# ANKYLOGLOSSIA SUPERIOR SYNDROME: A FORM OF THE OROMANDIBULAR-LIMB HYPOGENESIS SYNDROME

Danilo Augusto Nery Dos Passos Martins, Roseli Maria Zechi Ceide and Cristiano Tonello

Objectives:Under the spectrum of the oromandibular-limb hypogenesis syndrome, the ankyloglossia superior syndrome is an unusual condition defined by the coexistence of limb defects and glossopalatine ankylosis. This situation may promote important closure or limited access to the oral cavity and needs to be adequately managed from the point of view of the upper airway and dysphagia. We report an extremely rare case within this context.Resumed report:A boy was diagnosed at birth with narrow palpebral fissures, micrognathia and tongue adhered to the hard palate. Anomalies were also observed in the upper limbs, such as clinodactyly of the 5th finger of both hands, and in the lower limbs, with mesomelic shortening of the legs, clubfoot, rudimentary toes of both feet, with syndactyly (2nd to 5th) in the right and ecytrodactyly (2nd to 4th) in the left.The fibronasopharyngolaryngoscopy showed presence of submucous cleft of the soft palate, bifid uvula and omega-shaped epiglottis. There was no glossoptosis. Nutrition was performed via nasoenteral feeding tube until the surgery. At 32 days of age, the patient was intubated guided by nasofibroscopy and underwent glossoplasty. He presented a good postoperative evolution at 7 months of age with adequate coordination of swallowing.Conclusion:The etiology of these malformations remains unknown. The classification of the oromandibular-limb hypogenesis syndromes within a specific type is difficult due to the frequency of overlapping features.Particularly, the ankyloglossia superior syndrome represents a potential challenge in the management of the airway and dysphagia of the newborn.