Safe Use of Propofol Total intravenous anesthesia (TIVA) in Patients with Gorlin-Goltz Syndrome

P Owusu-Agyemang, A Van Meter, A Zavala, A Shah, S Moore, U Williams

University of Texas MD Anderson Cancer Center, Department of Anesthesia and Perioperative Medicine

Introduction

Gorlin-Goltz syndrome, also referred to as nevoid basal cell carcinoma or focal dermal hypoplasia, is a rare autosomal dominant disorder. It is diagnosed by major criteria: multiple basal cell carcinomas under the age of 20, odontogenic keratocysts of the jaw, plantar and palmar pits, falx cerebri calcification, bifid or fused ribs, and a first degree relative with this syndrome; and minor criteria: macrocephaly congenital malformations (frontal bossing), skeletal deformities, radiologic abnormalities, ovarian fibromas or medulloblastoma (1). It can be associated with difficult airways due to facial asymmetry, dental abnormalities, papillomas of the lips and tongue, and verrucous lesions in the hypopharynx (2).

Methods

Two five year old female twins with a history of Gorlin syndrome presented for MRI to rule out medulloblastoma. They had a history of multiple moles in the chest and back, and pits in their hands and feet. One of the twins had also recently complained of a headache. There was no history of airway issues. Both twins had normal vital signs prior to induction. Induction in the MRI suite was done with propofol 70 mg IV followed by a propofol infusion at 200 mcg/kg/min for both patients.

Results

A simple face mask was used during TIVA with propofol. There were no complications.

Discussion

While skin, nails, bones, hair, and teeth are all affected by the Gorlin syndrome, skin changes are seen most frequently (90% of patients) (3). Dental anomalies are seen in 46% of patients. Santana et al noted that 90% of patients develop odontogenic keratocystic tumors, and 20% develop multiple basal cell carcinomas (4). This multitude of issues most likely requires frequent screening and frequent use of anesthesia. There are no documented cases in the literature of propofol TIVA use in this patient population. We utilized propofol TIVA with no complications.

References