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Orthognathic Surgery in Patients With Congenital Myopathies and Congenital Muscular Dystrophies: Case Series and Review of the Literature.

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Abstract

PURPOSE: This case series examined preoperative findings and the surgical, anesthetic, and postoperative management of 6 patients with congenital myopathies (CMs) and congenital muscular dystrophies (CMDs) treated at a tertiary medical institution with orthognathic surgery over 15 years to describe pertinent considerations for performing orthognathic surgery in these complex patients.

MATERIALS AND METHODS: According to the institutional review board-approved protocol, chart records were reviewed for all orthognathic surgical patients with a clinical, genetic, or muscle biopsy-proved diagnosis of CM or CMD.

RESULTS: Six patients (5 male, 1 female) qualified, and they were treated by 4 surgeons in the division of oral and maxillofacial surgery from 1992 through 2007. Average age was 19.5 years at the time of orthognathic surgery. Five patients had Class III malocclusions and 1 patient had Class II malocclusion. All 6 patients had apertognathia with lip incompetence. Nasoendotracheal intubation with a difficulty of 0/3 (0=easiest, 3=most difficult) was performed in all cases. Routine induction and maintenance anesthetics, including halogenated agents and nondepolarizing muscle relaxants, were administered without malignant hyperthermia. All 6 patients underwent Le Fort level osteotomies; 4 also had mandibular setback surgery with or without balancing mandibular inferior border osteotomies. Five patients required planned intensive care unit care postoperatively (average, 18.4 days; range, 4 to 65 days). Postoperative respiratory complications resulting in major blood oxygen desaturations occurred in 5 patients; 4 of these patients required reintubation during emergency code response. Five patients required extended postoperative intubation (average, 4.2 days; range, 3 to 6 days) and ventilatory support. Average hospital length of stay was 21.8 days (range, 6 to 75 days). Average postoperative follow-up interval was 29.8 weeks (range, 6

to 128 weeks).

CONCLUSIONS: Patients with CMs or CMDs often have characteristic dentofacial malocclusions that contribute to functional problems with feeding and drooling and psychosocial problems. Orthognathic surgery, usually bimaxillary, can be judiciously considered in these patients; these procedures typically require multidisciplinary pre- and postoperative evaluation and care over lengthy hospital stays with a high risk of respiratory complications that bear consideration in treatment planning.

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