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"Cleft Lip and Palate Repairs in a patient with Epidermolysis Bullosa Simplex"

Background: Epidermolysis bullosa (EB) is a rare genetic condition resulting in extremely fragile skin that easily blisters with shearing or pulling. There are three major forms of EB which all present with sloughing of the skin due to mutations in genes involving the mechanical stability and adhesion of skin. The simplex form of EB results from mutations in keratin genes and causes blistering within the epidermis. This case reviews a patient with EB simplex and bilateral cleft lip and palate. To our knowledge, there are only two documented cases of a cleft lip or palate repair being performed on a patient with EB, the most recent being reported in 2005 (Cooper et al., 2003; Ozgur et al, 2005).

Description: Our patient is a now 10-month-old female with EB simplex, who received care for bilateral cleft lip and palate. A treatment plan for cleft lip and palate repair was created through communication with members of the local craniofacial team, dermatologist, and a national EB team, in cooperation with the patient's parents. Precautions were taken peri-operatively to minimize any shearing or pulling of the skin, such as wrapping Vaseline gauze underneath the blood pressure cuff, trimming ECG pads to minimize adhesive contact, suturing the endotracheal tube to the mandible to avoid the use of tape, and taking extra care whenever contact with the skin was required. The child underwent bilateral cleft lip adhesion at 4-months of age after limited upfront treatment with nasoalveolar molding by craniofacial orthodontics. The surgery resulted in no complications, and no new bullae were observed during or after her overnight admission. Outpatient testing by exposure to Dermabond skin adhesive was performed without untoward reaction. Definitive cleft lip repair was completed at 7-months of age using the Mulliken technique, as well as tip rhinoplasty with placement of nostril retainer stents. Otolaryngology also performed bilateral myringotomy and tympanostomy tube at this time. The child was again kept overnight for observation and discharged with no complications and no new bullae. A week later, the sutures were removed under anesthesia.

Results: Due to extensive planning and multidisciplinary teamwork our patient has had successful cleft lip repair with no complications and no new skin lesions upon discharge home or on post-operative follow-up. A palatoplasty is scheduled for the child and the same precautions will be taken.

Conclusion: Treating patients with cleft lip and palate along with EB requires careful planning and communication to prevent complications. Educating all team members and following established guidelines for patients with EB while modifying traditional cleft treatment plans is the best way to maximize outcomes.