea and the role of tongue reduction surgery in children with Beckwith-Wiedemann syndrome. *Paediatr Respir Rev.* 2018;25: 58-63. PubMed PMID: 28366681